ADVANCES IN SURGICAL TECHNIQUES FOR THE MANAGEMENT OF EARLY-ONSET SPINAL DEFORMITIES (EOSD)

by

M H H NOORDEEN MA, BM, FRCS(Tr & Orth)

Advances in Surgical Techniques for the Management of Early-onset Spinal Deformities

Mr. M H Hilali Noordeen

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-- **Mr. M H H Noordeen** MA, BM, MCh, FRCS (Tr & Orth)

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ABSTRACT

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Early-onset spinal deformity (EOSD) is characterised by detection of spinal deformity (scoliosis, kyphosis or multi-planar) in children at aged less than five years. The common causes could be classified under congenital, neuromuscular, syndromic and idiopathic etiologies. Early treatment is paramount in preventing rigid, severe and progressive deformities that can cause pulmonary compromise. The inability of lungs and thoracic cage to support normal ventilation at rest constitutes the pathophysiology in manifestation of thoracic insufficiency syndrome (TIS).

The treatment options have evolved from observation, serial casting, bracing to surgery. Early definitive spinal fusion is now obsolete and contra-indicated for EOSD. Growing rods (submuscular or subfascial) continues to be the standard of care in treating these challenging deformities. Vertebral expandable prosthetic titanium rib (VEPTR) continues to be an attractive option for TIS but is fraught with high complication rate.

I hereby present a theme on EOSD with a set of thirty three peer reviewed indexed publications and one surgical patent in treating such challenging conditions highlighting my original contribution as Consultant spinal surgeon spanning over two decades. My pioneering and ground-breaking research that has shaped the surgical management of EOSD and helped define ‘standard of care’ is presented. My novel and innovative concept of treating EOSD using magnet driven growing rod (MdGR) along with its preliminary results is discussed. MdGRs are an attractive alternative in eliminating need for repetitive anaesthesia facilitating normal cognitive development in comparison to growing rods (CGRs). They also improve pulmonary function in neuromuscular scoliosis. A brief one page summary of all my indexed publications with comments on originality and how they contributed to spinal surgery is enclosed at the end of each chapter. My current research update on MdGR project at Royal National Orthopaedic Hospital (Stanmore), my surgical patent, National institute of clinical excellence (NICE) position statement on MdGRs and clinical guidance documents are attached as appendices I - III.

Chapter 1

INTRODUCTION

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Scoliosis is derived from Greek word '*Scolios'* meaning crooked. It is characterised by a lateral curvature of the spine with the rotation of vertebrae in the axial plane. This curvature could be due to numerous etiologies, namely congenital deformations, neuromuscular causes, secondary sequelae of trauma, tumours and infections. When the cause was not known, despite extensive work-up, it was labeled as idiopathic (i.e diagnosis of exclusion). Idiopathic scoliosis is the most common type of spinal deformity. Scoliosis has traditionally been classified as infantile (zero to three years), juvenile (four to eight years), and adolescent (ten years and onwards until skeletal maturity) by JI James (Idiopathic Scoliosis: The prognosis, diagnosis, and operative indications related to curve patterns and the age of onset. J Bone Joint Surg 36B:36-49, 1954). This classification system enjoyed immense popularity as it was related to prognosis and was predictive of long-term outcome. No references were made for ages eight to ten years in his original article. Detecting the onset of spinal deformity can be difficult or impossible, which has led to the development of a separate classification system into two broad types: early-onset scoliosis (birth to five years) and late-onset scoliosis (five years and up). The rationale behind these distinctions being that late-onset curves have less risk of compromising pulmonary function.

The management of spinal deformities has evolved over the last few decades from non-operative modalities including traction, casting and bracing, to instrumented correction of the deformity with fusion. Advances in instrumentation for the spine have progressed from Harrington rods to segmental spinal fixation by pedicle screws. These pedicle screws and segmental spinal fixation have revolutionized the surgical management of scoliosis over the past two decades.

Early-onset spinal deformity is characterized by new diagnosis, where in one detects scoliosis, kyphosis, or multi-planar deformity in children under five years of age. The growing spine has been an area of increasing interest and research over the last few years. This

vulnerable age group has unique epidemiology, clinical features, management, and outcome parameters.

Treatment of progressive spinal deformity in a growing child poses a significant challenge and there is no universally agreed upon recommendations or treatment guidelines regarding standard of care. The chest cage deformation in early-onset scoliosis, and its impact on pulmonary function, has been extensively studied since the beginning of the new millennium. There has been an explosion in the development of newer techniques with the introduction of more sophisticated surgical implants, guided by a better understanding of pulmonary function and its impact on long-term quality of life, in addition to the correction of the spinal deformity. The main stay of the treatment options for these children is to correct and control their deformity while allowing them to have a near normal pulmonary maturation with growth. The aim is to ensure that this is achieved in the simplest way possible, has least impact on the child's cognitive, mental, behavioral, psychological and physical development, and has least number of complications. Ensuring these goals are met continues to be a formidable challenge to spinal deformity surgeons.

Furthermore, it is of the utmost importance to have a better understanding of the specific features and components of spinal deformity in a growing child, emphasizing the differences between younger children and the older adolescent age groups. Lung development, maturation, and pulmonary function are increasingly recognised as critical factors whose optimisation is paramount for successful long-term outcomes. Insight and awareness in to these factors has led to an understanding the etio-pathogenesis of thoracic insufficiency syndrome (TIS), and has created new surgical strategies to deal with this difficult problem. TIS is defined as the inability of the thorax (the combined biomechanical unit of thoracic spine, rib cage, sternum and the diaphragm) to support normal respiration or lung growth in a child. The three dimensional volume depletion deformity, as a result of progressive spinal and chest

cage deformation with growth and time, is responsible for TIS. It is associated with high mortality and morbidity, and the orthopaedic gold standard '*Cobb angle*' is of minimal relevance for a complex three dimensional problem.

A. Anatomy

The spine is the segmental column of thirty three vertebrae that provides strength, flexibility, and protection to the neural tube, and constitutes the major sub-cranial part of the axial skeleton. The precursors to the vertebrae are the somites arising from the paraxial mesoderm. Occipital (4), cervical (8), thoracic (12), lumbar (5), sacral (5) and coccygeal (4-5) pairs of somites eventually form the spinal column, and all of them are present by the fourth or fifth week of gestation. Its individual elements are united by a series of intervertebral articulations that form a firm, but flexible shaft supporting the trunk and protective covering for the neural elements.

The vertebral column and spinal cord of young children have their own unique features, differentiating them from skeletally mature spines. Appreciation of these differences and an understanding of biomechanics is the first step towards better care of these children. The length of the spine nearly triples between birth and adulthood. At birth, the vertebral column measures approximately 240mm in length. In newborns, only thirty percent of the spine is ossified with minimal difference in morphology between cervical, thoracic, and lumbar vertebrae. Subsequently the cervical, thoracic, and lumbar vertebrae acquire their individual identities gradually with time. This includes development of foramen transversarium for the vertebral artery, uncinate process, and unco-vertebral joints in the cervical region; unique sagittal orientation of facet joints with costal facets for articulation with the ribs in thoracic and mammillary processes with coronal orientation of facets for lumbar vertebrae respectively.

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The ossification of the spine is completed only in the third decade, unlike an appendicular skeleton (complete by the end of the second decade). The rate of growth also varies, and in the thoracic region, the posterior elements grow faster than anterior elements, imparting physiological thoracic kyphosis. The reverse occurs in the cervical and lumbar region, resulting in the development of cervical and lumbar lordosis.

B. Biomechanics

Background knowledge of material science and structural mechanics is necessary to understand and appreciate the fundamental biomechanics of spinal deformities. Leonhard Euler in 18th century described the mechanism of failure for an erect column. These engineering principles have stood the test of time for three centuries, and continue to be the foundational theories behind the design of spinal constructs to achieve sound fusion (arthrodesis). In addition, the biological tissue differences of a paediatric spine, which has growth potential and re-modeling, is different from that of an adult spine, which has completed its growth. This presents a unique set of problems that are yet to be fully understood, and the consideration of these problems is valuable in evaluating how best to manage paediatric spine deformities.

The main function of the axial skeleton is to provide a rigid support to the appendicular skeleton (i.e. limbs), and to withstand physiologically applied loads while facilitating a range of motion with nominal energy expenditure. Structural pathologies in the spine alter the equilibrium, and corrective strategies to treat spinal deformity attempt to restore this equilibrium by an application of force, either through bracing or internal fixation. One must first understand static mechanics principles, and the concept of force application and its effects, to better understand the treatment of spinal deformity. Common questions include:

1) What type of force to apply and how? (i.e. corrective bracing by three point fixation /

correction fulcrums; compressive vs. distractive forces; use of screws, hooks, rods plates, wires; segmental vs. end fixation anchors, and anterior vs. posterior scaffolds)

2) How does the force act in-vivo (i.e. in tension or in compression, and whether it is acting concentric or eccentrically?)
3) What are the consequences of such a force and in what plane? (i.e. coronal, sagittal, or multi-planar; what is the impact on local and global motion?).

C. Growth

Certain aspects of spinal growth are poorly understood despite the spine having a well-established pattern of growth throughout the years of maturation. The growth velocity (i.e. rate of growth) has a bimodal peak:

i) The first five years and

ii) The adolescent growth spurt.

The growth velocity is steady at about eight to ten mm a year between the ages of five to ten. A Dimeglio and F Bonnel studied the longitudinal growth of the thoracic and lumbar vertebrae and quantified it to be about eight to eleven mm a year (Dimeglio A, Bonnel, F: The Spinal Column in Growth. Springer, Paris: 1990). R Roaf studied the growth of intervertebral discs, and observed a mean increase in 0.2 to 0.6mm for thoracic and 0.3 to 0.8mm for lumbar discs per year respectively (R Roaf. Vertebral growth and its mechanical control J Bone and Joint Surg 42B(1): 40-59).

Severe scoliosis can cause pulmonary hypertension and Cor pulmonale. The chest cage deformation with progressive deformity results in costo-pelvic impingement, which compromises the space available for the lungs (SAL), and increases their work for breathing. With the diminished size of the thorax and a reduced excursion of the ribs, the accessory muscles of respiration are strained, causing tachypnoea and hypercapnia. The vicious cascade

finally culminates in right ventricular failure. These features are seen only in early-onset scoliosis and not in late-onset curve patterns. Numerous reports exist in literature correlating increased mortality and morbidity from cardio-pulmonary compromise in early-onset scoliosis.

Despite tremendous advances, very little is known regarding the natural history of early-onset scoliosis, particularly with regards to genetic defects. Thorough understanding of the natural history is of paramount importance, since it has a bearing on its management, especially with regards to surgical intervention.

D. Classification

Early-onset scoliosis has traditionally been classified according to etiology into: idiopathic, neuromuscular, congenital, and exotic types (which include syndromic, skeletal dysplasias, secondary to genetic disorders, and so forth). The natural history of many rare genetic and neurodegenerative conditions that produce scoliosis is not known. Given the rarity of such conditions in the general population, health bodies (NIH and DoH) have little interest in funding research for these areas. The Scoliosis Research Society (SRS) and the British Scoliosis Research Foundation (BSRF) continue to pioneer advances and excellence in understanding these conditions through research funding and international collaboration.

The first broadly applied classification system in instrumentation era for surgical treatment of idiopathic scoliosis was published by King and Moe (King HA, Moe JH et al: Selection of fusion levels in thoracic adolescent idiopathic scoliosis. J Bone Joint Surg Am 65(9):1302-1313, 1983). Their classification enjoyed immense popularity for more than two decades, and categorised AIS into five main types. It provided excellent treatment recommendations for the Harrington spinal instrumentation system and analysed only thoracic curve patterns in a two-dimensional plane. However, a better understanding of the sagittal

profile and its importance for long-term outcomes quickly exposed the inherent limitations of this classification. A robust, more comprehensive, and three-dimensional objective, which has a reliable and reproducible classification, that also provides fusion recommendations, was proposed by L Lenke (Lenke LG, Betz RR et al: Adolescent idiopathic scoliosis: a new classification to determine extent of spinal arthrodesis. J Bone Joint Surg Am 83-A(8):1169-1181, 2001). This classification was based on plain radiographs, and curves were classified into six types, with each curve type further sub classified into A, B, and C modifiers. The sagittal plane was also evaluated and +/- suffixes were attached to convey if the curves were hyper or hypokyphotic respectively. This yielded thirty six AIS curve characteristics. Despite this, the Lenke classification is not without its limitations:

i. Shoulder balance and symmetry are not taken into consideration

ii. It does not quantify rib-hump deformity

The DaVinci's three-dimensional classification system is a new research tool that accurately quantifies coronal, sagittal and axial plane deformities depicting them in a vectorial representation. First reported by L Doung and H Labelle, its use is confined largely for research purposes and is less used in day to day clinical practice. (Duong L, Labelle H et al. Three-dimensional classification of spinal deformities using fuzzy clustering. Spine 2006; 31:923–30). There continues to be a need for a clinically oriented, easily remembered, and consistently reproducible classification system based only on the factors that are of importance to patients and their caregivers. Such a classification should also directly affect the surgical treatment of these curves guiding the surgeon in choosing appropriate fusion levels and not be too time consuming for a busy clinician. It should also be able to meet patients' and their family's expectations, as well as meet the surgeon's definition of success with surgical intervention.

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E. Clinical Assessment

The initial evaluation of a child with suspected spinal deformity should begin with a detailed clinical history and careful physical examination with a detailed neurological evaluation. Antenatal and perinatal history, with special reference to birth presentation type and developmental milestones, should be diligently elicited as cognitive delay has been shown to correlate with curve progression. Idiopathic infantile scoliosis is seen more commonly with breech presentation and low birth weight males.

Physical evaluation should begin with search for a midline hairy patch (suggestive of spinal dysraphism), café-au-lait spots, axillary freckling (suggestive of neurofibromatosis), and other cutaneous markers (Shagreen's patch of Tuberous sclerosis, and so forth). Anthropometric measurements of sitting, standing height, and the upper: lower segment ratio is also recorded. Arm span and standing height, if disproportionate, suggests Marfanoid habitus syndrome. Chest cage circumference and chest expansion on maximum inspiration is also assessed. The flexibility of curvature can be studied by holding the child suspended under the arms, and looking for correction with lateral pressure or suspending the older child from a gymnastic bar.

Leg lengths are measured to rule out non-structural causes of scoliosis, especially in an ambulant child with a lumbar curve. Other features like plagiocephaly, developmental dysplasia of the hip (DDH), congenital heart diseases, inguinal hernia, anal atresia, and so on, are looked for and considered carefully. Joint laxity and hypermobility should be assessed in all patients. Evaluation of other systems and a study of facial features (low-set ears with hypertelorism, and the broad or depressed nasal bridge of Down's syndrome; the blue sclera of Osteogenesis imperfecta, and so forth) guides one in associating these features with syndromic diagnosis, thus giving insight into the natural history of underlying disorder.

Finally, careful evaluation of the neurological system is to be completed. Assessing

motor function and gait is imperative to evaluation of occult central nervous system abnormalities. Asymmetrical superficial abdominal reflexes may be the only manifestation of occult syringomyelia with Chiari I malformation. The superficial abdominal reflex is characteristically absent on the convexity of curvature. Deep tendon reflexes and coordination should be tested to evaluate long-tracts and cerebellar function.

Lastly, with a better understanding of pulmonary maturation in the new millennium and pathogenesis of thoracic insufficiency syndrome (TIS), consideration should be given for an evaluation by a pediatric pulmonologist, especially if in the presence of significant chest cage deformation and severe scoliosis. Though lung function is difficult to ascertain in children less than five years old, a dynamic 3D CT chest volumetric measurement gives enormous insight into the severity of the patient's condition, and the required timing and effect of surgical intervention. More recently, dedicated centers for thoracic insufficiency are also performing dynamic lung MRIs to better study the pulmonary biomechanics of TIS, and these centers are observing different types of volume depletion deformities of the thorax in order to formulate optimal surgical solutions.

F. Investigations

Plain radiographs are a cornerstone for diagnosis and the study of curve patterns and their characteristics. One of the first goals is to rule out congenital etiology by looking for an unsegmented bony bar or bridge on the concavity. Also, midline defects (absent spinous processes / spina bifida occulta) can be studied on plain x-rays. The Cobb angle measurement between the most titled vertebrae into the concavity of the curvature helps in quantifying the magnitude of the deformity. M Mehta in 1972 reported the relation of apical vertebra to the corresponding rib, and reported the RVAD (rib vertebral angle difference) to predict the progressing curve types from resolving ones. (MH Mehta. The rib-vertebral angle in the early

diagnosis between resolving and progressive infantile scoliosis. J Bone and Joint Surg. 1972, 54B: 230-43). Later, she also described an additional tool known as "phase of the rib head" for prognostication of infantile idiopathic scoliosis. Curves in phase I transition (no overlap of rib head or neck on apical vertebra) with a RVAD of less than twenty degrees resolved in 99 percent of instances. Curves in phase II transition with a RVAD of more than twenty degrees progressed in more than ninety eight percent of the cases.

In addition to plain radiographs, magnetic resonance imaging (MRI) is mandatory in many types of early-onset scoliosis to rule out split cord (diastematomyelia and diplomyelia) and Arnold-Chiari malformations. MRI is a non-invasive technique that provides multi-planar imaging without the use of ionizing radiation with superior soft-tissue characterization, facilitating the visualization of complex anatomy, and giving an accurate assessment of the entire neuraxis. Any patient with an atypical curve pattern, an abnormal neurology, or a painful, or rapidly progressing curve, should undergo an MRI. Up to forty percent of congenital spinal deformities have associated intra-spinal abnormality, but a routine pre-op MRI is not indicated in neurologically normal AIS patients with typical curve patterns. Since images may degrade at either end of the magnetic gradient, the areas of anatomic interest should be placed in the middle of the field gradient. In addition to providing direct evaluation of the spinal cord and cerebrospinal fluid (CSF), the MRI also gives multi-planar information on the surrounding soft tissue and osseous structures. Visualization and image characteristics may be augmented with intravenous administration of paramagnetic substances, such as gadolinium, that shortens both T_1 and T_2 relaxation times. This, to some degree, helps in differentiating infections from edema or tumours, though exceptions exist.

Although MRI is noninvasive, it may require sedation for young patients, or for those with claustrophobia. While open scanners have helped patients with claustrophobia,

their image quality lags behind that of closed scanners. Variations and choices in magnetic field strength (0.5T to 3.0T) may be considered in order to improve image characteristics and obtain the best quality images with maximal clinical information.

There is a small population of patients that exist who may have contra-indication for MRI (those with cardiac pacemakers, Berry aneurysm clips, cochlear implants, metallic foreign objects in the eyes, previous stainless steel instrumentation in the spine, and so forth). Contrast myelography or CT myelogram offers valuable dynamic information on cord compression and severity in such individuals. In severe multi-planar deformities, a dedicated CT scan with 3D reconstruction would facilitate a better understanding of local anatomy, especially with congenital deformities.

More recently rapid prototyping and reverse engineering principles are employed in creating a 1:1 ratio 3D model of the spine to better understand the anatomy, to plan or rehearse pedicle screw trajectories, and to perform resection or osteotomy prior to the surgical execution in the operating room to minimize neurological risks and complications. 3D printers are now available commercially, and such an approach is likely to be a standard of care in the coming years.

G. Management of most prevalent deformity types

Conservative management:

The treatment of early-onset scoliosis deformity (EOSD) revolves around one fundamental principle: the ‘degree of anticipated or actual curve progression’. Mehta’s criteria, though extremely useful for idiopathic infantile scoliosis, is of limited utility for EOS secondary to non-idiopathic etiologies. Curves in Phase II transition with a Cobb angle of twenty to thirty five degrees had a risk of progression, so bracing or casting is recommended. A total body cast with serial changes every six to twelve weeks is most popular for toddlers,

while the juvenile age group is prescribed braces (Boston underarm orthosis, or a Charleston brace, and so forth). Brace wear is encouraged full-time (at least twenty three hours), and should be discarded when there is no change in the Cobb angle for at least two years.

M McMaster has given enormous insight into the natural history and management of congenital scoliosis. He described the rate of progression for different curve patterns based on underlying congenital anomaly. Curves that had a unilateral unsegmented bar with contralateral hemivertebra had the highest risk of progression by ten to fifteen degrees, over a period of six months, where as those with block vertebra or hemi-metameric shift rarely progressed to a degree warranting surgical intervention. (MJ McMaster and K Ohtsuka. The natural history of congenital scoliosis. A study of two hundred and fifty-one patients. J Bone Joint Surg(Am), 1982, 64(8):1128-1147). In between these two ends of the spectrum exists the fully segmented and partially segmented hemivertebra (MJ McMaster and CV David. J Bone Joint Surg(Br) 1986, 68(4):588-595). Defects in segmentation resulting in posterior or postero-lateral quadrant producing kyphotic or kypho-scoliotic deformities were detrimental to cord function, resulting in paraparesis. Surgical management is recommended for such sagittal plane deformities.

The management of neuromuscular deformities is guided by the natural history of the underlying disorder. Bracing is largely ineffective, and it neither changes the nature of deformity nor its curative. It temporarily buys time in postponing the inevitable.

Surgical management:

The ultimate goal in surgical management of EOSD is to prevent the progression of deformity by guiding the growth of the spine by internal fixation and maximizing the normal development of the thorax, or chest cage with minimal complications. Though numerous methods and devices were proposed and are in use, an ideal one-off surgical solution that

achieves these desired goals is still not available. Therefore, the quest for such an ideal surgical option is the key crux and motivation for this dissertation. Surgery is usually entertained, especially when the Cobb angle is more than forty five degrees with a progressive curve (defined as more than five to six degrees of worsening in six months on at least two occasions) in a skeletally immature patient (Riser Gr.0–II).

Initial proponents believed a short, straight spine was better than a longer, bent spine, an early definitive spinal fusion was proposed. It was quickly evident, however, that this significantly compromised sitting height with a reduced upper: lower segment ratio, and it reduced pulmonary function or vital capacity. Exercise tolerance was unacceptable, so alternative modalities were explored. Isolated posterior fusion resulted in relentless anterior growth with a recurrence of deformity and the 'crankshaft phenomenon'. The surgical options for EOSD could be broadly divided into:

i) Fusion vs. non-fusion surgeries

ii) Anterior vs. posterior approach surgeries

i. Idiopathic

Roaf proposed that the deformity of early-onset idiopathic scoliosis was due to the asymmetrical rate of growth between the concave and convex sides. (R Roaf. The treatment of progressive scoliosis by unilateral growth-arrest. J Bone Joint Surg(Br) 45(6):637-51). The higher growth velocity on the convex side, in relation to the concave side, was an attractive option to perform anterior convex growth arrest (convex epiphysiodesis). This involved an ablation of the convex epiphyseal cartilage and adjacent vertebral endplates around the apex of the deformity. With time, instrumentation was added primarily to control the progression of the deformity and to achieve sound fusion over the operated area. In the new millennium, the same goal is achieved by employing a minimally invasive technique using staples which are popular amongst surgeons who perform video-assisted thoracoscopic surgeries (VATS).

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Posterior based surgical options include growth-sparing spinal instrumentation that started as a distraction-based technique with the use of Harrington rods and outrigger devices. Later with time, the growth-sparing surgical constructs became popularised as an expandable, segmental instrumentation technique for single or dual growing rod instrumentation with periodic lengthening, using a tandem end to end connector, or a domino side to side connector. Such rods were placed either sub-muscularly or sub-fascially.

ii. Congenital

There are several types of surgery available for congenital scoliosis, and surgical management is dictated by the underlying nature of the bony deformity. Co-existent tethered cord or spinal dysraphism mandates neurosurgical de-tethering and closure of meningocoele prior to scoliosis correction. Similarly for diastematomyelia, excision of a bony or cartilaginous spur is performed prior to attempting spinal surgery. The surgical options include in-situ fusion to hemivertebra excision with, or without, instrumentation. A fully segmented hemivertebra, especially in junctional areas (i.e. cervico-thoracic, thoraco-lumbar, or lumbo-sacral), requires complete resection either by all posterior or combined anterior-posterior approaches. The surgery is ideally performed before the age of five years to optimize outcomes with intact neurology. The choice of approach and the staging of surgery are guided by the surgeon's preference and the locally established hierarchy from the surgeon's pioneer predecessors.

The Vertical Expandable Prosthetic Titanium Rib (VEPTR) was developed by RM Campbell Jr. is a longitudinal rib to pelvis distraction device that is particularly useful in treating EOSD associated with thoracic insufficiency syndrome and fused ribs (RM Campbell, M Smith et al. The characteristics of thoracic insufficiency syndrome associated with fused ribs and congenital scoliosis. J Bone Joint Surg(Am) 85(3):399-408).

iii. Neuromuscular

The common neuromuscular disorders warranting surgeries are: neurofibromatosis, spinal muscular atrophy (SMA), cerebral palsy, Friedreich's ataxia, and Duchenne's muscular dystrophy (DMD). The surgical treatment is guided by the natural history of such disorders, as well as the expected survival rate. Scoliosis in neurofibromatosis could be either dystrophic (neuromuscular) or non-dystrophic (idiopathic type), and it is notorious for developing pseudoarthrosis. SMA presents with long C-shaped spinal deformity and a collapsing deformity of the chest wall (i.e. parasol deformity) with a vertical inclination of the ribs. Neuromuscular scoliosis secondary to cerebral palsy produces mainly two types of curves: i) thoraco-lumbar short segment deformity with hyperlordosis in hypertonic or spastic children, and ii) a long 'C' shaped collapsing curve in hypotonic or flaccid children. The most common cause of death in children with Friedreich's ataxia is cardiomyopathy, and surgery should ideally be performed before the onset of significant cardio-pulmonary compromise. Children with DMD should be operated as soon as they lose ambulant potential, or their curve approaches 40 degrees (whichever is earlier). Delaying surgery in DMD to a later date will pose significant challenges post-operatively, owing to rapidly deteriorating pulmonary function and vital capacity. The presence of pelvic obliquity mandates fixation to pelvis in such group of patients, in order to level the pelvis and optimize seating balance in a wheel-chair. My pioneering work on improvement in pulmonary function in six children with early-onset scoliosis secondary to neuromuscular disorders (EOS-NMD) following MdGR insertion with a minimum follow-up of two years was presented at the Scoliosis Research Society (SRS) annual meeting 2013 at Lyon, France. The same has also been published in a reputed peer-reviewed indexed scientific journal this year (Spine 2014, 39(15): 1196-1202). The published scientific article is attached as paper no. 8 in the *Innovations and Recent Advances* chapter (i.e. Chapter no. 5).

Chapter 2

LIST of PUBLISHED PAPERS

Advances in Surgical Techniques for EOSD

Growth & Generic (4)

1) Spinal growth and a histologic evaluation of the Risser grade in idiopathic scoliosis. Noordeen MH, Haddad FS, Edgar MA, Pringle J. *Spine* (Philadelphia, PA 1976). 1999 Mar 15; 24(6): 535-8. PMID: 10101816

2) Lordoscoliosis and large intrathoracic airway obstruction: Bartlett W, Garrido E, Wallis C, Tucker SK, Noordeen H. *Spine* (Philadelphia, PA 1976). 2009 Jan 1; 34(1): E59-65. PMID: 19127151

3) Spinal manifestations in a patient with congenital insensitivity to pain: Tsirikos AI, Haddo O, Noordeen HH. *J Spinal Disord Tech.* 2004 Aug; 17(4): 326-30. PMID: 15280764

4) Minimizing complications with single submuscular growing rods: a review of technique and results on 88 patients with minimum two-year follow-up: Farooq N, Garrido E, Altaf F, Dartnell J, Shah SA, Tucker SK, Noordeen H. *Spine* (Philadelphia, PA 1976). 2010 Dec 1; 35(25): 2252-8. PMID: 21102301

Innovations and Recent Advances (4)

5) In vivo distraction force and length measurements of growing rods: which factors influence the ability to lengthen?: Noordeen HM, Shah SA, Elsebaie HB, Garrido E, Farooq N, Al Mukhtar M. *Spine* (Philadelphia, PA 1976). 2011 Dec 15; 36(26): 2299-303. PMID: 21494191

6) Next generation of growth-sparing techniques: preliminary clinical results of a magnetically controlled growing rod in 14 patients with early-onset scoliosis. Akbarnia BA, Cheung K, Noordeen H, Elsebaie H, Yazici M, Dannawi Z, Kabirian N. *Spine* (Philadelphia, PA 1976). 2013 Apr 15; 38(8): 665-70. PMID: 23060057

7) Early results of a remotely-operated magnetic growth rod in early-onset scoliosis. Dannawi Z, Altaf F, Harshavardhana NS, Elsebaie H, Noordeen H. *Bone Joint J*. 2013 Jan; 95-B(1): 75-80. PMID:23307677

8) Magnetic growth rods improve pulmonary function in early-onset scoliosis secondary to neuromuscular disorders. Yoon WW, Sedra F, Shah SA, Noordeen MHH et al. *Spine* (Philadelphia, PA 1976). 2014 Jul 1; 39(15): 1196-1202. PMID: 24825149.

Surgical Patents (1)

1) Surgical Distraction Device Patent. (Internaitonal patent application number PCT/GB2001/001668 and (International publication number WO 01/78614 A1).

Congenital & Syndromic Scoliosis (6)

9) Congenital spinal deformity: a comprehensive assessment at presentation: Basu PS, Elsebaie H, Noordeen MH. *Spine* (Philadelphia, PA 1976). 2002 Oct 15; 27(20): 2255-9. PMID: 12394903

10) Short anterior instrumented fusion and posterior convex non-instrumented fusion of hemivertebra for congenital scoliosis in very young children: Garrido E, Tome-Bermejo F, Tucker SK, Noordeen HN, Morley TR. *Eur Spine J*. 2008 Nov; 17(11): 1507-14. E-pub 2008 Sep 27. PMID: 18820956

11) Anterior Instrumentation and Correction of Congenital Spinal Deformities Under Age of Four Without Hemivertebrectomy - A New Alternative: Elsebaie HB, Kaptan W, El Miligui Y, El Masry MA, Salaheldine M, Noordeen HM, Akbarnia BA. *Spine* (Philadelphia, PA 1976). 2010. 35(6): E218-222. PMID: 20195198

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Chapter 3

GROWTH & GENERIC

Advances in Surgical Techniques for EOSD

Paper: 1

Crankshaft phenomenon is a significant problem in the growing spine. It is defined as a progression of scoliosis and a rotational deformity after spinal fusion and instrumentation in the absence of 'adding-on' and pseudoarthrosis. Spinal growth potential depends on skeletal age and whether tri-radiate cartilages are open. Riser grading (i.e. appearance of iliac apophysis) has also been traditionally used to evaluate the residual growth potential of an immature spine.

Conventionally Riser grade zero to three is considered to be skeletally immature, and Riser grade four and five are considered to be close to completion of growth with minimal or absent residual growth potential. The longitudinal growth of the spine is through secondary ossification centers in the vertebral end-plates, and no study existed that correlated the histological characteristics of these ossification centers with the Riser grade. I was one of the first to conduct such an investigation. Thirty four patients (three males and thirty one females) who underwent anterior release for the correction of idiopathic scoliosis had superior and inferior end-plates harvested and assessed histologically for the degree of growth activity, correlating them with the Riser grade, chronological age, and pubertal status. Ten out of fourteen patients in Riser grade four had significant growth activity in end- plates and only Riser grade five was an absolute indicator of cessation of vertebral growth in AIS. This study challenged the previously believed notion that individuals with only Riser grade zero to three and open tri-radiate cartilages, developed crankshaft phenomenon following spinal fusion. Riser grade four, when coupled with chronological age and time since onset of menarche, was a better representative of growth cessation than the Riser grade on its own.

Spinal growth and a histologic evaluation of the Riser grade in idiopathic scoliosis. Noordeen MH, Haddad FS, Edgar MA, Pringle J. Spine (Philadelphia, PA 1976). 1999 Mar 15; 24(6): 535-8. PMID: 10101816

Advances in Surgical Techniques for EOSD

Paper: 2

Idiopathic scoliosis is characterized by the loss of thoracic kyphosis in the sagittal plane. Severe thoracic hypokyphosis may result in the reduction of the antero-posterior (AP) diameter of the chest, which may manifest as restrictive lung disease with reduced vital capacity. The effects of lordoscoliosis on the main-stem bronchi were not reported.

I observed two patients with severe lordoscoliosis and had right main-stem bronchus obstruction that was confirmed on ventilation-perfusion (VP) scans. Correction of their spinal deformity was imperative, not only for improvement in the coronal Cobb angle, but also to alleviate the bronchial obstruction and optimize lung function by increasing the AP diameter of their chest and the space available for lungs (SAL). Patient one had a growing rod inserted at four years old, which was successful in improving FVC from seventy nine percent of predicted to 100 percent and FEV_1 from sixty five percent of predicted to eighty four percent at their most recent follow-up. Patient two was sixteen years old and had a one-off posterior definitive spinal fusion with improvement in FVC from twenty eight percent to fifty four percent and FEV_1 from twenty five percent to forty six percent of predicted at the final follow-up. Comparison of post-op VP and 3D computed tomography (CT) scans with the pre-op ones demonstrated an alleviation of main-stem bronchus compression and an increased SAL in mm^3. I offered the caregivers early spinal deformity corrective surgery to prevent irreversible compromise in lung function and vital capacity. Evaluation for main stem bronchus compression has been a standard of care during pre-operative evaluation in all my patients over the years to minimize post-operative pulmonary complications by the aggressive institution of chest physiotherapy.

Lordoscoliosis and large intrathoracic airway obstruction: Bartlett W, Garrido E, Wallis C, Tucker SK, Noordeen H. Spine (Philadelphia, PA 1976). 2009 Jan 1; 34(1): E59-65. PMID: 19127151

Advances in Surgical Techniques for EOSD

Paper: 3

Congenital insensitivity to pain continues to be a diagnostic and therapeutic challenge. It can be inherited as an autosomal dominant, an autosomal recessive or a sporadic pattern. Co-existent peripheral neuropathy with an absence of potential withdrawal to painful stimulus is common. Charcot's arthropathy may develop and manifest as a spinal deformity, warranting a spinal surgeon's expertise.

A ten year old Russian boy born out of wedlock from a non-consanguineous marriage with self-mutilation tendencies was admitted under my care for an evaluation of a progressive thoraco-lumbar deformity, and investigations revealed L1-L2 vertebral destruction. Genetic testing and a detailed neurological assessment established it to be 'Hereditary motor sensory neuropathy (HMSN) type V', ruling out Riley-Day and Lesch-Nyhan syndromes. He was operated on at age eleven with combined instrumented anterior-posterior fusion, using a rib graft as an anterior strut. He did well for five years, only to present later with a L4-L5 destructive lesion and lumbar kyphosis. Another surgery in the form of single stage posterior spinal fusion from L3-L5 was undertaken to provide stability and arrest the progression of the deformity. His tingling, numbness, and radiculopathy also improved significantly post-operatively, and he was doing great at his most recent follow-up.

This was the first male patient with HMSN type V to present with a spinal deformity in published English medical literature. Only two other patients were reported earlier (both females, who were aged seventeen and twenty eight respectively).

Spinal manifestations in a patient with congenital insensitivity to pain: Tsirikos AI, Haddo O, Noordeen HH. J Spinal Disord Tech. 2004 Aug;17(4):326-30. PMID: 15280764

Advances in Surgical Techniques for EOSD

Paper: 4

The surgical management of early-onset scoliosis by instrumentation could be broadly categorised into three types: i) Self growing rods with wires (Luque-trolley), ii) Vertebral expandable prosthetic titanium rib (VEPTR) and iii) Growing rods (GR). They (i.e. GRs) act as an internal scaffold guiding the growth of the spine with age, and improving the Cobb angle and the space available for lungs.

I reported clinic-radiological parameters and surgical results of single submuscular growing rods with a minimum follow-up of two years along with another Consultant (S Tucker) in a series of eighty eight patients. The etiologies included: i) Idiopathic (28), ii) Syndromic (22), iii) Neuromuscular (20) and Congenital (18). The mean age at surgery was 7 years (range of 1.4 to 12.1 years), and the mean follow-up was 3.5 years old (range of 2 to 6.8 years). The mean Cobb angle at time of index surgery (GR insertion) was 73.3^0 and mean improvement in Cobb angle was 31.8^0 with a mean loss of 2.5^0 at final f/u. 30 of the 88 patients had definitive spinal fusion at the time of final follow-up. The complications included wound infections in 11 (superficial-8 and deep-3), rod fractures in 31 (which was the most common complication), and anchor failure or migration (proximal – 10 and deep – 6).

This is one of the largest cohorts of patients treated at a single center, with reports on the surgical results of single submuscular growing rods in the English language.

Minimizing complications with single submuscular growing rods: a review of technique and results on 88 patients with minimum two-year follow-up: Farooq N, Garrido E, Altaf F, Dartnell J, Shah SA, Tucker SK, Noordeen H. Spine (Phila Pa 1976). 2010 Dec 1;35(25):2252-8. PMID: 21102301

SPINE Volume 24, Number 6, pp 535–538

Spinal Growth and a Histologic Evaluation of the Risser Grade in Idiopathic Scoliosis

M. H. H. Noordeen, FRCS (Orth), Fares S. Haddad, FRCS (Orth), Michael A. Edgar, FRCS, and Jean Pringle, FRCP

Study Design. Thirty-four patients with idiopathic scoliosis who underwent anterior spinal surgery as part of the correction of spinal deformity were studied prospectively. Superior and inferior endplates were harvested and examined histologically for evidence of residual growth activity. This was then correlated with Risser grades, chronologic age, and pubertal status.

Objectives. To clarify the correlation between Risser grade and vertebral endplate growth potential in patients with idiopathic scoliosis.

Summary of Background Data. The importance of longitudinal spinal growth in patients with idiopathic scoliosis and its correlation with curve progression and the crankshaft phenomenon after posterior fusion are well recognized. The Risser grade, which shows the extent of excursion of the iliac apophysis on serial plain radiographs, is commonly used to estimate residual spinal growth. However, the correlation between the Risser grade and vertebral endplate growth potential in patients with idiopathic scoliosis remains unclear.

Methods. Superior and inferior endplates were harvested from these patients and examined histologically for evidence of residual growth. This was correlated with Risser grade, chronologic age, and pubertal status.

Results. Risser Grade 5 was found to be the only indicator of cessation of vertebral growth in idiopathic scoliosis. Of the 14 patients with Risser Grade 4, 10 showed significant growth activity in the vertebral endplates. The reliability of Risser Grade 4 increases when combined with chronologic age and time since menarche in female patients.

Conclusions. The crankshaft phenomenon is reported to occur only in patients with Risser Grade 2 or less, particularly those with open triradiate cartilages. Our findings of significant endplate growth activity, even in patients with Risser Grade 4, make it unlikely that the crankshaft phenomenon is caused purely by longitudinal spinal growth. [Key Words: scoliosis, Risser, spinal growth, endplates] **Spine 1999;24:535–538**

The prediction of curve progression in untreated patients is a major challenge to orthopedic surgeons. Progression of scoliosis and rotational deformity after spinal fusion and instrumentation, in the absence of "adding on" or pseudarthrosis, has been labeled the crankshaft phenomenon, and remains a significant problem in spinal surgery.[9,10,20]

From The Royal National Orthopaedic Hospital, Middlesex; and The Middlesex Hospital, London, United Kingdom.
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Device status category: 1.

These phenomena have been related to longitudinal spinal growth, and there are several parameters in current use that help in determining spinal growth.[15,16] The excursion of the iliac apophysis (the Risser grade) has been widely used since it was first described in 1958,[18] but there is considerable controversy regarding its reliability in predicting spinal growth.

We have been unable to identify any studies in which the Risser grade has been correlated with vertebral endplate histology. This prospective study was designed to ascertain the value of the Risser grade in predicting spinal growth, by comparing it with vertebral endplate histology in patients with idiopathic scoliosis.

Patients and Methods

From September 1991 through January 1995, vertebral endplates were harvested from patients with idiopathic scoliosis who underwent anterior spinal surgery for lumbar and thoracolumbar curves. The total number of patients was 43. The medical records of 7 patients were incomplete. On reviewing the records of the remaining 36 patients, 2 patients, both male, were found to have congenital scoliosis and were therefore also excluded. This left 34 patients, 31 females and 3 males. The average age of the female patients at the time of surgery was 17 years 8 months (range, 12–31.5 years), and of the male patients was 15 years and 7 months (range, 13 years 10 months to 18 years).

Markers of physiologic maturity including breast development, menarche, and the development of pubic and axillary hair were noted. Patients with previous illnesses, injuries, or musculoskeletal disorders that could affect vertebral development were excluded. Radiographs of the spine and pelvis obtained at the time of anterior spinal fusion were evaluated by two independent assessors to estimate the Risser grade, to determine the magnitude of scoliosis curve, and to rule out congenital vertebral anomalies. The triradiate cartilage of the acetabulum and the vertebral apophyses were also evaluated.

The superior and inferior endplates were harvested at surgery at every surgical level. The discs were excised before the vertebral endplates, which were excised intact using osteotomes. The superior, inferior, convex, and concave parts of the endplates were assessed at every level fused. The endplates were immediately fixed in 10% formal saline before they were transported to the pathology department. After 24 hours they were decalcified in buffered formic acid for 2–10 days. The end point of decalcification was radiographically assessed. After decalcification, the specimens were processed in paraffin wax. The embedded blocks were sectioned 4–5 μm in thickness, and 4–33 longitudinal sections of vertebral endplate were made in each patient and stained with hematoxylin and eosin. Specimens were examined under light microscopy for evidence of

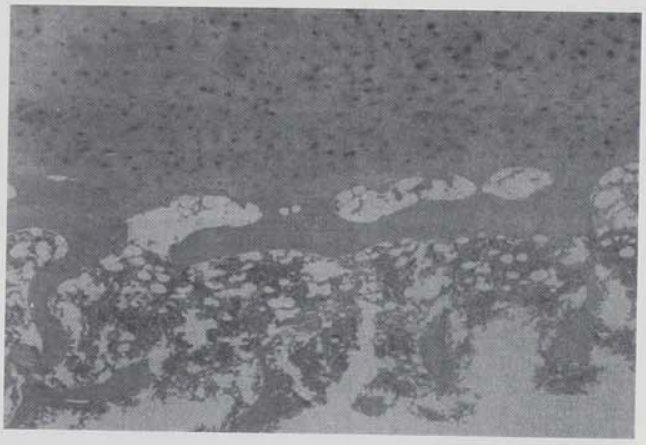

Figure 1. An example of histologic Grade 0 showing no proliferative cartilage zone and no growth activity.

and to determine the extent of residual growth plate activity and vertebral growth potential.

Histologic Grading of Vertebral Endplates. On the basis of the presence or absence and extent of the proliferative cartilage zone in the longitudinal sections of vertebral endplate, we identified four histologic grades as follows:

1. Grade 0: No proliferative cartilage zone and no growth activity (Figure 1).
2. Grade I: No growth activity, but areas of proliferative cartilage zones present (Figure 2).
3. Grade II: Areas of growth inactivity and areas of proliferative cartilage zones (Figure 3).
4. Grade III: A proliferative cartilage zone throughout the section (Figure 4).

Histologic Grade 0 and Grade I were not considered to represent significant vertebral growth. In histologic Grade II, inactive areas may represent the Hueter–Volkmann "epiphyseal pressure rule," which states that growth plate becomes inactive in areas of increased pressure.[18] These areas may represent the concave sides of the scoliotic curves. Active areas of the proliferative cartilage zones in Grade II were considered significant for continued vertebral growth.

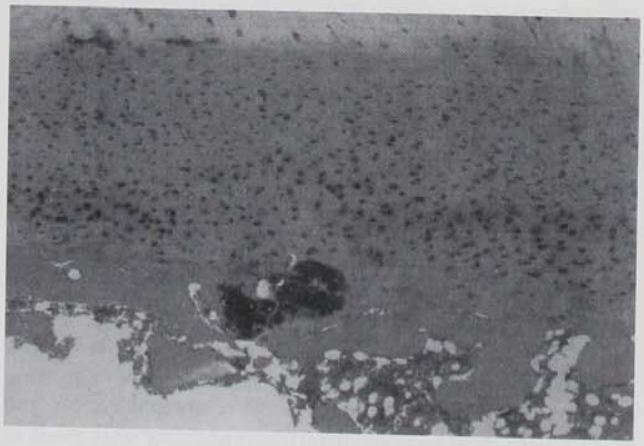

Figure 2. An example of histologic Grade I showing some proliferative cartilage zones but no growth activity.

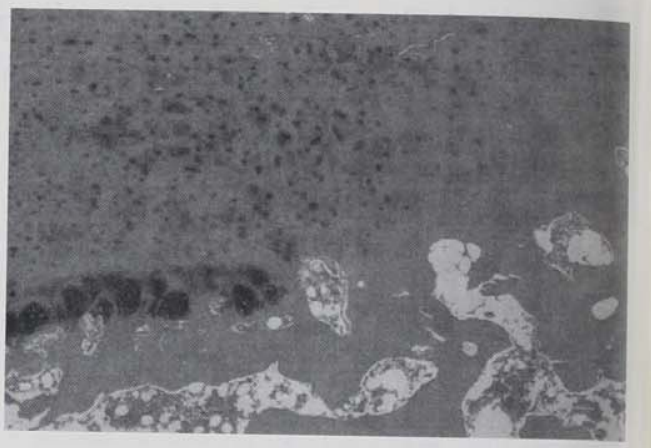

Figure 3. An example of histologic Grade II showing areas of growth inactivity and proliferative cartilage zones.

■ Results

At the time of anterior spinal fusion, there were 5 patients classified as Risser Grade 0, 2 as Risser Grade 1, 2 as Risser Grade 2, four as Risser Grade 3, 14 as Risser Grade 4, and 7 as Risser Grade 5. In 1 patient classified as Risser Grade 3, fusion with the ilium was achieved with this short excursion soon after surgery. This patient was classified as Risser Grade 4.

Histologic analysis of the vertebral endplates of all patients classified as Risser Grade 0 showed Grade III growth activity that was present uniformly in all the longitudinal sections of the endplates of each patient. Of the two patients classified as Risser Grade 1, one showed Grade II activity and the other Grade III activity. (The patient with Grade II activity was a 12-year-old girl. At the time of anterior spinal fusion, her Cobb angle was 63°, and she had had menarche at the age of 11 years and 6 months.) Of the two patients classified as Risser Grade 2 and 4 patients as Risser Grade 3, all showed Grade III growth activity, present uniformly in all histologic sections. Of the 14 patients classified as Risser Grade 4, all were female with a mean age of 16 years 7 months. One patient showed no growth activity on histologic analysis

Figure 4. An example of histologic Grade III showing proliferative cartilage zones throughout the section.

Table 1. Correlation of Clinical Parameters With Histologic Grades in Risser Grade 4 Patients

Histologic Grade	No. of Patients	Time From Menarche (yr)	Mean Age (yr)
0 and I	4	>4	17.92
II	10	<4	16.08

of her endplate, 3 patients showed Grade I growth activity, and 10 patients showed grade II growth activity in all histologic sections. All patients classified as Risser Grades 1, 2, and 3 in this study were premenarcheal.

The mean age of the patients in Risser Grade 4 and histologic Grade II was 16 years 1 month, (range, 14 years 2 months to 18 years 7 months). For those with histologic Grades 0 or I, the mean age was 17 years 11 months (range, 7 years 5 months to 18 years 3 months). These data are summarized in Table 1. All patients in Risser Grade 4 and histologic Grade II were operated on within 4 years of menarche. All patients in Risser Grade 4 and histologic Grades 0 and 1 were operated on at least 4 years after menarche. The mean age at menarche in patients in Risser Grade 4 was 13.9 years (range, 12–16 years). Five patients in Risser Grade 5 showed no growth activity; they were all more than 25 years of age. Two patients showed only focal activity (Grade I) and both were less than 19 years of age (Table 2).

No difference was detected between residual growth activity in lower thoracic and lumbar vertebrae. Radiographs of the pelvis showing the triradiate cartilage were available in only seven patients, all of whom showed closed triradiate cartilage. Vertebral ring apophyses were not clearly visible on any of the spine radiographs.

■ Discussion

Orthopedic surgeons involved in the management of patients with idiopathic scoliosis have a special interest in the assessment of spinal growth, because both the natural history of the disease and its management, operative or nonoperative, are related to the growth potential of the spine.

Although physeal growth occurs in a similar manner in the vertebrae as in the rest of the skeleton and is subject to the same governing factors, there is no completely satisfactory method of determining the degree of spinal maturity in children and adolescents. chronologic age is a poor determinant of subsequent spinal growth. The patient's weight and height and the development of secondary sexual characteristics are also inaccurate methods of determining skeletal maturity.

The maturation of vertebral apophyses is difficult to visualize. It also bears has no relation to longitudinal vertebral growth. Its fusion occurs over several years with different timing in different regions of the spine.[2,3] In the current series, the vertebral ring apophysis could not be visualized in any patient.

Although the Risser grade continues to be an accepted prognostic sign in the evaluation of patients with idiopathic scoliosis, there is considerable controversy about its reliability in predicting the cessation of vertebral growth.[4,7,8,14,21,22] Histologic analysis has shown considerable residual growth activity in adolescent idiopathic scoliosis patients with Risser Grades 1 to 4. This activity may well reflect residual growth potential.[5,11,12] Ten (71%) of 14 patients classified in Risser Grade 4 showed Grade II growth activity. This indicates that that Risser Grade 4 alone may be an unreliable predictor of vertebral growth. Its reliability increases when it is combined with other parameters, including the time since menarche. All patients in Risser Grade 4 who underwent surgery 4 years after menarche did not show significant growth activity in their endplates. All those in Risser Grade 4 who were operated on within 4 years of menarche showed significant Grade II growth activity.

Thus, in spite of difficulties in the prediction of physiologic activity from static histology, Risser Grade 5 appears to be the only indicator of the absolute cessation of vertebral growth. This confirms the clinical observations of Calvo,[6] Zaoussis and James,[25] Urbaniak et al,[23] and Karol et al.[13] In histologic Grade II, in which part of the vertebral endplate shows good growth activity and part of it shows complete fusion of the growth plate, the inactive areas may be because of a pressure effect on the vertebral endplate on the concave side of the scoliotic curve (Hueter–Volkmann epiphyseal pressure rule; Risser 1958). Arguments against this include the work of Bernick,[1] who was unable to show such a phenomenon in normal human vertebrae, only observing a gradual reduction in the width of the growth cartilage up to the age of 16–20 years.

For better understanding of Grade II activity, we need vertebral endplates from nonscoliotic Risser Grade 4 subjects as a control group. The width of the growth plate starts decreasing at the age of 10–15 years.[1] It is not possible, however, to predict accurately the number of years of growth left by looking either at the width or the histologic grade of the vertebral endplate, until and unless we have control subjects without scoliosis.

Although longitudinal vertical growth occurs primarily by growth through the endplate,[2] the neurocentral synchondrosis has been recognized as the major contributor to the growth of the posterior vertebral body.[19,24] The crankshaft phenomenon is reported to occur only in

Table 2. Correlation of Risser Grades and Histologic Grades

Risser Grade	Histologic Grade 0	I	II	III
0	–	–	–	5
1	–	–	1	1
2	–	–	–	2
3	–	–	–	4
4	1	3	10	–
5	5	2	–	–

patients classified in Risser Grades 0 to 2 and particularly in those with the open triradiate cartilages of the acetabulum.[17,20] Our findings of considerable residual growth activity in patients in Risser Grade 4 make it unlikely that longitudinal spinal growth alone could cause this phenomenon.

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Address reprint requests to

Fares S. Haddad, FRCS
46B Hanover Gate Mansions
Park Road
London NW1 4SN
England

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Lordoscoliosis and Large Intrathoracic Airway Obstruction

William Bartlett, MRCS (Ed),* Enrique Garrido, MRCS (Ed),* Colin Wallis, MD, FRCP,† Stewart K. Tucker, FRCS (Orth),* and Hilali Noordeen, FRCS (Orth)*

Study Design. Case series.

Objective. We report the treatment of 2 children with right main bronchus obstruction complicating thoracic lordoscoliosis.

Summary of Background Data. The preoperative investigation and treatment of large airway obstruction caused by lordoscoliosis has not been reported in the literature.

Methods. Obstruction of the right main bronchus was confirmed before surgery by ventilation-perfusion scans, bronchogram, and computed tomography scan. Deformity correction was achieved using a submuscular growth rod construct in one child, and posterior spinal fusion in the other. Clinical examination and repeat ventilation-perfusion scans were performed 8 weeks after surgery.

Results. In both children, ventilation to the right "convex" lung was reestablished after surgery. Lung function improved in both patients after surgery.

Conclusion. This is the first report of large airway obstruction associated with thoracic lordoscoliosis in which ventilation was reestablished after spinal deformity correction. Early deformity correction is indicated in such cases because of the risk of irreversible compromise to lung ventilation and perfusion.

Key words: ventilation, perfusion, bronchus, lordosis, lordoscoliosis, obstruction. **Spine 2009;34:E59–E65**

The deleterious effects of spinal deformity on pulmonary function are widely recognized. Thoracic lordoscoliosis, in particular, is associated with respiratory compromise.[1,2] Reduced anteroposterior diameter of the thorax decreases the total lung capacity, whereas loss of rib mobility and normal chest wall expansion reduce lung compliance. Torsion of large airways in kyphoscoliosis and extrinsic compression of the right main bronchus after compression instrumentation to correct thoracic hypokyphotic scoliosis has been described.[3,4] We report on 2 children who presented with obstruction of the right main bronchus due to thoracic lordoscoliosis. After deformity correction, the patient's chronic pulmonary symptoms resolved and ventilation and perfusion to the right lung was reestablished.

From the Departments of *Spinal Surgery and †Respiratory Paediatrics, Great Ormond Street Hospital for Children, London, UK.
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Address correspondence ad reprint requests to William Bartlett, MRCS (Ed), Great Ormond Street Hospital for Children, Great Ormond Street, London WC1N 3JH, UK; E-mail: william.bartlett@gmail.com

Case 1

After a normal pregnancy, the child was born vaginally at term. She reached milestones within normal limits and was well until age of 19 months. She developed a cough that rapidly led to respiratory compromise necessitating intubation. The child made a good initial recovery though continued to suffer from recurrent chest infections and poor respiratory function.

Examination of the child's chest aged 4 demonstrated an expiratory wheeze to the right of the sternum. Lung function tests confirmed severe restrictive lung disease. Bronchogram (Figure 1) demonstrated obstruction of the right main bronchus, not overcome with 15 mm H_2O. Ventilation-perfusion studies were then performed (Figures 2-3). These demonstrated absent ventilation and decreased perfusion in the right lung while lying supine. When the patient was positioned prone, a full return of ventilation to the right side occurred. Right lung perfusion was 27% of cardiac output when supine, improving by 10% after 10 minutes in the prone position.

A computed tomography (CT) scan of the chest confirmed a narrowing of the right main bronchus that appeared to be secondary to the thoracic lordoscoliosis (Figure 3). An orthopaedic opinion was then sought. Radiographs confirmed a coronal Cobb angel from T3 to T8 of 56 degrees. Thoracic lordosis from T5 to T9 measured 34°C with apex at T7 (Figure 4). In addition, Café au lait skin lesions and freckling were noted, consistent with neurofibromatosis.

The patient underwent posterior submuscular single growing rod insertion at the age of 4 years. Fixation was by pedicle hooks proximally and pedicle screw distally. Correction was achieved by distraction of the contoured growth rod (Downsize Synergy Spinal System, Parsippany, NJ). Lordosis was then further corrected by tensioning of 2 sublaminar cables around the growing rod construct (Titanium Atlas Cable System, Medtronic Sofamor Danek).

A repeat ventilation-perfusion scan performed 2 months postsurgery demonstrated a great improvement to both ventilation and perfusion in the right lung (right lung 45% of ventilation and 44% of cardiac output), which was almost comparable to the left lung (Figure 4). Forced vital capacities improved from 0.89l (79% predicted) before surgery to 1.14l (101% predicted) after surgery. Forced expiratory volume improved from 0.65 L (65% predicted) l to 0.84l (84% predicted).

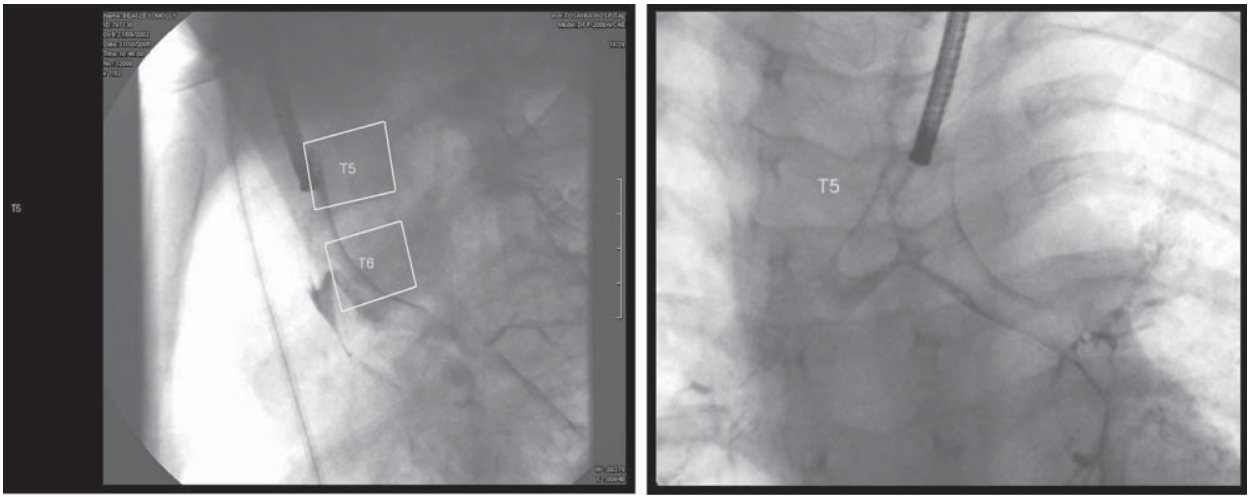

Figure 1. Bronchogram demonstrating oblique and AP views of the obstructed right main bronchus. The obstruction could not be overcome by pressure of 15 mm H_2O.

The patient has undergone 2 uncomplicated lengthening since growth rod implantation. Current coronal Cobb angle measures 20°C and thoracic lordosis 5°C. The patient has not experienced any further chest infections or nocturnal cough.

■ Case 2

A 15-year-old girl was referred to our spinal deformity unit after her mother had become aware of a rib hump and unbalanced shoulders. She was known to have Turner syndrome, although her medical history before adoption at the age of 7 was unknown. She had received growth hormone treatment for short stature from age 8. Her health had been good until the age of 14, when she began to suffer recurrent right lobar pneumonia and exertional dyspnoea. Auscultation of the right base revealed reduced air entry with expiratory wheeze. Lung function test demonstrated a restrictive pattern of lung disease. Radiographs confirmed the presence of a right thoracic lordoscoliosis with a coronal Cobb angel from T5 to L2 of 75°C and a thoracic lordosis T5 to T12 of 26°C (Figure 5). Spinal MRI confirmed the absence of intraspinal pathology. CT of the chest (Figure 6) demonstrated right lower lobe consolidation with obstruction of the right main bronchus that appeared draped over the apex of the lordoscoliotic spine. Ventilation-perfusion

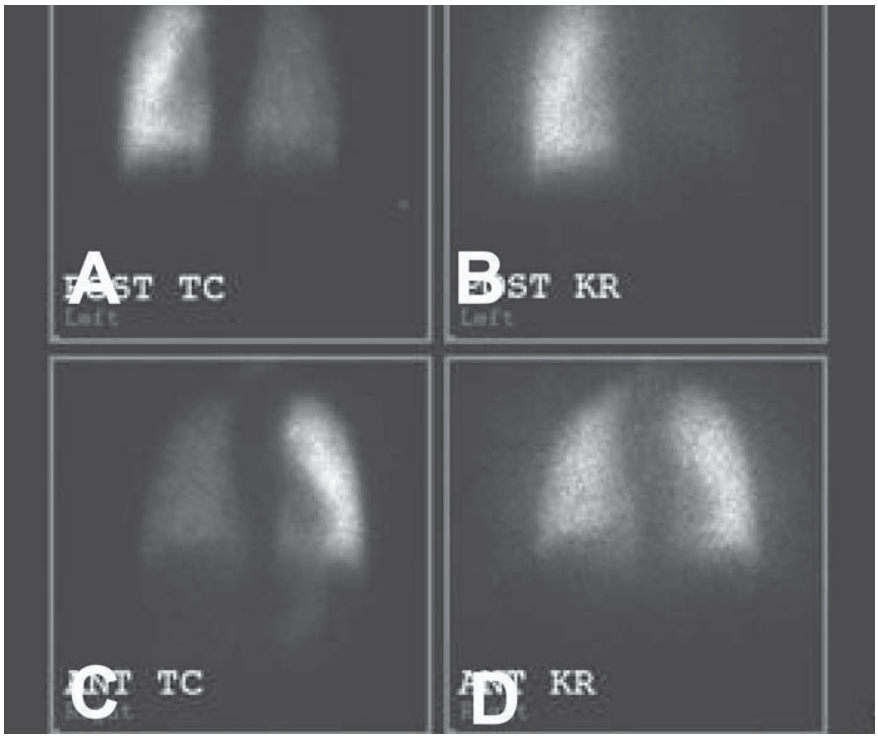

Figure 2. Preoperative ventilation-perfusion scan of case 1. Right lung perfusion is reduced whilst supine (**A**) and ventilation is absent (**B**). When prone, a full return of ventilation to the right side observed (**D**). Perfusion to the right lung was 27% of cardiac output when supine (A) and 37% when prone (**C**).

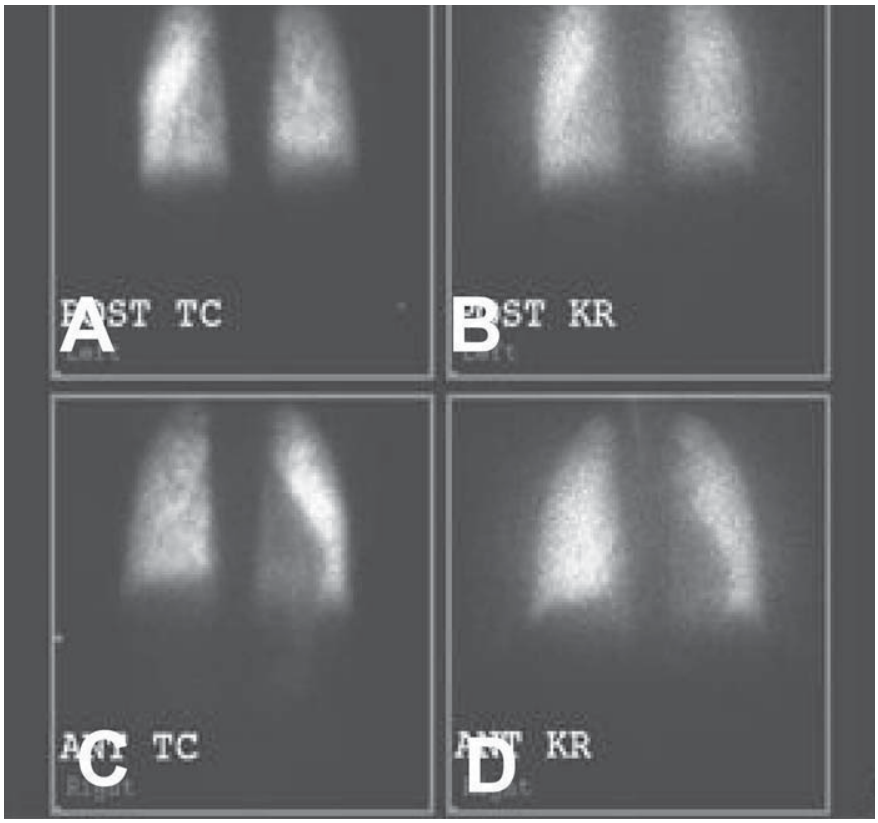

Figure 3. Ventilation-perfusion scan of case 1, performed 2 months postsurgery. Right lung ventilation is improved to 45% of total and is independent of supine (**B**) or prone (**D**) position. Perfusion when supine (**A**) and prone (**C**) was comparable to the left lung.

scan confirmed absent ventilation of the right middle and lower lobe and poor perfusion of the right lung, receiving only 24% of cardiac output. The left lung contributed 74% of total ventilation (Figure 7). Both ventilation and perfusion did not change in prone, standing, or supine position.

In view of the severity of the lordoscoliosis and the presence of obstruction of the right main bronchus obstruction, posterior spinal fusion with correction of the thoracic lordosis was performed. The spine was instrumented with pedicle screws at every level using a 6.3-mm diameter rod to achieve maximal correction of the sagital deformity (Synergy Spinal System).

Thoracic lordosis was corrected to a sagittal Cobb angle T5 to T12 of 3°C (Figure 7) and coronal Cobb angle T5 to L2 was corrected to 21°C. Exertional dyspnoea resolved and at 4 weeks postsurgery and nebulised bronchodilators were discontinued. A ventilation-perfusion scan was performed at 8 weeks postsurgery demonstrated improved ventilation to the right lung which received 37% of total ventilation and improvement in right lung perfusion (Figure 8). Forced vital capacities improved from 0.86 L (28% predicted) before surgery to 1.67 L (54% predicted) after surgery. Forced expiratory volume improved from 0.67 L (25% predicted) to l.22 (46% predicted).

■ Discussion

Respiratory compromise is a well recognized complication of spinal deformity.[1,2] Surgery is frequently indicated to prevent deformity progression and deterioration of pulmonary function. Initially, the deleterious effects of thoracic lordoscoliosis on respiratory function were considered a consequence of reduced lung capacities. Nash and Nevins[5] demonstrated that patients with scoliosis tended to have a decreased ratio of anteroposterior to lateral thoracic diameter and this decreased ratio correlated with pulmonary dysfunction. Winter *et al*[6] subsequently observed lordoscolisiosis to cause more respiratory compromise than kyphoscoliosis. Furthermore, improvements to pulmonary function after surgery were directly correlated to the amount of lordosis corrected. Recently, 3-dimensional CT volumetric analysis in patients with idiopathic scoliosis has shown that higher and more rotated thoracic curves restrict lung volume more than lower and less rotated curves, and that the concave lung is comparatively more affected.[7,8] Furthermore, lung scintigraphy has shown ventilation abnormalities in idiopathic scoliosis to be mainly localized in the concave lung.[9] In contrast, the 2 patients described in this report had severe impairment of ventilation and perfusion in convex lungs.

In addition to reduced lung capacities and altered chest mechanics, the deleterious effects of thoracic lordoscoliosis on respiratory function may result from direct bronchial obstruction. Borowitz[10] reported indirect evidence to suggest relief of left main bronchus obstruction after spinal release in a patient with idiopathic scoliosis. The patient's preoperative lung function tests suggested evidence of both restrictive and obstructive lung disease.

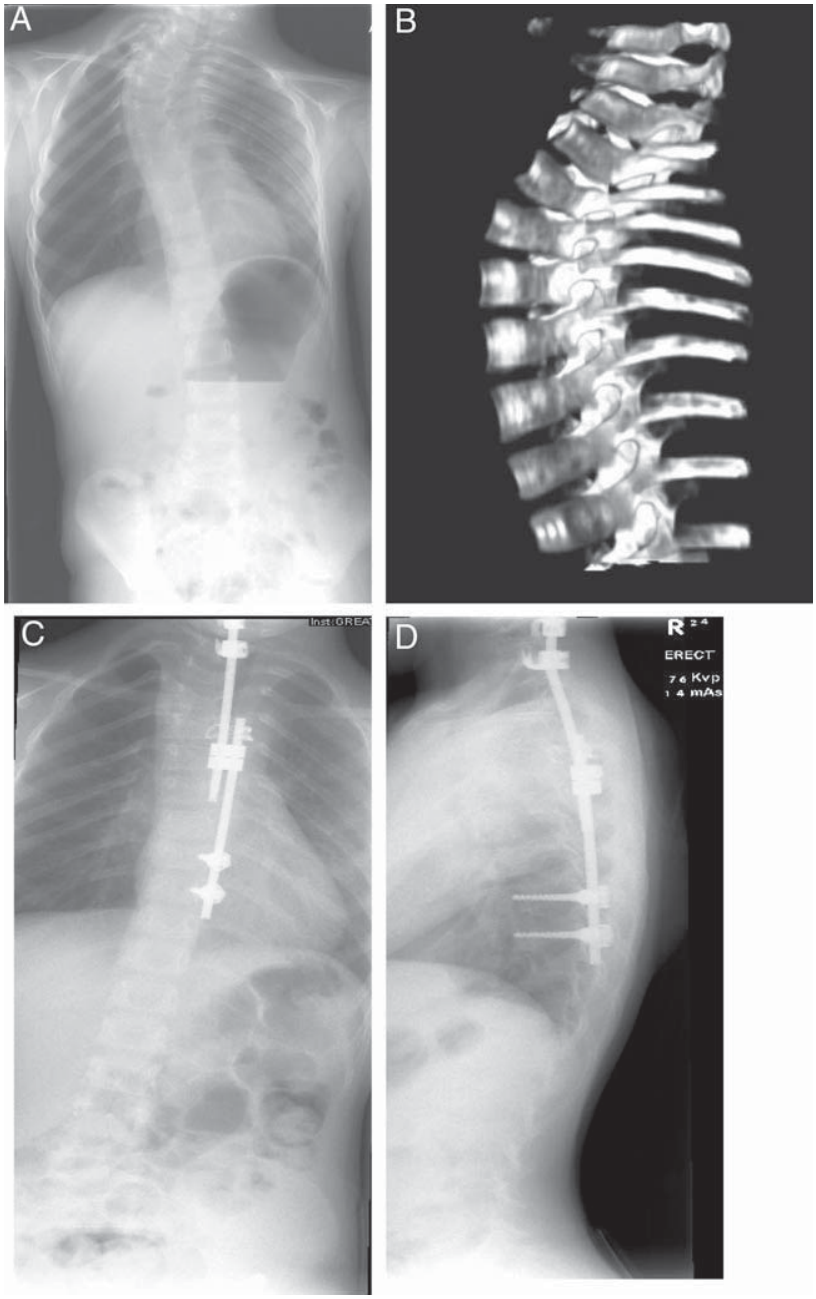

Figure 4. Preoperative AP radiograph (**A**) and sagittal CT (**B**) of case 1 demonstrating severe upper thoracic lordosis. Postoperative AP(**C**) and lateral (**D**) radiographs the correction achieved using submuscular growing rods.

At induction, a double lumen endotracheal tube could not be passed due to extreme deviation of the L main bronchus. After surgery, lung function test were reported to show improved flow, suggesting to the authors that the obstruction had lessened.

Position dependent changes in lung function test have been reported in 1 patient with lordoscoliosis. Holden *et al*[11] found that lung function improved when the patient adopted a forward-flexed position. Similarly, case 1 exhibited a position dependent reversible obstruction; Right lung ventilation and perfusion improved in the prone position. In the developing lung, even temporary airway obstruction may cause irreversible change to lung function.[12] Therefore, evidence of reversible obstruction on ventilation-perfusion scanning indicates the lung to be at risk. Early intervention should be considered in such cases.

The natural history of untreated airway obstruction secondary to spinal deformity is widely recognized. Wee *et al*[13] described the case of a 38-year-old man who presented with chronic cough and intermittent fevers. He was found to have a thoracic lordoscoliosis complicated by right lower bronchus obstruction and atelectasis, which required resection of the emphysematous right lower lobe. Obstruction of the right main bronchus leading to "destroyed lung" after Harrington compression instrumentation has also been recognized.[3,14] Described treatments for this complication have included partial

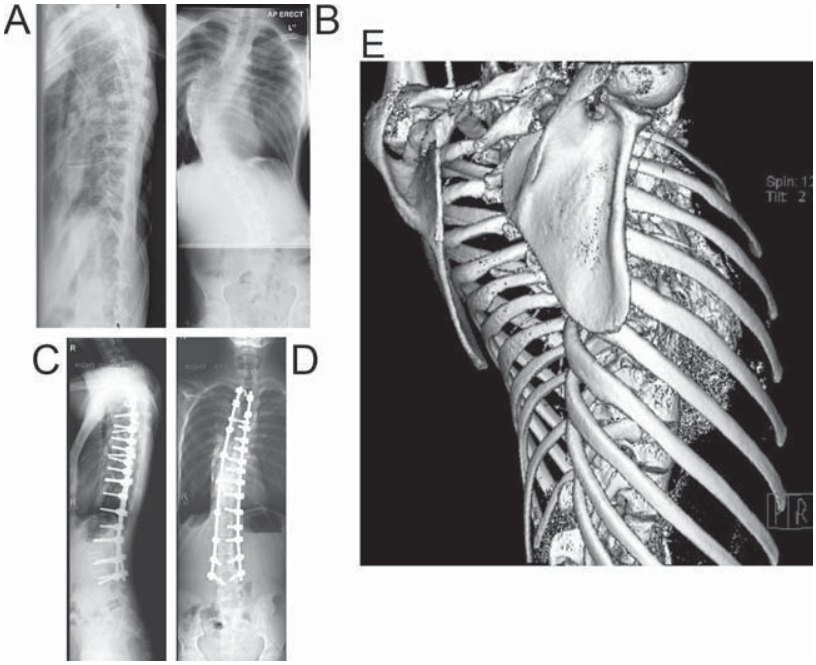

Figure 5. Preoperative lateral (**A**) and AP (**B**) radiographs of case 2. The preoperative thoracic deformity is also demonstrated on CT reconstruction (**E**). Postoperative lateral (**C**) and AP (**D**) radiographs show the position of the correction after posterior spinal fusion.

anterior resection of vertebral bodies[3] and bronchial stenting.[14]

Large airway obstruction due to thoracic lordoscoliosis is probably more common than generally appreciated. We believe that all children with lordoscoliosis require regular monitoring of respiratory function. If such patients develop pulmonary symptoms or abnormal lung function tests, a ventilation-perfusion scan should be promptly performed.[12,15] Abnormal ventilation patterns should be further investigated by 3-dimensional contrast CT. We have found this investigation to be extremely useful in precisely defining the anatomy of the obstruction and in assisting the

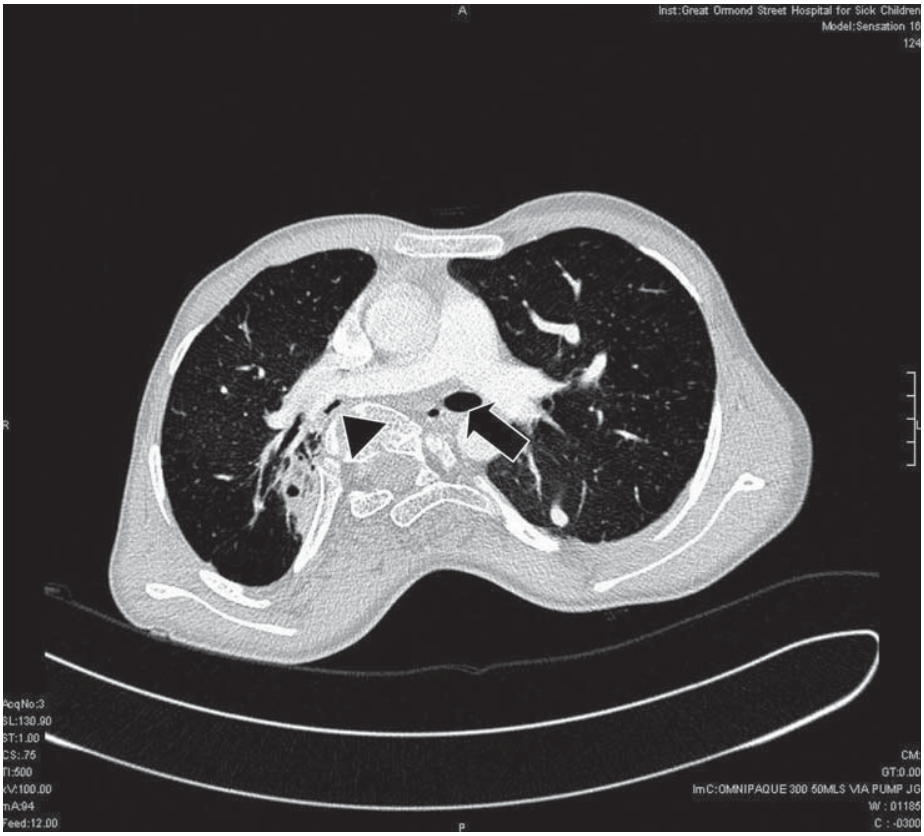

Figure 6. Axial CT of case 2 demonstrating collapse of the right lower lobe bronchus (arrowhead) by compression of the right pulmonary artery and spine. The larger diameter of the unobstructed bronchus to the left lower lobe bronchus is shown as (arrow).

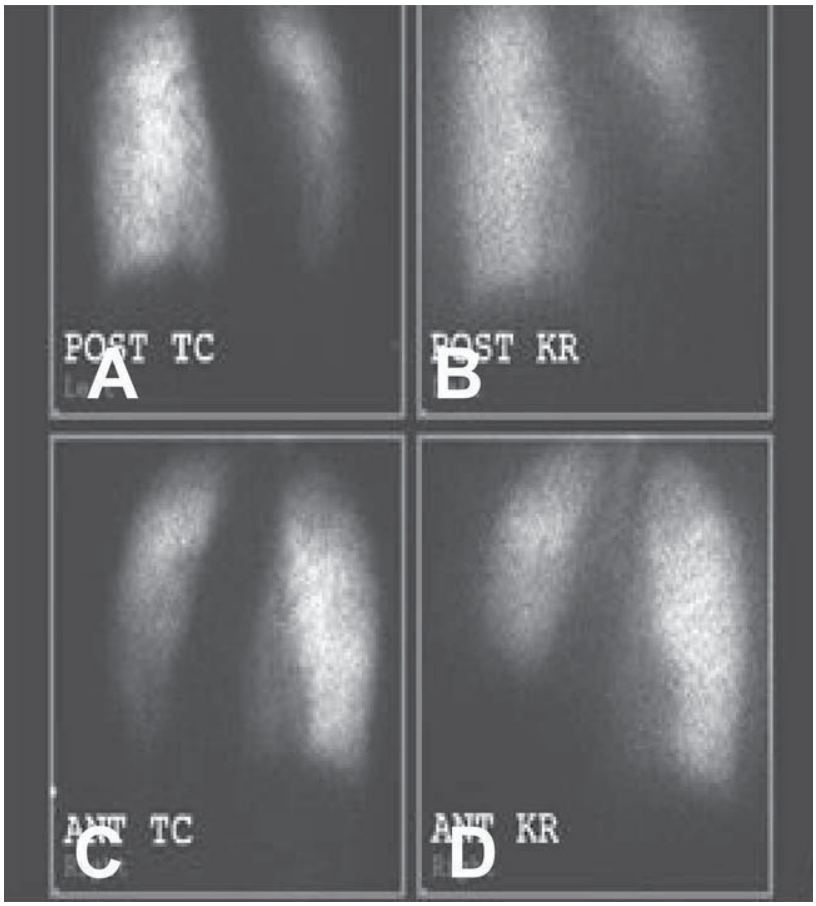

Figure 7. Preoperative ventilation-perfusion scan of case 2, demonstrating poor perfusion of right lower lobe and middle lobe when prone (**A**) and supine (**C**). Ventilation to the right lung was only 26% of total (**B**) and was not improved when prone (**D**) with absent ventilation (B and D).

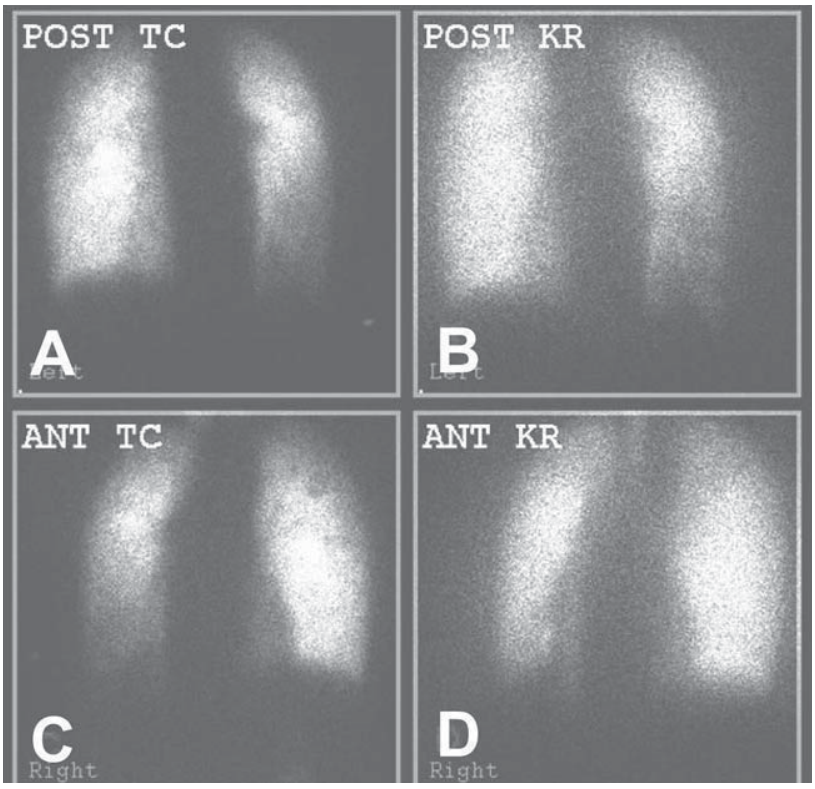

Figure 8. Ventilation-perfusion scan of case 2, performed 3 months postsurgery. Improved perfusion (**A** and **C**) and ventilation (**B** and **D**) of right middle and lower lobe. Right lung ventilation has increased to 37% of total.

surgeon to understand complex relationships within the thorax. Preoperative contrast CT may also lessen unanticipated problems with intubation at the time of surgery and eliminate the need for other invasive procedures such as bronchoscopy.

We believe that this is the first report of right main bronchus obstruction associated with thoracic lordoscoliosis in which ventilation and perfusion were reestablished after spinal deformity correction. Because of the risk of irreversible changes to the lung function, we recommend regular respiratory assessment of children with lordoscoliosis and early deformity correction in those with confirmed obstruction.

■ Key Points

- Focal bronchial obstruction may be associated with thoracic lordosis or lordoscoliosis.
- Bronchial obstruction can be relieved by correction of thoracic lordoscoliosis with return of function to the distal lung.
- Early correction is recommended in patients with large airway obstruction secondary to spinal deformity.

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CASE REPORT

Spinal Manifestations in a Patient with Congenital Insensitivity to Pain

Athanasios I. Tsirikos, MD, Omar Haddo, MD, and Hilali H. Noordeen, MD

Summary: Spinal manifestations in congenital insensitivity to pain are relatively uncommon and easily misdiagnosed. We report on a patient with absent protective pain sensation, who developed spinal neuropathic arthropathy. At age 11 years, he presented with a destructive lesion at the L1–L2 level, causing him tingling sensation in both lower limbs. He was treated with combined anteroposterior spinal fusion from T12 to L3 and had full recovery. Five years later, he presented with a long history of clicking in his low back, muscle weakness and paresthesia in both lower extremities during walking, and evidence of Charcot arthropathy at the L4–L5 level, resulting in junctional kyphosis and canal narrowing. Posterior spinal arthrodesis from L3 to the sacrum was performed, due to lack of patient and parental consent for combined anterior decompression/posterior fusion. The patient resumed normal muscle function and his previous level of activities. Spinal complications should be anticipated in this condition and create diagnostic and therapeutic dilemmas. However, surgical management can produce favorable clinical results.

Key Words: congenital insensitivity to pain, spine

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Congenital insensitivity to pain is a rare condition typically characterized by the absence of aversion or withdrawal response to normally painful stimuli. The condition can be inherited as autosomal recessive or dominant, and in many occasions it has a sporadic presentation.[1,2] These patients constitute an often-unrecognized population, and it was not until the early 1930s when this condition attracted medical attention. Dearborn's first report on a carnival entertainer calling himself the "Human Pin Cushion," who among his other achievements staged a self-crucifixion, provoked much interest.[3]

Historically, different classification systems have been proposed to encompass a number of pathologic entities, which have as a common primary manifestation the absence of perceived pain.[4,5] More recently, the presence of peripheral neuropathy has been considered a criterion for establishing diagnosis of congenital insensitivity to pain, in which the inability to perceive painful sensations is followed by nonresponsiveness to all noxious stimuli.[6] In contrast, individuals with congenital indifference to pain, a term that has been used in the past interchangeably with congenital insensitivity to pain, can identify pain-producing stimuli but lack the normal homeostatic mechanism of pain, while signal transmission is unimpaired.[2]

The purpose of this report is to describe a patient with congenital insensitivity to pain who developed an unusual pattern of recurrent Charcot arthropathy in his lumbar spine and associated neurologic involvement, with the aim to discuss the difficulties in establishing diagnosis and designing appropriate management.

CASE REPORT

Our patient was born in 1985 at term following an uneventful pregnancy, labor, and normal delivery. His birth weight was 3.5 kg. He was a Caucasian boy, the first child of a 25-year-old primigravida mother and nonconsanguineous father. Both parents were originally from Russia, and the maternal grandfather was Jewish. The family history was negative for congenital insensitivity to pain and other neurologic or neuromuscular diseases. He had two younger siblings who were both completely normal. During the neonatal period, he experienced respiratory stridor, which subsided spontaneously. With growth, he achieved normal early developmental milestones.

From infancy, he had been noted to have altered pain sensation and a tendency toward repetitive self-abuse. When teething, he expressed a self-mutilation behavior, constantly biting off the tip of his tongue and chewing his fingertips without experiencing pain. When crawling, he developed abrasions over his knees, which were complicated by superficial infections. At 4 years of age, he sustained a closed fracture of his left os calcis after jumping from a tree. This was treated conservatively and healed in a satisfactory manner. However, he developed cellulitis of the foot, diagnosed when the cast was removed, which required a long period of antibiotic treatment and eventually resolved without further complications and no evidence of osteomyelitis.

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From the Great Ormond Street Hospital for Children, London, UK.
Reprints: Athanasios I. Tsirikos, MD, Department of Orthopaedics, Great Ormond Street Hospital for Children, Great Ormond Street, London, UK WC1N 3JH (e-mail: atsirikos@hotmail.com).

At age 10 years, he was admitted to our hospital for further investigations to establish a diagnosis. On physical examination, he was noted to be between the 50th and 75th percentiles by weight and just above the 25th percentile by height. He was a pleasant, hyperactive child. He had multiple scars and bruises on his upper and lower limbs attributed to previous injuries. His right knee was in slight valgus alignment but had full range of movement and was clinically stable. He was also noted to have a 2-cm leg length discrepancy, with the right lower extremity being shorter than the left.

Central nervous system examination revealed a mildly diminished corneal reflex on the right side but was otherwise noncontributory. Peripheral nervous system examination confirmed normal muscle tone and intact power and coordination, whereas deep tendon, abdominal, and cremasteric reflexes were all present, brisk, and symmetrical. No pathologic reflexes were recorded. Touch, vibration, two-point discrimination, position and vibratory sensibility, and proprioception were normal. Skin surface pinpricking produced apprehension as touch but evoked no pain in any area. The patient was able to differentiate pressure sensation; however, pain and thermal sensitivities were absent. Attempts to generate pain responses by persistent muscular and bone compression, testicular, supraorbital, and sternal pressure, pinching the Achilles tendon, and pricking of the periosteum using a sterile needle were all ineffective. Fine touch sensation was also completely undisturbed. Clinical signs of autonomic dysfunction were also evident, with abnormal sweating and elevated core temperatures. Symptoms due to cerebellar dysfunction were not present, whereas stereognosis was intact. Psychological assessment placed him in the average intelligence range. There were also some behavioral issues, and even though proper education was offered to the patient in regard to his condition over the ensuing years, he did not seem to realize the importance of attempting to prevent painful stimuli from recurring and kept exposing himself to self-inflicted damage through vigorous sports activities.

The imaging studies during his hospitalization included plain radiographs of the lower limbs, which showed fusion of the right proximal tibial physis. This was thought to be the result of repetitive trauma in the absence of normal protective constraints. At that point, a right proximal fibular epiphysiodesis was performed.

Further investigations were carried out consisting of metacholamine eye test, histamine skin test, and nerve conduction studies, both motor and sensory, which did not elicit pathologic responses. Lumbar puncture was performed, and electroencephalogram and karyotype were examined, and all produced normal results. Riley–Day syndrome was excluded on the basis of normal deep tendon reflexes and normal metacholamine test. The diagnosis of hereditary sensory and autonomic neuropathy type V was determined by a geneticist. This was confirmed by a sural nerve biopsy, which showed severe loss of myelinated small-diameter C and A-delta fibers, the neural tracts that are responsible for transmitting pain sensation.

At the age of 11 years, he presented with tingling sensation in both lower limbs and what was thought to be a thoracic scoliotic deformity; however, the scoliosis was undetectable on clinical examination and radiologic review. His neurologic picture remained otherwise unchanged. There was negative history of previous injury. Plain radiographs highlighted a process of disintegration in the L1–L2 intervertebral disc space and adjacent end-plates (Fig. 1A and B). Magnetic resonance imaging (MRI) scans (see Fig. 1C) confirmed the existence of a destructive lesion at the same levels including L1 and L2 vertebral bodies and the L1–L2 disc with no intraspinal encroachment. The differential diagnosis included a traumatic, tumorous, infectious, or arthropathic pathologic process. The patient underwent a computed tomography (CT)–guided biopsy to assist diagnosis, which did not demonstrate any evidence of neoplasm or infection. In the absence of history of traumatic event and based on the imaging appearance, the diagnosis of Charcot arthropathy was made and a combined single-stage anterior–posterior spinal fusion was performed, extending from T12 to L3 with instrumentation used circumferentially. Rib bone autograft was applied anteriorly and autologous bone graft harvested from the spinous processes along with bone substitute posteriorly to enhance a solid 360° fusion. His postoperative recovery was unremarkable, and he was mobilized in the immediate postoperative period in a thoracolumbosacral orthosis, which was applied for 6 months to provide additional external support. Excellent bony fusion was achieved by 12 months after surgery (Fig. 2).

At 12 years of age, he sustained a fracture of the proximal phalanx of his left middle finger with minimal injury. This was treated conservatively and healed uneventfully. Two years later, at age 14, he sustained a Salter–Harris type II fracture of

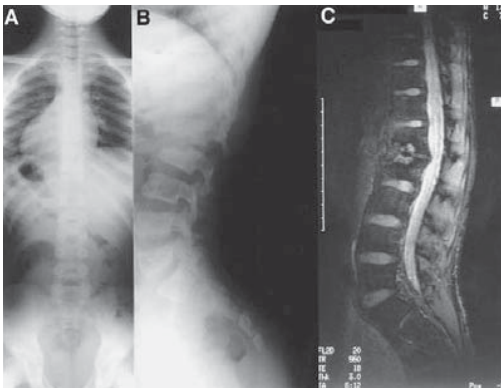

FIGURE 1. Anteroposterior (A) and lateral (B) radiographs and sagittal MRI view (C) of the spine, demonstrating a destructive lesion involving the L1 and L2 vertebral bodies and the L1–L2 disc. There is no evidence of scoliosis.

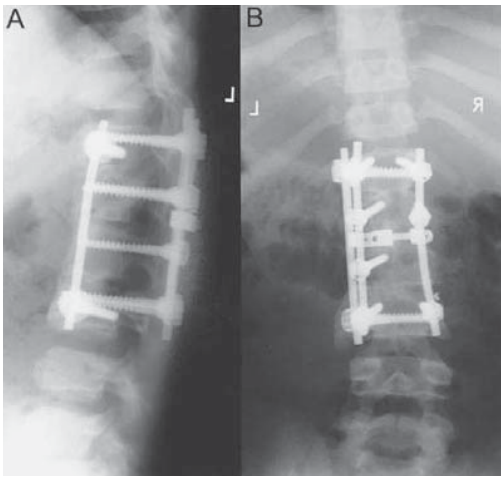

FIGURE 2. Lateral (A) and anteroposterior (B) radiographic views of the spine 1 year after the initial operative procedure confirmed a solid fusion extending from T12 to L3.

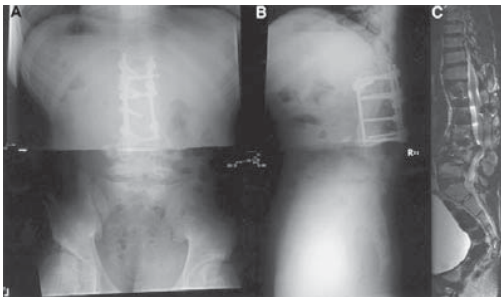

FIGURE 3. Anteroposterior (A) and lateral (B) plain radiographs of the spine revealed extensive degeneration at L4 and L5 levels, with associated anterior wedging, collapse, and extrusion of bony fragments anteriorly at L4 vertebral body. Posterior vertebral height loss at L5 level was also evident. Consequently, the L3–L5 levels were kyphotic, with increased compensatory lordosis at L5–S1 segment. Sagittal MRI view (C) shows the development of junctional kyphosis and spinal canal compression.

his left distal tibia with an associated Salter–Harris type I fracture of the distal fibula while playing soccer, again without significant trauma. Open reduction and internal fixation of both fractures were therefore performed with satisfactory results. At age 15 years, the patient underwent combined left distal femoral and proximal tibial/fibular epiphysiodesis to address a leg length inequality of 3.5 cm.

When he was 16 years old, he presented with a 6-month history of clicking in his back. He was also experiencing episodes of muscle weakness and paresthesia in the lower extremities associated with walking and limiting his physical activities. These symptoms would resolve completely with rest. There was no history of bladder or bowel dysfunction, and the patient denied any traumatic incident. Neurologic examination on admission was noncontributory, revealing normal muscle strength, tendon reflexes, and sensation to light touch in the legs with the patient at rest.

Plain radiographic views (Fig. 3A and B), CT images with three-dimensional reconstruction (Fig. 4A and B), and MRI scans (see Figs. 3C and 4C) demonstrated neurotrophic degeneration at L4–L5 vertebral bodies and adjacent L4–L5 disc, with significant anterior wedging, anterior protrusion of bony fragments, and collapse at L4 segment. In addition, the development of a severe junctional kyphosis could be documented at L3–L4 and L4–L5 levels, causing spinal canal stenosis (see Figs. 3B and C and 4C). There was also loss of height at the L5 vertebral body posteriorly with exaggerated lordosis at L5–S1 level. Due to the neural cord compromise, the patient was proposed an anterior decompression and posterior stabilization. However, due to his previous combined anteroposterior procedure 5 years ago, both he and his parents did not consent to the anterior stage of the operation; consequently, he underwent extension of the posterior spinal fusion from L3 down to the pelvis, which achieved good restoration of both coronal and sagittal spinal balance, using posterior spinal instrumentation, spinous process autogenous bone graft, and supplementary hydroxyapatite bone substitute (Fig. 5). His neurologic picture recovered completely, immediately after surgery, and a second-stage anterior decompression was not deemed to be necessary. The patient was mobilized in an underarm brace for 6 months.

At the most recent follow-up, 2 years after the operation, the patient was totally asymptomatic and had resumed his previous level of activities. Neurologic examination showed full recuperation of muscle power and paresthesias in his lower limbs. Radiographic studies confirmed a solid spinal fusion. His gait was very effective, with the hips, which had normal function and range of motion, compensating for the stiffness of the arthrodesed lumbosacral articulation.

DISCUSSION

Patients with underlying peripheral neuropathies lack normal peripheral afferents due to abnormal sensory nerves, thus exhibiting a constant impairment in discriminating noxious stimuli and appreciating their potentially damaging effect.

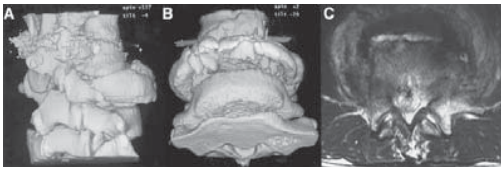

FIGURE 4. CT scans with three-dimensional reconstruction (A and B) and MRI scan (C), illustrating the degree of bony destruction with anterolateral and intracanal osseous fragment and disc material protrusion at L4 vertebra.

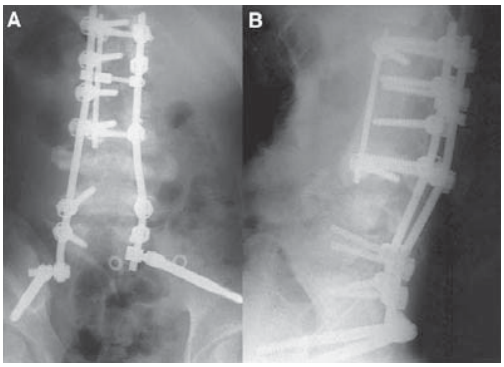

FIGURE 5. Anteroposterior (A) and lateral (B) radiographic images of the lumbar spine after the second surgical procedure, demonstrating extension of the spinal arthrodesis to include the lumbosacral joint with iliac fixation and resultant restoration of coronal and sagittal spinal alignment.

As the result of this inability to interpret stimulus presence and modality, these individuals lack normal pain avoidance and expose themselves to recurrent injuries. The fact that many of the patients with congenital insensitivity to pain represent complex orthopaedic cases or even die in childhood due to consistent failure to interpret traumatic incidents and illnesses has provided additional evidence that retaining an intact perceptual mechanism of pain has significant protective value in humans and determines survival rate and incidence of morbidity.

Other causes of regional or generalized pain perception deficit include space-occupying lesions in the spinal cord, such as syringomyelia, hydromyelia, hematomyelia, and neurofibromatosis, psychiatric pathologies, mental retardation, traumatic, neoplastic, or infectious lesions affecting the central or peripheral nervous system, diabetic neuropathy, alcoholism, leprosy, and tabes dorsalis.[7–9] Thrush[10] has proposed certain clinical criteria for establishing the diagnosis of congenital insensitivity to pain: 1) absent pain sensation since birth, 2) whole-body involvement, and 3) present deep tendon reflexes and other sensory modalities apart from pain apprehension. Bar-On et al[11] reviewed 13 patients with congenital insensitivity to pain and classified three different types of clinical presentation, which provide useful information in the prognosis and treatment of their musculoskeletal problems: type A, in which multiple infections were the most prominent symptom; type B, with numerous fractures, consequent growth disturbance, and avascular necrosis; and type C, with Charcot arthropathy, joint dislocations, infections, and fractures. However, further investigation will be needed, oriented more in the genetic, biochemical, and electrophysiologic patterns of the different subtypes, to establish satisfactory pathophysiology, allow genetic counseling, and provide means for prenatal diagnosis.

In pain sensation–deprived children, major skeletal complications arise early in life from the peculiar habit of self-mutilation, particularly about the face, and from repetitive traumatic events to the hands and feet.[7,12] Other principal orthopaedic manifestations of the disease include recurrent fractures, autoamputation, bone and joint infections, neurotrophic joint disease, mainly evident in the weight-bearing joints, joint dislocations, acro-osteolysis, limb length discrepancy, as the result of growth arrest, and scoliosis.[1,7–9,11,12,15–17] The benchmark of orthopaedic management in congenital insensitivity to pain focuses on preventing occurrence of musculoskeletal complications in patients who are otherwise physically fully active, through proper education aiming to increase awareness in the patients and their family environment of the possible risks related to the absence of pain constraints and the increased incidence of self-abusive behavior. Fortunately, with further maturation, the children learn to self-protect and develop a better understanding of their limitations. Therefore, these incidents tend to decrease in frequency and severity as the patients become more cautious.

A meticulous search of the literature identified only two patients with congenital insensitivity to pain that presented with spinal complications. Piazza et al[16] reported on a 28-year-old woman who developed neuropathic arthropathy in the lumbar spine, with resultant progressive spinal instability at the L1–L2 level with consequent segmental kyphosis and intact neurologic function. She was treated with a staged anterior–posterior spinal fusion; however, further neurotrophic changes appeared over time in the adjacent segments both distal and proximal to the area of the arthrodesis. Heggeness[14] presented the case of a 17-year-old pain-insensate girl who developed severe Charcot arthropathy at L3–L4 level, which deteriorated during pregnancy and caused considerable canal narrowing, resulting in neurologic symptoms from the lower extremities. The patient underwent a two-stage anteroposterior instrumented spinal fusion; she subsequently recovered full and symmetric motor power.

It is well known that individuals with decreased or absent pain sensation of any etiology are particularly predisposed to developing Charcot joints.[9,15,18,19] Petrie et al[20] treated a patient with paraplegia and consequent neuropathic arthropathy of the spine, associated with bowel and bladder incontinence, operatively resulting in partial neurologic resolution. Circumferential stabilization has been proposed by several authors to produce better results in the management of patients with Charcot arthropathy involving the spine.[20,21]

Our patient had a long history of self-inflicted, painless injuries with multiple fractures occurring since early childhood. When he presented initially with the destructive lesion at the L1–L2 level and no previous traumatic event, a CT-guided biopsy was deemed to be necessary with the aim to establish the diagnosis. It is worthy of note that in patients with this

condition, the differential diagnosis specifically between fractures and infections can be challenging, owing to the fact that they both present at an advanced stage with similar clinical and radiographic appearance, while body temperature as well as infectious markers such as erythrocyte sedimentation rate and C-reactive protein can be elevated secondary to concomitant infections at other sites.[11] Moreover, the diagnosis between a fracture and an arthropathic lesion can be dubious and will rely primarily on the presence or absence of a history of spinal injury and the different characteristics of the two pathologic entities on the imaging evaluation. A high index of awareness of the increased incidence of neurotrophic degeneration in this group of individuals should be able to guide the diagnostic process.

In this patient, combined anteroposterior approach and circumferential spinal arthrodesis were performed in the initial presentation, to better restore the developing segmental kyphotic deformity, prevent additional neurologic compromise, and achieve a solid fusion. We believe that a 360° spinal fusion is more adequate in patients with absent protective reflexes. The tingling sensation in the patient's lower extremities resolved after the surgery.

Secondary levels of arthropathy developed at a later age, below the distal instrumented vertebral body. Since the preoperative imaging assessment revealed the presence of neurotrophic destructive lesions at L4 and L5 levels and with the potential risk of further evolving deformity if the lumbosacral articulation were left unfused, it was decided to extend the arthrodesis distally to the sacrum with pelvic fixation. In addition, extending the fusion distally to include the lumbosacral joint achieved good restoration of lumbar sagittal alignment, which we believe accommodated for the recuperation of the neurologic function. Even though the patient did not undergo anterior decompression, the neurogenic intermittent claudication symptoms recovered and symmetrical, normal muscle power was restored in his lower limbs. Postoperative bracing was used to provide additional support and limit the patient's physical activities in the absence of pain constraints, until good healing of the spinal arthrodesis would be accomplished, especially since an anterior procedure was not performed. One year after surgery, the patient had resumed his previous level of activities.

Congenital insensitivity to pain is a relatively uncommon condition. Nevertheless, its wide range and long-term clinical implications should be well appreciated and call for an early diagnosis. Following the diagnosis, the key to minimizing skeletal deformity and avoiding permanent disability should concentrate on prevention. This unusual case serves to illustrate the complexity of spinal manifestations in patients with congenital insensitivity to pain, which pose a significant diagnostic and therapeutic challenge. Appropriate aggressive surgical treatment is often indicated to restore function and avoid unnecessary morbidity.

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Minimizing Complications With Single Submuscular Growing Rods

A Review of Technique and Results on 88 Patients With Minimum Two-Year Follow-up

Najma Farooq, FRCS(Tr & Orth),*† Enrique Garrido, EBOT, MRCS,‡ Farhaan Altaf, MRCS,*† Joanne Dartnell, MRCS,*† Suken A. Shah, MD,§ Stewart K. Tucker, FRCS(Orth),*† and Hilali Noordeen, MA, FRCS*†

Study Design. Retrospective clinical and radiologic review of consecutive series of patients treated with single submuscular growing rods from a single center with a minimum of 2-year follow-up.

Objectives. To describe the surgical technique and methods used to minimize complications and to report on the outcomes of a large consecutive series of patients treated with single submuscular growing rods for scoliosis in the immature spine from a single center.

Summary of Background Data. Previous studies have reported on the safety and efficacy of single and dual growing-rod constructs; however, these studies have been of small patient numbers with varying results.

Methods. Between 1999 and 2007, 88 patients underwent the insertion of a single, submuscular growing-rod construct for scoliosis. A clinical and radiologic review of these 88 consecutive patients with a minimum of 2-year follow-up was conducted. Diagnoses include idiopathic, neuromuscular, syndromic, and congenital. Data include Cobb angle measurements, T1–S1 heights, number, and frequency of lengthening as well as complications.

Results. The patients underwent single submuscular growing-rod insertion at an average age of 7.0 years. The mean follow-up period was 42 months. Twenty-eight patients had a simultaneous apical fusion. Growing-rod lengthening was performed on an average at 9-month intervals. The average initial Cobb angle was 73° (range: 40–117) and improved to 44° (range: 9–90) at final follow-up. T1–S1 height gain was 3.37 cm; this translates to 1.04 cm growth/yr. No significant difference was noted between those who had undergone apical fusion and those without. Complications noted in this series include 8 incidences of superficial infection and 3 of deep infection, proximal junctional kyphosis in 2 patients requiring early fusion, 31 rod fractures, 10 cases of proximal anchor failure, and 6 distal anchor failures. Thirty patients within study group have reached definitive fusion.

Conclusion. Favorable outcomes have been demonstrated in this large single-center series of growing-rod constructs used to treat scoliosis in the growing spine. Their safety and efficacy in controlling spinal deformity and allowing spinal growth along with an acceptable rate of complications would support the continued use of single growing-rod constructs as a scoliosis management option.

Key words: growing rod, immature spine, scoliosis, instrumentation, complications, outcomes. **Spine 2010; 35:2252–2258**

Scoliosis in the immature spine poses a number of challenges to the treating clinician. Management regimes must address the need to preserve pulmonary function, maintain spinal and truncal growth, and control spinal alignment. Conservative treatments in the form of orthoses and casting techniques have a valuable role to play. However, these options are not always effective or appropriate and surgical intervention must then be considered.

Growing-rod constructs have been used to correct the spinal deformity while allowing continued spinal growth and hence pulmonary development. Over the years, these constructs have taken a number of forms. Historically, Harrington rods have been used for this purpose.[1] The development and use of the Luque trolley in the growing spine has also been well described.[2] However, more recently single and dual growing-rod implants have become the mainstay of treatment in these patients.[3,4]

Studies pertaining to the safety and efficacy of growth rod variants for treatment of early onset scoliosis are well reported in the literature.[5,6] Publications have examined the effects of simultaneous apical fusion on the final result,[3] the optimum frequency of growth rod lengthening,[7] and the stability of different foundation anchor configurations.[8] Reported complication rates vary between studies, but in general, growing rod treatment has been associated with high complication rates.[5,6]

Multicenter studies have compared the outcomes of single- and dual-rod constructs.[9] The proponents of dual-rod constructs report better correction of deformity increased stability and a greater percentage of expected spinal growth compared with single rods.[9] However, these studies have involved a limited number of patients and different types of constructs. The purpose of this

From the *Department of Spinal Surgery, Great Ormond Street Hospital, London, United Kingdom; †Department of Spinal Surgery, Royal National Orthopaedic Hospital, London, United Kingdom; ‡Department of Spinal Surgery, Royal Hospital for Sick Children, Edinburgh, United Kingdom; and §Department of Orthopedics, Nemours/Alfred I. duPont Hospital for Children, Wilmington, DE.

The device(s)/drug(s) is/are FDA-approved or approved by corresponding national agency for this indication.

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Address correspondence and reprint requests to Najma Farooq, FRCS(Tr &Orth), 76 Dolphin Rd, Slough, Berkshire SL1 1TA, United Kingdom; E-mail: najmafarooq@doctors.net.uk

study was to describe the surgical technique of the single, submuscular growing-rod implant and to report on the clinical and radiologic outcomes of a large consecutive series of patients managed with such implants from a single center. The article will also outline methods used to minimize the complications commonly associated with this form of treatment.

■ Surgical Technique

The key points involved in the insertion of a single submuscular growing rod are as follows: insertion of anchors, preparation of a submuscular bed, contouring and insertion of proximal and distal rods connected with a side-to-side domino, and the initial distraction.

A single, longitudinal midline skin incision is used. The sites for the proximal and distal anchors are identified, and subperiosteal dissection of the spine is limited to these areas. Our preferred anchor configuration for the distal rod is 2 pedicle screws. For the proximal anchors, we most frequently use a "claw" configuration comprised of 2 pedicle hooks plus either a supralaminar or transverse process hook. Two pedicle screws and a hook are used less commonly. A limited fusion is performed at these foundations with decortication. A submuscular bed is prepared without exposure of the spine through an incision in fascia within the concavity of the deformity. A proximal and distal rod is contoured and inserted. The central overlap of the rods is bridged by a side-to-side domino. Each rod can be lengthened independently with distraction against the fusion points (Figure 1).

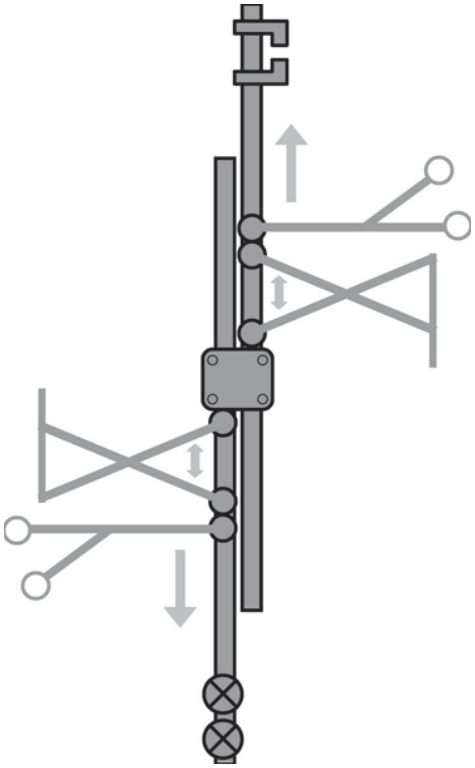

Figure 1. Shows a schematic representation of the single growing-rod construct. A side-to-side domino cross-connector is independently fixed to each rod at 2 consecutive points. This allows independent distraction of each rod against the level of anchor fixation. The placement of the rod holder and distracter is depicted here, enabling separate distraction of each rod during the lengthening procedure.

A Thoracolumbar Sacral Orthosis Is Used After Surgery

Growth rod lengthening was scheduled for 6 month intervals but varied between 6 and 9 months in practice. A small incision is made over the domino and adjacent rods. The domino locking nuts are loosened and a distraction force is used to lengthen the growing rod after which the locking nuts of the domino are retightened. The procedure involves a short overnight hospital stay.

In those patients also undergoing a combined apical fusion, the anterior procedure is undertaken first. A conventional transpleural thoracotomy approach is used. Access is gained to the apical segments on the convexity of the curve. A formal discectomy and anterior fusion is performed over these levels. At the end of the procedure, the pleura is closed, a chest drain is inserted, and the thoracotomy wound is sutured. The child is then repositioned prone, and the insertion of the growing rod is carried out as described previously.

■ Materials and Methods

Between 1999 and 2007, 88 consecutive patients were treated with a single, submuscular growing-rod construct for scoliosis and were followed up for a minimum of 2 years. Surgery was undertaken by the senior authors (M.H.H.N., S.K.T.) at 1 of 2 sites, the Royal National Orthopedic Hospital, Stanmore and Great Ormond St Hospital, London. The surgical technique has been described previously. Indication for surgery was a progressive spinal deformity, which was nonresponsive or not amenable to conservative management.

A retrospective review of the medical notes and radiographs was performed on patients enrolled into the growing-rod program.

Clinical data were collected to include patient details such as age at operation and diagnosis. Surgical data documented date of surgery, instrumented levels, types of anchors, whether an anterior apical fusion was performed, and the dates of distractions. Details of any complications including deep or superficial infections, rod fractures, anchor failures, junctional kyphosis, and deformity progression were recorded. The timing of definitive fusion was also noted.

Radiologic review was undertaken on radiographs taken just before the insertion of the growth rod, immediately postinsertion, and at latest follow-up. Cobb angle measurements of scoliosis were performed on each of these radiograph subsets. T1–S1 lengths were measured from radiographs taken immediately after insertion of the growing rod and from radiographs at latest follow-up. For those patients who had already undergone definitive fusion, the radiograph preceding the final fusion was analyzed to obtain the latest follow-up dataset (Figure 2).

■ Results

Eighty-eight patients with an average age of 7.0 years (1.4–12.1) underwent single, submuscular growing rod

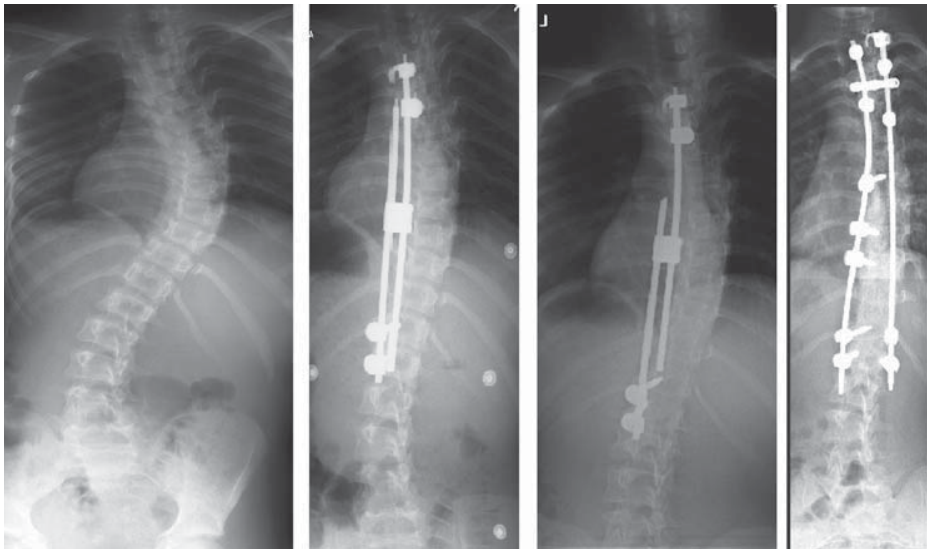

Figure 2. Case example. Shown below (left to right) are patient's preoperative radiographs, postoperative radiographs following growth rod insertion, those immediately preceding final fusion and radiographs after definitive fusion. T1–S1 gain was obtained by comparing T1–S1 height measurements from the immediate postoperative radiographs and those preceding definitive fusion (or latest follow-up if definitive fusion had not been performed).

insertion. The mean follow-up was 3.5 years (2–6.8). During this period, patients had on average 4.7 (0–12) growth rod lengthenings. Thirty patients reached definitive fusion within the study period.

The mean preoperative Cobb angle was 73.3° ± 15.6° (40.2–117); improving to 41.5° ± 14.6° (4.1–74) following insertion of the growth rod and progressing to 44.0° ± 15.2° (9.3–90) at the latest follow-up. T1–S1 lengths showed an average gain of 3.37 ± 2.53 cm (−1.9 to 8.76) at latest follow-up in this group of patients, translating to 1.04 ± 0.84 cm (−1.03 to 3.82) growth/yr.

Patients were categorized into the following diagnoses: idiopathic (28), congenital (18), syndromic (22), and neuromuscular (20). Twenty-eight of these patients also underwent a simultaneous anterior apical fusion.

A subgroup analysis was performed on the data according to the diagnostic category (Table 1). Comparison of the 4 groups found no significant differences between the age at insertion of growth rod, preoperative Cobb angle, and the average interval between growth rod lengthening (*P*: 0.83, 0.76, and 0.06, respectively).

A further comparison was carried out between those patients who had undergone a simultaneous apical fusion and those who had not (Table 2). The percentage breakdown of the diagnostic categories in each of these 2 groups is illustrated in Figure 3. This shows similar proportions of patients in the congenital and syndromic categories in both groups. However, there is preponderance of idiopathic patients in the apical fusion group and of neuromuscular patients in the nonapical fusion group. There were no significant differences between the 2 groups when comparing the age at insertion of growth rod, preoperative Cobb angles, percentage Cobb angle correction, and interval of growth rod lengthening. The growth per year was 1.06 cm in the apical fusion and 1.04 cm in the nonfusion group; again the differences were not significant.

Complications included 3 deep infections, necessitating removal of implants in 2 patients. Superficial infections requiring wound debridement occurred in 8 patients. The most common site for wound problems was in the vicinity of the domino.

Implant-related complications included 10 instances of proximal anchor failure and 6 of distal anchor failure requiring revision at the time of rod lengthening (Figure 4). A total of 31 rod fractures were noted. The caudal rod was found to be more frequently affected and the most common location to be adjacent to the distal an-

Table 1. Subgroup Analysis of Data According to Diagnostic Criteria

	All Diagnoses (n = 88)	Idiopathic (n = 28)	Syndromic (n = 22)	Neuromuscular (n = 20)	Congenital (n = 18)
Age at growth rod (GR) insertion (yr)	7.04 ± 2.62 (1.4–12.1)	7.42 ± 3.16 (1.4–12.1)	6.74 ± 2.26 (3.7–10.9)	7.02 ± 2.57 (3.2–11.3)	6.81 ± 2.26 (3.2–10.8)
Pre-GR Cobb	73.3 ± 15.6 (40.2–117)	73.0 ± 16.2 (43.2–112)	75.9 ± 17.2 (53–104.6)	74.5 ± 14.1 (58.7–117)	70.1 ± 15.9 (40.2–99.2)
Post-GR Cobb	41.5 ± 14.6 (4.1–74.4)	39.1 ± 10.8 (17.4–62)	47.2 ± 19.8 (4.1–71)	38.2 ± 13.8 (6–74.4)	43.9 ± 14.2 (24–70.9)
Final follow-up Cobb	44.0 ± 15.2 (9.3–90)	40.5 ± 15.4 (9.3–82)	50.6 ± 15.5 (28.2–90)	41.1 ± 12.8 (18.3–63.5)	46.6 ± 16.1 (15–70.7)
Cobb correction	29.3 ± 17.6 (−27–71.6)	32.5 ± 21.6 (−27–71.6)	25.3 ± 13.9 (5.8–63.8)	33.4 ± 15.7 (7.73–62.0)	23.5 ± 15.1 (−9–48.1)
% Cobb correction	38.7 ± 22.0 (−49–83)	41.9 ± 28.2 (−49–83)	32.9 ± 14.2 (11–61)	43.9 ± 17.6 (12–77)	33.4 ± 21.3 (−15–63)
T1–S1 gain (cm)	3.37 ± 2.53 (−1.9–8.76)	4.00 ± 2.37 (0.9–8.76)	3.64 ± 2.02 (0.22–6.7)	2.32 ± 2.68 (−1.9–8.7)	3.42 ± 2.76 (0.1–8.4)
Growth per year (cm)	1.04 ± 0.85 (−1.03–3.82)	1.20 ± 0.67 (0.19–2.68)	1.36 ± 1.07 (0.11–3.82)	0.77 ± 0.93 (−1.03–2.49)	0.84 ± 0.59 (0.03–1.89)
GR lengthening interval (mo)	9.75 ± 4.07 (3.45–23.51)	8.4 ± 2.59 (3.81–13.92)	9.73 ± 4.20 (3.45–16.42)	11.65 ± 5.21 (6.19–23.51)	9.32 ± 3.37 (5.86–19.95)

Table 2. Data Analysis and Comparison of Patients Who Underwent Simultaneous Apical Fusion With Those That Did Not

	All Diagnoses (n = 88)	Apical Fusion (n = 28)	No Fusion (n = 54)	*P*
Age at growth rod (GR) insertion (yr)	7.04 ± 2.62 (1.4–12.1)	7.11 ± 3.03 (2.1–12.0)	6.99 ± 2.47 (1.4–12.1)	0.85
Pre-GR Cobb angle	73.34 ± 15.64 (40.2–117)	74.66 ± 14.86 (43.2–104)	72.58 ± 16.17 (40.2–117)	0.59
Post-GR Cobb angle	41.54 ± 14.59 (4.1–74.4)	43.23 ± 13.21 (19.7–70.9)	40.56 ± 15.38 (4.1–74.4)	
Final follow-up Cobb angle	44.01 ± 15.2 (9.3–90)	44.30 ± 13.91 (18.7–90)	43.84 ± 16.04 (9.3–82)	
Cobb angle correction	29.3 ± 17.6 (−27–71.6)	30.36 ± 15.28 (2.3–56.3)	28.74 ± 18.93 (−27–71.6)	
% Cobb correction	38.7 ± 22.02 (−49–83)	39.44 ± 17.88 (5–75)	38.37 ± 24.25 (−49–83)	0.84
T1–S1 gain (cm)	3.37 ± 2.53 (−1.9–8.76)	4.19 ± 2.15 (0.06–8.4)	2.91 ± 2.62 (−1.9–8.76)	
Growth per year (cm)	1.04 ± 0.85 (−1.03–3.82)	1.06 ± 0.69 (0.03–3.0)	1.04 ± 0.94 (−1.03–3.82)	0.92
GR lengthening interval (mo)	9.75 ± 4.07 (3.45–23.51)	10.01 ± 3.82 (3.81–21.21)	9.6 ± 4.23 (3.45–23.51)	0.68

chors (Figure 5). The incidence of fracture was higher in the apical fusion group (42%) compared with the nonfusion group (35%), this effect may be the result of a stiffer spine placing more stress on the rod and causing subsequent failure.

Two patients developed a proximal junctional kyphosis that necessitated early fusion. In one of them, the proximal thoracic kyphosis only was fused and connected to a growing construct distally to allow continued spinal growth (Table 3).

■ Discussion

There is a varied opinion regarding the advantages and disadvantages conferred by single growing rods for the treatment of spinal deformity in the immature spine. Previous publications report that dual-rod constructs allow better control of the spinal deformity as well as achieving a greater percentage of expected spinal growth. Thompson *et al*[9] compared the outcomes in 3 patient groups; those treated with single growing-rod constructs with and without simultaneous apical fusion (n = 5 and 16 respectively) and those treated with dual-rod constructs (n = 7). Spinal growth was adequate in the single-rod group without apical fusion (0.53 cm/yr), better in the dual-rod group (1.1 cm/yr), and suboptimal in the single-rod group with simultaneous apical fusion (−0.2 cm/yr). The mean interval between lengthenings in this study did vary between the 3 groups. The patients in the dual-rod group were lengthened consistently at 6 month intervals, the patients in the single-rod groups were lengthened at 12 months (with apical fusion) and 9 months (without apical fusion). This disparity in the frequency of lengthening may account for the difference in growth rates seen between groups. The improved growth rates may not be entirely a result of the dual-rod configuration. This explanation becomes more plausible if we consider the findings of the study by Akbarnia *et al*,[7] 13 patients treated with dual growing-rod constructs were included. The T1–S1 growth per year was analyzed. Those undergoing lengthening at >6 monthly intervals (n = 6) achieved significantly less T1–S1 growth than those lengthened at <6 monthly intervals, 1.02 *versus* 1.84 cm/yr respectively. In our study, similar overall rates of growth (1.04 cm/yr) were achieved with single-rod constructs undergoing lengthening on average at 9 monthly intervals. The patients who underwent lengthening at ≤6 monthly intervals formed a relatively small subset of the entire group (n = 13) and when analyzed separately, the T1–S1 growth/yr was found to be 1.69 ± 1.16 cm/yr, which was significantly greater ($P = 0.002$) when compared with the remaining patients who underwent

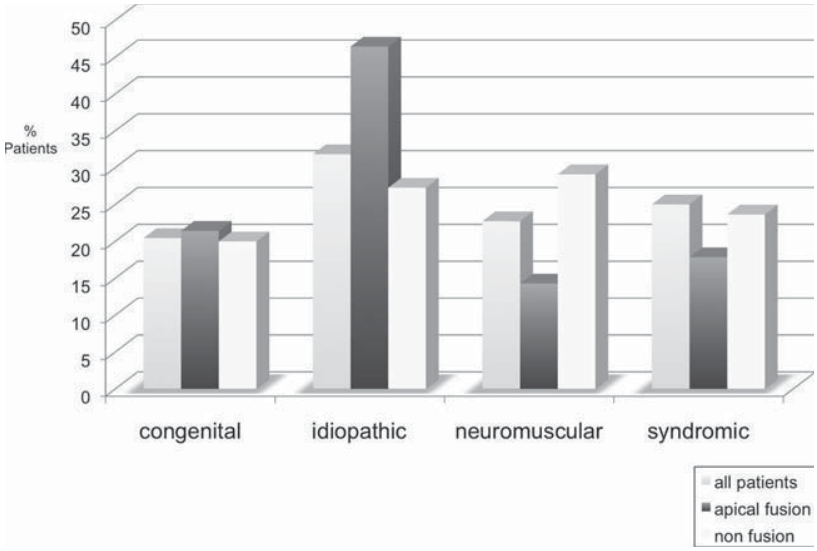

Figure 3. Comparison of simultaneous apical fusion and nonfusion groups and the percentage of patients within each diagnostic category.

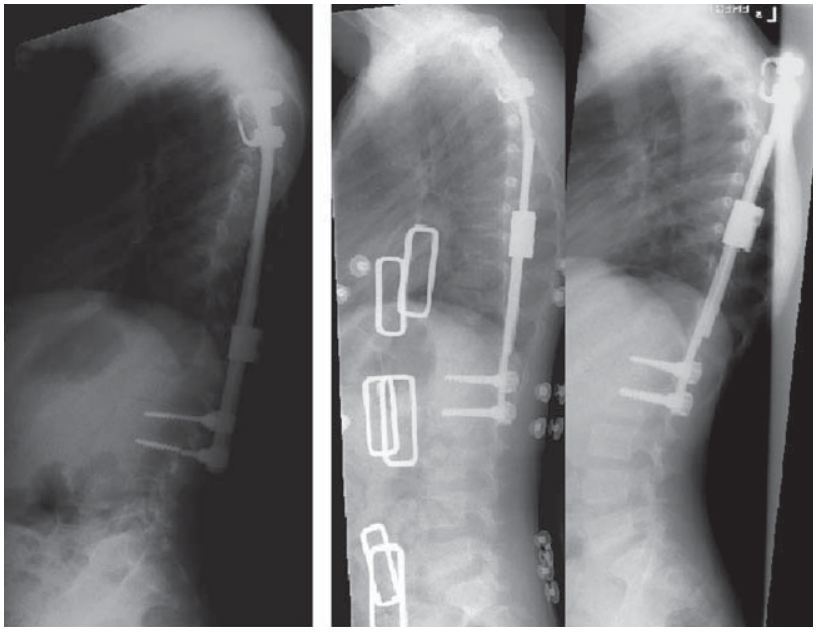

Figure 4. Examples of anchor failure. Pedicle screw breakage in distal anchor (left). Hook dislodgement of proximal anchors (right).

lengthening less frequently (>6 months) and subsequently achieved growth of 0.91 ± 0.71 cm/yr.

The effects of a simultaneous apical fusion have also been discussed in the literature. The use of an anterior convex hemiepiphysiodesis in conjunction with the Luque trolley construct has been reported to have a beneficial effect on outcome.[2] Studies examining the effects of apical fusion with growing rods have been less encouraging. Blakemore *et al*[3] published preliminary results on a series of 29 patients undergoing growing-rod procedures: 11 patients underwent a simultaneous apical fusion whereas the remainder did not. Their results were not able to demonstrate any significant improvement in deformity correction with the addition of the fusion. The publication by Thompson *et al*,[9] comparing 3 groups of patients with growing-rod constructs as we have discussed, reported significantly lower T1–S1 growth rates in those patients treated with an apical fusion. Furthermore, the correction of the spinal deformity was not maintained in this group.

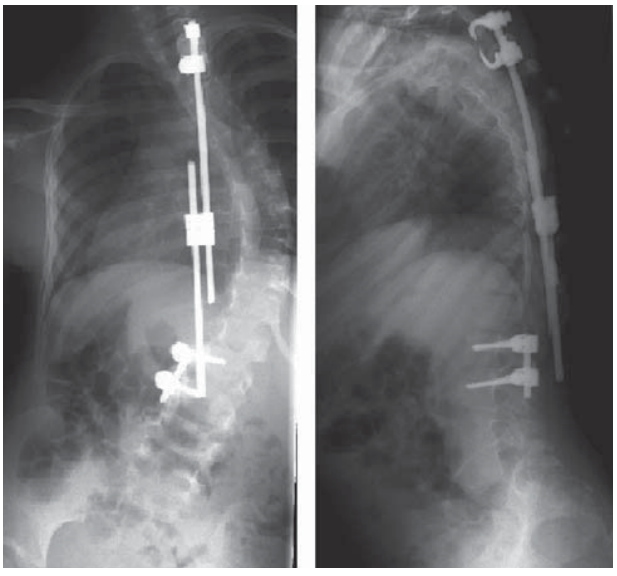

Figure 5. Example of rod fracture at commonest location.

In our study, 28 patients underwent a simultaneous apical fusion. During a timeframe within the study period, it was our standard practice to perform an apical fusion on all patients undergoing a growing-rod procedure. Therefore, the patient characteristics of this group are similar to the overall study population; however, as suggested before, there appear to be a greater proportion of idiopathic patients in the apical fusion group and overall greater proportion of neuromuscular patients in the nonfusion group (Figure 3). However, there are no significant differences in the age at insertion and the initial curve magnitude between the 2 groups (Table 2). Comparing the outcome in terms of curve correction and spinal growth again found no significant differences between these 2 sets of patients. Hence, in studies to date performing an apical fusion in patients treated with growing-rod constructs appear not to confer any additional advantage.

If we examine overall curve correction in our series of patients, the results achieved are comparable to and in some cases favorable to previously published series. Blakemore *et al*[3] reported on their series of 29 single-rod patients and noted Cobb angle improvements from 66° to 38° following insertion of the single growth rod. The curve then progressed to 47° at follow-up. The data from our series report a larger initial curve magnitude but similar end results. Results from Thompson *et al*[9] found the dual-rod group to fare far better with a greater percentage of curve correction compared with the single-rod groups. However, again there remains the confounding factor of increased frequency of lengthenings which Akbarnia *et al*[7] found to also improve curve correction.

Table 3. Shows Number and Types of Complications and the Distribution Across the Four Diagnostic Groups

	All Diagnoses (n = 88)	Idiopathic (n = 28)	Syndromic (n = 22)	Neuromuscular (n = 20)	Congenital (n = 18)
Infection—deep	3	0	2	1	0
Infection—superficial	8	2	2	1	3
Anchor failure—proximal	10	4	2	2	2
Anchor failure—distal	6	2	2	2	0
Rod fracture	31	9	9	9	4
Proximal junctional kyphosis	2	0	2	0	0

Growing-rod constructs are well renowned for their high rate of complications.[5,6] Implant failure, wound-related complications, and even death have been described in previous publications. Wound complications such as superficial and deep infection form a consistent feature of reports on growing rods. Mineiro and Weinstein's[5] series of 11 patients documented 1 superficial and 1 deep infection; Blakemore's series of 29 patients had 1 superficial infection; and that of Thompson *et al*, 2 superficial infections in 28 patients. In the study of Akbarnia *et al,* the series of dual-rod constructs found 4 superficial and 2 deep infections in a review of 23 patients.[4] Data presented from a multicenter review of complications in growing-rod surgery by Bess *et al*[10] reported 9 wound complications in a group of 73 single-rod patients and 21 in a group of 70 dual-rod patients. Our series found lower wound complication rates than those published for dual-rod constructs. The side-to-side domino and rod configuration of the growing-rod construct used in our study is of a comparatively lower profile than most dual-rod constructs; the additional submuscular placement of the implant further aids in reducing soft tissue complications and wound infections.

Optimum anchor configuration and prevention of anchor failure have been the subject of much debate. Early publications of growing-rod constructs report much higher rates of anchor pullout. The review by Klemme *et al*[6] found 21 incidences of anchor failure in 67 patients, 16 involved the proximal anchor. Subsequent studies have shown lower rates.[3,4,9] Thompson *et al* report no instances of anchor failure within the dual-rod group of 7 patients from their comparison of dual- and single-rod constructs, whereas they found 6 within the single-rod subset of 21 patients. Akbarnia's study of 23 patients with dual rods and only 2 anchor failures further supports the argument that dual-rod constructs might provide added stability to the anchors compared to single-rod cases, however, patient numbers remain small. Biomechanical experiments on the stability of anchor configurations found pedicle screw constructs or hook constructs with a cross connector to confer greater stability.[8] Our study of 88 patients produced anchor failure rates (10 proximal and 6 distal) higher than those reported in dual-rod series, however, they remain favorable in comparison with previous publications of single-rod constructs. In our study, the proximal anchors consisted almost invariably of hooks fashioned in a claw construct providing the necessary fixation without the risk of neurologic injury from anchor migration or plowing as can occur with pedicle screws. To reduce the risk of anchor failure, we performed a formal local fusion around the anchor bed to add further stability. Salvage of failed proximal anchors was achieved by relocating the claw construct to a more proximal level in most cases. Failure of distal anchors was relatively less common; conversion to constructs consisting of supra- and infralaminar hooks was used for revision purposes.

Rod fractures are inevitable occurrences in nonfusion techniques. Yang *et al*[11] identified several risk factors that increased the propensity of rod breakage in an analysis of 322 patients from a multicenter database of growing rods. These were single-rod constructs, rods of smaller diameter, stainless steel rods, and ambulatory patients. In contrast, Thompson *et al* showed higher rates of rod fracture in the dual-rod group despite the perceived greater stability provided by 2 rods *versus* 1. Thirty-one rod fractures were noted in our series despite the use of postoperative bracing in ambulatory patients. A higher incidence of rod fractures occurred in those patients treated with an apical fusion. The differential in stiffness of the spine might transfer further stress between the rods and fused regions making them more prone to breakage. It is no longer our practice to routinely perform a simultaneous apical fusion; this may lessen the frequency of rod fractures within our growing rod program. Modification of the material properties of the single-rod construct components may also reduce the number of rod fractures in future cases.

Our results of a consecutive, single-center series of 88 patients with an average of 3.5 years follow-up show that single, submuscular growing-rod constructs were adequate in maintaining spinal growth (1.04 cm/yr) and correcting deformities in early onset scoliosis, with a complication rate more favorable than that reported previously using single-rod constructs.

■ Key Points

- Single growing-rod constructs are safe and effective in controlling spinal deformity.
- Single growing-rod constructs allow spinal growth that is comparable to previous reports and to those of dual-rod constructs.
- The rate of complications is acceptable within this patient series.

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Chapter 4

INNOVATION & RECENT ADVANCES

Advances in Surgical Techniques for EOSD

Paper: 5

Growing rods are popular in treating early-onset scoliosis of all etiologies. In my surgical practice, I used to insert single submuscular growing rods during late 1990s and early 2000s. I observed over time that an increasing amount of forces had to be applied during progressive six to nine monthly distraction surgeries.

I undertook a systematic study, and objectively quantified the amount of distractive forces applied to obtain serial one mm lengthening in a group of twenty six patients who had sixty distraction procedures. The etiologies included: congenital (9), neuromuscular (9), idiopathic (4), syndromic (3) and neurofibromatosis (1). A transducer was attached to distraction calipers that recorded the force applied during distractions and at zero load status. The total length gained with each distraction procedure was recorded in mm. The applied distractive force at the first lengthening was 142N which increased to 165N by the third and 368N by the fifth lengthening. 608N force had to be applied on the tenth lengthening episode. Patients, who had apical fusion during index insertion of growing rods, needed 40 percent more force to achieve similar lengthening as compared to those who weren't fused around the apex. The forces applied doubled at the fifth lengthening (368N±54N) and a statistically significant force had to be applied at the fifth lengthening in comparison to the fourth ($p<0.01$). The average length gained with each distraction decreased over time (i.e. 'the law of diminishing returns') and by the fifth distraction onwards, the length gain was consistently <8mm. This was the first study to measure the amount of in-vivo distractive forces applied during growing rod surgeries.

In vivo distraction force and length measurements of growing rods: which factors influence the ability to lengthen?: Noordeen HM, Shah SA, Elsebaie HB, Garrido E, Farooq N, Al Mukhtar M. Spine (Phila Pa 1976). 2011 Dec 15;36(26):2299-303. PMID: 21494191

Advances in Surgical Techniques for EOSD

Paper: 6

A novel and innovative remote controlled growing rod system with a built-in magnet driven motor to induce distraction or lengthening on an outpatient basis was devised to minimize / eliminate the complications seen with use of conventional growing rods (CGRs). The magnet driven growing rods (MdGR) were successfully tested for safety in animals prior to human use. Also feasibility to perform MRI scans was also investigated.

I was a part of multi-centric prospective observational study group, reporting preliminary results of MdGR in a group of fourteen patients with progressive EOS secondary to diverse etiologies (seven males and seven females). The mean age at insertion was eight years and ten months (range: three years and six months to twelve years and seven months). The etiologies included idiopathic (5), neuromuscular (4), congenital (2), syndromic (2), and type 1 neurofibromatosis (1). MdGR were found to be safe and effective in controlling curve progression and probably cost-effective in the long-term. The MAGEC^{TM} rods (Ellipse Technology, Irvine, CA) were used in all fourteen patients, and sixty eight spinal distractions with an average of 4.9 distractions per patient were carried out successfully. Five patients had single-rod (SR), and nine had dual-rod (DR) constructs. The average follow-up was ten months. The mean time between index surgery and initiation of the first distraction was sixty six days. The mean interval between subsequent distractions after the first distraction was forty three days. The study found MdGRs to be a viable alternative to the CGRs for treatment of progressive EOS. There weren't any major implant-related complications.

Next generation of growth-sparing techniques: preliminary clinical results of a magnetically controlled growing rod in 14 patients with early-onset scoliosis. Akbarnia BA, Cheung K, Noordeen H, Elsebaie H, Yazici M, Dannawi Z, Kabirian N. Spine (Phila Pa 1976). 2013 Apr 15;38(8):665-70. PMID:23060057.

Advances in Surgical Techniques for EOSD

Paper: 7

I reported the largest series to date, with the use of MdGR in humans (Jan 2013). Between November 2009 and March 2011, Thirty four children with EOSD were prospectively recruited into the 'Growing rods program' at RNOHT, Stanmore. There were thirteen males and twenty one females. The mean follow-up was fifteen months (range: 12 to 18 months). The etiologies included idiopathic (11); neuromuscular (11); congenital (2); neurofibromatosis (3) and syndromic (4). A single-rod (SR) construct was used in twelve patients and twenty two had dual-rod (DR) constructs. Each patient had at least three out-patient distractions. The mean number of distractions per patient was 4.8 (range: 3 to 6). The mean time to the first distraction was sixty six days (range: 22 to 112 days). The mean interval between distractions was eighty seven days (range: 56 to 132 days). The mean pre-operative Cobb angle was sixty nine degrees (range: 46 to 108 degrees) which corrected to a mean 47 degrees (range: 28 to 91 degrees) post-operatively. The mean Cobb angle at final review was 41 degrees (range: 27 to 86 degrees). The mean pre-operative T1 to S1 length was 304 mm (range: 243 to 380) and increased to 335 mm (range: 253 to 400) in the immediate postoperative period. At final review the mean increase in T1 to S1 length was forty four mm. There were two implant failures and two wound infections (both superficial). There was loss of distraction owing to anchor dislodgement in two patients that warranted revision surgery. Proximal junctional kyphosis occurred in one patient warranting an extension of instrumentation. The preliminary results were promising and encouraging with fewer complications in comparison to historical series treated by using conventional growing rods.

Early results of a remotely-operated magnetic growth rod in early-onset scoliosis; Dannawi Z, Altaf F, Harshavardhana NS, Elsebaie H, Noordeen H, Bone Joint J 2013 Jan; 95-B(1): 75-80. PMID: 23307677

Advances in Surgical Techniques for EOSD

Paper: 8

The effect and impact on pulmonary function and vital capacity with use of MdGR is not known. I assessed pulmonary function in six patients with EOSD secondary to neuromuscular disorders (SMA II–2; Neurofibromatosis-2; William's Syndrome–1 and Congenital muscular dystrophy–1), studying their pre and post-op pulmonary function with a minimum follow-up of two years. The paper was presented at the SRS 2013 annual meeting as a free-paper and the manuscript was published in Spine (P'hila, 1976) this year.

There was a statistically significant improvement in FVC and FEV_1 / FVC ratio at both six months and at two years follow-up. The improvement was impressive for SMA type II. The mean pre-op FEV1 and FVC improved from twenty seven percent of predicted to forty one percent and forty five percent of predicted respectively at two years post-op. More importantly, all children had clinical benefit, with a reduced incidence of chest infections, unplanned admissions through the Accident and Emergency to pediatric or respiratory units for pulmonary issues. The caregiver satisfaction was immense, given that all patients continued to be healthy with adequate weight gains. Though MdGR may not alter the natural history of these neuromuscular disorders (i.e. progressive decline in pulmonary function and vital capacity with time), they markedly reduce the rate of decline beyond a doubt, and that gives the impression of immediate improvement in pulmonary function with surgical intervention.

Magnetic growth rods improve pulmonary function in early-onset scoliosis secondary to neuromuscular disorders. Yoon WW, Sedra F, Shah SA, Wallis C, Muntoni F, Noordeen MHH. Spine (Philadelphia, PA 1976) 2014; Jul 1, 39(15): 1196-1202. PMID: 24825149.

Spine

SPINE Volume 36, Number 26, pp 2299–2303

DEFORMITY

In Vivo Distraction Force and Length Measurements of Growing Rods

Which Factors Influence the Ability to Lengthen?

Hilali M. Noordeen, FRCS (Orth),* Suken A. Shah, MD,† Hazem B. Elsebaie, FRCS, MD,‡ Enrique Garrido, EBOT, MRCS,* Najma Farooq, FRCS (Tr & Orth),* and Mohannad Al Mukhtar, MRCS*

Study Design. Prospective, intraoperative force measurement in consecutive lengthening procedures in a series of growing-rod patients undergoing lengthening.

Objective. The purpose of this study was to measure the forces and amount of distraction over time in early onset scoliosis patients treated with growing rods.

Summary of Background Data. Growing rods are one of the current techniques used in the treatment of early onset scoliosis, and the goal of the growing-rod technique is to achieve deformity correction, maintaining spinal growth at the same time. Gradual stiffening or spontaneous fusion of the spine can interfere with the ability to lengthen. In addition, diminished acquired length with serial distraction are common observations and need to be evaluated and quantified.

Methods. Distraction forces were measured prospectively during 60 consecutive lengthening procedures in 26 patients. All patients had single submuscular rod constructs with side-to-side connectors. For each measurement, output from a transducer on a dedicated pair of distraction calipers was recorded at zero load status and the force was then recorded at every 1 mm lengthening; length was obtained at each event and was recorded in millimeters.

Results. The force required to distract the spine doubled at the 5th lengthening procedure (mean 368 N ± 54 N), and the distraction force was significantly higher at the fifth lengthening compared with the previous lengthening ($P < 0.01$). Mean length achieved at each distraction decreased over time such that by the fifth lengthening, consistently 8 mm or less was achieved.

Conclusion. Distraction forces increase significantly after repeated lengthening of growing-rod constructs, and the length obtained at each procedure exhibits a decreasing trend.

Key words: growing rods, force measurement, distraction, lengthening. **Spine 2011;36:2299–2303**

The treatment of children with progressive early onset scoliosis has proven to be difficult; standard methods such as orthosis or spinal fusion used in older patients are less effective in very young children with progressive curves. New surgical methods have evolved to allow near normal growth, at the same time, maintaining the correction achieved at initial surgery.[1] Growing rods are one of the current techniques used in the treatment of early onset scoliosis, and the goal of the growing-rod technique is to achieve deformity correction at the same time maintaining spinal growth; achieving this goal has led to some difficulties and complications and the best way to improve this technique is a better understanding of the details of the serial distraction process. In growing-rod technique, the rods are lengthened every 6 months regardless of curve progression.[2] The repetitive distractions under general anesthesia may increase the risk of instrumentation failures and infection; in an attempt to solve these problems, a spinal instrumentation system, consisting of a remote controlled growing-rod system with a built-in motor to induce distraction of the growing rods on an outpatient basis has been tried in animals.[3] The need to analyze the factors affecting the technique of serial distractions is important for any future modification of the system; these factors include the force needed to distract the rods apart, the length acquired in the system as a result of application of these forces, and the gradual stiffening of the spine that can interfere with the ability to lengthen the spine. The purpose of this study was to measure the forces applied and the amount of distraction over time in a consecutive series of early onset scoliosis patients treated with growing rods. To the best of our knowledge, this study elucidates and quantitates forces in modern spine correction that has only been evaluated once in the spine in the distant past.

METHODS

This is a prospective study analyzing the data recorded from 60 consecutive distractions done in 26 early onset scoliosis

From the *Great Ormond Street Hospital, London, United Kingdom; and Royal National Orthopedic Hospital, Stanmore, United Kingdom; †Department of Orthopedics, Nemours/Alfred I. duPont Hospital for Children, Wilmington, DE;·and ‡Cairo University Teaching Hospital, Cairo, Egypt.

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Address correspondence and reprint requests to Hilali Noordeen, Royal National Orthopedic Hospital, Stanmore, United Kingdom; E-mail: hnoordeen@aol.com.

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TABLE 1. Demographics

	Age Insertion (y)	Diagnosis	Preoperative Cobb Angle
1	3.16	Congenital	99.2
2	7.44	Neuromuscular	60.33
3	6.28	Neuromuscular	80.3
4	3.42	Idiopathic	54.8
5	3.17	Neuromuscular	75.71
6	5.61	Congenital	50.4
7	10.65	Neuromuscular	64.82
8	8.3	Congenital	72.3
9	4.45	Idiopathic	74.5
10	3.45	Neuromuscular	58.7
11	8.67	Idiopathic	70.3
12	10.36	Syndromic	61.4
13	9.42	Neuromuscular	68.6
14	3.18	Congenital	67.6
15	9.2	Congenital	60.4
16	4.53	NF	54.8
17	6.78	Neuromuscular	72.33
18	9.11	Neuromuscular	72.5
19	8.56	Congenital	40.2
20	5.88	Syndromic	104.6
21	7.04	Idiopathic	73.4
22	8	Congenital	63.3
23	4.35	Neuromuscular	83.6
24	6.11	Syndromic	90
25	4.47	Congenital	82.4
26	7.27	Congenital	81.6

NF indicates neurofibromatosis.

patients with an average age of 6.49 years (3.16–10.65) and an average Cobb angle of 70.23 (40–104) with different etiologies: four idiopathic, nine congenital, nine neuromuscular, and three syndromic (Table 1); during their treatment with serial distractions by growing rods. All patients had single submuscular rod constructs with side-to-side (domino) connectors and only three patients with severe stiff curves above 80° had anterior release and apical fusion. Distraction forces were measured with each millimeter of acquired length, and both values were recorded accordingly; the maximum force applied and its corresponding increase in length were recorded for each distraction.

Measurements were performed through a strain gauge applied to a custom-made pair of distraction callipers incorporating a load transducer that allowed measurement of load applied at the tip. Included in the design was a millimeter-scaled ruler to measure relative linear displacement between the jaws; using the ruler, the accuracy of the displacement measurement in this environment was considered ±0.25 mm.

For each measurement, output from a transducer on a dedicated pair of distraction calipers (Figure 1) was recorded at zero load status and the force was then recorded at every 1 mm lengthening:

1. Resting load prior to any distraction was recorded; the distraction was performed against a rod holder; a resting tension was deliberately put on to ensure that there was no loss of length already gained on previous lengthenings as each set screw was loosened and removed.
2. Loads were recorded with each 1-mm increase in length.
3. The maximum force resulting in distraction between the two rods at which no more length gain was seen was recorded as well.

The maximum increase in length obtained at each distraction was recorded in millimeters with its respective load applied; the measuring tool was checked after sterilization and before usage to confirm the precision of measurement. The data for the same number of distractions (first, second, …) were analyzed accordingly to determine the possible change in force applied and length acquired with the increased number of distractions.

RESULTS

Mean peak forces increased sequentially with each lengthening. The mean force to distract the spine during the first lengthening was 142, the 3rd was 165, the 5th was 368, the 7th was 390, and at the 10th was 608. The force required to distract the spine doubled at the 5th lengthening procedure (mean 368 N ± 54 N), and the distraction force was significantly higher at the 5th lengthening compared with the previous lengthening ($P < 0.01$); in addition, there

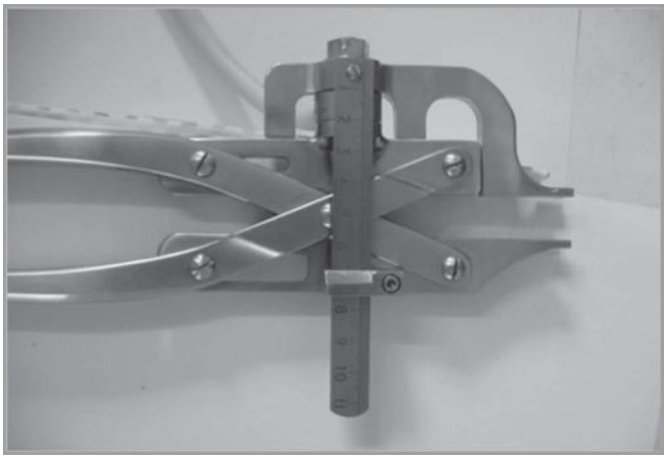

Figure 1. Intraoperative measurement device with custom-made pair of distraction calipers incorporating a load transducer allowing measurement of load applied at the tip. Included in the design was a millimeter-scaled ruler.

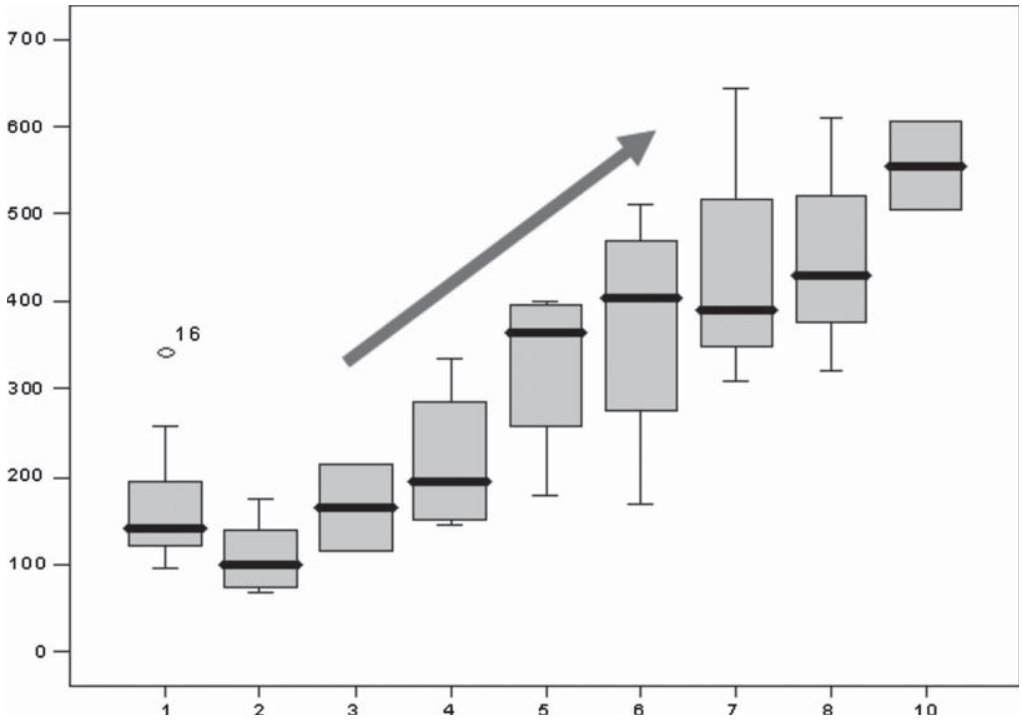

Figure 2. Graphical presentation of force *vs.* number of distraction.

is a trend for the force of distraction to increase with the increased number of distraction (Figure 2). Distraction forces were on average 40% higher in patients who had undergone an apical fusion at the time of primary implantation of the growing rod *versus* those who had no apical fusion ($P < 0.05$) (Figure 3).

The mean length acquired during the 1st distraction was 17.1 mm, the 3rd was 11.1, the 5th was 8.0, the 7th was 7.5, and the 10th was 6.1. Mean length achieved at each distraction decreased over time such that by the 5th lengthening, consistently 8 mm or less was achieved (Figure 4). The side-to-side connector between the cranial and caudal rods had a significant tethering effect; after lengthening the first rod, the resting pressure of the second rod was significantly lower than the recorded peak load of the first rod, as the connector had to be exposed of all the intervening soft tissue.

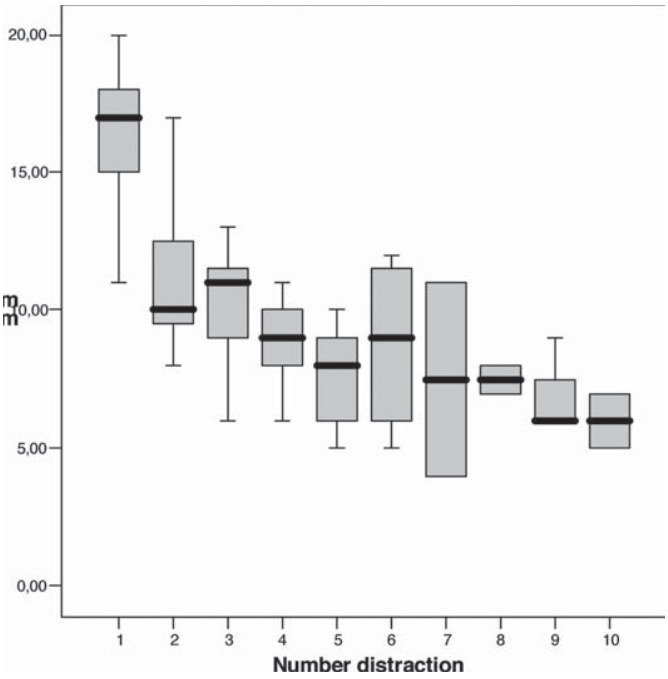

Figure 3. Graphical presentation of length *vs.* number of distraction.

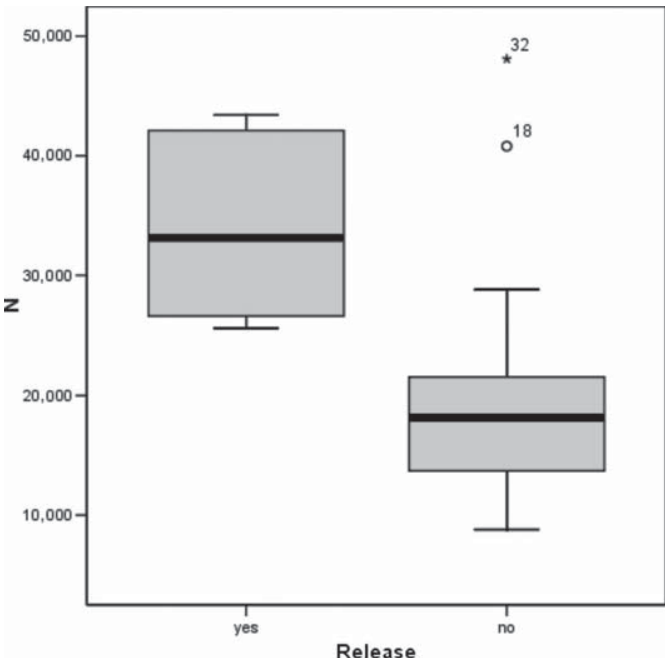

Figure 4. Graphical presentation of force measurement with and without apical fusion.

DISCUSSION

Expandable implants have been used for several decades in an attempt to control the spinal deformity and to allow for spinal growth until the child reaches an appropriate size or age for definitive fusion; these include a single growing rod with foundations proximally and/or distally,[4,5] dual growing rods with foundations proximally and distally,[5] and the vertical expandable prosthetic titanium rib implant.[6,7] The preliminary results indicate that either single rod or dual rods can be effective in controlling spinal deformity.[5] In a recently presented study from the same center, the results of a consecutive series of 88 patients, including the 26 patients of the current study, with an average of 3.5 years of follow-up showed that single submuscular growing-rod constructs with side-to-side connectors were adequate in maintaining spinal growth and correcting deformities in early onset scoliosis, with a complication rate comparable to previously reported double-rod constructs except in anchor failures, which were significantly less in double rods.[8]

It has been shown that with more frequent lengthening, the continued growth of the spine per year after the initial procedure equaled or surpassed the normal growth rate of the spine. The authors believed this is due to the effect of distraction on immature vertebral growth.[1] Other factors involved in the program of serial distraction are the length acquired during each distraction as well as the force applied to distract the rods; these factors become of great importance with the exploration of novel and less invasive treatments that achieve similar endpoints, at the same time reducing the number of surgical interventions necessary.

Force measurement has been an area of interest in many previous animal and human studies. *In vivo* human distraction forces have been measured in orthopedic surgery; in distraction epiphysiolysis, the initial force required to separate the physeal plate in humans was shown to be between 570 and 1500 N[9], and after separation the distraction force was found to be lower, between 100 and 200 N in the next 3 weeks.[10] In lower limb lengthening, the average peak values of force amounted to 300 N (maximum 737 N) in the femoral group and 305 N (maximum 592 N) in the tibial.[11]

In spinal surgery the distraction force generated by the Harrington rod is reported to be about 100 to 200 N.[12] A spinal instrumentation system, consisting of a remote-controlled growing-rod system with a built-in motor has been developed and was tried in animals; it can be used for repeated distraction of the spine and can be used to correct the deformity noninvasively after the instrumentation. When the controller is turned on, the torque generated by the motor is converted to distraction force by the inner gear, and therefore the growing rod stretches without rotating. The distraction force of the instrument was measured using a load cell, and the maximum distraction force of the instrument was 194 N.[3]

In the current human study, the distraction force used to move the two rods apart and elongate the system in the first four lengthening procedures averaged between 100 and 195 N; however, this force has doubled by the 5th distraction to 368 N and by the 10th distraction reached an average of 555 N. These high forces are recorded for the first time in humans intraoperatively, and these force measurements give an important primary understanding of the force needed to be generated to distract these rods remotely.

Distraction forces were on average 40% higher in patients who had undergone an apical fusion at the time of primary implantation of the growing rod *versus* those who had no apical fusion; we recommend limiting apical final fusion at index surgery to the absolutely indicated cases. It as it not only has the disadvantage of decreasing the number of vertebrae having the ability to grow with distraction, but it also increases the force needed to distract the rods with all its mechanical consequences. We also observed that using a side-to-side connecter, after lengthening the first rod, the resting pressure of the second rod was significantly lower than the recorded peak load of the first rod, as by the time we are distracting the first rod, the connector would have been exposed and freed from all intervening soft tissues with all their tethering effects.

Also there have been many observations by different investigators that the length acquired during serial distractions becomes less with time and with the increase of serial distractions. This is within the logical understanding of the growing-rod techniques being consistent with these curves becoming stiffer with the possibility of spontaneous fusion occurring at some levels. In this study, we have statistically confirmed these observations and we have quantified this decrease in millimeters for each distraction. We found the median length acquired during the 1st distraction was 17.1 mm, which became less than its half (8 mm) in the 5th distraction and reached only 6.1 mm by the 9th distraction.

The analysis of the results showing an increase of the force of distraction and a decrease in the acquired length with the increase in the number of distraction is another clear confirmation of the stiffness occurring in these curves with time and with serial distractions, and it has many clinical implications. It shows that the main increase in length acquired by serial distractions occurs during the first 3 to 4 years after the index surgery, and afterward there is a linear decrease in the length acquired, decreasing the benefit in continuing the distraction program. This shows the need to delay the index surgery as much as possible to get the maximum benefit of the use of growing rod. A second clinical implication is the increased stiffness of the curve with time, which can open the discussion for the need for final fusion surgery in acceptable curves reaching or approaching skeletal maturity. It also opens a new alternative of either leaving the growth rod *in situ* or removing the rods, leaving a stiff curve not amenable to progression.

CONCLUSIONS

Distraction forces increase significantly after repeated lengthening of growing-rod constructs, and the length obtained at each procedure exhibits a decreasing trend. The higher distraction forces measured on patients who had an apical fusion may explain the increased incidence of instrumentation failure reported in the literature. These findings help in understanding some of the limitations and complications of growing rods and help in the development of newer less invasive systems.

➢ Key Points

- ❑ There is a significant increase in distraction forces in growing-rod constructs with subsequent lengthening.
- ❑ Apical fusion increases the force needed to distract the rods significantly.
- ❑ The amount of length gained with each distraction decreases with time and with serial distractions.
- ❑ Instrumentation design must accommodate the forces generated and the trends observed in this study to minimize failure.

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DEFORMITY

Next Generation of Growth-Sparing Techniques

Preliminary Clinical Results of a Magnetically Controlled Growing Rod in 14 Patients With Early-Onset Scoliosis

Behrooz A. Akbarnia, MD,*† Kenneth Cheung, MD,‡ Hilali Noordeen, FRCS, (Orth),§ Hazem Elsebaie, MD, FRCS,¶ Muharrem Yazici, MD,‖ Zaher Dannawi, FRCS(Orth),§ and Nima Kabirian, MD*

Study Design. Prospective nonrandomized study.

Objective. To report the preliminary results of magnetically controlled growing rod (MCGR) technique in children with progressive early-onset scoliosis.

Summary of Background Data. The growing rod (GR) technique is a viable alternative for treatment of early-onset scoliosis. High complication rate is attributed to frequent surgical lengthening. The safety and efficacy of MCGR were recently reported in a porcine model.

Methods. Multicenter study of clinical and radiographical data of patients who underwent MCGR surgery and at least 3 distractions. Distractions were performed in clinic without anesthesia/analgesics. T1–T12 and T1–S1 heights and the distraction distance inside the actuator were measured after lengthening.

Results. Fourteen patients (7 girls, 7 boys) with a mean age of 8 years, 10 months (3 yr, 6 mo to 12 yr, 7 mo) had 14 index surgical procedures. Of the 14, 5 had single-rod (SR) surgery and 9 had dual-rod (DR) surgery, with overall 68 distractions. Diagnoses were idiopathic (N = 5), neuromuscular (N = 4), congenital (N = 2), syndromic (N = 2), and neurofibromatosis (N = 1). Mean follow-up was 10 months (5.8–18.2). The Cobb angle changed from 60° to 34° after initial surgery and 31° at latest follow-up. During distraction period, T1–T12 height increased by 7.6 mm for SR (1.09 mm/mo) and 12.12 mm for DR (1.97 mm/mo). T1–S1 height gain was 9.1 mm for SR (1.27 mm/mo) and 20.3 mm for DR (3.09 mm/mo). Complications included superficial infection in 1 SR, prominent implant in 1 DR, and minimal loss of initial distraction in 3 SR after index. Partial distraction loss observed after 14 of the 68 distractions (1 DR and 13 SR) but regained in subsequent distractions. There was no neurological deficit or implant failure.

Conclusion. Preliminary results indicated MCGR was safe and provided adequate distraction similar to standard GR. DR achieved better initial curve correction and greater spinal height during distraction compared with SR. No major complications were observed during the follow-up.

Key words: early-onset scoliosis, magnetic rod, growing rod, MCGR. **Spine 2013;38:665–670**

Growth-sparing techniques for treatment of progressive early-onset scoliosis (EOS) have evolved significantly during the past few decades. Originally, it was Paul Harrington,[1] the developer of Harrington instrumentation, in 1962 who recommended distraction instrumentation without fusion for children less than 10 years of age to allow continuous spinal growth. Moe *et al*[2] popularized instrumentation without fusion and included periodic construct lengthening to achieve both deformity correction and spinal growth using hooks and single distraction rod. The dual growing rod (GR) technique was introduced to provide stability, more predictable outcomes, and fewer complications than previous techniques.[3,4] During the past 5 years, other techniques have also been introduced as growth-friendly procedures.[5] Skaggs *et al*[6] have proposed a classification of the major growth-friendly techniques into 3 main categories: "distraction-based" (*e.g.*, GR, vertical expandable prosthetic titanium rib, and remote control), "tension-based" (*e.g.*, staple and tether), and "guided-growth" (*e.g.*, Luque and Shilla).

GR is currently the most commonly used distraction-based technique, which has the advantage of correcting spinal deformity, allowing normal growth, and may even have a potential for growth stimulation beyond the normal growth rate.[4] However, the technique requires frequent surgical procedures for lengthening to allow adequate spinal growth and to maintain scoliosis curve correction. Frequent surgical procedures

From the *Department of Orthopaedic Surgery, San Diego Center for Spinal Disorders; †Department of Orthopaedic Surgery, University of California San Diego (UCSD), La Jolla, CA; ‡Department of Orthopaedics and Traumatology, University of Hong Kong, Hong Kong, China; §Department of Orthopaedic Surgery, Royal National Orthopaedic Hospital NHS Trust, Stanmore; ¶Department of Orthopaedic Surgery, Cairo University, Cairo, Egypt; and ‖Department of Orthopaedics and Traumatology, Hacettepe University, Ankara, Turkey.

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The device(s)/drug(s) that is/are the subject of this manuscript is/are not FDA-approved for this indication and is/are not commercially available in the United States.

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Address correspondence and reprint requests to Behrooz A. Akbarnia, MD, San Diego Center for Spinal Disorders, 4130 La Jolla Village Dr, Ste 300, La Jolla, CA 92037; E-mail: akbarnia@ucsd.edu

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lead to an increased complication rate and more unplanned surgical procedures in severely involved children who often have other associated medical problems such as thoracic insufficiency syndrome. Bess *et al*[7] reviewed complications of GR surgery in 140 patients who underwent a total of 897 surgical procedures. The risk of complications increased by 24% for each additional surgical procedure performed.

Surgeons' knowledge regarding the various challenges in EOS surgery has also significantly improved during the past several years, and there has been an ongoing research to minimize complications associated with most growth-sparing procedures. Therefore, the idea of noninvasive multiple lengthening without the need for anesthesia and open surgery have long been appealing, given the direct relationship between high complications and repeated surgical procedures.

The concept of magnetic distraction is not new. Takaso *et al*[8] suggested that a remote magnet might be able to provide the driving force for a GR system and be used for correction of scoliosis. Soubeiran *et al*,[9] Miladi,[10] and Wilkins and Soubeiran[11] reported that a magnetically expandable GR can be used for distraction between ribs, vertebra, and the pelvis. We are reporting the preliminary results of a magnetically controlled growing rod (MCGR), which eliminates the need for repeated surgical procedures and anesthesia.

Preclinical studies examining the safety and efficacy of this MCGR system in an animal model has recently been reported.[12] This animal study confirmed that spinal distraction could be achieved safely with this technique, in the porcine model, without any adverse effect related to the MCGR system. The current study intends to report the safety and efficacy of this implant system (MAGEC Ellipse Technology, Irvine, CA) used in a human model.

DEVICE DESCRIPTION

The MCGR device was CE marked in Europe in October 2009 but is not yet approved for use in the United States. It is composed of an implantable rod, an external remote controller (ERC), and accessories (Figure 1A). The titanium rod includes a telescopic actuator portion that holds a small internal magnet. Rotation of the magnet remotely by use of the ERC causes the rod to be lengthened or shortened. The rod is implanted and secured to the spine using standard fixation components, such as hooks and/or pedicle screws as anchors. The implant system includes optional rod adapters (short straight adapter and dynamic adapter) to support physician preference of securing the rods to the construct foundations.

The handheld noninvasive ERC is electrically powered and contains 2 permanent magnets that can be rotated using gears. After the rod has been implanted, the ERC can be placed externally on the patient's spine at the location of the actuator portion of the rod. When activated, the ERC causes the magnet on the implantable device to rotate (Figure 1B). The implants provided were sterilized by gamma radiation. It is also worth mentioning, magnetic resonance compatibility of MCGR has not been examined yet, therefore at this point, its magnetic resonance compatibility is unknown, but we know that the presence of magnets within the actuator can cause artifact on magnetic resonance images.

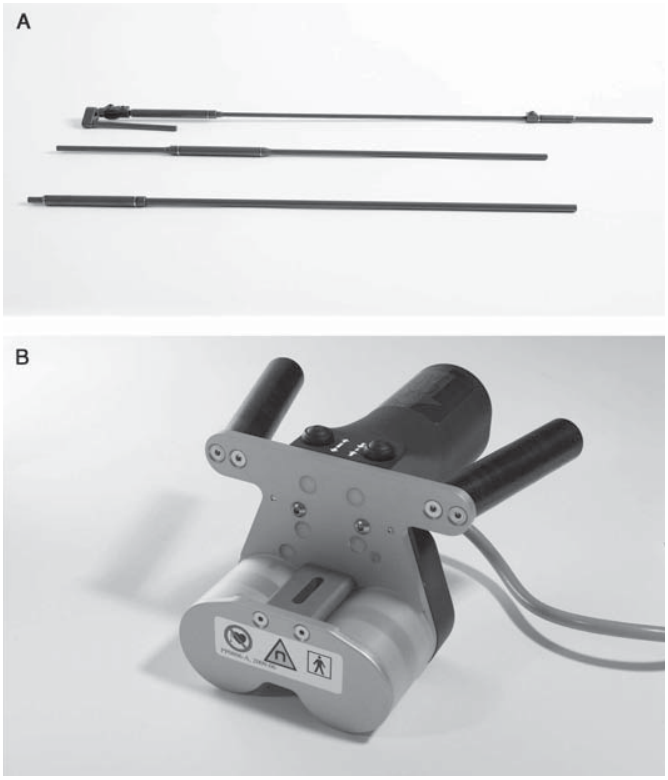

Figure 1. (**A**) MAGEC system rods. (**B**) MAGEC external remote controller.

MATERIALS AND METHODS

The study protocol was approved by the appropriate ethical committee at each study site, and patients gave consent for participation in the study. From November 2009 to December 2010, a total of 33 patients underwent the MCGR procedure at 4 centers. There were no study sites in the United States because the device had not been approved by the FDA. Clinical and radiographical data were collected prospectively before and after the initial surgery and before and after each spinal distraction. Preoperative work up and planning was similar to any pediatric spinal deformity assessment including upright posteroanterior and lateral scoliosis radiographs, supine bilateral bending and posteroanterior traction and hyperextension lateral radiographs when deemed appropriate by the treating physician.

Fourteen patients met the inclusion criteria, which included (1) EOS of any etiology; (2) a clear indication for an operative intervention (*e.g.*, documented failed nonoperative treatment and curve progression); and (3) having completed at least 3 outpatient distractions. Surgical technique for MCGR implantation, except the rod, is basically not different from traditional GR technique that has been well documented in previous literature.[3] Selecting the instrumentation length for each MCGR surgery was on the basis of the selection criteria surgeons normally used for their standard GR surgery. The length of MCGR instrumentation was usually long from the

TABLE 1. Summary of Radiographical Results of All Patients at Preoperative, Postoperative, and Latest Follow-up Time Points

	Mean Preop	Mean Postop	Latest Follow-up
Cobb angle	60°	34°	31°
Maximal thoracic kyphosis	39°	31°	48°
Total T1–T12 (mm)	178	198	208
Total T1–S1 (mm)	292	322	338

Preop indicates preoperative; Postop, postoperative.

upper thoracic vertebra to the mid- or lower lumbar levels, depending on the type, severity, and location of the curve. However, the technical pearl, similar to traditional GR, is to fuse only 1 to 2 motion segments at both upper and lower foundations to avoid unnecessarily exposure and possible iatrogenic auto-fusion.

Spinal distractions were performed on an outpatient basis with no general or local anesthesia or any pain medications. Distraction amount was measured using the end-to-end rod distance inside the actuator before and after each lengthening on posteroanterior radiographs. T1–T12 and T1–S1 heights were measured as the perpendicular between 2 parallel lines passing through the centers of upper end plate of the T1 and lower end plates of T12 and S1, respectively. T1–T12 and T1–S1 heights were recorded before and after each distraction.

The goal of the study was to identify the amount and rate of height change in the thoracic (T1–T12) and lumbar spine (T1–S1) after the MCGR surgery and to report any early complication that occurred during the course of study. Statistical analysis was done with SSPS 15 (2006) for Windows (Microsoft, Chicago, IL).

RESULTS

The mean age of the 14 patients (7 girls, 7 boys) included was 8 years, 10 months (3 yr, 6 mo to 12 yr, 7 mo). The patients had varied etiologies including 5 idiopathic, 4 neuromuscular, 2 congenital, 2 syndromic, and 1 with neurofibromatosis. The patients underwent a total of 14 initial surgical procedures and 68 spinal distractions with an average of 4.9 distractions per patient. The initial index procedures included 5 single-rod (SR) and 9 dual-rod (DR) insertions. All patients were kept in thoracolumbosacral orthosis brace for 3 to 6 months after index MCGR procedure but none underwent bracing after MCGR lengthening.

Patients were followed for an average of 10 months. Mean time between index surgery and the start of first distraction was 66 days. The mean interval between subsequent distraction procedures was 43 days.

There were 11 thoracic and 3 thoracolumbar/lumbar curves. The mean preoperative Cobb angle was 60° and was corrected to 34° immediately after initial surgery and was maintained at 31° at the time of latest follow-up. Similarly, mean preoperative maximum thoracic kyphosis was 39° and changed to 31° and 48° postoperatively and at latest follow-up, respectively (Table 1). The average coronal Cobb correction was also compared between patients who had SR and patients who had DR, and there was no significant difference in the initial and final correction between the 2 groups ($P = 0.91$ and $P = 0.85$) (Table 2) (Figures 2A–F).

Mean preoperative T1–T12 height for all 14 patients was 178 mm and increased to 198 mm ($P < 0.05$) and 208 mm ($P < 0.05$) after initial surgery and latest follow-up, respectively. Mean T1–S1 height also changed from 292 mm preoperatively to 322 mm ($P < 0.05$) after initial surgery and to 338 mm ($P < 0.05$) at the latest follow-up.

In patients with SR, the mean preoperative T1–T12 height was 178 mm, which increased to 196 mm immediately after index surgery ($P < 0.05$) and to 204 mm at the latest follow-up ($P < 0.05$). The average monthly T1–T12 height increase was 1.09 mm. The mean preoperative T1–S1 height was 295 mm, which increased to 322 mm immediately after index surgery and to 331 mm at the latest follow-up ($P < 0.05$). The average monthly T1–S1 height increase was 1.27 mm.

Similarly, in patients with DR, the mean preoperative T1–T12 height was 178 mm, which increased to 198.4 mm immediately after index surgery and to 210.5 mm at the latest follow-up ($P < 0.05$). The average monthly T1–T12 height increase was 1.97 mm. The mean preoperative T1–S1 height was 290.6 mm, which increased to 322 mm immediately after index surgery and to 342 mm at the latest follow-up ($P < 0.05$). The average monthly T1–S1 height increase was 3.09 mm.

The average monthly T1–T12 growth was not significantly different between the SR and DR groups ($P = 0.21$); however, there was a significant difference between average monthly T1–S1 growth in the 2 groups ($P < 0.05$).

TABLE 2. Comparison of Coronal Cobb Correction Between Patients With SR and Patients With DR

	Mean Preop	Mean Postop	Correction	Final	Correction (%)
Single rod	68°	38°	43%	36°	46%
Dual rod	56°	32°	42%	29°	48%

Preop indicates preoperative; Postop, postoperative.

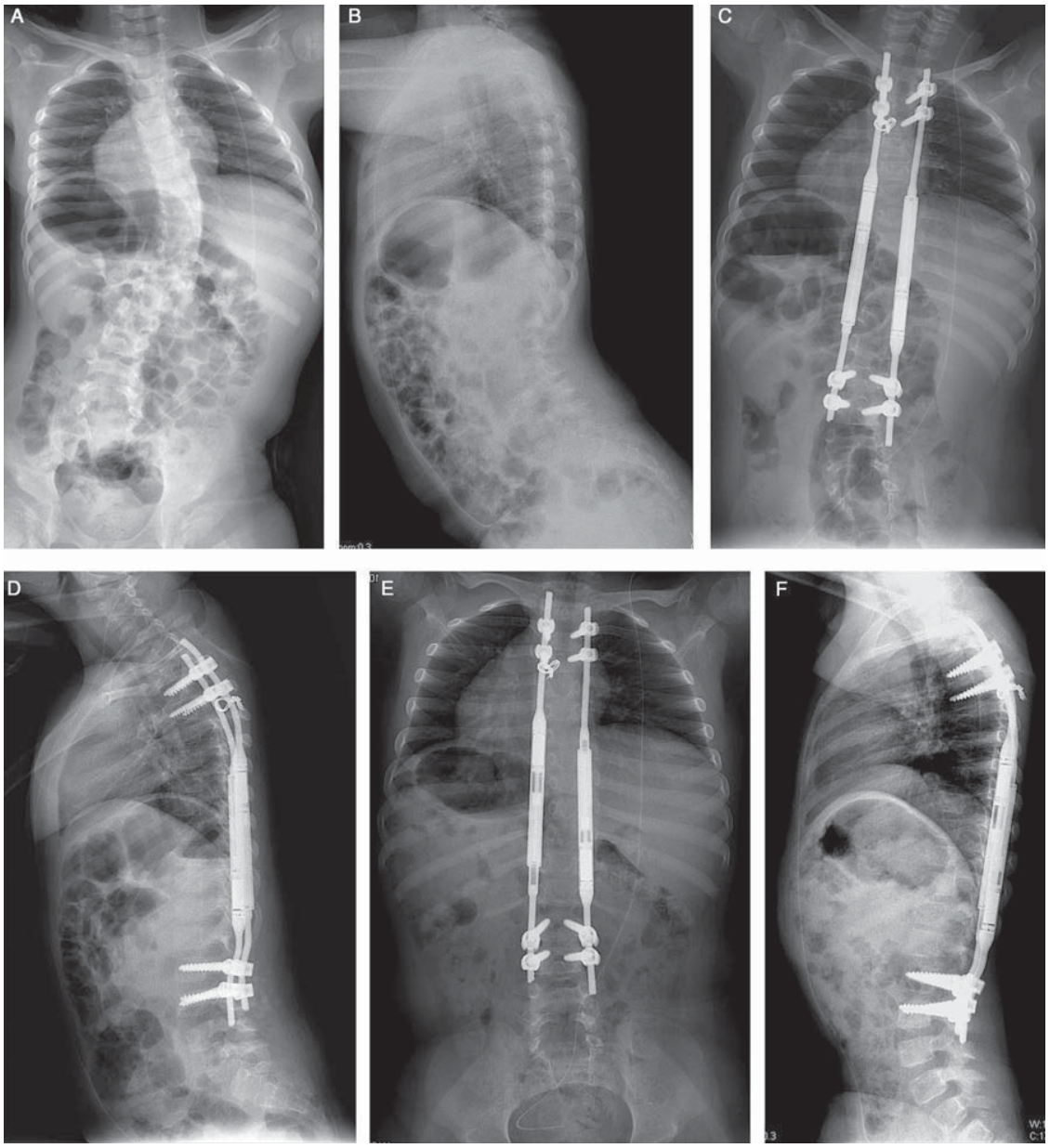

Figure 2. (**A–F**) Posteroanterior and lateral radiographs of a 5.5-year-old girl with neuromuscular scoliosis and right thoracic curve of 45° before (**A, B**) and after (**C, D**) dual MCGR. The patient has had monthly MCGR lengthening during the course of 7 months and achieved 12 mm and 31 mm of T1–T12 and T1–S1 height, respectively (**E, F**). MCGR indicates magnetically controlled growing rod.

Complications included superficial wound infection in 1 patient with SR (treated medically with antibiotics and frequent wound cleaning dressing), prominent implant in 1 patient with DR, and partial loss of initial height in 3 patients with SR (21%) after index surgery. Partial loss of distraction was the most common complication after 11 of the 68 distractions (2 DR and 9 SR). This loss was regained and maintained in subsequent distractions. No patient experienced any other implant-related complication and none needed rod exchange or revision of the anchors. Review of the radiograph at final follow-up did not show any proximal junctional kyphosis either. Finally, there was no neurological deficit at initial surgery or after lengthening.

DISCUSSION

Treatment of EOS with growth-sparing techniques ideally has 2 main goals: to correct the spinal deformity and allow for spinal (and indirect pulmonary) growth. These goals have to be met as we try to minimize complications.

Few but valuable studies have shown the effect of thoracic spinal fusion on pulmonary function. Karol *et al*[13] showed that the extent of the spine fused correlated with the forced vital capacity. Fusion in the upper thoracic spine was found to be associated with diminished pulmonary function because 8 of 12 patients with a fusion level of T1 or T2 had a forced vital capacity of less than 50%, but only 4 of 16 patients with a fusion beginning caudad to T3 had a forced vital capacity of

less than 50%. None of the 7 patients with a proximal level of fusion at T6 or caudad had a forced vital capacity of less than 50% at the time of follow-up.

Dimeglio *et al*[14] studied the growth of the spine and thoracic cage in children from birth to the onset of adulthood. They showed that T1–T12 had an average height of 110 mm at birth in both sexes and reached 180 mm at 5 years of age (average monthly growth of 1.16 mm and annual growth of 14 mm from birth to 5 yr) and then reached 220 mm at the age of 10 (average monthly growth of 0.67 mm and annual growth of 8 mm from 5–10 yr). Likewise, the average monthly T1–S1 growth from birth to 5 and from 5 to 10 years is 2 mm and 1.2 mm, respectively.

Previous studies on standard GR have shown that initial T1–S1 elongation achieved at index GR implantation is equal to T1–S1 height growth achieved during the subsequent distractions during the next 2 years (each is about 50% of the total T1–S1 achieved from preindex GR to 2-yr follow-up)[3]. In 2005, Akbarnia *et al*[3] reported that the average T1–S1 length increase was 12.1 mm per year for 23 patients with EOS who underwent dual GR surgery and were followed for at least 24 months. In a subsequent study published in 2008, they reported a mean T1–S1 increase of 14.6 mm/yr for a series of patients with dual GR with postoperative follow-up in the range of 3 to 11 years.[4] In a more recent study, GR has shown to significantly increase the T1–T12 height, and this increase was positively correlated with the number of lengthenings but did not depend on the underlying etiology.[15] In a study of 20 patients with EOS, Olgun *et al*[16] found that the growth of the vertebral body heights within the instrumented levels (7.0 ± 2.9 mm) is significantly more than those outside the instrumentation levels (5.2 ± 3.4 mm) and concluded that GR had stimulated the growth of the vertebral height compared with the instrumented levels.

Cheung *et al.* first reported two-year results of two patients with MCGR surgery and showed that the actual and predicted rod distractions were closely comparable. Patients experienced no MCGR-related complications and were satisfied with their treatment at the latest follow-up.[17] The average monthly T1–T12 and T1–S1 growth in our study is comparable, yet slightly higher than the traditional techniques. We think that this difference may be because of more frequent distractions, especially during the more rapid phase of growth; however, we expect that the spinal height achieved by MCGR will finally be similar to the above-mentioned numbers. A larger cohort of patients with longer postoperative follow-up needed to confirm this finding with more definitive conclusions.

Both single and dual standard GR have been commonly used with reproducible results. In our study, selection of single *versus* dual MCGR mainly depended on surgeon's preference. The literature on standard GR has shown that dual GR are more advantageous than single GR in terms of lower rate of rod fracture, better rate of deformity correction, and achieving a higher rate of spinal growth.[4,18] Despite being a small study, the current study confirms the same trends.

Timing of subsequent distractions mainly depended on the surgeon's preference based on the age, growth potential, curve flexibility, and diagnosis. The usual interval between distractions in the standard GR technique is between 6 to 9 months[19]; however, distraction in MCGR is noninvasively done and it can be done more frequently (with shorter intervals) if the physician prefers.

The correction of scoliosis in our study was 43% after initial MCGR and improved to 48% at final follow-up and both were comparable with the reported standard GR studies. The use of MCGR seems to be effective in correcting the coronal deformity and maintaining scoliosis curve correction (Tables 1 and 2).

We did not observe any major complication related to the MCGR in this series intraoperatively and perioperatively. Partial loss of distraction occurred in some patients with SR that were easily managed in subsequent distractions. It is important to mention that the observed distraction losses were from a first generation design. This issue was resolved by the addition of a magnetic lock that keeps rod length where the ERC positions it. The lock prevents the rod from shortening while it is implanted. Although we have not observed any implant-related complications that needed revision, we think that the revision surgery for rod exchange and anchor replacement is not different from similar procedures in the standard GR surgery. There are still some debates regarding proximal junctional kyphosis in growth-friendly procedures. Increased maximal thoracic kyphosis at final follow-up is thought to be the result of increase outside of the instrumented region but no clear proximal junctional kyphosis was observed in any of our patients.

Similar to traditional GR technique, longer span of the MCGR rods is usually preferred. The amount of deformity correction achieved in initial MCGR surgery is the result of internal bracing of the spine and almost pure distraction. This is to provide distraction over a length of unfused spine to stimulate growth. Also, the very young and growing spine tends to add to the deformity as the child grows. Extending the instrumentation in a later date will further affect the growth by creating more levels of unwanted fusion.

Autofusion in traditional GR technique has a complex pathophysiology but its main contributing factor is the technical error in unnecessary dissection of the uninstrumented, unfused vertebra. It can theoretically happen in MCGR, although we think that noninvasive distractions will reduce the incidence of this phenomenon in MCGR. We have not seen any autofusion incident in our reported patients until the latest follow-up.

Growth guided procedures (*i.e.*, Shilla) have been introduced to minimize the number of surgical procedures as they guide the spinal growth in direction of the implants.[5] The procedure does not apply any distraction to the spine. No study exists to compare the increase in length of T1–T12 and T1–S1 with the standard GR. We think that MCGR combines both advantages of reduction of surgical procedures and applying distraction at the same time.

Regarding the effects of electromagnetic field (<8–10 T) on humans, there currently is no evidence for any persistent or major side effect for repeated distraction especially in a local field; therefore, it is considered to be reasonably safe.[20]

Finally, longer follow-up and a larger group of patients are needed to determine the overall outcome of this technique and noninvasive method of lengthening and characterize its complications.

CONCLUSION

Our study shows that MCGR is a safe and effective growth-sparing modality in treatment of progressive EOS and may be considered a viable alternative to the traditional GR technique. It allows the similar growth of the T1–T12 and T1–S1 to what has been previously observed in normal children and those patients with EOS who underwent the standard GR procedure. We did not observe any major implant-related complication. Overall, the patients with DR showed better initial correction of coronal deformity and better monthly height increase of T1–T12 and T1–S1. A study with additional follow-up and a larger cohort is underway to confirm these initial findings.

➢ Key Points

- ❑ MCGR techniques (both SR and DR) achieved and maintained similar initial scoliosis Cobb correction compared with the standard GR surgery.
- ❑ Mean monthly T1–T12 and T1–S1 growths were comparable with both normal growth and the growth after the standard GR procedure.
- ❑ MCGR reduces the number of open surgical procedures required for lengthening and may significantly reduce the number of complications.

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■ SPINE

Early results of a remotely-operated magnetic growth rod in early-onset scoliosis

Z. Dannawi,
F. Altaf,
N. S. Harshavardhana,
H. El Sebaie,
H. Noordeen

From Royal National Orthopaedic Hospital, Stanmore, and Great Ormond Street Hospital, London, United Kingdom

Conventional growing rods are the most commonly used distraction-based devices in the treatment of progressive early-onset scoliosis. This technique requires repeated lengthenings with the patient anaesthetised in the operating theatre. We describe the outcomes and complications of using a non-invasive magnetically controlled growing rod (MCGR) in children with early-onset scoliosis. Lengthening is performed on an outpatient basis using an external remote control with the patient awake.

Between November 2009 and March 2011, 34 children with a mean age of eight years (5 to 12) underwent treatment. The mean length of follow-up was 15 months (12 to 18). In total, 22 children were treated with dual rod constructs and 12 with a single rod. The mean number of distractions per patient was 4.8 (3 to 6). The mean pre-operative Cobb angle was 69° (46° to 108°); this was corrected to a mean 47° (28° to 91°) post-operatively. The mean Cobb angle at final review was 41° (27° to 86°). The mean pre-operative distance from T1 to S1 was 304 mm (243 to 380) and increased to 335 mm (253 to 400) in the immediate post-operative period. At final review the mean distance from T1 to S1 had increased to 348 mm (260 to 420).

Two patients developed a superficial wound infection and a further two patients in the single rod group developed a loss of distraction. In the dual rod group, one patient had pull-out of a hook and one developed prominent metalwork. Two patients had a rod breakage; one patient in the single rod group and one patient in the dual rod group. Our early results show that the MCGR is safe and effective in the treatment of progressive early-onset scoliosis with the avoidance of repeated surgical lengthenings.

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■ Z. Dannawi, FRCS(Tr & Orth), Spinal Fellow
■ F. Altaf, BSc(Hons), Specialist Registrar Trauma and Orthopaedics
■ N. S. Harshavardhana, MS(Orth), Spinal Fellow
■ H. Noordeen, FRCS(Tr & Orth), Consultant Orthopaedic Spinal Surgeon
Royal National Orthopaedic Hospital, Brockley Hill, Stanmore, Middlesex HA7 4LP, UK.

■ H. El Sebaie, FRCS, MD, Professor of Orthopaedic surgery
Cairo University, School of Medicine, Al-Saray Street, El Manial, Cairo, PO Box 11956, Egypt.

Correspondence should be sent to Mr F. Altaf; e-mail: farhaanaltaf@hotmail.com

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Early-onset scoliosis (EOS) has been defined as significant spinal deformity beginning before the age of five years.[1] Its most common types are infantile idiopathic, congenital and neuromuscular.[2] Left untreated, it can rapidly progress causing cosmetic disfigurement and pulmonary insufficiency.[3] The goals of treatment are to control the deformity, allow growth of the spine and chest wall and to improve pulmonary function. Casting or bracing are commonly used for early intervention, but bracing is not effective in controlling the curves of neuromuscular or congenital scoliosis.[4-7] When the deformity is progressive and severe, surgical treatment is often indicated. Spinal fusion can be performed with or without instrumentation. If undertaken before growth is complete, it results in a short trunk with possible underdevelopment of the lungs and subsequent pulmonary complications.[8-10]

Procedures that allow or encourage growth of the spine and chest have recently gained popularity. These are variously referred to as 'growth-sparing' or 'growth-friendly'.[11] The most commonly used devices are single and dual growing-rod implants. The traditional growing rod is inserted across the spinal curvature under general anaesthesia and ideally distracted every six months or sooner.[12] This requires re-opening the surgical incision to distract the rod to mimic and maintain normal spinal growth. The disadvantages of using the traditional growing rod include the need for general anaesthesia and the invasiveness of repeated distractions. These implants have been associated with high rates of complication including wound infection, rod breakage, auto-fusion, anchor failure or prominence of the implant.[13,14]

There are also socioeconomic disadvantages to the use of traditional growing rods and their repeated surgeries, including the indirect costs to child and parents as a result of the time missed from school and absence from work.[15] Children exposed to multiple operations are also more likely to experience

Fig. 1

Photograph of a magnetically controlled growing rod with a telescopic actuator portion that holds a small internal magnet.

severe symptoms of post-traumatic stress disorder and depression than those who experience a single event.[16]

An earlier animal study by Takaso et al[17] suggested that the use of a remotely-controlled extending rod instrumentation system could decrease the number of procedures required. That study involved five beagles with induced scoliosis. The mean initial Cobb angle of 25° was corrected to a mean of 3° after four distractions over a period of 12 weeks. No complications were recorded.[17] Akbarnia et al[18] implanted remotely expandable rods in six immature Yucatan pigs. The mean distraction achieved was 39 mm over a seven-week period without any implant-related complications.

In the only study published so far on the use of a magnetically controlled growing rod (MCGR) it was shown possible in five children to correct a spinal curvature and encourage normal spinal growth.[19]

We present the largest series to date of the use of MCGR in humans.

Patients and Methods

Between November 2009 and March 2011, 34 children with EOS were treated using magnetically-controlled growing rods with prospective collection of data. The inclusion criteria were EOS from any cause, failed non-operative treatment with bracing or observation and progression of the curvature of > 10° over a six-month period with a Cobb angle[20] > 40°. Infected patients were excluded.

Two patients had previously undergone conventional treatment with growing rods before having an MCGR implant. The parents of the two children who underwent conversion from the conventional growing rod to the MCGR requested this new technique on the grounds that their children had significant remaining growth potential and felt that their children would benefit from non-invasive lengthening, having learnt of the treatment from other patients. One of these children, aged six years, had an infantile scoliosis with a left-sided thoracic curve and a Cobb angle of 75° at the time of conversion. A dual MCGR construct was implanted between T3 and L2. The other had a diagnosis of neuromuscular scoliosis and had a left lumbar curve with a Cobb angle of 80° for which a dual MCGR was implanted between T2 and the pelvis. The mean age of the children included in the series was eight years (5 to 12). There were 13 boys and 21 girls. The minimum length of follow-up was 12 months (mean 15 months (12 to 18)). The diagnosis was idiopathic scoliosis in 14 children, neuromuscular scoliosis in 11, congenital scoliosis in two, neurofibromatosis in three, and there were four syndromic patients. Skeletal maturity was assessed using Risser staging[21] and by evaluating the triradiate cartilage.

In all a single-rod construct was used for 12 patients and 22 required a dual-rod construct. All operations were performed by the senior author (HN). Each patient had at least three magnetically controlled lengthenings.

The decision to implant single or dual rods was dependent on the size of the patient and the preference of the surgeon. Single rods were implanted into slim patients to avoid the complications of symptomatic prominent metalwork. As our experience with the MCGR technique increased, we felt that the dual-rod construct provided better correction and greater stability and was therefore implanted into older patients with a higher body mass index.

The MCGR implant is made of titanium and includes a telescopic actuator portion that holds a small internal magnet (Fig. 1). Rotation of the magnet, by the use of remote control, causes the rod to lengthen or shorten.

Under general anaesthesia, with the patient prone, two separate posterior proximal and distal skin incisions were made over the foundation levels. In the revision cases we used the whole length of the previous scar to remove the previous metalwork and to implant the MCGR. A subperiosteal dissection was performed and the proximal and distal anchor sites prepared. A combination of hooks and pedicle screws were used as anchors. The magnetic rods (MAGEC Ellipse technology, Irvine, California) were contoured, tested and inserted submuscularly either as a single-rod (Fig. 2) or a dual-rod construct (Fig. 3). The single rod was always placed on the concavity of the curve. The levels of instrumentation did not vary between the single and dual-rod technique. When dual rods were used a connector was inserted between the two rods. Local bone graft was used at the exposed foundation levels to achieve a limited fusion and stability at the anchor sites.

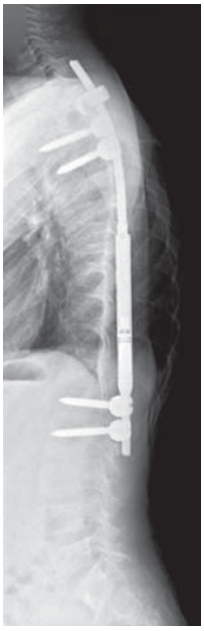

Fig. 2a

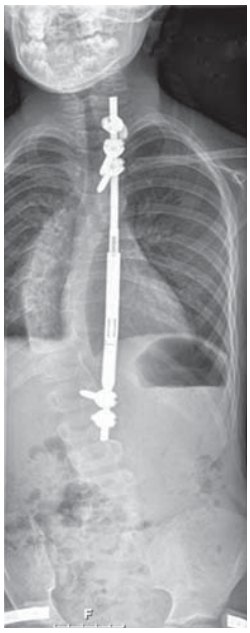

Fig. 2b

Lateral (a) and posteroanterior (b) radiographs in a ten-year-old girl at eight months after implantation of the single magnetic growth rod.

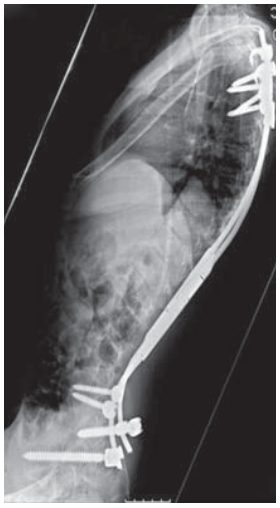

Fig. 3a

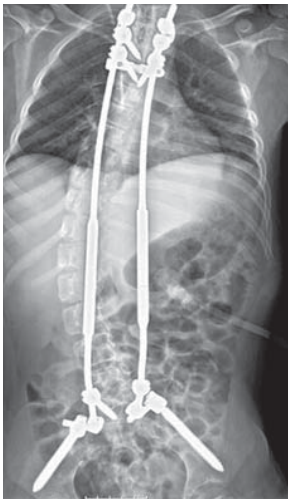

Fig. 3b

Lateral (a) and posteroanterior (b) radiographs in a seven-year-old boy at six months after implantation of the dual magnetic growth rod in a case of neuromuscular scoliosis.

The lengthenings were performed by an electrically powered remote controller. This contained two permanent magnets that could be rotated using gears. The remote control was placed externally over the patient's spine at the level of the actuator portion of the rod, which contained a magnet. The magnetic field from the latter was identified using an external magnet that was attracted to the rod magnet. When activated, the external remote control causes the magnet of the implanted device to rotate. The spinal distractions were performed on an outpatient basis without the need for anaesthesia or analgesia. All distractions were performed by the senior author (HN). The predicted lengthening was displayed on the external distraction device, which could also be used to retract the rod if the patient was uncomfortable or in pain. Pre- and post-distraction radiographs were taken for all patients to confirm that the rods had been lengthened.

With the dual-rod construct we inserted a standard rod on one side of the spine and an offset rod on the other side. The internal lengthening mechanism of the offset rod was located at the opposite end of the thickened portion of the rod from the standard rod in order to prevent interaction between the magnetic components of the rods during individual distractions. The levels of implantation and fixation were determined by the type of curve and the underlying pathology with the proximal fixation being usually at the level of T2-T4 and the distal instrumentation at the neutral vertebra intersected by the central sacral vertical line (CSVL). In neuromuscular cases, distal instrumentation was continued to the pelvis using iliac bolt screws.

The lengthening was performed over a mean of 87 days (56 to 132). Each patient was given an appointment at three-month intervals for distraction. Two patients who had an initial loss of distraction were seen at 56 and 61 days, respectively, to address this. These patients had a retainer magnet (externally applied to the skin at the site of the actuator) to maintain the distraction and prevent its collapse. The rod design was subsequently changed by the manufacturer to avoid this complication. One patient was lengthened at 132 days as he failed to attend his outpatient appointment at three months because of intercurrent illness.

Previous work has shown that patients lengthened at intervals of less than six months have a higher annual growth rate (1.8 cm *vs* 1.0 cm) than those lengthened at an interval between nine and 20 months.[22] Using this information we chose a distraction interval of three months to optimise the growth potential of the spine.

We calculated the length of predicted distraction based on the growth chart by DiMeglio and Bonnel[23] for healthy children. In general, the spine was distracted by approximately 1.5 mm per month with the aim to do this at a faster rate than the predicted spinal growth to allow for better curve correction. Based on our distraction protocol of three months, we aimed to achieve a distraction of approximately 4.5 mm per visit.

One spinal fellow (ZD) and one orthopaedic registrar (FA) measured the Cobb angles and T1-S1 length preoperatively, post-operatively, and before and after each distraction. Any discrepancy was discussed and a final measurement agreed by consensus. The authors measured the distraction as it pertained to the telescopic portion on radiographs. The distance between T1 and S1 was used as an

Table I. Mean Cobb angles, global thoracic kyphosis angles and T1-S1 lengths pre- and post-operatively and at final follow-up, and by single- and double-rod constructs

	Pre-operative	Post-operative	Mean improvement	p-value (pre-op *vs* post-op)*	Final follow-up	p-value (ANOVA)†
Mean Cobb angle (°) (range)	69 (46 to 108)	47 (28 to 91)	22	< 0.001	41 (27 to 86)	< 0.001
Single-rod	67 (46 to 108)	48 (33 to 91)	19	< 0.001	44 (32 to 86)	< 0.001
Double-rod	70 (56 to 95)	46 (28 to 60)	24	< 0.001	40 (27 to 56)	< 0.001
p-value (single- *vs* double-rod)‡	0.58	0.47	0.04		0.22	
Mean global thoracic kyphosis (°) (range)	33 (26 to 42)	29 (27 to 41)	4	0.13	32 (28 to 42)	-
Mean T1-S1 length (mm) (range)	304 (243 to 380)	335 (253 to 400)	31	< 0.001	348 (260 to 420)	< 0.001
Single-rod	307 (290 to 380)	335 (310 to 400)	28	< 0.001	347 (317 to 420)	< 0.001
Double-rod	302 (243 to 352)	335 (253 to 386)	33	< 0.001	349 (260 to 401)	< 0.001
p-value (single- *vs* double-rod)‡	0.62	0.89			0.94	

* paired *t*-test
† analysis of variance
‡ unpaired *t*-test

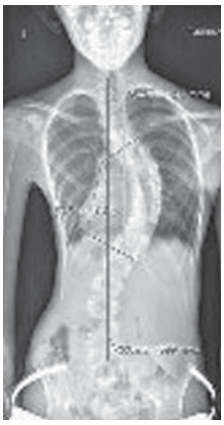

Fig. 4a

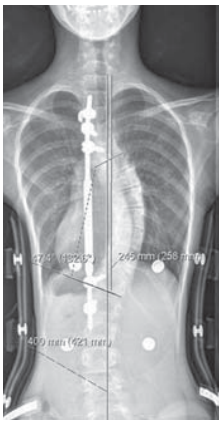

Fig. 4b

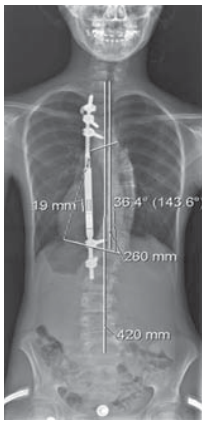

Fig. 4c

Postero-anterior radiographs showing an example of improvement in Cobb angle and lengthening after single-rod implantation, a) pre-operatively (Cobb angle 62°, T1-S1 length 380 mm), b) immediately post-operatively (Cobb angle 47°, T1-S1 length 400 mm), and c) at 12 months post-operatively (Cobb angle 36°, T1-S1 length 420 mm).

indication of growth and truncal height, and measured as the vertical distance between the two parallel lines that passed through the centres of the upper end-plates of T1 and S1. Complications that were intra-operative or implant-related and general complications, such as wound infections, were recorded.

Statistical analysis. Comparison of the pre-operative and post-operative Cobb angles and the distance between T1 and S1, was evaluated using a paired-samples *t*-test, unpaired *t*-test, and analysis of variance (ANOVA) using SPSS v20 software (IBM, Armonk, New York). An independent samples *t*-test was used to compare patients who underwent treatment with single magnetic growth rod with those with dual magnetic growth rods. A p-value < 0.05 was considered statistically significant.

Results

There were a mean of 4.8 distractions (3 to 6) per patient. The mean time to the first distraction was 66 days (22 to 112). The mean time between distractions was 87 days (56 to 132). The minimum follow-up was 12 months from the time of initial surgery, with a mean of 15 months (12 to 18).

There was improvement in the mean Cobb angle and T1-S1 lengthening, and to a lesser extent the global thoracic kyphosis (Table I, Fig. 4).

In the single-rod group there was a significant difference between the mean pre-operative, immediate post-operative and final Cobb angles (ANOVA, p < 0.001). In addition, there was a significant reduction in the mean Cobb angle between the pre-operative and immediate post-operative measurements (paired *t*-test, p < 0.001)

(Table I). There was a significant difference between the mean pre-operative, immediate post-operative and final distance between T1 and S1 (ANOVA, $p < 0.001$), and a significant increase in the mean T1-S1 distance between the pre-operative and immediate post-operative measurements (paired *t*-test, $p < 0.001$) (Table I).

In the dual rod group there was a significant difference between the mean pre-operative, immediate post-operative and final Cobb angles (ANOVA, $p < 0.001$) and significant reduction in the mean Cobb angle between the pre-operative and immediate post-operative measurements (paired *t*-test, $p < 0.001$) (Table I). There was a significant difference between the mean pre-operative, immediate post-operative and final distance between T1 and S1 (ANOVA, $p < 0.001$). Additionally, there was a significant increase in the mean distance between T1 and S1 between the pre-operative and immediate post-operative measurements (paired t-test, $p < 0.001$ (Table I).

The difference in the mean pre-operative Cobb angles between the single and dual rod groups was not significant (unpaired *t*-test, $p = 0.58$). The mean improvement in Cobb angle (immediate post-operative to pre-operative Cobb angle) was significantly different between the single and the dual rod groups (unpaired *t*-test, $p = 0.04$).

One patient in each group developed a superficial wound infection that was successfully treated with oral antibiotics. There were also two patients in the single-rod group who had a loss of distraction that was rectified by subsequent lengthening. In each group there was one rod breakage that required revision of the construct. The breakage in the single-rod construct occurred in a patient who had previously undergone conventional growing rod treatment. The breakage was noticed before her second lengthening. Hook pull-out occurred in one patient in the dual rod group. Another patient had prominent metalwork that required trimming of the rod.

No patient encountered more than one complication. Post-operative fusion did not occur in any patient and no patient developed a neurological deficit.

Discussion

Treatment of children with an early onset scoliosis should focus not only on the spinal deformity but also on the growth of the chest wall and development of the lungs. If surgical correction is required and fusion is performed before the completion of growth, the child is left with a short trunk and a disproportionate body habitus. This may adversely affect lung development and result in respiratory insufficiency in the very young.[24,25] There is also a possibility of developing further deformity from the 'crankshaft' phenomenon.[24,25]

In order to allow continuous spinal growth and overcome the complications arising from spinal fusion, Harrington[26] advocated distraction instrumentation without fusion in young children. Moe et al[27] subsequently developed the technique of periodic lengthening of the construct, using single distraction rods without fusion, to correct the deformity and maintain spinal growth. Over the last decade many growth-sparing procedures have gained popularity. Sankar et al[11] classified the various growth-friendly procedures into three types: 1) distraction-based devices; 2) tension-based devices; and 3) growth-guided procedures. The most commonly used spine-based distraction technique is the 'growing rods' method. This technique follows the concept of periodic distraction to catch up and keep pace with the demands of growth. Single and dual growing rod constructs have been used in an attempt to control spinal deformity and allow continued spinal growth. The proponents of the dual rod constructs report better correction of deformity, increased stability and a greater proportion of expected spinal growth than that achieved with single rods.[28]

Conventional growing rods allow spinal growth to continue and prevent curve progression but need multiple interventions that increase the risk of infection,[14,29,30] complications of general anaesthesia, and stress to both parents and children.

In our study, only two of 34 patients developed a wound infection, which is better than a conventional growth rod series.[14,29,30]

Implant-related complications such as rod fracture, anchor failure or prominence of the implant, were the most frequent complications in one series of growth rods.[31] Rod fractures are inevitable with non-fusion techniques. Yang et al[32] identified several risk factors that increased the probability of rod breakage in a multicentre analysis of 322 patients with growing rods. These were single-rod constructs, rods of smaller diameter, stainless steel rods and ambulatory patients. By contrast, Thompson et al[28] showed a higher rate of rod fracture in the dual-rod group despite the perceived greater stability provided by the construct. Klemme et al[33] reported 33 implant-related problems in 25 patients (37%) including one death during rod exchange through a subfascial tunnel when the rod was deflected and transgressed the retroperitoneal space and thorax. Bess et al[13] showed a linear decrease in survivorship for each operation undertaken with 49% chance of having a complication after seven procedures.

In our preliminary series, there were two rod breakages in 34 patients, a lower incidence than that reported in a conventional growing-rod series,[31] although our follow-up was shorter. The two cases of rod breakage occurred in patients with a residual coronal imbalance of > 2 cm at the time of the original surgery. It is likely this resulted in increased stress and bending moments on the rod and subsequent rod breakage. The loss of distraction that occurred in our first cases was again related to excessive stress caused by the bending moment on the rod, leading to slippage of the magnetic mechanism. We eliminated the slippage by the addition of a retainer magnet and the rod design was subsequently changed by the manufacturer, which has eliminated this problem.

Non-invasive extendible endoprostheses have been used successfully to reconstruct the lower limb after resection of malignant bone tumours in children.[34] Miladi and Dubousset[35] reported the use of a magnetically expandable growing rod that can be used for distraction between ribs, vertebra and pelvis. The idea of non-invasive multiple lengthening in the treatment of scoliosis without the need for open surgery under anaesthesia is appealing given the direct relationship between repeated operations and a high rate of complication. There is no evidence that the electromagnetic field causes any persistent or major side effect with repeated distraction, especially in a local field.[36]

Limitations of this study include the variation in the diagnosis, curve types, gender, post-operative distraction period and the number of distractions, as well as short duration of follow-up.

Our preliminary results show that the use of MCGR in treatment of progressive EOS is safe and effective in correcting deformity while allowing continuous spinal growth. Overall, we found fewer complications than series that had reviewed conventional growing rods.[14,31] Lengthening of the MCGR can be carried out as an outpatient procedure and results in comparable spinal growth to the conventional growing rod procedure. A larger cohort of patients with a longer follow-up is required to determine the long-term outcomes of this technique.

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DEFORMITY

Improvement of Pulmonary Function in Children With Early-Onset Scoliosis Using Magnetic Growth Rods

Wai Weng Yoon, MBBS, BSc (Hons), FRCS (Tr & Orth),* Fady Sedra, MD,* Suken Shah, MRCS,† Colin Wallis, MD,‡ Francesco Muntoni, MD,§ and Hilali Noordeen, FRCS*

Study Design. Case series.

Objective. To determine whether there is improvement in pulmonary function in children with early-onset scoliosis (EOS) using magnetic growth rods (MGRs).

Summary of Background Data. EOS deformities have large impacts on lung function and volumes. Deterioration of pulmonary function in scoliosis is multifactorial, including severity, location of apex vertebra, and medical comorbidities. MGR insertion has benefits including reduction in operative procedures with repeated anesthetics, cost-effectiveness, and minimizing surgical and psychological distress. Pulmonary function tests provide objective and quantitative information about functional impairment caused by scoliosis. This is the first study that observes the MGR lengthening and changes in pulmonary function during a minimum period of 2.2 years.

Methods. Six cases of EOS secondary to neuromuscular disease were identified. Mean age at diagnosis was 2.8 year (2.1–4.9 yr), mean age at surgery was 7.5 year (5–10 yr), and mean follow-up was 2.5 year (2.2–2.8 yr). Pulmonary function test (forced vital capacity [FVC] + forced expired volume in 1 second [FEV1] both % predicted) was measured before and after insertion of MGR and at every lengthening clinic subsequently for a minimum 2 years. Coronal and sagittal Cobb angles were measured pre- and postoperatively as were length extension of growth rods. All except 1 patient had dual MGRs inserted (the other had a single rod). Lengthening was commenced and data was collected at 6-month intervals.

Results. Average correction was 34° ± 18° and 36° ± 15° for coronal and sagittal Cobb angles, respectively. Mean lengthening achieved was 24.9 mm. Mean improvement in postoperative FVC and FEV1 was 14.1% and 17.2%, respectively. There was significant difference between the median preoperative and postoperative Cobb angle, $P = 0.028$.

Conclusion. This study demonstrates early intervention using MGR in patients with EOS is associated with significant improvement in postoperative pulmonary function tests; and significant improvement in deformity correction with use of MGR with added benefits of reduction in repeat anesthesia, reduction in surgical and psychological distress, and cost-effectiveness.

Key words: early-onset scoliosis, neuromuscular, magnetic growth rod, respiratory function.

Level of Evidence: 4

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From the *Spinal Deformity Unit, Royal National Orthopaedic Hospital, Stanmore, Middlesex, United Kingdom; †Spine and Scoliosis Center, Nemours/Alfred I. duPont Hospital for Children, Wilmington, DE; ‡Respiratory Paediatric Department, Great Ormond Street Hospital, London, United Kingdom; and §Dubowitz Neuromuscular Centre, Great Ormond Street Hospital, London, United Kingdom.

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The manuscript includes unlabeled/investigational uses of the products/devices listed below and the status of these is disclosed in the manuscript: Magec Rod.

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Address correspondence and reprint requests to Wai Weng Yoon, MBBS, BSc (Hons), FRCS (Tr & Orth), Spinal Deformity Unit, Royal National Orthopaedic Hospital, Brockley Hill, Stanmore, Middlesex HA7 4LP, United Kingdom; E-mail: wwyoon@me.com

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Early-onset scoliosis (EOS) is defined as primarily a coronal plane spinal deformity commencing before the age of 5 years[1] and as it progresses it has significant impact on pulmonary function.[2] Growing rods are among the most common of the distraction-based techniques employed for correction of the deformity and maintenance of spinal and thoracic growth, but have the downside of requiring frequent open surgical distractions under general anesthesia with the associated complications; as much as 19% per procedure.[3] In patients with neuromuscular scoliosis, this is particularly relevant as they are more susceptible to infection and associated restrictive lung disease due to thoracic muscle weakness, poor cough function and aspiration.[4] This medically complex group of patients frequently demonstrate an FVC less than 30% (predicted) and in

some circumstances have been considered too sick for repeated anesthetics required for multiple lengthening procedures associated with conventional growth rod or posterior distraction-based treatment.

Magnetic growth rods (MGRs) are a relatively new technology that allow posterior distraction of the growing child's spine in the clinic or office setting, noninvasively, while the patient is awake and hence offer marked benefits including a reduction in the number of surgical procedures.[5–7]

Pulmonary function tests provide objective and quantitative information about the type, severity, and functional impairment caused by scoliosis and the severity of scoliosis is associated with diminishing pulmonary function, especially in young children. Furthermore, the detriments of fusing a young child's spine and limiting thoracic spinal height and chest volume growth are coming to light.[8]

In this study, we aimed to determine if MGR distraction demonstrates an improvement in pulmonary function.

MATERIALS AND METHODS

Six patients with EOS secondary to neuromuscular disease who had failed conventional nonoperative treatment, such as casting or bracing were prospectively treated between 2010 and 2013. Two patients had such a significant lung function deficit that they were excluded from our conventional growth rod program. Every patient underwent preoperative pulmonary function testing and preoperative FEV1 and FVC were compared with the postoperative FEV1 and FVC. Percent-predicted values were derived from Stanojevic equations.[9] Patients subsequently underwent MGR implantation (Ellipse Magec growth rods; Ellipse Technologies, Irvine, CA) with sequential remote lengthening at between 3- and 6-month intervals for a minimum of 2 years with an average of 7 (6–9) distractions with repeat pulmonary function testing in a lung function laboratory. The coronal and sagittal Cobb angles were measured pre- and postoperatively, as were the length extension of the growth rods and T1–S1 height (measured from the middle of the upper endplate of T1 to the middle of the upper endplate of S1). The growth rods were magnetically lengthened and measured by calibrating the radiographs (obtaining radiographs immediately before and after lengthening) using identifiable markings on the extendable portion of the rod (Figure 1). All except 1 patient had dual MGRs inserted. The technique favored a consolidation period of 3 months before lengthening in all cases in order for fusion to be established at the anchor points.

The Wilcoxon signed rank test was used to compare the pre- and postoperative outcomes. Correlation analysis performed to assess the relationships between the amount distracted and the change in outcome was conducted using Spearman rank correlation coefficient (ρ). All statistical analysis was performed using Stata/IC, version 12.0 (STATACORP LP, College Station, TX). Throughout, a $P < 0.05$ was considered statistically significant.

RESULTS

Six patients included 4 males and 2 females. Two patients had spinal muscular atrophy-II, 2 had neurofibromatosis-2, 1 had Williams syndrome, and 1 had congenital muscular dystrophy. The mean age of the patients at diagnosis was 2.8 years (2.1–4.9 yr) and the mean age at surgery was 7.5 years (5–10 yr). The mean follow-up period was 2.5 years (2.2–2.8 yr).

The mean preoperative Cobb angle was 87° ± 23° in the coronal plane and 66° ± 14° in the sagittal plane. The mean coronal and sagittal correction was 34° ± 18° and 36° ± 15°, respectively. Kyphosis was corrected to a normal range (20°–50°),[10] and coronal plane deformity was significantly improved ($P < 0.03$) at the last lengthening. The mean lengthening achieved was 24.9 mm (10.9–26.6 mm). The mean height gain between T1 and S1 (immediately postoperatively to the last follow-up) was 27.2 mm. The mean height gain was 15 mm, 1 year after lengthening and 12.1 mm at year 2 (Tables 1, 2). MGR improved pulmonary function in these respiratory compromised children (Tables 1, 2): the Wilcoxon signed rank test identified a significant difference between the median pre- and postoperative FVC ($P = 0.028$) and FEV1 ($P = 0.027$). There was also a significant difference found between the median pre- and postoperative Cobb angle, $P = 0.028$ (Figure 2A, B).

There was a moderate positive trend between the amount distracted and improvement in FVC P = 0.486 ($P = 0.329$). Greater changes in the FVC were associated with larger amounts of distraction in this sample of 6 patients; however this relationship was not significant.

Complications included a reoperation for trimming of a prominent rod in 1 case and 1 rod breakage in the other (at

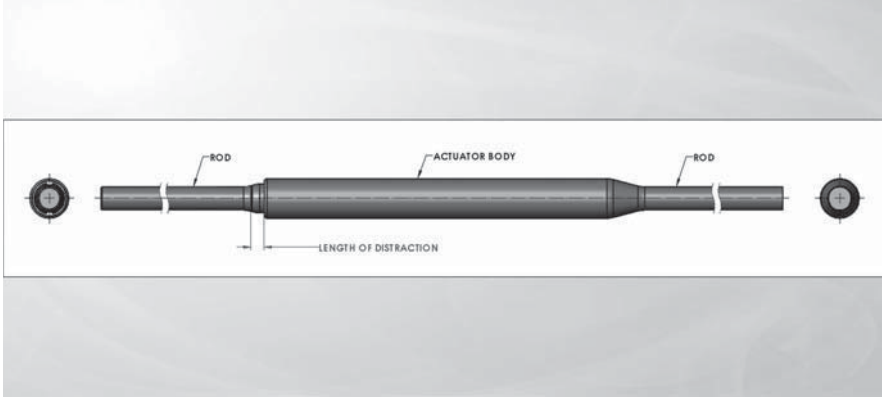

Figure 1. Magnetic growth rod—diagram with measurement points.

TABLE 1. Pulmonary Function Data for the Study Patients Preoperative and at Postoperative Follow-up of 31 ± 2 Months Data are Presented as Median (IQR)

Outcome	Preop (n = 6)	Postop* (n = 6)	$P < 0.05$
%-predicted FVC, mean (range)	27 (15–45)	41 (28–60)	0.028
%-predicted FEV1, mean (range)	27 (15–43)	45 (25–66)	0.027

**Average postoperative follow-up of 31± 2 months.*

Preop indicates preoperative; postop, postoperative; IQR, interquartile range; FVC, forced vital capacity; FEV1, forced expired volume in 1 second.

the motor region of the MGR), which required reimplantation (this was the single-rod construct). There was no neurological deficit postoperatively or at each lengthening procedure.

DISCUSSION

The treatment of EOS and pulmonary function has been studied in the past[11] and it has a multifaceted association. This is the first study of the use of MGRs in early-onset neuromuscular scoliosis specifically evaluating pulmonary function. Many groups have focused on the high incidence of complications in this cohort with increased risk of wound complications (33%) and pulmonary complications postoperatively.[12,13] Our study did not encounter these problems. We used the same protocols and level selection as for our conventional growth rod system with proximal and distal fusion sites allowing for minimal periosteal segment exposure to prevent spontaneous fusion (Figures 3–8). Submuscular placement of the rods is also standard in our practice, which has been shown to reduce wound complications.[3] We are aware that this patient population has increased risk for surgery as well as postoperative complications but the results have shown a significant improvement in pulmonary function (a first in this patient population). This may be due to the fact that these patients have such poor pulmonary function at baseline that moderate changes in thoracic spinal height and chest wall volume have great effects. Our study shows a correlation with lengthening and improvement in pulmonary function but more patients are required to make this significant association. In this study, we had 1 rod breakage in the motor segment of the MGR. This case was a single-rod breakage where only 1 rod was employed. There were no complications in the revision of this rod.

In our study the intervals of lengthening were between 3 and 6 months. Some have suggested that spontaneous spinal fusion may result from multiple distractions and this may result in a reduction in T1–S1 lengthening over time,[14] however in all subjects in this study, growth continued with distraction. The mechanism by which fusion occurs has been

TABLE 2. Preoperative and Postoperative Patient Characteristics

Patient	Age at MGR Insertion	Apex Vertebrae	Diagnosis	Preop Cobb Angle (°)	Immediate Postop Cobb Angle (°)	2-Year Follow-up Cobb Angle (°)	No. of Lengthenings	Length of Distraction (mm)	Immediate Postop T1–S1 (mm)	2-Year Follow-up T1–S1 (mm)	PFT %-Pred Preop FVC	PFT %-Pred Preop FEV1	PFT %-Pred Postop FVC	PFT %-Pred Postop FEV1
1	10	T8	Williams syndrome	78.8	48.1	59	8	26.6	358	380	50	54	86	82
2	5	L1	SMA II	80.5	39.6	14.3	7	52.7	332	365.9	32	33	49	60
3	6	T12	SMA II	105.9	58.9	49.1	7	29.1	327.6	343.2	15	16	27	26
4	5	T6	CMD	121.8	76.8	94.5	6	13.9	273	295	14	13	28	23
5	6	T9	NF-2	69.8	57.8	49.5	9	16.4	301.5	342	43	40	51	56
6	10	T10	NF-2	63.2	59.8	52.8	7	10.9	321.1	350	22	21	32	33

SMA indicates spinal muscular atrophy; CMD, congenital muscular dystrophy; MGR, magnetic growth rod; Pred, predicted; NF, neurofibromatosis; Preop, preoperative; Postop, postoperative; PFT, pulmonary function test; FVC, forced vital capacity; FEV1, forced expired volume in 1 second.

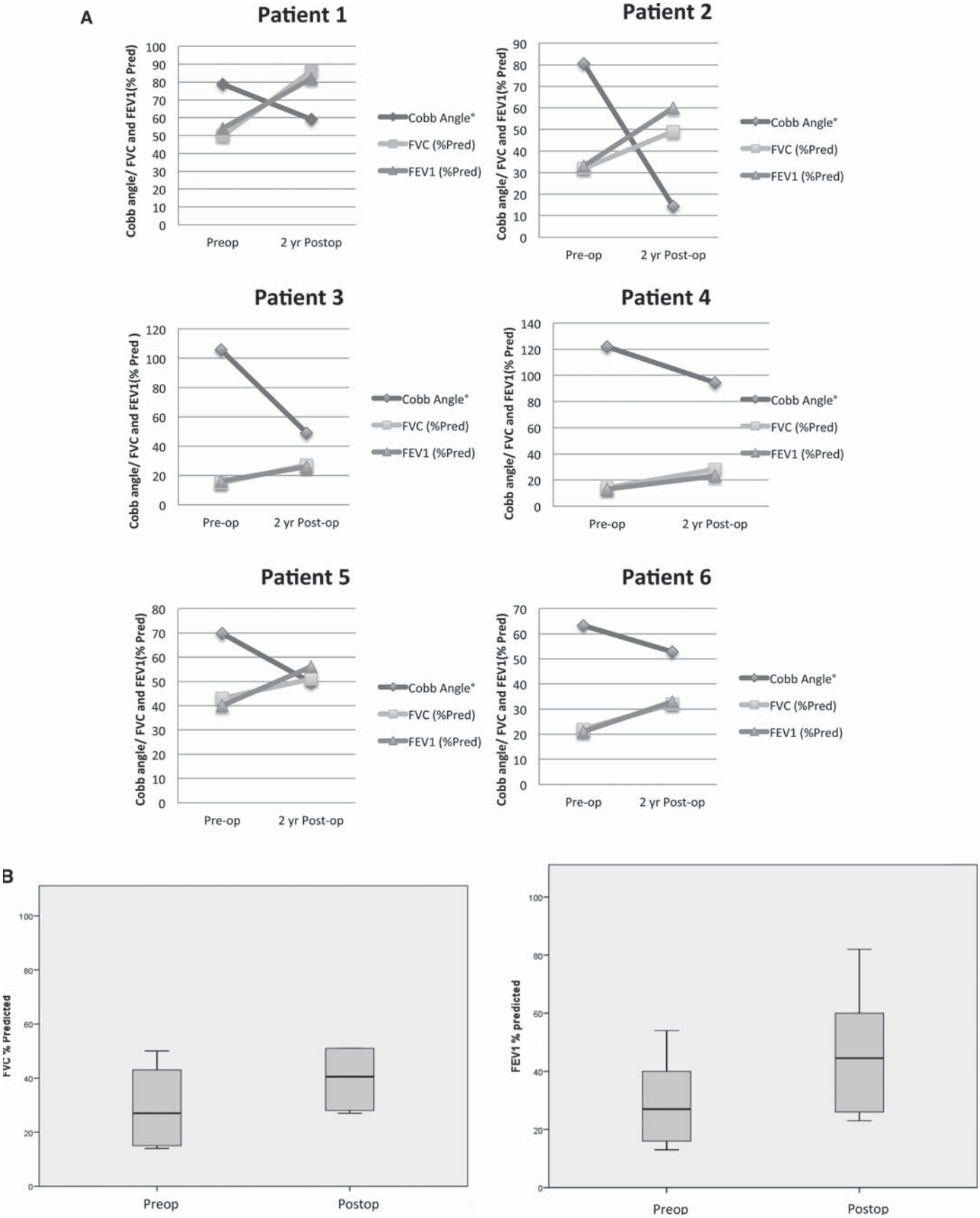

Figure 2. A, Individual patient comparison between change in Cobb angles and pulmonary function tests. **B**, Graphs illustrating the outcome over time (preoperative vs. postoperative) (plots illustrate the median and IQRs of the observed data). IQR indicates interquartile range.

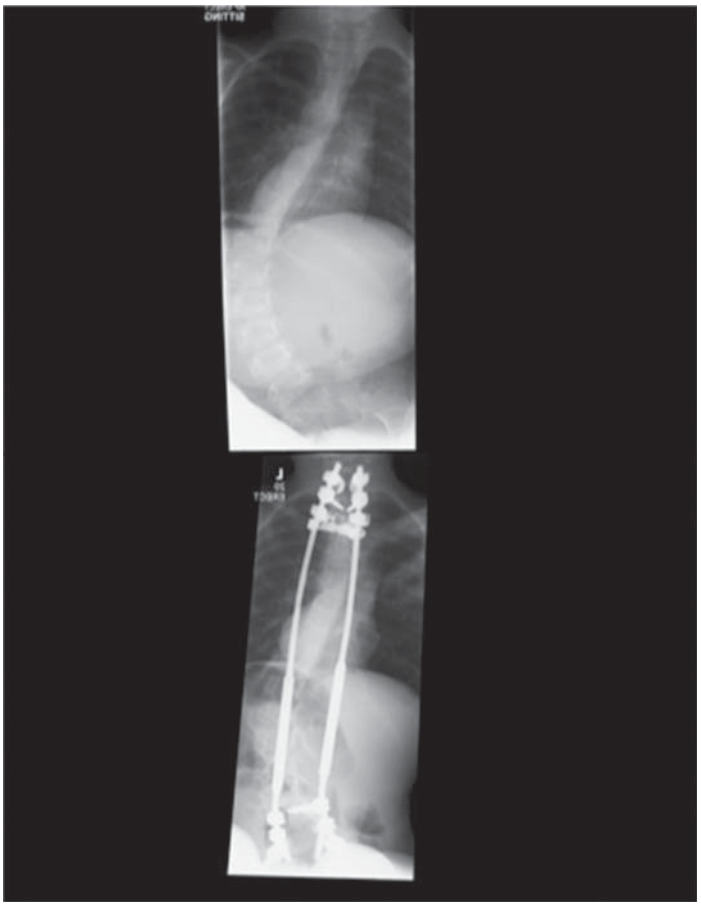

Figure 3. Preoperative and postoperative radiographs of magnetic growth rod.

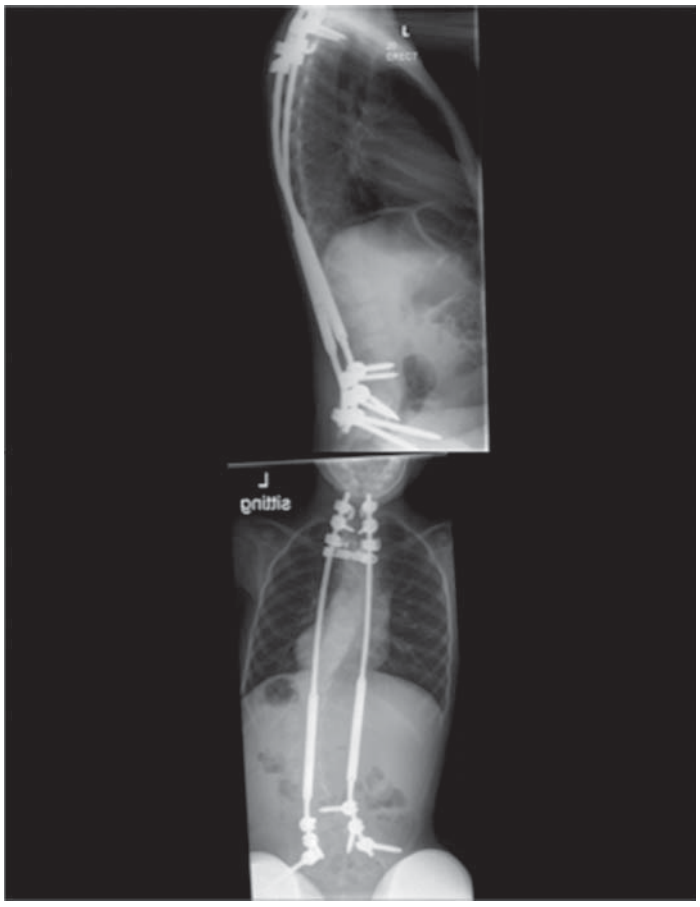

Figure 4. AP and lateral radiographs after first distraction—3 months. AP indicates anteroposterior.

a topic of popular debate and powerful distractions up to 6 months apart still seem to allow the spine to grow. It has recently been suggested that the growth may be a fusion mass that has a thin/weak layer that is fractured at every distraction or a bioactive tissue that grows under distractive forces and animal studies have similarly shown preservation of growth potential in the spine with distractive forces.[15]

Our study has used the Stanojevic equation[9] for percent-predicted values and there is current debate as to which pediatric reference equation is best. These extended models provide more accurate reference ranges for spirometry with transition into adulthood and also incorporate age-related differences in between-subject variability, improving the definition of lower limits of normal.[9]

Although many equations exist, very few include children younger than 5 years.[16] Of the preschool equations that are available, lack of details regarding either the population characteristics or measurement techniques, together with a failure to link results to school age equations, may limit their clinical usefulness.[17] Furthermore, there is debate as to whether expiratory volume in 1 second is an appropriate measurement for very young children. In 2007, Piccioni *et al*[18] published a study including reference equations for FEV in 0.75 seconds ($FEV_{0.75}$) in 3- to 6-year-old children. This may be a more appropriate outcome measure for this age group, because young children have large airways relative to their lung volumes and as such, during forced expiration, emptying may be virtually complete in 1 second. This is one further area where we intend to elaborate with further research but currently the Stanojevic equation remains the "gold standard" in our tertiary referral center.

The natural history for lung function deterioration in the conditions in our series is very variable but longitudinal improvement is not expected in any group. In children with spinal muscular atrophy, there is inevitably a slow decline in lung function as part of their condition during a 10-year period.[19] Similarly, in congenital muscular dystrophy, although a more heterogeneous group, there is an expected decline with increasing age and weight through childhood. Children with neurofibromatosis are likely to show a drop in lung function that commensurate with the changes in spinal curvature but may have normal longitudinal tracking if there is no thoracic spinal or neurological involvement.

We encountered an average increase in percent-predicted FVC of 14%, which is greater than that shown by any other study. There may be multiple factors that are responsible for this. One may be the severe nature of the pulmonary compromise in the cohort of patients studied, in that a modest increase in trunk height may lead to disproportionate increase in percent-predicted FVC. The

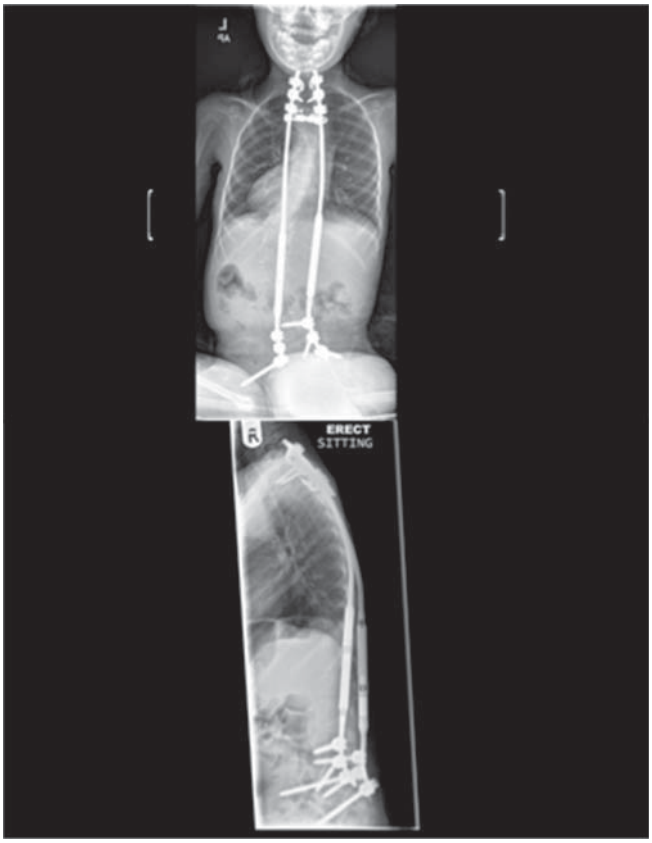

Figure 5. AP and lateral radiographs after second distraction—6 months. AP indicates anteroposterior.

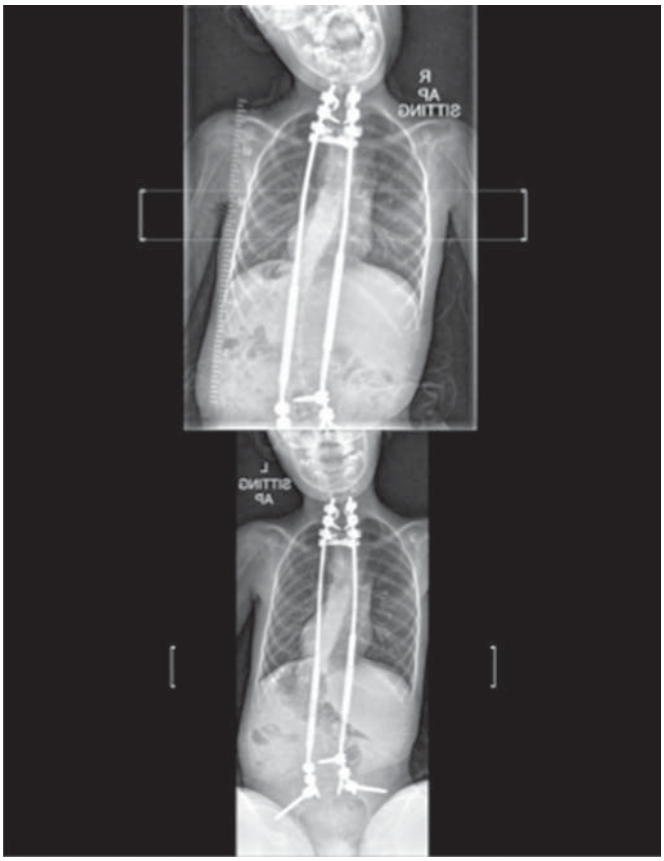

Figure 7. AP radiographs after fifth and sixth distractions (19 and 23 mo). AP indicates anteroposterior.

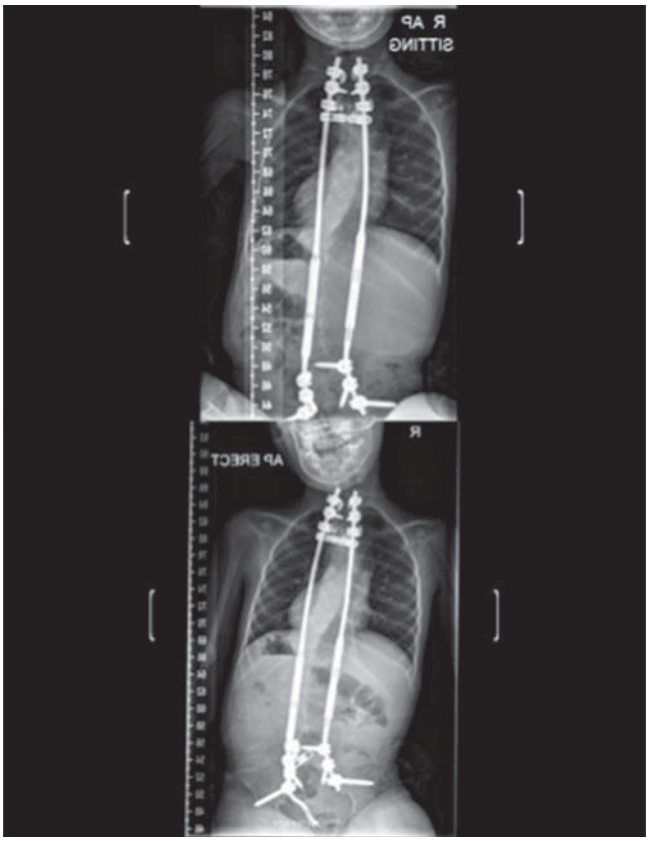

Figure 6. AP radiographs after third and fourth distractions (11 and 16 mo). AP indicates anteroposterior.

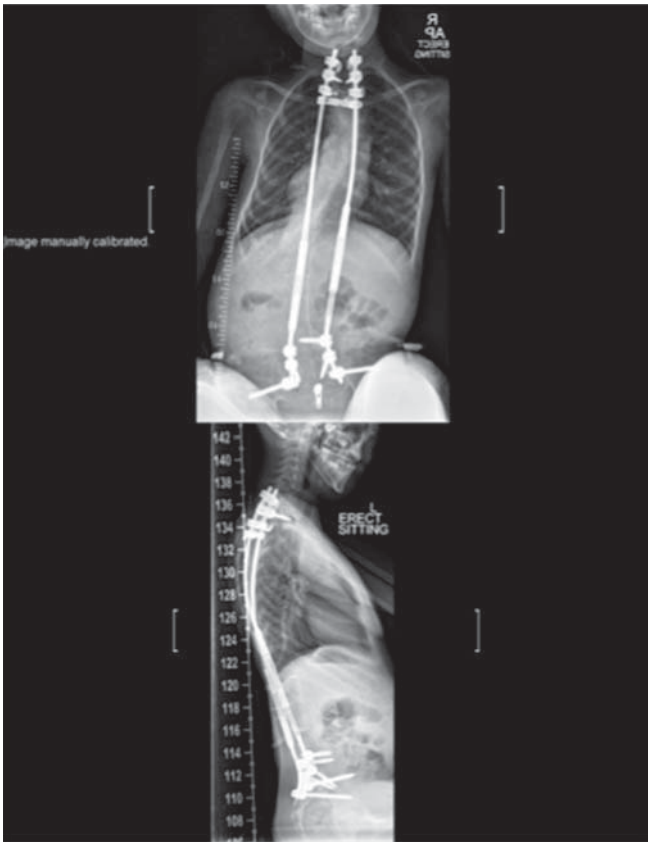

Figure 8. AP and lateral radiographs at seventh distraction—26 months. AP indicates anteroposterior.

MGR allows increase in length of the rod without need for invasive surgery and this may also play a part; however normal development in trunk height, and aging of these children may also have led to gains in percent-predicted FVC, but the small numbers in this study make it difficult to attribute to any specific factor. The structure-respiratory function relationship is complex[20] and needs further research.

CONCLUSION

This study demonstrated that early intervention using posteriorly-based distraction with MGRs in patients with EOS and neuromuscular disease was associated with a significant improvement in postoperative FVC + FEV1 and deformity correction with the added benefits of reduction in repeat anesthetics and open surgery required for lengthening in this high-risk population.

MGR insertion is less invasive, has less complications. In our study we had a single-rod breakage in the motor portion of the rod and this was replaced. This was an early generation rod and this portion was examined and redesigned to minimize breakage. It is also likely to be a more cost-effective solution during the long-term than conventional growth rod treatment, which has been claimed to be a 10-year projected savings of £47,896.[21] This will have to be confirmed in future published studies.

➢ Key Points

- ❑ This study demonstrates that early intervention with MGR lengthening in EOS significantly improves pulmonary function.
- ❑ A significant improvement in deformity correction can be achieved with MGR lengthening.
- ❑ MGRs have the added benefit of reduction in repeat anesthesia and reduction in surgical and psychological distress.

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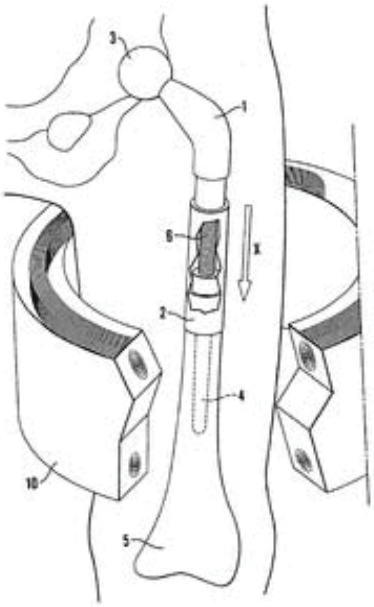
Advances in Surgical Techniques for EOSD

Surgical Patent: 1

Surgical Distraction Device Patent

The surgical distraction device has an application for treating scoliosis, kyphosis, and multi-planar deformities. In this innovative procedure, the components of the device will be anchored to the appropriate vertebrae or to a rib as a proximal anchor, and either to the lumbar vertebrae or sacrum / pelvis as a distal anchor. The distraction force is applied using an externally generated electro-magnetic field to overcome the resistance of the tissue, together with the friction of the linkage.

The surgical distraction device comprises first and second extension rods, each having a firm attachment means at one end to the axial skeleton (vertebra / rib / sacrum). The other end is connected to a motor, driven by a magnetic field. Subjecting the motor to an external strong magnetic source causes the magnet to turn or rotate, and this rotational movement is transformed into linear motion resulting in lengthening or distraction of the two rods by causing the attachment means of one rod to move against the attachment means of the other rod, thus guiding the growth of the spine by acting as an internal brace.



Advances in Surgical Techniques for EOSD

Inventors:

Blunn, Gordon (North Hertfordshire, GB) Perry, John (Hertfordshire, GB) Noordeen, Hilali (London, GB) Meswania, Jay (Hertfordshire, GB) Cobb, Justin (London, GB)

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Chapter 5

CONGENITAL & SYNDROMIC

Advances in Surgical Techniques for EOSD

Paper: 9

Congenital scoliosis could be broadly classified into defects of formation, segmentation, and mixed anomalies. Often they are part of a syndrome (VATER and VACTERL syndromes, and so forth). I undertook an observational study to evaluate the incidence of intra-spinal anomalies and organ or system defects in patients presenting with congenital spinal deformity.

A consecutive series of 126 patients (sixty four males and & sixty two females) were seen by me and all patients were extensively investigated by MRI, echocardiography and renal ultrasound, in addition to a comprehensive clinical and neurological examination. The average age at presentation was 8.7 years (range: two to fifteen years). The most common cause of spinal deformity was a failure in formation (n=67), followed by mixed anomalies (n=33). Intra-spinal anomalies were found in forty seven patients (37 percent). The most common intra-spinal anomaly detected was a tethered cord (n=13), followed by syrinx (n=10). Patients with congenital kyphosis had the highest incidence of intra-spinal anomaly which was also statistically significant ($p<0.05$). Of the patients with hemivertebrae (HV), those with cervical and thoracic HV, and children with mixed and segmentation defects had a higher incidence of intra-spinal anomalies.

Organ defects were found in sixty six patients (55 percent), and the most common organ system affected was the musculoskeletal system (34 percent). Cardiac anomalies were seen in twenty six percent of instances, and urogenital anomalies in twenty one percent of patients. Craniofacial involvement was seen in ten percent of patients.

Congenital spinal deformity: a comprehensive assessment at presentation: Basu PS, Elsebaie H, Noordeen MH. Spine (Philadelphia, PA 1976). 2002 Oct 15; 27(20): 2255-9. PMID: 12394903

Advances in Surgical Techniques for EOSD

Paper: 10

I have reported surgical results of congenital scoliosis treated by short anterior instrumented fusion (one segment above and below the hemivertebra [HV]) in children less than six years of age. There were thirty one patients (fifteen males and sixteen females) with three three HV. The mean age at surgery was two years and ten months (range: eight months to one year and nine months). There were twenty seven fully segmented HV, two semi-segmented HV, and two fully segmented HV at two separate levels.

My preference was to partially preserve the HV, in an effort not to compromise the blood supply of the spinal cord, as any ischemic insult to the neural tissue would be irreversible with permanent neurodeficit. Third generation anterior instrumentation with mono-axial screws was used, following anterior column reconstruction by strut grafts. The mean follow-up was 6.1 years. The Cobb angle of the major or structural curve improved from forty one to seventeen degrees with segmental improvement in scoliosis by sixty two percent. The segmental kyphosis improved from sixteen to eleven. There were two wound infections, and no neurological complications. One patient had an intra-operative fracture of the vertebral body that necessitated spanning an additional level. Screw loosening without any symptoms was also observed in one patient. The presence of thoraco-lumbar kyphosis, despite anterior surgery warranted second stage posterior instrumented correction or fusion.

Short anterior instrumented fusion and posterior convex non-instrumented fusion of hemivertebra for congenital scoliosis in very young children: Garrido E, Tome-Bermejo F, Tucker SK, Noordeen HN, Morley TR. Eur Spine J. 2008 Nov;17(11):1507-14. Epub 2008 Sep 27. PMID: 18820956

Advances in Surgical Techniques for EOSD

Paper: 11

I devised a novel technique for addressing severe congenital spinal deformities secondary to hemivertebra, and reported clinic-radiological results of the same in twelve very young children aged less than four years. The procedure included combined anterior + posterior surgery (partial hemivertebra [HV] resection by anterior approach with anterior instrumentation and posterior uninstrumented fusion).

There were eight females and four males. The mean age at surgery was two years and seven months (range: one year and nine months to three years and one month), and the mean follow-up was three years and one month (range: two years to four years and five months). There were nine fully segmented HV, and three semi-segmented. The Cobb angle of scoliosis improved from 48.3 to 17.2 degrees. The kyphosis improved from 23.2 to 11.7 degrees.

The release of a concave tether facilitated the correction safely, and all patients were subjected to a wake-up test intra-operatively at the end of the correction or instrumentation. They subsequently wore a brace for 12 weeks. Though conventionally it was reported in literature for wake-up test to be less reliable in children aged less than 7 years, I had consistent reliability in all 12 children in this series. Additional neuromonitoring was used in only 4 patients. There was one superficial wound infection, and no neurological complications. Preserving part of the hemivertebra also protects the spinal cord from ischemic injury, which could be devastating and cause a permanent neurodeficit.

Anterior Instrumentation and Correction of Congenital Spinal Deformities Under Age of Four Without Hemivertebrectomy - A New Alternative: Elsebaie HB, Kaptan W, El Miligui Y, El Masry MA, Salaheldine M, Noordeen HM, Akbarnia BA. Spine (Philadelphia, PA 1976). 2010 Feb 26. [E-pub ahead of print] PMID: 20195198

Advances in Surgical Techniques for EOSD

Paper: 12

The posterior quadrant hemivertebra (PQ-HV) or postero-lateral quadrant (PLQ-HV) may cause kyphosis or kyphoscoliotic deformity. I published the surgical results of fifteen patients under the age of three years, who were operated by me for congenital kyphosis. My technique included stand-alone anterior instrumented correction and fusion. Bony rib harvested from costectomy and performed for surgical access was used to create an anterior column support i.e. strut graft). Instrumented posterior fusion was rarely performed. There were ten males and five females and the mean age at surgery was twenty two months (range: eight to three three months). The most common location of deformity was the thoraco-lumbar region. 13 patients had PLQ-HV, while 2 had PQ-HV.

The mean follow-up was 6.8 years. The mean improvement in kyphosis correction was forty three percent, and in scoliosis correction was sixty four percent. The mean blood loss was fifteen percent of estimated blood volume (EBV). A second stage posterior surgery was reserved for residual unacceptable deformity, in the form of unilateral instrumentation with fusion on the convex side.

I also observed precarious vascularity in congenital spinal deformities, with surgery causing a compromise in cord perfusion, manifesting as an irreversible neurodeficit. I therefore advocated against a complete HV excision, especially at the apex of the deformity.

The surgical treatment of congenital kyphosis: Noordeen MH, Garrido E, Tucker SK, Elsebaie HB. Spine (Phila Pa 1976). 2009 Aug 1;34(17):1808-14. PMID: 19644332

Advances in Surgical Techniques for EOSD

Paper: 13

Mucopolysaccharidosis (MPS) are a group of inherited metabolic disorders affecting the musculoskeletal system and causing skeletal dysplasia. Most of them are acquired by autosomal recessive transmission, though Hunter's syndrome is transmitted by an X-linked recessive. Thoracolumbar gibbus is a common deformity, owing to defective anterior vertebral elements that are flattened or beaked. The intervertebral discs are large or bulging into the canal, compressing the thecal sac.

I have published the surgical results of seven patients of MPS with thoraco-lumbar gibbus treated with anterior surgery, or decompression and instrumented fusion. All patients were between four and ten years at the time of surgery, and the minimum follow-up was two years. The surgery involved a thoraco-abdominal approach through the bed of the left ninth or tenth rib, and a peripheral release of the diaphragm. Four patients had a neurodeficit. All of them improved neurologically post-operatively, and three patients returned to their previous level of activity. The key advantages of anterior surgery were:

i) Reduced incidence of infection by anterior approaches

ii) Larger bed or area for sound arthrodesis

iii) The vertebral body is stronger anteriorly, as compared to posterior elements

iv) Complete discectomy with good decompression of thecal sac.

Anterior instrumented fusion for thoracolumbar kyphosis in mucopolysaccharidosis. Dalvie SS, Noordeen MH, Vellodi A. Spine (Phila Pa 1976). 2001 Dec 1;26(23):E539-41. PMID: 11725253

Advances in Surgical Techniques for EOSD

Paper: 14

Jarcho Levin syndrome is one of the skeletal dysplasias characterized by congenital scoliosis with fused vertebra or ribs, presenting with a short or small thorax, which causes thoracic insufficiency syndrome (TIS). It could be classified into two broad types:

- Spondylocostal dysostosis (SCD)
- Spondylothoracic dysostosis (STD)

I have reported clinical manifestation of SCD in one of the largest series reported in English medical literature (there were thirteen patients; seven males and six females). Congenital scoliosis was seen in ten of these thirteen patients, whereas the prevalence of SCD in the general population is 1/400,000. The mode of inheritance could be either autosomal dominant or recessive. The follow-up duration varied from two to six years. The survival rate in this series was 100 percent. Only one patient had to be operated on for a rapid progression of congenital scoliosis, and that was treated with a posterior spinal fusion. My recommendations were:

- Chest physiotherapy to be mandatory for pulmonary toilet and to prevent infections
- Early prenatal USG diagnosis is possible, and genetic counseling is important for parents to make informed decisions.
- Surgery should be done to address severe or progressive scoliosis and TIS.
- Chest reconstruction increases the space available for the lungs (SAL) and lung growth.

Spondylocostal dysostosis: thirteen new cases treated by conservative and surgical means. Teli M, Hosalkar H, Gill I, Noordeen H: Spine (Philadelphia, PA 1976). 2004 Jul 1; 29(13): 1447-51. Review. PMID: 15223937

SPINE Volume 27, Number 20, pp 2255–2259

Congenital Spinal Deformity

A Comprehensive Assessment at Presentation

Partha S Basu, FRCS Orth,* Hazem Elsebaie, FRCS,† and MHH Noordeen, ChM, FRCS‡

Study Design. A series of 126 consecutive patients with congenital spinal deformity is presented.

Objective. To assess the incidence of intraspinal anomaly and other organic defects associated with different types of spine deformity at presentation.

Summary of Background Data. A high incidence of intraspinal abnormalities and other organ defects is reported in relation to congenital spine deformity. The prevalence of these problems with different types of deformities is to be determined.

Methods. All patients had MRI, echocardiography, renal ultrasound, and a thorough clinical assessment.

Results. Intraspinal abnormalities were found in 47 patients (37%). These abnormalities were significantly more common in patients with congenital kyphosis ($P = 0.0048$), and in those with scoliosis resulting from mixed and segmentation defects. Scoliosis patients with cervical and thoracic hemivertebrae had significantly more intraspinal abnormalities ($P = 0.0253$) than those with lumbar hemivertebrae. In 64 (55%) patients other organic defects were found. These defects were more common in patients with congenital scoliosis resulting from mixed defects ($P = 0.002$). Cardiac defects were detected in 26% and urogenital anomalies in 21% of the patients.

Conclusions. Magnetic resonance imaging and echocardiography should be an essential part in the evaluation of patients with congenital spinal deformity, and special attention should be paid to patients with segmentation abnormalities, mixed defects, and kyphosis. [Key words: congenital spinal deformity, congenital heart disease, hemivertebra, intraspinal abnormality, segmentation defects] **Spine 2002;27:2255–2259**

Congenital scoliosis often is associated with intraspinal abnormalities and other organ defects. The embryonic development of vertebrae is closely related with that of the spinal cord and the organs of the mesoderm.[1,3,6,9,12] The intraspinal anomalies can cause progressive neural loss with growth and curve progression. In addition, they greatly increase the risk of neurologic injury during surgical correction of the deformity.

From the * Orthopaedic Specialist Registrar, Freeman Hospital, Newcastle-Upon-Tyne, UK, the †Cairo University Hospital, Cairo, Egypt, and the ‡Hospital for Sick Children, London, United Kingdom.
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McMaster[9] reported intraspinal abnormality in 18% of 251 patients with myelography. Magnetic resonance imaging (MRI) is noninvasive and more sensitive in detecting the intraspinal abnormalities. Using MRI, Bradford et al[3] reported that 38% of the 42 patients in their study had an intraspinal anomaly.

Using intravenous urography (IVU), MacEwen et al[7] found genitourinary abnormality in 20% of patients with congenital scoliosis. Guerrero et al[4] found an incidence of genitourinary abnormalities of 34% using ultrasound and IVU. The exact incidence of congenital heart disease associated with congenital scoliosis has not been reported. Hensinger et al[5] found a 14% incidence of congenital heart disease among patients with Klippel–Feil syndrome.

The authors have retrospectively reviewed their series of 126 consecutive patients with congenital spinal deformity. Their aims were to assess the overall incidence of intraspinal anomaly and other organ defects; to study the associations between the different types of deformity, the intraspinal abnormalities, and other organ defects; and to develop a clinical guideline for investigating these difficult patients.

Materials and Methods

Between July 1995 and June 2000, 126 patients (62 boys and 64 girls) with congenital scoliosis were seen at the authors' hospital. Of these, 27 patients were referred by physicians from other specialities to the authors' hospital. The remaining 99 patients were referred from other hospitals. All these patients underwent a full clinical examination, plain radiograph, neural axis MRI (from the brain stem to the sacrum), renal ultrasound, echocardiography, and assessment by pediatricians. Almost all of them underwent MRI while under sedation or general anesthesia. The indications for performing MRI with the patient under general anesthesia were failure to obtain a scan under sedation, age younger than 5 years (because of difficulty achieving adequate sedation), and a brain MRI for other indications.

All these patients currently are being followed up until maturity. Their average age at presentation was 8.7 years (range, 2 weeks to 15 years). The congenital spine deformity was classified as congenital scoliosis resulting from failure of formation, failure of segmentation, mixed and complex lesions, or congenital kyphosis.[12] Progression of the spinal deformity, defined as an increase in the curvature by at least 5°, was noted in 78 patients (62%). Of these patients with progressive deformity, 71 underwent surgery, and 5 are waiting to have surgery. One patient refused surgery, and another child had multiple congenital anomaly with severe respiratory insufficiency, rendering him unsuitable for surgery. Although plain radiograph and MRI were available for all 126 patients, parts of patient record were missing for 9 patients. The remaining 117 patients were

Table 1. Ultraspinal Anomalies

Intraspinal Anomalies	Number
Tethered Cord	16
Syrinx	13
Thickened & Fatty Filum	11
Low Conus	10
Diastematomyelia	8
Intradural Mass/Lipoma	5
Extradural Mass	2
Chiari Malformation	2
Arachnoid Cyst	1
Dandy-Walker Malformation	1

(Some patients had multiple abnormalities).

assessed for the presence of other congenital defects and syndromes. The intraspinal anomalies and other organic defects are not likely be normally distributed. The Mann–Whitney *U* test, the Kruskall–Wallis test, and the χ^2 test (as appropriate for the data type) were used for these nonparametric variables.

■ Results

In the current series of 126 patients, 110 were found to have congenital scoliosis. Of these, 67 had failures of formation: hemivertebra (n = 65), wedge vertebra (n = 1), butterfly vertebra (n = 1). Failure of segmentation was found in 10 patients, and 33 patients had mixed and complex lesions. Among 16 patients with congenital kyphosis, 4 had congenital dislocation of the spine.

Neurocutaneous stigmas of intraspinal lesions were found in 24 patients. The findings in these patients showed cutaneous stigmas overlying the spine (n = 5), bladder symptoms (n = 4), paraesthesia in one leg (n = 3), foot deformity (n = 4), obvious wasting of one leg (n = 4), asymmetrical abdominal reflexes (n = 1), and abnormality of posterior column sensation (n = 2). Of these 23 patients, 5 had a normal MRI.

Intraspinal abnormality was found in 37% (n = 47) of these 126 patients (Table 1). Among the scoliosis patients, intraspinal anomaly was present in 29% of the patients with failure of formation, in 40% of the patients with failure of segmentation, and in 40% of the patients with mixed lesions. Of the patients with congenital kyphosis, 56% had intraspinal anomaly ($P = 0.0048$) (Table 2). In the patients with scoliosis who had failures of formation, 37 were thoracic, 9 were cervical, and 21 were lumbar. Of the 46 patients with cervical and thoracic hemivertebrae, 15 (33%) had intraspinal anomaly. On the other hand, 4 of 21 patients (20%) with lumbar hemivertebrae had an abnormal MRI ($P = 0.0253$).

Table 2. Incidence of Different Types of Spinal Deformities

Anomaly	Total	Intraspinal
Failure of Formation	67	20 (30%)
Failure of Segmentation	10	4 (40%)
Mixed Defects	33	13 (40%)
Congenital Kyphosis	16	9 (56%)

Table 3a. Associated Syndromes

Syndromes	Number
VATER	13
Goldenhar	5
Poland's	3
Noonan	1
CHARGE	1
Melanocytic Naevus	1

Sixteen patients had congenital kyphosis. Of the 4 patients with congenital dislocation of the spine, 3 had intraspinal abnormality (75%), as did 4 of 10 patients with failures of formation (posterior or posterolateral hemivertebrae) (40%). Two patients with kyphosis resulting from anterior unsegmented bar both had intraspinal anomaly.

Of 64 patients (55%) who had other congenital defects, 24 patients had a constellation of abnormalities as part of a syndrome. The remaining 40 patients had an isolated congenital abnormality affecting one or more organ systems (Table 3). Of the patients with congenital kyphosis, 50% had other organ defects, as did 47% patients with scoliosis caused by failures of formation. Among patients who had scoliosis with segmentation defect, 33% had associated organ defects, as did 72% patients with mixed defects (Table 4). The patients with mixed defects had a significantly higher incidence of other organ defects ($P = 0.002$). Intraspinal abnormalities were present in 28 patients (44%) with other organ defects and 19 patients (36%) without other organ defects ($P = 0.103$).

The incidence of congenital heart disease in patients with congenital spine deformity was 26% (Table 5a). Table 5b shows the relative incidence of congenital heart disease among the different groups. The proportion of congenital heart disease was higher in those with congenital kyphosis (33%) and in those with scoliosis caused by mixed defects (37%) ($P = 0.058$). Nineteen patients with thoracic spinal deformity (28%) had congenital heart disease, as did 10 patients with lumbar spinal deformity (21%) and 2 patients with cervical spinal deformity (15%) ($P = 0.661$). Of the 31 patients with congenital heart disease, 11 (33%) had surgical treatment, and a further 4 needed medical therapy. The remaining 16 patients (50%) did not need active treatment, but were under observation. For 21 of these patients, the diagnosis of congenital heart disease had been made before their

Table 3b. Associated Isolated Congenital Defects

Defects	Number
Musculoskeletal	18
Cardiac	15
Craniofacial	6
Tracheo-oesophageal Fistula	6
Ano-Rectal	5
Renal	5

Table 4. The Incidence of Other Organic Defects in Different Spinal Deformities

	Organic Defects	Normal	Unknown
Failure of Formation	30 (47%)	34	3
Failure of Segmentation	3 (37%)	6	1
Mixed Defects	22 (73%)	8	3
Congenital Kyphosis	7 (50%)	7	2

referral to us. The remaining 10 patients were found to have cardiac defects on echocardiography and clinical examination as part of the authors' protocol. These patients were referred subsequently to the specialist physicians. Two of these 10 patients were receiving medical treatment, whereas active treatment was not indicated in the remaining 8 patients. All the patients were under regular observation by the physicians.

The spinal deformity in nine patients with congenital heart disease did not require surgery. Surgical correction of the spinal deformity was performed in the remaining 22 patients with congenital heart disease. Among these 22 patients, 9 had cardiac surgery and 2 were receiving medical treatment before the deformity correction. All the patients with cardiac defects were admitted a few days before surgery to ensure preoperative optimization by cardiologists. No change of the surgical or monitoring protocol was necessary.

The incidence of urogenital abnormality was 21% in the current series of patients (Table 6). Among the 25 patients with urogenital anomalies, 6 (25%) needed surgery and 2 had abnormal renal function requiring medical therapy, including dialysis. The remaining 17 patients had abnormalities that did not affect renal function or did not require treatment. Musculoskeletal abnormalities were present in 34% and craniofacial abnormalities in 10% of the patients.

■ Discussion

Four reports have been published on the association of intraspinal abnormality with congenital spine deformity.[2,3,9,11] McMaster[9] reported an incidence of 18.3%. In his series, 106 of 251 patients underwent myelography, and diastematomyelia was diagnosed in 8 patients on the basis of midline bony spur on plain radiograph. In his series, because of the toxic effects from oil-based contrast, myelography was performed only on patients with clear neurologic signs in the lower limbs and before surgical correction. Subsequently, Blake et al[2] performed myelography with water-soluble contrast on a series of 108 patients with congenital spinal deformity. They reported an incidence of 58%. Such a high incidence probably results from too broad a radiologic criterion for assessing tethered cord and "tight" filum.

Bradford et al[3] reported an incidence of 38%, using MRI, in a series of 42 patients. Prahinski et al[11] found a 30% incidence of intraspinal anomaly in a series of 30 patients. Both studies, using MRI, included small numbers of cases, and consequently could not investigate the association between intraspinal anomaly and different types of spinal deformities. Bradford et al[3] noted the higher association of intraspinal anomalies with the two congenital kyphosis cases.

The current study conclusively showed that the incidence of intraspinal abnormality is higher among patients with congenital kyphosis than among those with congenital scoliosis. Among the patients with scoliosis, the incidence of intraspinal anomaly in segmentation and mixed defects was higher than among patients with failures of formation. Both McMaster[8–10] and Bradford et al[3] noted the association between diastematomyelia and unsegmented bar with contralateral hemivertebra. In the current study, unsegmented bar with contralateral hemivertebra was present in eight patients: five with diastematomyelia and one with an associated syrinx. The cervical and thoracic hemivertebrae had a significantly higher incidence of intraspinal abnormalities than lumbar hemivertebrae. This has not been described previously. The relative incidence of the intraspinal abnormalities was similar to that in the study of Bradford et al,[3] with teth-

Table 5a. Congenital Heart Disease

Defect	Number
Ventricular Septal Defects	14
Atrial Septal Defects	5
Patent Ductus Arteriosus	3
Fallot	3
Transposition of Great Arteries	1
Pulmonary Stenosis	1
Sick Sinus Syndrome	1

Table 5b. The Incidence of Heart Defects in the Different Groups

Failures of Formation	18%
Failures of Segmentation	10%
Mixed Defects	37%
Congenital kyphosis	18%

Table 6. Urogenital Abnormalities

Defect	Number
Renal Hypoplasia	5
Horseshoe Kidney	3
Single Kidney	3
Congenital Megaureter	2
Ectopic Kidney (pelvic)	2
Hypospadius	2
Pelviureteric Junctional Obstruction	2
Posterior Urethral Valve	2
Cloacal Anomaly	1
Epispadius	1
Exstrophy of the Bladder	1
Hydronephrosis	1
Undescended Testis	1

ered cord the most common pathology and syrinx the second most common pathology. In line with the studies of McMaster[8–10] and Bradford et al,[3] the current authors found that the presence of neurocutaneous stigmas are unreliable indicators of intraspinal abnormality.

The performance of MRI in young children involves administration of sedation or general anesthesia, with the attendant risks of respiratory complications. As a result, a selective approach probably is wise. The current authors believe that an MRI scan must be obtained in all cases before surgical correction of the spinal deformity, in cases with established or developing neurologic signs, and probably also in cases with progressive deformity, in which surgery is to be considered sooner. For children 5 years of age or older, they recommend MRI scan with the patient under sedation if the deformity is progressive or neurologic signs develop. In children younger than 5 years, MRI with the patient under general anesthesia is to be considered if surgery is imminent or neurologic signs develop.

In this study, congenital heart disease was found to be associated with congenital spinal deformity in one fourth of the patients. This has not been described previously. Two thirds of these patients had already received the diagnosis of congenital heart disease before referral to the authors. The authors' assessment protocol was able to detect congenital heart defects in another one third of the previously undetected patients. A proportion of these hitherto undetected patients had already needed medical therapy, and probably would require surgery for the cardiac condition in the future. This underscores the importance of a systematic clinical cardiac assessment using echocardiography for these patients.

The authors believe that all patients in whom surgery is planned for correction of congenital spinal deformity should have echocardiography as part of preoperative workup. In addition, patients with congenital scoliosis resulting from mixed bony defects and those with congenital kyphosis should have a routine echocardiogram because of the higher risk for congenital heart disease. Patients with congenital scoliosis caused by failures of formation and segmentation, who are not surgical candidates, should have clinical screening for heart disease and an echocardiography if they are clinically abnormal or there is suspicion of congenital heart disease.

The incidence of urogenital abnormality in patients with congenital spinal deformity ranges from 22% to 34%,[1,4,5,7] and was 21% in the current study. The authors found that most urogenital anomalies are benign in nature and need no treatment, as in the study of MacEwen et al.[7] The patients with mixed and segmentation defects had a consistently higher incidence of defects in other organs, probably indicating a more widespread embryonic injury. In the current study, no association between the presence of intraspinal anomaly and other organ defects was found.

■ Conclusion

Intraspinal anomalies were associated with 37% the congenital spinal deformity cases, according to MRI. Such anomalies were significantly more common in the patients with congenital kyphosis than in the patients with congenital scoliosis. Among the patients with scoliosis, those with mixed defects had a higher incidence of intraspinal abnormality. Also, in the patients with scoliosis, the hemivertebra located higher up in the spine (cervical and thoracic) had higher risk of intraspinal abnormalities.

Overall, 55% of the patients had other congenital defects, and these were significantly more common in the patients with scoliosis caused by mixed defects. Congenital heart disease was found in 26% of the patients. Half of the patients with heart defects needed treatment, and the remainder were under observation. Urogenital abnormalities were noted in 21% of the patients. Most were benign, but one third of them needed active treatment.

All patients with congenital spinal deformity should undergo MRI and echocardiography before any surgical correction. In addition, children with neurologic signs and those older than 5 years with progressive deformity should have MRI. Patients with scoliosis caused by mixed defects and those with congenital kyphosis should have echocardiography. Patients with scoliosis resulting from failures of formation and segmentation should have clinical screening for cardiac defects.

■ Key Points

- Intraspinal anomalies were found in 37% of the patients.
- Mixed and segmentation defects and thoracic hemivertebrae were common in patients with congenital kyphosis.
- Other organic defects were found in 55% of the patients.
- Congenital heart disease was present in 26% of the patients.
- Magnetic resonance imaging and echocardiography should be part of routine assessment.

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Address reprint requests to

Partha S. Basu, FRCS
5 Sandringhan Rd.
Gosforth
Newcastle Upon Tyne
United Kingdom

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ORIGINAL ARTICLE

Short anterior instrumented fusion and posterior convex non-instrumented fusion of hemivertebra for congenital scoliosis in very young children

E. Garrido · F. Tome-Bermejo · S. K. Tucker · H. N. N. Noordeen · T. R. Morley

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Abstract A variety of treatments has been described in the literature for the treatment of HV. We report the results of early surgical anterior instrumented fusion with partial preservation of the HV and posterior non-instrumented fusion in the treatment of progressive congenital scoliosis in children below the age of six. Between 1996 and 2006, 31 consecutive patients with 33 lateral HV and progressive scoliosis underwent short segment fusions. Mean age at surgery was 2 years and 10 months. Mean follow-up period was 6.1 years. The major scoliotic curve improved from 41° preoperatively to 17° on follow-up. Preoperative segmental Cobb angle averaging 39° was corrected to 15° after surgery, being 15° at the last follow-up (62% of improvement). Compensatory cranial and caudal curves corrected by 47 and 45%, respectively. The angle of segmental kyphosis averaged 16° before surgery, 11° after surgery, and 11° at follow-up. There were two wound infections requiring surgical debridment, one intraoperative fracture of the vertebral body and one case lost correction due to implant failure. All went on to stable bony union. There were no neurological complications. Early diagnosis and early and aggressive surgical treatment are mandatory for a successful treatment of congenital scoliosis and prevention of the development of secondary compensatory deformities. Anterior instrumentation is a safe and effective technique capable of transmitting a high amount of convex compression allowing short segment fusion, which is of great importance in the growing spine.

Keywords Hemivertebra · Congenital scoliosis · Fully segmented hemivertebra · Congenital spine deformity · Anterior instrumented fusion

Introduction

The natural history of congenital scoliosis has been well documented [6, 7, 11, 14, 21, 36, 39]. Foetal ultrasound scan has made antenatal diagnosis of hemivertebra (HV) possible [32]. Children with HV are, therefore, referred much earlier before severe local and compensatory curves develop. Surgical treatment of patients with congenital scoliosis carries a risk of neurological injury greater than that in patients with idiopathic spinal deformities [12, 19, 38]. Early treatment of progressive deformities helps in minimising the risks of surgery and allows better correction and prevents the development of structural and compensatory curves [7, 21, 38].

The current study describes the result of anterior partial resection and short-segmented instrumented fusion of hemivertebra in a series of 31 consecutive patients below the age of 6 years with progressive congenital scoliosis due to a lateral hemivertebra.

To date, there are no published reports that examine the safety and efficacy of anterior instrumented fusion for the treatment of congenital scoliosis due to HV in such a young patient population.

Study conducted at the Great Ormond Street Hospital for Children and the Royal National Orthopaedic Hospital, Stanmore, London, UK.

E. Garrido (✉) · F. Tome-Bermejo · S. K. Tucker · H. N. N. Noordeen
Department of Spinal Surgery, Great Ormond Street Hospital for Children, Great Ormond Street, London WC1N 3JH, UK
e-mail: Garrie@gosh.nhs.uk; macgarrido@gmail.com

S. K. Tucker · H. N. N. Noordeen · T. R. Morley
Spinal Deformity Unit, Royal National Orthopaedic Hospital, Stanmore, London, UK

Materials and methods

From 1996 to 2006, 31 consecutive patients with progressive congenital scoliosis due to 33 lateral hemivertebrae underwent short segment anterior instrumented fusion using a single solid rod construct with simultaneous convex non-instrumented posterior fusion corresponding to the levels of the anterior surgery.

Of the 31 patients in the study, 16 were girls and 15 boys. Their mean age at the time of surgery was 2 years and 10 months (range, 8 months to 5 years and 9 months). The mean follow-up period was 6.1 years (range, 2 to 11.3 years).

In total, 19 HV were located in the thoracolumbar junction (T10–L2), 11 in the lumbar spine (L3–L4) and 3 in the thoracic spine.

There were 27 children with single fully segmented HV, 2 semisegmented midlumbar HV, and 2 children had two ipsilateral fully segmented HV.

Only the vertebra above and below the hemivertebra were instrumented in 26 fusions. An additional level was included in seven cases: in three cases with thoracic HV due to the higher rigidity of the spine, in one case were the vertebral body fractured during screw insertion and in three cases with a higher magnitude curve.

Webb–Morley Spine System (Biomet-Merck Ltd, Bridgend, UK) was used on five patients and Downsize Synergy^{TM} Spinal System, (Parsippany, NJ, USA) on 26 patients.

Children were evaluated clinically and by retrospective chart and radiographic review. Spinal cord anomalies were present in four cases (13%). Prior to deformity correction one patient underwent Arnold Chiari decompression and another patient untethering of a lipomyelocele. The remaining two patients had a cervical syrinx and a thoracolumbar syrinx. Other congenital vertebral anomalies were present in 9 patients (29%). Congenital heart disease was found in 6 cases (19%), genitourinary anomalies in 5 (15%) and gastrointestinal abnormalities in 3 (10%) patients. There was one Goldenhar syndrome.

Preoperative radiological imaging included anterior–posterior and lateral radiographs of the whole spine, MRI, echocardiogram and renal ultrasound.

Standing posteroanterior and lateral radiographs of the full spine were analysed and angles measured using the Cobb method. The angles of total main scoliosis, segmental scoliosis, compensatory cranial and caudal curves, segmental and total kyphosis, sagital alignment of the thoracolumbar junction and lordosis were measured and recorded. The total angle of kyphosis was measured from T4 to T12. Lordosis was measured from L1 to L5. The thoracolumbar junction was measured from T9 to L2. MediCAD software (Hectec GmbH, Niederviehbach, Germany) was used to make all the measurements.

Curve correction was statistically analysed using paired Student's t test.

Surgical technique

All the patients are placed in the lateral decubitus position with the hemivertebra facing upward. Spinal cord monitoring is used for all patients. Care must be taken to keep the posterior spine exposed. A transthoracic or thoracoabdominal approach is chosen depending on the level of the HV.

Once the vertebral column is visualised, intraoperative radiographs are taken to confirm levels. Segmental vessels are preserved if possible. After exposure of the HV (Fig. 1), the disc material proximal and distal to the HV including the concave part of the disc is removed. Further resection of the HV is reserved until later stage to minimise bleeding.

Now a posterior midline incision is made. The posterior elements on the convexity are carefully exposed. In the first year to 2 years of life, cartilage forms a significant part of the posterior elements and correction of the deformity can be obtained without a formal posterior release. In older children, facectomies of the HV are performed. Convex onlay morselised autogenous rib graft augmented with ProOsteon (Interpore Cross International, Irvine, CA) is applied after decortication of the laminas and spinous process corresponding to the levels of the anterior instrumentation.

If there is a failure of segmentation of the posterior elements, an osteotomy of the lamina can be done to further improve mobility at the segment.

The superior and inferior endplates of the HV are now removed to bleeding bone. The posterior body of the HV is partially decancellated. The body of the hemivertebra is

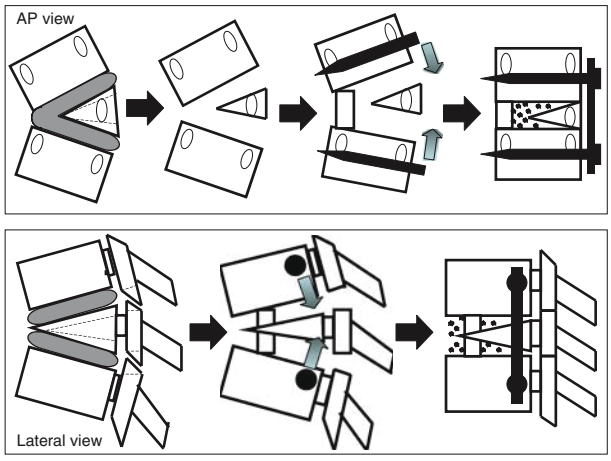

Fig. 1 Anterior–posterior and lateral sequence of the operative technique

resected to a stump corresponding to the pedicle location to achieve union with the adjacent endplates. Partial preservation of the body of the HV prevents nerve root impingement and minimises interference with local blood supply to the spinal cord.

At this point, anterior instrumentation begins. Bicortical transbody monoaxial screws are placed posteriorly and parallel to the endplates above and below the HV. Bicortical purchase of all screws is documented by manual palpation of the contralateral side of the spine. Washers were used in 6 of the 32 patients. A single straight rod is then engaged and the inferior screw is locked. Manual posterior pressure is applied to the spine to open the disc space and rib strut graft is inserted into the anterolateral portion of the concave disc space. Final position of the struts can be adjusted with a small bone punch. The intact posterior longitudinal ligament prevents over distraction during this manoeuvre.

Then careful intersegmental compression is applied. The strut graft prevents inducing kyphosis during anterior compression and serves as a fulcrum to obtain coronal correction. The disc space is further filled with morselized rib autograft.

Intraoperative radiographs are obtained before closure of the wound.

Patients were braced in a TLSO for 6 months postoperatively.

Results

The average operating time in this procedure was 133 min (range 80–210 min). The average blood loss 169 mL (range 50–550 mL), equivalent to a mean external blood volume loss of 10% (range 5–24%).

Segmental scoliosis (Table 1) was corrected from 39° (range 29°–56°) before surgery, to 15° (range 2°–27°) after surgery, and 15° (range 3°–32°) on the average, at the last follow-up. The major curve was 41° (range 29°–60°), 19° (range 4°–40°), and 17° (range 6°–35°), respectively. This represents a 62% of improvement for the segmental curve, and a 59% of improvement for the total main scoliosis curve at last follow-up.

About 47% of correction was obtained in the upper and 45% correction in the lower compensatory curves at the last follow-up.

Table 1 Correction of main and segmental scoliosis curves, compensatory curves, kyphosis and sagittal measurements

	Preoperative	Postoperative	Follow-up	Paired Student's test at follow-up
Total main (mayor) curve (°)	41	19	17	$P < 0.001$
Segmental curve (°)	39	15	15	$P < 0.001$
Cranial curve (°)	15	9	8	$P < 0.001$
Caudal curve (°)	20	12	11	$P < 0.001$
Segmental kyphosis (°)				
Thoracic HV	11	10	9	
Thoracolumbar HV	20	16	13	
Lumbar HV	10	6	8	
Total number HV	16	11	11	
Total kyphosis T4–T12 (°)				
Thoracic HV	17	15	35	
Thoracolumbar HV	18	21	25	
Lumbar HV	11	12	22	
Total number HV	15	17	25	
Thoracolumbar Junction T9–L2 (°) (+ Kyphosis) (− Lordosis)				
Thoracic HV	7	3	2	
Thoracolumbar HV	16	11	10	
Lumbar HV	4	4	−2	
Total number HV	12	9	6	
Total Lordosis L1–L5 (°)				
Thoracic HV	27	28	44	
Thoracolumbar HV	28	30	43	
Lumbar HV	19	21	33	
Total number HV	26	28	40	

In the sagittal plane, the segmental kyphosis averaged 16° (range 3°–47°) before surgery, 11° (range 2°–29°) postoperatively and 11° (range 3°–29°) at last follow-up.

Follow-up focal kyphosis of four patients with a segmental kyphosis of more than 20° preoperatively (average 31°, range 22°–46°) had a residual average segmental kyphosis of 24.5° (range 21°–29°) on the latest follow-up. The total thoracic kyphosis T4–T12 averaged 25° (range 4°–44°) at the latest follow-up. Lumbar lordosis L1–L5 was 40° (range 15°–58°) at the latest follow-up.

There is a significant range of normal values with respect to the sagittal alignment of the spine [2, 4, 8, 10, 24, 27, 30, 33]. Accepted normal ranges of thoracic kyphosis and lumbar lordosis are 20°–50°, and 20°–60°, respectively. In our present study, seven patients were outside this range: five patients with thoracolumbar HV and one patient with a lumbar hemivertebra developed a thoracic hypokyphosis on follow-up, and one patient with a lumbar HV developed lumbar hypolordosis and thoracic hypokyphosis. The average segmental kyphosis for these patients was 17°.

Twenty-nine patients showed a sagittal plumb line with in normal range on follow-up (Figs. 2, 3).

Complications

One patient had an intraoperative fracture of the vertebral body during screw insertion, and required fusion of one additional segment.

Loosening of the screw rod interface (Webb–Morley Spine System, Biomet-Merck Ltd, Bridgend, UK) occurred in one patient. Intraoperative correction was lost in this patient, although went on to stable bony union without further intervention.

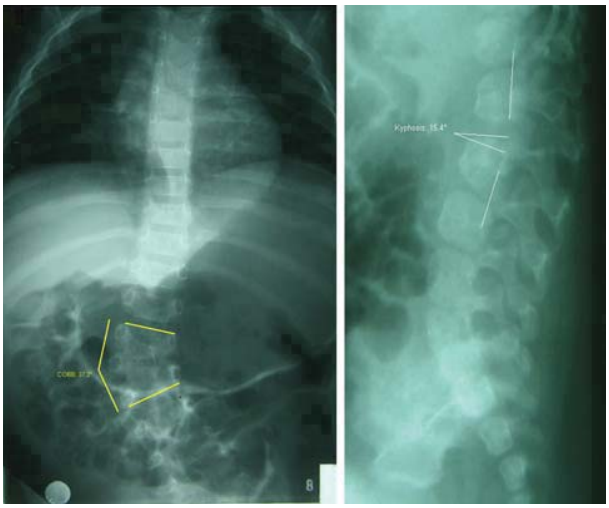

Fig. 2 Preoperative anterior–posterior and lateral radiograph in a 19-month-old girl with congenital scoliosis due to fully segmented hemivertebra

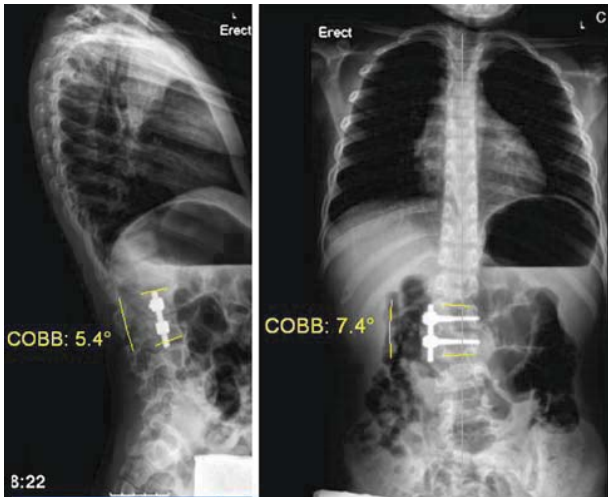

Fig. 3 Follow-up radiographs of the spine 7 years later showing satisfactory fusion and deformity correction in both coronal and sagittal planes

There were no neurological complications. Pseudoarthrosis was not observed.

Two patients had a deep posterior postoperative wound infection which required wound debridement, and resolved successfully with a course of antibiotics. No other reoperations were performed.

Discussion

Surgery is often necessary for progressive congenital scoliosis due to HV in the early growth cycle [7, 11, 13, 21, 25]. The severity of the curve, the type of HV, the location and the age of the patient are important factors in choosing the type of surgical intervention needed [3, 6, 22, 39].

Surgery is indicated with severe curvature at presentation, documented progression or predictable course. A corrective procedure to prevent the development of secondary structural curves and keeping the fused segment short to allow spinal growth is preferable [15, 16, 25, 28].

However, surgical treatment for congenital scoliosis carries a higher risk of paralysis [12, 19, 38]. Abnormal vascular supply to the spinal cord in congenital scoliosis has been postulated as one of the reasons for the higher incidence of neurological injury after surgical treatment for congenital deformities [5]. Normotensive anaesthesia and spinal cord monitoring are mandatory [18, 35].

An ideal treatment for congenital scoliosis due to HV is controversial [39] (Table 2).

Posterior spinal fusion alone with or without instrumentation carries a high risk of crank shafting, non-union and curve progression and most surgeons would reserve

Table 2 Summary of previous reports of surgical treatment for progressive congenital scoliosis

	Year of publication	No. of patients	Mean follow-up (years)	Age at surgery (years)	Approach + instrument.	Scoliosis main curve			Segmental kyphosis		Complications
						Preoperative (degree)	Follow-up (degree)	Improvment at FU (%)	Preoperative (degree)	Follow-up (degree)	
Winter et al. [37]	1984	290	6	11	Post. arthrodesis (127 with Harrington rod)	55	44	20	–	–	26% more than 10° loss of correction 20 pseudoarthrosis 2 paraplegias
Winter et al. [36]	1988	13	6.6	3.6	Ant. + post. convex epiphysiodesis and hemiarthrodesis	46	31	32	–	–	1 extension of fusion 5 (38%) ephiphysiodesis effect with improvement more than 5°
Dubousset et al. [9]	1993	10	2.4	4.1	Ant. + post. convex epiphysiodesis and hemiarthrodesis	46	37	20	–	–	1 epiphysiodesis effect with improvement more than 5°
Holte et al. [13]	1995	37	6	12	Ant. + post. HV resection Post. hooks + rod in 28 patients (18 had previous arthrodesis)	54	35	35	–	–	8 nerve root lesions (1 permanent) 3 pseudoarthrosis 3 Infections 6 extension of fusion Reoperation rate 35%
Callahan et al. [6]	1997	10	4.5	3.9	Ant. + post. resection 9 spinous process wiring 1 Harrington rod	40	16	60	–	–	1 wire removal for pain 1 rod breakage
Lazar and Hall [16]	1999	11	2.3	1.5	Ant. + post. HV resection Post. hooks + rod	47	14	70	30	17	1 transient leg weakness 3 metal work removals Short follow-up
Klemme et al. [15]	2001	6	3.4	1.6	Ant. + post. HV resection Sublaminar suture	38	11	71	–	–	Suture cutting through lamina
Shono et al. [28]	2001	12	5.9	14	Post. resection Post. hooks + screws Long instrumentations	49	18	64	40	17	Long instrumentation Concave disc material not completely removed
Nakamura et al. [23]	2002	5	12.8	10	Post. resection Post. hooks + rod Long fixations	49	26	52	48	17	1 spinal decompensation
Ruf and Harms [25]	2003	25	3.5	3.3	Post. resection Transpedic. screws 20 unisegmental instrumentations	45	13	72	22[a]	8[a]	3 implant failures requiring revision 2 pedicle fractures 2 curve progression at the operation site requiring revision 1 Infection and revision
Bollini et al. [3]	2006	34	6	3.5	Ant. + post. HV resection Post. hooks + rod	40	27	33	20[a]	14[a]	5 pseudoarthrosis 6 curve progression Length of instrumentation unspecified 1 transient paraparesis

[a] Segmental angle of kyphosis

this procedure for patients presenting late in the growth cycle with no decompensation of the spine [37].

Combined anterior and posterior hemiarthrodesis was designed to benefit of the growth potential on the concave side of the curve [9, 20, 31, 36]. Results are unpredictable but it remains a viable option on young children with low magnitude curves with enough discs above and below to prevent unbalancing of the spine.

An anterior or posterior HV excision lead to instability and it is necessary to apply compression instrumentation for adequate arthrodesis [15, 28]. A number of reports describe the difficulty of closing the wedge created by means of hooks, wiring techniques and sublaminar suture tape requiring frequently longer posterior fusions [3, 15, 16, 23].

HV usually produce acute angular deformity and posterior compression instrumentation can end up in the midline or concavity of the curve, increasing the deformity even further [3]. Spinal cord injury and nerve root compression when the pedicles of the vertebra above and below are approximated has been reported in HV resection [17, 29, 38].

A recent development has been the excision of HV through a posterior only approach [23, 25, 28]. The posterior-only approach for hemivertebra excision is a technically demanding procedure especially above the level of the cauda equina.

Anterior column visualisation and concave disc resection is difficult. Spinal cord and dural manipulation increases the risk for neurological injury [12].

Ruf et al. [25] reported good sagittal and coronal correction on 28 patients with posterior HV resection and pedicle screw instrumentation. He also reported difficulties achieving secure posterior instrumentation.

High intraoperative blood loss has been reported with posterior HV resection [23, 25]; thus the ability of maintaining normotensive anaesthesia in children where the incidence of congenital heart disease averages 26% [1], can be compromised.

Focal kyphosis can improve with further growth after single stage posterior resection of HV with transpedicular fixation [26]. This phenomenon has not been observed in series using a combined approach with posterior instrumentation [3]. In our series, we have not observed an anterior tethering effect with progressive focal kyphosis during further growth.

Our technique described earlier has a low intraoperative blood loss. No dural manipulation is needed and interference with local blood supply to the spinal cord is minimal. The partially preserved HV is the ideal vascularised bone graft to achieve solid fusion.

Short anterior instrumentation is able to transmit higher convex compressive forces to the vertebral body than a posterior tension band system like pedicle screws, providing good correction and stability [34]. This technique is particularly effective for correction of adjacent ipsilateral hemivertebrae (Figs. 4, 5)

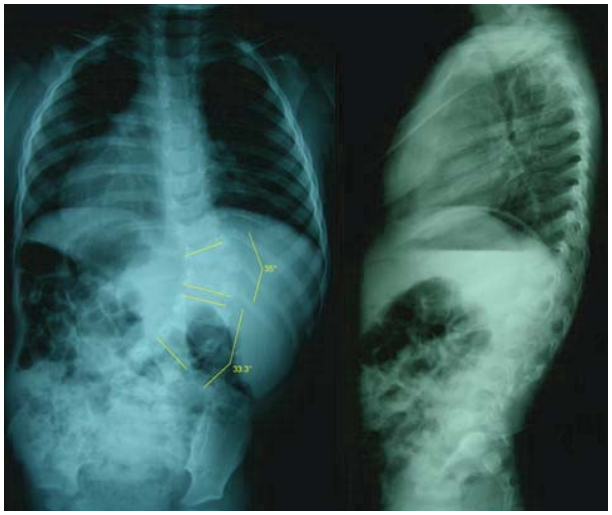

Fig. 4 Preoperative anterior–posterior and lateral radiograph in a 26-month-old boy with congenital scoliosis due to two fully segmented ipsilateral hemivertebrae

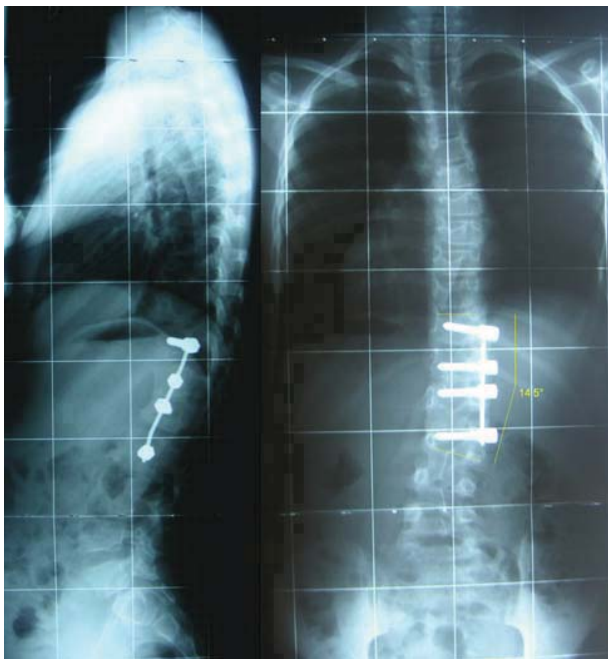

Fig. 5 Follow-up radiographs of the spine 8.5 years later showing a balanced spine with satisfactory fusion in the coronal plane and residual thoracolumbar kyphosis

Our technique has many advantages but has only limited ability to correct focal kyphosis. In cases with significant kyphosis, additional posterior instrumentation is required to close the posterior gap.

Conclusions

A range of procedures has been proposed for the treatment of congenital scoliosis. Trend is more towards the radical procedures in young children to correct this condition with significant morbidity. Our series of short anterior instrumented fusions with partial preservation of the HV show that this is a safe and effective technique with minimal blood loss, reliable fusion rate and low morbidity to treat congenital scoliosis in very young children.

It also provides excellent coronal correction and balanced growth in the coronal plane. In the saggital plane, especially in the thoracolumbar junction persistant kyphosis during correction may require the addition of posterior instrumentation.

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SPINE Volume 35, Number 6, pp E218–E222

Anterior Instrumentation and Correction of Congenital Spinal Deformities Under Age of Four Without Hemivertebrectomy

A New Alternative

Hazem B. Elsebaie, FRCS, MD,* Wael Kaptan, MD,* Yasser El Miligui, FRCS, MD,* Mohamed A. El Masry, MRCS, MD,* Motaz Salaheldine, MSc(Orth),† Hilali M. Noordeen, FRCS(Orth),‡ and Behrooz A. Akbarnia, MD§¶

Study Design. Retrospective clinical and radiologic evaluation of a single-stage partial corpectomy of the hemivertebra with anterior instrumentation and simultaneous posterior noninstrumented fusion.

Objective. To determine the safety and efficacy of a new technique in the management of progressive congenital spinal deformities due to failure of formation in the very young age.

Summary of Background Data. Several techniques have been reported for the surgical treatment of young children with congenital spinal deformities. There have been concerns regarding epidural bleeding, neurologic complications, pedicle screws placement, implant failure, and prominence of posterior constructs in this very young age group. A single-stage partial corpectomy of the hemivertebra with anterior instrumentation and simultaneous posterior noninstrumented fusion can offer a new alternative which can avoid these concerns.

Methods. Twelve patients with progressive congenital spinal deformities due to failure of formation were retrospectively reviewed after adopting the above mentioned technique. All patients included in the study presented with a single hemivertebra. The mean age at time of surgery was 2 years 7 months (range, 1 year and 9 months to 3 years and 10 months). The average follow-up period was 3 years and 1 month (range, 2 years to 4 years and 5 months).

Results. There were no cases of intra or postoperative neurologic or implant related complications. There was 1 superficial infection. All patients showed solid radiologic fusion. The mean scoliosis angle improved from 48.3° (range, 34°–58°) preoperative to 17.2° (range, 11°–25°). The mean angle of kyphosis improved from 23.2° (range, 16°–57°) before surgery to 11.7° (range, 4°–16°).

Conclusion. A single-stage partial corpectomy of the hemivertebra with anterior instrumentation and simultaneous posterior noninstrumented fusion offers a safe alternative method in treating patients with congenital hemivertebra under the age of 4 years.

Key words: congenital scoliosis, hemivertebra, anterior instrumentation. **Spine 2010;35:E218–E222**

The natural history of congenital scoliosis is usually progressive and surgery is required in most cases. Hemivertebrae are the most frequent causes of congenital scoliosis.[1]

Previously described surgical procedures include *in situ* posterior or anterior–posterior fusion with or without instrumentation, combined anterior and posterior convex hemiepiphysiodesis and hemiarthrodesis, hemivertebra excision with fusion,[2] and more recently posterior hemivertebra resection with transpedicular instrumentation.[1] Fusion *in situ*, hemiepiphysiodesis and hemiarthrodesis while successful in preventing the progression of deformity, is unable to produce significant correction of the existing deformity and moreover the effect on further growth is unclear.[3]

Hemivertebra resection which is a wedge osteotomy was first described by Royle in 1928.[4] Immobilization in a body cast or a brace for approximately 6 months has been customary. Further reports on resections by combined anterior and posterior approaches in 1-stage or 2-stage procedures with or without instrumentation followed.[5–7] A recent interest in posterior pedicle screw fixation of these curves after hemivertebrae excision yielded better results with significant correction, using a combined anterior and posterior approach[7] or a posterior only approach.[1,8,9] There have been concerns regarding complications such as epidural bleeding, neurologic complications, pedicle screws placement, implant failure, and prominence of posterior constructs in this very young age group.

The aim of this study was to report on a single-stage partial corpectomy of the hemivertebra with anterior instrumentation and simultaneous posterior noninstrumented fusion in the treatment of congenital scoliosis caused by a hemivertebra in children below the age of 4 years. To the best of our knowledge, the use of such technique has not been previously reported in the literature.

From the *Cairo University Teaching Hospitals, Cairo, Egypt; †Abulreish Student's Insurance Hospital, Cairo, Egypt; ‡Great Ormond Street Hospital, London, United Kingdom; §University of California, San Diego, CA; and ¶San Diego Center for Spinal Disorders, La Jolla, CA.

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Address correspondence and reprint requests to Hazem B. Elsebaie, FRCS, MD, 22 Degla St, P.O. Box Office 12411, Mohandessine, Giza, Egypt; E-mail: hazembelsebaie@yahoo.com

■ Materials and Methods

Twelve consecutive patients were evaluated retrospectively. All patients underwent surgery at our institutions between 2002 and 2005.

There were 8 girls and 4 boys. The mean age at time of surgery was 31.2 months (range, 21–46 months).

All patients presented with a single level hemivertebra. The hemivertebra located at the thoracic spine in 4 cases and the thoracolumbar region in 8 cases. Seven hemivertebra were right sided, and 5 were left sided. Nine hemivertebra were fully segmented and 3 were semisegmented. Radiographic measurements were performed with the patient standing using coronal and lateral radiographs of the spine. Cobb's method was used to assess the scoliosis angle.[10] The local kyphosis angle was measured to assess the degree of kyphosis.

All patients underwent preoperative MRI, renal ultrasound, and echocardiography. Of the 12 patients, none had intraspinal anomaly, 5 showed associated anomalies in the following locations: genitourinary system in 2, and cardiopulmonary system in 3. None of the patients had undergone a prior operation.

Surgical Technique

A single-stage partial hemivertebrectomy with anterior instrumentation using different rod screw systems with screws diameters of 4.35, 4.75, and 5 mm and with rod diameters of 4.75 and 5.5 mm; simultaneous posterior noninstrumented fusion was used in all the patients.

The patient was placed in the lateral decubitus position with the hemivertebra facing upward. Both the thoracic and abdominal regions extending from the axilla to the greater trochanter and extending anterior to the sternum and umbilicus were sterilized and prepared together with the posterior spine from the cervicothoracic junction to the gluteal fold. Depending on the level of the deformity; a thoracic approach was used for hemivertebrae at and above T10 and a retroperitoneal thoracoabdominal approach was used below T10.

After identifying the hemivertebra using its shape and the internal counting of the ribs using the preoperative Plain Radiographs as a guide for identification of the level, full exposure of the discs and vertebral bodies above and below was done (Figure 1). Segmental vessels at the level of the hemivertebra have to be cauterized for adequate preparation to the following steps, the intervertebral discs above and below the hemivertebra were completely excised followed by removal of the adjacent cartilaginous endplates and hence exposing the underlying bone. The convex-side cortex of the hemivertebra was removed exposing the cancellous bone. Partial anterior corpectomy was done using a rongeur and/or a high-speed burr reaching the concave tether on the other side of the hemivertebra and posteriorly leaving the posterior cortex with its overlying cancellous shell and pedicle intact to avoid exposure of the spinal canal and epidural space. Partial corpectomy is important as an additional type of release, it gives full exposure of the endplates of the bodies above and below the hemivertebra to give a better guide for the anterior screws insertion, and finally it gives a full exposure to the tough concave tether allowing for its safe rigorous release.

Release of the concave side soft tissue tether was then achieved greatly aided by a manual posterior to anterior push at the apex of the deformity putting this tether in tension and therefore identifying the amount of maximum release needed for better correction. At this stage insertion of the body screws at the vertebrae above and below the hemivertebra was done, the entry point was identified, the bone at the entry point was opened by a sharp awl, the direction of the screw was guided by the well exposed endplate, and the screw was then inserted with a special attention to go as posterior in the body as possible to have a better hold without using a tap in order to get a bicortical purchase in the body. The position of the screws was checked by an image intensifier in an anteroposterior and lateral views. The rod was gently bent before insertion to fit into the deformity then rotated into a lordotic position to facilitate correction of the kyphosis; the amount of rod bent is dependent on the degree of kyphosis and the site of the deformity with its expected normal sagittal contour. The rod was applied first to the cephalad screw and correction of the deformity was achieved by bringing the rod onto the caudal screw trying to get the superior and inferior endplates (around the hemivertebra) as parallel as possible, this is greatly helped by a gentle gradual manual push on the posterior elements at the apex of the deformity to open up the space at the concave side and to correct the kyphosis at the same time, a strut rib graft was then inserted to keep the space opened and decrease the stress on the implant (Figure 2). Following the insertion of the rod the screws are compressed, to achieve better correction before final tightening

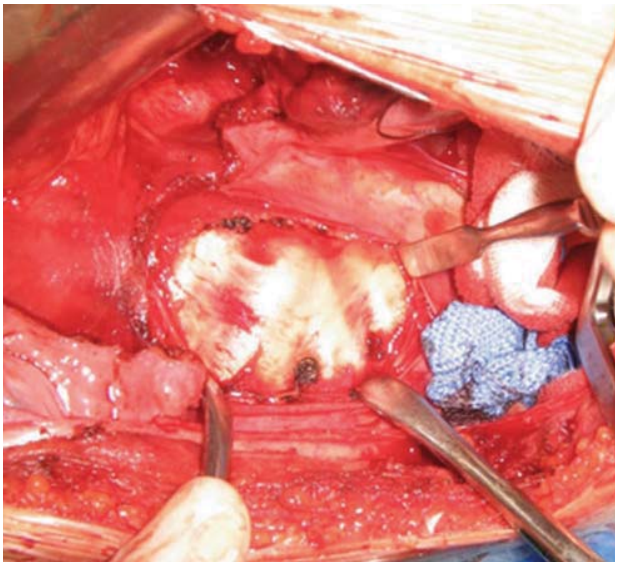

Figure 1. Intraoperative picture showing the hemivertebra after full exposure.

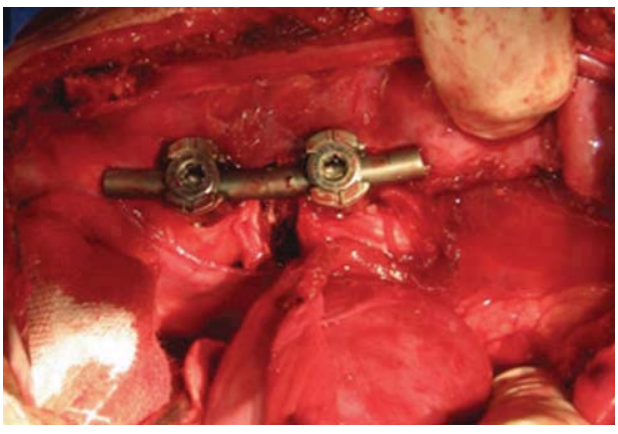

Figure 2. Intraoperative picture after insertion of the screws and placement of the anterior rib graft.

Table 1. Preoperative and Postoperative Data

Patient Number	Age at Operation (mo)	Preoperative Cobb°	Preoperative Kyphosis°	Follow-up Cobb°	Follow-up Kyphosis°
1	23	44	n	22	9
2	21	42	27	13	6
3	26	58	29	25	16
4	23	34	n	12	7
5	42	52	23	11	10
6	43	50	38	19	15
7	46	56	22	15	13
8	32	40	16	16	4
9	28	41	n	17	12
10	25	52	34	21	15
11	31	55	32	18	14
12	34	56	57	17	19
Average	31.2	48.3	23.2	17.2	11.7

n indicates normal.

of the screws. Autologous strut rib graft was then packed into the void space left after the partial corpectomy and correction (Figure 5). Closure of the wound in layers with a routine use of a chest drain was used in all patients.

The patient was then turned to the prone position; a routine posterior approach was used exposing the posterior elements of the vertebra above and below the hemivertebra identified by image intensifier in an anteroposterior view. Decortication of the posterior elements of the hemivertebra with the vertebra above and below was achieved using a high speed burr followed by the application of autologous rib graft.

Wake up test was performed for all patients, and additional neuromonitoring has been used in 4 cases, and this was decided per surgeon's discretion. All patients had prophylactic antibiotic coverage for 24 hours. The drains were removed at 48 hours. All patients were mobilized in a brace for 12 weeks. They were routinely discharged from hospital 5 days after surgery, and reassessed in the clinic at 6 weeks. They were routinely reviewed again with further plain radiographs at 3, 6, and 12 months, and then yearly after surgery.

■ Results

The average follow-up period was 3 years and 1 month (range, 2–4 years and 5 months).

The mean blood loss was 150 mL (range, 80–500 mL). The mean operating time for the anterior and posterior procedures was 120 minutes (range, 98–180 minutes). There were no cases of intra or postoperative neurologic complications. There were no cases of deep infection; 1 patient developed a superficial wound infection. There was no obvious pseudarthrosis, decompensation, or implant failure at the last follow-up.

The mean preoperative was 48.3° (range, 34°–58°). The mean kyphosis angle was 23.2° (range, 16°–57°) (Figures 3A, B). The mean postoperative Cobb angle for scoliosis was 17.2° (range, 11°–25°) with no loss of correction at the latest follow-up. This represents a 64.5% improvement in the curve magnitude. The mean postoperative kyphosis angle was 11.7° (range, 4°–16°) which was maintained at the latest follow-up (Table 1). This represents a 49.5% improvement (Figures 4A, B). One patient developed a junctional kyphosis and another patient had an adding on (elongation of the distal scoliotic curve).

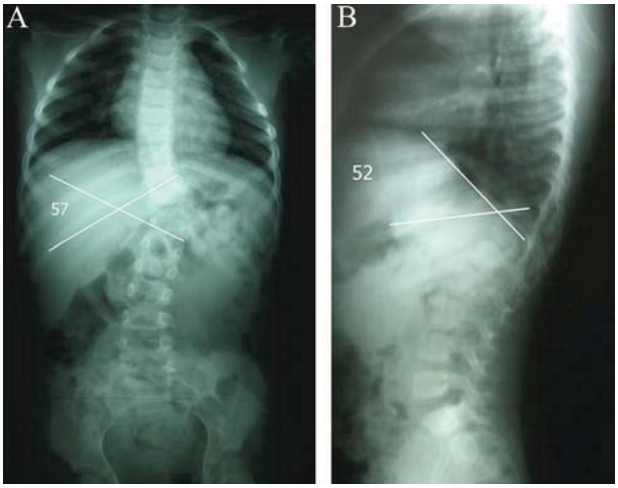

Figure 3. **A, B,** Preoperative anteroposterior and lateral radiographs showing a scoliotic curve measuring 56° and a kyphosis angle measuring 52°.

■ Discussion

Untreated congenital scoliosis caused by hemivertebrae usually leads to unacceptable deformities.[11,12] McMaster and Ohtsuka[13] reported that approximately 75% of congenital scoliosis progress significantly and that congenital kyphosis due to a hemivertebra always showed rapid progression.[14]

Fusion *in situ*, hemiepiphysiodesis and hemiarthrodesis while successful in preventing the progression of deformity, is unable to produce significant correction of the

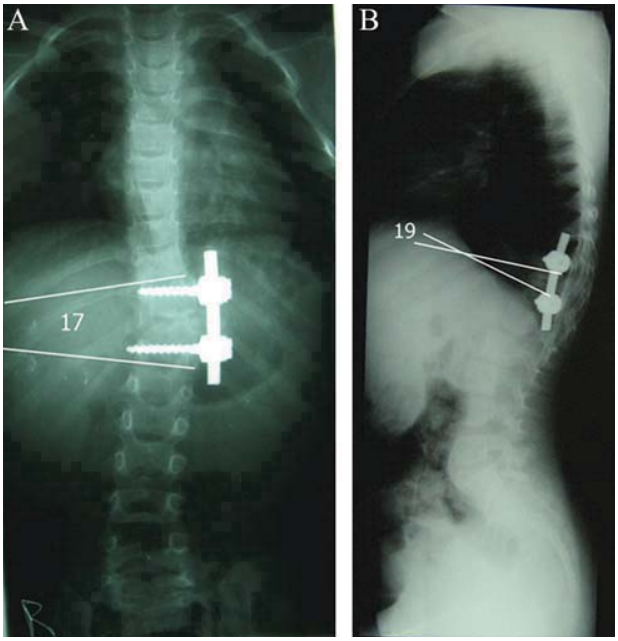

Figure 4. **A, B,** Two years and 8 months follow-up anteroposterior and lateral radiographs showing a residual scoliotic deformity measuring 17° and a kyphotic deformity measuring 19°.

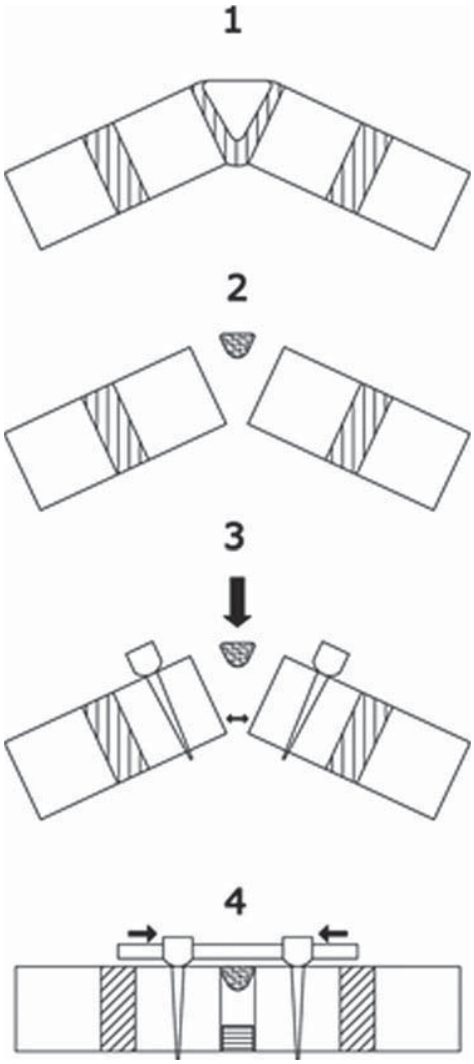

Figure 5. Diagram for description of the surgical technique: (1) Exposure of the hemivertebra. (2) Excision of the intervertebral discs with partial anterior corpectomy. (3) Insertion of body screws. Manual push on the posterior elements at the apex of the deformity to open up the space. (4) Autologous strut graft packed; the rod inserted and the screws are compressed.

existing deformity and moreover the effect on further growth is difficult to estimate.[3]

Early description of hemivertebra excision was from Royle[4] in 1928, further reports followed.[15,16] The most notable problems were pseudarthrosis, kyphosis and neurologic deficits.

Leatherman and Dickson[17] developed the concept of two-stage surgery with shortening of the spine during correction to reduce the risk of neurologic complications. They reported on 50 patients treated with 2-stage hemivertebra excision, with a mean correction of 43%.

Slabaugh *et al*[18] reported on 8 lumbosacral resections of hemivertebrae, again as a 2-stage procedure with a mean correction of 35%. Bergoin *et al*[19] reported on 19 resections of hemivertebrae in 9 children who underwent a single-stage posterior and anterior procedure and were able to achieve a mean correction of scoliosis from 30° to 16° and mean correction of kyphosis from 33° to 12°. Holte *et al*[6] reported on a series of 37 patients with congenital scoliosis treated with thoracic and lumbar hemivertebrectomy but reported 8 neurologic complications (seven were transient and 1 permanent).

A combined anteroposterior fusion reduces the incidence of pseudarthrosis and theoretically reduces crank shaft phenomena.

A recent interest in posterior instrumentation with hemivertebra excision in children below the age of 5 years yielded better results with significant correction. Lazar and Hall[7] reported on a simultaneous anterior and posterior hemivertebra excision with posterior instrumentation in 11 patients with a mean age of 18 months, they achieved an average correction of 70% with 1 patient having a postoperative transient left leg weakness. Ruf and Harms[1] published their results with posterior hemivertebra excision with transpedicular instrumentation in children aged 1 to 6 years, with mean curve of 45°. The average blood loss was 496 mL. They had 1 infection, 2 pedicle fractures, 3 implant failures and 2 new developing curves. They achieved a considerable correction of 68.8% for the main scoliosis angle and 60% of the local kyphosis angle.

There has been a concern regarding epidural bleeding, neurologic complications, pedicle screws placement, implant failure, and prominence of posterior constructs in this very young age group. The new alternative technique described in this study consisting of partial corpectomy of the hemivertebra leaving the posterior cortex, pedicle and posterior elements intact without exposing the dura with anterior instrumentation and posterior fusion avoids such problems. Moreover, it allows excision followed by correction and instrumentation at the site of pathology. The anterior instrumentation will allow direct compression of the graft, obviates the need for postoperative casting allowing early mobilization with a removable brace, and avoids the use of posterior pedicle screw fixation with its potential difficulties and risks. A great part of the correction in this technique is achieved by closing the discs above and below the hemivertebra and opening the contralateral concave side where the strut graft is placed making the 2 endplates as parallel as possible, all while leaving the posterior cortex intact.

The results reported in this study of 64.5% mean correction of the scoliosis angle and 49.5% mean correction of the kyphosis angle correlates well with the best results reported in the literature.

In our series, there were no implant related complications, and despite concerns for the relative bulkiness of the head of the screws for this age group this did not seem to cause any additional problems, there were no neurologic complications, no decompensation, and no deep infections. One patient developed superficial wound infection which responded well to oral antibiotic treatment.

A short fusion increases the risk of newly developing deformity and the need for reoperation; however, this should be accepted in order to minimize the compromise

of spinal growth.[9] We had 1 patient who developed a junctional kyphosis, and 1 patient who had an adding on (elongation of the distal scoliotic curve) which increased despite bracing and was subsequently scheduled for growing rods; it would of course be valuable to have a much longer follow-up for these patients through maturity to get a more comprehensive picture for the long-term behavior of the whole spine.

■ Conclusion

Early instrumented correction and fusion in the very young age group is the recommended treatment for progressive congenital scoliosis produced by a hemivertebra. A single-stage partial corpectomy of the hemivertebra with anterior instrumentation and simultaneous posterior noninstrumented fusion is a safe and effective alternative technique in such patients. It avoids the risks and disadvantages of complete excision of the hemivertebra and pedicle screws instrumentation with comparable efficacy. Smaller size implant systems specially designed for this age group would make their fixation easier and decrease the concerns related to the bulkiness of the currently available systems.

■ Key Points

- Surgery is required in most cases of congenital scoliosis.
- Fusion *in situ* is unable to produce significant correction of the existing deformity.
- The most notable problems with excision of the hemivertebra are epidural bleeding, pseudarthrosis, kyphosis and neurologic deficits.
- A single-stage partial corpectomy of the hemivertebra with anterior instrumentation and simultaneous posterior noninstrumented fusion offers a safe and alternative method.

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The Surgical Treatment of Congenital Kyphosis

M. H. H. Noordeen, MA, FRCS,*† Enrique Garrido, EBOT, MRCS,*
Stewart K. Tucker, FRCS (Orth),*† and Hazem Bayoumi Elsebaie, FRCS

Study Design. Retrospective study with clinical and radiologic evaluation of 15 patients with congenital kyphosis or kyphoscoliosis who underwent anterior instrumented spinal fusion for posterolateral or posterior hemivertebra (HV). The management of congenital kyphosis has been described in the literature using a variety of techniques. The presentation of patients at diagnosis is discussed. The question of when to begin treatment is reviewed. The pitfalls in the management and how to avoid these are discussed. The different published techniques are reviewed. We present our own techniques and our results of treatment of congenital kyphosis in very young children.

Objective. To evaluate the safety and efficacy of early surgical anterior instrumented fusion with partial preservation of the HV in the treatment of progressive congenital kyphosis in children below the age of 3. We discuss the management of patients presenting with neurologic compromise. We aim to systematically review the literature and to present our own experience in the management of these deformities, so that the issues common to treating physicians may be explored.

Summary of Background Data. A variety of treatments have been described in the literature for the treatment of congenital kyphosis due to HV. We report the results of our technique.

Methods. Between 1997 and 2005 we have treated 15 consecutive patients with progressive congenital kyphosis with anterior instrumented fusion and strut grafting. Thirteen patients had a single posterolateral HV and 2 patients had a single posterior HV. Of the 15 patients in the study, 5 were girls and 10 boys. Mean age at surgery was 22 months (range, 8–33). Mean follow-up period was 6.8 years. Thirteen HV were located in the thoracolumbar junction (T10–L2) and 2 in the thoracic spine.

Results. The average operating time of procedure was 150 minutes (range, 130–210 minutes). The average blood loss was 180 mL (range, 100–330 mL), equivalent to a mean external blood volume loss of 15% (range, 11%–24%).

Preoperative segmental Cobb angle averaging 34° at last follow-up. Compensatory coronal cranial and caudal curves were corrected by 50%. The angle of segmental kyphosis averaged 39° (range, 20°–80°) before surgery and 21° (range, 11°–40°) at last follow-up. This represents a 43% of improvement of the segmental kyphosis, and a 64% of improvement of the segmental scoliosis at last follow-up. One case with initial kyphosis of 80° continued to progress and required revision anterior and posterior surgery. There were no neurologic complications.

Key words: hemivertebra, congenital kyphosis, congenital spine deformity, anterior instrumented fusion, segmentation mismatch. **Spine 2009;34:1808–1814**

From the *Department of Spinal Surgery, Great Ormond Street Hospital for Children, London, United Kingdom; and †Spinal Deformity Unit, Royal National Orthopaedic Hospital, Stanmore, London, United Kingdom.
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Study conducted at the Great Ormond Street Hospital for Children and the Royal National Orthopaedic Hospital, Stanmore, London, United Kingdom.
Address correspondence and reprint requests to Enrique Garrido, EBOT, MRCS, Department of Spinal Surgery, Great Ormond Street Hospital for Children, Great Ormond St, London WC1N 3JH, United Kingdom; E-mail: Garrie@gosh.nhs.uk

Natural History and Classification

The estimated prevalence of vertebral malformations is approximately 0.5 to 1 in 1000. Beals *et al* have shown that the most common vertebral anomaly is the multiple hemivertebra (HV) (44%) followed by anterior segmentation defects (32%) and single HV (18%).[1] Congenital kyphosis is far less common than congenital scoliosis. The deformity is caused by developmental anomalies that impair longitudinal growth anterior or anterolateral to the transverse axis of vertebral rotation in the sagittal plane.[2] Winter *et al* concluded that congenital kyphosis is progressive without surgical intervention. The apex of the deformity occurs most commonly in the thoracolumbar junction. Winter *et al* classified the deformities into 3 types.[3]

Type 1

Anterior failure of vertebral body formation is the most common type of congenital kyphosis. This deformity if left untreated can develop into severe rigid angular deformity, causing spinal cord compression in 25% of cases.[2,4]

In North America, congenital kyphosis is the most common cause of paraplegia due to spinal deformity.[5] Paralysis occurs most commonly during the pubertal growth spurt.

Type 1 deformity due to posterolateral quadrant HV progresses at a median rate of 2.5° per year until the age of 10 and then 5° thereafter. Posterior hemivertebrae have slightly better prognosis followed by butterfly and wedge vertebrae.[2]

Type 2

Anterior failure of vertebral body segmentation are anterior or anterolateral bars. These curves have a smooth kyphosis and usually involve several segments. Anterior bars in McMaster series showed a slowly increasing deformity by about 1% per year before the age of 10, with anterolateral bars having a worse prognosis.[2]

Other authors have reported higher deformity progression in failures of vertebral body segmentation.[6] The main problem of these malformations is spinal imbalance. Specially if located in the thoracolumbar junction it produces a compensatory hyperlordosis causing low back pain.[6]

Type 3

A combination of anterior failure of formation and segmentation and is the least common malformation. Kyphoscoliosis due to anterolateral unsegmented bar combined with posterolateral quadrant HV progress at 5° per year until the age of 10 and 10° thereafter.[2] In practice, deformities present for treatment either with severe deformity with or without neurologic compromise, or as deformity with the propensity to progress.

■ Patient Evaluation

Associated Congenital Anomalies

Vertebral malformations have been shown to be associated with hemifacial microsomia, Alagille, Jarcho-Levin, Klippel-Feil, Goldenhar, Joubert syndrome, and VACTERL sequence as well as basel cell nevus, trisomy 18, and diabetic embryopathy.[7]

Associated anomalies both within and outside the spine in children with congenital deformity ranges from 50% to 60%.[1,8] In patients with congenital spinal deformity, the prevalence of intraspinal anomalies that are detected with MRI has been reported to be approximately 37%.[9]

Treatment of intraspinal pathologies is usually performed before deformity correction. However, a recent study has reported simultaneous neurosurgical treatment and correction of deformity without adverse events.[10]

Congenital heart disease is present in 7% to 26% of children. Urogenital anomalies occur in 21% to 27% of patients with vertebral malformations but obstructive uropathy is found only in 3% of this patients.[9,11,12] All patients with congenital spinal deformity should be investigated with whole spine MRI, echocardiography, and renal ultrasound before any surgical intervention.

Anatomical Pitfalls

Accurate radiologic evaluation is essential. Morphologic evaluation is more difficult in congenital kyphosis than in scoliosis, especially in young children. Plain radiographs have shown poor sensitivity identifying anomalies in the posterior elements of the spine.[13] Preoperative planning and intraoperative localization of the congenital anomaly is facilitated by a detailed knowledge of the anatomy. Three-dimensional computed tomography allows precise correlation of anterior and posterior anatomy to plan osteotomy planes and anticipate problems with placement of the instrumentation.[13,14] The authors have performed osteotomies of single posterior segmentation failures during HV resection based on the preoperative imaging. Multiplanar reformatted images are particularly useful in recognizing complex segmentation failures.[14]

Classification

Moe and Winter *et al* classified congenital scoliosis according to morphologic characteristics on plain radiographs into formation failure, segmentation failure, and a mixed type. Earlier authors have recognized the frequent lack of correlation between anterior and posterior elements in congenital vertebral malformations.[13,15] Nakajima studied the morphology of the posterior elements and classified these "discordant" anomalies as mismatch malformations and mixed complex malformations. We feel that a classification system that describes the anomaly accurately is necessary to study the natural history as well as to guide treatment in the future.

Deformities can be very complex and it is best to describe them individually by their anterior and posterior formation and segmentation anomalies using Winter classification system. As Nakajima proposed one can classify the congenital deformity into type A where there is correlation of anterior and posterior malformation and type B where there is a mismatch.

Classification would start by describing the anterior elements into Type 1 (anterior failure of vertebral body formation), Type 2 (anterior failure of vertebral body segmentation), and Type 3 (combination of anterior failure of formation and segmentation) followed by description of the posterior morphology.

Vascular Pitfalls

Several reports show that single-stage HV excision through anterior/posterior or posterior only approach is safe in the young child.[16–24] However, there is a great concern regarding the risk of spinal cord injury during HV excision, especially in older children.[25] Abnormal vascular supply to the spinal cord in congenital deformity has been postulated as one of the reasons for the higher incidence of paralysis after surgical treatment for congenital deformities.

Loss of spinal cord monitoring signal occurs in 20% of children undergoing kyphosis correction, with spinal osteotomy.[26] Loss of SSEP and paraplegia has been reported during segmental vessel ligation in congenital kyphoscoliosis before any correction maneuver is performed.[27]

Tanaka and Uhtoff showed in a study on 11 embryos and fetuses with vertebral malformations that that there is a relationship in the genesis of aberrant vertebrae and vasculature.[28] Abnormal segmental blood supply has been reported on adults with congenital kyphosis.[29] Our own observations with CT angiography have shown aberrant vasculature and abnormal course of vessels.

The extramedullary blood supply to the spinal cord comes from many branches derived from segmental arteries. The segmental arteries divide into numerous branches at the intervertebral foramen that supply the anterior and posterior part of the vertebral canal. Only at some levels this arteries further divide into arteries that join the anterior and posterior roots to reach the me-

Table 1. Data on Patients Who Presented to Us With Significant Neurological Deficit Improved With Surgical Stabilisation and No Correction of the Deformity

Case	Sex	Age at Presentation	Associated Anomalies	Level	Preoperative Neurologic Condition	Follow-Up (yr)	Procedure	Current Neurological Status
1	M	16	No	T8 posterior HV	Paraparesis only residual movement in right foot. Incontinent Frankel C	10.3	Anterior release and rib strut grafting. Posterior instrumented fusion T4–T12. Somatosensory evoked potentials could not be recorded	Walk indoors unaided, outdoors 2 crutches. Regained bowel and bladder control
2	M	14	No	T5 posterolateral HV	Paraparesis lower limbs MRC 3 with hyperreflexia and normal sensation. No bowel or bladder dysfunction	6	Anterior in situ fusion and posterior instrumented fusion T3–T8 Somatosensory evoked potentials could not be recorded	Full recovery working in the building trade
3	M	3	No	L1 posterior HV	Hyperreflexive, falls and reduced motor power MRC 4	10	Anterior strut grafting and posterior *in situ* fusion. Plaster jacket. Augmentation of posterior bone graft 1 yr later	Full recovery except MRC 4+ weakness in right quadriceps
4	F	6 mo	Bilateral clubfeet	Segmental spinal dysgenesis T12–L1	Spastic paraparesis. MRC 2	7.3	Anterior grafting and instrumentation with cervical plate	Able to ambulate with a walker
5	M	7 mo		Segmental spinal dysgenesis T12–L1	Hyperreflexia, decreased lower extremity movements	8.5	Anterior grafting and instrumentation with cervical plate	Full neurological recovery. Residual kyphosis of 55°

dulla. In the thoracolumbar junction where kyphosis is most common one of the anterior radicular arteries is always dominant in caliber and is termed the great radicular artery or artery of Adamkiewicz. These radicular branches have a typically cranially directed course (due to the ascension of the cord). The intrinsic arterial system of the spinal cord is composed of a central (centrifugal) system and peripheral system (centripetal). The central system is composed of 5 to 9 sulcal or central arteries per spinal segment, arising from the anterior spinal artery. The peripheral system consists of numerous small arteries which originate in the pial network covering the spinal cord and pass through the white matter in a radial course. In the normal spinal cord numerous extra- and intraspinal anastomoses guarantee sufficient collateral supply in the presence of extravertebral occlusion.[30] However, cadaver studies have shown that bilateral ligation of segmental arteries can lead to a decreased blood supply. Chwajol and Houten reported a case in which the segmental arteries failed to develop in concert with the anterior failure of formation of the L1 and L2 body.[29]

In such circumstances interference with the anastomotic network at the apex of the deformity by vertebral resection or osteotomy may lead to cord ischemia.

We conclude that in severe congenital kyphotic deformities blood supply is possibly compromised by 3 factors:

Elongation and compression of the longitudinal arterial trunks, with kinking or stretching of the sulcal arteries.

Instability causing microcirculatory perfusion deficits.

Congenitally deficient segmental blood supply at the level of the deformity.

■ Discussion

Nonoperative treatment has been ineffective in controlling progressive deformity.[3] Operative correction of congenital kyphoscoliosis is still a challenging problem in the growing spine. Normotensive anesthesia and spinal cord monitoring are mandatory. It is important to use frequent spinal cord monitoring during corrective procedures.

Early Treatment

Noninstrumented Fusion. Posterior quadrant HV and posterior HV or type 3 deformities can cause sharp angular kyphosis with spinal cord compression, and early prophylactic surgical intervention is recommended (Appendix 1, Supplemental Digital Content 1; available at: http://links.lww.com/A1325). Winter *et al* outlined the principles of posterior *in situ* arthrodesis to halt the progression of the kyphosis due to unbalanced growth of the spine (Table 1).[3]

However, correction relies on anterior growth potential which is unpredictable. Pseudarthrosis rates approximate 20% and long posterior fusions are needed interfering with further spinal growth.[3,31] Winter *et al* also noted that kyphosis greater than 55° in children older than 5 years required anterior and posterior fusion to halt progression.[31]

Anterior and Posterior HV Excision. Modern pediatric instrumentation and increasing familiarity with anterior and posterior column surgery have allowed more aggressive treatment of these deformities presenting early in the growth cycle. The aim is now not only to prevent future spinal cord compression but to achieve a balanced sagittal profile with a short fusion segment. A prerequisite to achieve this goal is early diagnosis and early surgical

intervention before the curve becomes severe and rigid. HV resections with short segment instrumented fusions can be performed for segmental kyphosis not exceeding 40° in young children.[17,19] The excision of a HV may be performed by using staged or simultaneous anterior and posterior procedures.[16,24]

An anterior or posterior HV excision lead to instability and it is necessary to apply compression instrumentation for adequate arthrodesis.[20,22] A number of reports describe the difficulty of closing the wedge created by means of hooks, wiring techniques, and sublaminar suture tape, requiring frequently longer instrumented posterior fusions.[19,21–23]

HV usually produce acute angular deformity and posterior compression instrumentation can end up in the midline or concavity of the curve, increasing the deformity even further.[19] Spinal cord injury and nerve root compression when the pedicles of the vertebra above and below are approximated have been reported in HV resection.[18,24,25,32]

All-Posterior Approach for HV Excision. A recent development has been the excision of HV through a posterior-only approach.[17,20,23] The posterior-only approach for HV excision is a technically demanding procedure especially above the level of the *cauda equina*. The disadvantages of posterior-only procedures include the difficulty with anterior column visualization and incomplete resection and the need for spinal cord and dural manipulation creating further risk for neurologic injury.[20]

Ruf and Harms reported good sagittal and coronal correction on 28 patients with posterior HV resection and pedicle screw instrumentation.[17] They also reported difficulties achieving secure posterior instrumentation. In their study, 18% of the patients had an implant failure or intraoperative pedicle fracture.

Higher intraoperative blood loss has been reported with posterior HV resection; thus the ability of maintaining normotensive anesthesia in children where the incidence of congenital heart disease is high, can be compromised.[9,17,23]

Anterior Instrumentation. It is our preference to treat congenital kyphosis very early before the deformity becomes rigid and more severe. Anterior instrumentation with anterior strut grafting incorporating 3 spinal segments is our preferred method of treatment for children younger than 3 years of age. We believe that anterior instrumentation with bicortical purchase of the vertebral body offers the most secure fixation in the very young child. Blood loss is less because no epidural bleeding is encountered. Partial preservation of the vertebral body prevents compression of the exiting nerve root.

Between 1997 and 2005 we have treated 15 consecutive patients with progressive congenital kyphosis with anterior instrumented fusion and strut grafting. Thirteen patients had a single posterolateral HV and 2 patients a single posterior HV. Of the 15 patients in the study, 5 were girls and 10 boys. Mean age at surgery was 22 months (range, 8–33). Mean follow-up period was 6.8 years (range, 2–10 years). Thirteen HV were located in the thoracolumbar junction (T10–L2) and 2 in the thoracic spine.

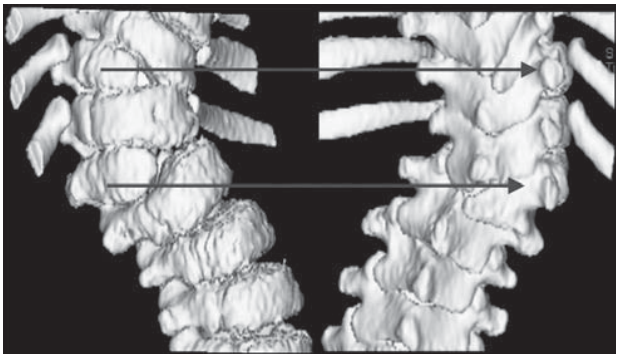

Figure 1. Patient with double ipsilateral hemivertebrae. Posterior elements of the T11 hemivertebra form a complete vertebral arch with the spinal segment above. During surgery it would be easy to misidentify the hemivertebra.

Surgical Technique

All the patients are placed in the lateral decubitus position with the HV facing upward. Spinal cord monitoring is used for all patients. Care must be taken to keep the posterior spine exposed. A transthoracic or thoracoabdominal approach is chosen, depending on the level of the HV.

Once the vertebral column is visualized, intraoperative radiographs are taken to confirm levels. Segmental vessels are preserved if possible. After exposure of the HV (Figures 1–3), the disc material proximal and distal to the HV including the disc on the concave side is removed. The superior and inferior endplates of the HV are now prepared to bleeding bone.

The body of the HV is partially preserved keeping part of the posterior vertebral body wall and PLL intact. Transbody screws are inserted into the 2 vertebrae above

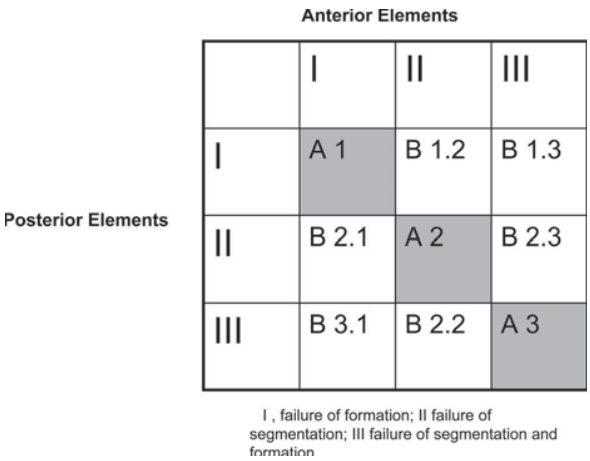

Figure 2. Classification system to take into account mismatch of anterior and posterior elements. Anterior elements of the involved spinal segment are classified first followed by the type of anomaly present posteriorly.

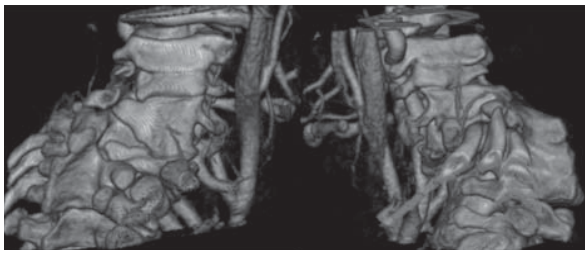

Figure 3. Complex cervicothoracic malformation with hypoplastic vertebral artery.

and 2 vertebrae below the HV. Screws are inserted posterior as possible and parallel to the endplates. Bicortical purchase of all screws is documented by manual palpation of the contralateral side of the spine. Insertion of multiple rib strut grafts or a small cage is facilitated by manual anterior directed pressure over the gibbus. A small bone punch can be used to position the struts as far as possible into the concave disc space.

In young patients the deformity is relatively flexible and elongation of the spinal cord is prevented by the intact PLL and the posterior wall of the HV. A single straight rod is then engaged and the inferior screws are locked. Careful intersegmental compression is then applied using the rib graft as a fulcrum against the body of the HV to obtain coronal correction.

The patient is turned to the prone position and posterior midline incision is made. Onlay bone graft is applied after gentle decortication of the laminas, transverse process, and spinous process, with a high-speed burr. If there is residual kyphosis after anterior instrumentation a single posterior compression rod is added on the convex side. All patients were braced in a thoracolumbosacral orthosis for 6 months (Figures 4, 5).

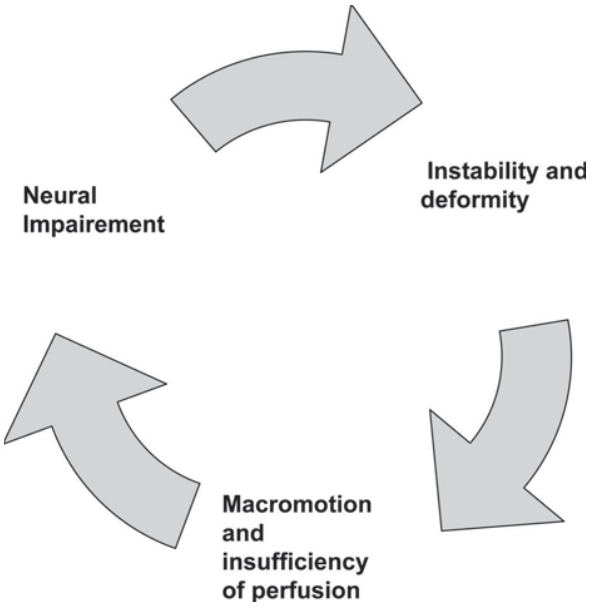

Figure 4. Pathogenesis of neurologic injury in congenital kyphosis.

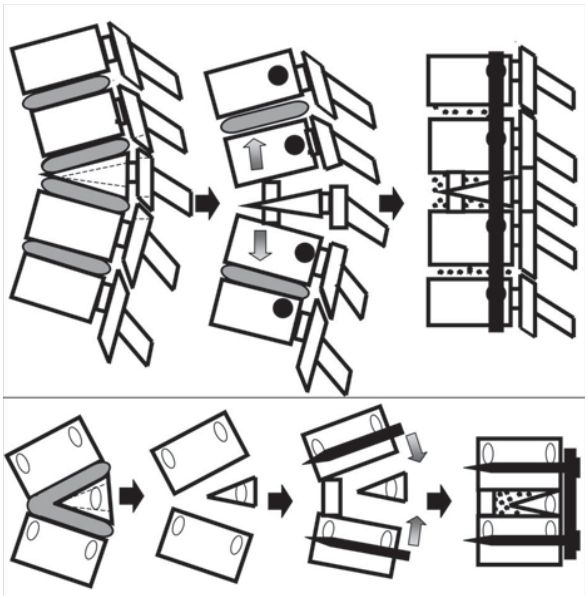

Figure 5. Lateral and AP sequence of correction technique.

■ Results

The average operating time of the procedure was 150 minutes (range, 130–210 minutes). The average blood loss was 180 mL (range, 100–330 mL), equivalent to a mean external blood volume loss of 15% (range, 11%–24%).

Preoperative segmental Cobb angle averaging 34° (range, 0°–58°), was corrected to 18° (range, 0°–31°) at last follow-up. Compensatory coronal cranial and caudal curves were corrected by 50%. The angle of segmental kyphosis averaged 39° (range, 20°–80°) before surgery and 21° (range, 11°–40°) at last follow-up. This represents a 43% of improvement of the segmental kyphosis and a 64% of improvement of the segmental scoliosis at last follow-up.

One case with initial kyphosis of 80° continued to progress and required revision anterior and posterior surgery. There were no neurologic complications or changes in monitored spinal cord potentials.

Late Treatment

Correction of a rigid kyphoscoliotic deformity in the older child is a difficult problem. Neurologic injury is much more likely than in the young child. Preoperative traction is not recommended as it only corrects the more mobile compensatory curves but not the rigid gibbus of the deformity. Elongation of the spinal cord with traction can result in neurologic deterioration.[31]

Anterior spinal release and strut graft insertion followed by posterior instrumented fusion provides reliable results.[31] Variable amounts of vertebral resection are necessary to obtain a satisfactory release and secure placement of the grafts. The posterior HV can be difficult to visualize during an anterior exposure and the spinal cord is compressed against the gibbus. In the authors experience it is usually not necessary to perform a full formal anterior decompression. Lengthening of the an-

terior column with an intact medial column results in indirect decompression of the spinal canal.

The senior author (M.H.H.N.) avoids whenever possible resecting the body of the HV fully to avoid instability and interference with the blood supply at the apex of the deformity.

Multiple strut grafts to fill the angle of the kyphosis effectively build an anterior bridge to support the spine. The most anterior strut should lie at the level of the gravity line but the further anterior these grafts are placed the more likely they are to fracture.[34] Posterior instrumentation is applied by use of compression and leverage. There have been several reports on HV resections in older children with moderate kyphotic deformities.

Shono *et al* reported the results of HV resections with an average kyphosis of 40° through a posterior-only approach. Despite that the anterior column and disc on the concave side could not be fully resected sufficient correction could be achieved.[20] One patient reported persistent dysesthesias in his lower extremities after surgery. Substantial blood loss should be anticipated.[35] Only few studies include patients with severe angular congenital kyphosis.

Shimode *et al* reported the results of circumferential posterior spinal osteotomy on 5 patients with congenital kyphosis. Kyphosis was corrected from 104° to 48° on average. Segmental vessels were clamped before ligation and controlled shortening across the resection gap was performed under frequent spinal cord monitoring. One patient with preoperative neurologic dysfunction improved by 1 Frankel grade and other suffered transient muscle weakness after surgery. The authors concluded that correction with too much shortening is dangerous and considered the procedure acceptably safe.[36] Although these techniques provide effective deformity correction the reviewer feels that they have to be considered high-risk procedures.

■ The Child Presenting With Spinal Cord Compromise

There may be several mechanisms responsible for neurologic dysfunction in patients with congenital kyphoscoliosis. They are as follows:

1. Mechanical instability of the spine, which may result in chronic trauma to the spinal cord,
2. Gradual impingement on the cord,
3. Associated anomalies of the spinal cord.[4]

McMaster reported on 6 patients who presented with significant neurologic deficit. On 3 patients an indirect decompression by partial correction and stabilization was performed. Two patients made a complete recovery and 1 made a partial recovery. On 3 patients on whom a formal anterior or anterolateral decompression was performed, 1 patient was worse, 1 made a partial recovery, and 1 recovered.[31] The question if neurologic decompression should be performed is controversial. Zeller *et al* recommended this procedure while McMaster had good results with circumferential fusion only.[31,37]

Adequate decompression requires removal of the body of the apical vertebra but also frequently resections on adjacent vertebrae. This is difficult to achieve without interfering with segmental blood supply to the spine, which can result in further deterioration.

With increasing familiarity with osteotomy procedures, surgeons have begun to develop techniques for decompression and correction of kyphotic deformities through an all-posterior approach.[20,36,38,39] One-level closing wedge osteotomies restrict sagittal correction to 25° in the thoracic spine and 35° in the lumbar spine.[40] Correction of greater angles requires shortening of the spinal cord and seems dangerous in the presence of compromised neural tissue.

We believe that in evolving paraplegia stability through circumferential fusion is essential for neurologic recovery. Stability rather than correction of deformity is paramount. In a recent review of 52 patients who had surgical treatment of severe angular kyphosis, of whom 17 had a myelopathy, anterior decompression, and fusion with posterior fusion and no attempted correction of deformity found that this was a safer modality with better results than previously reported.[41]

■ Conclusion

Congenital kyphosis remains a challenging problem for the deformity surgeon. Better understanding of the pathophysiology of neurologic dysfunction in congenital kyphosis may help us in the future to develop safer treatment methods. Morphologic classification will need to evolve to study the natural history of more complex anomalies. A new technique to treat young children is described in which the HV is partially preserved, reducing the intraoperative blood loss, avoiding dural manipulation, and minimizing the risk of interfering, with local blood supply. The preserved HV is the ideal vascular graft to achieve solid fusion.

■ Key Points

- In progressive congenital kyphosis, early diagnosis and aggressive surgical treatment are mandatory for a successful result.
- Early treatment minimizes the risks of surgery.
- Anatomic and physiologic pitfalls in the treatment of congenital kyphosis are discussed.
- Anterior instrumented fusion of congenital kyphosis provides sagittal and coronal correction in very young children with low risk of complications.

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SPINE Volume 26, Number 23, pp E539–E541

Anterior Instrumented Fusion for Thoracolumbar Kyphosis in Mucopolysaccharidosis

Samir S. Dalvie, MS, M. H. H. Noordeen, FRCS, and Ashok Vellodi, FRCP

Study Design. A case series of seven children who had a thoracolumbar gibbus related to mucopolysaccharidosis treated with anterior instrumentation were reported retrospectively.

Objective. To describe a new technique for treating progressive thoracolumbar kyphosis in children with mucopolysaccharidosis.

Summary of Background Data. Management of this condition is not well represented in the literature. Isolated reports on the surgical management of this disorder appear, but there is no previous report of correction performed anteriorly.

Methods. Seven patients underwent anterior instrumentation for correction of a thoracolumbar gibbus not arrested by brace treatment. Preoperative kyphosis ranged from 42° to 64° (average, 52.5°). Data on all seven patients were collected prospectively. The technique and its principles are described.

Results. A good correction of the kyphosis was obtained, with postoperative angles of 3° to 29° (average, 15°), and maintained through the follow-up period. There were no complications from the procedure.

Conclusion. Anterior instrumented correction and fusion of the spine is effective in treating thoracolumbar kyphosis associated with mucopolysaccharidosis. [Key words: kyphosis, mucopolysaccharidosis, spinal instrumentation] **Spine 2001;26:E539–E541**

The mucopolysaccharidoses are a group of inherited metabolic disorders characterized by abnormal storage of mucopolysaccharides in various tissues of the body. In the past, most patients with this disorder did not survive beyond early childhood. Patients with Morquio Type A disease died of sudden atlantoaxial dislocations, and those with Hurler, Hunter, and Maroteaux-Lamy disease died of systemic complications. However, there has been increased survival of these patients related to early "prophylactic" fusion of the occipitocervical complex and bone marrow transplantation for the systemic disorders. This has resulted in a presentation of problems arising from dysplasia of the thoracolumbar spine in the surviving children.

The vertebral bodies of these patients are deficient anteriorly, and thus are flattened, beaked, or notched.[3,4] The intervertebral discs are thick and bulging, often larger than the bodies. Thus, in time, the thoracolumbar spine collapses into a long-segment kyphosis. If confined to a short segment, this leads to a progressive localized kyphos with the appearance of an anterolisthesis. This deformity is progressive, leading to poor trunk balance and possible neurologic deterioration.[1] The gibbus is quite flexible in childhood, but with progression becomes increasingly rigid.

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Methods

Seven children with a previous diagnosis of mucopolysaccharidosis were treated with an anterior instrumented correction and fusion for a progressive thoracolumbar kyphosis (Table 1). These children had dysplastic vertebrae and a " round-back" kyphosis with a T12 or L1 apex. Cobb angles for the kyphosis ranged from 42° to 64° (average, 52.5°) in the erect lateral radiographs. The kyphosis was flexible, as demonstrated by the difference in the kyphosis noted on the magnetic resonance scans obtained with the patient in the supine position, and in the standing lateral radiographs. All children with Morquio Type A disease and thoracolumbar gibbus were treated with a molded thoracolumbar spinal orthosis brace. However, this was ineffective in halting curve progression in these seven children. All children were ambulant at the time of surgery.

Four of the children (patients 1 to 4) presented with neurologic deterioration manifesting as poor coordination and clumsiness of gait resulting from motor weakness. In addition, patient 1 had bladder dysfunction, and patient 3 had sensory disturbances. These children had magnetic resonance scans of their entire spine. These scans showed bulging discs causing canal compromise over the kyphotic area. Differential evoked potentials were obtained, and these confirmed the level of neurologic dysfunction to be at the thoracolumbar spine.

Indications for surgery were progression of the curve despite bracing and neurologic deterioration. Surgery was performed to improve and restore trunk balance and stability and to prevent or improve neurologic dysfunction. Another benefit of surgery was avoidance of long-term brace wear.

Technique. The number of levels to be included in the fusion is determined. These include all the vertebrae that form part of the curve. A few additional wedged vertebrae at the thoracic level may be included. Some patients have a single-level problem similar to a congenital kyphosis resulting from anterior failure of formation. These patients require instrumentation above and below the deficient vertebra.

An extensile thoracolumbar approach is performed through the appropriate rib bed in the thorax, extending retroperitoneally into the abdomen and connected by peripheral incision in the diaphragm. The bulging discs are excised to the posterior longitudinal ligament, and the cartilaginous endplate are excised on either side. The vertebral body is defined, with care taken to reflect the psoas posteriorly in the lumbar spine. The vertebral bodies typically are wedge-shaped, giving a larger

Table 1. Results of Treatment With an Anterior Instrumented Correction and Fusion for a Progressive Thoracolumbar Kyphosis in Seven Children With a Previous Diagnosis of Mucopolysaccharidosis

Patient	Age (years)	Gender	Diagnosis	Previous Treatment	Preoperative Kyphosis (°)	Postoperative Kyphosis (°)	Fusion Levels	Follow-up Period (months)
1	10	M	Morquio A (MPS IV)	OC Fusion at age 4	59	6	T10–L3	36
2	8	M	Morquio A (MPS IV)	OC Fusion at age 5	45	3	T7–L4	19
3	13	F	Morquio A (MPS IV)	OC Fusion at age 8	42	8	T10–L3	11
4	7	F	Morquio A (MPS IV)	OC Fusion at age 5	44	24	T12–L2	9
5	14	M	Martoux-Lamy (MPS VI)	BMT at age 8	55	17	T10–L3	16
6	4	F	Martoux-Lamy (MPS VI)	BMT awaited	64	29	T10–L3	8
7	5	M	Hurler's (MPS I)	BMT at age 4	59	18	T11–L2	5

MPS = mucopolysaccharidosis; OC = occipitocervical fusion; BMT = bone marrow transplant.

area for screw placement. A 4-mm pediatric screw is placed in the middle, with care taken to ensure bicortical purchase.

After instrumentation of all levels, a 4.75-mm solid rod is inserted, engaging the screw heads sequentially, thus correcting each level. A three-point corrective force is used to aid the correction. The screw heads then are distracted gently to open the disc spaces farther.

Interbody fusion is performed by placing struts fashioned from the excised rib. Interestingly, the ribs of these patients are very stout. The graft is loaded by compressing the screws gently before final torque-tightening of the top nuts. All the children are braced after surgery in a total contact thoracolumbar spinal orthosis brace for 6 months. They are allowed to ambulate.

■ Results

All the patients had a good correction of kyphosis (Table 1) Postoperative Cobb angles ranged from 3° to 29° (average, 15°). The average operating time was 1 hour and 55 minutes (range, 1 hour 25 minutes to 2 hours 35 minutes), and the average blood loss was 150 mL (range, 125–250 mL). The longest follow-up period (patient 1; Figure 1) at this writing is more than 3 years. There has been no loss of correction in any of the patients.

All four patients with a neurologic deficit recovered their baseline neurology after surgery. Patient 1 had improvement in bladder function, beginning at 3 months, with motor improvement beginning at 6 months and completed by 18 months. The three other patients had early onset of recovery, with complete recovery by 6 months. All the children experienced restoration of their previous gait. No significant postoperative complications from this procedure occurred in these patients.

■ Discussion

Children with Morquio Type A disease are significantly handicapped because of their short stature and multiple skeletal dysplasias. A progressive thoracolumbar gibbus further adds to their disability. These patients require

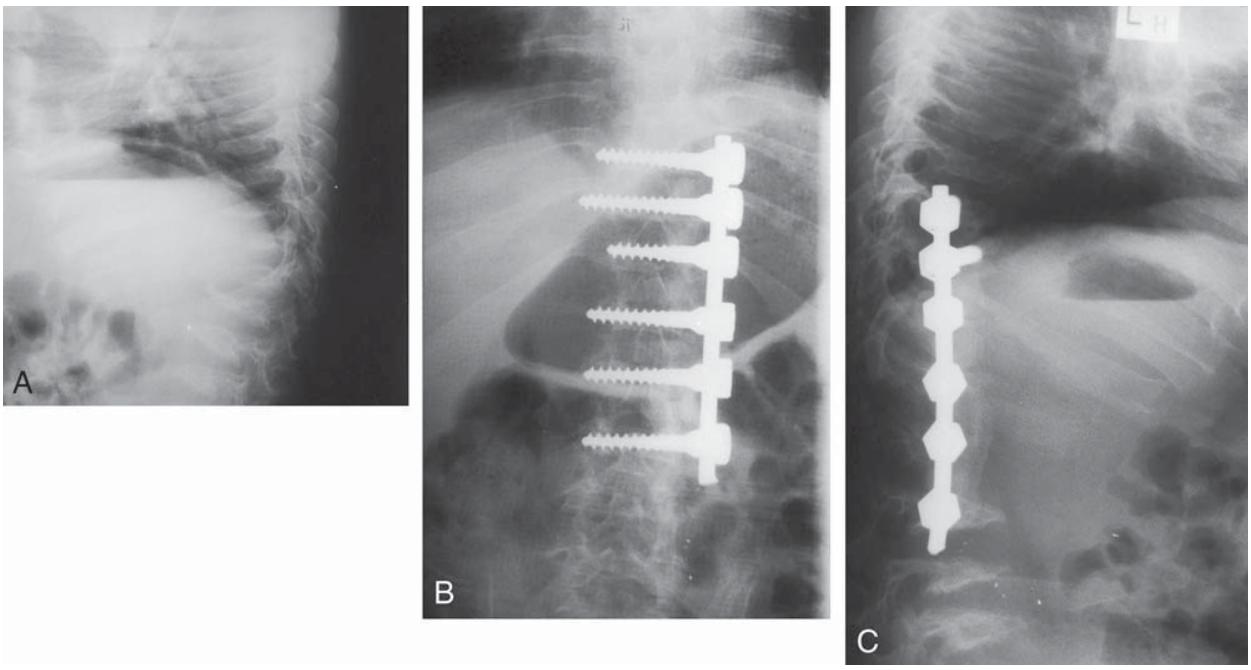

Figure 1. **A,** Preoperative radiographs of patient 1. **B,** Anterioposterior radiographs showing the instrumentation in place. **C,** Radiograph at 3 years, showing correction of the gibbus with the instrumentation and a solid bony fusion.

correction and stabilization of their thoracolumbar gibbus to preserve trunk balance, prevent further progression, and avert or reverse neurologic deterioration. In the past, they were treated with prolonged bracing or *in situ* posterior fusion.[4] However, it is well recognized that a kyphos in a dwarf must be fused early. Now that the life expectancy of these patients is improving, it is imperative that correction at the thoracolumbar spine be obtained and maintained.

The authors have used the described anterior technique because they believe it offers certain distinct advantages:

1. There is an opportunity for anterior decompression by excision of the bulging discs before correction of the kyphosis.

2. The number of levels necessarily included in the fusion is always less than required posteriorly, thus sparing lumbar levels, which is of particular importance in these patients because of the skeletal dysplasia.

3. The posterior elements in these children are not strong enough to hold the instrumentation. Furthermore, there is canal stenosis because of soft tissue deposition, making sublaminar wires unsafe.

4. The interbody fusion obtained is of an excellent quality, using the maximum fusion area loaded in compression.

5. Anterior surgery can be performed quickly and dissects fewer muscle planes.

Intraoperative technical difficulties can be encountered during screw placement and correction. The vertebral bodies are very small, and great care must be taken to ensure central placement after the exact dimension of the bony corpus is determined. If the correction maneuver places excessive stress on the implants, they may cut through the bone. Thus, the correction maneuver must include an external corrective force.

In these seven patients, the authors intended to instrument 41 vertebrae, but were successful in instrumenting only 37. Anterior column stability in cases of fixation loss is restored by strut-grafting the intervertebral spaces.

Traditionally, kyphosis is corrected using posterior instrumentation, although anterior surgery is used for a release or arthrodesis. To the authors' knowledge, this is the only report describing anterior instrumentation for correction of a kyphosis. The short-term results of this procedure have been satisfactory, but the issue of late changes at nonfused levels will be clarified only after a longer follow-up period.

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Address reprint requests to

M. H. H. Noordeen, FRCS
Great Ormond Street Hospital for Children
London WC1N 3JH
United Kingdom
E-mail hilali.noordeen@virgin.net

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Spondylocostal Dysostosis

Thirteen New Cases Treated by Conservative and Surgical Means

Marco Teli, MD, Harish Hosalkar, MD, Inder Gill, FRCS, and Hilali Noordeen, FRCS

Study Design. Prospective assessment of a cohort of patients affected by spondylocostal dysostosis.

Objective. To report on the results of conservative and operative management of spondylocostal dysostosis and, based on this, to propose an assessment and treatment protocol for the condition.

Summary of Background Data. Spondylocostal dysostosis and spondylothoracic dysostosis are subtypes of Jarcho-Levin syndrome, a hereditary condition manifested by vertebral body and related rib malformations. Mortality prevails in spondylothoracic dysostosis because of more severe respiratory compromise.

Methods. Details of prenatal and postnatal diagnosis, history, and management of 13 patients with spondylocostal dysostosis are presented. All patients were treated postnatally with repeated chest physiotherapy. Two patients refractory to conservative treatment underwent surgical intervention: the first had a chest wall reconstruction *via* a latissimus dorsi flap, the second a posterior spinal instrumented fusion for progressive scoliosis.

Results. Prenatal ultrasound in 4 of 13 cases showed full details of vertebral and rib anomalies. Thoracic and lumbar hemivertebrae were most common, leading to congenital scoliosis in 10 of 13 cases. A number of extraskeletal abnormalities were also identified. At an average follow-up of 4.5 years, the survival rate was 100% with a remarkable decrease of the rate of respiratory complications. Surgical treatment in selected cases led to satisfactory results.

Conclusions. Prenatal diagnosis of spondylocostal dysostosis allows exclusion of spondylothoracic dysostosis and aids genetic counseling in quantifying the risk to siblings. Postnatally, prompt management of these patients with physiotherapy leads to prolonged survival. Surgical intervention may then be indicated to stabilize chest wall or spine deformities, with promising results.

Key words: congenital scoliosis, spondylocostal dysostosis, operative treatment. **Spine 2004;29:1447–1451**

In 1938, Jarcho and Levin described a syndrome affecting individuals with short neck and short-trunk dwarfism resulting from multiple vertebral and associated rib anomalies. In Jarcho-Levin syndrome (JLS), hemivertebrae, butterfly vertebrae, and unsegmented, hypoplastic vertebrae may almost affect the entire thoracic and lumbar spine at alternate levels.[1] Cervical spine involvement may be in the form of hemi- and unsegmented vertebrae, causing shortness of neck. The ribs can vary in number and shape often showing fusion, primarily near the costovertebral ends. Multiple rib and vertebral anomalies may determine scoliosis, kyphosis, and pectus deformities. The widespread distribution of the vertebral anomalies and their association with rib anomalies differentiate JLS from congenital scoliosis.[1] In 1978, Solomon *et al* classified cases of JLS into two types: spondylothoracic dysotosis (STD) and spondylocostal dysotosis (SCD).[2]

Individuals with STD have vertebral anomalies and severely deformed and fused ribs, resulting in the "crablike" appearance of the chest on plain radiographs. The inheritance pattern is autosomal dominant. Patients affected by STD have a higher fatality rate as a result of posterior tethering of the ribs, leading to progressive thoracic restriction with growth.

Individuals with SCD have vertebral anomalies and intrinsic rib defects or malformations (hypoplasia) of varied patterns. The inheritance pattern is either autosomal dominant or recessive, the latter being described as the most aggressive form. Patients with SCD survive more often than those with STD despite gross thoracic abnormalities, possibly because the lungs are not restricted. SCD has a reported prevalence of $0.25/10^5$.[3]

The aim of this study is to outline a protocol for the assessment and treatment of SCD based on the results of conservative and surgical treatment in 13 new cases of this syndrome and the pertinent literature. We also describe the results of surgical treatment in 2 of our patients, including the use of the latissimus dorsi flap for costal defect reconstruction in one and instrumented posterior spinal fusion in the other.

To the best of our knowledge, the present series is the largest ever reported on the condition. From 1938 to date, 41 cases of SCD and 20 cases of STD have been described in the English medical literature, the largest single series reporting on no more than 5 and 6 patients, respectively.[2]

Materials and Methods

We performed a prospective follow-up study of 13 pediatric patients affected by SCD and treated at two Pediatric Orthopedic Institutions.

Five patients had consanguineous parents. Four patients were seen from birth and prenatal ultrasound diagnosis had been made in all of them. All patients had full spine and chest plain radiographs; six had ultrasound[4] evaluation of segmental defects and three had full spine MRI scans. A thorough clinical and radiologic assessment was done in all cases for cardiovascular,[5] genitourinary, parenchymal organ defects and neural tube defects.[6]

Our protocol in managing these patients postnatally was based on chest physiotherapy (CPT) and regular clinical eval-

From the Great Ormond Street Hospital for Sick Children, London, UK. Dr. Hosalkar is formerly with Wadia Children's Hospital, Mumbai, India.
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Address correspondence to Marco Teli, MD, Residence Grancia, flat 112, 7, Via Tevere, 20020 Lainate, Italy; E-mail: marcoteli@hotmail.com

Table 1. Patients' Details

Case No.	Age First Seen	Sex	Consanguinity	Prenatal Diagnosis	Vertebral Defect Site	Associate Anomalies	No. of Absent Ribs R	No. of Absent Ribs L	Rib Fusion	Rate of LRTI per Month Pre-Rx	Rate of LRTI per Month Post-Rx
1	Newborn	M	+	+	TL	ASD	5	0	–	3	1:3–6
2	Newborn	F	–	–	T	Jaundice, left renal agenesis, clubfeet	4	0	–	3–4	1:3
3	6 mo	F	+	–	T + L	Hydrocephalus, spina bifida, bifid uvula, supemumerary nipple	1	5	7,8,9 (L) 4,5 (R)	5–6	1:2
4	3 mo	M	–	–	TL	Sprengel's deformity	3	0	–	1–2	1:4
5	2 mo	F	–	+	T	Polycystic kydney	0	2	–	3	1:3
6	5 mo	F	+	–	T	–	2	0	5,6,7 (R)	4–5	1
7	5 mo	M	–	–	CT + TL	ASD, supemumerary nipple	4	0	6,7 (R)	3–4	1:3
8	Newborn	M	–	+	TL	ASD, diaphragmatic hernia	0	4	–	1	1:8
9	2 mo	M	–	–	T	Hydrocephalus, spina bifida, clubfeet	0	3	–	4–5	1
10	Newborn	F	+	–	TL	VSD, left renal agenesis	4	0	–	3–4	1:3
11	1 mo	M	+	+	T	Cleft palate, supemumerary nipple, DDH	2	3	–	4	1:3
12	3 mo	F	–	–	CT + TL	Jaundice, clubfeet	4	0	–	2–3	1:3
13	10 yr	M	–	–	T	–	3	3	6,7 (R)	4	1:2

CT = cervicothoracic; T = thoracic; TL = thoracolumbar; L = lumbar; VD = vertebral defect; ASD = atrial septal defect; VSD = ventricular septal defect; DDH = developmental dysplasia of the hip; LRTI = lower respiratory tract infection.

uations. Chartered physiotherapists treated patients on a daily basis in hospital and at weekly intervals after discharge, until improved clinical conditions allowed monthly appointments. Treatment consisted of a modified CPT routine followed by breathing games to perform diaphragmatic breathing in infants, and of positive expiratory pressure therapy plus chest wall oscillation in children.

The frequency of lower respiratory tract infections (LRTIs) was recorded. In the clinical setting of an LRTI, *i.e.*, worsening general conditions, oxygen desaturation, raised white cell count, and positive chest radiograph, patients were admitted to hospital to receive broad-spectrum intravenous antibiotic coverage. Antibiotics were then targeted to sputum culture results and discontinued at the reversal of the above clinical indexes.

Bracing was used in those cases warranting treatment of a progressive spinal deformity, provided it was tolerated by patients and on the grounds of a satisfactory pulmonary function.

A rib defect reconstruction procedure was performed in patient 8 (Table 1) who had frequently recurrent LRTIs, paradoxic chest wall movements, and a diaphragmatic hernia. Ultrasound showed herniation of the lower left lung lobe with the diaphragm and the upper pole of the spleen with respiratory movements. The rib defect was repaired with an autologous, vascularized latissimus dorsi flap to stabilize the chest wall and contain the lung and spleen herniation.[7]

Posterior instrumented spinal fusion was done in patient 13 (Table 1)[2,8] who was referred to our service at 10 years of age for progressive scoliosis. At the time of the first observation, he already had features of lower thoracic paraplegia,[2] restrictive lung disease, and recurrent and severe chest infections. His sitting balance had been deteriorating because of increasing spinal deformity and pelvic obliquity. Figure 1 shows his preoperative plain radiographs. Because of his poor general health, posterior instrumented spinal fusion with limitation of fusion levels from the lower thoracic vertebrae to the pelvis was the only management option at that time. Figure 2 shows his 3-year follow-up films, with adding-on of the upper thoracic curve and satisfactory maintenance of the lower spinal and pelvic deformity. At this time, his general health had improved to the stage that proximal extension of the spinal fusion was deemed feasible.

■ Results

There were 7 males and 6 females. Patient age varied from 1 day to 10 years when first seen. Final follow-up ranged from 26 to 72 months, averaging 56 months. No patient was lost to follow-up.

The nature of the vertebral defects was delineated in all cases. One or more vertebral anomalies were present in all patients and included all together 1 hypoplastic sacral vertebra, 3 absent thoracic vertebrae, 7 butterfly vertebrae (4 thoracic, 3 lumbar), and 18 hemivertebrae. Hemivertebrae, the most common vertebral anomalies, were more commonly located in the thoracic and thoracolumbar region than in the cervical region. Congenital scoliosis was identified in 10 of 13 patients (75%): average anteroposterior Cobb angles varied from 23° at the beginning of the observation to 38° at the latest follow-up.

Both unilateral (10 cases) and bilateral (3 cases) rib defects were noted. Rib fusion was found in 3 cases. Rib anomalies included posterior fusion, absence, and irregular size and shape.[9]

The extraskeletal associated anomalies[10] included supernumerary nipples (3 cases), clubfoot deformity (3

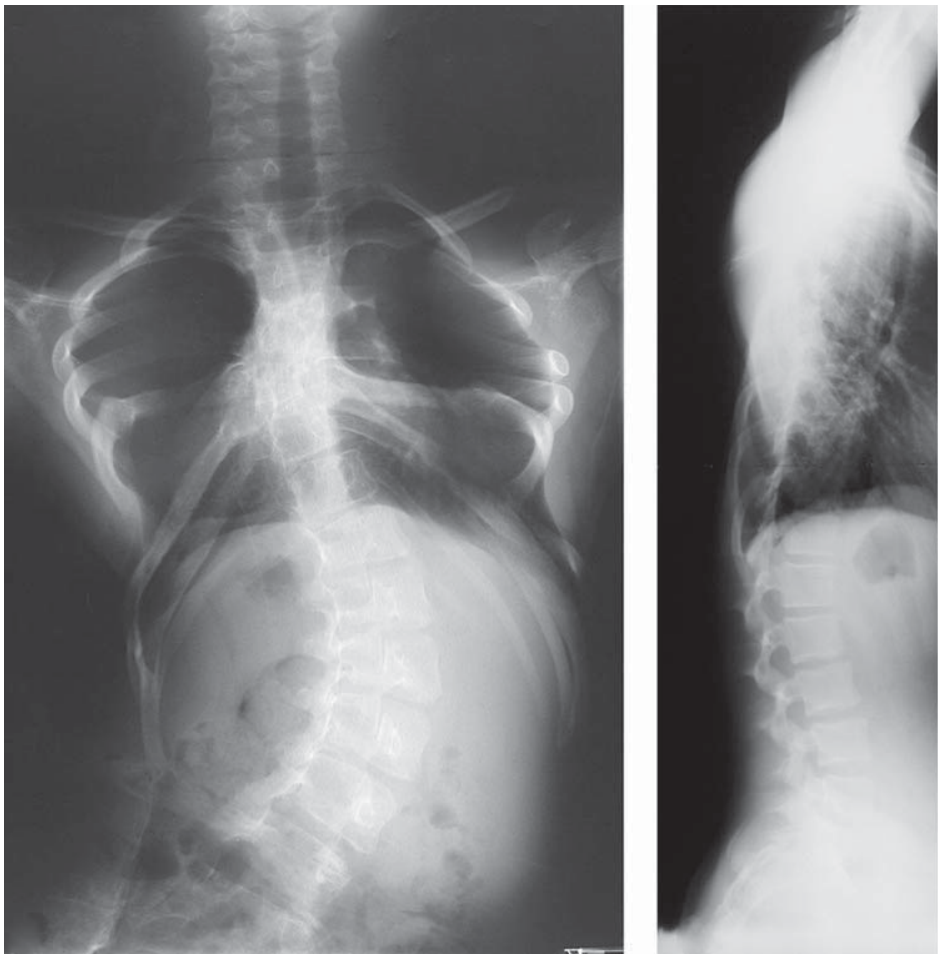

Figure 1. Case No. 13 preoperatively, aged 10 years. PA + lateral sitting films.

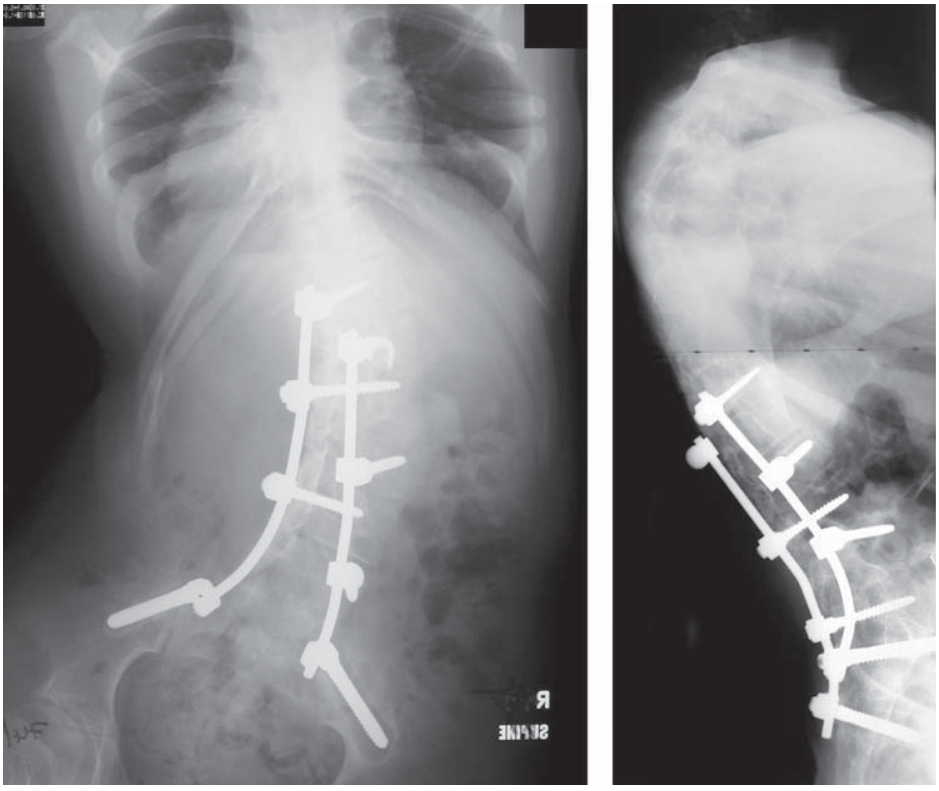

Figure 2. Case No. 13 2 years after posterior spinal fusion, aged 13 years. PA + lateral sitting films.

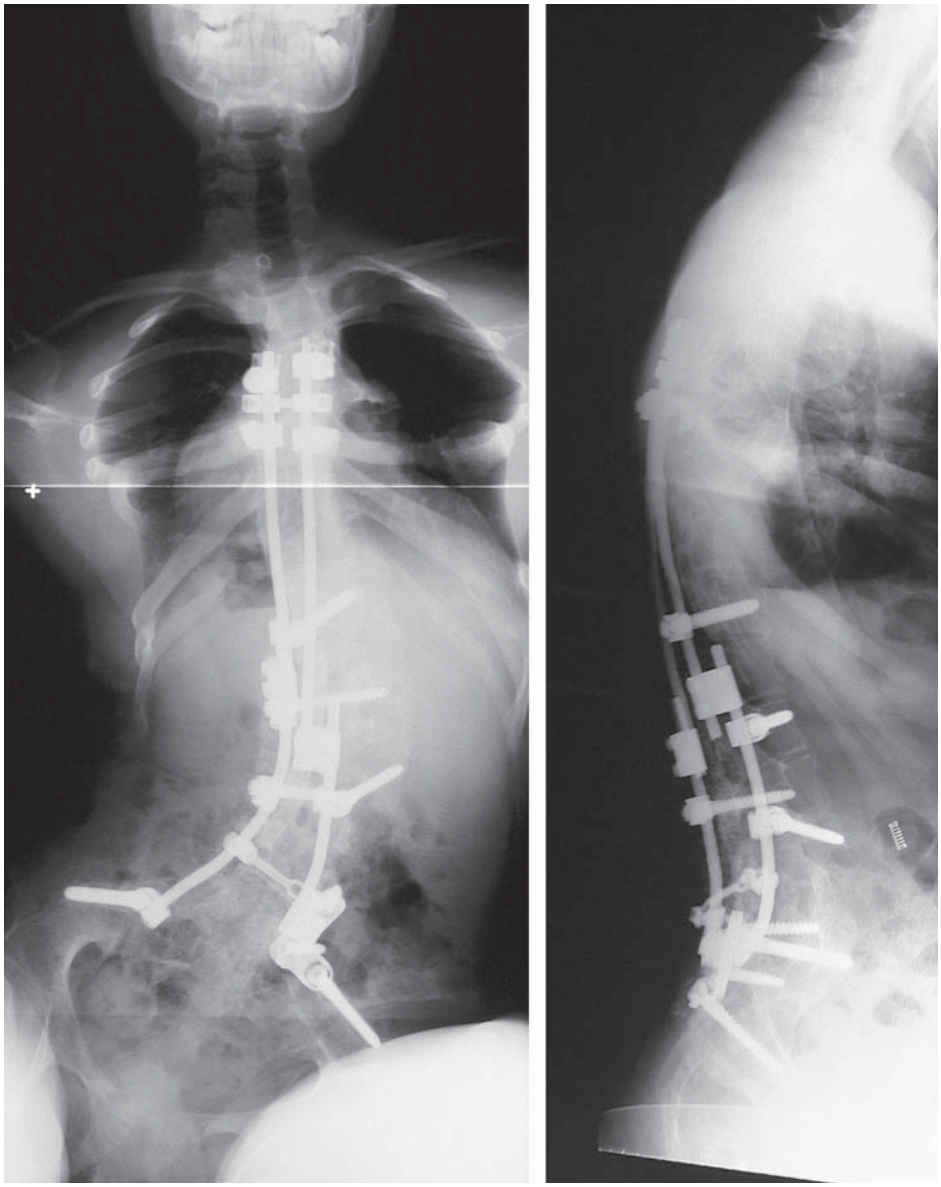

Figure 3. Case No. 13 2 years after thoracic extension of spinal fusion, aged 15 years. PA + lateral sitting films.

cases), Sprengel's deformity (1 case), strabismus (1 case), bifid uvula (1 case), cleft palate (1 case), congenitally dislocated hip (1 case), neonatal jaundice (2 cases), myelomeningocele and hydrocephalus (2 cases), renal anomalies (4 cases), atrial septal defect (1 case), and ventricular septal defect (1 case).

The presenting complaints in all patients were recurrent LRTIs, with an average of 3 episodes per month (range, 1–6), poor general condition, and failure to thrive. After birth, patients were hospitalized on average once a month (range, 0.3–4) mostly for treatment of LRTIs. Mechanical respiratory support was necessary once every three admissions to hospital for an average of 2 days (range, 1–5) during hospitalization. Mean height and weight growth curves were under the fifth and around the 10th centiles, respectively, before treatment.

This picture was reversed at follow-up, with a reduction of the incidence of LRTIs to an average of 1 episode every 3 months (range, 1:8 to 1 a month) and hospitalization to an average of once every 6 months (range, 1:8 to 1:3 months). We observed a mean reestablishment of the weight curves to the 25th centile and fairly stable height curves just under the fifth centile.

Patient 8, treated with autologous rotation latissimus dorsi muscle flap, at follow-up reveals containment of herniation, a supple scar, and a static rib defect. His pulmonary function improved readily after surgery[7] and currently, 24 months after surgical intervention, he experiences 1 episode of respiratory infection every 8 months. His paradoxical chest movements have stopped and his diaphragmatic hernia is well contained.

The follow-up for patient 13 is now 2 years following the revision procedure. Currently, the patient is pleased by his sitting balance and by the absence of uneven pressure areas or spinal instrumentation prominence. Although he his still frequently admitted to our Hospital

for respiratory care, the rate of his chest infections has fallen from 4 a month to 1 a month. Figure 3 shows satisfactory fusion and absence of instrumentation failure in his latest follow-up spine radiographs.

■ Discussion

SCD is a rare[3] disorder of vertebral and rib formation of unknown etiology. In our series, there was history of maternal diabetes in one of the patients only, confirming previous reports of low incidence of diabetic embryopathy in JLS.[3,8] Regarding its pathogenesis, the vertebral anomalies observed in this syndrome could be secondary to a developmental disturbance at the fourth to eight week of fetal life. At that time, the multiple centers of chondrification formed about the notochord unite to form a complete cartilaginous pro-vertebra composed of centra in the vertebral body and transverse, articular, and spinous processes. Failure of these chondrification centers to develop or unite could result in the development of hemivertebrae and butterfly vertebrae. The anomalous rib development is apparently secondary to the vertebral malformations.[11]

Second trimester prenatal diagnosis[6] was done in 4 of our 13 cases. A deformed spine and incompletely formed ribs were demonstrated in all cases. Prenatal diagnosis is thus feasible and important to detect the more aggressive and lethal form of JLS, *i.e.*, STD.[1]

Genetic counseling is likewise fundamental. In the recessive form of SCD, the risk to siblings of an affected child is 25%. In the dominant form, this risk is negligible but rises to 50% for the offspring of a long survivor.[1,12]

The survival rate seen in our 2- to 6-year follow-up was 100%. This applies to a series of patients affected by both the dominant and the recessive form of SCD. The recessive form of the disease resulted from a number of patients being born to consanguineous parents (Table 1).

The outcome of conservative treatment was noteworthy in all cases, especially considering the dramatic decrease in the incidence of respiratory infections. Both CPT and hospitalization had to be repeated at regular intervals. Noncompliance was noted in some cases, especially after the reestablishment of the weight curves observed with improved general conditions.

Prolonged survival thus leads to issues of chest wall or spinal deformity management. Chest wall reconstruction has been performed by using either polypropylene mesh[13] or, in our case, autologous muscle flap[7] to improve chest wall compliance. The results have been so far favorable. Spinal fusion in our patient adds to the few other relative descriptions[2,8,14] and eventually succeeded in stabilizing a progressive scoliotic deformity with pelvic obliquity.

■ Conclusion

Based on the above findings we feel that assessment and treatment of SCD should be based on the following guidelines:

- Early prenatal ultrasound diagnosis of SCD is feasible and genetic counseling thus plays an important role.
- In view of the concomitant nonskeletal anomalies, a thorough clinical and radiologic screening of every patient with these skeletal defects is essential.
- Early and repeated CPT forms the mainstay of treatment.
- Prolongation of life span with CPT opens up new possibilities for surgical intervention for reconstruction or deformity correction.
- Surgical intervention for correcting costal defects and spinal surgery with instrumentation seem to be beneficial in these cases.

■ Key Points

- Spondylocostal dysostosis is a form of hereditary scoliosis amenable to prenatal ultrasound diagnosis.
- A mainstay of treatment is conservative with chest physiotherapy plus prevention and treatment of respiratory tract infection.
- Surgery is reserved to selected cases of untreatable respiratory deterioration or scoliosis progression.

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Chapter 6

NEUROMUSCULAR SCOLIOSIS

Advances in Surgical Techniques for EOSD

Paper: 15

Segmental posterior spinal instrumented correction and fusion is the current 'standard of care' for treating scoliosis secondary to neuromuscular scoliosis (NMS). I undertook a retrospective comparative clinical study (Level of Evidence [LoE] III: Therapeutic), evaluating the clinico-radiological results of second vs. third generation instrumentation systems, in matched cohort of patients, (i.e. Luque-trolley or Unit rods vs. Cotrel-Dubosset and segmental instrumentation systems). All children were under twenty one years old, and were operated between 1995 and 2000. The study group comprised of fifty six patients (twenty nine males & twenty seven females) who were closely followed-up for a mean of fifty two months (range: twenty four to eighty five months). The minimum follow-up was two years in all patients. The etiologies included cerebral palsy (36), Duchenne's muscular dystrophy (8), and miscellaneous (12), which comprised of Friedreich's ataxia, Spina bifida, Spinal muscular atrophy, Syringomyelia, and Poliomyelitis.

NMS is characterized by the highest incidence of complications amongst all spinal deformities. Efforts are underway to reduce complications without a compromise in clinical or surgical results. This study was one such effort from my end wherein I had convincingly showed that there were reduced complication rates with use of the third generation instrumentation systems. This was also one of the first studies that had objectively evaluated the cumulative benefit, using a validated outcome measure. The overall incidences of major and minor complication in this series were ten percent (7/56) and sixteen percent (9/56) respectively. There were only two deep, and one superficial, wound infection. There was one death, unrelated to surgery in this series (progressive cardiomyopathy in Friedreich's ataxia).

Neuromuscular scoliosis treated by segmental third-generation instrumented spinal fusion.
Teli M, Elsebaie H,Biant L, Noordeen H. J Spinal Disord Tech. 2005 Oct; 18(5): 430-8.
PMID: 16189456

Advances in Surgical Techniques for EOSD

Paper: 16

The most common orthopaedic manifestation in neurofibromatosis-1 is scoliosis. I had published a review article in the European Spine Journal summarizing all the salient findings and treatment challenges in this unique group of patients. The scoliosis could be broadly divided into dystrophic and non-dystrophic types. NF-1 scoliosis is notorious for psuedoarthrosis. There may also be co-existent dural ectasias and dumbbell-shaped neurofibromas. The dystrophic curves are very rigid, and commonly present with multi-planar deformities (i.e apex of kyphosis and scoliosis to be at different levels). Bracing is often unsuccessful, warranting aggressive surgical intervention. Implant failure secondary to pseudoarthrosis is also common. BMP (i.e. bone morphogenetic protein) is often used, especially in revision surgeries, to achieve a solid arthrodesis.

I have extensive surgical experience in treating all types of spinal deformities by growth sparing and fusion surgeries. The use of magnet driven growing rods has been an innovative concept and boon to such children presenting with severe or rigid deformities at very young age (under five years old). Early surgery should be offered for dystrophic curves when the Cobb angle is less than twenty degrees. Kyphosis, when seen, is most common in the cervical spine, and may cause a neurodeficit. Atlanto-axial instability should also be ruled out in pre-op assessment, prior to subaxial spinal surgical fixation and fusion.

Spinal deformity in neurofibromatosis type-1: diagnosis and treatment. Tsirikos AI, Saifuddin A, Noordeen MH. Eur Spine J. 2005 Jun; 14(5): 427-39. E-pub 2005 Feb 15. PMID: 15712001

Advances in Surgical Techniques for EOSD

Paper: 17

Duchenne's muscular dystrophy (DMD) is characterised by muscle dysfunction, and is inherited as an X-linked recessive disorder (which is lethal in boys). The pathognomonic feature is the absence of dystrophin, an integral membrane protein responsible for the contractility of the skeletal, cardiac, and smooth muscles. Affected children have mild mental retardation with an intelligent quotient (IQ) of one standard deviation (SD) below the mean. DMD patients also develop progressive rapid decline in pulmonary function and Many surgeons have also observed increased intra-operative bleeding in DMD.

I was one of the first authors to formally document the increased bleeding tendency objectively, in a series of forty eight out of 102 patients with DMD treated with first and second generation instrumentation systems operated over a decade (1983 to 1993). The diagnosis of DMD was histologically proven by a quadriceps biopsy in forty one patients (85 percent). There was a statistically significant increase in blood loss compared to spinal muscular atrophy (SMA) and scoliosis secondary to other neuromuscular etiologies. The possible causes and hypothesis proposed were:

i) Intrinsic abnormality of smooth muscles affecting the blood vessels' contractility

ii) Absence of dystrophin in skeletal and smooth muscles, causing excessive bleeding

This is a key contribution to the spinal community, in an attempt to understand the pathophysiology of DMD and emphasise the importance of meticulous attention to haemostasis in such high-risk patients while performing scoliosis correction.

Blood loss in Duchenne muscular dystrophy: vascular smooth muscle dysfunction? Noordeen MH, Haddad FS, Muntoni F, Gobbi P, Hollyer JS, Bentley G: J Pediatr Orthop B. 1999 Jul; 8(3): 212-5. PMID: 10399127.

Advances in Surgical Techniques for EOSD

Paper: 18

Multi-modal neuromonitoring (SSEP, SCEP and TcMEP) are now the standard of care in spinal deformity corrective surgery for all etiologies. However, neuromuscular scoliosis (NMS) constitutes a special category of patients who have a high incidence of false positive and true negative signals. I was one of the early pioneers in using multi-modal sensory monitoring, in an attempt towards making these surgeries as safe and reliable as possible. I have published a review article summarising the role of spinal cord monitoring for NMS. I had also described a novel technique of obtaining reliable somatosensory evoked potentials (SSEP) and spinal cord evoked potentials (SCEP) signals, using epidural electrodes. The fifty percent rule (i.e. the fall in amplitude by more than fifty percent from baseline) correlated very well with impending spinal cord injury (eighty seven percent sensitivity and forty four percent specificity), and has been universally accepted in the contemporary era.

The operative step of passing sublaminar wires was found to be at the highest risk of impending spinal cord injury. Other factors that may influence neuromonitoring signals are:

i) Hypotensive anaesthesia,

ii) Hypothermia, and

iii) Anaesthetic agents administered.

Spinal cord monitoring in neuromuscular scoliosis. Tucker SK, Noordeen MH, Pitt MC: J Pediatr Orthop B. 2001 Jan; 10(1): 1-5. Review. PMID: 11269804

Advances in Surgical Techniques for EOSD

Paper: 19

I reviewed ninety nine patients with NMS treated with Luque-Galveston rods and sub-laminar wires, using SSEPs under supervision of Prof. G Bentley in the mid 1990s. It is a well-known fact, from numerous published studies, that neuromonitoring for neuromuscular scoliosis (NMS) is less reliable, and more prone for false positive and true negative episodes in comparison to the adolescent idiopathic scoliosis (AIS). All patients were operated on at a single institute between 1983 and 1994 who were comprehensively reviewed and their neuromonitoring data analysed. The etiologies included Duchenne's muscular dystrophy (DMD)-55, spinal muscular atrophy (SMA)-30 and miscellaneous-14.

Of the ninety nine patients, the pre-operative baseline could not be obtained in only two patients (DMD and myelomeningocoele). The sensitivity and specificity of each etiology at twenty five, fifty and seventy five percent loss of amplitude is shown below:

	25%		50%		75%	
	Sensitivity	Specificity	Sensitivity	Specificity	Sensitivity	Specificity
DMD	90	29	87	44	70	52
SMA	93	19	93	42	82	52
Misc	Not reported		86	54	Not reported	

I advocate the use of a fifty percent fall in baseline amplitude to be of optimal sensitivity and specificity. Twenty five percent change was associated with a high incidence of false positive episodes, and a seventy five percent change in amplitude yielded too many false negatives. Hence, the fifty percent change in amplitude rule made sense. This has now been a benchmark for over two decades for safety during scoliosis surgery.

Spinal cord monitoring in operations for neuromuscular scoliosis: Noordeen MH, Lee J, Gibbons CE, Taybr BA, Bentley G. J Bone Joint Surg Br. 1997 Jan; 79(1): 53-7. PMID: 9020445.

ORIGINAL ARTICLE

Neuromuscular Scoliosis Treated by Segmental Third-Generation Instrumented Spinal Fusion

Marco Teli, MD, Hazem Elsebaie, FRCS, Leela Biant, FRCS, and Hilali Noordeen, FRCS

Abstract: We aimed to investigate whether the outcome and complications of surgical treatment of neuromuscular curves with segmental third-generation instrumentation could compare with those reported with standard second-generation instrumentation. The clinical and radiologic data of a single surgeon's consecutive series of patients with neuromuscular scoliosis treated with two types of newer-generation instrumentation and posterior or anteroposterior approaches were retrospectively and independently reviewed. The results of this study support the concept that third-generation instrumentation is able to provide at least as good results as second-generation instrumentation in the treatment of neuromuscular scoliosis patients, at the expense of a lower complication rate.

Key Words: neuromuscular scoliosis, spinal fusion, third-generation instrumentation

(*J Spinal Disord Tech* 2005;18:430–438)

The term "neuromuscular scoliosis" (NMS) describes a spectrum of spinal disorders that have in common the resulting disturbance of the musculoskeletal system because of altered innervation and/or muscle tone. As a consequence, the spine in these patients tends to lack its supportive properties, which makes ambulating and sitting problematic. Patients affected by NMS have a poor response to conservative treatment of spinal deformities, and the consequences of spinal deformity itself are more serious for them than for patients with idiopathic scoliosis, namely, in the cardiovascular and respiratory systems.[1] Surgical treatment, in the form of segmental instrumented spinal fusion (ISF) with second-generation instrumentation (eg, Luque–Galveston and unit rod constructs), has been considered the standard surgical technique for NMS surgery in the last two decades.[2] These techniques generally lead to improved quality of life with correction of coronal deformity and of pelvic obliquity in the range of 50–75% of preoperative values. Major complications, most commonly deep infection, instrument failure, and pseudoarthrosis, affect 7–45% of treated patients.[3–7]

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From the Great Ormond Street Hospital for Sick Children, London and Royal National Orthopaedic Hospital, Stanmore, UK.
Dr. M. Teli's current address is Galeazzi Orthopaedic Institute, Milan, Italy. Dr. H. Elsebaie's current address is Cairo University Hospital, Cairo, Egypt.
Reprints: Dr. M. Teli, Galeazzi Orthopaedic Institute, Via Galeazzi 4, 20161 Milan, Italy (e-mail: marcoteli@hotmail.com).

The widespread use of third-generation instrumentation (Cotrel-Dobousset and related implants) has changed our approach to the surgical treatment of both adolescent[8] and adult[9] idiopathic scoliosis. Surgery for idiopathic scoliosis using third-generation instrumentation techniques provides significant deformity correction and patient satisfaction, with an acceptable complication rate. Nevertheless, almost 20 years after the introduction of such devices, reports are lacking on the results of third-generation instrumentation for the treatment of NMS. Therefore, the aim of this study is to verify the hypothesis that treatment of NMS with third-generation ISF may provide at least as good results as second-generation ISF, with fewer complications. To do so, we report on a single surgeon's consecutive series of patients treated at two tertiary referral centers for spinal disorders and compare the results with the pertinent literature.

MATERIALS AND METHODS

Inclusion criteria for the study were a diagnosis of progressive scoliosis due to a neurologic or muscular disorder, treatment by posterior-only or anteroposterior spinal fusion by third-generation instrumentation, and a minimum follow-up of 2 years from surgery. Exclusion criteria were incomplete clinical and/or radiologic documentation at follow-up. The medical records and imaging of all patients operated on by the senior author from 1995 to 2000 who satisfied the above criteria were reviewed.

Patients aged up to 15 years were treated at a pediatric orthopedic institution, whereas older patients were treated at a separate spinal deformity unit. All patients were assessed preoperatively by the senior author and followed up postoperatively in his outpatient clinic. At every appointment, sitting (standing when possible) posteroanterior and lateral 50 × 115–cm radiographs were obtained. Posteroanterior traction (bending when possible) films were requested preoperatively. The coronal and sagittal components of the spinal deformity were measured by the Cobb method. Pelvic obliquity was measured as the angle described by a line tangential to both iliac crests and a line perpendicular to the spinous processes of L4 and L5.[10] When instrumentation, fusion mass, or removal of spinous process made this comparison unreliable, we drew a line intersecting the middle of the pedicles of L4 and L5 on the posteroanterior view.

Indications for surgery were based on (a) consideration of the magnitude and progression of the spinal deformity; (b) presence of functional deterioration; (c) evaluation of cardiopulmonary status, including echography where necessary (eg,

in myopathic patients) and pulmonary function tests in cooperative patients; (d) evaluation of nutritional status, including weight/height ratio, lymphocyte count, and serum albumin.[1–3,5–7] These evaluations were carried out by a team composed of a spinal deformity surgeon (senior author) with his fellow and resident, a dedicated anesthesiologist, a referring pediatrician/neurologist, specialist nurses, physiotherapists, nutritionists, and occupational therapists. Procedures were planned as posterior-only, anteroposterior on the same day under one anesthetic session (combined) or at a time interval of 1 week (staged), depending on etiology and flexibility of the curve, general health status, skeletal maturity, and anticipated intraoperative blood loss. Indications for posterior-only instrumented spinal fusion were given to patients affected by flaccid NMS due to Duchenne muscular dystrophy (DMD), to those whose cardiopulmonary function was not deemed suitable for a double approach, and to those skeletally mature patients whose deformities were flexible (above 50% correction on traction/bending films). Indications for anteroposterior spinal fusion were given, with the above exceptions, to patients featuring rigid coronal curves (below 50% correction on traction/bending films) and/or fixed pelvic obliquity, deficiency of posterior elements, and skeletal immaturity. Anterior instrumentation was used to correct fixed thoracolumbar and lumbar coronal and/or sagittal deformities whenever bone was judged of sufficient quality by the surgeon. The decision whether to perform an anterior instrumented versus noninstrumented fusion was taken both on the results of the preoperative screening (nutritional status, walking ability) and on the operative findings (bone quality and ability to withstand implants). Bone densitometry was not part of the screening tests of the study population. Combined procedures were selected by the surgical team whenever possible. Staging was reserved to those cases where significant intraoperative factors (ie, amount of blood loss and operating time) were anticipated to contraindicate same-day surgeries or when halo-tibial traction was used to progressively correct fixed sagittal deformities. Anterior procedures always preceded posterior ones. Two types of third-generation titanium instrumentation systems were used anteriorly and posteriorly as available in the two public health care institutions where patients were treated. The two systems used were the Synergy (Interpore Cross, Irvine, CA) and the Colorado (Medtronic Sofamor Danek, Memphis, TN). Indications for fusion to the pelvis by iliac screws were given to nonambulatory patients with sitting unbalance and to patients who had curves including the pelvis and/or fixed pelvic obliquity.

Surgical Technique

Anterior thoracic or thoracoabdominal approaches to the spine were done on the convex side of the deformity, through the rib bed correspondent to the highest spinal level to reach. The selected rib was excised, morselized, and used as autograft for the posterior spinal instrumented fusion. The diaphragm was divided peripherally in thoracolumbar approaches. Ligature of segmental vertebral vessels was done only in instrumented procedures. It was the surgeon's effort to perform release of the anterior longitudinal ligament, discectomy, and removal of vertebral endplates. This aimed to enhance mobilization of the spinal segments and chances of interbody fusion. Twenty to 40 mL of hydroxyapatite derived from marine coral was used as interbody graft material. Anterior instrumentation included segmentally the entire curve as measured preoperatively. Instrumentation consisted of 5.0- to 7.0-mm-diameter screws with washer and bicortical purchase in the vertebral bodies and a single 5.5-mm-diameter rod, tightened to each screw and derotated to achieve the necessary coronal and sagittal contour of the regional vertebral column. In thoracolumbar approaches, the diaphragm was closed in a continuous layer with the pleura over a suction drain.

Posterior approaches were done through a straight midline incision and subperiosteal exposure of the vertebral levels to be included in the fusion. Facetectomy was done at the same levels. Vertebral levels encompassed the entire thoracic (from T1 or T2) and lumbar spine according to the morphology of the deformity. Laminar, pedicle, and transverse process hooks were preferred at thoracic levels. Pedicle screws were positioned at lumbar and thoracolumbar levels. Sublaminar wires were used at lumbar and thoracolumbar levels in case of insufficient bone–implant purchase to achieve further fixation. Two 5.5-mm-diameter rods linked by cross-connectors were implanted in each patient. The technique of ileospinal fixation involved bilateral exposure of the posterior superior iliac spine through the same midline incision used for the lumbar exposure, perforation of the iliac crest with a pedicle finder directed toward the superior and lateral rim of the acetabulum, and use of 5.5- to 7.0-mm-diameter, 40- to 80-mm-long iliac screws. Bilateral S1, L5, or L4 screws and dedicated connectors completed the construct. Deformity correction was obtained by contouring of the rods, careful translation, and segmental compression/distraction. Morselized autografts, from harvested spinous processes, facet joint bone, and rib (where previously harvested), were all applied after decortication of the posterior elements. This was routinely augmented with 20–40 mL of hydroxyapatite derived from marine coral. The fascia was closed in a continuous layer followed by subcuticular and intradermal layers. No wound drains were used. No cell-saver was used intraoperatively on the patients included in the study. Patients received homologous blood transfusions intra- and postoperatively as needed by clinical conditions. Controlled hypotensive anesthesia (with a mean blood pressure above 60 mm Hg) was carried out to maintain a dry surgical field and to control blood losses, a feature of utmost importance in NMS surgery and ensuring critical perfusion of the vital organs. Recording of the somatosensory spinal-evoked potentials (SSEPs) was done routinely: A prolonged 50% decrease in SSEP trace amplitude was regarded as significant.[4] Intravenous antibiotics, a combination of aminoglycosides and β-lactamates for prophylaxis against gram-negative and gram-positive bacteria, were started perioperatively and continued for 72 hours.

Postoperative Care and Follow-Up

Postoperatively, patients were transferred to an intensive care unit (ICU) if they were still ventilated or to a high-dependency unit (HDU) if they were not ventilated and needed only specialized postoperative care. Patients were transferred from these units to their wards after extubation and fluid

balancing. Oral feeding was stimulated as soon as tolerated, whereas enteral or parenteral hyperalimentation was used in case oral feeding was not possible. Mobilization to a molded wheelchair or ambulation was also allowed as soon as tolerated, with the assistance of specialized physiotherapists. No brace was prescribed postoperatively. Clinical and radiologic outpatients' follow-up was done at 3, 6, and 12 months postoperatively and yearly thereafter with the modalities described above. Fusion and graft incorporation were routinely judged upon examination of posteroanterior and lateral radiographs. Oblique views and computed tomography scans were requested in cases of suspected pseudoarthrosis, namely, local tenderness, loss of correction, implant failure, or a combination of these factors.

An outcome questionnaire validated for flaccid NMS[5] only was adopted for review of spastic NMS cases too and mailed to patients' parents/caregivers at the final follow-up. The questionnaires were filled in by patients themselves or by parents or caregivers depending on the patients' intellectual abilities. The questionnaire includes a total of 20 questions concerning satisfaction with the surgical outcome, cosmesis, function, self-image, pain, patient care, pulmonary function, and quality of life. Every answer is graded −2 (worse) to +3 (major improvement) for a cumulative score ranging from −36 to +52.

Analysis of Data

Clinical and radiographic data were subdivided among treatment groups, that is, posterior-only (group 1), anteroposterior combined (group 2), and anteroposterior staged (group 3) surgeries. We considered sex, age, and weight at surgery, functional status, number of levels operated, operative time, intraoperative blood and estimated blood volume (EBV) loss, instrumentation type, ICU/HDU stay, hospital stay, complications, follow-up length, preoperative/postoperative/follow-up scoliosis, kyphosis, lordosis, and pelvic obliquity angles. Complications were divided into major and minor according to their effect on the course of recovery in hospital and on their potential life-, limb-, or function-threatening effect.[6,11,12] Major complications included pneumonia, pneumothorax requiring prolonged chest tube, tracheostomy, persistent neurologic deficit, deep wound infection, bowel ischemia, congestive heart failure and injury to a major vessel, urinary sepsis and gross hematuria, disseminated intravascular coagulation, rod breakage, implant loosening requiring revision, and pseudoarthrosis. Minor complications included upper respiratory tract or tracheostomy infections, spontaneously resolving pneumothorax, paresthesia, cerebrospinal fluid leak, superficial wound infection, wound dehiscence, pressure sore, ileus, gastritis, colitis, transient cardiac arrhythmia, urinary tract infection, and implant loosening not requiring revision.

Pearson correlation analysis was applied to data by means of an SPSS program (Chicago, IL). To test persistent effects, preoperative, postoperative, and follow-up scores were evaluated by the Wilcoxon signed rank test. Differences between groups were evaluated by the χ^2 test. For all comparisons, a P value of <0.05 (two tailed) was considered significant. Descriptive statistics were used for these and other outcomes, and the results were expressed as means ± SD, range, percentage, or total number of cases.

RESULTS

Of a group of 62 patients who satisfied the inclusion criteria, 5 were excluded because their follow-up data were incomplete owing to change of address and hospital care area. One (affected by arthrogryposis) was excluded because at the time of the latest follow-up, he had been treated with a long anterior instrumented fusion only and was being observed. Of the 56 patients (27 females, 29 males) included in the study cohort, mean age at surgery was 14 (range 8–21) years, mean weight at surgery was 46 (range 30–70) kg, and mean follow-up was 52 (range 24–85) months. Etiology of spinal deformity included spastic quadriplegic cerebral palsy (CP) in 36 patients, and DMD in 8 patients. Spina bifida, Friedreich ataxia, syringomyelia, spinal muscular atrophy (SMA), and poliomyelitis were the etiologies in the remaining 12 patients. Functional level was graded according to the World Health Organization classification of impairment, activity, and participation.[13] Forty-five patients (80%) included in this study were at functional level 5. Patients' functional levels and etiologies of spinal deformity are displayed in Table 1.

At the time of surgery, spinal growth in 35 patients (62.5%) was considered complete. This was defined on preoperative posteroanterior full spine radiographs as Risser 5 ossification of the iliac apophysis and closure of the triradiate cartilages.

Table 2 summarizes the main clinical features regarding the three treatment groups. There were 23 (41%) patients in group 1 (posterior-only), 24 (42%) in group 2 (anteroposterior combined), and 9 (17%) in group 3 (anteroposterior staged).

TABLE 1. Patients' Functional Level[13] and Etiology of Spinal Deformity

	No. of Patients (n = 56)	Etiology/No. of Patients
WHO functional level 1 = walkers without devices	—	—
WHO functional level 2 = walkers with limitations in community walking	6	CP/4, Friedreich ataxia/1, Syringomyelia/1
WHO functional level 3 = walkers with mobility devices	5	CP/2, DMD/2, syringomyelia/1
WHO functional level 4 = self-mobilized with either transport or power mobility outdoors	2	CP/1, DMD/1
WHO functional level 5 = self-mobilized with severe limitations	43	CP/29, DMD/5, spina bifida/4, Friedreich ataxia/2, syringomyelia/1, SMA/1, poliomyelitis/1

WHO, World Health Organization.

TABLE 2. Main Clinical Data

Group	Gender (M:F)	Age at Surgery (y)	Weight at Surgery (kg)	Operated Levels	Operative Time (min)	Blood Loss (mL) [EBV]	ICU/HDU Stay (h)	Hospital Stay (d)	Major Complications (no. of patients)	Minor Complications (no. of patients)	Follow-up Length (mo)
1 (n = 23)	14:9	14 ± 2	49 ± 12	16 ± 4	273 ± 67	2,815 ± 953 [1.1 ± 0.5]	34 ± 6	9 ± 8 $P < 0.05$	2	5 $P < 0.05$	56 ± 23
2 (n = 24)	9:15	14 ± 2.5	43 ± 9	24 ± 6	443 ± 175 $P < 0.05$	3,228 ± 1,253 [1.2 ± 0.6]	41 ± 10	13 ± 7	2	1	50 ± 14
3 (n = 9)	4:5	15 ± 3.3	44 ± 10	25 ± 6	383 ± 78	2,626 ± 1,584 [0.9 ± 0.5]	43 ± 15	17 ± 5	2	3	50 ± 16

Values are means ± SD.

Among the 23 patients in group 1, there were 11 affected by CP, 8 by DMD, 3 by Friedreich ataxia, and 1 by SMA; the last 7 were not deemed medically fit for a double approach. Among the 24 patients in group 2, 19 were affected by CP, 1 by poliomyelitis, and 4 by spina bifida. Among the nine patients in group 3, six were affected by CP and three by syringomyelia. Two patients in group 3, both affected by spastic quadriplegic CP and having a stiff thoracolumbar kyphosis as the main feature of their deformity, underwent a 1-week period of halotibial traction between stages. Sex, age, and weight at surgery, functional levels, and follow-up length were not significantly different among the three groups. On average, 8 levels (range 5–13) were operated on the 37 anterior and 16 levels (range 9–19) on the 56 posterior procedures. Of 37 anterior procedures, 25 (64%) were instrumented. Of 56 posterior procedures, 48 (86%) included the pelvis. The longest mean operative time was recorded in group 2, that is, combined approach (443 minutes; $P < 0.05$). The highest blood loss was recorded in group 2 (1.2 EBVs) and the lowest (0.85 EBVs; $P < 0.05$) in group 3, that is, staged approach. Blood loss was correlated with the degree of pelvic obliquity ($r = 0.29$) and number of operated levels in all groups: 1 ($r = 0.42$, $P < 0.05$), 2 ($r = 0.39$, $P < 0.05$), and 3 ($r = 0.42$, $P < 0.05$).

Significant intraoperative SSEP changes, that is, drops to below 50% of baseline trace amplitude, were observed in seven patients (12.5%). These were all false-positive intraoperative records; that is, they did not correlate with newly developed postoperative neurologic deficits.

The longest ICU/HDU stay (46 hours on average) was recorded in group 3, but this value did not reach statistical significance compared with the two other groups. Hospital stay in group 1 (9 days on average) was significantly shorter ($P < 0.05$) compared with the two other groups.

Major complications affected six patients (10.7%). Of a total of seven major complications, there was one pneumonia in a patient in group 2 who eventually required a long-term tracheostomy, one persistent pleural effusion due to chylothorax in a patient in group 3, two deep wound infections (both in group 1 patients, resolved with repeat operative debridement), two cases of ileosacral screw loosening requiring removal (one in group 1 same patient who had the deep wound infectioń[and one in group 2), and one pseudoarthrosis at the midthoracic level requiring a local revision and fusion procedure in a group 3 patient. No perioperative death occurred in this study cohort.

Minor complications affected nine patients (16.1%). Of a total of nine minor complications, there was one superficial infection that settled with intravenous antibiotics in group 1, one ischial pressure sore requiring a local flap procedure 6 weeks postoperatively in group 3 (same patient who had chylothorax), and four cases (four patients, two in group 1 and one each in groups 2 and 3) of urinary tract infection treated with oral antibiotics. Furthermore, there were three cases of implant prominence (proximal hooks and ileosacral screws) in thin patients, two in group 1 and one in group 3, not requiring removal at the date of follow-up. Major complications were not statistically different in the three groups of treatment. Minor complications prevailed significantly in group 1 (5 patients affected; $P < 0.05$).

One patient in the study group died of cardiac failure at age 19, 41 months after posterior-only spinal fusion. He had cardiomyopathy and restrictive lung disease, his primary diagnosis being Friedreich ataxia.

Outcome questionnaires[5] (Table 3) were returned by 46 of 56 (82%) parents/caregivers. The parents of the deceased patient refused to respond. Nine other patients had changed their address at the time of the postal survey, and we have been unable to track their new address. Their follow-up data are thus based only on recorded evidence of outpatient and radiology appointments made until a minimum follow-up of 2 years. The mean cumulative score of the outcome questionnaire was 26.3 (range 21–31) for 8 patients with flaccid NMS (7 by DMD and 1 by SMA) and 20.9 (range 10–37) for 38 patients with spastic

TABLE 3. Evaluation Questionnaire Scores[5]

	Flaccid NMS (n = 10)	Spastic NMS (n = 36)
Cumulative	26.1 ± 5.0	20.9 ± 6.5
Cosmesis	4 ± 1.4	4.2 ± 1.9
Function	6 ± 3.6	3.8 ± 3.8
Self-image	0.7 ± 0.9	0.7 ± 1.3
Pain	0.7 ± 0.9	0.7 ± 1.3
Patient care	1 ± 1.4	1.6 ± 3.9
Pulmonary function	0.7 ± 0.9	0.5 ± 1.6
Quality of life	4 ± 0.5	3 ± 3.2
Satisfaction	9 ± 2.5	6.4 ± 4.4

Note: Validated for flaccid NMS only. Scores are means ± SD.

NMS whose parents/caregivers completed the questionnaire. The highest scores in both flaccid and spastic patients were recorded in the areas of satisfaction, cosmesis, function, and quality of life. In flaccid patients, satisfaction and function scores were definitely higher than in spastic ones (on average 9 and 6 compared with 6.4 and 3.8, respectively). No statistical test was applied to these data because the questionnaire is not validated for spastic NMS cases. The lowest questionnaire score was recorded in the above-described patient affected by CP who required a long-term tracheostomy.

Table 4 summarizes the main radiographic data of the three study groups. On average, preoperative coronal deformity was lower in group 1 (58°) than in group 2 (72°; $P < 0.05$) and group 3 (86°; $P < 0.001$), whereas no significant differences were noted in preoperative kyphosis and lordosis angles. Again, main preoperative pelvic obliquity was lower in group 1 (8°) than in group 2 (19°; $P < 0.05$) and group 3 (18°; $P < 0.05$). Correction of scoliosis compared with preoperative values was statistically significant within groups, but no differences were significant among groups. At follow-up, loss of correction of the coronal deformity varied from 0% (group 1) to 2% (group 2). Variation of kyphosis values observed after treatment prevailed in group 1; differences with preoperative values were all statistically significant within groups, but no differences were significant among groups. More importantly, in all groups, an average return to values of kyphosis close to normality was observed postoperatively, without significant changes at follow-up. Variation of lordosis angles prevailed in group 2 (5°; $P < 0.05$ versus groups 1 and 3), with a corrective effect on lordosis (−2°) recorded in group 1. At follow-up, change of lordosis angles prevailed in group 1 (3°). Correction of pelvic obliquity compared with preoperative values was statistically significant in groups 2 and 3. At follow-up, loss of correction of pelvic obliquity prevailed in group 1 (3° or 38%). Figure 1 (A and B) illustrates a neuromuscular curve with pelvic obliquity in a 14-year-old girl with paraplegia due to spina bifida. Follow-up spinal radiographs (see Fig. 1, C and D) show the correction of deformity achieved with anteroposterior instrumented fusion.

DISCUSSION

There are no published randomized studies on conservative versus surgical treatment of progressive neuromuscular curves, spinal surgeons being divided between strong opponents and proponents of the treatment. Nevertheless, reports are increasingly being published on the favorable results of surgery on NMS patients. Areas of particular benefit are better nursing, fewer respiratory complications, improved feeding, and better cosmesis compared with the preoperative status. It is therefore not only a matter of curve correction and stabilization that may provide a satisfactory outcome in these surgically treated patients, but also of overall improved quality of life.

The use of postoperative means of external support (casts, braces) after second-generation ISF for NMS is common.[16] Third-generation segmental ISF has boosted our capability to correct spinal deformities and allow early ambulating/nursing

TABLE 4. Main Radiographic Data

Group	Scoliosis Reop. (Cobb) [% flexibility on bending/traction films]	Scoliosis Postop. (Cobb) [% correction]	Scoliosis f/Up (Cobb) [% lost]	Kyphosis Preop. (Cobb)	Kyphosis Postop. (Cobb)	Kyphosis Follow-up (Cobb) [° lost]
1 (n = 23)	69 ± 30	24 ± 16	24 ± 11	52 ± 23	32 ± 19	31 ± 19
	[64%]	[65%]	[0%]		$P < 0.001$	[−1°]
		$P < 0.001$				
2 (n = 24)	72 ± 21	29 ± 11	31 ± 18	54 ± 19	41 ± 15	44 ± 17
	[60%]	[59%]	[2%]		$P < 0.001$	[3°]
	$P < 0.05$	$P < .05$				
3 (n = 9)	86 ± 16	40 ± 19	41 ± 16	50 ± 30	38 ± 11	41 ± 25
	[48%]	[53%]	[1%]		$P < 0.001$	[3°]
	$P < 0.001$	$P < 0.001$				

Group	Lordosis Preop. (Cobb)	Lordosis Postop. (Cobb)	Lordosis Follow-up (Cobb) [° lost]	Pelvic Obliquity Preop. (Cobb)	Pelvic Obliquity Postop. (Cobb) [% correction]	Pelvic Obliquity Follow-up (Cobb) [% lost]
1 (n = 23)	31 ± 22	29 ± 15	32 ± 15	8 ± 2	4 ± 4	7 ± 2
			[3°]		[50%]	[38%]
2 (n = 24)	22 ± 23	27 ± 15	27 ± 18	19 ± 10	9 ± 8	10 ± 8
		$P < 0.05$	[0°]	$P < 0.05$	[47%]	[6%]
					$P < 0.05$	
3 (n = 9)	30 ± 26	32 ± 16	33 ± 18	18 ± 14	9 ± 11	10 ± 12
			[1°]	$P < 0.05$	[50%]	[5%]
					$P < 0.05$	

Values are means ± SD, expressed as degrees.

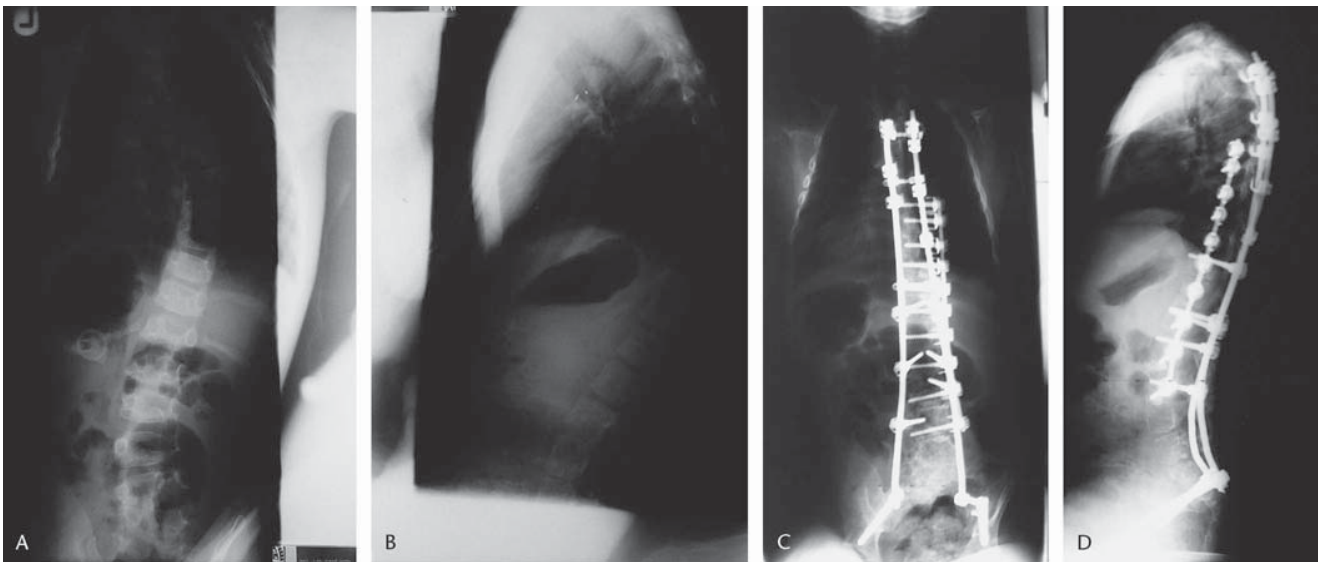

FIGURE 1. A–D, Posteroanterior, lateral preoperative, and follow-up radiographs following anteroposterior combined instrumented fusion in 12-year-old boy with spina bifida (see text).

without external supports in idiopathic scoliosis surgery.[8,9] There is so far no evidence that the same is true in NMS surgery.

It is widely reported that anteroposterior spinal fusion increases deformity correction and decreases the risk of pseudoarthrosis in NMS surgery with second-generation implants.[3,6,7,11,12,14–16] In our practice, anterior instrumentation of lumbar and thoracolumbar deformities with third-generation implants gives the spine satisfactory solidity, adding to the strength of posterior instrumentation, with the final result of a low loss of correction and pseudoarthrosis rate (see Tables 2 and 4; Fig. 1). This seems to reflect the experience of other authors with second-generation posterior implants.[16,17] Conversely, we did not find that anterior procedures with or without instrumentation increase deformity correction, as our data show that the greatest correction was achieved in the group of patients treated with posterior-only approaches. A likely explanation for this finding is that this group of patients had the greatest curve flexibility, because flaccid deformities prevailed in patients treated only posteriorly. A few studies have compared combined versus staged anteroposterior second-generation ISF in NMS, with results in terms of outcome and complications that are variably favorable to combined[7,15] or staged[12,14] procedures. The use of halo traction between stages has not been reported to provide higher correction rates.[16] Nevertheless, in the authors' experience, it can be used after anterior surgical release to make stiff sagittal deformities more flexible and less demanding to correct by posterior surgery.

Comparison of the results of this study with previous-generation instrumentation series was not performed because such a patient population was not available to the authors. Limitations of the present study include the retrospective study design and the diversity of diagnoses in the study group. Previous reports on second-generation ISF for NMS have similar biases, thus making the results of the present study comparable from the point of view of operative time, blood loss, hospital stay, deformity correction, and complications. Two points have to be acknowledged when comparing second-generation studies with the current one: (a) Patients in our series possibly received improved preoperative assessment and postoperative care, improved anesthetic techniques, and dedicated rehabilitation programs; (b) only crude numbers can be compared, statistical analysis being impossible owing to different data reports in the previous studies. Table 5 summarizes the main indexes of clinical and radiographic outcome by comparing the results of the current study (considering cohort values) with those of second-generation anteroposterior ISF in NMS patients.[5,6,11,12]

This retrospective analysis on a series of 56 patients consecutively treated by a single surgeon shows that at an average follow-up of >4 years, segmental ISF with third-generation implants in NMS patients provides improved quality of life with satisfactory correction of coronal deformity, sagittal deformities, and pelvic obliquity. There were statistically significant differences in the main indexes of clinical outcome (Table 2) among patients treated posteriorly only or anteroposteriorly: Patients treated with posterior-only procedures had the shortest hospital stay but the highest incidence of minor complications, whereas patients treated with anteroposterior combined procedures had the longest operating time and highest blood loss, the latter not being statistically significant. Blood loss correlated significantly with the number of operated levels (a parameter of the extent of surgery) in all types of procedures, but not with the number of complications. Blood loss also correlated with the degree of pelvic obliquity, a finding that is explained by the necessity for more extensive anterior and posterior surgeries with increasing pelvic obliquity.

The major complication rate (10.7% of patients) in this study cohort was remarkably lower than that generally reported for second-generation instrumentation (Table 5) with

TABLE 5. Comparison of Main Clinical and Radiographic Data of Current Study with Similar Reports on Second-Generation ISF in NMS

Report	No. of Patients	Operative Time (min)	Blood Loss (mL) [EBV]	Hospital Stay (d)	Questionnaire[5] Mean Cumulative Score	Scoliosis % Correction
Current study, cohort data	56	370	3250 [1.1]	14	26.1 (flaccid NMS)	59
Benson et al[6]	50	402	1839 [unknown]	20	Not applicable	65
Bridwell et al[5]	54	Unknown	Unknown	Unknown	17.5 (flaccid NMS)	51
Sarwahi et al[11]	111	Unknown	660 [unknown]	Unknown	Not applicable	Unknown
Tsirikos et al,[12] cohort data	45	420	2600 [1.6]	26	Not applicable	67

Report	Kyphosis (°), Follow-up Mean	Lordosis (°), Follow-up Mean	Pelvic Obliquity % Correction	Major Complications (% of patients)	Minor Complications (% of patients)	Mean Follow-up Length (mo)
Current study, cohort data	37	32	47	10.7	16.1	53
Benson et al[6]	Unknown	Unknown	75	14	28	40
Bridwell et al[5]	9 (thoracolumbar)	Unknown	57	7.3	1.8	92
Sarwahi et al[11]	unknown	Unknown	Unknown	21.6	22.5	Unknown
Tsirikos et al,[12] cohort data	41	40	75	43.3	50	38

the single exception of the above-referenced study on flaccid NMS patients.[5] A lower complication rate possibly resulted from improved preoperative assessment, intraoperative management, and postoperative care compared with previous reports. Major complications were equally distributed among patients treated with the three different procedures, although deep wound infections were observed only in the group of patients treated with posterior-only procedures. In the current authors' practice, deep wound infections tend to affect NMS patients treated with any procedure. The pseudoarthrosis rate detected at an average follow-up of 4 years (1 of 56 patients or 1.8%) in the present series is in the lower range compared with previous reports on second-generation ISF (1.5–10%).[3,5,6,11,12,14,15] The low pseudoarthrosis rate, possibly due to the increased stability provided by third-generation instrumentation, is reflected in a minimal loss of correction of scoliosis, kyphosis, and lordosis angles (0–6% in the different groups) observed at follow-up (Table 4). Nevertheless, we noted that in the group of patients treated with posterior-only procedures, loss of correction of lordosis was noticeable at

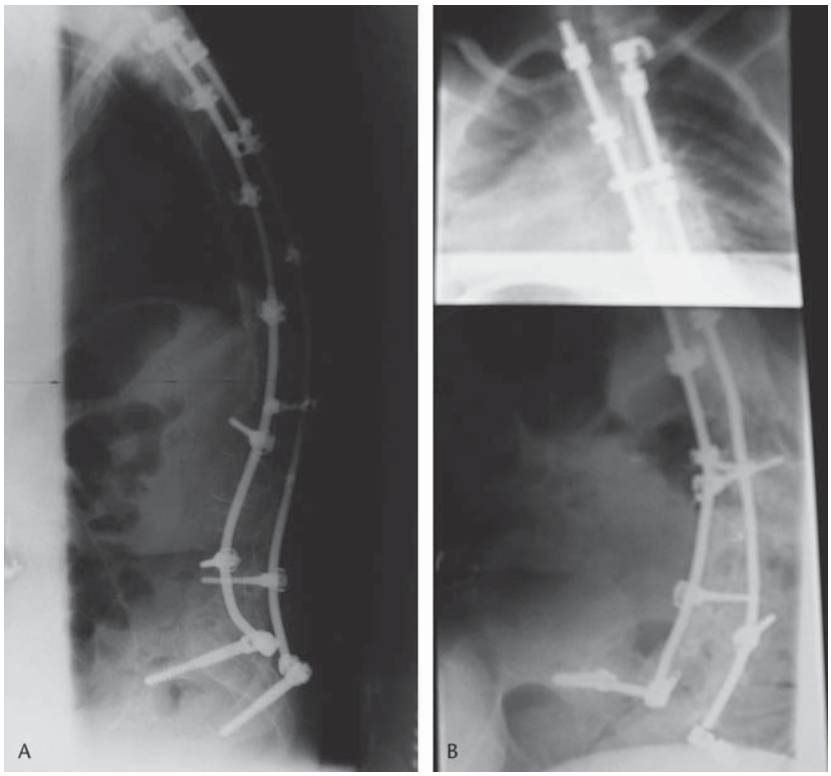

FIGURE 2. A–B, Posteroanterior and lateral preoperative and follow-up films in a 15-year-old boy with DMD. There are adequate lumbar fixation points following posterior-only instrumentation.

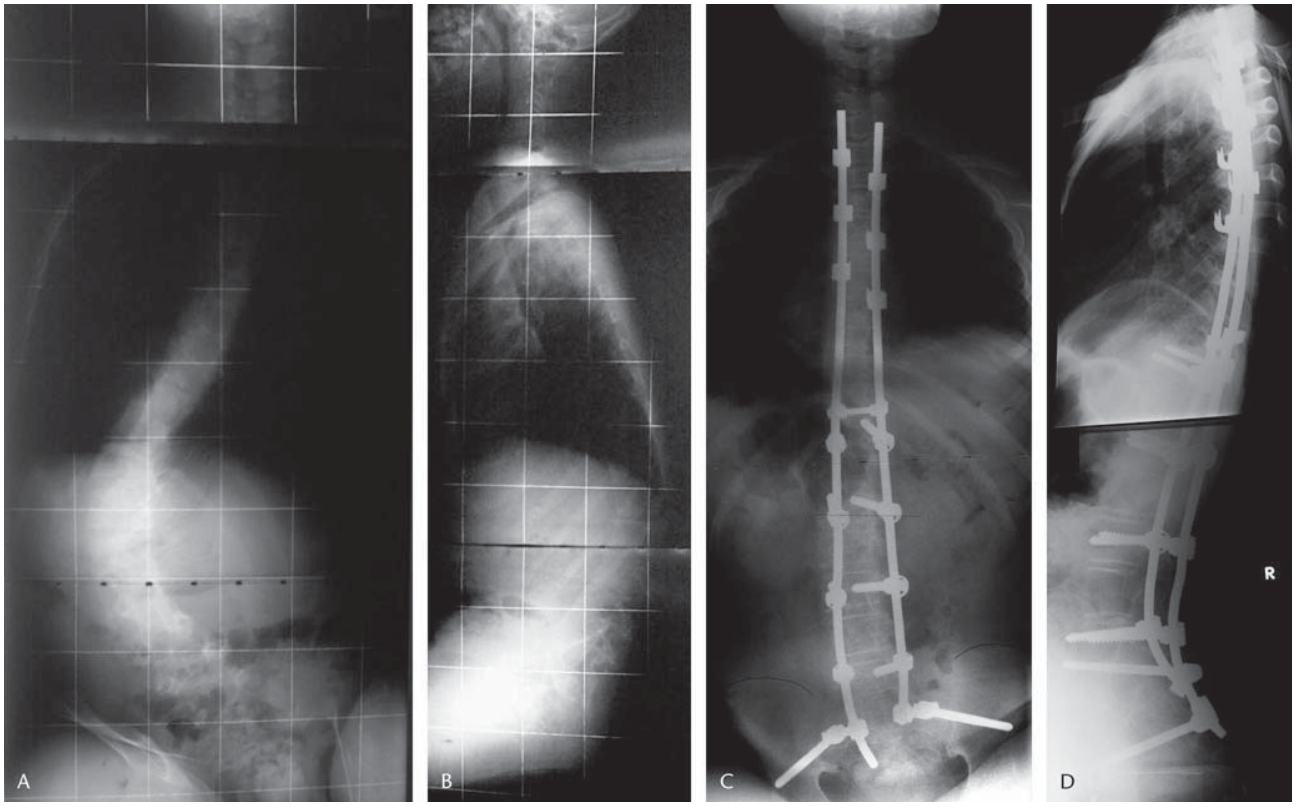

FIGURE 3. A–D, Posteroanterior and lateral follow-up films in a 13-year-old boy with spastic quadriplegic CP. Note the paucity of lumbar fixation points following anterior release/fusion and posterior-only instrumentation. The right rod has disengaged from the iliac bolt, indicating local motion and possibly lumbosacral pseudoarthrosis.

follow-up. Also, correction of pelvic obliquity and loss of correction of the same at follow-up were worse in the group of patients treated with posterior-only procedures than in patients treated with anteroposterior combined and staged procedures, a finding that could lead to deterioration of the patients' conditions at a later date. During the review of the postoperative and follow-up films, it became apparent that in a number of early patients, posterior instrumentation was not applied segmentally (Fig. 2). The paucity of fixation points at the apex of coronal and sagittal curves might cause loss of correction through failure of instrumentation and pseudoarthrosis at a later date. Therefore, we suggest that a strictly segmental concept of fixation should be applied to posterior third-generation instrumentation in neuromuscular deformities (Fig. 3).

The administration of an outcome questionnaire[5] for the evaluation of results of NMS treatment was an effort to move on from the subjective evaluation models that previous authors have reported. This questionnaire was used for evaluation of spastic cases too because no validated questionnaires exist for these patients to date. The designers of the adopted questionnaire[5] validated it only for evaluation of flaccid cases (DMD and SMA) treated with second-generation instrumentation, reporting an average cumulative total of 17.5 points in their patients (Table 5). We recorded a 26.3-point mean score in our flaccid NMS patients and, for descriptive purposes only, 20.9-point mean score in our spastic NMS patients. Still, dissatisfaction might have been expressed by the parents/caregivers who could not receive the outcome questionnaire. Scores in the specific areas of cosmesis and quality of life (Table 3) of our flaccid NMS patients were remarkably similar to those reported for flaccid NMS patients treated with second-generation instrumentation.[5] In both our spastic and flaccid patients, "major improvements" were recorded on average in the areas of satisfaction, cosmesis, function, and quality of life, although higher scores were recorded in flaccid than in spastic patients in the specific areas of satisfaction and function. This finding might be explained by the fact that flaccid cases were treated when functional deterioration was starting to be significant. On the other hand, it might suggest that the course of this deterioration was altered by surgical intervention.

Based on the results of this study, we conclude that segmental third-generation ISF is able to offer NMS patients improved quality of life through satisfactory and lasting correction of their spinal deformity, at the expense of a lower complication rate than that generally reported for second-generation ISF.

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REVIEW

Athanasios I. Tsirikos
Asif Saifuddin
M Hilali Noordeen

Spinal deformity in neurofibromatosis type-1: diagnosis and treatment

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A. I. Tsirikos · M. H. Noordeen
Spinal Deformity Unit,
The Royal National Orthopaedic Hospital NHS Trust, Brockley Hill,
Stanmore, Middlesex, HA7 4LP, UK

A. Saifuddin
Department of Radiology, The Royal National Orthopaedic Hospital NHS Trust,
Brockley Hill, Stanmore,
Middlesex, HA7 4LP, UK

A. Saifuddin (✉) · M. H. Noordeen
Institute of Orthopaedics and Musculoskeletal Disorders,
University College London, London, UK
E-mail: asaifuddin@aol.com
Tel.: +44-20-89095443
Fax: +44-20-89095281

Abstract Spinal deformity is the commonest orthopaedic manifestation in neurofibromatosis type-1 and is categorized into dystrophic and non-dystrophic types. Management should be based on a meticulous assessment of the spine with plain radiography and magnetic resonance imaging (MRI) to rule out the presence of dysplastic features that will determine prognosis and surgical planning. MRI of the whole spine should also be routinely obtained to reveal undetected intraspinal lesions that could threaten scheduled surgical interventions. Non-dystrophic curvatures can be treated with similar decision-making criteria to those applied in the management of idiopathic scoliosis. However, close observation is necessary due to the possibility of modulation with further growth and due to the increased reported risk of pseudarthrosis after spinal fusion. The relentless progressive nature of dystrophic curves necessitates aggressive operative treatment, which often has a significant toll on the quality of life of affected patients through their early childhood. Bracing of dystrophic curves has been unsuccessful. Combined anterior/posterior spinal arthrodesis including the entire structural component of the deformity is indicated in most cases, particularly in the presence of associated sagittal imbalance. This should be performed using abundant autologous bone graft and segmental posterior instrumentation to minimize the risk of non-union and recurrence of the deformity.

Keywords Neurofibromatosis · Spine deformity · Management · Radiography · MRI

Introduction

Neurofibromatosis is the most frequent single-gene disorder affecting mankind [12]. It is a disorder of neural crest cells defined as a spectrum of multifaceted diseases, probably hamartomatous in origin, involving neuroectoderm, mesoderm and endoderm [16]. Its clinical manifestations have in common the presence of neurofibromas, schwannomas and cafè-au-lait macules, which can potentially appear within any organ system of the body, involving primarily the skeleton, skin and soft tissues.

Individuals with the disorder have attracted in the past significant public interest mainly due to the dramatic cutaneous manifestations, the overgrowth of an extremity as part of a gigantism process and the development of severe spinal deformities causing devastating cosmetic results. ''The Hunchback of Notre Dame'', a novel written by Victor Hugo, portrays Quasimodo as a man with a deformed back who was probably affected by neurofibromatosis [33]. The condition drew public attention when Joseph Carey MacDonald Merrick (John Merrick), who was portrayed in

the book and play "The Elephant Man", was thought to have neurofibromatosis. However, it is now believed that Proteus syndrome was a much more likely diagnosis [4, 61].

Historically, the clinical symptomatology of neurofibromatosis has been described as early as the 14th century [52]. Virchow, a well-known German pathologist, reported more than 150 years ago clinical features of the condition in several members of the same family, while 35 years later his student von Recklinghausen investigated the histological characteristics of the disorder that was later named after him [70, 72].

Classification and diagnosis

Two distinct clinical forms of neurofibromatosis have been described; neurofibromatosis-1 (NF-1) or peripheral neurofibromatosis and neurofibromatosis-2 (NF-2) or central neurofibromatosis. Other investigators also accept two additional clinical types namely segmental and mixed neurofibromatosis [16]. Central neurofibromatosis affects 1:50,000 individuals and is characterized by bilateral acoustic neuromas. It is not associated with primary skeletal disorders and orthopaedic complications [43, 55]. The most common form of neurofibromatosis is NF-1, which affects 1:3000–4000 individuals and 1,000,000 people worldwide, being seen in all racial and ethnic groups [2, 9, 27, 32, 39]. Inheritance of the disorder is autosomal dominant with high penetrance. Nevertheless, approximately 50% of cases arise sporadically due to de novo mutations [7, 67, 54, 71]. Advanced paternal age appears to predispose to new mutations in the NF-1 gene [54].

NF-1 is caused by a defect in the gene responsible forthe production of the protein neurofibromin. This is a tumour suppressor gene linked to the long arm of chromosome 17 [16, 67, 71]. Neurons, oligodendrocytes, non-myelinated Schwann cells, adrenal medulla, testes and leukocytes are the primary sites where neurofibromin demonstrates its highest expressivity [18, 31]. The major function of the gene product appears to be regulation of the ras protein [44, 71]. Absence of neurofibromin expression results in tumourgenic effects and the development of NF-1 associated neoplasms, including malignant peripheral nerve sheath tumours, pheochromocytomas, malignant myeloid dysplasias and benign neurofibromas [46, 71]. The role of ras in the pathogenesis of tumours in NF-1 has suggested an approach to treatment using ras inhibitors, some of which are likely to begin clinical trials in patients with NF-1 in the near future [44]. Moreover, neurofibromin is a large protein with 2,818 amino acids and has many locations probably interacting with other intracellular proteins. Therefore, one could postulate that interruption of this combined activity could be contributory, apart from the formation of neoplastic lesions, to the development of the other numerous clinical manifestations evident in individuals with NF-1 [19, 30].

The NF-1 gene is fairly large, with approximately 300,000 base pairs [16]. It also demonstrates a considerably high mutation rate, lacks hotspots where these mutations arise, has a variable expressivity from patient to patient and does not show a clear genotype-phenotype correlation [71]. These are the reasons why, despite the identification of the neurofibromin gene, prenatal genetic diagnosis still remains clinically impractical [16, 29]. The detection of new cases is, therefore, still based on clinical criteria defined by the 1987 Consensus Development Conference of the National Institutes of Health on NF-1 (Table 1) [53]. Diagnosis requires at least two of the described criteria to be present [53].

Clinical manifestations of NF-1

Patients with NF-1 may present with a wide variety of clinical manifestations that may not all be readily apparent at birth. Café-au-lait spots are present in over 90% of all individuals affected by NF-1, usually in early childhood and classically take on a Coast of California appearance [16, 71]. Axillary or inguinal freckling also bears a high specificity for confirming the diagnosis of NF-1 in children. Lisch nodules of the iris are common in children above the age of 6 years [48] and are evident in most patients older than 30 years [54]. In contrast, optic gliomas constitute an infrequent finding [39]. However, a limited percentage of these tumours may enlarge rapidly and cause exophthalmos and visual compromise [16]. Neurofibromas appear around puberty and can be cutaneous or deep, infiltrating the sur-

Table 1 Diagnostic criteria defined by the 1987 Consensus Development Conference of the National Institutes of Health for the diagnosis of neurofibromatosis [53]. Two or more criteria present confirm the diagnosis of NF-1

Diagnostic criteria for NF-1
Six or more cafe-au-lait macules >5 mm in greatest diameter in pre-pubertal individuals and >15 mm in greatest diameter in post-pubertal individuals
Two or more neurofibromas of any type or more than one plexiform neurofibroma
Freckling in the axillary or inguinal regions
Optic glioma
Two or more Lisch nodules (iris hamartomas) by slit lamp examination
A distinctive osseous lesion, such as sphenoid dysplasia or thinning of a long bone cortex, with or without pseudoarthrosis
A first-degree relative (parent, sibling, or offspring) with NF-1 by the above criteria

rounding tissues and affecting peripheral nerves or the spinal cord. These lesions can potentially increase in size and number due to puberty or pregnancy and are quite frequent in patients over 30 years old [14, 54].

Approximately 50% of patients with NF-1 will develop severe orthopaedic complications during childhood with spinal deformity and congenital pseudarthrosis of the tibia creating the most challenging therapeutic dilemmas [12, 71]. A recent study reported that 70% of affected individuals would require hospitalization to address surgical or medical issues directly related to neurofibromatosis [79]. Familiarity with the various manifestations of NF-1 in different anatomic locations is, therefore, critical in making an early diagnosis and optimizing treatment. The mainstay of care for these patients focuses on the symptomatic management of disease complications. Counselling of patients and their families should provide adequate information on the possible disease complications, which may impede the quality of patients' life at a very early age, while emphasizing that most individuals with NF-1 have normal life expectancy and lead a productive life.

Spinal deformity in NF-1: classification and imaging evaluation

Scoliosis is the most frequent musculoskeletal manifestation in NF-1, usually occurring in the thoracic region [2, 16, 58]. Almost a century ago, both Gould [28] and Weiss [73] called attention to the high incidence of spinal deformity in patients with neurofibromatosis. However, the true prevalence of spinal deformity in NF-1 remains unknown, with figures in the literature ranging from 2 to 69% [2, 12, 16, 21, 26, 35, 38, 45, 50, 57, 58, 63, 64, 66]. Conversely, 2–3% of all scoliotic patients with significant curves have neurofibromatosis [57]. Suggested aetiological theories for the development of spinal imbalance in this condition include erosion or infiltration of bone by localized neurofibromas, primary mesodermal dysplasia, osteomalacia and endocrine disturbances [8, 16, 26, 43].

Coronal spinal decompensation in neurofibromatosis is generally classified into non-dystrophic and dystrophic types based on the absence or presence of skeletal dysplasia on plain radiographic evaluation, with the clinical and radiological features in the non-dystrophic type being similar to idiopathic scoliosis [26, 75] (Fig. 1). A meticulous search for evidence of dysplastic changes should be performed in all patients since prognosis and management of the scoliotic curve depend largely on the presence of dystrophic features (Table 2). Dystrophic features include vertebral scalloping (posterior, lateral or anterior) (Fig. 2), rib pencilling (Fig. 3) or spindling of the transverse processes, wedging of one or more vertebral bodies (Fig. 4), paraspinal or intraspinal soft tissue masses, a short curve with significant apical rotation, occasionally leading to subluxed or dislocated vertebral bodies, foraminal enlargement and defective pedicles. Dystrophic changes are thought to be either intrinsic in origin or associated with intraspinal anomalies, namely abnormalities of the dura mater such as dural ectasia, or dumbbell neurofibromas extending through the intervertebral foramina and causing foraminal enlargement [16, 43]. As a general rule, the more severe the dystrophic changes identified in the vertebral bodies, the higher the likelihood that the scoliotic curvature will deteriorate. In a previous study investigating the evolution of spinal deformity in NF-1, when a combination of three or more dysplastic features was present, the risk of curve progression was significantly increased in 85% of the patients, while rib pencilling was the only singular

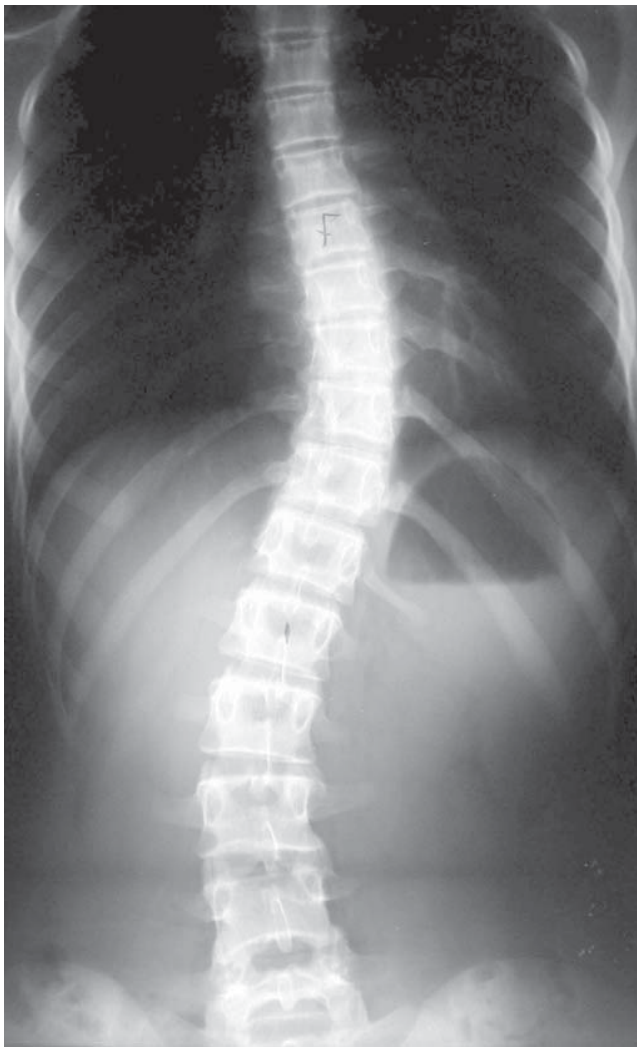

Fig. 1 PA radiograph of the spine demonstrating a non-dystrophic curve pattern in NF-1

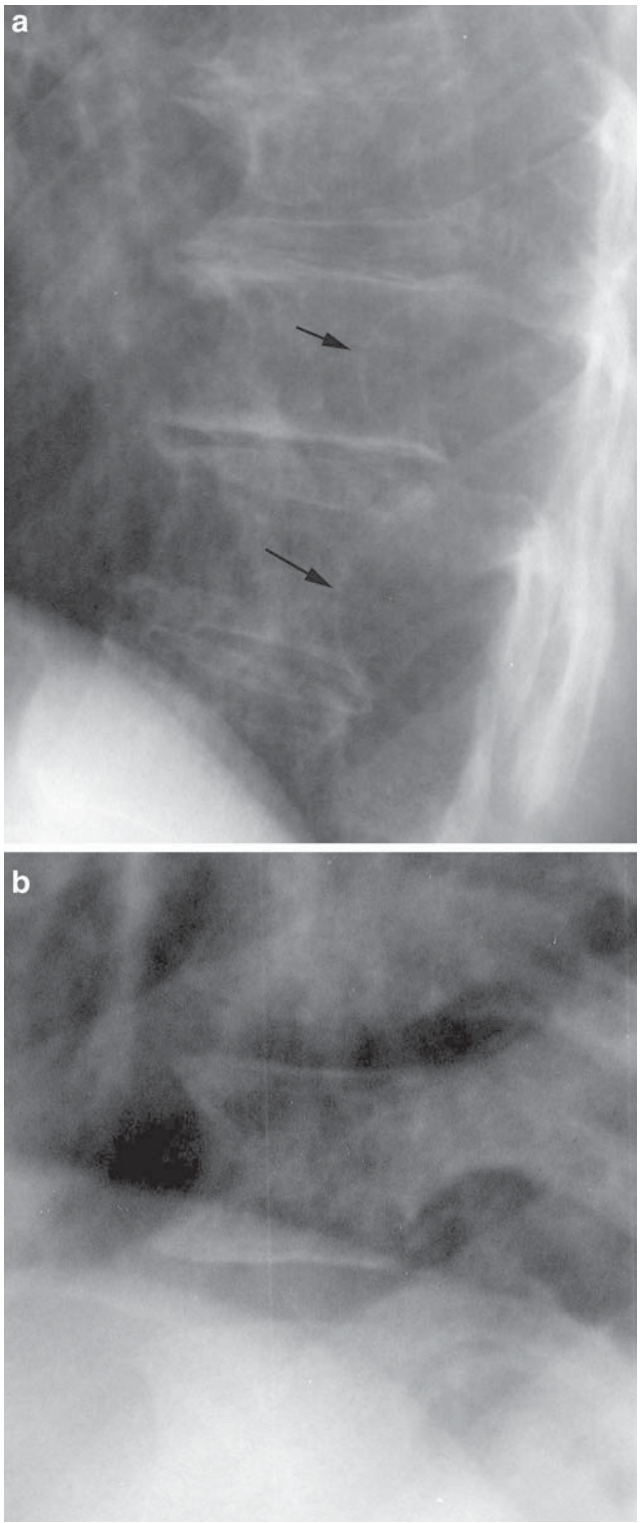

Fig. 2 Vertebral scalloping in NF-1. **a** Lateral coned radiograph of the lower thoracic spine demonstrating posterior vertebral scalloping (*arrows*). **b** Lateral coned radiograph of the lower thoracic spine demonstrating anterior vertebral scalloping

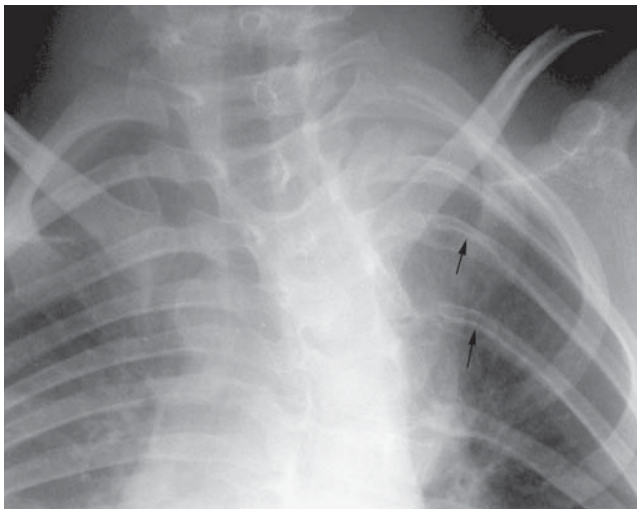

Fig. 3 AP radiograph of the upper chest wall demonstrating rib pencilling (*arrows*)

dystrophic factor statistically influencing risk of scoliosis deterioration [22].

A finding characteristic of NF-1 is dural ectasia, an expansion of the thecal sac at the expense of the bony and ligamentous structures. This may result in posterior vertebral scalloping (Fig. 5) and lateral thoracic meningocele formation (Fig. 6), often causing destabilization of the vertebrae leading to spontaneous subluxation or dislocation, as well as penetration of the spinal canal by protruding ribs separated from their costotransverse attachments [25, 49, 60]. Canal widening, as the result of dural ectasia, is the reason why even tremendous angular deformities may not be accompanied by spinal cord compromise and neurologic deficit. On the contrary, if an intraspinal neurofibroma is the aetiological factor related to the development of canal expansion (Fig. 7), as with any other space-occupying lesion, it can provoke cord compression.

If dystrophic changes are noted on plain radiographs, magnetic resonance imaging (MRI) has been considered absolutely essential to further investigate the intraspinal contents [16], particularly when surgical management of scoliosis is anticipated [16, 43]. MRI of the entire spine is recommended as part of the routine preoperative assessment in all patients with neurofibromatosis to detect intraspinal mass lesions [16, 57, 67]. It is worth noting that the interpretation of MRI can occasionally be difficult in patients with complex deformities including significant vertebral rotation and acute kyphosis.

In our institution, patients with NF-1 who present with spinal curvatures are routinely screened at initial presentation with the use of plain radiographs in order to characterize the deformity in both coronal and sagittal planes and identify associated dystrophic changes. MRI is obtained as part of the regular imaging evaluation,

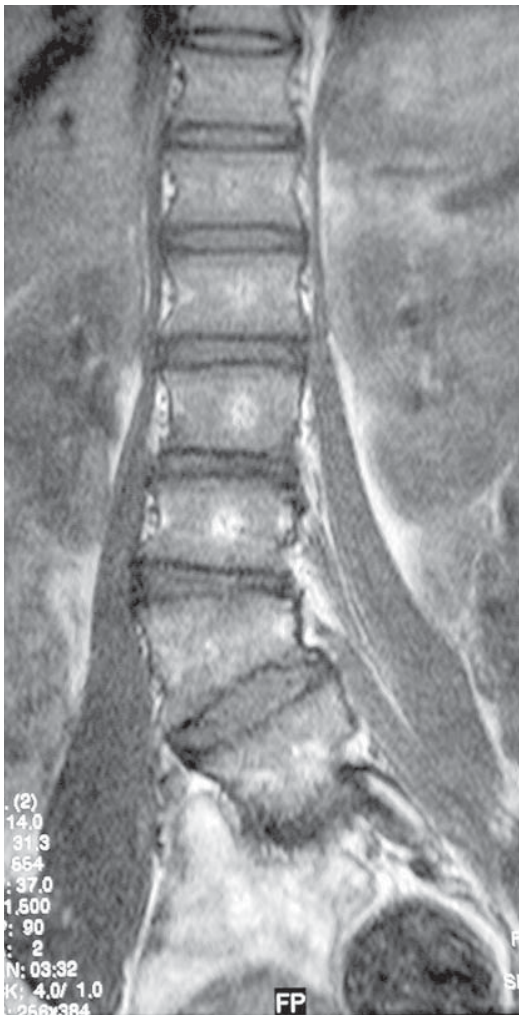

Fig. 4 Coronal T1W MRI showing wedging of the L3 vertebra

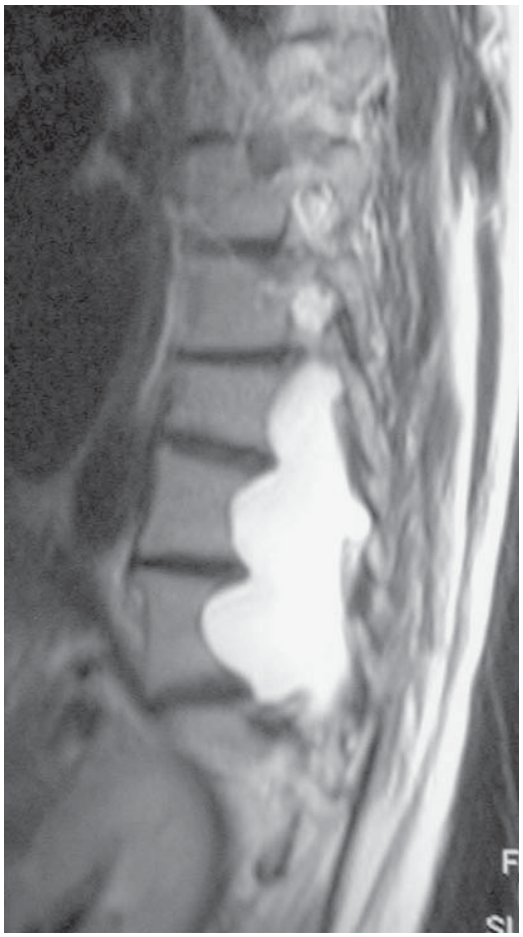

Fig. 5 Sagittal T2W MRI showing dural ectasia in the lower thoracic region associated with posterior vertebral scalloping

Table 2 Typical dysplastic changes evident on plain radiographs in patients with NF-1 [22, 26, 37, 43]

Dysplastic changes
Vertebral scalloping (considered to be present when the depth of scalloping is more than three millimetres in the thoracic spine or more than four millimetres in the lumbar spine)—this is either associated with dural ectasia or neural tumour
Rib pencilling (considered to be present when the width of the rib was smaller than that of the narrowest portion of the second rib)
Transverse process spindling
Vertebral wedging
Paravertebral soft tissue mass
Short curve with severe apical rotation
Intervertebral foraminal enlargement
Widened interpediculate distances
Dysplastic pedicles

regardless of the presence of neurological symptoms, with the aim of delineating intraspinal and paraspinal lesions and better illustrating the components of the deformity. In our experience, MRI of the whole spine identified vertebral dysplasia in 36.3% of cases of NF-1 initially classified on plain radiography as having non-dystrophic curves, while 25% of this subgroup of patients required early surgical correction of the curvature due to rapid progression. This finding indicates the potential value of whole spine MRI at presentation in patients with NF-1 and spinal deformity in clarifying the classification of curve type and assisting management planning.

The need for preoperative assessment with whole spine MRI to exclude underlying intraspinal pathology cannot be overemphasized. However, the requirement for radiological surveillance of spinal tumors in patients with NF-1 that are not scheduled for spine surgery is controversial. Previous investigators have used whole spine MRI, in conjunction with plain radiographs, as a

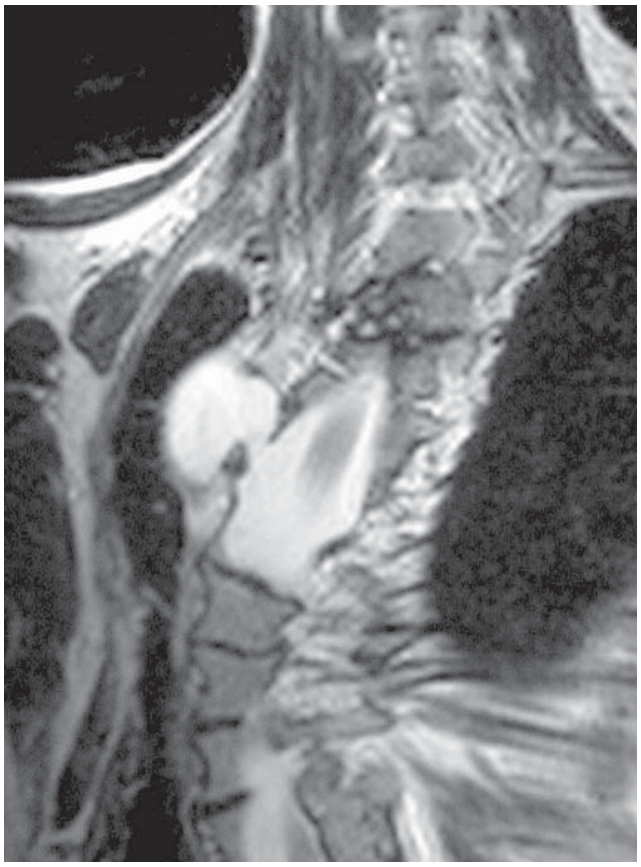

Fig. 6 Coronal T2W MRI showing dural ectasia and a right lateral thoracic meningocele in the upper thoracic region

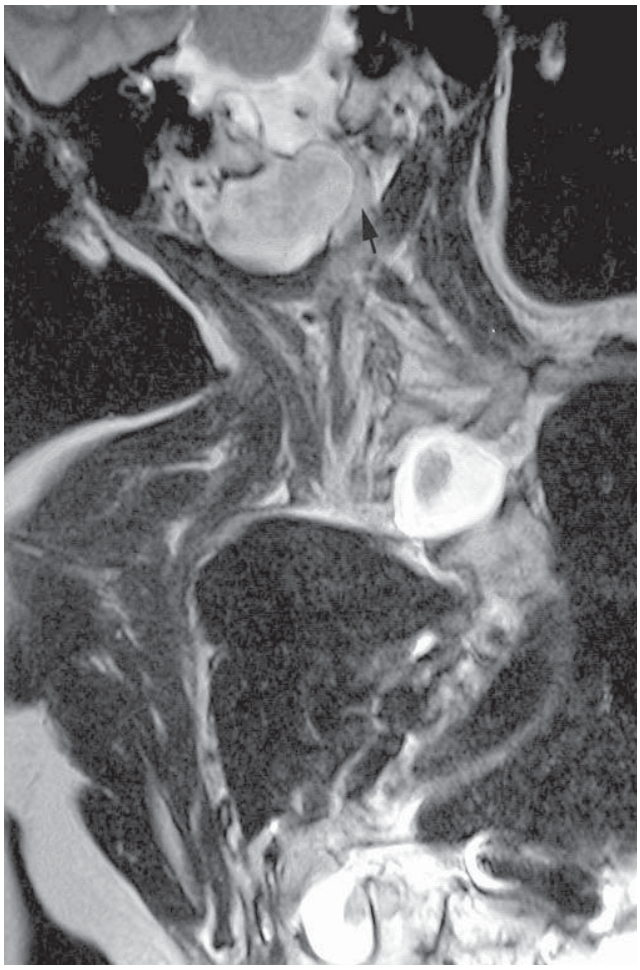

Fig. 7 Coronal T2W MRI in the cervicothoracic region showing a dumb-bell neurofibroma compressing the cervical spinal cord (*arrow*)

diagnostic tool to detect the presence of spinal tumors in patients with NF-1 [23, 42, 69]. Egelhoff et al. [23] identified a high incidence of spinal tumors on MRI (35.7%) in a mixed population of adult and pediatric patients with NF-1 without spinal deformity, and suggested that MRI of the spine should be performed as a routine test in this patient group. In contrast, other investigators do not advocate routine MR imaging and emphasize that MRI should be indicated by clinical necessity [32]. Khong et al. [42] examined 62 children with NF-1 with whole spine MRI and reported an incidence of 13.2% for spinal neurofibromas, closely associated with an increased incidence of scoliosis, localized cutaneous neurofibromas and massive soft-tissue neurofibromas. Thakkar et al. [69] conducted an MRI study including 1,400 children and adults and detected symptomatic spinal tumors in only 23 patients (1.6%).

In our experience, the prevalence of intraspinal and extraspinal neurofibromas was relatively higher compared to that previously documented [23, 42, 69], with a cumulative incidence of 37% in a combined group of patients with dystrophic and non-dystrophic curves. However, none of these tumors was associated with any neurological impairment and all patients were completely asymptomatic on serial clinical examination. Intraspinal or paraspinal neurofibromas were identified in almost half of the patients in the dystrophic group. In this group, there was a tendency for the neurofibromas to develop adjacent to the convexity of the curve (Fig. 8).

Management of spinal deformity in NF-1

Non-dystrophic curves

Non-dystrophic curves can be managed similarly to idiopathic scoliosis and demonstrate comparable response to treatment [3, 13, 15, 43, 55, 62, 75]. If the magnitude of the scoliotic deformity is less than 20–25°,

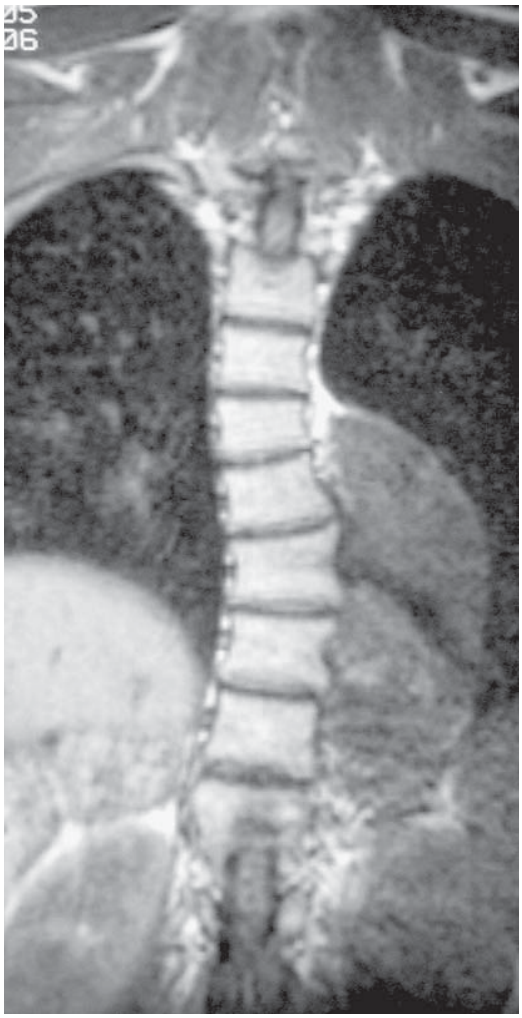

Fig. 8 Coronal T1W MRI in the mid-thoracic region showing a neurofibroma adjacent to the convexity of the curve and associated with minor lateral vertebral scalloping

the patient may be observed closely at regular 6-monthly clinic visits. Brace treatment can be applied for curves between 20° and 40° if the patient still has significant remaining growth. When bracing is selected as the preferred management option, it should be noted that compliance can be particularly challenging, since children with NF-1 may often have cognitive dysfunction, intellectual handicap, attention deficit disorders, seizures and a greater degree of social, emotional and psychological problems compared to their unaffected siblings [24, 41].

If the deformity exceeds 40°, it should be treated surgically by a posterior spinal fusion and segmental instrumentation. The use of autologous iliac crest graft is recommended to enhance a solid bony fusion, especially since there is evidence of a higher incidence of non-union after attempted instrumented spinal fusion in patients with NF-1 in comparison to those with idiopathic scoliosis [1, 13, 15, 16, 43, 55]. For curves of more than 55–60°, where increased rigidity should be anticipated, combined anterior release and bone grafting followed by posterior spine fusion with the use of instrumentation is often necessary to achieve restoration of spinal balance [13, 15, 40, 43, 55].

Close observation of the evolution of deformity is critical due to the possibility of modulation of spinal deformity from non-dystrophic to dystrophic curves and the development of spinal canal neurofibromas, giving rise to pressure-induced expansion of the canal and secondary dysplastic changes in the vertebral bodies [16, 22]. Modulation of non-dystrophic to dystrophic scoliotic curvatures is unique to children with neurofibromatosis, with a reported incidence that varies from 81% in patients diagnosed before 7 years of age to 25% in those detected after the age of 7 years [22]. When observing patients with NF-1 initially classified as having non- dystrophic curves over an extended period of time, there is a higher propensity for developing progressive deformity compared to the idiopathic scoliosis population. In these patients dystrophic changes may develop with growth as part of the modulation phenomenon, but do not show a consistent pattern across the neurofibromatosis population [13, 15, 26, 43]. It is possible that the dystrophic features in this subgroup of skeletally immature patients with idiopathic-like curves have not yet been developed. Another explanation, however, could be that at least certain patients in the non- dystrophic group had occult dystrophic changes initially missed on plain radiography. This concept would call into question the theory of modulation as postulated by Durrani et al. [22].

Dystrophic curves

The dystrophic type of scoliosis is less common, but much more resistant to management [6, 67]. This type of deformity is characterized by short segment, sharply angulated, single thoracic curves that involve four to six vertebral levels and present with three or more dystrophic elements [67]. Dysplastic scoliotic curves may be associated with sagittal plane deformities, namely an angular apical thoracic kyphosis (Fig. 9), or less commonly thoracic lordosis. These concomitant deformities should be recognized early, as they play a significant role in surgical planning.

Dystrophic curves should be treated aggressively, as there is a strong tendency for curve progression even following spinal fusion [3, 6, 16, 43, 75, 77]. The natural history of untreated dysplastic curves, particularly between the ages of 6 and 18 years, is that of relentless deterioration [5]. Passive observation of dystrophic

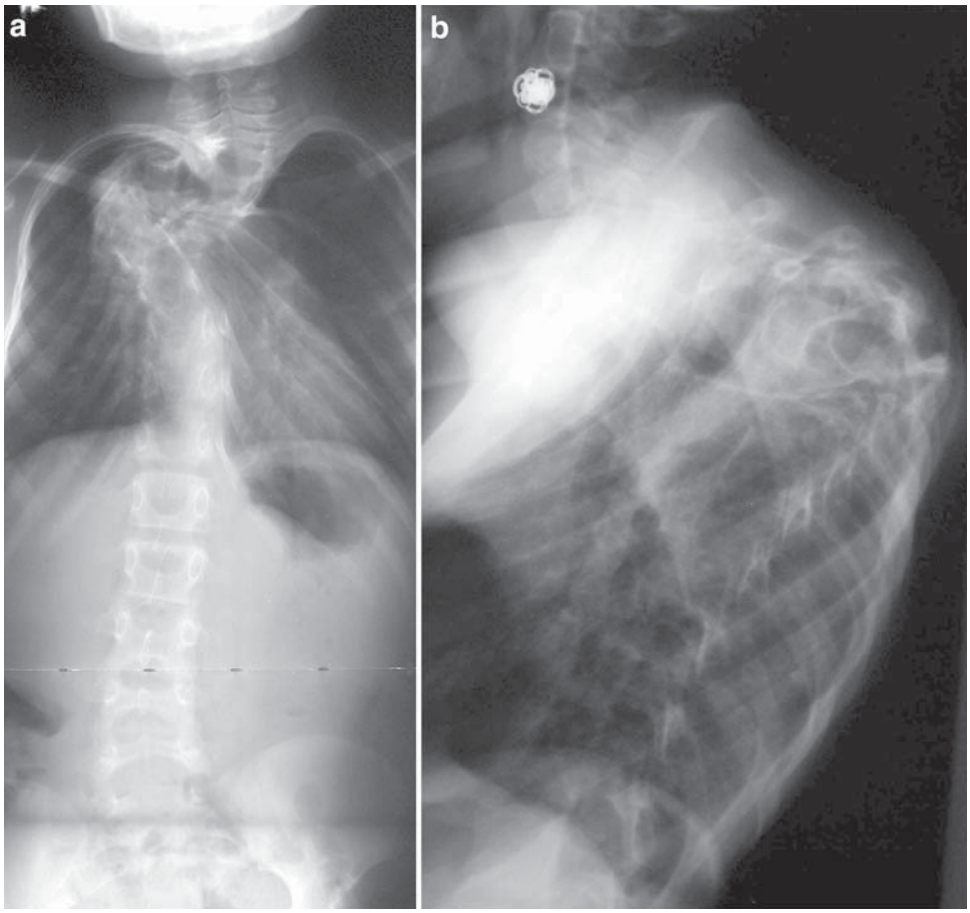

Fig. 9 **a** AP and **b** lateral radiographs of the spine showing the classical acute upper thoracic kyphoscoliosis

curves as they progress throughout childhood is inadequate and unjustifiable [16, 75]. Brace therapy has been ineffective [62, 76] and the need for early aggressive surgical intervention is well documented even in young children [3, 6, 8, 13, 15, 43, 55, 62, 66, 75, 77]. Early spinal fusion does not lead to loss of trunk height since the developing curves are usually short-segment, with limited growth potential. Therefore, loss of height should be anticipated if the deformity is allowed to progress rather than if premature fusion is performed. Apart from the presence of dystrophic changes, other factors that increase substantially the risk of curve deterioration include a young age and a high magnitude of deformity at initial presentation, pathological kyphosis of greater than 50°, location of the apex of the curvature in the mid to caudal thoracic region of the spine, severe apical vertebral rotation of more than 11°, and a severely notched anterior vertebral body [6, 13, 15, 26, 37, 43, 55, 66, 75].

Dystrophic scoliotic curves less than 20° should be closely observed at 6-month intervals to identify any sudden rapid progression and thus prompt surgical management. For patients with scoliotic deformities measuring 20–40° with less than 50° of kyphosis, posterior spinal arthrodesis using segmental fixation with either multiple sublaminar wires or dual rod-multiple hook constructs and the application of autologous iliac crest graft is strongly indicated [16, 43]. Pedicle screws can be occasionally used to provide more stable fixation in the thoracolumbar and lumbar spine in the absence of pedicular dysplasia. Apart from this selective group of patients that can be treated with isolated posterior instrumented fusion, for most types of progressive dystrophic curvatures regardless of the degree of sagittal imbalance, antero-posterior fusion is recommended and provides more reproducible results.

Skeletally immature patients with curves that deteriorate in an uncontrolled fashion require an additional anterior spinal fusion in conjunction to the posterior surgery, with the aim of preventing the development of crankshaft phenomenon. This is produced by continuing unbalanced anterior vertebral growth, which results in increasing rotation of the spine in the presence of a posterior tether caused by the fusion [55]. When the dystrophic scoliotic curve exceeds 40°, combined anterior/posterior spinal fusion including anterior discecto-

my, intervertebral bony fusion with autograft followed by posterior instrumented arthrodesis provides more consistent results in both correcting the deformity and reducing the risk of pseudarthrosis [16]. Both procedures can be performed under the same anesthetic session, unless there is a medical contraindication such as excessive hemorrhage.

Patients with coronal deformity and a dystrophic angular kyphosis also respond poorly to posterior fusion alone. Most authors recommend combined anterior/posterior spinal arthrodesis as the most reliable surgical option in the presence of associated thoracic hyperkyphosis that exceeds 50° [6, 16, 55, 56, 67]. In the latter situation, a structural bone autograft, either rib or fibula, should reinforce the anterior intervertebral fusion followed by posterior instrumentation and arthrodesis using a copious amount of autologous iliac crest bone.

When a neurological deficit is present in a young patient with neurofibromatosis, it is usually caused by increasing kyphosis. Other contributory factors include penetration of the ribs into the spinal canal, structural instability of the vertebral column, progressive dystrophy or destruction of the vertebrae, fibrofatty tissue reaction, intraspinal tumour or dural ectasia [15, 16, 20, 40, 43, 68, 75, 78]. Kyphosis results considerably more than scoliosis in neurologic impairment by creating a pathological spinal flexion, which produces excessive attenuation and deformation of the spinal cord parenchyma and gives rise to neurological symptoms [17, 47, 51]. If the cord is compressed due to the development of a progressive kyphotic deformity, treatment should consist of anterior decompression through a vertebrectomy in the concavity of the deformity followed by combined circumferential bony fusion. Laminectomy has been shown to be ineffective to release pressure in a sharply angulated spinal cord [77]. In the presence of an angular kyphotic deformity, shown to be flexible on extension radiographs, with associated mild neurologic involvement, preoperative halo-dependent traction may be considered, in order to maximise curve correction during the anterior decompression and facilitate placement of the strut graft [15, 43].

Intraspinal tumours can also contribute to spinal cord encroachment and neurologic compromise, particularly in older patients [16, 43]. Treatment consists of laminectomy in combination with tumour resection. Hemilaminectomy is preferable when feasible with the aim of preserving as much bone stock as possible. Removal of the lesion must be accompanied by prophylactic instrumentation and spinal arthrodesis to stabilize the vertebral segments that have been decompressed and prevent the development of post-laminectomy kyphosis [10, 16, 43, 55].

The development of thoracic lordosis is relatively infrequent in patients with NF-1. However, it is often associated with significant respiratory compromise and mitral valve prolapse [34, 74]. Anterior discectomy and fusion followed by posterior fusion using segmental instrumentation, either sublaminar wires or rod-multiple hook constructs should be performed to restore normal sagittal alignment.

There are several difficulties that a spine surgeon involved in the management of patients with neurofibromatosis should be prepared to encounter. Excessive bleeding can hamper particularly anterior approaches to the vertebral bodies and can occur due to the presence of paraspinal neurofibromas and plexiform venous channels in the soft tissues surrounding the spine (Fig. 10). The disproportionate vascularity of neurofibromatous soft tissue is also responsible for the increased frequency of postoperative haemorrhage and haematoma formation [37, 50]. Meticulous haemostasis and wound drainage are necessary to address this problem. Moreover, patients with NF-1 often suffer from hypertension, occasionally associated with renal artery stenosis or pheochromocytoma [59, 71]. Therefore, thorough investigation is required to identify the aetiology of elevated blood pressure.

In patients with neurofibromatosis the fusion area has to be generous. The usual reason for failure of spine surgery is the implementation of technically inadequate anterior procedures, such as performing a short fusion or using a limited amount of bone graft [16, 43, 75, 77]. The entire structural area of the deformity should be fused anteriorly with complete disc resections, interposition of iliac crest and rib morselized autograft and additional strong strut grafting using fibular or rib autologous graft in the presence of kyphosis. All grafts

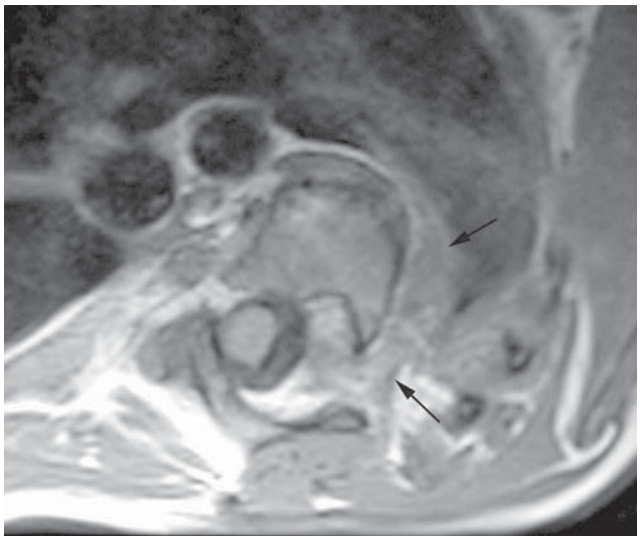

Fig. 10 Axial T1W MRI through the upper thoracic spine demonstrating plexiform neurofibromatous tissue (*arrows*) around the vertebral body, entering the left intervertebral foramen and associated with posterior vertebral scalloping

should have direct contact with the spine and with each other, while any intervening soft tissue should be meticulously excised. Bone grafts surrounded by abnormal neurofibromatous soft tissue demonstrate an increased tendency to resorb in the midportion [75]. Following the anterior stage of the procedure, posterior segmental instrumentation and iliac crest autogenous graft should be used to secure fixation and enhance a solid arthrodesis. The fusion should be extended to include the neutral vertebra above and below the curve [16]. During the posterior exposure of the spine, the surgeon should be particularly careful to avoid invading the spinal canal and injuring the cord in areas where the posterior bony elements are weakened due to the presence of intraspinal tumours or dural ectasia.

Segmental instrumentation can occasionally be challenging, since severely deformed vertebrae constitute poor anchorage points for fixation. If internal fixation is not technically feasible because of poor bone stock, in situ fusion with bone autograft and application of a postoperative cast or brace is necessary [43, 55]. If excessive angular kyphosis is present or if the vertebrae are weak due to the bony dysplasia, postoperative orthotic immobilization is recommended even if instrumentation has been successfully applied, in order to remove excessive strains at the proximal hook sites and prevent dislodgement of the implants [36, 43, 55, 62, 65]. Due to the high incidence of pseudarthrosis, evaluation of the fusion mass should be routinely performed at 6 months. If there is evidence of weakness of the fusion mass, posterior re-exploration and augmentation of the fusion should be undertaken [16, 55, 75, 77].

Scoliosis in the lumbar spine is relatively uncommon in NF-1. However, the principles of treatment do not differ. In the operative management of a lumbar curvature, it is important to rule out the presence of spondylolisthesis, which shows an incidence similar to that in patients without neurofibromatosis. In NF-1, spondylolisthesis can be the result of an increased diameter of the spinal canal, causing pathologic elongation and thinning of the pedicles and giving rise to an abnormal forward displacement of the anterior bony elements of the spine [16]. Distortion of the pedicles precludes reduction maneuvers using pedicle screws, while posterior fusion even after the application of autologous bone graft may be delayed, necessitating further reinforcement of the fusion mass at 6 months, if imaging studies show evidence of poor healing.

Deformities of the cervical spine in NF-1 have received little attention in the literature. They can cause neck pain and occasionally neurological complications, including nerve root compromise and complete or incomplete spinal cord deficits [11, 17, 55, 80]. However, in a large percentage of patients, cervical deformities are asymptomatic, therefore, anteroposterior and lateral radiographs should always be obtained, especially for patients scheduled to undergo instrumented fusion of the thoracic or lumbar spine or halo-dependent traction. If dystrophic features are identified, oblique radiographs can illustrate the presence of dumpbell lesions. Instrumentation and manipulation of the spine in the presence of undetected cervical intraspinal lesions can be extremely dangerous.

Kyphosis is the most common deformity occurring in the cervical spine in neurofibromatosis (Fig. 11). It is often the result of a previous excision of an intraspinal tumorous mass, which necessitated resection of the laminae and posterior elements creating secondary destabilization of the vertebral column and post-laminectomy kyphosis [13, 16, 43]. Anterior fusion with a combination of iliac crest and fibular autograft supplemented by halo vest or cast can achieve satisfactory results [55]. Alternatively, combined anterior-posterior spinal fusion with segmental fixation can provide adequate stability and avoid postoperative external immobilization. Atlantoaxial instability may also be present and evaluation for this condition is necessary using lat-

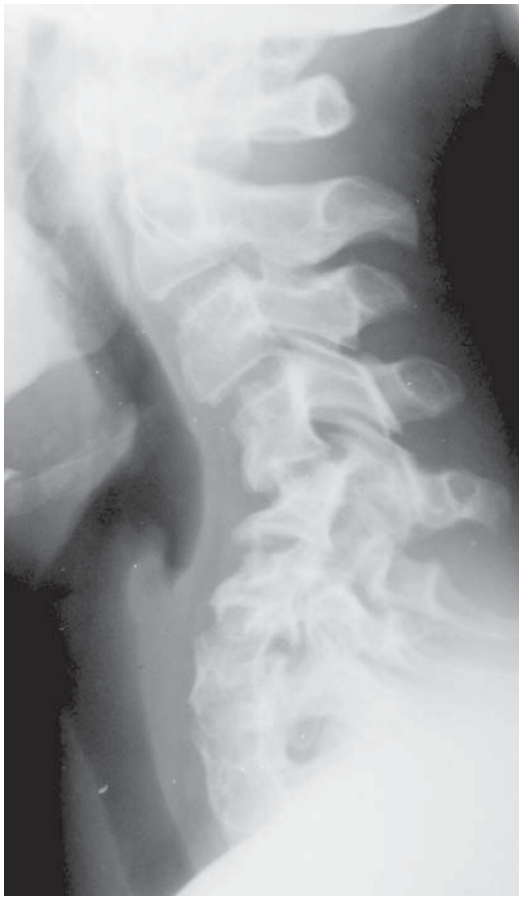

Fig. 11 Lateral radiograph demonstrating the classical cervical kyphosis of NF-1

eral cervical radiographs in flexion and extension, particularly if application of halo traction is anticipated.

In conclusion, spinal deformity in patients with neurofibromatosis type-1 poses a significant diagnostic and therapeutic dilemma. A thorough search for evidence of dysplastic changes on plain radiography and MRI is mandatory and will clarify prognosis and management options. MRI of the entire spine will illustrate the intraspinal contents and unveil the presence of intracanal anomalies that might interfere with any attempt for surgical correction of the deformity. Non-dystrophic curves are treated using the same principles applied in idiopathic scoliosis. On the contrary, dystrophic scoliotic curvatures or multiplanar spinal deformities with significant sagittal decompensation necessitate early aggressive surgical management, which should include an anteroposterior spinal arthrodesis with the use of segmental instrumentation and plentiful bone autograft. The primary goal of surgery in those cases is to stabilize the vertebral column and halt further progression of the deformity rather than perform heroic attempts for correction that could potentially result to permanent neurological injuries.

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Blood Loss in Duchenne Muscular Dystrophy: Vascular Smooth Muscle Dysfunction?

*M. H. H. Noordeen, F.R.C.S. (Orth), *F. S. Haddad, F.R.C.S., †F. Muntoni, F.R.C.P., †P. Gobbi, M.R.C.P., †J. S. Hollyer, Ph.D., and *G. Bentley, Ch.M., F.R.C.S.

**The Royal National Orthopaedic Hospital, Brockley Hill, Stanmore, Middlesex, U.K.; and †The Hammersmith Hospital, London, U.K.*

Summary: Patients with Duchenne muscular dystrophy (DMD) tend to bleed more during surgery than do patients with other conditions. A retrospective analysis of blood loss after spinal surgery for scoliosis was therefore undertaken in 102 patients undergoing surgery in the senior author's unit. These included 48 patients with DMD, 26 patients with spinal muscular atrophy, and a miscellaneous group of 28 other patients most of whom had idiopathic scoliosis. For each patient the age at surgery, estimated blood volume, duration of operation, Cobb angle, and number of vertebrae fused were recorded and compared. Expression of dystrophin in skeletal muscle and the underlying gene mutation were also determined. The estimated blood loss in patients with DMD was significantly higher than that in patients with spinal muscular atrophy undergoing the same or similar procedure ($P < 0.005$) and was also significantly greater than that of the third group, which consisted mostly of patients with idiopathic scoliosis ($P < 0.0005$). Blood loss in the patient group with DMD showed a significant relationship with duration of surgery ($P < 0.05$). As most patients expressed no dystrophin, this did not correlate with the estimated blood loss. There was also no correlation between the estimated blood loss and the type of gene mutation found causing DMD. The authors' previous observations confirm the increased blood loss in patients with DMD who undergo scoliosis surgery. Because children with DMD lack dystrophin in all muscle types, including smooth muscle, the excessive blood loss may be because of a poor vascular smooth muscle vasoconstrictive response due to a lack of dystrophin. *Key Words:* Blood loss—Duchenne muscular dystrophy—Scoliosis—Neuromuscular—Smooth muscle—Dystrophin.

Duchenne muscular dystrophy (DMD) is the most common lethal genetic disorder in boys. It is an X-linked recessive disease that ultimately leads to premature death. Although it has been known to be a distinct clinical entity for over a hundred years, the cause was unknown until the identification of dystrophin (8). Dystrophin, an integral membrane protein, is normally present in skeletal, cardiac, and smooth muscle as well as in cortical neurons but is absent in the corresponding tissues in boys with DMD. As a result, there is a severe and progressive degenerative process of all muscles that leads to a complete loss of contractile function. The lack of dystrophin in the central nervous system is the most likely cause for the mental retardation in DMD, with average IQ 1 SD below the normal range and mental retardation present in a third (10).

Although there is less information on the effect of dystrophin deficiency in smooth muscle, there have been reports on delayed gastric emptying (2) and fatal gastric distension (17) in patients with DMD. Constipation (16) and impaired bladder function (9) have also been described. Dystrophin is also expressed in arterial smooth muscle (8,11) and in particular the tunica media of blood vessels larger than arterioles such as muscular arteries, which possess thick smooth muscle layers (13). The absence of dystrophin from the arteries of children with DMD has been documented in the few patients who have been studied to date, but scant attention has been paid to its functional significance.

Children with DMD develop a progressively severe scoliosis that often requires instrumentation. The authors of the present study, along with other investigators (3,20), have noted that patients with DMD tend to bleed more during surgery than do patients with other conditions, including other neuromuscular disorders. The aims of this study were to formally document this observation and to investigate the hypothesis that dystrophin deficiency in arterial smooth muscle in DMD may lead to an increased bleeding tendency during surgery.

MATERIALS AND METHODS

A total of 102 consecutive patients who underwent scoliosis surgery in the senior author's unit were studied. All had spinal correction and stabilization with Harring-

Address correspondence and reprint requests to Dr. F. S. Haddad, 46B Hanover Gate Mansions, Park Road, London NW1 4SN, U.K.

ton-Luque distraction instrumentation with posterior fusion (36 patients) or using Luque instrumentation including fusion to the pelvis with crossed rods using the Galveston technique (66 patients).

The patients were divided into three groups: group 1—DMD (48 patients); group 2—spinal muscular atrophy (SMA, 26 patients); group 3—idiopathic or other etiology of scoliosis (miscellaneous group, 28 patients). For each patient the estimated blood volume (EBV), estimated blood loss (EBL), duration of surgery, Cobb angle, and the number of vertebral bodies involved were evaluated.

In all 48 patients, DMD was diagnosed according to the standard clinical and biochemical criteria (7). Screening for the deletion in the dystrophin gene was performed in all patients, and the majority (85%) also had a quadriceps needle biopsy as part of the diagnostic work-up; this was processed in line with standard techniques (6,7) including immunohistochemistry with antidystrophin antibodies. In addition, the quantity and quality of skeletal muscle dystrophin expression were determined.

In the SMA group, 20 were classified as having "intermediate" SMA, and the remaining 6 had "mild" SMA. In the third group of 28 patients that was used as a further control, 24 patients (85%) had idiopathic scoliosis. The other four patients had diagnoses of congenital muscular dystrophy, minicore disease, rigid spine syndrome, and Charcot-Marie-Tooth disease.

DNA Analysis

Screening for the dystrophin gene deletion was performed on genomic DNA using multiplex polymerase chain reaction methods and cDNA probe hybridization to Southern blot (7).

Gel Electrophoresis and Western Blotting

Muscle biopsy specimens were frozen in liquid nitrogen, crushed, and treated with a solubilization buffer containing protease inhibitors (5). A sample of solubilized muscle containing 60 μg of total protein was electrophoresed on 4% to 20% linear gradient polyacrylamide gels. The separated proteins were electroblotted onto nitrocellulose membrane; blots were then incubated with dystrophin antisera and diluted in phosphate buffered saline 0.05%, and bound antibody was visualized.

Antibodies

Antiserum P6 was polyclonal whole rabbit antiserum raised to a fusion protein (19). Monoclonal antibodies DYS 1 and DYS 2 were obtained from Novo Castra Laboratories (Newcastle upon Tyne, U.K.).

Immunocytochemistry

Muscle biopsy specimens were mounted transversely and frozen in isopentane cooled in liquid nitrogen. Unfixed frozen sections (5 μm) were incubated with the various dystrophin antisera for 30 minutes. Bound antibodies were detected as previously described (14,18).

Operative Intervention

All surgical procedures were performed by one surgeon (G.B.) between April 1983 and November 1993.

TABLE 1. *Baseline data for the Duchenne group*

	Range	Mean	Median	SD
Age at surgery (yr)	11–18	14.08	14.5	1.30
Preoperative Cobb angle	10–120	69.67	70.0	22.42
Number of vertebrae	11–17	13.02	14.00	2.31
EBV (mL)	1,500–6,150	3,477	3,368	1,208
EBL (mL)	700–8,000	2,977	2,500	1,667
Duration of surgery (min)	155–310	214	295	45.1

EBL, estimated blood loss; EBV, estimated blood volume; SD, standard deviation.
In the study group, n = 48.

The two procedures performed in this study were Luque instrumentation and Harrington-Luque instrumentation with posterior fusion (4). Indications for the former generally included patients that were wheelchair bound. Indications for the latter included patients with less severe neuromuscular disorders or those who were thought likely to remain ambulant. As a result, of the 74 patients in the DMD and SMA groups, all had Luque-Galveston instrumentation except for 8 who underwent the Harrington-Luque procedure. The less severely affected patients in the miscellaneous group all had Harrington-Luque procedures except for one.

Normal adult blood volume is roughly equal to 75 mL/kg but may vary by 10% depending on body morphology (1). Estimated blood volume was therefore calculated using this equation where the weight (in kilograms) was multiplied by 75 to determine EBV (in milliliters).

Estimated blood loss values were retrieved from the anesthetic records at time of operation and from surgical postoperative notes. The duration of the operation was defined as the time from incision to closure and was also determined from anesthetic and surgical records.

The Cobb angle was derived from spinal radiographs. The angles were all measured from the views taken with the patient seated, not suspended. Only the dominant curve's Cobb angle was used in the statistical analysis. The number of vertebral bodies involved was inferred from the same radiograph, taking into account the highest and lowest vertebrae involved.

TABLE 2. *Baseline data for the spinal muscular atrophy group*

	Range	Mean	Median	SD
Age at surgery (yr)	5–13	9.46	9.0	2.32
Preoperative Cobb angle	45–150	91.15	90.00	23.16
Number of vertebrae	11–17	14.04	14.00	1.34
EBV (mL)	700–4,500	1,936	1,500	1,012
EBL (mL)	350–3,500	1,437	1,150	782
Duration of surgery (min)	120–275	203.2	205	36.56

EBL, estimated blood loss; EBV, estimated blood volume; SD, standard deviation.
In the study group, n = 26.

TABLE 3. *Baseline data for the miscellaneous group*

	Range	Mean	Median	SD
Age at surgery (yr)	8–21	14.5	14.5	3.20
Preoperative Cobb angle	30–128	78.14	77.5	23.2
Number of vertebrae	10–18	11.93	12.00	2.18
EBV (mL)	1,500–5,400	3,121	3,100	994.6
EBL (mL)	700–3,500	1,568	1,350	758.2
Duration of surgery (min)	125–290	209.81	200	60.33

EBL, estimated blood loss; EBV, estimated blood volume; SD, standard deviation.

In the study group, n = 28.

Statistical Analysis

A stepwise linear regression was performed on the data. A log transformation was required for EBL and cubes were taken of the number of vertebral bodies. The EBL was taken as the variable of interest and the others as explanatory variables. A P value of less than 0.05 was taken as the level for statistical significance.

RESULTS

The baseline patient data for the three groups are presented in Tables 1, 2, and 3. The significant results of the multiple linear regression analysis are summarized in Table 4.

The EBL in the DMD group is significantly higher than values in the two other groups (medians: 2500 versus 1150 and 1350; $P < 0.005$). The analysis is complicated by the fact that, in the SMA group, the age at operation and hence the EBV are notably lower with respect to the other two groups. In the miscellaneous group, although the EBV is similar to the DMD group, the EBL is considerably lower. The results show that the group the patient belongs to is a significant explanatory variable for EBL in all models that include DMD, but not in the model containing the SMA and miscellaneous groups.

Neither age nor the number of vertebrae fused has a significant association with EBL, when considered with the other variables, in any of the models. Duration of the surgery has a significant association with the EBL only in the smaller models containing the patients with DMD. Analysis of the DMD group alone shows that duration of surgery was significantly associated with EBL within this group ($P = 0.0023$).

Although the preoperative Cobb angle has some value as an explanatory variable, it is significant only in the models containing both DMD and miscellaneous groups, perhaps as a consequence of a significant difference in the angle in patients in these groups. Similarly, EBV is significant only in the models that include the SMA group. As EBV is calculated by measurement of body weight, this is significantly lower in the SMA group, as is the patient age, which may account for this.

Thus, the EBL has a predominant dependence on patient group and is significantly greater in the DMD group than in the SMA or miscellaneous groups. The other variables have differing predictive values depending on the model used and the groups included. Estimated blood loss also shows a significant relationship to duration of surgery within the DMD group.

Dystrophin immunocytochemistry performed with the various antidystrophin antibodies showed that no patients with DMD produced dystrophin at the periphery of their skeletal muscle fibers. In approximately one third of the patients with DMD, the occurrence of rare revertrant fibers, a common phenomenon in DMD, was noted. Western blot analysis confirmed the lack of dystrophin expression in the skeletal muscle of patients with DMD.

Forty-eight percent of patients with DMD had a deletion of one or more exons of the dystrophin gene. The majority (75%) of the patients had a deletion in the central region of dystrophin, and only a minority had a deletion at the 5′ end, a trend that is almost identical to the one observed in the general DMD population (7). All patients had out-of-frame deletions. There was no correlation between the location of the intragenic deletion and the EBL.

DISCUSSION

The absence of dystrophin in skeletal and cardiac muscle in DMD leads to a number of clinical sequelae: all children with DMD are wheelchair bound by the age of 12 years and ultimately die of cardiac or respiratory failure. Patients with DMD require surgery for progressive scoliosis. A tendency to excessive blood loss has been noted before but not formally documented.

This study was undertaken to ascertain whether the

TABLE 4. *The stepwise multiple linear regression analysis*

	All groups	DMD and SMA	DMD and Miscellaneous	SMA and Miscellaneous
Preoperative Cobb angle	0.022		0.008	23.2
Group	<0.0005	0.005	<0.0005	
EBV (mL)	<0.0005	0.005		0.002
Duration of surgery (min)		0.007	0.012	

DMD, Duchenne muscular dystrophy; EBV, estimated blood volume; SMA, spinal muscular atrophy.

Where there is no P value or where the variable is omitted (age, number of vertebrae), the variable did not make a significant contribution to the fit of the model and was therefore omitted.

apparent increase in blood loss in patients with DMD was a consistent finding. The EBL in a group of patients with DMD was compared with values in a group of patients with SMA, a condition affecting exclusively the anterior horn motor neurons. A third miscellaneous group, consisting mainly of patients with idiopathic scoliosis, was also studied and compared with both the DMD and the SMA groups. The results indicate that there is a statistically significant increase in EBL in the DMD relative to the other two groups who have similar blood losses. This confirms that patients in the DMD group have a greater predisposition to blood loss during surgery. Because the type of surgical intervention, the duration of surgery, and the number of vertebral bodies fused did not differ significantly between the DMD and SMA groups, this suggests that DMD is a condition associated with increased blood loss.

Because patients with DMD do not produce a physiologic protein, dystrophin, that is produced in all muscle types, it is possible that the increased blood loss observed in these patients is related to a lack of dystrophin in arterial smooth muscle. It is unclear whether dystrophin-deficient smooth muscle cells have a primary contractility problem or whether this lack of contractility is related to degeneration of the smooth muscle cells and subsequent replacement by connective and adipose tissue, as occurs in skeletal and cardiac muscle. There is evidence that the smooth muscle histology of the gastrointestinal tracts in patients with DMD resembles the abnormalities found in skeletal and cardiac muscle (2). However, to date, no structural abnormalities, other than the absence of dystrophin, have been noted in the vasculature (15). Mechler et al. (12) reported their experience with Becker muscular dystrophy, a milder variant of DMD in which dystrophin can be partially produced. Adrenergic beta-receptor vascular smooth muscle responses were normal, but alpha-receptor responses were abnormal. This also suggests an intrinsic problem in vessel contractility in patients with DMD.

Children with DMD have been shown to lose significantly more blood during surgery when compared with children with other neuromuscular conditions and this blood loss is dependent on the duration of surgery. Although there is no direct explanation for this, the fact that dystrophin is physiologically expressed in arterial smooth muscle, and that it is absent in patients with DMD, allows us to hypothesize that increased blood loss is a consequence of vascular smooth muscle dysfunction. Although further studies are necessary to define the precise mechanisms responsible for this excessive bleeding, clinicians should be aware that patients with DMD experience more severe blood loss than other scoliosis patients. This should be taken into account when planning surgery in this condition.

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Spinal Cord Monitoring in Neuromuscular Scoliosis

Stuart K. Tucker, F.R.C.S. (Orth.),* M. H. H. Noordeen, F.R.C.S. (Orth.),*† and Matthew C. Pitt, M.D., F.R.C.P.†

**The Royal National Orthopaedic Hospital Trust, Stanmore; and †The Hospital for Sick Children, Great Ormond Street, London, UK*

Summary: This article reviews the use of spinal cord monitoring in neuromuscular scoliosis, a condition having a higher incidence of true positive results than idiopathic scoliosis. While somatosensory cortical evoked potentials (SCEP) are unreliable, somatosensory spinal evoked potentials (SSEP) are possible to obtain in most cases and a method using an epidural electrode is described. The '50% rule' is satisfactory having good specificity and sensitivity with it rare for post-operative paralysis to have occurred undetected. The spinal cord in these cases appears to have increased susceptibility particularly during the passage of sublaminar wires with the incidence of complications reduced using modern instrumentation. **Key Words:** Spinal cord monitoring—Scoliosis—Neuromuscular Scoliosis.

Intraoperative monitoring of spinal cord function is now a routine accompaniment to complex spinal surgery, providing a continuous means to assess the integrity of the spinal cord. The recovery of postoperative neurologic deficits shows a directly proportional relationship to the speed of removal of instrumentation (11). Theoretically, earlier detection of spinal cord dysfunction allows immediate intervention and the possible prevention of any neurologic deficit.

In most spinal surgery units, spinal cord monitoring has replaced the Stagnara wake-up test. The latter, however, is still of clinical importance should there be a significant change in intraoperative monitoring. However, the wake-up test is not always practical in patients with neuromuscular scoliosis, owing to muscle weakness and mental retardation (18).

Somatosensory cortical evoked potentials (SCEP) have been found to be unreliable and nonspecific in neuromuscular scoliosis. Somatosensory spinal evoked potential (SSEP) monitoring, however, is useful for the detection of neurologic injury. The detection of true-positive results is higher in neuromuscular scoliosis, when compared with idiopathic scoliosis. In neuromuscular patients, SSEP monitoring is less sensitive and specific, and there are more false-negative results. An amplified loss, less than 50% of baseline, may still be associated with significant neurologic deficit.

Address correspondence to M.H.H. Noordeen FRCS, 107 Harley Street, London, WIN 1DG.

METHOD

The recording technique currently in use at The Hospital for Sick Children is described. Evoked potential recordings are made, at the time of operation, using a commercial electromyography (EMG) and evoked potential machine. Two disposable, adhesive and pre-gelled electrodes are applied to each popliteal fossa with the cathode proximal, and an earth electrode placed on the posterior thigh. These are connected to the stimulator outputs of the preamplifier.

The evoked potentials are recorded with a 4 F bipolar electrode, inserted into the epidural space, cephalad to the highest proposed level of fusion, by either direct incision of the ligamentum flavum, through a large bore intravenous cannula or via an epidural needle placed by the anaesthetist before skin incision.

The legs are stimulated alternately with the stimulus intensity increased until a reproducible signal could be identified. If no signal is obtained, technical failure must be excluded before concluding that the response is absent. The stimulation duration is 0.5 ms and the rate 20 Hz, with 100 stimuli per test. The recording parameters are: low-frequency filter, 200 Hz; high frequency, 2 kHz; input sensitivity, 50 μV cm; and average sensitivity, 0.5 μV cm.

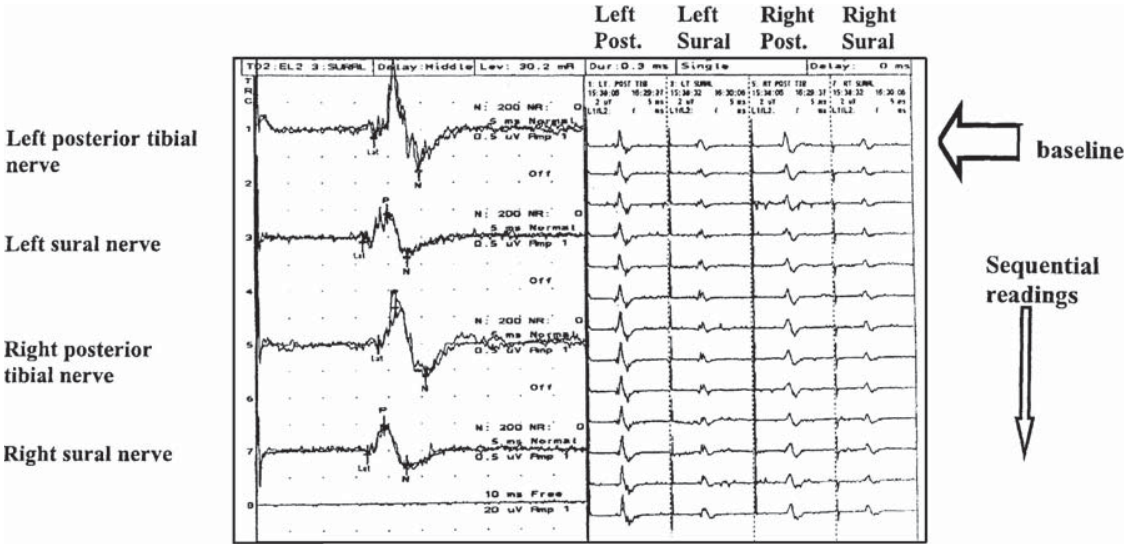

FIG. 1. Typical visual display.

Cursors are applied manually and the maximum amplitude difference of the polyphasic waveform measured, usually of the second or third waveform. The latency is measured to the start of the response. Recordings are made repeatedly, with the stimulus alternating between the two lower limbs (Fig. 1).

A failure of monitoring is represented by no recordable responses after all known cases of technical error have been excluded. Achievement of responses from only one lower limb is not considered a technical failure.

INTERPRETATION OF SPINAL CORD MONITORING

Approximately one-quarter of monitored patients show a fall of 10% to 50% in sensory evoked potential (SEP) amplitude during surgery. A 60% fall in peak-to-peak amplitude of the evoked potential wave carried a 10-fold increase in the risk of neurologic complication (10). The latency of the evoked potential and the duration of the recorded waveform are of no significance in the prediction of neurologic outcome (8).

The present rule is that an amplitude fall of 50% from the initial baseline should be considered significant (3,9,16) and carries a risk of spinal cord injury (Table 1).

False-positive SSEPs are not infrequent during intraoperative monitoring and are usually caused by technical problems or environmental factors unrelated to surgical manipulation. These include equipment failure, electrode movement, alterations in blood pressure, depth of anaesthesia and temperature changes.

TABLE 1. *Classification of sensory evoked potentials* (15)

Classification	Definition
True-negative	No change in trace No neurologic sequelae
False-negative	No change in trace Neurologic sequelae
True-positive	Change in trace that can be related to surgery; does not necessarily mean neurologic sequelae
False-positve	Change in trace cannot be related to surgical or anaesthetic events No neurologic sequelae

Hypotension

Hypotensive anaesthesia has significantly reduced operative blood loss and blood-product requirement. It does, however, have a deleterious effect, especially on SCEPs, but may affect SSEPs to a lesser degree. Anaesthesia appears to have a significant effect on the amplitude of SCEPs; especially those recorded from patients undergoing surgery for neuromuscular scoliosis.

Decreased body temperature will typically increase the latency of a response because of slowing in conduction velocity. A gradual increase in latency

TABLE 2. *Modes of neurologic injury*

Direct
Sublaminar wire hook passage
Indirect
Distraction instrumentation
Derotation instrumentation
Ischaemia

is common during an operative procedure owing to cooling of the patient (7). Latency increase without significant loss of SEP amplitude is not associated with neurologic complications (Table 2).

DISCUSSION

SSEPS are primarily an indication of dorsal column integrity and do not provide information about the more clinically relevant motor pathways. However, motor evoked potential (MEP) monitoring has not yet been refined sufficiently to facilitate its use routinely. SSEP monitoring therefore relies on intraoperative events affecting descending motor pathways producing concurrent changes in dorsal column activity.

Postoperative paralysis has been reported despite normal SCEP monitoring perioperatively (2,6). In the majority of these cases, the operated deformity was principally kyphosis, which accounts for 30% of cases of major neurologic complication (11). It is likely that the cause of neurologic injury in such cases is vascular, with infarction, initially of the anterior portion of the cord, explaining the immediate postoperative finding of motor paralysis despite normal SEPs during monitoring. Experimentally, motor cordotomy has been performed without SEP change (13).

It is generally assumed that, in the absence of a 50% fall of epidural SEP amplitude, which is not rapidly reversible, the risk of clinically significant motor tract damage must be exceedingly low. However, the need for concurrent, reliable MEP and SSEP monitoring is clear.

Ashkenaze et al. (1) found SCEP monitoring to be unreliable and nonspecific in 101 patients undergoing neuromuscular scoliosis surgery, and considered it nonefficacious in preventing or detecting spinal cord injury when used alone. Of note in their study was the inability to obtain adequate baseline tracings in 28% of their patients.

Owen et al. (14) found that, with the use of multiple recording sites, a reliable cortical or subcortical SEP could be obtained in 90% of neuromuscular patients. Furthermore, with the addition of MEPs, a reliable response was present in 96% of patients. It was possible to obtain reliable myogenic or neurogenic MEPs in all patients with some motor function demonstrable before surgery. Owen et al. (14) considered that the combined use of multiple recording sites and multiple modalities improved their capability to monitor spinal cord function successfully in patients with neuromuscular scoliosis.

Williamson and Galasko (18) documented 60 neuromuscular patients 30 Duchenne muscular dystrophy (DMD), 6 myelomeningocele, 6 spinal muscular atrophy (SMA), 7 cerebral palsy, 11 miscellaneous conditions . They obtained satisfactory SSEP traces in all but two patients (one FA, one myelomeningocele). Using the classification of Szalay (15), true negative responses occurred in 43 patients and true-positive responses in 15 patients, invariably associated with the tightening of a sublaminar wire. In 11 of these patients, the SEP trace returned to its control latency and amplitude after loosening of the wire, which was later tightened uneventfully. In one patient, the SEP trace did not recover until all the sublaminar wires had been loosened. Instrumentation was therefore abandoned and an in situ fusion performed, without consequence. Szalay (15) found no false-positive and no false-negative results, and in no case did the postoperative neurologic state differ from the preoperative state.

The largest reported series is that of Forbes et al. (5). They detailed 1168 consecutive cases with SSEP monitoring in scoliosis surgery, of which 21% were congenital or neuromuscular curves. No technically adequate tracing could be obtained in 26 patients (2.2%).

In neuromuscular scoliosis and, particularly, the cerebral palsy and Charcot–Marie–Tooth groups, altered neural pathways between the applied stimulus and the sensory cortex may account for the high rate of nonreproducible tracings. The higher success rate of obtaining a reproducible waveform in DMD and polio patients reflects the underlying integrity of the sensory pathway being tested.

One hundred and nineteen (10.5%) patients showed a drop in amplitude of greater than 50% of the initial SEP on one or both sides at some time during the operation. In 35 of these patients, the loss of trace resolved spontaneously or when the recording electrode was repositioned. None of this group suffered neurologic complications. The remaining 84 patients with persistent depression of more than 50% included all but one of the 33 patients (2.7%) who suffered neurologic complications. Of the group with persistent SSEP decrements, 38% had neuromuscular scolioses.

The overall incidence of SSEP fall was 7.2% and that of neurologic complication was 2.7%. These incidences were significantly higher in patients with neuromuscular scoliosis, most due to DMD or SMA, than for patients with idiopathic and osteogenic curves (P 0.01).

All patients who sustained neurologic complications at operation recovered to some degree. Most of the serious episodes were related to distraction of the spine or to wire placement and tightening. Of the 84 patients with significant SEP decrements, 53 received instrumentation with an applied distraction force. All the neuromuscular patients received Luque instrumentation.

This series has a highly significant false-negative rate (P 0.001), suggesting that, in the absence of

technical failures, SSEP traces that remain above 50% of their initial value will not be associated with clinically detectable neurologic complications.

An SSEP fall of 50% or greater in initial SSEP amplitude should be considered significant. Greater falls in SSEP amplitude are generally associated with a higher incidence of neurologic complications. Although recovery of amplitude is encouraging, it does not guarantee that there will be no neurologic complications.

Noordeen et al. (12) retrospectively reviewed 99 consecutive patients who underwent reconstructive spinal surgery for neuromuscular scoliosis (55 DMD, 30 SMA, 14 miscellaneous conditions), with SSEP intraoperative monitoring. Traces could not be obtained in only two patients (2%), one with DMD and the other with myelomeningocele and paraplegia.

They studied the significance of any diminution in amplitude with three levels; namely, 25%, 50% and 75% of the initial baseline. In the DMD group, 25% loss of amplitude showed a 90% sensitivity, with 295 specificity and 2 false-negative results; 50% loss of amplitude had 87 sensitivity with 44% specificity and 3 false-negative results; and 75% loss of amplitude had a 70% sensitivity with 52% specificity and 7 false-negative results. Results were comparable in the SMA and miscellaneous groups.

Noordeen et al. concluded that a loss of amplitude of greater or equal to 50% of the initial baseline should be used to define abnormality in neuromuscular scoliosis, as with idiopathic scoliosis.

The incidence of neurologic injury in neuromuscular scoliosis is much higher than in idiopathic scoliosis. In the neuromuscular group, an amplitude recovery greater than 50% may still be associated with significant, although temporary, neurologic sequelae. Both these findings suggest an increased sensitivity of the spinal cord to injury in the neuromuscular patient.

An amplitude loss of greater than 50% in the DMD group seen in 35 patients (69%) was attributable to sublaminar wiring in all cases (34 during wire tightening, 1 during wire passage). Neurologic injury was detected in 22 cases but was temporary in all.

Within the SMA group, 73% had a loss of amplitude greater than 50%, attributable to sublaminar wiring in 95% (81% wire tightening, 14% wire passage). Fifteen (of 22) patients had some degree of neurologic impairment.

In the authors' experience with third-generation instrumentation (pedicle screw hook rod constructs), there has been no loss of amplitude in 20 consecutive patients operated on for Duchenne muscular dystrophy. This is additional evidence to the previous reporting of the sensitivity of neuromuscular patients to the passage of sublaminar wires (4,17).

CONCLUSION

Spinal cord monitoring can reliably be obtained in approximately 98% of patients with neuromuscular scoliosis, using SSEPs with epidural placed electrodes rather than SCEPs. A decline in amplitude of 50% of the initial baseline reading should be considered significant.

The occurrence of postoperative motor paralysis, despite normal intraoperative SEPs, although reported, is extremely low. However, this reinforces the urgency for the development of reliable MEP monitoring in accompanied of SSEP monitoring.

The spinal cord appears to have an increased susceptibility to injury in neuromuscular patients, especially during the passage and tightening of sublaminar wires. The rates of diminution in SSEP amplitudes previously reported are much lower using modern pedicle screw hook rod constructs.

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SPINAL CORD MONITORING IN OPERATIONS FOR NEUROMUSCULAR SCOLIOSIS

M. H. H. NOORDEEN, J. LEE, C. E. R. GIBBONS, B. A. TAYLOR, G. BENTLEY

From the Royal National Orthopaedic Hospital, Stanmore, England

We reviewed retrospectively the role of monitoring of somatosensory spinal evoked potentials (SSEP) in 99 patients with neuromuscular scoliosis who had had operative correction with Luque-Galveston rods and sublaminar wiring.

Our findings showed that SSEP monitoring was useful and that a 50% decrease in the amplitude of the trace optimised both sensitivity and specificity. The detection of true-positive results was higher than in cases of idiopathic scoliosis, but the method was less sensitive and specific and there were more false-negative results. In contrast with the findings in idiopathic scoliosis, recovery of the trace was associated with a 50% to 60% risk of neurological impairment.

Only one permanent injury occurred during the use of this technique, and any temporary impairment resolved within two months.

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Intraoperative monitoring of the spinal cord is commonly used to help to diminish the risk of neurological injury. Somatosensory recording of the spinal evoked potential (SSEP) is a popular method which offers continuous surveillance and is generally used during operations for idiopathic scoliosis.[1] Its role in neuromuscular scoliosis, however, is less well defined.[2,3] We have therefore assessed the value of SSEP monitoring in a large series of patients with this condition.

PATIENTS AND METHODS

We retrospectively reviewed 99 consecutive patients who had reconstructive surgery of the spine for neuromuscular scoliosis between April 1983 and December 1994. The operations had all been performed by one surgeon in the same unit with the same anaesthetist. All the patients had a one-stage Luque-Galveston procedure with crossed rods and sublaminar wires. Of the 99 patients, 55 had Duchenne's muscular dystrophy (DMD), 30 had spinal muscular atrophy (SMA), and 14 had miscellaneous conditions including congenital myopathies, spinal dysraphism and other rare syndromes. Table I gives the details of the groups.

We reviewed the records of each patient and recorded the age at operation, gender, diagnosis, the operative approach, the instrumentation and the levels fused. In addition, we noted the magnitude, flexibility, percentage correction and level of coronal and sagittal spinal curves fused, the anaesthetic details and intraoperative events, the neurological function after operation and any adverse postoperative symptoms.

Spinal cord monitoring. Measurement of the SSEP was used to monitor the function of the spinal cord throughout the operation. Stimulation of the posterior tibial nerve with supramaximal constant-voltage single-pulse stimuli of 0.2 ms, at 20 per second, was applied alternately to both legs. The SSEP was recorded with a 4F bipolar electrode (Orthoexpress, Amersham, UK) from the epidural space at the level above the highest proposed level of fusion. The signals were amplified, conditioned and displayed on a Medelec MS91 (Medelec, Old Woking, UK) and a PA89 preamplifier. The filters were set to BER (200 to 2000 Hz) and sweeps were averaged to obtain clear recordings at intervals coinciding with steps in the operation or changes in the signal. Hard copies of the traces were made on calibrated paper.

The SSEP was measured as the maximum peak-to-peak value, and its amplitude related to different points in the operation. The amplitude at insertion of the electrode was taken as the control level and the amplitudes at the insertion of sublaminar wires, correction (tightening of the wires) and at closure of the skin were measured and expressed as a percentage of the control value.

Three levels of loss of signal, namely more than 25%, 50% or 75% of the control were used to judge the sig-

M. H. H. Noordeen, FRCS, Consultant Orthopaedic and Spinal Surgeon
J. Lee, FRCA, Anaesthetic Registrar
B. A. Taylor, FRCS, Consultant Orthopaedic and Spinal Surgeon
G. Bentley, FRCS, Professor of Orthopaedic Surgery
The Institute of Orthopaedics and the Royal National Orthopaedic Hospital Trust, Brockley Hill, Stanmore, London HA7 4LP, UK.

C. E. R. Gibbons, FRCS, Orthopaedic Registrar
Royal Surrey County Hospital, Egerton Road, Guildford, Surrey GU2 5XX, UK.

Correspondence should be sent to Mr M. H. H. Noordeen.

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Table I. Details of the patients with neuromuscular scoliosis

	Group		
	DMD	**SMA**	**Miscellaneous**
Mean age at operation in years (range)	14.08 (11 to 18)	9.46 (5 to 13)	13.78 (8 to 21)
Mean Cobb angle in degrees (range)	69.67 (10 to 120)	91.15 (50 to 150)	74.10 (30 to 128)
Mean number of vertebrae instrumented (range)	13.02 (11 to 17)	14.04 (11 to 17)	11.99 (10 to 18)

nificance of any diminution in amplitude.

The SSEP traces were designated as false-positive, false-negative, true-positive or true-negative, defined as follows:
False-positive. A change in the trace unrelated to the position of the electrode or an anaesthetic event but with no neurological injury detected.
False-negative. No change in the trace but a neurological injury was produced.
True-positive. A change in the trace unrelated to the position of the electrode or an anaesthetic event with neurological injury detected.
True-negative. No change in the trace and no neurological injury detected.

The neurological injuries included change in muscle power in the lower limb, change in sensation in the trunk or in the lower limb, or a change in bladder control.

Anaesthesia. Similar techniques of anaesthesia were used. Induction was either by intravenous injection of thiopentone or by inhalation of halothane and was followed by maintenance with enflurane and nitrous oxide in oxygen. Vecuronium or pancuronium was given for paralysis and doses of opioid were given peroperatively for analgesia. The systolic blood pressure was maintained at about 70 mmHg, and labetolol or trimetephan was occasionally given to facilitate this. All patients had intra-arterial monitoring, and most also had a central venous pressure transducer.

Statistical methods. We analysed the relationship between reduction of the trace to less than 50% of the control value and the neurological injury, the flexibility of the curves, the Cobb angle before operation and the percentage correction obtained using the Mann-Whitney U test. Comparison of the systolic blood pressures and the temperatures were made by Student's two-tailed *t*-tests.

RESULTS

Traces could not be obtained in two of the 99 patients; one had DMD and the other was paraplegic due to meningomyelocoele.

DMD group. There were 51 sets of traces for the 55 patients available for analysis (Fig. 1); 40 (79%) had lost trace amplitude by more than 25% of the control, 35 (69%) by more than 50% and 30 (59%) by more than 75% of the control. The results were as follows: 25% loss of amplitude showed a 90% sensitivity with 29% specificity and two false-negative results; 50% loss of amplitude had 87% sensitivity with 44% specificity and three false-negative results; and 75% loss of amplitude had a 70% sensitivity with 52% specificity and seven false-negative results.

Neurological injury was detected in 22 cases (43%) (Fig. 2)

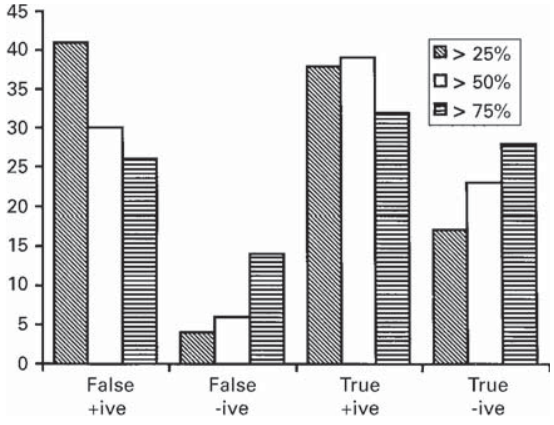

Fig. 1

Accuracy of SSEPs in the DMD group (n = 51) at loss of amplitude of more than 25%, 50%, and 70% of the control.

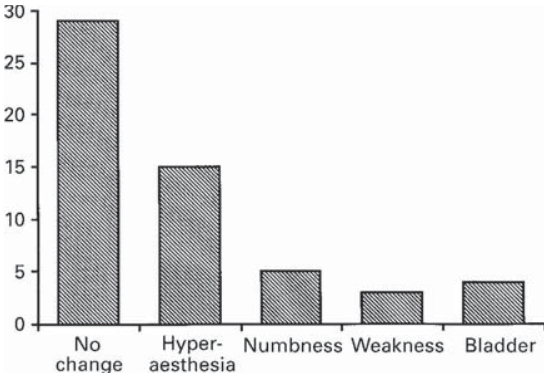

Fig. 2

Postoperative neurological status in the DMD group (n = 51).

but was temporary in all of them. There were three false-negative results at 50% amplitude loss: one patient had hyperaesthesia on the abdomen for one week and in the legs and feet for three weeks, one had hyperaesthesia in a leg for two weeks and another had hyperaesthesia of the lower abdomen for two days.

Of the 35 patients with SSEP loss of over 50%, this occurred during wire tightening and correction in 34 and during passage of the wire in one. In 13 patients although the SSEP amplitude improved between correction and skin closure, six of these had neurological impairment.

SMA group. Examination of 30 sets of traces available for the patients with spinal muscular atrophy (Fig. 3) showed that 26 (87%) had a loss of amplitude of more than 25%, 22 (73%) of more than 50% and 20 (67%) of more than 75%. The results were as follows: 25% loss of amplitude had a sensitivity of 93% with specificity of 19% and one false-negative result, 50% loss of amplitude had a sensitivity of 93% with specificity of 42% and one false-negative result, and 75% loss of amplitude had a sensitivity of 82% with a specificity of 52% and three false-negative results.

There were 14 cases of neurological injury (47%) (Fig. 4), but all except one resolved. Loss of amplitude of 50% occurred on wire tightening and correction in 24 (81%) and on passing wires in four (14%). Of the 22 patients with loss of amplitude greater than 50%, 15 had some degree of neurological impairment.

Miscellaneous group. The miscellaneous group consisted of patients with a variety of syndromes and congenital myopathies. There were 13 traces available for analysis in the 14 patients; nine had lost amplitude by more than 50%, giving a sensitivity of 86% and a specificity of 54% (Fig. 5). There was one false-negative result. The pattern of neurological damage is shown in Figure 6, but there were no cases of permanent injury.

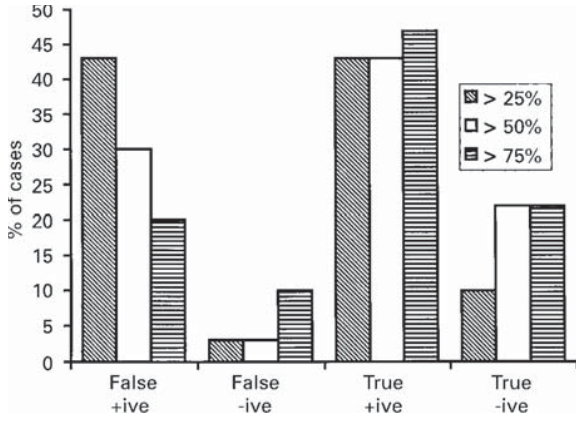

Fig. 3

Accuracy of SSEPs in the SMA group (n = 30) at loss of amplitude of more than 25%, 50% and 70% of the control.

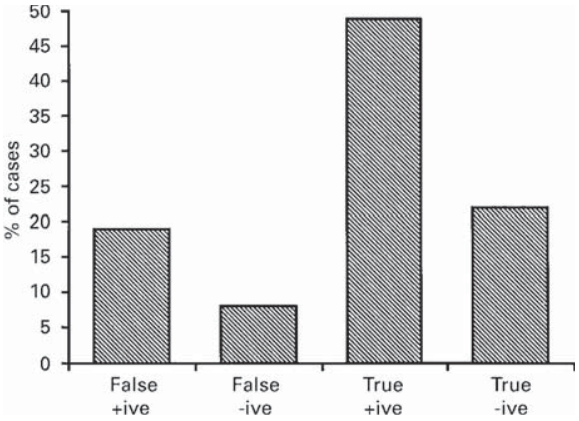

Fig. 5

Accuracy of SSEPs in the miscellaneous group (n = 13) at loss of amplitude of more than 25%, 50% and 70% of the control.

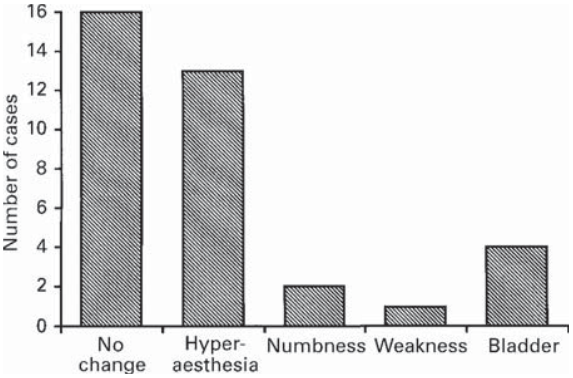

Fig. 4

Postoperative neurological status in the SMA group (n = 30).

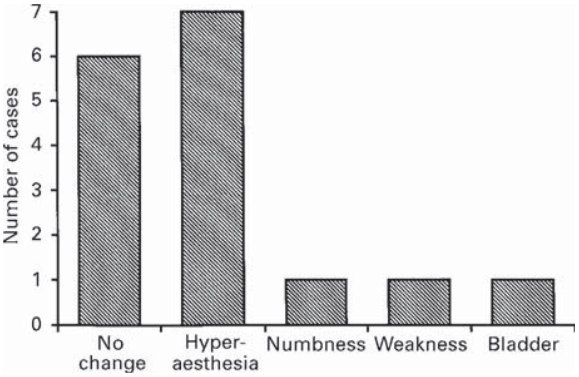

Fig. 6

Postoperative neurological status in the miscellaneous group (n = 13).

Table II. Characteristics of the curves in degrees (mean, range) in the DMD and SMA groups

Group	Preoperative Cobb angle	Flexibility	% correction obtained
DMD (n = 51)	69.3 (10 to 122)	24.8 (0 to 70)	57.2 (15 to 100)
SMA (n = 30)	93 (40 to 100)	20 (5 to 70)	48.4 (18 to 80)

Table III. Physiological measurements (mean, SEM) in patients with neurological injury compared with those with no injury

	Systolic blood pressure (mmHg)		
Group	Lowest	Highest	Core temperature at skin closure (°C)
No neurological injury (n = 28)	71.7 (2.1)	112.4 (3.3)	36.2 (0.2)
Neurological injury (n = 45)	68.5 (1.4)	107.6 (1.9)	36.2 (0.2)

In all three groups of patients there was a 100% correlation between the side on which the amplitude decreased and the side of the neurological deficit.

In both the DMD and SMA groups, the Mann-Whitney U test showed no significant correlation between amplitude loss of greater than 50% or neurological injury and the preoperative Cobb angles, the flexibility curve or the percentage correction of the curve obtained during the operation (Table II).

Most patients with bladder dysfunction had retention of urine; those with permanent damage had intermittent retention.

Physiological details were available for 73 (74%) of the patients. Those who had neurological impairment were compared with those who did not, using Student's two-tailed *t*-test (Table III). No significant difference was obtained when comparing the highest or the lowest systolic blood pressures or the core temperatures at skin closure.

DISCUSSION

Segmental instrumentation of the spine carries a high risk of neurological injury.[4] Intraoperative monitoring of the function of the spinal cord includes wake-up techniques and the recording of evoked potentials. Some patients, however, continue to suffer neurological complications despite monitoring.[5]

A study of SSEP monitoring of the spinal cord in 1168 consecutive patients with idiopathic scoliosis using techniques identical to those in our series showed an inability to obtain adequate tracings in 26 patients (2.2%) which compares well with our figure of 2.1%.[1] The incidence of technical failure was 28% using cortical somatosensory evoked potentials in a series of patients with neuromuscular scoliosis.[6]

In the series of Forbes et al[1] 119 patients (10.2%) with an amplitude loss greater than 50% of the control value had detectable neurological injury with a sensitivity of 100% and a specificity of 57.8% (Table IV). In 35 (29.4%) of these, however, the SSEP improved spontaneously to more than 50% of the control level and none had neurological injury. There were no false-negative results. In our series there was a high rate of trace loss, temporary neurological injury and some false-negative results, as shown by the reduced sensitivity. Forbes et al,[1] however, used broader categories than in our study and the results of neurological impairment are therefore difficult to compare directly.

An amplitude loss greater than 25% diminished the rate of false-negative results but increased the false-positives, diminishing specificity. The use of loss of amplitude greater than 75% reduced the number of false-positive results, but resulted in an unacceptable number of false-negatives. We therefore suggest that a loss of amplitude of 50% or more should be used to define abnormality in neuromuscular scoliosis.

The incidence of neurological injury in neuromuscular

Table IV. Comparison of idiopathic and neuromuscular scoliosis

	Group			
	Forbes et al [1]	DMD	SMA	Miscellaneous
Number of patients	1168 (26 failures; 2.2%)	55	30 (2 failures for all three groups; 2.1%)	14
Loss of trace >50%	119 (*10.2%*)	35 (*69%*)	22 (*73%*)	9 (*69%*)
Recovery of this degree of loss (%)	*29.4* of which none had neurological complications	*37* of which 46% had neurological complications	*36* of which 68% had neurological complications	*33* of which 66% had neurological complications
Neurological complication	32 (*2.8%*)	22 (*43%*)	14 (*47%*)	6 (*46%*)
Long-term neurological impairment	6 (*0.5%*)		1 for all three groups; 1%	
True-positive results (%)	*26.9*	*37.0*	*44.0*	*46.0*
False-negative results (%)	*0.0*	*6.0*	*3.0*	*7.0*
Sensitivity (%)	*100.0*	*80.0*	*88.5*	*77.0*
Specificity (%)	*57.8*	*52.8*	*59.5*	*66.0*

scoliosis seems to be much higher than in idiopathic scoliosis. In the former group an amplitude recovery above 50% may still be associated with significant, although temporary, neurological sequelae which may be due to an increased sensitivity of the spinal cord to injury, caused by the segmental wiring or to both. Luque-Galveston instrumentation using sublaminar wires has been associated with a significantly higher incidence of trace changes.[6]

We conclude that SSEP monitoring in neuromuscular scoliosis is useful for the detection of neurological injury in contrast to the use of somatosensory cortical evoked potentials which have been found to be unreliable and non-specific. Although the detection of true-positive results is higher in neuromuscular scoliosis when compared with idiopathic scoliosis, the method is, however, less sensitive and specific.

No benefits in any form have been received or will be received from a commercial party related directly or indirectly to the subject of this article.

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Chapter 7

IDIOPATHIC SCOLIOSIS

Advances in Surgical Techniques for EOSD

Paper: 20

Adolescent idiopathic scoliosis (AIS) is the most common spinal deformity and is most commonly seen amongst girls (male:female ratio being 1:10). The exact cause is not known, though recent studies have identified many genes associated with causation of AIS. I have published a review article in British Medical Journal (BMJ) to guide general practitioners, and other healthcare professionals in evaluating patients presenting to their practice with scoliosis. The article describes the manifestations and indications for surgery, guided by clinical evaluation, comprehensive assessment and special investigations (magnetic resonance imaging [MRI] and integrated shape and imaging system [ISIS] scans).

Curves less than twenty degrees are treated by close observation with serial six monthly x-rays, whereas twenty to fifty degrees are treated with bracing. The brace is worn full-time, and it provides a rigid, three-point fixation, supporting the growth of the spine over time, which minimizes the progression of the deformity. Surgery is offered for curves greater than fifty degrees and posterior spinal fusion with segmental fixtion using pedicle screws and judicious use of hooks and wires, has been a gold standard in the contemporary era.

I have also developed a simplified algorithm to aid surgeons in choosing fusion levels, while performing posterior spinal fusion, which takes into consideration the shoulder heights. Unlike Lenke's classification, which has six main curve patterns with a total of fifty four sub-types that are very cumbersome to apply, the new algorithm has only three main curve patterns, with a total of six curve sub-types. The validated algorithm in 177 cases was presented at International Meeting on Advanced Spinal Techniques (IMAST) in 2010.

Adolescent idiopathic scoliosis. Altaf F, Gibson A, Dannawi Z, Noordeen H. BMJ. 2013 Apr 30; 346: f2508. PMID:23633006

Advances in Surgical Techniques for EOSD

Paper: 21

Idiopathic scoliosis could be complicated with abnormalities in neuraxis (Chiari malformation, syrinx, and so forth). A whole spine MRI is undertaken prior to surgery in most patients. Asymmetrical or absent superficial abdominal reflexes (SAR) has been reported to be associated with an occult syrinx or a neuraxial anomaly. However, true incidence of a SAR abnormality, and its relevance with respect to neuraxial anomaly in scoliotic groups, was unknown until a prospective study was undertaken under my supervision.

Amongst seventy three patients comprising of infants (3), juveniles (23), and adolescents (47), who were prospectively evaluated in my out-patient clinic with idiopathic scoliosis (eleven males and sixty two females), eight of them had abnormal SAR (an incidence of eleven percent). Two out of eight patients (twenty five percent) had an absence of SARs (one patient each had unilateral and bilateral absence). Both patients had Chiari-I malformation with syrinx on their magnetic resonance imaging (MRI) scans. Absence of SAR on convexity of curve or bilateral absence was found to be highly associated with the underlying neuraxial anomaly warranting a mandatory MRI prior to surgical intervention to minimize the risk of post-op neurodeficit or intra-op loss of neuromonitoring signals.

This was one of the first studies in English medical literature to emphasize the importance of a meticulous clinical examination, and how subtle clinical examination findings may have wide surgical implications in the assessment of patients presenting with spinal deformities.

Prevalence and clinical significance of superficial abdominal reflex abnormalities in idiopathic scoliosis: Saifuddin A, Tucker S, Taylor BA, Noordeen MH, Lehovsky J. Eur Spine J. 2005 Nov; 14(9): 849-53. E-pub 2005 Mar 9. PMID: 15756608

Advances in Surgical Techniques for EOSD

Paper: 22

Syringomyelia is characterised by an abnormal dilatation of the central canal, resulting in a *balaclava* pattern of loss of sensations in the scalp and upper extremities. It can be present in any segment of the spine (i.e. cervical, thoracic, or lumbar), and may be associated with Chiari malformation.

The erstwhile and now obsolete Harrington instrumentation system was less tolerated by the neural tissues, and the presence of syringomyelia was a risk factor for spinal deformity correction by use of such distraction systems. I had published a case of an eighteen year old male with scoliosis and co-existent syringomyelia, who had a reduction in somatosensory evoked potentials (SSEP) and spinal cord evoked potenitals (SCEP) amplitude during his corrective surgery in 1994, warning other surgeons of this intra-op neurological event, and offering valuable insight into early preventive steps that can be taken to minimize neurological risk:

i) Remove all distraction

ii) Conversion of hypotension to normotension to prevent ischemia to the cord

I recommended a fifty percent reduction in amplitude from baseline to be a threshold to take aggressive intra-op steps to facilitate neurological recovery. The index patient had a temporary retention of urine, and he did not suffer from any permanent neurological deficit at the final follow-up following excellent post-op care.

Syringomyelia. A potential risk factor in scoliosis surgery. Noordeen MH, Taylor BA, Edgar MA. Spine (Philadelphia, PA 1976). 1994 Jun 15; 19(12): 1406-9. PMID: 8066525

Advances in Surgical Techniques for EOSD

Paper: 23

Idiopathic scoliosis has traditionally been classified as infantile, juvenile, and adolescent (James JI, 1954). Juvenile idiopathic scoliosis (JIS) is characterized by the presence of a spinal deformity between three to eight years old. I undertook a prospective clinical study (Level of Evidence [LoE] II; Prognostic study) evaluating all JIS patients by a detailed clinical assessment, a comprehensive neurological examination, and magnetic resonance imaging (MRI) scans of the neuraxis in a series of thirty one patients (seven males and twenty four females). The mean age at first assessment of scoliosis in a specialist unit was 8.6 years in males and 9.1 years in females.

Of these thirty one patients, nineteen of them had single, and the remaining twelve had double structural curves with a mean Cobb angle of forty degrees (range: ten to one hundred degrees). All patients had pre-operative or pre-bracing MRI scans with a 1.5T scanner. Eight patients (two males and six females) had neuraxial anomalies (six patients Chiari I malformation with syrinx, one patient each had isolated Chiari malformation and astrocytoma with syrinx.) Surprisingly six of the eight patients (seventy five percent) had normal neurology, and only two patients had abnormal neurology on clinical examination. One patient had a unilateral absence of superficial abdominal reflexes (SAR), and one with astrocytoma had ataxia with nystagmus. Four of the six patients with Chiari Malformation had left sided curves.

This was one of the first studies to emphasize the need for mandatory MRIs in all patients presenting with JIS despite normal neurology given the risks associated surgery. Left sided curves and absence of SAR were more likely to have neuraxial anomalies.

MRI of 'idiopathic' juvenile scoliosis. A prospective study. Evans SC, Edgar MA, Hall-Craggs MA, Powell MP, Taylor BA, Noordeen HH. J Bone Joint Surg Br. 1996 Mar; 78(2): 314-7. PMID: 8666649.

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CLINICAL REVIEW

Adolescent idiopathic scoliosis

Farhaan Altaf *specialist registrar trauma and orthopaedics*, Alexander Gibson *consultant spinal surgeon*, Zaher Dannawi *spinal fellow*, Hilali Noordeen *consultant spinal surgeon*

Spinal Surgery Unit, Royal National Orthopaedic Hospital, Stanmore, London HA7 4LP, UK

Scoliosis is a three dimensional deformity of the spine defined as a lateral curvature of the spine in the coronal plane of more than 10°.[1] It can be categorised into three major types—congenital, syndromic, and idiopathic. Congenital scoliosis refers to spinal deformity caused by abnormally formed vertebrae. Syndromic scoliosis is associated with a disorder of the neuromuscular, skeletal, or connective tissue systems; neurofibromatosis; or other important medical condition. Idiopathic scoliosis has no known cause and can be subdivided based on the age of onset—infantile idiopathic scoliosis includes patients aged 0-3 years, juvenile idiopathic scoliosis includes patients aged 4-10 years, and adolescent idiopathic scoliosis affects people aged >10 years.

Adolescent idiopathic scoliosis (AIS) is the most common spinal deformity seen by primary care physicians, paediatricians, and spinal surgeons.[2] This review is focused on AIS and reviews the diagnosis, management, and controversies surrounding this condition based on the available literature.

What causes adolescent idiopathic scoliosis?

The diagnosis of AIS is one of exclusion, and is made only when other causes of scoliosis, such as vertebral malformations, neuromuscular disorders, and other syndromes have been ruled out. According to epidemiological studies, 1-3% of children aged 10-16 years will have some degree of spinal curvature, although most curves will not require surgical intervention.[3 4]

Suggested causes of AIS include mechanical, metabolic, hormonal, neuromuscular, growth, and genetic abnormalities.[5 6] These factors are not yet well accepted as a direct cause for this condition. The current view is that AIS is a multifactorial disease with genetic predisposing factors.

What is the natural course of adolescent idiopathic scoliosis?

The natural course of scoliosis was studied in a prospective case series of 133 patients. The patients were followed for an average of 40.5 years (range 31-53 years), and 68% of adolescent idiopathic curvatures were found to progress beyond skeletal maturity. Thoracic curvatures greater than 50° progressed at an average of 1° a year, thoracolumbar curves progressed at 0.5° a year, and lumbar curves progressed at 0.24° a year. Thoracic curvatures of less than 30° did not progress.[7]

Previous long term retrospective observational studies of idiopathic scoliosis presented a poor prognosis (respiratory failure, cardiovascular risk, and mortality).[8] This has created a misinterpretation that all types of idiopathic scoliosis inevitably lead to disability from back pain and serious cardiopulmonary compromise. These studies included patients with mixed diagnoses, which could explain the poor outcomes reported. In a more recent prospective case-control study describing the 50 year natural course of untreated idiopathic scoliosis, there was no evidence linking untreated AIS with increased rates of mortality in general, and cardiopulmonary compromise in particular.[9]

Progressive scoliosis can result in the development of a worsening deformity and cosmesis.[10] The physical deformities seen include the development of chest wall abnormality, rib prominences, asymmetry in shoulder height, and truncal shift.

How does adolescent idiopathic scoliosis present?

Patients with AIS most often present with unlevel shoulders, waist line asymmetry (one hip “sticking out” more than the other), or a rib prominence. This is usually first identified by the patient, family member, general practitioner, or a school nurse.

Back pain is sometimes the presenting complaint. The association between scoliosis and back pain has been demonstrated in a retrospective study of 2442 patients with idiopathic scoliosis,[11] which found that 23% of patients with AIS had back pain at initial presentation, and another 9% developed back pain during the study. An underlying pathological condition was identified in 9% (48/560) of the patients with back pain, mainly spondylolysis and spondylolisthesis and only one case of an intraspinal tumour.[11]

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Summary points

- Scoliosis is a lateral curvature of the spine measuring >10° in the coronal plane
- Several different types of scoliosis exist, and idiopathic scoliosis occurs in 0.5-3.0% of the paediatric population
- Initial evaluation should involve a focused history and physical examination. The Adam's forward bend test is particularly useful for detection
- Factors predicting curve progression include maturity (age at diagnosis, menarchal status, and the amount of skeletal growth remaining), curve size, and position of the curve apex
- Bracing is used to treat scoliosis in many European countries, but practice is divided in the UK and US, and elsewhere
- Surgery is recommended in adolescents with a curve of a Cobb angle more than 45°-50°

Sources and selection criteria

We searched Medline and the Cochrane Library using MeSH terms "adolescent idiopathic scoliosis", and "scoliosis bracing". We included systematic reviews, randomised controlled trials, and good quality prospective observational studies mainly from the past 15 years but did not exclude seminal papers from before this time.

How is adolescent idiopathic scoliosis diagnosed?

On presentation of a patient with scoliosis to primary care, a detailed history, examination, and radiological investigations should be undertaken before referral to a specialist.

The history should include a detailed birth history, developmental milestones, family history of spinal deformity, and assessment of physiological maturity. Difficulties during labour can be associated with a diagnosis of cerebral palsy, which can lead to neuromuscular scoliosis. A history of developmental delay can be indicative of a non-idiopathic cause for the scoliosis.

Assessment of maturity includes inquiry about the growth spurt and the menarchal status in girls, as menarche indicates a point at which the growth starts to decrease over a period of two years from its onset.[12]

The patient's presenting complaint should be elicited, including back pain, neurological symptoms, and any concerns regarding cosmesis. The presence of constant pain, night pain, or radicular pain indicates that further investigations are required to exclude underlying pathology.[13]

When examining a patient with suspected scoliosis, adequate exposure is required to assess the spine appropriately. Boys should be examined in their underwear or shorts; girls should be wearing underwear and a bra. Gait and posture should be evaluated, looking in particular for a short-leg gait due to leg length discrepancy and listing to one side seen in severe curves.

The patient's upright posture should be evaluated from the front, back, and sides. The relative heights of the iliac crests and the shoulders should be observed for any asymmetry that could be indicative of curve severity. The pelvis should be level and any lower limb discrepancy compensated with a lift (a series of wooden blocks may be placed under the short leg until the hips are level). If a curvature of the spine is seen, the location and direction of the curve(s) should be noted. The curve is designated according to the direction of the curve convexity.

The back should be inspected for the presence of cafe au lait spots, subcutaneous nodules, and axillary freckles, which are seen in neurofibromatosis. The presence of hairy patches or skin dimples over the lower back can be an underlying sign of spinal dysraphism (a constellation of congenital abnormalities including defects of the spinal cord and vertebrae).

The balance of the thorax over the pelvis is assessed by dropping a plumb line from the C7 spinous process, which normally falls within the gluteal cleft. In cases of coronal imbalance the distance from the plumb line to the gluteal cleft is measured in centimetres and the direction of deviation noted.

The Adam's forward bend test[14] is carried out to assess the degree of rotational deformity associated with the scoliosis. The patient is asked to bend forward at the waist with the knees straight and the palms together (fig 1⇓). The examiner looks down the back for the presence of asymmetry in the rib cage (rib prominence) or deformities along the back indicative of a structural scoliosis. A non-structural curve (postural scoliosis) normally disappears on bending forwards.

A scoliometer is an instrument that is placed on the back and can be used to provide an objective measure of curve rotation.[15] In primary care the use of a scoliometer is not required for the diagnosis of scoliosis, and suspected cases should be referred for specialist opinion on diagnosis.

A detailed neurological examination should be performed testing motor and sensory function and reflexes. Asymmetries in reflexes can be a sign of an intraspinal disorder.[16] The abdominal reflex refers to the neurological reflex stimulated by stroking the abdomen around the umbilicus. This usually involves a contraction of the abdominal muscles, resulting in the umbilicus moving towards the source of the stimulation. An abnormal abdominal reflex may be suggestive of an intraspinal disorder and is often absent on the convex side of the curve.

What imaging is required?

Full length standing posteroanterior and lateral radiographs of the spine are required in order to assess the degree of deformity. These are taken with the patient in a standing position in order to assess the effect of gravity on the deformity. Patients are instructed to remove their shoes, and any lower limb discrepancy is compensated with a shoe lift before the radiograph is taken. Radiographs are taken with the patient looking straight ahead, legs apart for stability and with their hands on clavicles. If a radiograph is normal the patient and family can be reassured that there is no scoliosis. A referral can still be made if there is concern about pain, axial tenderness, or neurological abnormalities. If x ray facilities are not available, the patient may be referred directly to the specialist without radiographs.

On a full length posteroanterior plain radiograph, the magnitude of a scoliosis curvature is determined with the Cobb technique (fig 2⇓). Firstly, it is important to identify the superior and the inferior end vertebrae—the vertebrae with the greatest tilt at the proximal and distal ends of the curve. The angle between them is measured by drawing a line from the top of the superior end vertebra parallel to the upper endplate, and another line from the bottom of the inferior end vertebra parallel to the lower

endplate. Perpendicular lines are then constructed at right angles to the lines along the endplates. The angle formed by the intersection of the perpendicular lines defines the Cobb angle (fig 2⇓).

If surgery is considered, films of lateral bending view (full length posteroanterior plain radiographs with patient bending to the right and to the left) are first taken to determine curve flexibility, which is important in the preoperative evaluation and surgical planning.

The presence of a left thoracic curve or an abnormal neurological finding are most predictive of the presence of an underlying disease and warrant referral for further imaging.[11] Magnetic resonance imaging is useful for the identification of tumours and other pathological lesions—associated neural axis abnormalities such as syrinx (a fluid filled cavity within the spinal cord) and Arnold-Chiari malformations.[18]

What are the risk factors for curve progression?

For decisions about choosing conservative or surgical treatment, the child's maturity and the severity of the curvature are the two most important factors. It is important to evaluate maturity because the younger the child the greater is the likelihood of curve progression, equally the larger the curve magnitude the greater is the risk of progression.[9]

Scoliosis with a high risk for rapid progression must be detected as early as possible. In a retrospective case series of 205 patients (163 girls and 42 boys) with idiopathic scoliosis at skeletal maturity, the surgical risk for a curve of 20° at the onset of puberty was at 16%. This surgical risk increased to 100% for curves ≥30° at the onset of puberty.[19] The table⇓ summarises the risk factors for curve progression.

Scoliosis curve progression increases markedly at the time of the adolescent growth spurt in idiopathic curves and markedly slows or ceases at the time of completion of growth.[20-22] Spinal growth is closely associated with increase in height, but the measurement of height velocity at sequential visits is often associated with inaccuracies. Other maturity markers are therefore often used to measure the growth rate. The use of these maturity markers allows us to determine which curves are at risk of progression. This information allows the clinician to differentiate between curves that require careful regular monitoring and ones that require active treatment.

The total growth spurt has a duration of about 2.5-3.0 years,[3] with the mean age for peak height velocity being about 14 years in boys and 12 years in girls.[23]

Sexual maturity can be evaluated with the Tanner grading scale,[24] which is based on the extent of development of secondary sexual characteristics. It is important to ask about menarche because curve progression is less common after its onset.

Skeletal age is a more accurate marker of maturity. The Risser sign,[25] which refers to the appearance of the iliac apophysis of the pelvis, can be used to determine skeletal age. There are six Risser stages, from zero to five, denoting the course of the apophysis from the anterior to the posterior iliac spine, and then the fusion with the iliac bone (fig 3⇓).[23] The incidence of progression of untreated AIS has been correlated with Risser sign and curve magnitude.[26] For curves of 20°-29° in a immature child with a Risser sign of 0 or 1, the incidence of progression was 68%. For curves <19° in a mature adolescent with a Risser sign of ≥2, the incidence of progression was 1.6%. For small curves <19° in an immature child (Risser sign 0 or 1), and larger curves (20°-29°) in a mature child (Risser sign ≥2), the incidence of progression was about the same, at 22% and 23% respectively.[26] The disadvantages of the Risser sign are that it correlates with skeletal age differently in boys and girls and it typically appears after the peak height velocity.

Skeletal age can also be assessed by evaluating the development of the left hand and wrist on a radiograph: the bones are compared with those of a standard atlas compiled by Greulich and Pyle.[27] Sanders found that the scoring of the metacarpals and phalanges more closely related to scoliosis progression than other maturity indicators, including Tanner stage and Risser sign.[23] Dimeglio et al described elbow maturation as being more precise than hand maturation.[28]

How is adolescent idiopathic scoliosis managed?

Observation for AIS is the most common approach used for patients with mild deformity (such as a Cobb angle measurement <25°). Depending on the degree of skeletal maturity, patients are assessed every four to six months at a specialist clinic to watch for curve progression. The interval of follow-up will be determined on an individual basis, based on the age of the patient, degree of curve, and skeletal maturity. Posteroanterior radiographs only are taken during each follow-up visit in order to minimise the exposure to radiation.

Bracing

Bracing in AIS is controversial, with treatment effectiveness remaining questionable based on available evidence, with most published studies being of low methodological quality. The rationale for the use of braces has been that external forces can guide the growth of the spine. Brace treatment is not necessarily benign in terms of the psychosocial and body image concerns it causes for many patients and their families. Bracing is used for the treatment of scoliosis in many centres in continental Europe, but practice is divided in the UK and US, and elsewhere.

Advocates of bracing quote level 2 evidence based information from prospective controlled studies[29-31] as well as other studies with level 3 and 4 information[32-34] in support of bracing efficacy. In a meta-analysis a total of 1910 patients had non-operative treatment for idiopathic scoliosis, with 129 patients managed with observation only.[34] The analysis concluded that bracing was effective in altering the natural course of scoliosis. In 1995, a prospective, multicentre, non-randomised, non-blinded study also showed the effectiveness of bracing in girls with curves of 25°-35°.[30]

Other studies have shown less positive results. A prospective case series of 102 immature patients with idiopathic scoliosis reported that bracing provided curve correction in only 15% of patients, while 42% later became surgical candidates.[35]

The primary goal of bracing for scoliosis is to halt curve progression. The most widely accepted practice for brace treatment suggests that patients with curves of 25°-45° and in the most rapidly growing stage (Risser stage 0 or 1) should be offered a brace on initial evaluation. Curve progression is defined as an increase in the magnitude of the deformity by more than 5° at consecutive follow-up appointments of between four and six months.

Various factors can hinder successful brace treatment. Poor adherence is common. A meta-analysis reported that a protocol of 23 hours/day was more successful than protocols of 16 hours/day or night time use.[34] A multidisciplinary team approach involving the patient's general practitioner, surgeon, orthotist,

physiotherapist, and parents is needed to improve adherence. Families must be counselled that there is a risk that bracing may not be successful, but that the chances of success are improved with discipline and adherence to wearing the brace for the recommended time. Patients who have passed the peak height velocity, are within a year of skeletal maturity, or are a year or more after menarche are unlikely to benefit from use of a brace.

When should surgery be considered?

About 10% of adolescents with idiopathic scoliosis will progress to a level requiring consideration of surgery.[36] Surgery is generally indicated to treat a significant clinical deformity or to correct a scoliotic deformity that is likely to progress. Surgery is recommended in adolescents with a curve that has a Cobb angle greater than 45°-50°. This recommendation is derived from studies that have shown that curves >50° tend to progress slowly after maturity.[11] The decision to proceed with surgical correction therefore needs to take into consideration the clinical assessment, comorbid conditions, the wishes of the patient, and the effects the scoliosis has on the patient's quality of life. It is not clear that surgery is an effective treatment for back pain associated with scoliosis.

The aims of surgery may be to arrest curve progression by achieving a solid fusion, to correct the deformity, and to improve cosmetic appearance. If the decision is taken to operate, the usual approach in AIS is posterior (fig ⇓). In this approach a longitudinal posterior midline incision is used. Pedicle screws are inserted into the spine and two metal rods are measured and contoured. Curve correction is achieved as the two metal rods are attached and tightened on to the pedicle screws. An anterior fusion is used in AIS either as the sole approach in thoracolumbar or lumbar curves or in conjunction with posterior fusion in special cases.

Surgical treatment of AIS has a low rate of non-union and other complications. The incidence of neurological complications for spinal deformity surgery has been estimated by the Scoliosis Research Society at <1%.[10] A more recent prospective clinical case series of 1301 patients reported a neurological complication rate of 0.69%.[37] A long term case-control study of scoliosis curves fused to the lumbar spine evaluated pain and functional status of AIS patients with a minimum of 10 years' follow-up (average 19 years).[38] These patients were compared with a control population matched for work, age, and recreational activities. The two groups did not differ with respect to functional status or pain.

After surgery it is important to check for abnormal neurology and for bowel and bladder symptoms. Back pain after surgery is not uncommon, especially if it is mechanical in nature. In the presence of continuous or night pain, infection or non-union should be considered, and referral to a specialist is advised.

Postoperative follow-up often involves clinical and radiological reviews at six weeks, three months, six months, and one year. These intervals and will vary between institutions, but follow-up until completion of growth is common.

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Additional educational resources

Resources for healthcare professionals

Scoliosis Research Society website. www.srs.org

AAOS American Academy of Orthopaedic Surgeons. Adolescent idiopathic scoliosis: etiology, anatomy, natural history, and bracing. *Instructional Course Lectures* 2005;54:529-36.

Resources for patients

Scoliosis Association United Kingdom (SAUK). www.sauk.org.uk—Provides patient information on the condition and treatments

Scoliosis Research Society. www.srs.org/patient_and_family—Patient and family section provides information on the condition, treatments, and outcome

Tips for non-specialists

Postural scoliosis can be differentiated from structural scoliosis with the Adam's forward bend test: the curvature will disappear on forward bending in postural scoliosis

If scoliosis is seen in a premenarchal female there is a higher risk of curve progression, and early referral to a specialist is advised

Patients undergoing brace treatment for scoliosis must be encouraged to adhere with brace treatment. Patients must be informed that the brace can be removed for washing and swimming

Table

Table 1| Risk factors for curve progression in adolescent idiopathic scoliosis

Risk factor	Comment
Age	The younger the age at diagnosis, the greater potential for curve progression at the onset of adolescent growth spurt
Sex	Progression is more common in girls
Menarche	Progression is least common after menarche
Remaining skeletal growth	More skeletally immature the greater risk of curve progression
Curve pattern	Double curves are more likely to progress than single curves
Curve magnitude	The risk of progression increases with curve magnitude

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Figures

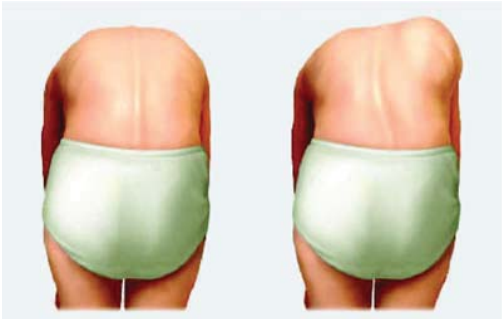

Fig 1 The Adam's forward bend test performed by (left) a patient without scoliosis, and (right) a patient with scoliosis showing a rib prominence

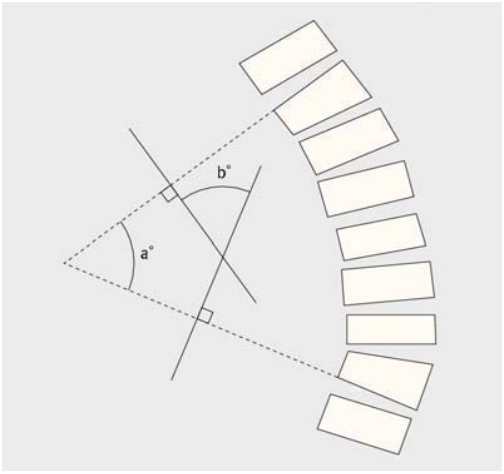

Fig 2 Cobb technique for determining size of a scoliosis curvature. On a posteroanterior view of the spine, tangents (dashed-dotted lines) are drawn along the superior endplate of the superior end vertebra and the inferior endplate of the inferior end vertebra. The angle formed (angle a) by the intersection of these two lines is the Cobb angle. This is more conveniently measured as the angle (b) formed by the intersection of two lines drawn perpendicular to the tangents. Adapted from Kim et al[17]

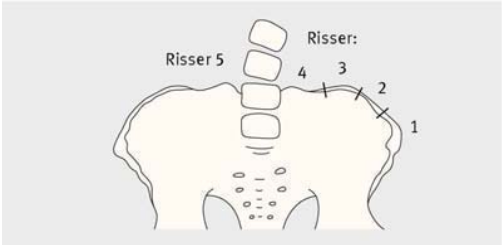

Fig 3 Illustration of the six Risser stages of skeletal age, from 0 to 5, denoting the course of the apophysis from the anterior to the posterior iliac spine, and then the fusion with the iliac bone

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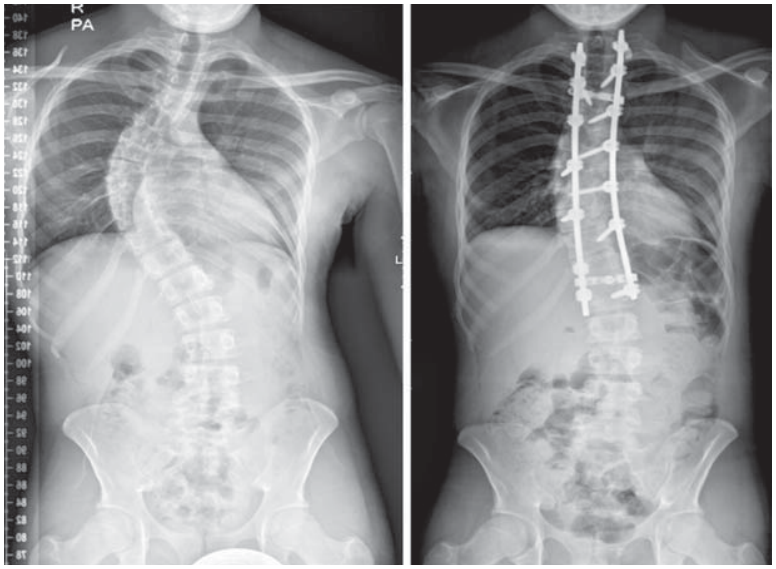

Fig 4 Preoperative (left) and postoperative (right) radiographs of an adolescent boy with idiopathic scoliosis, showing correction of the scoliosis by posterior instrumented fusion of the spine

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ORIGINAL ARTICLE

Asif Saifuddin
Stuart Tucker
Benjamin A. Taylor
M. Hilali Noordeen
Jan Lehovsky

Prevalence and clinical significance of superficial abdominal reflex abnormalities in idiopathic scoliosis

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A. Saifuddin (✉)
The Department of Radiology, The Royal National Orthopaedic Hospital NHS Trust, Brockley Hill, Stanmore, HA7 4LP Middlesex, UK
E-mail: ASaifuddin@aol.com
Tel.: +44-20-89095443
Fax: +44-20-89540281

S. Tucker · B. A. Taylor
M. H. Noordeen · J. Lehovsky
The Department of Spinal Surgery, The Royal National Orthopaedic Hospital NHS Trust, Brockley Hill, Stanmore, HA7 4LP, Middlesex, UK

Abstract To determine prevalence and significance of abnormal superficial abdominal reflexes (SARs) in idiopathic scoliosis. Study of 73 patients with presumed idiopathic scoliosis referred for magnetic resonance imaging (MRI), either as a routine pre-operative assessment ($n=42$) or because of abnormal symptoms or neurological signs ($n=31$). All patients were examined prior to magnetic resonance imaging (MRI), and the presence of abnormal SARs was noted. All patients then underwent MRI of the whole spine from the foramen magnum to the sacrum. The presence of Chiari 1 malformation and syrinx was recorded. The study group consisted of 11 males and 62 females with a mean age at time of MRI of 18 years (range 5–51 years) and a mean Cobb angle of 48° (range 10–104°). Abnormality of the SARs was recorded in eight cases (prevalence 11%). An abnormal MRI study was recorded in nine cases (12.3%), all patients having a syrinx and four having in addition, a Chiari 1 malformation. Of the patients with abnormal SARs, only 2 (25%) had an abnormal MRI study; 1 had unilateral absence of the reflexes whereas the other had complete absence of SARs. Of patients referred for MRI as a routine pre-operative assessment, 5 (11.6%) had an abnormal MRI study. In patients with idiopathic scoliosis, abnormality of the SARs was recorded in 11% of cases. Unilateral absence was present in one case only and was associated with the presence of syrinx. Other patterns of abnormality were not a useful indicator of underlying cord abnormality.

Keywords Scoliosis · Abdominal reflexes · Syrinx · Chiari malformation · MRI

Introduction

A proportion of children and adolescents with presumed 'idiopathic' scoliosis have an underlying abnormality of the neuraxis, particularly Chiari 1 malformation and syrinx [2, 7, 9, 13, 18]. Several studies have indicated the value of magnetic resonance imaging (MRI) in the investigation of 'idiopathic' scoliosis in selected groups of patients. Factors considered to be important indicators of underlying brainstem and spinal cord abnormality include infantile and juvenile onset [6, 8, 11], left thoracic curves [4], severe curves [3] and abnormal neurological examination [1]. Abnormal superficial abdominal reflexes (SARs) have been reported to be a specific early sign of occult syrinx. Zadeh et al. [21] described 12 children with abnormal SARs, of whom 10 had a syrinx identified by means of MRI. Ghanem et al. [10] reported 12 children with Chiari 1 malformation

and syrinx, 7 of whom had scoliosis and all of whom had abnormal SARs. These reports are retrospective and complicated by the fact that there were additional risk factors for cord abnormality, particularly early age of onset and other abnormal neurological signs. For these reasons, it is difficult to know the true relevance of abnormal SARs as an isolated sign of underlying syrinx. The problem is compounded by reports of 'abnormality' of the SARs in normal individuals [20].

The aim of the present study was to determine the prevalence and clinical relevance of abnormal SARs in a consecutive group of patients with presumed idiopathic scoliosis who underwent whole spine MRI.

Materials and methods

This was a study of patients with presumed idiopathic scoliosis who were referred for whole spine MRI. Patients were imaged as a routine prior to corrective surgery, as part of standard clinical practice, despite normal neurological examination (group 1), or due to the presence of atypical clinical symptoms or examination findings (group 2). Prior to MRI referral, patients underwent a complete neurological examination by experienced scoliosis surgeons, particularly noting the status of the SARs. Other data collected included the sex of the patient, the age of onset of the curve, presence of symptoms such as headache and spinal pain and the age at which MRI was performed. All patients in group 1 were asymptomatic, had normal neurological examinations, including SARs, and had typical idiopathic curves. Patients in group 2 had symptoms, abnormal neurological signs or atypical curve patterns, typically left thoracic curves. Patients were also classified into infantile (IIS), juvenile (JIS) and adolescent (AIS), depending on age at onset, with infantile being 0–3 years, juvenile 4–11 years and adolescent over 12 years. Cobb angle was recorded from the radiograph obtained at the time of referral for MRI.

MRI studies were performed at 1.0 T field strength with the whole spine imaged from the foramen magnum to the sacrum using a combination of sagittal, axial and coronal T1-weighted spin echo (W SE) and T2-weighted fast spin echo (W FSE) sequences. All MRI studies were reported by experienced spinal radiologists, with particular note made of the presence of Chiari I malformation and syrinx. The relationship between abnormalities of the SARs and MRI findings was determined.

Results

A total of 73 patients were included in the study, 62 females and 11 males. Patients presented over a period of 21 months. The mean age of the whole group at time of referral for MRI was 18 years (range 5–51 years). Three children were classified as IIS, 23 as JIS and 47 as AIS. Of the patients, 42 were asymptomatic and had normal neurological examination, being referred for MRI as a routine pre-operative assessment (group 1); 31 were referred for assessment of a variety of symptoms and signs (group 2). Of these 31, 8 had an abnormality of the SARs, giving a prevalence of SAR abnormalities of 11%. Details of these patients are presented in Table 1. Other indications for MRI were spinal pain ($n = 23$), headaches ($n = 1$), abnormal lower limb reflexes ($n = 5$) and an unusual curve pattern ($n = 5$).

The mean Cobb angle at the time of referral was 48° (range 10–104°) and 64 MRI studies were normal. Of the 9 that were abnormal, 4 showed the presence of a Chiari I malformation, and all 9 had a syrinx. MRI abnormalities are detailed in Table 2. Of the 42 routine preoperative referrals, 4 (9.5%) had abnormal MRI studies (1 Chiari I malformation and 4 syrinxes). Of the 8 patients referred because of abnormal SARs, 2 (25%) had abnormal MRI studies, both with Chiari 1 malformation and syrinx. Both were girls with JIS. In one case, there was unilateral absence of SARs while, in the other, there was complete absence of the SARs.

Table 1 Details of patients with abnormal superficial abdominal reflexes (SARs) and relationship to magnetic resonance imaging (MRI) findings

No.	Sex	Age at presentation (years)	SAR abnormality	Age at MRI (years)	MRI findings
1	Male	19	Asymmetric	20	Normal
2	Female	16	Absent	29	Normal
3	Female	4	Asymmetric	5	Normal
4	Female	9	Absent left side	16	Chiari 1, syrinx C2–T12
5	Female	14	Asymmetric	15	Normal
6	Female	8	Absent	10	Chiari 1, syrinx C4–C6 and long thoracic
7	Female	12	Asymmetric	12	Normal
8	Female	12	Absent	17	Normal

Table 2 Details of magnetic resonance imaging (MRI) abnormalities

No.	Age onset curve (years)/ sex	Referral group	Chiari I malformation	Syrinx	Syrinx level
1	15/Male	1	Yes	Yes	C5–7
2	12/Female	2 (back pain)	Yes	Yes	Cervical down to T7
3	2/Female LBP	2	No	Yes	C1–T11
4	9/Female	2 (L thoracic curve. Abnormal SAR)			
5	6/Male	1	No	Yes	C5–6, T3–5
6	10/Female	2	Yes	Yes	C3 to T9/10
7	8/Female	2 (shoulder pain absent SARs)	Yes	Yes	C4–6, long thoracic
8	15/Female	1	No	Yes	C6
9	13/Female	1	No	Yes	T7

Discussion

The need to identify abnormalities of the hindbrain and spinal cord, particularly syrinx, prior to scoliosis correction surgery has been highlighted [15]. MRI has been established as the technique of choice for assessing the spinal cord in patients with scoliosis [16], but it is a relatively costly examination and the routine use of MRI prior to surgery has been questioned. In the setting of asymptomatic children with typical AIS and normal neurological examination, the pick-up rate and clinical significance of Chiari 1 malformation and syrinx is small, suggesting that routine pre-operative MRI is not indicated in this group of patients. Shen et al. [17] identified two Chiari 1 malformations in 72 patients with AIS. In a group of 140 children with typical AIS, Winter et al. [19] identified only four abnormalities, one small thoracic syrinx and three Chiari 1 malformations. All patients underwent scoliosis surgery without complications. Maiocco et al. [14] reported 45 consecutive children with AIS, finding one Chiari 1 malformation and one syrinx. Most recently, Do et al. [5] reported on 327 consecutive patients with AIS who underwent MRI prior to surgical correction and found only two syrinxes and four Chiari 1 malformations. In the current study, 4 of 42 (9.5%) patients with asymptomatic typical curve patterns (group 1) had an MR abnormality; 3 of these were considered to be typical cases of AIS (the other being a boy with JIS). The incidence of MR abnormality in asymptomatic AIS is therefore 7%, which is slightly higher than previously reported.

The situation is different in the case of IIS and JIS onset scoliosis, and in the presence of symptoms and/or abnormal neurological examination. Dobbs et al. [6] identified MRI abnormalities in 21% of neurologically normal children with IIS, including five Chiari 1 malformations, eight syrinxes, one low lying conus and one brainstem tumour. Evans et al. [8] performed a prospective MRI study of 31 patients with juvenile onset 'idiopathic' scoliosis. Of these, 8 (26%) had significant abnormalities, including 7 Chiari 1 malformations, 6 syrinxes and 1 cervical cord astrocytoma. It is notable that only 2 of these patients had abnormal neurological examinations. Unilateral absence of the SARs was recorded in 3 patients, only 1 of whom had an abnormal MRI. Gupta et al. [12] reported on 25 consecutive children with atypical features including age <11 years at presentation, presence of pain, hyperkyphosis, severe curves and the presence of the left thoracic or thoracolumbar curves, finding abnormalities in 7 patients (28%).

The potential relationship between abnormality of the SARs and underlying syrinx was promoted by Zadeh et al. [21]. They reported 12 children presenting over a period of 8 years who had abnormal SARs. The mean age at diagnosis of scoliosis was 4.3 years (range 1 year 8 months–6 years 7 months). The majority had other abnormal examination findings and 5 of the curves were left sided. Of the children, 10 had syringomyelia and had either the complete absence of the SARs or unilateral absence on the side of the convexity of the curve. In 2 cases, the abnormal SAR was the only neurological finding. They concluded that an absent SAR on the side of the convexity of the curve was an early and sensitive indicator of underlying syringomyelia. However, the presence of several other risk factors makes it difficult to assess the relevance of the abnormal SAR. Also, it is not known how many children with normal SARs presenting to their Unit over the same time period had abnormal MRI findings. Therefore, the true sensitivity and specificity of an abnormal SAR cannot be determined from their study.

The prevalence of abnormal SARs in children and adolescents with 'idiopathic' scoliosis has not been reported, although in the study of JIS by Evans et al. [8], 3 of 31 (10%) children had an abnormal SAR. In contrast, the prevalence of 'abnormal' SARs in otherwise normal children, adolescents and young adults has

been studied. Yngve [20] studied the variation of SARs in 65 normal subjects, 30 in the 10- to 20-year age range and 35 in the 21- to 30-year age range. In the first group, the SARs were present and equal in 73% of cases, present but asymmetric in 10% and absent in some quadrants in 3%. Complete absence was seen in 13% whereas unilateral absence was never seen. In the second group, the SARs were present and equal in 49% of cases, present but asymmetric in 17% and absent in some quadrants in 17%. Complete absence was seen in 17% whereas unilateral absence was again never seen. This report therefore calls into question what constitutes abnormal SAR.

In our Spinal Deformity Unit, any asymmetry or absence of the SARs has been considered an indication for MRI to rule out underlying syrinx. In the present study, the prevalence of 'abnormal' SARs was 11%, consistent with the findings of Evans et al. [8]. In four patients, the SARs were present but asymmetric, and none of these cases had an abnormal MRI study. In one patient, there was unilateral absence of the SARs and this patient had an abnormal MRI study. She was a girl with JIS, a left thoracic curve and left-sided absence of the SARs. In three patients, the SARs were completely absent and one of these had an abnormal MRI. She was a girl with JIS, upper limb pain and abnormal neurological signs. Our study therefore supports the view that unilateral absence of SARs on the convexity of the curve is a significant, albeit uncommon, clinical finding in children with 'idiopathic' scoliosis and is consistent with the findings of Zadeh et al. [21]. Complete absence of the SARs may also be associated with syrinx. However, any other 'abnormality' of SARs was not associated with abnormal MRI findings and is consistent with the normal variation in abdominal reflex patterns as reported by Yngve [20].

Conclusions

We have identified a prevalence of abnormality of the SARs in 11% of patients with suspected idiopathic scoliosis undergoing whole spine MRI. Unilateral absence of SARs on the convexity of the curve is a significant clinical finding suggestive of underlying syrinx. Complete absence of SARs may also be associated with neuraxis anomalies. However, asymmetry of SARs is a normal variation, which in itself is not and indication for whole spine MRI.

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Syringomyelia

A Potential Risk Factor in Scoliosis Surgery

M. H. H. Noordeen, FRCS,† B. A. Taylor, FRCS,† and M. A. Edgar, FRCS*

Study Design. An 18-year-old patient with "idiopathic" adolescent scoliosis is presented. A thoracic syrinx was detected as an incidental finding during magnetic resonance imaging of the spine.

Objectives. Syringomyelia may be a risk factor for neurologic injury during correction of scoliosis, and in these cases, spinal cord monitoring may be of particular value.

Background Data. Spinal distraction and instrumentation carry a risk of neurologic damage in patients with scoliosis and associated syringomyelia. Syringomyelia is a cause of scoliosis, and although neurologic problems are the usual symptom, scoliosis may be the only sign at initial examination. A higher risk of neurologic injury has been reported in corrective surgical treatment of patients with syringomyelia. The mechanism of cord damage is unclear. Monitoring of spinal cord function is recommended to detect intraoperative neurological injury, which may be reversed on removing distraction and implants.

Results. Intraoperative somatosensory-evoked potential (SSEP) spinal cord monitoring detected possible cord damage during outrigger distraction. Reduction of distraction led to a recovery of SSEPs and a satisfactory operative outcome.

Conclusion. Syringomyelia may be a risk factor for neurologic injury during correction of scoliosis, and SSEP spinal cord monitoring may identify and prevent intraoperative spinal cord injury. [Key words: surgery, scoliosis, syringomyelia, complications] **Spine 1994;19: 1406–1409**

Spinal distraction and instrumentation carry a risk of neurologic damage in patients with scoliosis and associated syringomyelia, and three cases of permanent paraplegia have been reported after correction of scoliosis in these patients.[7,9,13] Syringoyelia has been recognized as a neurogenic cause of scoliosis, and the incidence of scoliosis in patients with syringomyelia is between 20% and 85%, with a higher incidence in those with skeletal immaturity.[6,7,18] Although neurologic problems are the usual symptom, scoliosis may be the only sign at initial examination.[2,6,19] Syringomyelia is often associated with other abnormalities, particularly the Chiari type I malformation. Magnetic resonance imaging (MRI) should be used to evaluate syringomyelia.[9]

Neurologic complications during surgery for spinal deformity vary between the populations treated and the procedure performed and are reported between 0.7% and 5%.[1,10] A higher risk of neurologic injury has been reported in corrective surgical treatment of patients with syringomyelia.[13,19] The mechanism of cord damage is unclear, although risk factors have been identified.[12] Monitoring of spinal cord function is recommended to detect intraoperative neurologic injury, which may be reversed by the reduction of distraction or the removal of implants.

An 18-year-old adolescent with "idiopathic" scoliosis is presented; a Chiari type I malformation and a thoracic syrinx were detected as an incidental finding during MRI spinal scanning trial. Intraoperative somatosensory-evoked potential (SSEP) spinal cord monitoring detected possible cord damage during outrigger distraction. Reduction of distraction led to the recovery of SSEPs and a satisfactory operative outcome. Syringomyelia may be a risk factor for intraoperative neurologic injury during correction of scoliosis, and in these cases, spinal cord monitoring may be of particular value.

Case Report

An 18-year-old male had scoliosis, which was detected 1 year previously, at initial examination. He had occasional discomfort over the right scapula for 2 years. He had no history of illness or injury. Birth, perinatal, and early development were within normal limits; there was no family history.

On examination, he was 5 feet and 5 inches tall, and he had right thoracic scoliosis, with a high right shoulder, and a prominent right scapula. There was a full range of spinal movements, and the deformity corrected with lateral flexion. A neurologic examination including examination of power, tone, coordination, light touch sensation, pin- prick sensation, joint position sensation, reflexes, and the plantar response, did not reveal any abnormality. Measurement of rotational deformity using the Bunnell method yielded 17°, in the thoracic spine.

Radiographic films (Figure 1) showed a right-sided thoracic curve from the T5–T11 levels, measuring 55° by the Cobb method. The iliac epiphyses had fused. Longitudinal traction reduced the curve to 35°. A technetium 99m bone scan did not showed any significant abnormality. MRI scans of the whole spine showed a small syrinx extending from the T6–T8 ver-

From the *The Middlesex Hospital, London, and the †The Royal National Orthopaedic Hospital Trust, Stanmore, Middlesex, United Kingdom.
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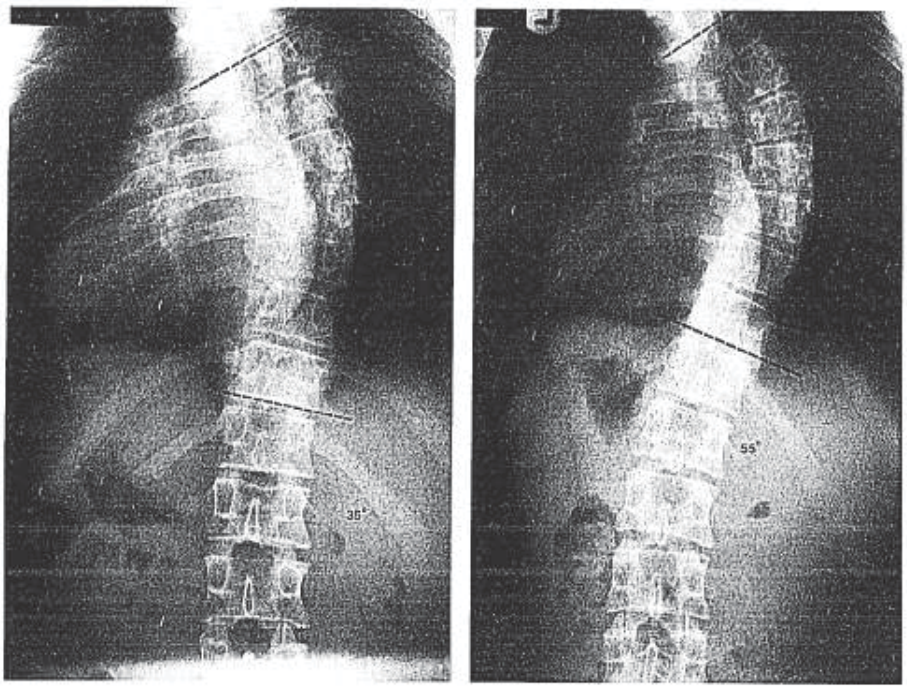

Figure 1. Preoperative anteroposterior and lateral radiographic films, demonstrating the deformity before and after horizontal traction.

tebral levels, crossing the apex of the scoliosis (Figure 2). A Chiari type I malformation was identified in the cervical spine. The patient had a Moe fusion performed with a contoured Harrington rod and sublaminar wiring, through a posterior midline incision. SSEP spinal cord monitoring was used throughout the operation.

Spinal Cord Monitoring. Posterior tibial nerve stimulation with supramaximal constant voltage single pulse stimuli of 0.2 ms, at 20 per second were applied alternately to left and right legs. The SSEP was recorded with a 4F bipolar electrode (Orthoexpress, Amersham, Bucks, England) from the epidural space at the T2 level. Signals were amplified, conditioned, and displayed on a Medelec MS91 (Medelec, Old Woking, Surrey, England) and PA89 preamplifier. Filters were set to BER (200–2000 Hz), and sweeps were averaged to obtain clear recordings. Recordings were of good technical quality throughout.

After exposure of the spine and insertion of upper and lower Harrington hooks, outrigger distraction was applied initially to 12.5 lb. After facet joint excision, distraction was increased to 17.5 lb, and simultaneous changes in pulse, blood pressure, and the SEP occurred on the right side, which dropped to 36% of the control amplitude (Figure 3). Distraction was discontinued completely, and pulse and blood pressure rapidly returned to previous levels. The SSEP from the right leg rapidly recovered to 150% of control amplitude. Redistraction to 12.5 lb did not alter the SSEP, which remained stable for 30 minutes until insertion and distraction of the Harrington rod, which again caused a rapid decrease to

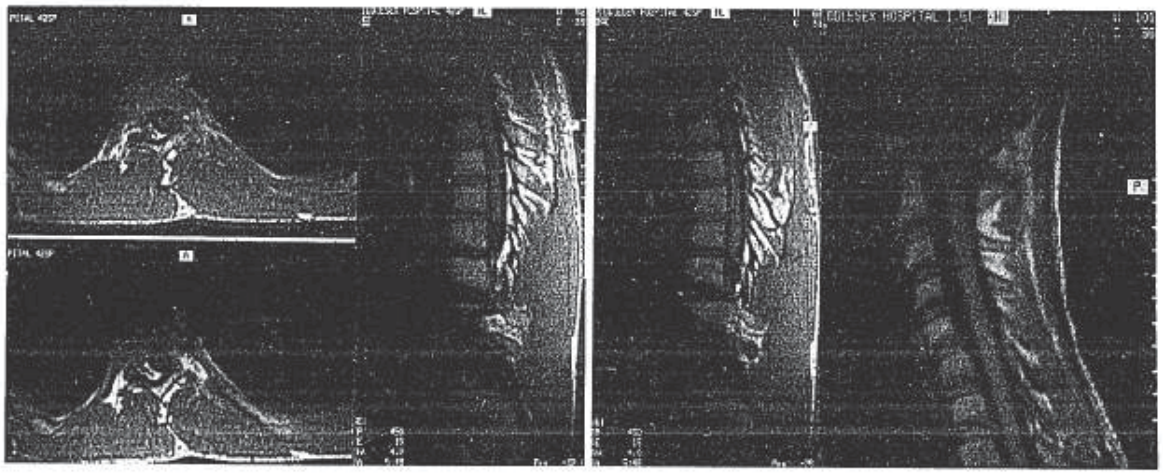

Figure 2. MRI imaging of thoracolumbar spine demonstrating a small syrinx at the T6–T8 vertebral levels, transverse section at the T7 vertebral level demonstrating the syrinx, and at the level of the foramen magnum, demonstrating the Chiari type I malformation.

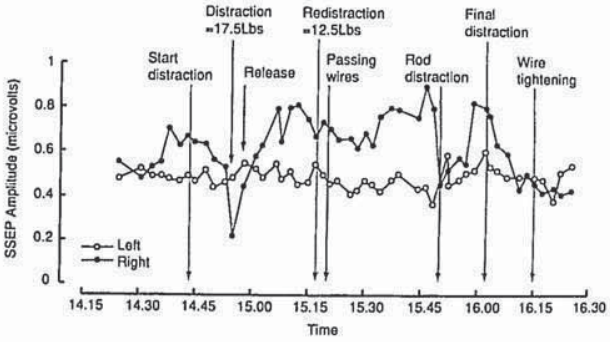

Figure 3. Chart showing SEP potentials obtained, the relationship between time, the SEP potentials on each side, and intraoperative events.

50% of peak amplitude. This recovered over 10 minutes but again decreased (to initial control amplitude) after sublaminar wire tightening.

Postoperatively, the patient had urinary retention and had to be catheterized. Neurologic examination including examination of power, tone, coordination, light touch sensation, pin-prick sensation, joint position sensation, reflexes including abdominal reflexes, and the plantar response, revealed loss of the right-sided abdominal reflex. At 48 hours, both the ability to void urine, and the abdominal reflexes, had returned. Neurologic examination did not reveal any abnormalities. Postoperative erect radiographic films (Figure 7, 8) showed correction to 30°, measured by the method of Cobb. The patient was mobilized with a brace and is included in follow-up without any evidence of problems.

Discussion

Unusual curve types, painful curves, and rapidly progressive curves despite skeletal maturity, have been recognized as heralding the presence of syringomyelia in scoliosis.[2,6] MRI studies on patients with idiopathic scoliosis have shown that syringomyelia may be more common than previously thought, and MRI imaging of the spinal cord should be mandatory before bracing or operative correction of scoliosis.[17] In patients with neurologic symptoms, scoliosis, and syringomyelia, drainage of the cyst is recommended because it stabilizes or improves neurologic function, and although it does not arrest progression of the curves, it temporarily does so in immature patients. It may make scoliosis surgery less dangerous.[15] The recommendations for patients with small cysts presenting with scoliosis only, are less clear, though scoliosis may precede neurologic symptoms by several years.[3,16] Patients with Chiari type I malformations who do not have syringomyelia are unlikely to develop scoliosis, and the effect of Chiari surgery on scoliosis is inconclusive.[17] If surgical correction is anticipated without syrinx decompression, constant intraoperative monitoring may be particularly indicated.

Intraoperative monitoring of spinal cord function includes wake-up techniques and recording of SSEP. A small number of patients, however, continue to have neurologic complications despite monitoring.[5] A study of SSEP spinal cord monitoring in 1168 consecutive patients using techniques identical to that used in the present case[4] showed no false-negative cases of intraoperative spinal cord damage.

When decreases in SSEP amplitudes were more than 50%, as observed in this instance, 38% had clinically detectable postoperative neurologic changes. Those patients with a decrease of amplitude greater than 80% had a high risk of significant neurologic deficit (57%). There was a 100% correlation between the side of the amplitude decrease and the side of the neurologic loss in the trunk or limb. Those patients in whom SSEP amplitudes did not return to levels above 50% had an increased risk of neurologic complication. Abdominal reflexes are cutaneous reflexes consisting of a brisk unilateral contraction of a part of the abdominal wall in response to a cutaneous stimulus, such as light scratch with a pin. It is a polysynaptic, multisegmental reflex, localized in the spinal cord from the 7th to 12th dorsal segments. Although lower motor neurone damage may cause dimunition or loss of the reflex, it is normally dependent on the integrity of the ipsilateral corticospinal tract,[8] thus indicating ipsilateral upper motor neuron injury in the case reported.

Spinal cord injury may occur intraoperatively after "moderate" distraction in "idiopathic" scoliosis. The case reported was an "idiopathic" scoliosis until MRI scanning revealed an unsuspected thoracic syrinx. MRI scanning has improved the detection of syringomyelia, and this case strengthens the case for routine MRI screening of all "Idiopathic" scolioses. Syringomyelia may be a risk factor for neurologic injury during correction of scoliosis; SSEP spinal cord monitoring may identify and prevent intraoperative spinal cord injury.

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Address reprint requests to

M. H. H. Noordeen, FRCS
Department of Trauma & Orthopaedic Surgery
The Middlesex Hospital
Mortimer Street
London W1N 8AA, United Kingdom

MRI OF 'IDIOPATHIC' JUVENILE SCOLIOSIS

A PROSPECTIVE STUDY

S. C. EVANS, M. A. EDGAR, M. A. HALL-CRAGGS, M. P. POWELL, B. A. TAYLOR, H. H. NOORDEEN

From The Middlesex Hospital, London, England

In a prospective trial we performed MRI of the spine and hind brain in 31 patients with scoliosis of onset between the ages of four and 12 years.

In eight patients (26%) there was a significant neuroanatomical abnormality; there were six cases of Chiari-1 malformation associated with a syrinx, one isolated Chiari-1 malformation and one astrocytoma of the cervical spine. Four of these patients had left-sided curves.

There were no clinical features which could reliably identify those patients with abnormalities on MRI. In particular, the unilateral absence of abdominal reflexes was found to be non-specific (1 of 8 of patients with neuroanatomical abnormalities (12.5%) *v* 2 of 23 with normal scans (8.7%)).

In view of the established risks of surgical correction of scoliosis in the presence of undecompressed syringomyelia and the possible improvement that may follow decompression of the foramen magnum, we feel that MRI of all patients with scoliosis of juvenile onset should be obligatory.

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S. C. Evans, FRCS, FRCS Orth, Senior Orthopaedic Registrar
M. A. Edgar, MChir, FRCS, Consultant Orthopaedic Surgeon
M. A. Hall-Craggs, MD, MRCP, FRCR, Consultant Radiologist
M. P. Powell, FRCS, Consultant Neurosurgeon
H. H. Noordeen, FRCS, FRCS Orth, Consultant Orthopaedic Surgeon
The Middlesex Hospital, Mortimer Street, London W1N 8AA, UK.

B. A. Taylor, MCh Orth, FRCS, FRCS Ed, Consultant Orthopaedic Surgeon
Royal National Orthopaedic Hospital NHS Trust, Brockley Hill, Stanmore, Middlesex HA7 4LP, UK.

Correspondence should be sent to Mr S. C. Evans at 70 Durlston Road, Kingston upon Thames, Surrey KT2 5RU, UK.

0301-620X/96/21140 $2.00

In 1954 James classified idiopathic scoliosis into three groups according to the age of onset: infantile, juvenile and adolescent. In his study the juvenile group included only 16 of 134 thoracic curves. Although the age criteria for classification vary between different studies, juvenile scoliosis remains relatively uncommon (Tolo and Gillespie 1978; Dickson 1985). Some juvenile cases result from undiagnosed infantile curves and, conversely, a proportion of adolescent scolioses will represent previously undetected juvenile cases. Doubt over the true significance of the juvenile group led Dickson (1985) to use the original classification of Ponseti and Friedman (1950) into early and late onset, with five years as the dividing point.

The association of scoliosis with syringomyelia is well established (Woods and Pimenta 1944; McRae and Standen 1966). In the past the identification of abnormal neurological signs prompted further, usually invasive, investigation. Baker and Dove (1983) were the first to report scoliosis as the presenting sign of otherwise asymptomatic syringomyelia and this has since been confirmed by others (Raininko 1986; Isu et al 1992; Lewonowski, King and Nelson 1992). With the advent of MRI, sporadic accounts of syringomyelia, often associated with the Chiari-1 malformation, have identified a preponderance within the juvenile period (Arai et al 1992).

Zadeh et al (1995) found ten cases of syringomyelia in 12 patients with idiopathic scoliosis who had been selected for MRI after the identification of asymmetrical or absent superficial abdominal reflexes. This review, and that of Schwend et al (1995), emphasised the need for a prospective study of MRI in idiopathic scoliosis and we have performed such a study on the juvenile age group.

PATIENTS AND METHODS

We performed a prospective study of 31 consecutive patients who presented with idiopathic scoliosis first detected between the ages of four and 12 years inclusive. All patients were assessed clinically with a full history and examination including neurological evaluation.

There were 24 girls and seven boys. The mean age at which scoliosis was first noted was 8.6 years (6.6 years for boys and 9.1 years for girls; range 4.0 to 12.5; Fig. 1).

All patients had MRI with a 1.5 Tesla Siemens Magnetom 63SP system (Siemens, Erlangen, Germany). Images of the whole spine were produced by a linearly-polarised

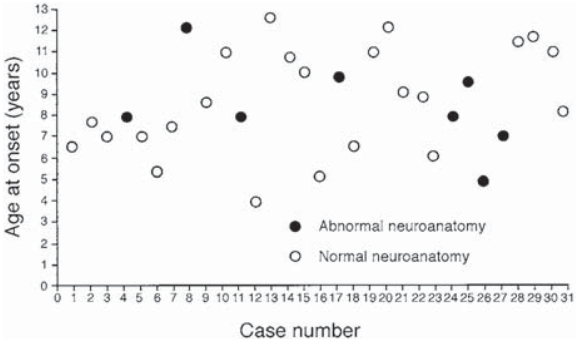

Fig. 1

Scattergram showing the age at onset of scoliosis in 31 patients.

cervical spine coil for the cervical spine and a four-element phased array spine coil for the thoracic and lumbar spine. A combination of sagittal, coronal and axial T1-weighted scans was made in all patients. Those in whom a syrinx was found without an associated Chiari malformation were also examined after intravenous injection of gadolinium-DPTA (Magnevist; Schering, Burgess Hill, UK) to exclude an underlying neoplasm of the cord. These scans were prospectively studied by experienced radiologists. The hind brain and spinal cord were examined for syrinx formation, features of spinal dysraphism and cord neoplasms.

RESULTS

There were 19 single and 12 double curves (Fig. 2). The thoracic curves were right-sided in 24 and left-sided in 7. The Cobb angles of the major curves at presentation ranged from 10° to 100° with a mean of 40°.

In eight patients a considerable neuroanatomical abnormality of the hind brain or spinal cord was identified; there were six cases of Chiari-1 malformation associated with a syrinx, one of an isolated Chiari-1 malformation and one astrocytoma with a syrinx. In four of these patients the thoracic curve was left-sided, whereas this was seen in only three of the 23 patients with normal neuroanatomy ($p = 0.05$).

There was no significant difference in the mean age of onset between the groups with and without neuroanatomical abnormalities (8.3 years and 8.7 years, respectively). The mean curve at presentation was 45° in the group with abnormal neuroanatomy and 40° in the normal group. The proportion of abnormal neuroanatomical findings was similar for both sexes (6 girls and 2 boys, 25% and 29%, respectively).

At presentation only two of the eight patients with neuroanatomical abnormalities had neurological signs. These were marked in one and included nystagmus and ataxia; in the other there was unilateral absence of the abdominal reflex. Both patients had a syrinx with a Chiari-1 malformation. Two patients with normal neuroanatomy had unilateral absence of the abdominal reflex.

Twenty patients were observed for at least six months before surgery. They had a mean progression of the curve of 7.6°/year (–10 to 44). For the five patients with neuroanatomical abnormalities who were observed for this period the mean progression was 9.8°/year and for the 15 normal cases 6.9°/year.

All six patients with a Chiari malformation and syrinx had decompression of the foramen magnum (Fig. 3). Three with curves greater than 50° at the time of neurosurgery subsequently had two-stage spinal fusion. One of the three remaining patients showed an improvement from 33° to 19° over nine months after decompression. Follow-up of the last two patients, with curves of 28° and 44° respectively, has been too short to make a useful observation.

The patient with an astrocytoma and syrinx had a biopsy and radiotherapy. The patient with an isolated Chiari-1 malformation had a two-stage fusion 47 months after presentation.

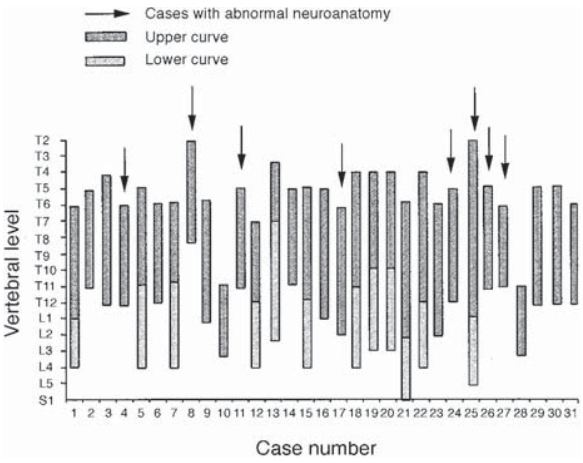

Fig. 2

Chart showing the site of scoliosis in 31 patients.

Of the 23 patients with normal neuroanatomy six were managed by simple observation, four had bracing and 13 had fusion (1 anterior, 1 posterior and the remainder two-stage).

DISCUSSION

We have used MRI prospectively to screen a consecutive group of 31 children with idiopathic scoliosis of juvenile onset. We found abnormalities of the hind brain or cord in eight (26%). Since the clinical presentation did not discriminate between those with and without CNS abnormalities, the underlying abnormalities would have remained undiagnosed without the routine use of MRI.

The unilateral absence of abdominal reflexes did not

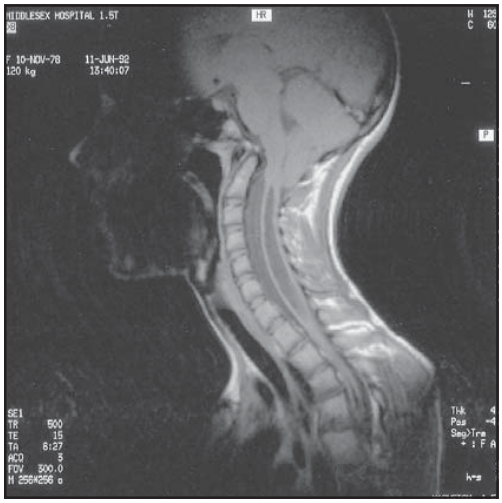

Fig. 3a

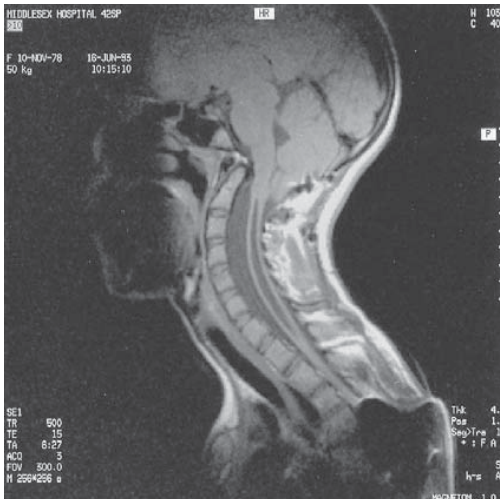

Fig. 3b

MRI showing a Chiari malformation associated with a large syrinx in the cervical and thoracic cord before decompression of the foramen magnum (a) and eight months after decompression (b) with reduction in the size of the syrinx.

prove to be a specific finding, being identified in one of eight patients with neuroanatomical abnormalities as against two of 23 with normal neuroanatomy. This appears to conflict with Zadeh et al (1995) who concluded that "an absent superficial abdominal reflex on the same side as the convexity of the curve is an early and sensitive indicator of underlying syringomyelia". The patients whom they report, however, were selected for scanning on the basis of abnormal abdominal reflexes; no comment can be made on the incidence of syringomyelia in patients who had not been scanned. The age distribution of their patients was also younger and we doubt the significance of this reflex when it is absent bilaterally rather than unilaterally.

We found that 50% of the neuroanatomical abnormalities were associated with left-sided curves. In other reviews of syringomyelia Isu et al (1992) and Muhonen et al (1992) had similar findings of approximately 50%, but Schwend et al (1995) found a greater association with left-sided curves (77%). If MRI is done solely on the basis of the direction of the scoliosis then half of all such abnormalities will be missed.

Although only one of our four patients managed by decompression of the foramen magnum has so far shown a reduction of the curve, several reports have indicated either arrest or improvement of the scoliosis after this procedure (Dure et al 1989; Phillips, Hensinger and Kling 1990; Muhonen et al 1992). Early detection of a syrinx by prompt MRI may therefore be of particular value.

The risk of neurological injury during instrumented correction of scoliosis without prior decompression of an associated syrinx has already been documented (Huebert and MacKinnon 1969; Diaz and Lockhart 1987; Noordeen, Taylor and Edgar 1994). The cause is uncertain, but sensitivity to direct trauma, a tenuous blood supply in the presence of an expanded cord and changes in the pressure of the CSF have all been suggested (Phillips et al 1990).

The identification of a spinal tumour lends weight to the value of MRI. There have been several reports of scoliosis as the presenting feature of an intramedullary cord tumour without neurological signs or symptoms (Allen and Kahn 1933; Dalloz et al 1963; Citron et al 1984). Early detection and treatment of our patient allowed the preservation of complete neurological function.

In a prospective study of routine MRI of 'idiopathic' scoliosis in 26 patients of all ages Samuelsson, Lindell and Kogler (1991) identified two with syrinxes and concluded that "MR should be mandatory before bracing or operative correction of scoliosis". In our study we have failed to identify any clinical features which may reliably distinguish affected cases. For all new patients with scoliosis the cost of routine MRI may be prohibitive in many units, but since we have found significant neuroanatomical abnormalities in 26% of curves of juvenile onset, we feel that in this age group MRI should be obligatory.

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Chapter 8

SPONDYLOLYSIS, TUMOURS & TRAUMA

Advances in Surgical Techniques for EOSD

Paper: 24

Isthmic spondylolysis most commonly at L5 is a common cause of back pain in children. Sporting activities involving repetitive hyperextension (viz. gymnastics, fast bowlers in cricket, and so forth) are prone to have spondylolysis. It is characterized by a defect in pars interarticularis. Surgical intervention is offered for those who have refractory pain, despite activity modification and non-operative treatment. Pars repair, or spinal fusion, is one of the most commonly performed surgeries.

I have described an innovative technique of treating symptomatic back pain that has failed all conservative modalities of management in such young children, by using a modular cross-link and pedicle screws without spinal fusion. A series of twenty such adolescents and teenagers between nine and twenty one years of age were treated by this novel technique and prospectively followed-up for a mean of four years (range: 2.3 to 7.3 years) was published. The validated outcome measures used were Oswestry disability index (ODI) and visual analog scale (VAS) score. The fusion rate at the pars defect area assessed by post-operative CT scans was eighty percent. All patients had a statistically significant improvement in back pain with post-op mean ODI of only eight percent ($p<0.001$).

The use of a modular 'U' cross-link without spinal fusion has other advantages as the incidence of adjacent segment degeneration (ASD) is eliminated. Anatomical repair and union of the pars defect restores normal biomechanics at the lumbo-sacral junction which is desired for the long-term success.

Repair of spondylolysis using compression with a modular link and screws: Altaf F, Osei NA, Garrido E, Al-Mukhtar M, Natali C, Sivaraman A, Noordeen HH. J Bone Joint Surg Br. 2011 Jan; 93(1): 73-7. PMID: 21196547

Advances in Surgical Techniques for EOSD

Paper: 25

Osteoid osteomas are characterized by a nidus surrounded by dense sclerotic bone, and are a common cause of back pain in young individuals. It most commonly affects the posterior elements (i.e. lamina, pedicles, and the spinous processes), and is invariably associated with a painful scoliotic list. The secretion of proteoglycans and chemical mediators is responsible for pain. Prompt relief of this pain, with an intake of aspirin and other NSAIDs, is pathognomonic of this condition.

A ten year old girl who was referred by her GP for persistent back pain and painful scoliosis was seen in my out-patient clinic. A CT scan confirmed the diagnosis of an osteoid osteoma located in the postero-lateral aspect of the left T6 vertebral body. The NSAIDs therapy was so effective that an interval CT scan, performed at sixteen months from the date of her first clinical evaluation, revealed a complete resolution of the scoliosis with a less conspicuous radiolucent nidus. The patient had a complete resolution of all her symptoms within two years, and had returned to full sporting activity.

This goes to emphasise that a surgical excision of a nidus, though an attractive option, may not necessarily be needed in selective cases of osteoid osteoma causing back pain and scoliosis.

Symptomatic resolution of spinal osteoid osteoma with conservative management: imaging correlation. Jayakumar P, Harish S, Nnadi C, Noordeen H, Saifuddin A. Skeletal Radiol. 2007 Jun; 36 Suppl 1: S72-6. E-pub 2006 Sep 12. PMID: 16967288

Advances in Surgical Techniques for EOSD

Paper: 26

Langerhans cell histiocytosis (LCH) is a spectrum of disorders characterized by a pleomorphic infiltration of tissues by Langerhan's giant cells. The severity may vary from a solitary bone to diffuse systemic skeletal involvement. The most common type is eosinophilic granuloma (EG). LCH also affects the skull, causing a '*geographical skull*' appearance, and the spinal cord. Vertebral body involvement, in the form of '*vertebra plana*', is the most common feature. Skin lesions or multiple vertebral involvement is also not uncommon, and active lesions may be associated with back pain, warranting surgical interventional when all conservative modalities have failed. The most common region of the spine affected is the thoracic, and incidence of vertebra plana is only 15 percent.

I have published a series of two patients with LCH, and have studied the MRI features and characteristics differentiating the active and inactive lesions, correlating them with histological features at the time of surgery. The presence of a hyperintense lesion on a T_2 weighted MRI was suggestive of an active EG, a key observation that was hitherto unknown to spinal surgeons until this publication.

Patient 1: A seven year old female with an L5 lesion that was positive on S100 and CD1a special stains on biopsy.

Patient 2: A ten year old male with T6-T9 collapse and angular kyphosis showing an increased uptake at T9 (i.e. hyperintense T2 images).

Langerhans' cell histiocytosis of the spine: use of MRI in guiding biopsy: Kaplan GR, Saifuddin A, Pringle JA, Noordeen MH, Mehta MH. Skeletal Radiol. 1998 Dec; 27(12): 673-6. PMID: 9921928

Advances in Surgical Techniques for EOSD

Paper: 27

Very little is known about traumatic lumbo-sacral dislocation, and if it is unrecognized, a L5 lytic or isthmic spondylolysis can rapidly progress to become a Gr IV spondylolisthesis or spondyloptosis. I have published one such case in a sixteen year old girl, who was a victim of a vehicular accident and was hit by a truck from behind.

Despite being a rear seated, chest and shoulder strap restrained, passenger, she sustained significant abdominal injuries, in addition to spinal trauma, requiring an urgent laparotomy. Over a few months, despite being in a lumbar corset with activity modification, she had persistent back pain and left leg sciatica or radicular pain. An interval CT scan at one month post laparotomy revealed a fracture of the S1 articular process with a degenerative L5-S1 disc. She successfully underwent a combined anterior + posterior instrumented spinal fusion with anatomical restoration of her lumbo-sacral junction and biomechanics.

A series of two cases (another patient being an adult) was published in Spine in 2004. The most common mechanism hypothesised was hyperflexion with compression, which can also result in bilateral L5-S1 facet fractures. Other radiological and clinical features which raise a strong suspicion of L5-S1 dislocation are:

i) Perineal lacerations

ii) Transverse process fractures

Traumatic lumbosacral dislocation: report of two cases. Tsirikos AI, Saifuddin A, Noordeen MH, Tucker SK. Spine (Philadelphia, PA 1976). 2004 Apr 15; 29(8): E164-8. PMID: 15083005

Advances in Surgical Techniques for EOSD

Paper: 28

Multi-modal neuromonitoring is now a standard of care for all spinal deformity corrections surgery of the cervical and lumbar spine. I have published a comprehensive review article explaining the role of somatosensory evoked potentials (SSEPs) in the setting of spinal trauma, and reported results from a personal series of eighty two patients operated on over five years (1995 to 2000) for trauma where SSEPs were used. The duration of the follow-up was 2 to 7.5yrs. They included cervical (20), thoracic (8), thoraco-lumbar (8), and lumbar fractures (48). Forty of them had a pre-operative incomplete neurodeficit (group I), and 42 patients had no pre-op neurodeficit (group II). Eleven patients underwent combined anterior + posterior spinal fusion, whereas forty seven patients had posterior only, and twenty four patients had stand-alone anterior spinal fusions.

A fifty percent fall in amplitude in comparison to baseline recordings was associated with sixty seven percent sensitivity, and seventy one percent specificity in predicting neurological deficit. Patients, who had greater than twenty percent improvement in amplitude at end of surgery in group I, were associated with neurological improvement to functional status. Anterior surgeries were most commonly associated with a fall in amplitude by more than fifty percent (forty four percent of cases), compared to posterior surgeries (seventeen percent of cases), which was also statistically significant ($p<0.001$). This article continues to be one of my key contributions to the spinal surgical community, emphasising safety issues, and discussing the potential application of neuromonitoring data in predicting prognosis in patients with spinal trauma.

Spinal cord monitoring using intraoperative somatosensory evoked potentials for spinal trauma. Tsirikos AI, Aderinto J, Tucker SK, Noordeen HH. J Spinal Disord Tech. 2004 Oct; 17(5): 385-94. PMID: 15385878.

Repair of spondylolysis using compression with a modular link and screws

F. Altaf,
N. A. Osei,
E. Garrido,
M. Al-Mukhtar,
C. Natali,
A. Sivaraman,
H. H. Noordeen

From the Royal National Orthopaedic Hospital, Stanmore, United Kingdom

We describe the results of a prospective case series of patients with spondylolysis, evaluating a technique of direct stabilisation of the pars interarticularis with a construct that consists of a pair of pedicle screws connected by a U-shaped modular link passing beneath the spinous process. Tightening the link to the screws compresses bone graft in the defect in the pars, providing rigid intrasegmental fixation. We have carried out this procedure on 20 patients aged between nine and 21 years with a defect of the pars at L5, confirmed on CT. The mean age of the patients was 13.9 years (9 to 21). They had a grade I or less spondylolisthesis and no evidence of intervertebral degeneration on MRI. The mean follow-up was four years (2.3 to 7.3). The patients were assessed by the Oswestry Disability Index (ODI) and a visual analogue scale (VAS). At the latest follow-up, 18 patients had an excellent clinical outcome, with a significant ($p < 0.001$) improvement in their ODI and VAS scores. The mean ODI score at final follow-up was 8%. Assessment of the defect by CT showed a rate of union of 80%. There were no complications involving the internal fixation.

The strength of the construct removes the need for post-operative immobilisation.

■ F. Altaf, BSc(Hons), MRCS, Specialist Registrar in Trauma and Orthopaedics
■ N. A. Osei, FRCS(Tr & Orth), Spinal Fellow
■ E. Garrido, MRCS, EBOT, Spinal Fellow
■ M. Al-Mukhtar, MRCS, Registrar in Orthopaedics
■ H. H. Noordeen, MD, FRCS(Orth), Consultant Spinal Surgeon
Royal National Orthopaedic Hospital, Brockley Hill, Stanmore, Middlesex HA7 4LP, UK.

■ C. Natali, BSc, FRCS(Orth), Consultant Spinal Surgeon
■ A. Sivaraman, FRCS(Orth), Consultant Spinal Surgeon
Royal London Hospital, Whitechapel Road, Whitechapel, London E1 1BB, UK.

Correspondence should be sent to Mr F. Altaf; e-mail: farhaanaltaf@hotmail.com

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A spondylolysis is a defect of the pars interarticularis. Usually there is a complete break in the pars on one or both sides of the neural arch. The condition is often asymptomatic, but may be the cause of pain in the back. Free nerve endings with nociceptive function have been identified in the area of the lysis by Schneiderman et al.[1] Neuropeptides which mediate in the sensation of pain have been described at the site of the defect by Eisenstein et al.[2]

The surgical management of symptomatic spondylolysis or isthmic spondylolisthesis was first described by Albee,[3] Hibbs,[4] and Bosworth et al[5] using posterior fusion to stabilise the defect by eliminating the movement segment. Cleveland, Bosworth and Thompson[6] and Watkins[7] described the technique of posterolateral fusion.

Kimura[8] was the first to describe the direct repair of the pars defect using an isolated bone graft without instrumentation. Patients who underwent this procedure had to remain recumbent for a number of weeks, followed by external immobilisation in a cast for up to six months. Buck[9] first described the technique of direct repair by filling the gap in the defect with iliac cancellous autograft and placing screws directly through the defect itself.

A number of techniques have since been described for direct repair of the defect, but have been associated with complications such as loosening and breakage of the internal fixation, technical difficulty, extensive muscle and tissue dissection, and variable rates of consolidation of the defect.

We describe a prospective case series evaluating a technique of direct repair of the pars stabilised with a construct consisting of a pair of pedicle screws connected by a U-shaped modular link passing beneath the spinous process. Tightening the link to the screws compresses the bone graft in the defect, providing rigid intrasegmental fixation. This technique was first described by the senior author (HHN).

Patients and Methods

Patients were selected who had chronic disabling pain located in the lower back without sciatica, which had been resistant to conservative treatment for at least 12 months, including modification of activity and a trial of bracing and physiotherapy. All had defects in the pars at L5 on CT scans. Other inclusion criteria were the absence of adjacent degenerative disc disease confirmed by normal T2-weighted MRI, and no more than a grade I spondylolisthesis.[10]

Of the 20 patients in this series, 12 were male and eight were female, and nine had grade I spondylolisthesis. There was one

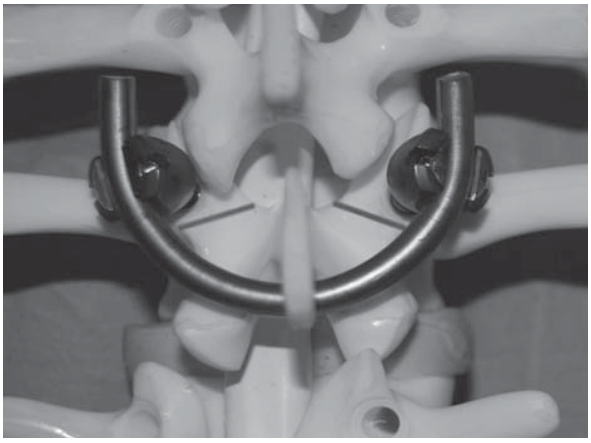

Fig. 1

Photograph showing the construct in a model spine. The shaded lines illustrate the location of the pars defects.

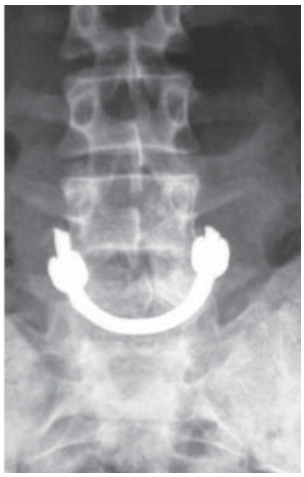

Fig. 3

Anteroposterior radiograph showing the construct at L5.

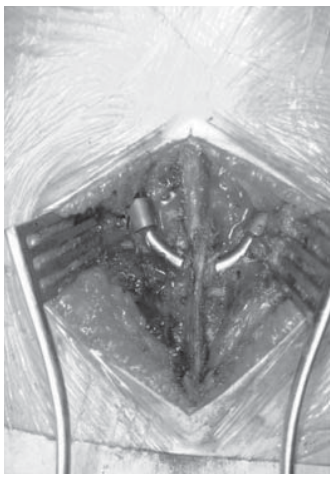

Fig. 2

Photograph showing the construct *in situ* from above. The rod is firmly fixed against the spinous process and the laminae, which promotes compression of the graft in the defect and stabilises the posterior arch.

smoker, who was male. The mean age of the patients was 13.9 years (9 to 21). None had a motor or sensory deficit.

Bony union of the pars was assessed by plain lateral radiographs and CT scans in all patients. The clinical outcome was evaluated using the Oswestry Disability Index (ODI),11 and a visual analogue scale (VAS)[12] with 0 representing no pain and 10 maximum pain), both pre-operatively and at follow-up. Complications were also recorded.

Statistical analysis used the paired *t*-test and the Wilcoxon's signed-ranks test. Significance was set at $p < 0.05$.

Surgical technique. The patient is positioned prone. A midline incision is made and the paraspinal musculature elevated laterally to expose the lamina, the pars and the base of the transverse process. Care is taken not to injure the capsule of the facet joint. The defect in the pars is exposed and the fibrocartilaginous element curetted. A burr is used to decorticate the defect and the corresponding lamina and transverse process. Anatomical landmarks and fluoroscopy are then used to determine the starting point for the pedicle screw. A starting hole is burred and a pedicle finder used to enter the pedicle. The walls and floor are assessed with a ball-tipped probe, and the hole is tapped and prepared for a 5 mm pedicle screw. Bone graft is harvested from the iliac crest, placed in the defect and impacted before insertion of the screw. After placement of the screw, a rod is contoured in a U-shape, placed just caudal to the spinous process, deep to the interspinous ligament of the affected level, and attached to each pedicle screw (Figs 1 and 2). When tightened to the screws, the rod compresses the defects. It is firmly fixed against the spinous process and the laminae, which promotes compression of the graft in the defect and stabilises the posterior arch.

Finally, fluoroscopic imaging (Fig. 3) confirms correct placement of the screw and link. Post-operative instructions include the administration of intravenous antibiotics until the wound is dry. The patient is mobilised without a brace.

Results

The mean follow-up was four years (2.3 to 7.3). None of the patients was lost to follow-up. The mean in-patient stay was three days (2 to 5). Post-operative complications

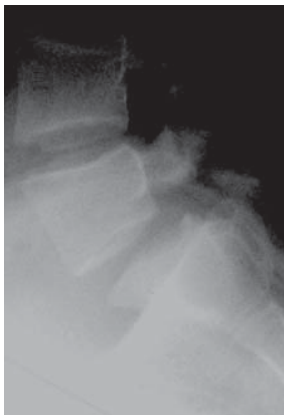

Fig. 4a

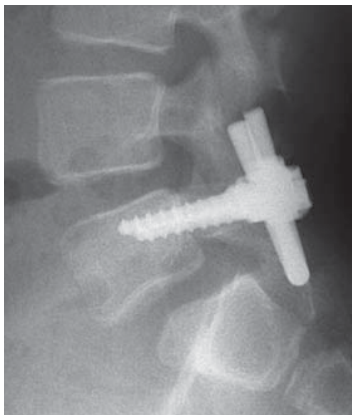

Fig. 4b

Lateral radiographs showing a) the pars defect at L5 pre-operatively and b) union of the pars defect at six months post-operatively.

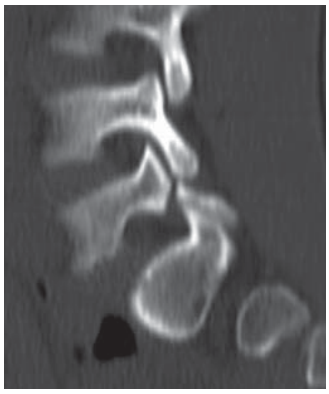

Fig. 5a

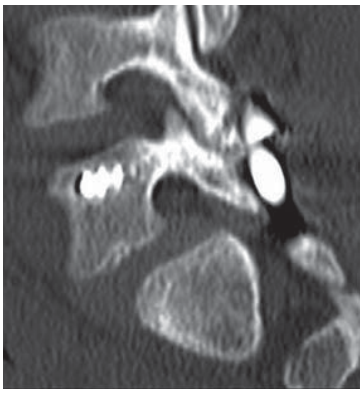

Fig. 5b

Lateral CT scans showing a) the pars defect at L5 pre-operatively and b) union of the pars defect at six months post-operatively.

included one superficial wound haematoma which did not require evacuation, and two superficial wound infections which responded to antibiotic treatment.

Bony union of the defect (Figs 4 and 5) occurred in 16 of the 20 patients. Three of the four patients with nonunion had a grade I spondylolisthesis. At the latest follow-up, 18 of the 20 patients had a significant ($p < 0.001$) improvement in their ODI (Table I) and in their VAS scores compared with the pre-operative levels. The mean ODI improved from 54% (42 to 78) pre-operatively, denoting severe disability, to 8% (0 to 42), indicating minimal disability. The mean VAS improved from 8.1 (6.5 to 10) pre-operatively to 1.6 (0 to 8.2) after operation. The mean ODI and VAS of the 16 patients with union of the defect was 3% and 0.4 respectively, at final follow-up.

The two patients who did not have a significant improvement in outcome had no evidence of union of the pars defect on the CT scan. The other two patients with nonunion were asymptomatic. Of the two patients who developed a symptomatic nonunion, one was classified as having severe disability at follow-up. He was a smoker, and was offered a further grafting procedure. The second patient was classified as having moderate disability, was a non-smoker, and had grade I spondylolisthesis. This patient had some subjective improvement and wanted no further intervention.

No breakage or loosening of the internal fixation was encountered, and none was removed.

Discussion

The aetiology of spondylolysis remains unknown, but current opinion is that there is an element of weakness which is genetically determined, or a degree of dysplasia at the pars interarticularis which renders it susceptible to the stresses of normal activity, resulting in a stress fracture.

Table I. Post-operative clinical assessment using the Oswestry Disability Index11

	Score (%)	Number of patients
Excellent (minimal disability)	0 to 20	18
Good (moderate disability)	21 to 40	1
Fair (severe disability)	41 to 60	1
Poor (complete disability)	61 to 80	0
Very poor (bedbound)	81 to 100	0

A minority of those affected need treatment and only a few require surgery. Most patients considered for surgical treatment have had the time for adjacent disc disease to develop, or have a spondylolisthesis. Lumbosacral fusion is the most common operation performed in these cases. For a grade I or less spondylolisthesis, a direct repair of the pars defect may be considered. This has the advantage of preserving adjacent movement segments and dealing directly with the anatomical defect responsible for any listhesis.

Buck[9] initially carried out his technique of direct repair in 16 patients. None developed nonunion, which was assessed radiologically, but three patients had a poor outcome. He suggested that this technique should only be used in cases where the gap in the neural arch was < 3 mm to 4 mm. In 1979[13] he described a larger series of 75 patients, 88% of whom had a satisfactory result. The accurate placement of the screw in this technique was found to be difficult, with a lengthy learning curve, and complications of this procedure most commonly arose secondary to screw loosening or misplacement.[14]

The Scott technique[15] involved stabilisation of the loose posterior arch by cerclage wiring of the transverse process to the spinous process of the involved vertebra with fusion of the defect. This technique requires greater surgical exposure, with extensive stripping of the muscle in order to expose the transverse process completely. Placement of the wires under the transverse processes is difficult and can lead to substantial bleeding. There is also a risk of injuries to the nerve root. Several cases of wire breakage have been reported.[16] Salib and Pettine[17] described a modification of this technique that consisted of a tension band wire around the posterior spinous process, with a screw in the pedicle. The proposed advantages were that there was no need to pass the wire around the transverse process, with its associated risk of injury to the nerve root, and the better anchorage of the wire using the pedicle screw. The strength and linkage of wires and the diversion of the compression force away from the graft mass are disadvantages. Songer and Rovin[18] described a small series in which the cerclage wire was replaced by a cable passing underneath the lamina, fixed on a pair of pedicle screws, in a technique that requires specific instrumentation.

In 1984, Morscher, Gerber and Fasel[19] described a new repair technique that used a laminar hook positioned in the defect and a compressive force placed upon it with a spring held against a screw within the articular process. The proposed advantages of this technique were that the fixation did not depend on the shape of the defect; it allowed for maximal grafting of the defect; and it would exert a compressive force across the lamina. Numerous problems have been encountered with this technique, including difficulty in screw placement, screw loosening[20] and breakage,[21] and a high rate of failure. Tokuhashi and Matsuzaki[22] described a modified technique that stabilised bone grafted to the pars defect with a pedicular screw, a hook and a rod used in combination. They described significant relief of symptoms in six patients over a limited period of follow-up.

Kakiuchi[23] used a variable-angle pedicle screw, a rod and a laminar hook with bone graft placed across the defect. The proposed advantage was fixation which was more rigid. He reported resolution of symptoms in 13 of 16 patients and bony union in all at a mean follow-up of 25 months.

Gillet and Petit[24] described direct repair by placing screws on the pedicles of the involved vertebra and fixing the loose posterior arch with a solid rod bent into a V-shape, taking purchase on the spinous process and laminae. They achieved excellent results in six of ten patients, with no complications.

In our series of 20 patients we achieved an excellent clinical outcome in 18, with a mean follow-up of four years. Union of the pars defect was achieved in 16 patients (80%). Contradictory opinions have been reported as to the effect of nonunion on outcome. According to Nicol and Scott,[25] Debnath et al,[26] Wu, Lee and Chen[27] and Dai et al,[28] patients with a pseudarthrosis have a poor or fair outcome. However, Hefti, Seelig and Morscher,[29] Johnson and Thompson[30] and Pellisé et al[31] could not prove this relationship. In our series, of the four patients with a nonunion,32 two had a poor and two an excellent outcome.

The treatment of defects of the pars that appear cold on a bone scan is associated with a higher rate of nonunion.[32] Although the patients in our study did not have a preoperative bone scan, this may be a useful investigation to identify the rate of nonunion in those patients who have a cold lesion. Biomechanical studies may also be useful to further evaluate the distribution of compressive forces across the pars defect in our construct. Most studies[9,16,18,19,23] on spondylolysis have used plain radiographs for the assessment of union, with its associated limited sensitivity.[33] In our study we used CT imaging, which is more sensitive and specific when used to assess union.[33] This may give an impression of a higher rate of pseudarthrosis in our study as compared with other studies which have used radiographs to assess union.

Some authors[30,34,35] consider that a slight degree of spondylolisthesis does not affect the outcome of patients undergoing direct repair of the pars. Others[36,37] carry out direct repair of the pars in patients with a spondylolisthesis of up to 25%. Jeanneret[38] found no difference in outcome

between patients with a spondylolisthesis of between 0% and 25%. In our series, nine of the 20 patients had grade I spondylolisthesis, three of whom developed a nonunion of the defect.

Our patients were aged between nine and 21 years, with no evidence of adjacent level disc disease on MRI, and spondylolisthesis not greater than grade I. Numerous authors have found that patients in the age range 20 to 30 years have worse results than younger patients.[38-40] The primary reason for this failure is thought to be associated with the appearance of degenerative disc disease.

Our technique uses readily available instrumentation to provide a strong construct. The bone graft in the pars defect is not hindered by screws, allowing for high rates of union. Post-operative recovery is made easier as the strength of the construct removes the need for post-operative immobilisation.

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CASE REPORT

Prakash Jayakumar
S. Harish
Colin Nnadi
Hilali Noordeen
Asif Saifuddin

Symptomatic resolution of spinal osteoid osteoma with conservative management: imaging correlation

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P. Jayakumar · C. Nnadi · H. Noordeen
Spinal Surgery, The Royal National Orthopaedic Hospital,
London, UK

S. Harish
Radiology, The Royal National Orthopaedic Hospital,
London, UK

A. Saifuddin
Department of Imaging, The Royal National Orthopaedic Hospital,
London, UK

P. Jayakumar (✉)
The Whitehouse,
51 Bycullah Road, Enfield,
Middlesex, EN2 8PH, UK
e-mail: pjay007@doctors.org.uk
Tel.: +44-7866-424589
Fax: +44-208-3670303

Abstract A 10-year-old girl presented with a history of painful scoliosis. Imaging performed, including computed tomography (CT) and magnetic resonance imaging (MRI), demonstrated a lesion with radiological features consistent with an osteoid osteoma (OO) of the 6th thoracic vertebra. The patient was treated conservatively with non-steroidal anti-inflammatory drugs (NSAIDs). Over eight months of clinical and radiological surveillance, she became entirely asymptomatic and demonstrated complete resolution of the scoliotic curve. The CT and MRI features of the osteoid osteoma during the period of surveillance are presented and are correlated with the corresponding clinical features.

Keywords Osteoid osteoma · Spine · MRI · CT · Conservative management · Spontaneous resolution

Introduction

Osteoid osteomas (OOs) of the spine present as the most common cause of painful scoliosis in children and young adults [1]. Surgical excision has been considered to be the treatment of choice, particularly given the potential risks of structural spinal deformity [2]. Minimally invasive treatments, such as radiofrequency (RF) and interstitial laser ablation, have also been applied and continue to advance [3]. Conservative treatment with non-steroidal anti-inflammatory drugs (NSAIDs) has been reported to be extremely effective in the management of OOs, particularly in the spine [4]. However, aspects such as the duration of treatment and the frequency of surveillance remain unclear.

We report a case of OO of the 6th thoracic vertebra (T6) managed conservatively and describe the correlative changes on computed tomography (CT) and magnetic resonance imaging (MRI). A case of spontaneous remission of OO in the axis with CT follow-up has been reported in the literature [5]. To the best of our knowledge, this is the first report that discusses the follow-up MRI features in a case of conservatively treated spinal OO.

Case report

A 10-year-old girl presented to her local General Practitioner with a history of mild back pain of two months duration. Clinically, the pain appeared to be poorly localised to the thoracic spine region. Initial spine radiographs performed did not show any definite abnormality. However, her back pain persisted and a repeat radiograph

performed six months after the initial consultation revealed a right thoracic scoliosis. This prompted a referral to the Spinal Surgery Unit. The patient had severe pain localised to the mid-thoracic spine, including night pain, but was otherwise systemically well. She did not take any medications. Clinically, no motor weakness, sensory deficit or bladder or bowel problems were present. An MRI study of the thoracic spine was performed eight months from the initial presentation. Sagittal Spin Echo T1-weighted (SE T1W) and Fast Spin Echo T2-weighted (FSE T2W) MR images of the spine demonstrated bone marrow oedema in the posterior two-thirds of the T6 vertebral body towards the left side (Fig. 1a,b). CT of the thoracic spine subsequently demonstrated a nidus in the cortex of the posterolateral vertebral body of the T6 vertebra on the left side with intense perinidal sclerosis (Fig. 2a,b). A diagnosis of spinal OO was made on the basis of the above findings. She was subsequently placed on NSAIDs, as both her and her family declined operative treatment. Over the course of our surveillance, her pain began to improve early on in her treatment course. Moreover, improvements were observed in the degree of curvature, to the extent that both the pain and scoliotic curves had completely resolved at the last follow-up consultation, 16 months after the initial presentation. CT scanning localised to the T5–T7 levels performed 14 months after the initial presentation demonstrated a radiolucent nidus and persistent perinidal sclerosis (Fig. 3). CT scanning localised to the T5–T7 levels performed 16 months after the initial presentation at the final follow-up visit demonstrated only mild perinidal sclerosis and a less conspicuous radiolucent nidus (Fig. 4a,b). Sagittal SE T1W and STIR MR images performed at the final consultation, 16 months after the initial presentation, showed complete disappearance of the bone marrow oedema, but there was persisting low signal intensity in the T6 vertebral body on both sequences, in keeping with residual sclerosis (Fig. 5a,b). At this stage, the patient was completely asymptomatic and had returned to full activity.

Discussion

OO has been reported in patients ranging from 2 to 56 years of age [6–9]. The lower incidence in the elderly population may suggest a spontaneous natural regression over time. However, long persistent cases without regression have also been reported [8].

There are two contemporary case series and one isolated case report in the literature describing actively observed spontaneous resolution of OO [4, 5, 10]. In the series by Ilyas and Younge, of 11 OOs treated conservatively, one occurred in the lamina of the lumbar spine [10]. They

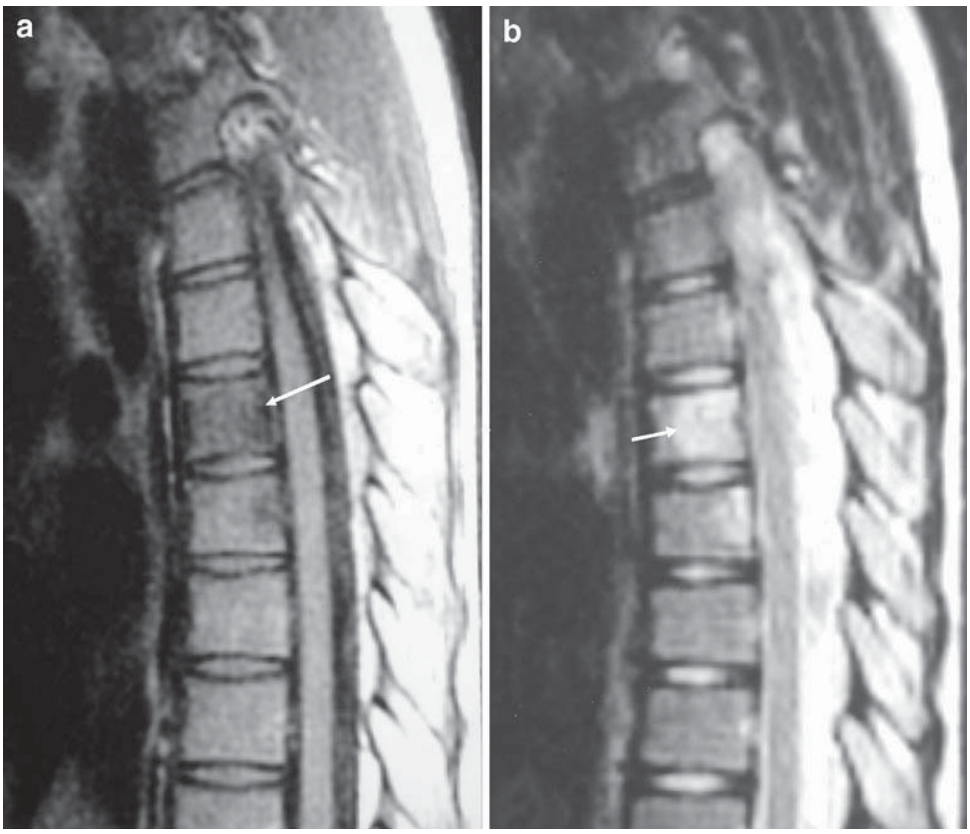

Fig. 1 **a** Sagittal Spin Echo T1-weighted (SE T1W) MRI: eight months post initial presentation. **b** Fast Spin Echo T2-weighted (FSE T2W) MRI: eight months post initial presentation

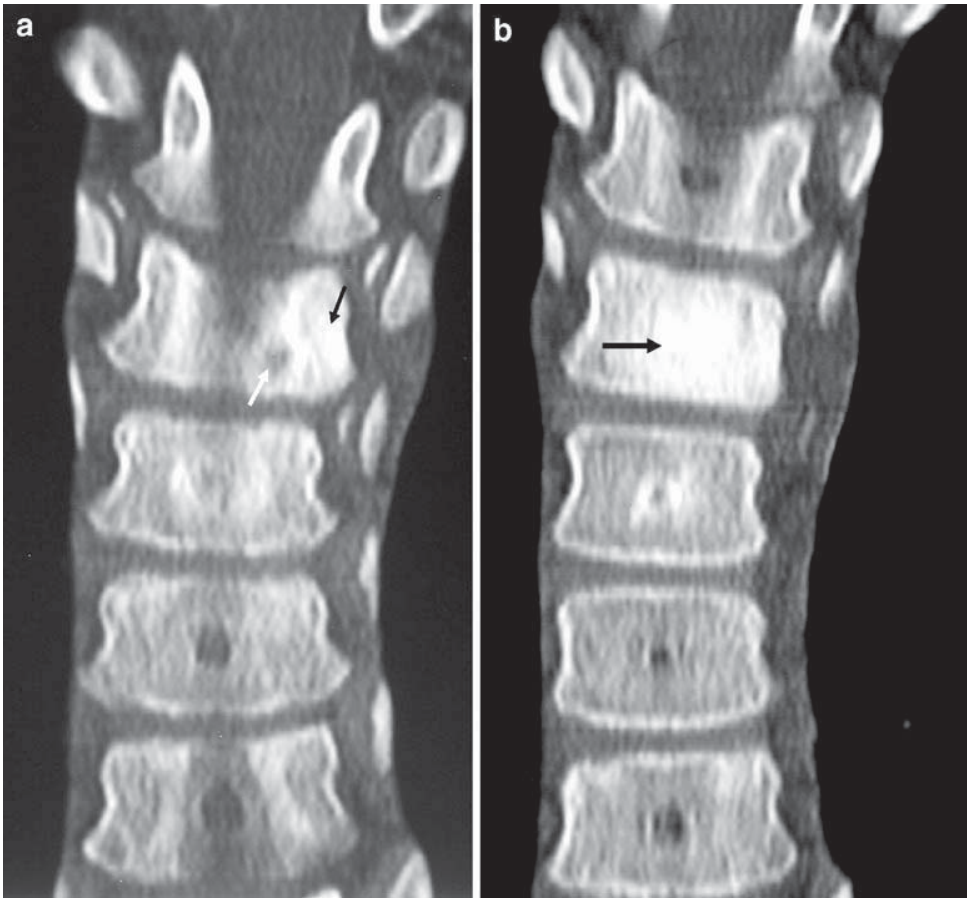

Fig. 2 **a** CT: eight months post initial presentation. **b** CT with contrast: eight months post initial presentation

reported continued presence of the radiolucent nidus and perinidal sclerosis on radiographs in the majority of their patients after the resolution of symptoms. No cross-sectional imaging follow-up was detailed in their series. From a series of nine patients with OO treated conservatively compared with 15 treated by surgical excision, Kniesl and Simon concluded that the administration of NSAIDs could be *as effective* as operative excision for the treatment of OO [4]. They advocated conservative treatment, particularly for complex lesions with potentially significant surgical morbidity. Coulier et al. reported the only case in the literature of the spontaneous resolution of spinal OO with imaging follow-up in a 17-year-old girl with OO of the lateral mass of the axis [5]. In their case, the lytic lesion almost completely disappeared with some residual hyperostosis on CT after treatment with NSAIDs for eight months. However, no MRI follow-up was presented in their case.

These studies support the theory that OOs are clinical entities that are non-progressive in nature. They state that conservative management with the long-term administration of NSAIDs may be of symptomatic and therapeutic benefit, posing as an alternative management option to surgical excision. The dramatic response of pain to NSAIDs is thought to be due to the high levels of prostaglandin (PGE_2) and prostacyclin (PGI_2) produced within the nidus [11]. Furthermore, the presence of extensive perinidal oedema in OO, as seen on MRI, is thought to be due to high levels of cyclo-oxygenase-2 expression in neoplastic osteoblasts in the nidus [12]. NSAIDs may reduce the inflammation, which is considered to be a major contributing factor in exacerbating vascular pressure and the stimulation of autonomic nerves in the vicinity of the lesion. These factors may, subsequently, cause the pain-provoked, asymmetric muscle spasm, triggering a reactive scoliosis [13, 14].

Surgical excision of the nidus has long been considered to be the gold standard in the treatment of spinal OOs [2, 15, 16]. However, complex lesions that can be difficult to localise intra-operatively may require wide excision of a nidus, resulting in variable levels of post-operative morbidity. RF and laser ablation are attractive alternatives, considered by some authors not only to be a safe and effective option, but, potentially, the treatment of choice for OOs [9, 15]. However, several authors have highlighted that this technology may have deleterious effects if applied to spinal lesions in the proximity of neural structures within the spinal canal, lateral recesses and neural foramina [9].

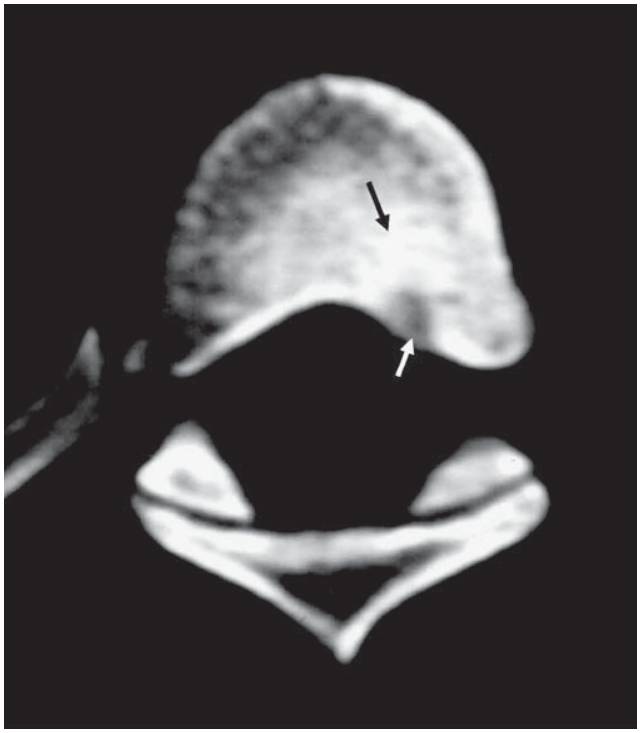

Fig. 3 CT: 14 months post initial presentation

Furthermore, they have recommended that percutaneous RF ablation should be contraindicated in spinal OOs if the nidus is <5 mm from neurological structures [15]. A study by Rosenthal et al. stated that safety can generally be assured if the electrode is at least 1 cm away from major neural structures [9]. As developments continue to advance, proponents of this technique argue that medical management only has a role in those patients with OOs who refuse operative surgery or percutaneous ablation [17].

This interesting case highlights a potential therapeutic role for expectant conservative therapy via the long-term administration of NSAIDs in the management of OOs of the spine. However, further research is essential in order to determine whether this is a reliable and appropriate treatment option. Conservative therapy may be of value in carefully selected cases where early symptomatic control is achieved. However, caution must be taken in patients with an immature skeleton, significant skeletal deformity or those presenting after a long delay before diagnosis, especially given the potential chronicity of these painful lesions. Failure to exert adequate control may lead to significant compromise in the quality of life, despite ongoing medical management. A further caution must be placed on the chronic use of NSAIDs and the increased risk of mucosal disruption of the gastrointenstinal tract. In the case of spinal tumours, it may be a suitable option in complex or inaccessible lesions due to the attendant risks of percutaneous ablation or surgical excision. It is important to note that these are less frequent. At present, surgical excision remains the gold standard in managing these lesions, with percutaneous RF ablation presenting a dynamic and attractive alternative.

From an imaging perspective, we demonstrate the radiological evolution and eventual resolution of a conservatively treated lesion using serial CT and MRI. To our knowledge, this has not been reported previously in the literature. Furthermore, the lesion presented is located in the posterolateral vertebral body, which is a less common location in terms of spinal OOs. Fine-cut CT has been considered the modality of choice in demonstrating OO [3, 13]. However, MRI demonstrates characteristic inflammatory changes in the neural arch, suggesting a diagnosis of spinal OO [18]. Thus, CT and MRI may both be useful surveillance modalities in demonstrating the radiological regression of a lesion treated by a conservative approach. In particular, we suggest that MRI is used for follow-up as the disappearance of previously observed oedema could be considered as a radiological sign of resolution. However, more detailed studies are required to correlate the length of conservative treatment and the disappearance of oedema with the resolution of pain and the regression of the spinal OO.

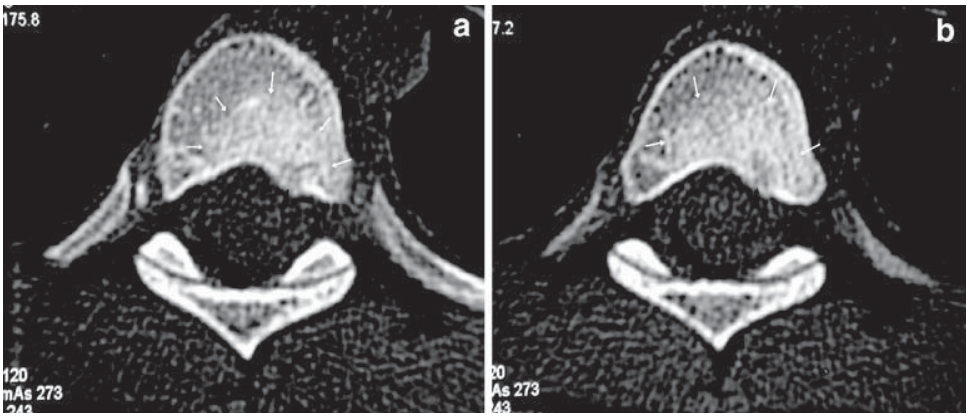

Fig. 4 **a** CT: 16 months post initial presentation. **b** CT with contrast: 16 months post initial presentation

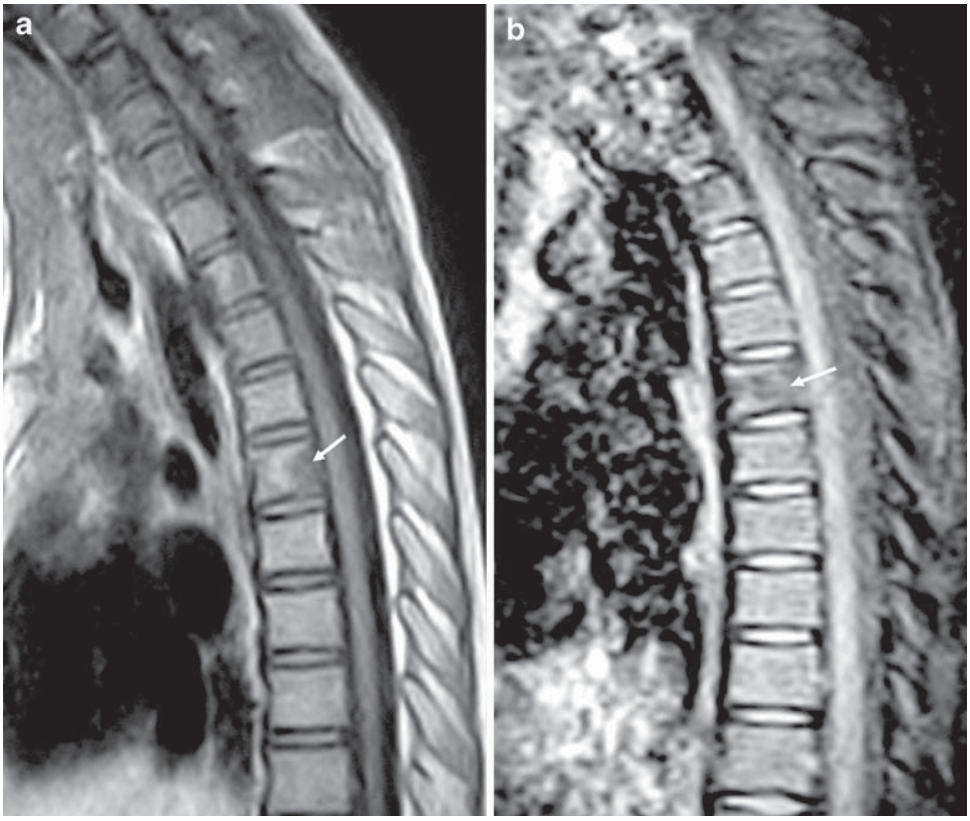

Fig. 5 **a** SE T1W MRI: 16 months post initial presentation. **b** STIR MRI: 16 months post initial presentation

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ARTICLE

G.R. Kaplan
A. Saifuddin
J.A.S. Pringle
M.H. Noordeen
M.H. Mehta

Langerhans' cell histiocytosis of the spine: use of MRI in guiding biopsy

G.R. Kaplan, FRCR
A. Saifuddin, FRCR (✉)
Department of Diagnostic Imaging,
The Royal National Orthopaedic Hospital Trust, Brockley Hill, Stanmore, Middlesex HA7 4LP, UK

J.A.S. Pringle, FRCS
Department of Morbid Anatomy,
The Royal National Orthopaedic Hospital Trust, Brockley Hill, Stanmore, Middlesex HA7 4LP, UK

M.H. Noordeen, Mch FRCS (Orth)
M.H. Mehta, FRCS
Department of Spinal Deformities,
The Royal National Orthopaedic Hospital Trust, Brockley Hill, Stanmore, Middlesex HA7 4LP, UK

Abstract The MRI features of two cases of spinal Langerhans' cell histiocytosis with multilevel involvement are presented in which MRI was of help in differentiating active from inactive healing lesions by the demonstration of signal changes in the vertebral body marrow of the active lesion, manifest as low signal intensity on T1-weighted sequences and high signal intensity on T2-weighted sequences. This distinction could not be made by plain radiography or bone scintigraphy. In cases where biopsy is required for diagnosis, MRI is recommended to guide the biopsy towards levels suggestive of active involvement.

Key words Langerhans' cell histiocytosis · Spine · MRI · Biopsy

Introduction

Langerhans' cell histiocytosis (LCH) encompasses a spectrum of conditions characterised pathologically by the invasion of various tissues by a pleomorphic infiltrate containing Langerhans' cells, which are normally found only in the skin and draining lymph nodes [1]. The commonest form of the disease is restricted to the skeletal system (a condition previously termed eosinophilic granuloma of bone or histiocytosis X). The spine is involved in approximately 28% of cases of eosinophilic granuloma [2]. In one study [3], only three of 28 patients with spinal involvement had multilevel disease at presentation.

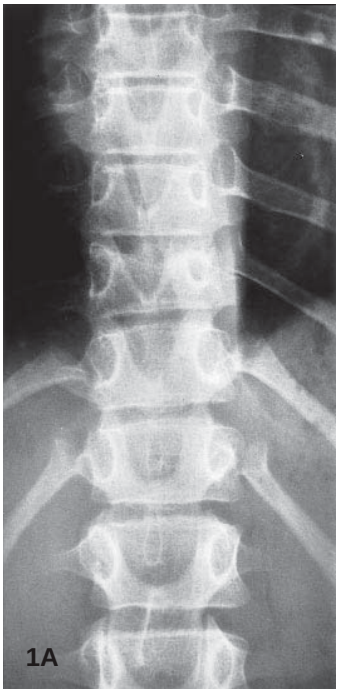
In LCH, a characteristic feature of vertebral involvement is that the endplates are not involved. For this reason, the vertebrae typically regain some height once the disease becomes inactive [4]. This presents a dilemma to the radiologist or clinician when faced with a child whose spinal radiographs show multiple partially collapsed vertebrae. Which vertebra is actively involved and collapsing and which one is already beginning to reconstitute its height?

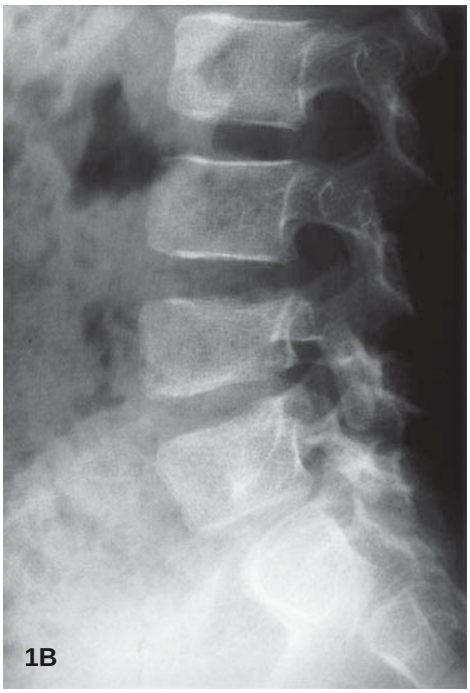
We present two patients with multilevel disease limited to the spine in whom MRI suggested different degrees of vertebral activity. Biopsies of the relevant vertebrae supported the MRI findings.

Case reports

Case 1

A 7-year-old girl presented with a 2-month history of thoracic and lumbar back pain. Examination of the spine was unremarkable. Plain radiography of the lumbar spine demonstrated collapse of T4, T10 and L5 (Fig. 1). Whole-body bone scintigraphy (Fig. 2) identified increased activity in the mid-thoracic spine, T10 and the sacrum. The patient's symptoms continued intermittently. MRI of the lower thoracic and lumbar spine was performed and demonstrated collapse and signal intensity (SI) changes in both T10 and L5 (Fig. 3). Col-





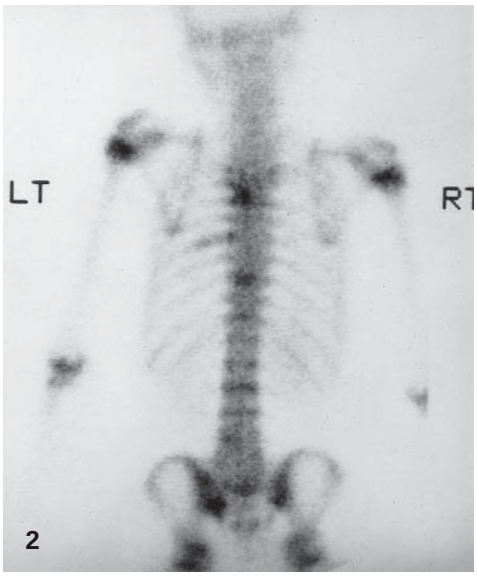
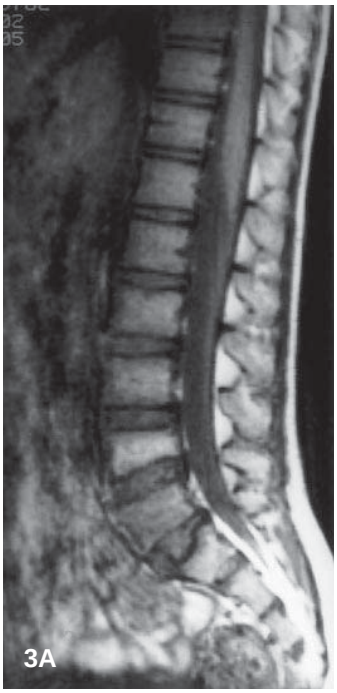


Fig. 1A, B Case 1. **A** Anteroposterior radiograph of the lower thoracic spine demonstrating collapse of T10. **B** Lateral radiograph of the lumbar spine showing reduced height of L5 (the patient has six lumbar vertebrae)

Fig. 2 Case 1. Posterior view 99^{m} Tc bone scan showing increased activity in the mid-thoracic spine, T10 and in the sacrum



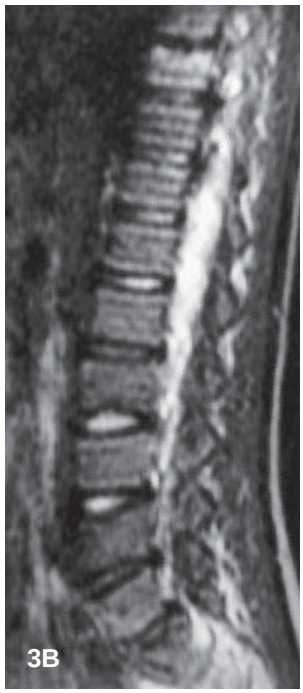


Fig. 3A, B Case 1. **A** Sagittal T1-weighted spin echo (SE) and **B** T2-weighted SE MR images of the lower thoracic and lumbar spine. L5 shows increased signal intensity (SI) on the T1-weighted sequence and isointensity on the T2-weighted sequence, indicating fatty replacement consistent with a healed lesion. T10 shows decreased SI on the T1-weighted sequence and increased SI on the T2-weighted sequence, indicating oedema consistent with an active process (note that the patient has six lumbar vertebrae). Collapse of the superior endplate of the sacrum is also identified, indicating previous involvement

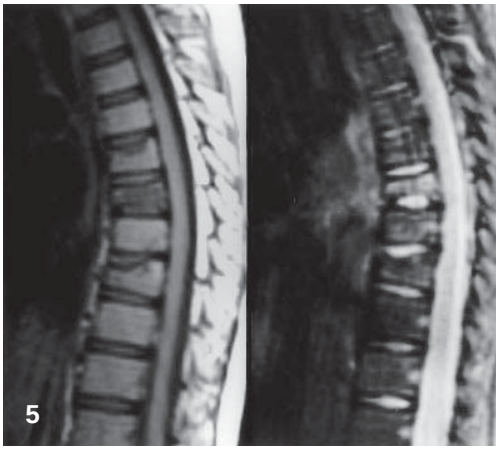
lapse of the superior endplate of the sacrum was also identified but marrow SI in the sacrum was normal.

CT-guided biopsy of L5 was performed revealing haematopoietic and fatty marrow with evidence of increased vascularity. The bone trabeculae showed active surface osteoblasts with the majority having parallel cement lines and layers of appositional reactive bone. The features indicated a healing response but the cause of the abnormality was not evident. Since the patients symptom's were progressing, a further percutaneous needle biopsy was undertaken, this time at the T10 level. Histological examination of the tissue core showed trabeculae of lamellar bone, marrow spaces and blood clot containing a mixed inflammatory infiltrate including numerous eosinophils together with large cells with reniform nuclei and copious cytoplasm. These showed positive immunohistochemical staining with S100 and CD1a, confirming the diagnosis of LCH. The patient was treated conservatively. At latest follow-up she was entirely asymptomatic.

Case 2

An 11-year-old boy presented with a 6-month history of recurrent mid-thoracic back pain. Examination revealed tenderness and limitation of movement with a minor focal kyphosis. Plain radiographs of the thoracic spine showed wedge collapse of T7 and T9. Whole-body bone scintigraphy (Fig. 4) revealed increased activity in the T6–7 region and at T9. MRI of the thoracic spine (Fig. 5) showed collapse of T6, T7 and T9 with SI changes limited to T7.

Percutaneous needle biopsy of T7 was therefore undertaken. Histological examination confirmed a diagnosis of LCH. The patient was treated in a brace and prescribed analgesics. At final follow-up, the patient was asymptomatic as regards the spine. Repeat MRI at this stage demonstrated hyperintensity in T7 on T1-weighted images consistent with fatty replacement and healing (Fig. 6).



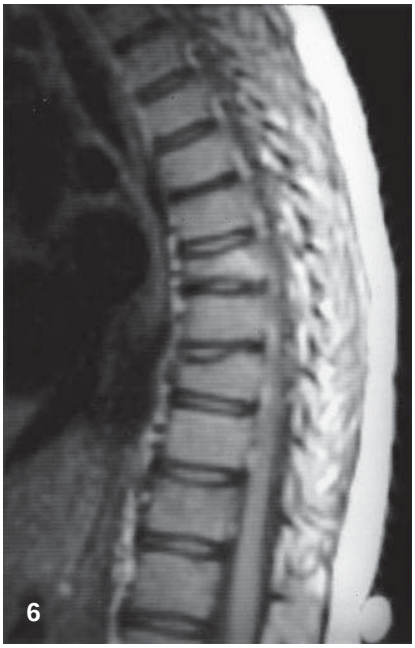


Fig. 4 Case 2. Posterior view 99^{m} Tc bone scan of the thoracic spine showing increased activity in the T6–7 region and in T9

Fig. 5 Case 2. Sagittal T1-weighted SE (*left*) and T2-weighted SE (*right*) MR images of the thoracic spine. T9 shows normal SI on both sequences suggesting an inactive lesion, whereas T7 shows reduced SI on the T1-weighted sequence and increased SI on the T2-weighted sequence consistent with an active lesion. There is also a minor region of decreased SI on the T1-weighted sequence under the superior endplate of T1, which shows a normal SI on the T2-weighted sequence. This may represent a partially healed lesion.

Fig. 6 Case 2. Sagittal T1-weighted MR image of the thoracic spine at final follow-up. The SI in T7 is now increased consistent with fatty replacement and healing. The SI in T6 has also returned to normal

Discussion

In the spine, LCH most commonly affects the thoracic vertebrae, followed by the lumbar and cervical regions. In the series of Robert et al. [3], 46 of 54 lesions involved the body while only seven lesions involved the posterior elements. In only one case was simultaneous involvement of the vertebral body and posterior elements present. The vertebral body may collapse in a symmetrical or asymmetrical fashion and associated paravertebral soft tissue mass is uncommon [4], although it has been reported [5, 6]. Extraosseous disease with cord compression is occasionally demonstrated, but is also a non-specific feature.

The classical appearance of "vertebra plana" is only seen in approximately 15% of cases [4]. Therefore, in the absence of this finding, a differential diagnosis needs to be considered. This includes lymphoma, leukaemia, metastases from solid tumours such as neuroblastoma, Gaucher's disease and osteogenesis imperfecta [4]. In such cases, vertebral biopsy may be required to achieve a diagnosis.

The natural history of LCH in the spine is one of partial restoration of vertebral height following either spontaneous resolution of the disease or various treatments. Nesbit et al. [4] studied serial radiographs of 17 lesions for between 2 and 11 years and found that 13 of these lesions showed partial reconstitution of vertebral body height. This was not influenced by the treatment patients received, regardless of whether this was radiotherapy, chemotherapy or corticosteroids. Partial reconstitution of vertebral body height was also shown in 15 of 20 vertebral lesions in the study by Sartoris and Parker [2]. The rate of healing was again unaffected by the mode of therapy but was related to the age of the patient, with the rate of healing being inversely proportional to the patient's age at presentation. Up to 85% of normal height can be achieved in some cases. Healing with fusion across the disc space is also a feature.

It is clear that when a child presents with multilevel spinal involvement plain radiography may not be able to distinguish whether a partially collapsed vertebra is in a stage of active collapse or in a stage of healing with reconstitution of its height. Some vertebrae show a "bone within a bone" appearance as they heal, but this finding is only seen following collapse to the degree of vertebra plana [4].

Bone scintigraphy is less sensitive than plain radiography for identifying lesions of LCH. Parker et al. [7] found that only 35% of lesions visible on radiographs were seen on radionuclide studies. This is contrary to earlier studies but agrees with the findings of Siddiqui et al. [8], who found that seven of 20 bone scans were completely normal despite extensive disease shown by plain radiography. In only one patient was the bone scan abnormal with a normal radiograph. It has been suggested that radionu-

clide uptake may be related to disease activity [8]. This may well be true, but case 2 in the present series indicates that a vertebra which has healed (as manifest by normal MR signal intensity within the vertebral marrow) may still show increased uptake on scintigraphy. This is presumably due to the increased osteoblastic activity associated with active reconstitution of vertebral height. Thus scintigraphy cannot be considered a sensitive technique for identifying disease activity or, therefore, for guiding biopsy.

There are several studies that have described the MRI features of LCH, mostly in lesions not involving the spine [9,10]. Those cases involving the spine, however, show non-specific features including vertebral collapse and reduced marrow signal intensity on T1-weighted sequences with increased signal intensity on T2-weighted sequences [9, 11, 12]. The two cases involving the spine in the series of de Schepper et al. [10] showed intermediate to high signal intensity compared with muscle on T1-weighted sequences and high signal on T2-weighted sequences. These authors could not confirm a relationship between high signal on T1-weighted sequences and increased lipid content or stagnant blood within the lesion at pathological examination.

As far as we are aware, the MRI features of multilevel spinal involvement have not previously been reported. Both patients in the present series had a chronic history of recurrent back pain and plain radiography showed multilevel involvement in the spine with no evidence of extraspinal disease. In case 1, T4, T10, L5 and the sacrum were shown to be involved at various stages. MRI of the lower thoracic and lumbar spine showed reduced SI in T10 on T1-weighted sequences and increased signal on T2-weighted sequences, while there was mild increased SI in L5 on T1-weighted sequences with normal marrow signal on T2-weighted sequences. Both vertebrae showed evidence of collapse. Biopsy of L5, chosen because of the abnormal signal characteristics and the easier location for percutaneous biopsy, showed evidence of a non-specific healing response in the marrow spaces and trabeculae. Biopsy of T10 at a later stage revealed typical histological features of LCH, allowing a definitive diagnosis. We assume that the signal changes in L5 were consistent with a healed lesion of LCH. Similar SI changes were seen in T7 in case 2 at a stage when the child had no significant back pain. Marrow can show evidence of increased SI on T1-weighted sequences following various treatments such as radiotherapy and chemotherapy [13]. Successful treatment of spinal osteomyelitis can also result in increased SI in the affected marrow on T1-weighted sequences [14]. A similar process presumably occurs following spontaneous healing of lesions of LCH. The cases of de Schepper et al. [10] also showed mild hyperintensity on T1-weighted sequences but differed in that there was also increased marrow signal on T2-weighted sequences. In case 2, two of the three abnormal levels on radiography and scintigraphy showed normal marrow SI on both T1- and T2-weighted sequences. The vertebral body showing abnormal SI (low on T1 and high on T2) was therefore chosen for biopsy, revealing typical histological features of LCH. Although biopsy of one of the other lesions would have been of interest to determine the activity of the disease at sites with normal marrow SI, it was not felt justified based on the experience of case 1.

The value of MRI in the local staging of diseases involving the spine is well known. The two cases presented suggest that MRI may be of value in guiding biopsy of patients with suspected multilevel spinal involvement with LCH. We propose that MRI is useful not only in assessing associated reactive bone marrow and soft tissue changes, but also in assessing the activity of the vertebral body lesion, and therefore providing a guide to the optimal biopsy site.

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Traumatic Lumbosacral Dislocation

Report of Two Cases

Athanasios I. Tsirikos, MD,* Asif Saifuddin, FRCR,† M. Hilali Noordeen, FRCS,* and Stewart K. Tucker, FRCS*

Study Design. A retrospective study of 2 patients with traumatic lumbosacral dislocation.

Objectives. To discuss the difficulty in making diagnosis and the effect of surgical treatment.

Summary of Background Data. Traumatic lumbosacral dislocation is an uncommon injury, which creates diagnostic difficulty and is typically managed by open reduction internal fixation of the lumbosacral spine.

Methods. Medical notes and imaging of the 2 patients were reviewed.

Results. Both patients were engaged in high-energy accidents and had concomitant injuries. Patient 1 was initially misdiagnosed as having L5 lytic spondylolisthesis and was treated with a lumbar corset. She developed progressive low back and left leg pain. Eleven months after the accident, a bilateral lumbosacral dislocation with right S1 superior facet fracture, disc rupture, posterior soft tissue disruption, and a resultant Grade 4 L5–S1 traumatic spondylolisthesis was identified. She underwent open reduction, followed by a staged anteroposterior spinal arthrodesis using instrumentation with excellent results. Patient 2 sustained a unilateral L5–S1 facet dislocation without neurologic deficit, which reduced spontaneously. The evaluation demonstrated a grossly disturbed posterior ligamentous complex adjacent to the lumbosacral articulation. A combined anteroposterior spinal fusion with instrumentation was performed with favorable outcome.

Conclusion. Meticulous clinical examination and careful imaging assessment, including CT and MRI, assist an early diagnosis in cases of lumbosacral dislocation. Open reduction and circumferential bony fusion restore segmental stability and painless function. [Key words: spine, trauma, lumbosacral dislocation] **Spine 2004;29:E164–E168**

Traumatic lumbosacral dislocation constitutes an unusual injury pattern, which results from violent trauma and is frequently associated with adjacent spinal fractures. The direction of the displacement may be anterior, posterior, or lateral depending on the vector of the deforming force. The dislocation may be either unilateral or bilateral. We report 2 patients involved in severe road traffic accidents, who had lumbosacral disarticulation.

From the Departments of *Spinal Surgery and †Diagnostic Imaging, Royal National Orthopaedic Hospital NHS Trust, Middlesex, U.K.
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Address correspondence to Athanasios I. Tsirikos, MD, c/o Spinal Unit, Royal National Orthopaedic Hospital, Brockley Hill, Stanmore, Middlesex, HA7 4LP, U.K.; E-mail: atsirikos@hotmail.com

Case Reports

Patient 1. A 16-year-old girl was a rear-seated, shoulder and lap strap seat-belted passenger in a car, which was hit from behind by a truck. She sustained a right clavicular fracture and multiple intra-abdominal injuries. Initial investigation revealed that multiple sections of her bowel had become devascularized, and she underwent an urgent laparotomy with partial resection of the small intestine, from which she made a satisfactory recovery. However, in the following days, she started complaining of constant low back pain. Imaging evaluation, including plain radiographs (Figure 1A) and computed tomography (CT) of the lumbar spine (Figure 1B), was performed and was initially reported to show a right pars interarticularis defect at L5–S1. Magnetic resonance imaging (MRI) was subsequently obtained, which revealed a Grade 1 L5–S1 spondylolisthesis (Figure 1C). A rigid lumbar corset was prescribed and the patient was discharged from the hospital.

Her back pain persisted and with the addition of radiation to the left leg with associated numbness in the calf and dorsum of the foot. At that point, 11 months after the accident, the patient was referred to our institution. Clinical examination revealed a palpable step-off from the spine of the fifth lumbar vertebra to the sacrum, with anterior translation of L5 on S1, and a positive straight leg-raising test at 45° on the left side resulting in paresthesia. Repeat imaging, including radiographs (Figure 2A), CT with multiplanar reconstruction (Figure 2B), and MRI (Figure 2C), demonstrated Grade 4 spondylolisthesis due to left L5–S1 facet dislocation and fracture of the right superior S1 articular process. Degeneration of the L5–S1 disc and disruption of the posterior ligamentous complex were also identified.

The patient underwent staged circumferential bony fusion of the lumbosacral joint. The first stage comprised reduction of the L5–S1 dislocation with application of bilateral pedicle screws at the L5 and S1 levels through a posterior midline approach, and precontoured oversized rods to provide distal corrective force. Realignment was achieved by slow traction on L5 after the placement of the screws. Intraoperative findings showed disruption of the interspinous ligaments between L5 and S1. Bilateral facet joint dislocation and the fractured right superior S1 facet were confirmed. The left L5–S1 facet was excised to facilitate reduction. After securing the rods at L5, both rods were pushed caudally to achieve further correction and restore the sagittal alignment of the previously dislocated segment. Bilateral posterolateral arthrodesis was performed with onlay autologous bone graft harvested from the posterior iliac crest. The procedure was performed under spinal cord monitoring using epidural recording of somatosensory-evoked potentials, with normal traces obtained and maintained throughout the operation.

One week later, the patient underwent anterior spinal fusion with the use of a threaded metallic cage and iliac crest bone autograft retained from the first stage. The lumbosacral articulation was exposed by a transperitoneal approach through the previous laparotomy scar.

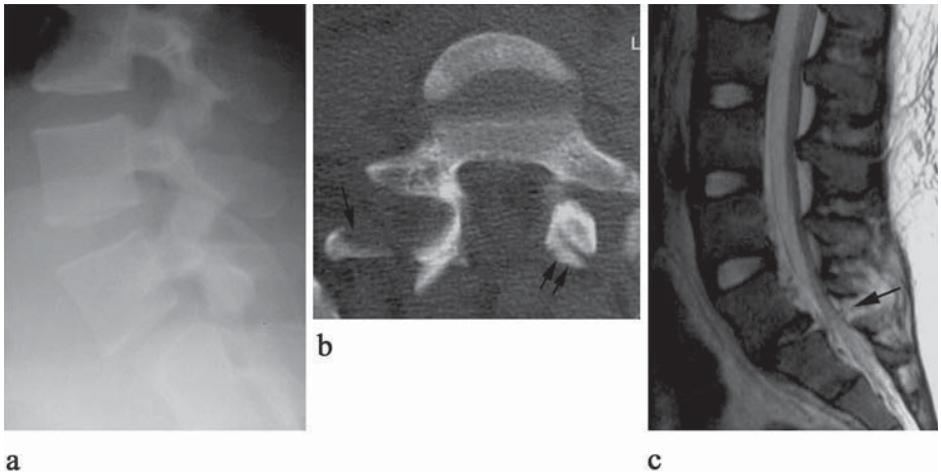

Figure 1. Patient 1. Imaging immediately postinjury. **a:** Lateral radiograph of the lumbosacral junction showing Grade 1 L5–S1 spondylolisthesis. **b:** Axial CT through the lumbosacral junction demonstrates disruption of the neural arch. Note fracture of the right transverse process (arrow) and the "naked" facet sign (double arrow). **c:** Sagittal T2-weighted MRI demonstrates avulsion of the L5–S1 disc from the sacrum and disruption of the ligamentum flavum (arrow).

Satisfactory reduction of the facet dislocation and the spondylolisthesis was achieved and maintained at follow-up. The patient's postoperative course was uneventful and her neurologic picture recovered completely. She was mobilized in a lumbar orthosis with extension above the knee on the right side for 3 months.

Two years after the surgery, the patient had her first baby through a vaginal delivery without complications. Her spine remained asymptomatic throughout her pregnancy. At the most recent examination, 3 years after surgery, she was pain free with normal neurologic function and radiographic evidence of a stable fusion (Figure 3).

Patient 2. A 42-year-old patient was the victim of a road traffic accident, sustaining a closed head injury from which he recovered uneventfully. However, during his hospitalization, he experienced low back pain without neurologic deficit. CT demonstrated the presence of a right unilateral L5–S1 facet dislocation with associated ipsilateral L5 transverse process fracture (Figure 4). While being transferred for an MRI scan, he had an acute, intense pain in the lumbosacral region. The ensuing MRI confirmed spontaneous reduction of the dislocation and showed normal bilateral L5–S1 facet joints. The L5–S1 disc appeared normal, although there was evidence of posterior ligamentous complex rupture. The neurologic examination remained normal.

Because of the significant risk of residual lumbosacral instability and recurrence of the dislocation, the patient was treated surgically by a combined single-stage anteroposterior spinal arthrodesis. The spine was first approached through a posterior midline incision with the patient in the prone position. The posterior spinal elements were dissected and a hematoma was found in the paraspinal musculature, with disruption of the interspinous and supraspinous ligaments on the side of the dislocation. The contralateral facet joint, exposed for the purpose of arthrodesis, appeared normal. Posterolateral bony fu-

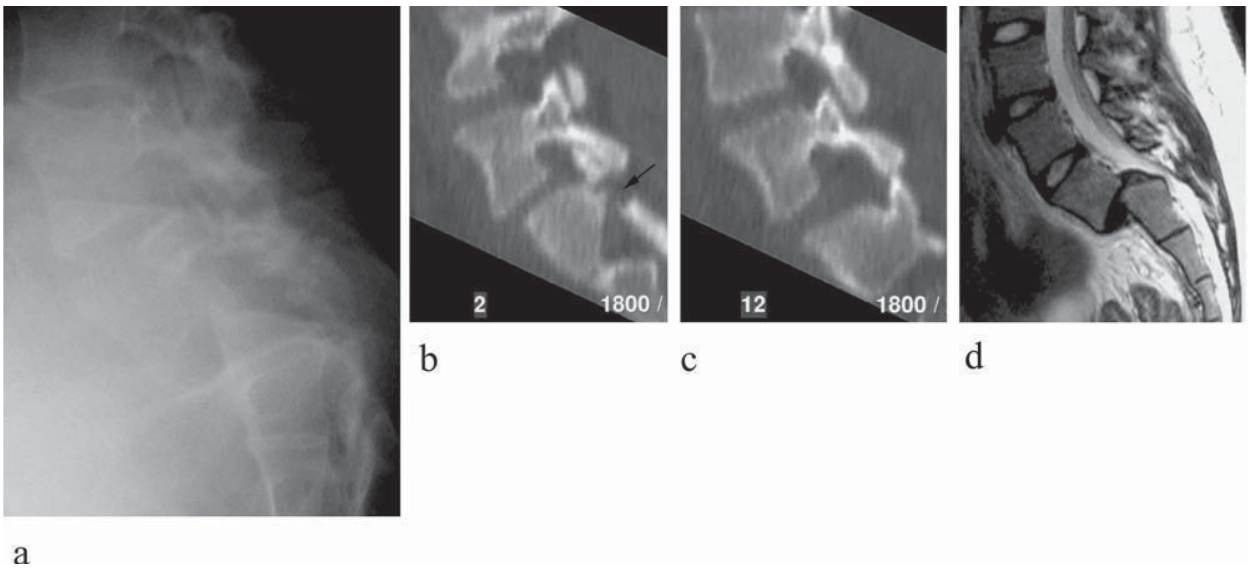

Figure 2. Patient 1. Delayed imaging. **a:** Lateral radiograph of the lumbosacral junction shows Grade 4 L5–S1 spondylolisthesis. **b:** CT of the lumbosacral junction. Right parasagittal multiplanar reconstruction showing fracture of the S1 superior articular facet (arrow). **c:** CT of the lumbosacral junction. Left parasagittal multiplanar reconstruction showing dislocation of the left L5–S1 facet joint. **d:** Sagittal T2-weighted MRI demonstrates Grade 4 L5–S1 traumatic spondylolisthesis with compression of the cauda equina and degeneration of the L5–S1 disc.

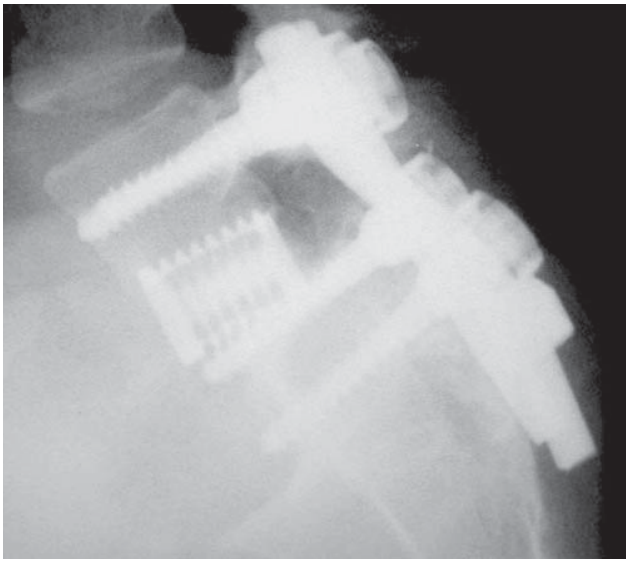

Figure 3. Patient 1. Postoperative imaging. Lateral radiograph demonstrates solid instrumented L5–S1 circumferential fusion.

sion was performed and instrumentation was used in the form of a pedicle screw-rod construct securing fixation on the L5 and S1 segments. Hydroxyapatite bone substitute was applied to enhance fusion. A recording electrode was introduced to the epidural space by an open procedure and monitored normal spinal cord function during the posterior stage of the surgery. The patient was then rolled to the supine position, the lumbosacral joint was exposed through a Pfannenstiel incision and a transperitoneal approach, and a threaded metallic cage was inserted into the disc space to achieve anterior interbody fixation and arthrodesis. During the anterior procedure, the L5–S1 disc seemed to have a normal appearance. The patient had an uncomplicated recovery from his operation. Postoperative management consisted of a lumbar corset worn for 3 months.

Two years postsurgery, he was completely asymptomatic, with normal neurologic findings, and had resumed his previous level of physical activities. Radiographic evaluation confirmed a solid spinal fusion.

■ Discussion

Watson-Jones[1] was the first to describe lumbosacral dislocation, which he attributed to a hyperextension mechanism. However, most authors consider a combination of hyperflexion with compression as the commonest mechanism of injury to produce bilateral L5–S1 dislocation.[2–4] Dewey and Browne[5] postulated that a severe force is applied to the lumbar spine and results in forward shifting on the fixed sacrum. The presence of a presumed unilateral rotatory element is implicated in unilateral facet dislocations.[6–8] Roaf[9] showed experimentally that a combination of hyperflexion with vertical loading and rotation is necessary to create this injury. Hyperflexion alone is not capable of producing either pure dislocation or fracture–dislocation in the lumbar spine.[9] Other investigators have assumed hyperflexion with various degrees of distraction and sheer forces to be the most frequent mechanism of lumbar facet dislocation.[10–12] There are also reports of direct traumatic vec-

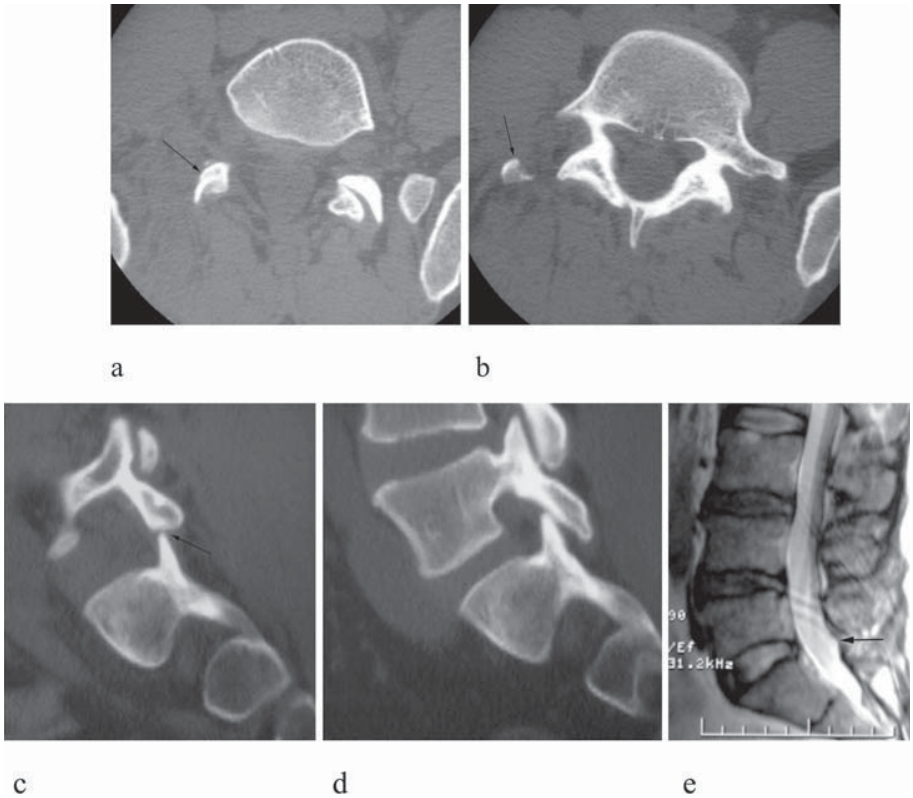

Figure 4. Patient 2. **a:** Axial CT at the lumbosacral junction demonstrating unilateral right L5–S1 facet dislocation manifest as the "naked" facet sign (arrow). **b:** Axial CT at the lumbosacral junction demonstrating right L5 transverse process fracture (arrow). **c:** CT at the lumbosacral junction. Right parasagittal multiplanar reconstruction showing L5–S1 facet dislocation. **d:** CT at the lumbosacral junction. Left parasagittal multiplanar reconstruction showing normal L5–S1 facet joint. **e:** Sagittal T2-weighted MRI. The lumbosacral disc appears normal, although there is evidence of ligamentum flavum rupture (arrow).

tors that act as a tangential force applied to the L5–S1 apophyseal joints and cause the dislocation to occur.[13,14]

Radiologic diagnosis of this injury is dependent on good quality initial radiographs, which will demonstrate the altered association of the lumbosacral facet joints. However, emergency room radiographs are frequently inadequate and can be easily misdiagnosed as normal such that the lesion is missed on original presentation.[4,15,16] Our first patient was misdiagnosed as having an L5–S1 lytic spondylolisthesis, and the correct diagnosis was not established until 11 months postinjury. However, careful assessment of the original MRI study demonstrated avulsion of the L5–S1 disc from the sacrum and disruption of the posterior ligamentous complex.

Aihara *et al*[17] reviewed 7 patients with fracture–dislocations of the fifth lumbar vertebra and analyzed 50 previously reported cases. They developed a classification system with the aim at planning operative management. According to the proposed classification system, Patient 1 would be categorized as having a Type 2 injury and Patient 2 as Type 1.

Herron and Williams[18] suggested that the presence of transverse processes fractures should lead to the suspicion of traumatic lumbosacral dislocation. The presence of a transverse process fracture appears to be a clear association with lumbosacral dislocation in consecutive historical reports.[2–5,8,10,11,13,15,18–22] In addition, the existence of a perineal laceration should alert radiographic examination of the lumbar spine to exclude the occurrence of L5–S1 dislocation.[2,6]

Unilateral facet dislocation is an injury classically localized to the mobile cervical spine.[23] However, its rarity in the lumbosacral spine is attributed to the more vertical sagittal orientation of the facet joints and the presence of robust muscular and capsuloligamentous structures that provide substantial stability in the lower lumbar region.[8,22] Unilateral dislocations are unlikely to cause neurologic compromise.[8,24] On the contrary, bilateral dislocations, especially those associated with a significant degree of spondylolisthesis, can produce neurologic deficit. The most commonly affected nerve root is S1, and even with reduction of the dislocation, only partial recovery of nerve function should be anticipated.[13]

The management of lumbosacral dislocation has varied. Newell[14] and Zoltan *et al*[22] reported success with conservative treatment in the unreduced position. Beguiristain *et al*[13] treated a 5-year-old boy with pure traumatic anterior L5–S1 dislocation with traction, followed by a lumbar plaster jacket. A favorable outcome was obtained 8 years postinjury, even though the dislocation was only partially reduced. In the case of subacute or inveterate dislocation, previous authors have suggested conservative management, in view of the complexity of surgical treatment and reported excellent clinical results.[15] Boger *et al*[6] presented a case of a 24-year-old male patient who sustained a spontaneously reducing unilateral facetal dislocation and was treated nonsurgically with good outcome.

Holdsworth[25] postulated that spinal dislocations without associated fracture should be treated by open reduction and fusion particularly in unreduced dislocations. Irrespective of the type of injury, lumbosacral dislocation is a three-column injury and consequently highly unstable.[26] Open reduction internal fixation and bone grafting as soon after the injury as possible will ensure complete correction of the deformity, prevent later deterioration, and should be considered the treatment of choice for unilateral or bilateral lumbosacral dislocations.[2–4,8,11,17–21]

Closed reduction of bilateral lumbosacral dislocation has not been advised in previous reports, citing the risk of increasing the neurologic deficit and the apparent difficulty in reducing the displacement.[3,5,7,10]

Partial facetectomy can be performed in cases of pure dislocation to facilitate reduction and does not jeopardize segmental stability provided internal fixation is used.[2,5,7,24] However, if reduction can be attained without facet resection, the intact apophyseal joints serve as a block against deforming forces that can cause redislocation. Decompressive laminectomy is not indicated in the absence of neurologic signs and can create further instability.[3,10] It has, however, been recommended in cases of cauda equina compromise or delayed reduction.[7,18,20,27]

Injury to the intervertebral disc is a prominent feature of L5–S1 facet dislocation and can be reliably assessed with preoperative MRI.[10,27] Attention should be drawn to the disc disruption in order to prevent the development of cauda equina syndrome or nerve root compression after operative reduction of the dislocation.[7,10,11,27] In our first case, the use of intraoperative spinal cord monitoring confirmed intact peripheral neural function during reduction of the dislocation, and a decompressive laminectomy was not deemed to be necessary.

Even though in previous reports, interspinous wiring has been used after reduction of lumbosacral dislocations with good reported results,[2–4,11] pedicle screw instrumentation is now the preferred technique for achieving posterolateral fixation.[7,10,19] Aihara *et al*[17] recommended anterior lumbar interbody fusion after posterior reduction of the dislocation with pedicle screw instrumentation. Verlaan *et al*[27] treated a patient with bilateral dislocation with a posterior interbody fusion procedure combined with a posterolateral instrumented fusion. We also believe that lumbosacral dislocation is significantly unstable; therefore, we supplemented the posterolateral fusion with an additional anterior interbody fusion. This combined anteroposterior surgical intervention produced a solid circumferential segmental arthrodesis and restored stability of the lumbosacral junction. Consequently, both patients returned to painless function and had an excellent clinical outcome.

Finally, associated bony injury must be identified. Ebraheim *et al*[28] described the case of an 18-year-old girl who sustained a traumatic unilateral lumbosacral dislo-

cation associated with a vertical shear fracture of the sacrum and instability of the symphysis pubis.

■ Key Points

- Early recognition of a lumbosacral dislocation is challenging and depends on a careful imaging evaluation.
- Identification of all the different components of the injury guides adequate surgical treatment.
- Reduction of the dislocated lumbosacral facet joints requires open techniques.
- Combined anteroposterior L5–S1 arthrodesis with compression instrumentation achieves stability.

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ORIGINAL ARTICLE

Spinal Cord Monitoring Using Intraoperative Somatosensory Evoked Potentials for Spinal Trauma

Athanasios I. Tsirikos, MD, Joseph Aderinto, MD, Stewart K. Tucker, MD, and Hilali H. Noordeen, MD

Background: Intraoperative spinal cord monitoring is commonplace in scoliosis surgery as an adjunct to evaluate functional integrity of the cord; however, limited information is available on its applicability in spinal trauma.

Methods: We investigated the efficacy of somatosensory evoked potential (SEP) recording during reconstructive procedures in 82 patients who sustained 20 cervical, 8 thoracic, 6 thoracolumbar, and 48 lumbar vertebral fractures or fractures–dislo-cations. Seventy-one patients underwent single anterior or posterior operations and 11 combined anterior–posterior procedures. Forty patients had incomplete injuries, and 42 had no preoperative neurologic deficit. SEP trace amplitude at insertion of electrode was considered as the baseline value and was compared with the lowest intraoperative signal amplitude and the amplitude at completion of operation.

Results: Fifty-nine patients had a depression in wave amplitude of >25% during surgery; in 25 patients, the trace fell by >50%, and in 7 cases, a >75% diminution was recorded. A loss of 50% in SEP signal amplitude showed 67% sensitivity and 71% specificity in predicting neurologic outcome. Increasing trace deterioration threshold from 50% to 60% improved specificity to 81% without compromising sensitivity. A loss of >50% in SEP amplitude occurred with significantly increased incidence during the anterior compared with the posterior spinal procedures. More than 20% recovery in signal amplitude at the conclusion of the procedure in patients with incomplete injuries was correlated with favorable neurologic function.

Conclusion: Persistent intraoperative decrement in SEP amplitude and poor restitution at completion of surgery increase the risk for postoperative neurologic compromise.

Key Words: spinal cord monitoring, trauma, spine

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The risk of spinal cord injury is always present during corrective surgery involving the spine, particularly when instrumentation is used to achieve optimum spinal alignment and allow for a solid fusion. The prevalence of neurologic complications following instrumented fusion for spinal deformity has been previously reported to range between 0.7% and 2.7%.[1,2] If neural cord injury occurs, this can result in permanent neurologic damage. Intraoperative monitoring of the spinal cord provides continuous surveillance and has helped diminish the incidence of neurologic impairment.[2,3]

The efficiency of somatosensory evoked potential (SEP) screening varies depending on the type of spinal deformity. SEP recording during operations for idiopathic scoliosis has proven to be a helpful diagnostic tool with consistent sensitivity, reproducibility, and reliability in the early detection of cord compromise; acute intervention at this stage can prevent irreversible cord injury.[2,3] On the contrary, the efficacy of SEPs in the surgical management of neuromuscular scoliosis has been controversial.[4,5] Noordeen et al[6] demonstrated that, despite a lower sensitivity and specificity, the application of this method in neuromuscular scoliosis can still provide useful information and predict neurologic deficits. A number of studies have indicated a decrement of 50% in SEP signal amplitude during scoliosis surgery as the criterion for amplitude change that carries increased risk of cord dysfunction.[2,3,6] Animal studies have also confirmed the predictive value of a fall of 50% in the amplitude of SEP waves in detecting potential neural cord damage.[7–9] The role of SEP monitoring during reconstructive procedures in patients who have sustained spinal trauma is, however, less certain.

The purpose of this study was twofold: primarily, to define possible benefits from monitoring continuously electrical activity in the sensory pathways using mainly spinal epidural SEPs in predicting the ultimate neurologic outcome in a group of patients who underwent surgery following spinal column injury; and second, to determine if the threshold of 50% in SEP trace amplitude diminution, described in deformity surgery as a predictive factor for neurologic compromise, could be reliably applied during operative procedures to address spinal trauma.

MATERIALS AND METHODS

This is a retrospective study including a population of patients treated in our institution for spinal injuries between 1995 and 2000. Our study cohort comprised 82 patients who

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From the Royal National Orthopaedic Hospital, Stanmore, Middlesex, UK.
Reprints: Athanasios I. Tsirikos, MD, Spinal Unit, Royal National Orthopaedic Hospital, Brockley Hill, Stanmore, Middlesex, HA7 4LP, UK (e-mail: atsirikos@hotmail.com).

underwent 83 reconstructive procedures (Table 1). Seventy-one patients underwent isolated posterior or anterior spinal surgery, whereas combined anterior–posterior operative procedures were necessitated in 11 patients. In all but one patient in the combined procedures group, the operations were performed in one session; one patient required staged anterior–posterior approaches, and therefore he had two consecutive SEP trace recordings. The medical records of the 82 patients were reviewed to document the type of injury and the surgical treatment offered, the method of anesthesia, the posttraumatic, preoperative, and postoperative neurologic status of each individual patient and any adverse sequelae, and the fluctuation of SEP monitoring during surgery. Inclusion in this study required availability of complete intraoperative spinal cord monitoring signals and accurate documentation of the level of neurologic function before and after surgery.

All surgical procedures were performed by consultant spinal surgeons in our unit, applying a consistent perioperative management protocol with the same anesthetic team. Physiologic measurements such as lowest and highest intraoperative systolic blood pressures and body temperature at completion of the operation were also documented. Detailed information on the age and gender of patients, date and level of injury, and surgical approach and procedure were recorded. Similar techniques of anesthesia were applied. Induction was done by intravenous thiopentone or inhalation of halothane, followed by maintenance with enflurane and nitrous oxide in oxygen. Vecuronium or pancuronium was used for muscle paralysis and opioids for perioperative analgesia. During the operation, the systolic blood pressure was maintained at levels above 85 mm Hg, occasionally with the administration of labetalol or trimetaphan. Intra-arterial monitoring and central venous pressure transducers were used in all patients.

Neurologic Assessment

Neurologic outcome measures were based on changes in motor grade strength, sensory function, tendon reflexes, and gait pattern. Neurologic examination results in the preoperative and postoperative period were available for all 82 patients. Motor activity was scored according to the grading system proposed by the American Spinal Injury Association.[10] This scale quantifies muscle strength from 0 (no strength) to 5 (normal) for each one of the five muscle groups in the upper and lower extremities individually. Documentation of upper and lower extremity tendon reflexes, sensory chart diagrams, and assessment of both bladder and bowel function were available for all patients. Thus, improvement or deterioration in their postoperative neurologic picture could be determined. A minimum 2-year clinical follow-up (range 2–7.5 years) was available for all patients who participated in the study.

Spinal Cord Monitoring

Eighty-three sets of complete SEP traces from 82 patients were available for analysis. Epidural measurement of the SEPs was used to monitor the function of the spinal cord throughout the operation. The consensus technique for monitoring was the unilateral stimulation of the posterior tibial, median, or ulnar nerves with supramaximal constant-current single-pulse stimuli of 15- to 50-mA intensity, pulse duration of 0.2 milliseconds, at a rate of 14–16 stimuli/s for the lower limbs and 3–7 stimuli/s for the upper limbs applied alternately to both upper or both lower limbs. The SEP was recorded with a 4F bipolar electrode (Orthoexpress, Amersham, UK) inserted into the epidural space above the highest proposed level of fusion in 69 patients. In the patients who underwent posterior spinal procedures, the intervertebral space one to two levels above the most cephalad proposed vertebra to be instrumented was exposed through a direct incision of the ligamentum flavum, and the recording electrode was introduced in the epidural space. In patients undergoing anterior surgery, the epidural electrode was placed percutaneously through a guiding needle. In 19 patients, scalp electrodes were used to measure cortical evoked potentials. In 6 of these 19 patients, recordings were obtained from both scalp and epidural elec-

TABLE 1. Type of Operative Procedures Performed in the Patients Included in Our Series

Single Surgical Procedures (Posterior: 47 Patients, Anterior: 24 Patients)	Number of Procedures	Neurological Complications
Posterior instrumented fusion	46	1 (radicular motor deficit)
Cervical laminectomy/posterior instrumented fusion	1	1 (spinal cord sensory deficit)
Anterior corpectomy/instrumented fusion	23	0
Cervical anterior instrumented fusion	1	0
Combined Surgical Procedures (11 Patients)	**Number of Procedures**	**Neurological Complications**
Anterior corpectomy/posterior instrumented fusion (one stage)	10	2 (spinal cord motor deficit/ radicular sensory deficit)
Anterior/posterior instrumented fusion (two-stage)	2	0

trodes. In the remaining 13 patients, good cortical baseline signals could be monitored so the use of an epidural electrode was not deemed to be necessary. In 4 patients of the total number of 11 patients who underwent combined anterior and posterior procedures, scalp electrodes were used to measure SEPs during the anterior and epidural recording electrodes during the posterior stage of the operation. In most of the patients who underwent anterior cervical surgery, SEP monitoring was performed using cortical evoked potentials. In the cases when cortical SEP traces were not satisfactory, recording was obtained using epidural electrodes placed percutaneously at the C7–T1 level, and the correct position of the electrode in the spinal canal was confirmed using fluoroscopic imaging.

The signals were amplified, conditioned, and displayed on a Medelec MS91 electromyograph (Medelec, Old Woking, UK) and a PA89 preamplifier. The amplifier filters were set to BER (200 Hz to 2 kHz), and sweeps were averaged to obtain clear recordings at intervals coinciding with steps in the operation or changes in the signal. Filter settings were kept constant during the procedure, as changes would cause alteration of SEP waveforms. After 1996 a Viking (Nicolet) display unit and preamplifier were used with filter settings 200 Hz to 2 kHz. SEP trace was measured as the maximum peak-to-peak value, and hard copies of the signals were obtained on calibrated heat-sensitive paper to document deviations at different points of the surgery. The SEP wave at insertion of the electrode was taken as the control value. The lowest intraoperative SEP amplitude and trace amplitude at conclusion of operation right before closure of the skin were recorded and expressed as a percentage of the initial control measurement. Three levels of SEP signal loss, namely, >25%, >50%, or >75% of the baseline level, were used to determine significance of any alteration in amplitude. The SEP traces were designated according to the classification developed by Szalay et al[4] as false positive, false negative, true positive, or true negative (Table 2).

Statistical Methods

The Pearson χ^2 statistic ($P < 0.05$) was used to compare loss of trace amplitude and neurologic outcome. Statistical significance of the association between patients' age, gender, and change of SEP signal amplitude was determined with the application of the Mann–Whitney *U* test. The positive and negative predictive values for SEP changes during surgery were also calculated. Comparison of intraoperative systolic blood pressure and core temperature at skin closure between patients' subgroups was performed using the two-tailed Student *t* test.

RESULTS

Twenty-eight (34%) of the patients in our series were females and 54 (66%) males, with an average age at surgery of 33 years (range 8–87 years). The distribution of spinal trauma included 20 cervical, 8 thoracic, 6 thoracolumbar (T11–L1), and 48 lumbar vertebral fractures or fractures–dislocations. Surgery to address consequent spinal malalignment and restore function was performed at an average of 10 days after the injury (range 1–133 days). This was the average time required for the patients to be referred to our institution, which is a tertiary referral center. Forty-two patients (51.2%) had normal preoperative neurology. Forty of the 82 patients (48.8%) in this study had neurologic deficit prior to operation, as the result of incomplete spinal cord injuries in all cases. Thirty-six of these 40 patients (90%) were treated with either anterior decompression–fusion or combined anterior decompression and posterior fusion.

Intraoperative Fall in SEP Trace Amplitude

Fifty-nine patients (71.9%) had an intraoperative fall in trace amplitude of >25% compared with the baseline value at insertion of the electrode; in 25 patients (30.5%), the SEP signal fell by >50%, and in 7 patients (8.5%), a diminution of >75% was recorded during surgery (Table 3). When sensitivity and specificity were calculated, a decrement of 25% in waveform amplitude was associated with 67% sensitivity, 29% specificity, and one false-negative result in predicting neurologic outcome. A loss of 50% showed 67% sensitivity, 71% specificity, and one false-negative result. Seventy-five percent depression of the signal had sensitivity of 33%, specificity of 91%, and two false-negative results in predicting postoperative function. In most of these cases, the loss of trace resolved rapidly, either when the recording electrode was repositioned or spontaneously. The recording electrode in the epidural

TABLE 2. Classification of Somatosensory Evoked Potential Traces Introduced by Szalay et al[4]

Classification	Definition
True positive	Change in the signal that can be related to surgical or anesthetic events/does not necessarily imply neurologic deficit
True negative	No alteration in the signal/no neurologic complication
False positive	Change in the trace unrelated to surgical or anesthetic events/no neurologic sequelae detected
False negative	No change in the trace/neurologic injury is produced

TABLE 3. Decrement in SEP Wave Amplitude During the Surgery and Incidence of Associated Neurologic Compromise

Intraoperative Loss of SEP Trace Amplitude	No. of Recordings	Neurologic Sequelae
58%	28	1 (1.7%)
58%, recovery at completion of surgery	19	0
58%, nonrecovery at completion of surgery	6	2 (33%)

space had become dislodged or surrounded by hematoma, and its adjustment or replacement caused the signal to return. Patients with a fall in SEP amplitude of >50% that did not recover at completion of the surgical procedure demonstrated an increased risk of neurologic compromise ($P < 0.01$). However, there was no significant correlation between the incidence of SEP waveform amplitude decrease of >50% and the age or gender of the patients. Forbes et al[2] first postulated that a diminution of <50% in signal amplitude from the initial measurement during the surgical correction of spinal deformity is not accompanied by clinically detectable damage to the spinal cord and runs an exceedingly low risk of considerable motor tract complications. In this study, we found that increasing the threshold for what is considered significant trace deterioration from 50% to 60% reduced the rate of false-positive results, thus improving specificity from 71% to 81%, without compromising sensitivity, which remained at 67% (Fig. 1). Positive and negative predictive values for SEP signal changes are presented in Table 4. There was also 100% correlation between the side of the amplitude drop and the side of neurologic loss in the trunk or limb ($P < 0.001$).

A decrement in SEP recording amplitude of >50% occurred more often during the anterior than the posterior procedures (Fig. 2). Trace amplitude loss of >50% occurred in 16 of 36 patients who underwent anterior spine surgery (44%) compared with 10 of 59 patients (17%) in whom posterior-only procedures were performed. This difference was statistically significant ($P < 0.001$).

Increase in SEP Trace Amplitude at Completion of Operation

Trace amplitude at completion of operation was compared with neurologic outcome in the 40 patients with incomplete injuries and recorded preoperative neurologic deficits. A total number of 22 patients had improved SEP recordings before skin closure; 19 of these patients demonstrated an improved neurologic function after the operative procedure (Table 5). In these 19 patients, a positive statistical association could be documented between the signal changes and the neurologic outcome ($P < 0.05$). In 12 patients, the trace amplitude increased by >20% before the end of the surgery; none of these patients showed evidence of an impaired spinal cord function,

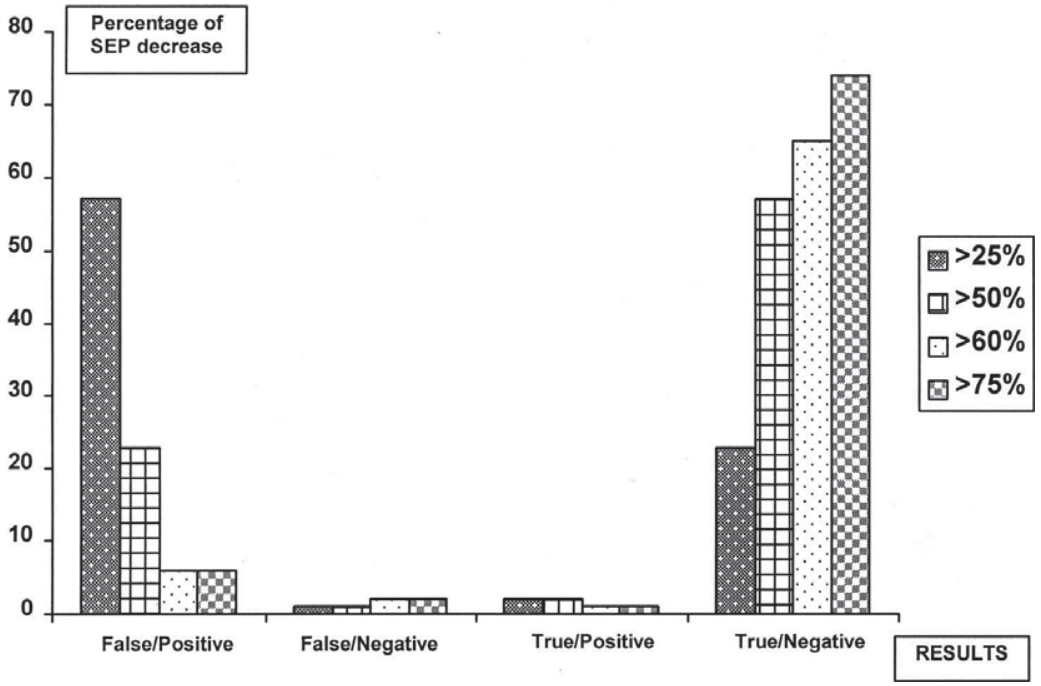


FIGURE 1. Distribution of results in relation to different percentages of SEP trace amplitude diminution during spinal surgery.

TABLE 4. Positive and Negative Predictive Values for Intraoperative SEP Changes in Regard to Neurologic Outcome

	Positive Predictive Values (%)	Negative Predictive Values (%)
>50% intraoperative loss of SEP signal amplitude	6.9	98.1
>60% intraoperative loss of SEP signal amplitude	11.1	98.5
>20% recovery SEP at completion of surgery	91.7	14.3
>40% recovery SEP at completion of surgery	100	15.1

which was either better than or the same as compared with their preoperative state. In seven patients, the trace amplitude at completion of the spinal procedure had increased by >40% of the baseline initial value. None of the patients in this group had neurologic complications, and they all demonstrated improvement in their neurologic status. Nevertheless, interestingly enough, two of the patients with up to 20% improvement in the trace amplitude compared with the original control measurement presented deterioration in their neurologic picture in the postoperative period.

In 17 patients, the SEP waveform amplitude was unchanged at the conclusion of the operation; in those cases, the neurologic functional level after surgery was equally unaltered. However, in one case, even though the end recording was consistent in amplitude with the control value, aggravation in neurologic picture was recorded after spine surgery. No significant difference was obtained when comparing systolic blood pressures or core temperatures at skin closure between the group of 18 patients with neurologic impairment and those with a documented increase in signal amplitude at completion of the surgery ($P > 0.05$). Equally, no statistical difference was noted for the same parameters between the group of 19 patients with an intraoperative rise in SEP trace amplitude and an improved postsurgical level of function and the 3 patients whose neurologic state remained the same or even worsened, although the signal recovered to a certain extent before the end of the operation ($P > 0.05$).

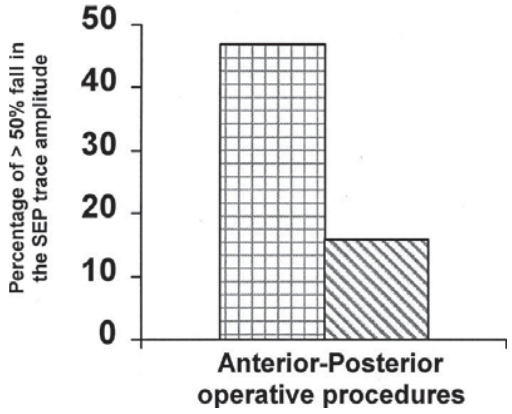


FIGURE 2. Percentage of cases of >50% depression in SEP signal amplitude during the isolated anterior versus the single posterior surgical procedures.

Neurologic Evaluation

Neurologic deterioration after surgery was detected in four patients; one patient in this group had <50% and two patients >50% diminution in SEP signal amplitude that did not recover at completion of the operation. Two of these patients developed spinal cord compromise postoperatively, even though they had a normal neurologic picture prior to surgery. We believe these four cases are worthy of further description.

The first patient with normal function before the procedure was a 31-year-old man who sustained a midcervical fracture, developed in the postoperative period right upper extremity weakness, and was found to have penetration of both C3–C4 and C4–C5 intervertebral foramina by posterior fixation screws in the right side, causing nerve root compression and accounting for his symptoms. This could not have been discovered with the epidural SEP monitoring method that we applied during the operation, where the highest recordable level is C6 and C7 (median nerve innervation). Therefore, he was excluded from the calculations of sensitivity and specificity of SEP for detection of neurologic injury.

The second patient with no preoperative neurologic abnormality was a 25-year-old man who sustained an L4 burst fracture and underwent posterior L3–L5 instrumented fusion, followed by subsequent anterior corpectomy and titanium cage insertion filled with bone autograft. Intraoperative spinal cord monitoring indicated 85% reduction in signal amplitude on the left side and 20.8% on the right, which did not recover before completion of the procedure on the left side (Fig. 3). In the immediate postoperative period, he developed transient numbness in his left lower limb, which eventually resolved spontaneously.

A 70-year-old man sustained a hyperflexion injury to his cervical spine following a fall. He presented to his local hospital with bilateral hand weakness. Plain radiographs were normal, and magnetic resonance imaging scans showed increased signal in the cord at the C7 level. He was initially treated conservatively with the application of a hard cervical collar. Four weeks later, he developed further symptoms including left leg

TABLE 5. Improvement of SEP Trace Amplitude at Skin Closure in Relation to Postoperative neurologic Outcome

SEP Signal Amplitude Recovery at Conclusion of Operation (% Above Initial Baseline Recording)	Unchanged Neurologic Status	Improved Neurologic Picture	Deterioration of Neurologic Function
<20%	0	8 (80%)	2 (20%)
20–40%	1 (20%)	4 (80%)	0
>40%	0	7 (100%)	0

clonus, bilateral lower limb hyperreflexia, and patchy sensory loss around his left heel. Repeat cervical spine radiographs revealed C6–C7 bilateral facet dislocation. Seven weeks after injury, he underwent posterior C6 laminectomy followed by C5–C7 posterior spinal fusion using lateral mass instrumentation and iliac crest bone autograft. Intraoperative SEP amplitudes fell by <25% bilaterally. SEP monitoring was performed by stimulation of the ulnar and median nerves. Cortical SEPs were also measured using scalp electrodes. At conclusion of operation, the trace amplitudes were 8% and 4% above the initial control value on the left and right sides, respectively. Postoperatively, he had persistent right-sided weakness of elbow flexion and associated weakness of elbow extension, wrist flexion, finger abduction, and hip extension bilaterally. Seven months after the injury, his symptoms had considerably improved, but he still had residual bilateral hand weakness.

A 47-year-old woman sustained an L1 burst fracture and presented on admission with numbness of S3, S4, and S5 distribution on the right side. Six days after the injury, she underwent anterior L1 corpectomy, insertion of a titanium cage filled with bone autograft, followed by posterior T12–L2 instrumented fusion. Intraoperative spinal cord monitoring showed signal reduction of 61% on the right side and 63% on the left. SEP trace amplitude at the end of the operation had a persistent decrement of 55% on the left side and had completely recovered on the right. One week after surgery, she complained of numbness in the right S1–S2 distribution, which was confirmed by examination.

DISCUSSION

Electrophysiologic monitoring during spinal surgery continues to be an important modality with the aim to identify at an early stage potentially reversible sensory or motor injury. An ideal neurologic function monitoring technique should have high sensitivity and specificity, should produce real-time feedback, should not interfere with anesthetic methods allowing access to the patient for the anesthetist, should not impede the operative field, should not prolong excessively the surgery, and should have good reliability and reproducibility in patients with or without pre-existing neurologic impairment.[11]

Evoked potentials are essentially electrical fields that are generated by a nervous structure in response to a stimulus. A basic principle applied in evoked potential recording is that the area at risk for neurologic injury must lie in the pathway between the stimulus and the response. The tracts that can be monitored using the current neurophysiologic methods are the dorsal somatosensory columns and the lateral corticospinal motor tracts. Somatosensory tracts carry information from mostly large, myelinated fibers in the periphery that subserve position sense, vibration, and fine touch discrimination.[12] SEPs have been used with great success in predicting sensory abnormalities that may occur in patients who undergo corrective surgery for spine deformity and have normal preoperative neurologic findings.[13] Nash et al[14] pioneered the development of scalp-recorded cortical SEPs for electrophysiologic monitoring. Worldwide, this is the most widely used spinal cord recording technique. Shimoji et al[15] introduced the epidural recording of SEPs after the stimulation of peripheral nerves; this technique has given very promising results in serial reports.[2,6,15–17] The epidural method of monitoring SEPs provides more accurate results, as it records responses directly from the cord, and these responses are more resistant to anesthesia than cortical SEPs, which are amenable to signal amplitude depression and peak latency increase, particularly as the result of inhalation anesthetic agents, systemic hypotension, or hypothermia.[18,19] An additional reason why we preferred using epidural as opposed to cortical SEP recordings in most of the patients included in our study is their relative stability and speed of acquisition.[20–22] Epidural electrodes carry the potential risk of infection or cord trauma during placement; none of these complications was encountered in any of our patients.

SEPs have been used extensively in the neurophysiologic assessment of spinal cord integrity, reflecting mainly the function of the dorsal column. In patients with spinal trauma, however, the predictive value of SEPs in detecting potential for neurologic recovery after the injury has been controversial. York et al[23] studied the prognostic significance of cortical SEPs in patients with complete and incomplete spinal injuries; while the absence of recordable SEP was associated with no clinical recovery, the presence of an SEP was of lesser value in predicting the neurologic state at the time of examination or the possibility for recovery. McGarry et al[24] investigated scalp-recorded SEPs after peroneal stimulation in patients with long-standing incomplete spinal cord injuries; the

results could not be accurately correlated with the clinical functional outcome, and the authors concluded that this spinal monitoring technique does not reliably predict function after spinal cord trauma.

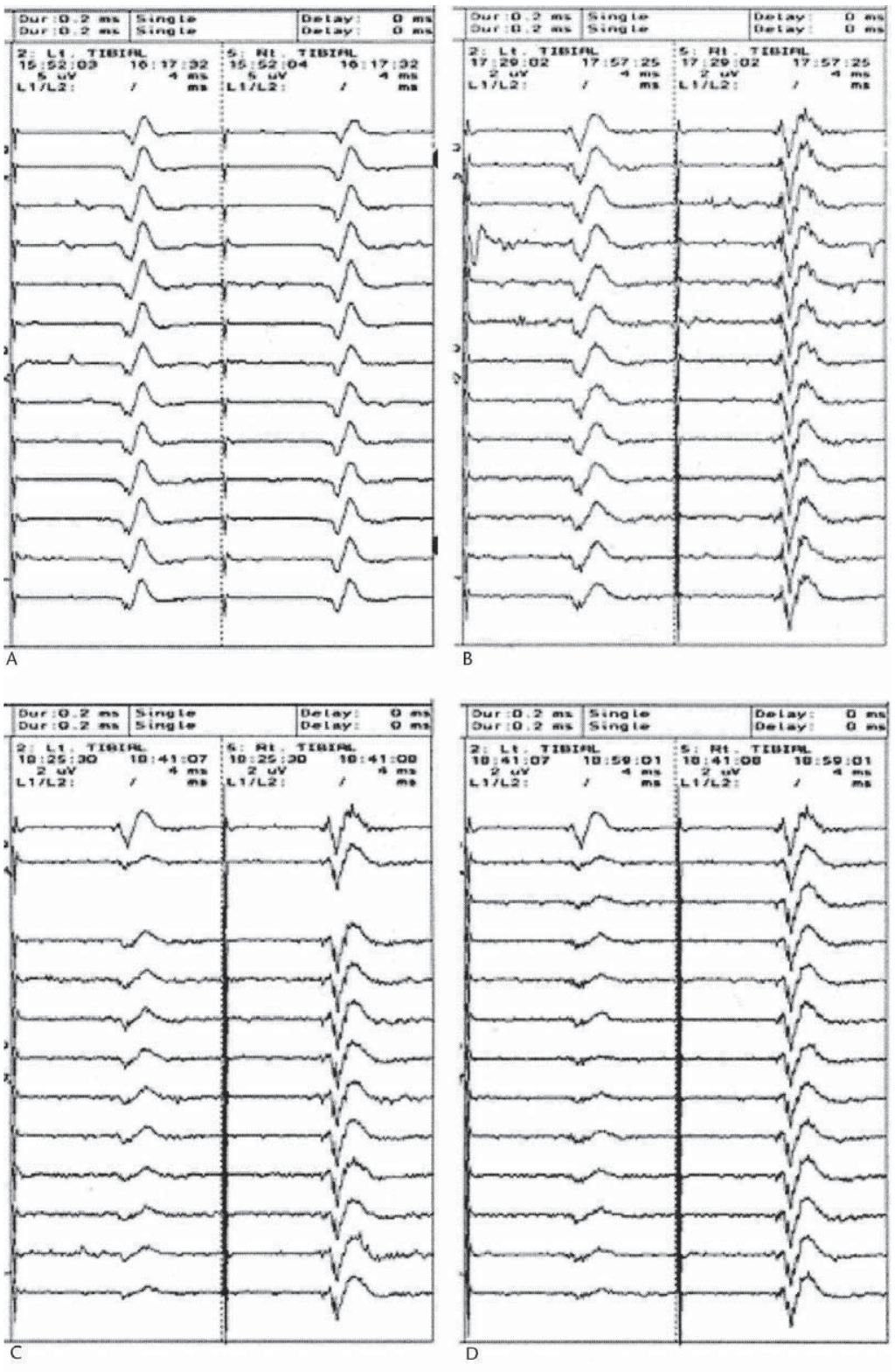
On the contrary, Rowed et al[25] suggested that when the spinal injury is incomplete, alterations in SEPs may be elicited from stimulation of a nerve entering the cord distal to the level of trauma; the recording of such potentials soon after injury or their premature return and progressive normalization of the waveform are sensitive early indications of favorable prognosis indicating recovery of posterior column function. Katz et al[26] examined the ability of SEPs and dermatomal-evoked potentials to predict motor return after acute spinal cord injury; they concluded that both the initial physical examination and the evoked potentials were reasonable predictors of further motor improvement. Curt and Dietz[27] monitored median and ulnar nerve SEPs in patients who sustained cervical spinal cord injury; they showed that SEP recording was valuable to indicate the level of injury, outline the degree of sensory impairment, and predict the outcome of hand function even in unconscious patients. Curt[28] suggested that the clinical examination of patients with spinal cord injury can be effectively supplemented by electrophysiologic techniques (SEPs, motor evoked potentials, and electroneuromyographic recordings) to assess the extent and severity of the injury, monitor the course of neurologic deficits, and predict functional outcome. Fehlings et al[29] showed in an experimental rat model of spinal cord injury that changes in axonal conduction in both motor and somatosensory tracts of the cord had a significant correlation to the extent of cord injury and could be accurately monitored by combined recording techniques of motor evoked potentials and SEPs, with the aim to assess posttraumatic functional integrity of the spinal cord.

Picozzi et al[30] recorded SEPs in patients with cervical spinal cord trauma at admission, after surgical treatment, and at follow-up; they suggested that the SEP monitoring method is well suited to the noninvasive study of spinal cord condition and the serial recording in patients with neural cord injury, with the presence of well preserved SEP traces in the posttraumatic period indicating the best prognosis for motor and sensory functions. Dorfman et al[31] monitored cerebral SEPs in patients with incomplete localized spinal cord lesions, including traumatic injuries, and demonstrated that the latency and amplitude characteristics of the signal recordings permitted quantitative evaluation of the functional status of the spinal somatosensory system. A study conducted by Pierlovisi-Lavaivre et al[32] evaluated cortical and cervical early SEP signals in patients with traumatic paraplegia or quadriplegia and found a good correlation between the presence of SEP, even of low amplitude or long latency, and a favorable prognosis. Consequently, the authors recommended investigation of SEPs soon after the injury and for a period of 6 months, with the aim to clearly establish the irreversibility of the lesion and the clinical recovery of posterior column function.[32] Cheliout-Heraut et al[33] assessed patients with severe cervical trauma to determine the efficacy of motor and sensory evoked potentials before and after surgical management; a good correlation was found between the severity of the spinal cord injury and SEP grading, whereas the combined electrophysiologic exploration of motor and sensory potentials proved to be the most useful tool for monitoring patients with significant neural cord damage. Delamarter et al[34] observed SEP trace amplitude improvement following immediate decompression in an animal model of experimentally induced spinal cord compression causing paraplegia.

To the authors' knowledge, the use of intraoperative spinal cord monitoring with epidural recording of SEP traces in cases of spinal trauma necessitating surgical intervention has not been previously investigated. In the current study, we used this different approach of intraoperative screening, compared with the reports mentioned above in which SEP was examined before and after the operative procedure, with the aim to determine the efficacy of SEP recording in predicting neurologic outcome after corrective surgery to address spinal injuries. The ultimate functional result after these reconstructive procedures is clearly correlated primarily to the extent of initial cord deficit but also to the surgical maneuvers that could potentially create further damage to an already compromised neural cord.

Recognizing the value of intraoperative SEP monitoring in scoliosis and other spinal surgery, we applied prospectively continuous SEP recording in 82 patients who sustained spinal injuries requiring surgical stabilization. In our series, intraoperative loss of SEP trace amplitude of >50% occurred frequently and was poorly predictive of postoperative neurologic deterioration. These attenuations in SEP trace, interpreted as false-positive results, are most likely the result of transient cord compromise, representing mild, reversible neurologic impairment during surgery with subclinical neurologic sequelae but without detectable clinical consequences. Szalay et al[4] correlated intraoperative SEP fall with surgical events and reported significant SEP waveform changes in 20% of patients, which were associated with these events but did not result in postsurgical neurologic deficits. We found that increasing the threshold for loss of signal from 50% to 60% reduced significantly the false-positive rate that reflects a tendency to false alarms, therefore improving specificity from 71% to 81% without compromising sensitivity. Moreover, the side of the SEP trace amplitude fall during the procedure always corresponded to the side in which a neurologic deficit was documented.

We presented the case of a patient with <50% decrement in SEP wave amplitude, which recovered during surgery, and who experienced postoperative neurologic complications; this case represents a false-negative result. The occurrence of such false-negative results is rare. Forbes et al[2] found no false-negative results when they reviewed 1168 patients who under-



went surgery for spinal deformity and were monitored with SEPs; Nuwer at al[3] reported false-negative rates of <0.2% in a large series of patients who underwent instrumented fusion for scoliosis. Jones et al[35] reported two cases of temporary quadriparesis following anterior cervical discectomy and instrumented fusion with normal perioperative SEP recordings, which represent false-negative results and a failure of SEPs to detect iatrogenic lesions during surgery. Our patient exhibited deterioration only in his motor function and may therefore correspond to a case where isolated injury to the motor tracts has occurred with sparing of sensory pathways allowing maintenance of normal SEPs. In this patient, SEP monitoring was performed by stimulation of the median and ulnar nerves and cervical cord injury was at the C6–C7 level, which could potentially have compromised conduction of sensory stimuli, reducing the sensitivity of SEP monitoring. Fisher et al[36] postulated that even though SEP recording may be effective in predicting and reducing the rate of complications, such neurologic sequelae still occur and are not reflected by preoperative SEP signal changes. These complications appear to be the product of partial neural cord lesions impairing primarily motor function and sparing the sensory pathways.[36] This is the reason why previous authors have supported the use of intraoperative monitoring by stimulating motor instead of sensory pathways, with the aim to increase accuracy and reproducibility of the method.[37–40]

In this study, nonrecovery of trace amplitude at conclusion of surgery was associated with significant risk of neurologic compromise. Most (86.3%) of the patients who had increased SEP trace amplitude before skin closure had an associated improved functional outcome after the operation. Most of the patients (91.6%) with >20% rise in signal amplitude before completion of the procedure demonstrated a better neurologic picture compared with their preoperative status. An intraoperative recovery in SEP wave amplitude of >40% compared with the initial control value was correlated with postsurgical improvement in neurologic state in all patients. However, in two cases, a <20% increase in trace amplitude was associated with a less favorable outcome. Moreover, the 47-year-old patient with the L1 burst fracture developed in the postoperative period neurologic symptoms on the right side, which was the side where the SEP trace amplitude had completely recovered in the end of surgery. We believe that these three cases could represent a degree of disparity in the reproducibility of the monitoring method.

A standardized anesthetic regimen was used in all cases with the aim to minimize anesthetic effects on intraoperative SEP signal changes, and no statistical difference could be detected between the different groups of patients in regard to systolic blood pressure and core temperature at completion of the operation.

Patients undergoing anterior procedures were particularly prone to loss of SEP trace amplitude of >50%. However, all the patients who required anterior procedures had impaired preoperative neurologic pictures. Epstein et al,[41] in a study of 100 patients undergoing cervical surgery for stenosis, spondylosis, and ossification of the posterior longitudinal ligament, also observed that SEP attenuation was common during anterior procedures but found little variation in frequency between anterior and posterior procedures. The increased incidence of SEP fall during anterior operations in our series may be related to the original neurologic compromise caused by the injury but may also reflect the surgical environment; therefore, it could be attributed to transient cord compression while manipulating vertebral fragments in close proximity to an edematous spinal cord. Ligation of the segmental vertebral vessels to facilitate the approach to the anterior surface of the vertebral body could also account for this difference. Another potential reason that could justify this discrepancy between the rate of SEP decrement during the anterior compared with the posterior procedures is displacement of the epidural electrode away from the spinal cord when anterior surgery was performed. However, in this patient group, when the epidural electrode was positioned percutaneously, it was tightly secured to the skin to minimize the risk of pulling off the neural cord.

CONCLUSIONS

Persistent intraoperative depression in SEP trace amplitude of >60% occurring in patients with normal preoperative neurology or incomplete injuries as well as limited (<20%) or absent recovery of signal amplitude at completion of surgery in patients with incomplete injuries were identified, in this study, as factors that increased considerably the risk of postoperative neurologic deficit. In patients with incomplete neurologic damage, a >20% restitution in the SEP wave amplitude from the initial baseline recording before conclusion of the procedure was positively correlated with superior prognosis in regard to spinal cord function. Patients with incomplete spinal cord damage who underwent anterior surgical procedures had greater incidence of significant somatosensory potential loss

FIGURE 3. The first SEP trace always indicates the baseline recording. A, The SEP traces during the posterior instrumented spinal fusion have been stable throughout. B, The baseline traces have been reset before the anterior stage of the combined procedure. The signals were stable during the first part of the anterior surgery. C, The SEP waveforms showed significant diminution on the left side and slight decrement on the right side. D, Recovery of the signal amplitude occurred on the right but not on the left side before conclusion of the anterior operative procedure, which was associated with the development of neurologic complications on the left lower extremity.

during the operation. In this series, continuous intraoperative SEP recording appeared to be adequately reproducible, sufficiently reliable, and therefore a practical tool in monitoring operative procedures for spinal trauma. Even though, compared with deformity surgery, the method is less sensitive and specific, it may help reduce the incidence of devastating neurologic injury during the operation on an already compromised neural cord and can provide a good prediction in terms of postoperative neurologic outcome. Thus, it could be considered a useful surgical adjunct in the management of patients with spinal trauma.

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Chapter 9

COMPLICATIONS and M&M

Advances in Surgical Techniques for EOSD

Paper: 29

Infections have been classified as 'never events' by the department of health (DoH) and NHS to improve patient experiences and surgical outcomes. Me being an active member of Scoliosis Research Society's (SRS) Mortality and Morbidity (M&M) committee and as a part of commitment to patient care took on the mammoth task of reporting the infection rates of 108,419 spinal surgeries from the SRS M&M database. The infection rates for children (younger than twenty one years) and adults (older than twenty one years) were analysed separately. The incidence of superficial, deep, and total infection rates were evaluated for all etiologies by logistic regression. The breakdown of infection rates for children is summarised in the table below (Table 1):

TABLE 1. Rate of Infection Based on Patient Age and Primary Diagnosis*

Primary Diagnosis	No. Cases	Superficial Infection (%)	Deep Infection (%)	Total Infection (%)
Pediatric (<21 yr)				
Degenerative disease	654	5 (0.8)	1 (0.2)	6 (0.9)
Scoliosis	20,424	195 (1.0)	341 (1.7)	536 (2.6)
Spondylolisthesis	827	9 (1.1)	12 (1.5)	21 (2.5)
Fracture	623	5 (0.8)	9 (1.4)	14 (2.2)
Kyphosis	1555	35 (2.3)	49 (3.2)	84 (5.4)
Other	1273	11 (0.9)	23 (1.8)	34 (2.7)
Primary tumor†	152	0 (0.0)	5 (3.3)	5 (3.3)
Implant revision/removal†	308	1 (0.3)	6 (1.9)	7 (2.3)
Not recorded	76	1 (1.3)	1 (1.3)	2 (2.6)
Total	25,432	261 (1.0)	436 (1.7)	697 (2.7)

*Age not recorded for 905 patients. These patients are not included in this table.

†Selected subgroup within the "other" category. Note that numbers of cases for these subgroups are part of the total other cases indicated.

Rates of infection after spine surgery based on 108,419 procedures: a report from the Scoliosis Research Society Morbidity and Mortality Committee: Smith JS, Shaffrey CI, Sansur CA, Berven SH, Fu KM, Broadstone PA, Choma TJ, Goytan MJ, Noordeen HH, Knapp DR Jr, Hart RA, Donaldson WF 3rd, Polly DW Jr, Perra JH, Boachie-Adjei O; Scoliosis Research Society Morbidity and Mortality Committee. Spine (Philadelphia, PA 1976). 2011 Apr 1; 36(7): 556-63. PMID: 20739916.

Advances in Surgical Techniques for EOSD

Paper: 30

Being an active member of SRS and part of its M&M committee, I retrospectively reviewed the database of 605 consecutive paediatric isthmic (542) and dysplastic (62) spondylolisthesis cases that were operated on between 2004 and 2007. The overall complication rate was 10.4 percent (63 out of 605). The incidence of a neurodeficit was five percent (31 out of 605). Deep wound infection was seen in twelve cases (2 percent), and a dural tear in eight patients (1.3 percent). The overall complication was double in patients who had attempted reduction, as compared to those who had fusion in-situ. This difference was also statistically significant *p=0.01* (8 out of 376 in unreduced vs. 23 out of 229 in reduced groups). There was no statistically significant difference in instrumented vs. uninstrumented group. Most of the neurodeficit was transient, and high grade spondylolisthesis (Myerding grading III and above) was more commonly associated with a new onset of a neurodeficit.

This study is the largest to date published in spinal surgical literature, and it discusses, very comprehensively, the role of reduction, instrumentation, and incidence of complications or infections in paediatric patients undergoing spondylolisthesis surgeries. This will help healthcare professionals to appropriately counsel parents and families in optimising expectations and outcomes.

Morbidity and mortality in the surgical treatment of six hundred five pediatric patients with isthmic or dysplastic spondylolisthesis: Fu KM, Smith JS, Polly DW Jr, Perra JH, Sansur CA, Berven SH, Broadstone PA, Choma TJ, Goytan MJ, Noordeen HH, Knapp DR Jr, Hart RA, Donaldson WF 3rd, Boachie-Adjei O, Shaffrey CI. Spine (Philadelphia, PA 1976). 2011 Feb 15; 36(4): 308-12. PMID: 20739916

Advances in Surgical Techniques for EOSD

Paper: 31

Deep vein thrombosis (DVT) and Pulmonary embolism (PE) could be fatal following spinal surgeries. Me being an active member of Scoliosis Research Society's (SRS) Mortality and Morbidity (M&M) committee and as a part of commitment to patient safety and minimize complication, I along with colleagues of the M&M committee evaluated 108,419 spinal surgeries from the SRS M&M database (2004-07) reporting the incidence of PE, deaths due to PE and incidence of DVT following three commonly performed spinal surgeries i.e. lumbar microdiscectomy (LMD), anterior cervical discectomy with fusion (ACDF) and decompression for lumbar canal stenosis (DLCS). Adults (≥21 years) and pediatric (≤21 years) patients were analysed separately. There were 25,432 pediatric cases and the etiology-wise break-up of the incidence of PE, deaths due to PE and DVT rate are in the table below:

Table 1. Thromboembolic Complications Reported for 25,432 Pediatric Spine Procedures From the Year 2004 to 2007*

	n	PE Rate (per 1000 Cases)	PE Deaths (per 1000 Cases)	DVT Rate (per 1000 Cases)
Pediatric (<21 yr old)				
Degenerative disease	20	0.00	0.00	0.00
Fracture	623	1.61	0.00	1.61
Kyphosis	1555	0.64	0.64	2.57
Spondylolisthesis	827	1.21	0.00	2.42
Scoliosis	20,424	0.39	0.05	0.15
Other	1983	0.00	0.00	0.00
Total pediatric cases	25,432	0.43	0.08	0.39

It is evident that the incidence of PE was highest in patients with fractures (1.61 per 1000). However the deaths due to PE was highest in the kyphosis group (0.64 deaths per 1000). The DVT rate was highest in kyphosis group followed by those undergoing surgery for spondylolisthesis correction.

Complication rates of three common spine procedures and rates of thromboembolism following spine surgery based on 108,419 procedures: a report from the Scoliosis Research Society Morbidity and Mortality Committee. Smith JS, Fu KM, Polly DW Jr, Sansur CA, Berven SH, Broadstone PA, Choma TJ, Goytan MJ, Noordeen HH, Knapp DR Jr, Hart RA, Donaldson WF 3rd, Perra JH, Boachie-Adjei O, Shaffrey CI. *Spine* (P'hila Pa 1976). 2010, Nov 15; 35(24): 2140-9. PMID: 20581760.

Advances in Surgical Techniques for EOSD

Paper: 32

Unintended durotomy (UD) during spinal surgery is associated with adverse outcomes viz. prolonged hospital stay, increased incidence of infection and onset of new neurological deficit. I evaluated 108,478 cases from SRS M&M (2004-07) along with other colleagues of M&M committee in an attempt to analyse factors associated with UD and report age and etiology-wise reliable incidence of durotomy rates from a large pool of patients. There were 19,953 pediatric patients (aged ≤20 years). The age group-wise break-up in the incidence of durotomy rates are summarised in Table 1 below.

Table 1: Incidence of Unintended Durotomy in Spine Surgery Based on 19,953 Paediatric Cases, Stratified by Patient Age Groups

Patient Age, y	Cases, n	Durotomies, n	Incidence of Durotomy, %
0-9	3791	43	1.1[a]
10-19	16162	282	1.7

The overall incidence of UD ranged from 1.1 to 1.9 percent. 0-9 yrs. age group had the lowest incidence of durotomy at 1.1 percent as compared to elderly patients who had twice the increased risk (i.e. 2.2 percent in those aged ≥80 yrs.). Etiology-wise, kyphosis and spondylolisthesis correction were associated with highest incidence of UD (1.9 percent). Other characteristics that were associated with high incidence of UD were revision surgeries and degenerative spinal deformities. There was a statistically significant association between UD and new onset of neurological deficit ($p<0.001$).

Incidence of unintended durotomy in spine surgery based on 108,478 cases. Williams BJ, Sansur CA, Smith JS, Berven SH, Broadstone PA, Choma TJ, Goytan MJ, Noordeen HH, Knapp DR Jr, Hart RA, Zeller RD, Donaldson WF 3rd, Polly DW Jr, Perra JH, Boachie-Adjei O, Shaffrey CI. *Neurosurgery* 2011, Jan; 68(1):117-23. PMID: 21150757

Advances in Surgical Techniques for EOSD

Paper: 33

Halo traction is commonly used in children with spinal deformity to make the rigid spine more flexible, and eliminate the need for combined anterior and posterior surgery at times. However, they are not without complications; the most common being pin track infections with loosening. Cranial nerve paresis is very uncommon and is rarely observed. The cranial nerve that is most vulnerable for paresis is the Abducens nerve (VI cranial nerve), owing to its longest intra-cranial course. I observed and reported two such cases in children who were treated with halo, and had both Trochlear (IV cranial nerve) and Abducens nerve (VI cranial nerve) palsies that recovered to a varying extent following the cessation of halo treatment.

Patient 1: A nine year old female with traumatic atlanto-axial dislocation and torticollis was noted to have diplopia with anisometropia. The halo was removed and treated in a hard collar. The VI cranial nerve recovered completely and the IV cranial nerve had only partial recovery.

Patient 2: A fifteen year old female with osteogeneis imperfecta (OI) with rigid scoliosis underwent a staged anterior and posterior surgery. She had halo traction following anterior release which resulted in IV and VI cranial nerve paresis. Complete resolution of IV and VI cranial nerve paresis was seen at two months post halo traction removal.

This was the first ever report or Trochlear nerve (IV cranial nerve) involvement with a halo traction. My team is credited for highlighting this rare complication. The mechanism of Trochlear nerve (IV cranial nerve) involvement and reasons for poor recovery despite removal of halo is unknown even to this day.

Fourth and sixth cranial nerve injury after halo traction in children: a report of two cases. Pinches E, Thompson D, Noordeen H, Liasis A, Nischal KK. J AAPOS. 2004 Dec; 8(6): 580-5. PMID: 15616508.

Spine

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DEFORMITY

Rates of Infection After Spine Surgery Based on 108,419 Procedures

A Report from the Scoliosis Research Society Morbidity and Mortality Committee

Justin S. Smith, MD, PhD,* Christopher I. Shaffrey, MD,* Charles A. Sansur, MD,† Sigurd H. Berven, MD,‡ Kai-Ming G. Fu, MD, PhD,* Paul A. Broadstone, MD,§ Theodore J. Choma, MD,¶ Michael J. Goytan, MD,|| Hilali H. Noordeen, MD,** Dennis R. Knapp, Jr., MD,†† Robert A. Hart, MD,‡‡ William F. Donaldson III, MD,§§ David W. Polly Jr, MD,|| || Joseph H. Perra, MD,*** Oheneba Boachie-Adjei, MD†††

Study Design: Retrospective review of a prospectively collected database.

Objective. Our objective was to assess the rates of postoperative wound infection associated with spine surgery.

Summary of Background Data. Although wound infection after spine surgery remains a common source of morbidity, estimates of its rates of occurrence remain relatively limited. The Scoliosis Research Society prospectively collects morbidity and mortality data from its members, including the occurrence of wound infection.

Methods. The Scoliosis Research Society morbidity and mortality database was queried for all reported spine surgery cases from 2004 to 2007. Cases were stratified based on factors including diagnosis, adult (≥21 years) *versus* pediatric (<21 years), primary *versus* revision, use of implants, and whether a minimally invasive approach was used. Superficial, deep, and total infection rates were calculated.

Results. In total, 108,419 cases were identified, with an overall total infection rate of 2.1% (superficial = 0.8%, deep = 1.3%). Based on primary diagnosis, total postoperative wound infection rate for adults ranged from 1.4% for degenerative disease to 4.2% for kyphosis. Postoperative wound infection rates for pediatric patients ranged from 0.9% for degenerative disease to 5.4% for kyphosis. Rate of infection was further stratified based on subtype of degenerative disease, type of scoliosis, and type of kyphosis for both adult and pediatric patients. Factors associated with increased rate of infection included revision surgery ($P < 0.001$), performance of spinal fusion ($P < 0.001$), and use of implants ($P < 0.001$). Compared with a traditional open approach, use of a minimally invasive approach was associated with a lower rate of infection for lumbar discectomy (0.4% *vs.* 1.1%; $P < 0.001$) and for transforaminal lumbar interbody fusion (1.3% *vs.* 2.9%; $P = 0.005$).

Conclusion. Our data suggest that postsurgical infection, even among skilled spine surgeons, is an inherent potential complication. These data provide general benchmarks of infection rates as a basis for ongoing efforts to improve safety of care.

Key words: spine surgery, complications, infection, instrumentation, pediatric, minimally invasive, deformity. **Spine 2011;36:556–563**

Despite substantial advancements in the surgical care of spine disease, postoperative wound infections remain a relatively common source of morbidity and increased costs.[1,2] Although several potential risk factors for the development of postoperative spinal wound infections have been suggested,[3–17] the etiology of the vast majority remains unclear.

The rate of wound infection after spine surgery is dependent on many factors, including the complexity of the procedure, health status of the patient, and potentially the experience and technique of the operating surgeon and facility related factors. This multifactorial nature of spinal wound infection complicates the ability to determine rates of wound infection that have generalizable, clinically meaningful applicability.

Documentation of the rates at which postoperative spine infections occur is important for several reasons, including preoperative patient counseling, quality improvement, and potentially medicolegal issues. The reported rate of postoperative spine infection ranges from 0.3% to 20%.[4–10,18–22] However, the

From the *Departments of Neurosurgery, University of Virginia Medical Center, Charlottesville, VA; †Department of Neurosurgery, University of Maryland Medical Center, Baltimore, MD; ‡Department of Orthopedic Surgery, University of California—San Francisco, San Francisco, CA; §Spine Surgery Associates, Chattanooga, TN; ¶Orthopedic Surgery, University of Missouri, Columbia, MO; || Health Sciences Centre, Winnipeg, Canada; **The Royal National Orthopedic Hospital and the Great Ormond Street Children's Hospital, London, UK; ††Arnold Palmer Children's Center, Orlando, FL; ‡‡ Department of Orthopedic Surgery, Oregon Health and Science University, Portland, OR; §§ Departments of Orthopedic Surgery, University of Pittsburgh Physicians, Pittsburgh, PA; || || Departments of Orthopedic Surgery and Neurosurgery, University of Minnesota, Minneapolis, MN; ***Twin Cities Spine Center, Minneapolis, MN; and †††Hospital for Special Surgery, New York, NY.

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No external funding was received for this project.

This project was submitted to the Hospital for Special Surgery (New York, NY) institutional review board and was determined to be exempt from institutional review board approval based on the use of deidentified data (institutional review board number 29045).

Address correspondence and reprint requests to Christopher I. Shaffrey, MD, Department of Neurosurgery, University of Virginia, PO Box 800212, Charlottesville, VA 22908; E-mail:CIS8Z@hscmail.mcc.virginia.edu.

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majority of these reports are based on relatively small patient populations, single surgeon or single institution experiences, or include a heterogeneous collection of spine procedures.

The Scoliosis Research Society (SRS) is a group of predominantly fellowship-trained spine surgeons and pediatric orthopedists dedicated to the study of spinal deformity. As part of its mission, the SRS collects surgical case data from its members, including morbidity and mortality (M&M). The resulting database consists of a consecutively collected series of cases, representing a broad range of procedures, from simple lumbar discectomy to complex deformity correction. In addition, these cases are submitted by spine surgeons with a broad range of experience and were performed at multiple institutions spanning the country and world. We have previously provided validation of this database for the assessment of complications associated with spine surgery.[23]

Our objectives in the present study were twofold. First, we sought to assess the rates of postoperative wound infection associated with spine surgery based on primary diagnosis, using cases submitted to the SRS M&M database over four consecutive years. Second, we stratified cases based on multiple factors, including subdiagnosis, patient age, spinal level, primary *versus* revision surgery, and surgical approaches, to determine the corresponding rates of infection and to assess for potential factors associated with the occurrence of infection.

MATERIALS AND METHODS

Patient Population

Before application for active membership in the SRS, surgeons must complete 5 years of candidate membership. International membership status is also available for surgeons outside of the United States. Candidate members are required to collect and submit data on all spine cases performed, including all associated M&M. Reported complications do not influence whether a candidate is offered membership, since the SRS membership committee is only provided with indication as to whether each candidate member has completed the required case submission process, not the number or types of complications for each candidate. Active members are also encouraged to submit their cases. Deidentified data are collected using a questionnaire developed by the SRS M&M committee in the 1990s and updated to a secure Internet-based data entry form in 2001. The SRS has invested substantial resources in this database and emphasizes to its membership the importance of accurate and consistent reporting. In addition, data submission includes a process in which members formally attest that submitted data are true and complete.

The SRS M&M database was queried for all cases reported from the years 2004 through 2007. Extracted data included patient age, diagnosis, whether the surgery was primary *versus* revision, whether a spinal fusion was performed and if so the type of fusion, whether implants were used, whether a minimally invasive approach was used, and whether a superficial or deep infection occurred after surgery. Patients were stratified based on age as either pediatric (younger than 21 years) or adult (aged 21 years or older).

This project was submitted to the Hospital for Special Surgery (New York, NY) institutional review board and was determined to be exempt from institutional review board approval based on use of deidentified data (institutional review board #29045).

Statistical Analyses

Frequency distributions and summary statistics were calculated for all clinical and demographic data. For categorical variables, cross-tabulations were generated and Fisher exact tests were used to compare distributions. All statistical analyses were two sided. $P < 0.05$ was considered significant.

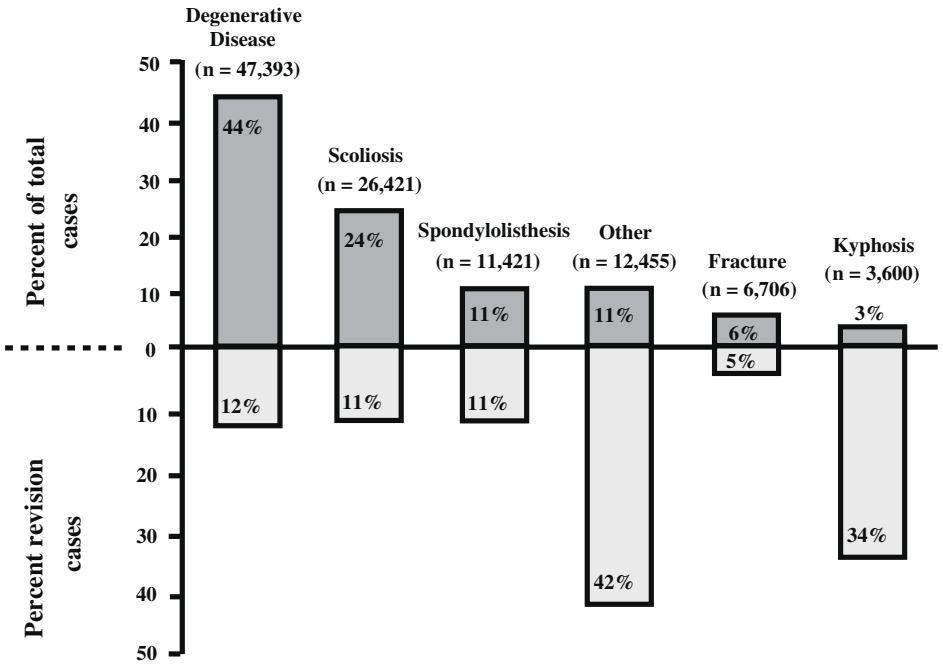

Figure 1. Distribution of cases based on primary diagnosis and percentage of revision cases. The top half of the plot shows the percentage of cases represented by each primary diagnosis, The 423 cases for which primary diagnosis was not reported are not included. The bottom half of the plot shows the percentage of cases within each primary diagnosis that were revison cases.

RESULTS

A total of 108,419 surgical cases were reported to the SRS M&M database from 2004 to 2007. The mean patient age was 47 years (median: 48, range: 1 month–97 years), with 76% of cases performed in adult and 24% in pediatric patients. The distribution of cases by primary diagnosis and the percentage of revision cases within each diagnosis are shown in Figure 1. Procedures using a minimally invasive approach comprised 13% of cases. Of the total cases, spinal fusion was performed in 67%. Active members contributed the majority of cases (71%), followed by candidate members (23%) and international members (6%).

The overall rates of superficial and deep infection were 0.8% and 1.3%, respectively. Superficial and deep infections occurred in 0.8% and 1.2% of adult patients (n = 82,082), respectively, and in 1.0% and 1.7% of pediatric patients (n = 25,432), respectively (Table 1). Based on primary diagnosis, the rate of infection ranged from 1.4% for degenerative disease to 4.2% for kyphosis among adults and ranged from 0.9% for degenerative disease to 5.4% for kyphosis among pediatric patients (Table 1). Cases reported as pediatric degenerative disease predominantly consisted of disc herniations, stenosis, or degenerative disc disease, with the vast majority involving the lumbar region. Notably, metastatic tumor and acute osteodiscitis, two subgroups within the "other" category for adults, each had infection rates of approximately 5%.

The overall rate of infection among adult patients with a primary diagnosis of degenerative disease varied based on spinal location, with the highest rate for thoracic procedures (2.1%), followed by lumbar (1.6%) and cervical (0.8%) procedures (Table 2). Within each spinal location, the rate of infection varied based on the diagnosis. For each spinal location, the rate of infection was lowest for disc herniations, ranging from 0.5% for cervical to 1.7% for thoracic herniations. The

TABLE 1. Rate of Infection Based on Patient Age and Primary Diagnosis*

Primary Diagnosis	No. Cases	Superficial Infection (%)	Deep Infection (%)	Total Infection (%)
Adult (≥21 yr)				
Degenerative disease	46,434	308 (0.7)	342 (0.7)	650 (1.4)
Scoliosis	5801	66 (1.1)	146 (2.5)	212 (3.7)
Spondylolisthesis	10,529	93 (0.9)	130 (1.2)	223 (2.1)
Fracture	6025	31 (0.5)	92 (1.5)	123 (2.0)
Kyphosis	2012	28 (1.4)	57 (2.8)	85 (4.2)
Other	11,089	113 (1.0)	237 (2.1)	350 (3.2)
Primary Tumor†	404	4 (1.0)	7 (1.7)	11 (2.7)
Metastatic Tumor†	726	9 (1.2)	27 (3.7)	36 (5.0)
Acute osteo/discitis†	866	12 (1.4)	32 (3.7)	44 (5.1)
Implant revision/removal†	1720	16 (0.9)	39 (2.3)	55 (3.2)
Not recorded	192	2 (1.0)	2 (1.0)	4 (2.1)
Total	82,082	641 (0.8)	1006 (1.2)	1647 (2.0)
Pediatric (<21 yr)				
Degenerative disease	654	5 (0.8)	1 (0.2)	6 (0.9)
Scoliosis	20,424	195 (1.0)	341 (1.7)	536 (2.6)
Spondylolisthesis	827	9 (1.1)	12 (1.5)	21 (2.5)
Fracture	623	5 (0.8)	9 (1.4)	14 (2.2)
Kyphosis	1555	35 (2.3)	49 (3.2)	84 (5.4)
Other	1273	11 (0.9)	23 (1.8)	34 (2.7)
Primary tumor†	152	0 (0.0)	5 (3.3)	5 (3.3)
Implant revision/removal†	308	1 (0.3)	6 (1.9)	7 (2.3)
Not recorded	76	1 (1.3)	1 (1.3)	2 (2.6)
Total	25,432	261 (1.0)	436 (1.7)	697 (2.7)

*Age not recorded for 905 patients. These patients are not included in this table.

†Selected subgroup within the "other" category. Note that numbers of cases for these subgroups are part of the total other cases indicated.

TABLE 2. Rate of infection among adult patients with a primary diagnosis of degenerative disease, stratified based on spinal location and subtype of disease

Spinal Region/Diagnosis	No. Cases	Superficial Infection (%)	Deep Infection (%)	Total Infection (%)
Cervical				
Postlaminectomy syndrome	35	1 (2.9)	1 (2.9)	2 (5.7)
Spinal stenosis	3466	13 (0.4)	27 (0.8)	40 (1.2)
Spondylotic radiculopathy	1859	9 (0.5)	5 (0.3)	14 (0.8)
Degenerative disc disease	1769	5 (0.3)	7 (0.4)	12 (0.7)
Disc herniation	4472	11 (0.2)	12 (0.3)	23 (0.5)
Not recorded	73	0 (0.0)	0 (0.0)	0 (0.0)
Total	11,674	39 (0.3)	52 (0.4)	91 (0.8)
Thoracic				
Degenerative disc disease	95	0 (0.0)	3 (3.2)	3 (3.2)
Spinal stenosis	161	2 (1.2)	2 (1.2)	4 (2.5)
Disc herniation	235	3 (1.3)	1 (0.4)	4 (1.7)
Spondylotic radiculopathy	17	0 (0.0)	0 (0.0)	0 (0.0)
Not recorded	16	0 (0.0)	0 (0.0)	0 (0.0)
Total	524	5 (1.0)	6 (1.1)	11 (2.1)
Lumbar				
Spondylotic radiculopathy	949	22 (2.3)	14 (1.5)	36 (3.8)
Spinal stenosis	12,270	116 (0.9)	153 (1.2)	269 (2.2)
Postlaminectomy syndrome	573	6 (1.0)	4 (0.7)	10 (1.7)
Degenerative disc disease	7213	55 (0.8)	58 (0.8)	113 (1.6)
Disc herniation	12,694	62 (0.5)	51 (0.4)	113 (0.9)
Not recorded	211	2 (0.9)	2 (0.9)	4 (1.9)
Total	33,910	263 (0.8)	282 (0.8)	545 (1.6)
Spinal location not recorded	326	1 (0.3)	2 (0.6)	3 (0.9)

highest rate of infection among cases performed for cervical degenerative disease was associated with a diagnosis of postlaminectomy syndrome (5.7%). However, this estimate should be interpreted with caution, since only 35 cases and two infections were reported for this category (Table 2).

Scoliosis cases were further stratified by subtype and age group (Table 3). The rate of infection was higher for scoliosis cases than degenerative disease cases, and the proportion of deep wound infections was considerably greater. Among adult patients, the rate of infection ranged from 2.8% for idiopathic to 8.9% for neuromuscular scoliosis. The most common subgroup, degenerative scoliosis, had an intermediate rate of infection of 4.1%. Among pediatric cases, the most common subtype, idiopathic scoliosis, had the lowest rate of infection (1.4%). Although cases of posttraumatic scoliosis had relatively high rates of infection for both adult and pediatric patients, the numbers of cases reported are limited.

Kyphosis cases were also stratified based on the etiology of the deformity (Table 4). Postlaminectomy kyphosis was associated with the highest rate of infection among adults (5.1%). A wide range of infection rates was observed for pediatric kyphosis, ranging from 1.5% for congenital to 14% for neuromuscular kyphosis (Table 4).

Multiple surgical factors were assessed for an association with the occurrence of infection (Table 5). Cases that included spinal fusion had a 33% higher rate of infection (2.4% *vs.* 1.8%, $P < 0.001$). Among cases with spinal fusion, the rate of infection varied based on the type of fusion. Compared with the overall rate of infection associated with fusion cases, the rate of infection for anterior-only cases (0.6%) was significantly lower ($P < 0.001$), and the rates of infection for combined anterior-posterior fusions (3.2%), posterolateral-only fusions (3.0%), and interlaminar/facet-only fusions (2.8%) were significantly higher ($P < 0.001$, $P < 0.001$, and $P = 0.003$, respectively) (Table 5). In addition, the overall infection rate for cases with implants was 28% higher than the rate for cases without implants (2.3% *vs.* 1.8%, $P < 0.001$).

TABLE 3. Rate of Infection Among Patients with a Primary Diagnosis of Scoliosis, Stratified Based on Patient Age and Subtype of Scoliosis

Age Group/Type of Scoliosis	No. of Cases	Superficial Infection (%)	Deep Infection (%)	Total Infection (%)
Adult (≥21 yr)				
Neuromuscular	292	8 (2.7)	18 (6.2)	26 (8.9)
Posttraumatic	30	0 (0.0)	2 (6.7)	2 (6.7)
Degenerative	2533	31 (1.2)	73 (2.9)	104 (4.1)
Congenital	137	1 (0.7)	4 (2.9)	5 (3.6)
Idiopathic	2488	23 (0.9)	46 (1.8)	69 (2.8)
Other	139	3 (2.2)	2 (1.4)	5 (3.6)
Not recorded	182	0 (0.0)	1 (0.5)	1 (0.5)
Total	5801	66 (1.1)	146 (2.5)	212 (3.7)
Pediatric (<21 yr)				
Posttraumatic	35	1 (2.9)	1 (2.9)	2 (5.7)
Neuromuscular	4855	83 (1.7)	184 (3.8)	267 (5.5)
Congenital	2045	27 (1.3)	18 (0.9)	45 (2.2)
Idiopathic	11,741	65 (0.6)	100 (0.9)	165 (1.4)
Other	1464	16 (1.1)	31 (2.1)	47 (3.2)
Not recorded	284	3 (1.1)	7 (2.5)	10 (3.5)
Total	20,424	195 (1.0)	341 (1.7)	536 (2.6)

TABLE 4. Rate of Infection Among Patients with a Primary Diagnosis of Kyphosis, Stratified Based on Patient Age and Subtype of Kyphosis

Age Group/Type of Kyphosis	No. Cases	Superficial Infection (%)	Deep Infection (%)	Total Infection (%)
Adult (≥21 yr)				
Postlaminectomy	198	5 (2.5)	5 (2.5)	10 (5.1)
Osteoporosis	104	3 (2.9)	2 (1.9)	5 (4.8)
Scheuermann	227	2 (0.9)	8 (3.5)	10 (4.4)
Posttraumatic	365	3 (0.8)	12 (3.3)	15 (4.1)
Fixed sagittal plane deformity	553	6 (1.1)	15 (2.7)	21 (3.8)
Neuromuscular	42	0 (0.0)	1 (2.4)	1 (2.4)
Congenital	48	0 (0.0)	1 (2.1)	1 (2.1)
Other	396	9 (2.3)	13 (3.3)	22 (5.6)
Not recorded	79	0 (0.0)	0 (0.0)	0 (0.0)
Total	2012	28 (1.4)	57 (2.8)	85 (4.2)
Pediatric (<21 yr)				
Neuromuscular	264	10 (3.8)	27 (10.2)	37 (14.0)
Fixed sagittal plane deformity	18	1 (5.6)	0 (0.0)	1 (5.6)
Postlaminectomy	64	3 (4.7)	0 (0.0)	3 (4.7)
Scheuermann	634	12 (1.9)	15 (2.4)	27 (4.3)
Posttraumatic	31	1 (3.2)	0 (0.0)	1 (3.2)
Congenital	329	3 (0.9)	2 (0.6)	5 (1.5)
Other	200	4 (2.0)	5 (2.5)	9 (4.5)
Not recorded	15	1 (6.7)	0 (0.0)	1 (6.7)
Total	1555	35 (2.3)	49 (3.2)	84 (5.4)

TABLE 5. Summary of Surgical Characteristics Assessed for Association with Infection

Parameter	No. Cases	Superficial Infection (%)	Deep Infection (%)	Total Infection (%)	*P**
Spinal fusion performed					
Yes	72,534	642 (0.9)	1072 (1.5)	1714 (2.4)	<0.001
No	35,877	267 (0.7)	377 (1.1)	644 (1.8)	
Not recorded	8	0 (0.0)	0 (0.0)	0 (0.0)	
Type of fusion					
Post-ant-post	271	2 (0.7)	7 (2.6)	9 (3.3)	
Anterior-posterior	7887	91 (1.2)	161 (2.0)	252 (3.2)	<0.001†
Posterolateral	19,710	221 (1.1)	379 (1.9)	600 (3.0)	<0.001†
Interlaminar/facet	16,192	167 (1.0)	281 (1.7)	448 (2.8)	0.003†
All fusion cases	72,534	642 (0.9)	1072 (1.5)	1714 (2.4)	
TLIF/PLIF	12,267	104 (0.8)	182 (1.5)	286 (2.3)	
Anterior only	15,336	46 (0.3)	48 (0.3)	94 (0.6)	<0.001†
Not recorded	871	10 (1.1)	14 (1.6)	24 (2.8)	
Implants					
Yes	74,114	647 (0.9)	1092 (1.5)	1739 (2.3)	<0.001
No	34,305	262 (0.8)	357 (1.0)	619 (1.8)	
Revision surgery					
Yes	16,503	183 (1.1)	359 (2.2)	542 (3.3)	<0.001
No	91,916	726 (0.8)	1090 (1.2)	1816 (2.0)	
Minimally invasive approach					
Yes	14,301	43 (0.3)	35 (0.2)	78 (0.5)	<0.001
No	94,115	866 (0.9)	1414 (1.5)	2280 (2.4)	
Not recorded	3	0 (0.0)	0 (0.0)	0 (0.0)	

**P-values are based on statistical comparisons of total infection rates.*

†Anterior-posterior, posterolateral, and interlaminar/facet fusions had significantly higher and anterior only had significantly lower rates of infection compared with the overall rate of infection for all fusion patients (2.4%).

TLIF indicates transforaminal lumbar interbody fusion; PLIF, posterior lumbar interbody fusion.

Revision cases had a 65% higher overall rate of infection compared with primary cases (3.3% *vs.* 2.0%, $P < 0.001$) (Table 5). This difference was more evident for deep wound infections (2.2% *vs.* 1.2%) compared with superficial wound infections (1.1% *vs.* 0.8%).

The rate of wound infection, especially deep wound infection, was significantly lower for cases performed using a minimally invasive approach compared with those using more traditional open approaches (0.5% *vs.* 2.4%, $P < 0.001$) (Table 5). Two specific procedures that are frequently performed minimally invasively were further assessed. The overall rate of infection associated with minimally invasive transforaminal lumbar interbody fusion (TLIF) (n = 848) was significantly less than that of the traditional open approach (n = 6,241; 1.3% *vs.* 2.9%; $P = 0.005$; Figure 2). Notably, this difference was almost entirely accounted for by the marked differences in the rate of deep wound infection (0.4% *vs.* 1.9%,

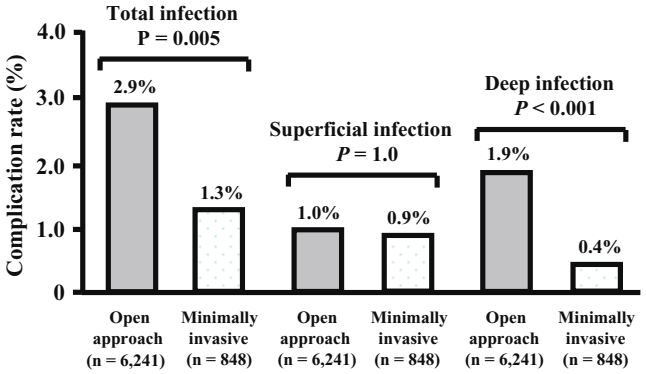

Figure 2. Rates of infection for transformaminal lumbar interbody fusions (TLIF) performed using a traditional open approach compared with a minimally invasive approach. The minimally invasive approach is associated with a significantly lower overall rate of infection and rate of deep wound infection.

$P < 0.001$; Figure 2). The overall rate of infection for lumbar discectomy was also significantly lower for cases performed minimally invasively compared with those performed using traditional open techniques (0.4% *vs.* 1.1%). This difference was also primarily due to the differences in the rate of deep wound infection (0.08% *vs.* 0.5%, $P < 0.001$).

DISCUSSION

This study provides the rates of postoperative wound infection after a broad range of spinal procedures, based on cases performed predominantly by fellowship-trained spine surgeons. The large number of cases enabled assessment of infection rates for relatively uncommon procedures, including those performed on pediatric patients. The SRS M&M database also enabled stratification of cases and assessment of corresponding infection rates based on operative factors, including primary *versus* revision status, use of implants, fusion approach, and whether minimally invasive techniques were used. In addition, the database distinguished between superficial and deep infections, allowing separate reporting of these complications.

The overall superficial and deep wound infection rates for the present series of 108,419 patients were 0.8% and 1.2%, respectively. The 2% total rate of infection in this series is comparable to previously reported series that include a diverse representation of spine procedures, in which the rate of infection ranges from 0.9% to 4.4%.[9,15,24–26] Prior reports have documented the rates of infection for select spinal procedures, and the rates in the present study are generally comparable to these prior reports. For example, the rate of postoperative infection for lumbar discectomy is 0.9% in the present series, which is comparable to recent prior reports in which the rate ranges from 0% to 1.6%.[27–31] As another example, the rate of wound infection after surgery for adolescent idiopathic scoliosis has been previously reported to range from 0.7% to 3%,[21,32] which is comparable to the present series in which the rate is 1.4%. As a third example, the rate of wound infection after surgery for degenerative scoliosis has been reported to be 4.3%,[33] which is comparable to the 4.1% rate in the present series.

Although for some of the more common procedures, the infection rates reported herein are comparable to those previously reported, for the vast majority of the other diagnoses and subdiagnoses presented, the literature provides very limited or no comparison. That some of the common procedures in the present series have comparable infection rates as in the literature, in our view, adds to the credibility of the rates of infection that are reported herein for many of the other diagnoses and subdiagnoses for which the literature fails to provide comparison.

Based on the SRS M&M database, the rates of postoperative wound infections are significantly higher for cases that included fusion or implants than those that did not. It is important to recognize that these data do not necessarily suggest a causation link between infection and performance of fusion or inclusion of implants, but rather likely reflect greater complexity and associated risk for cases that require fusion or the use of implants.

The overall infection rate for procedures performed using a minimally invasive approach was significantly less compared with those not doing so. Importantly, many of the procedures that are commonly performed using a minimally invasive approach, such as lumbar discectomy or TLIF, generally have relatively low infection rates, while more complex procedures that were typically only performed using a traditional open approach, such as degenerative scoliosis or neuromuscular kyphosis, generally have substantially higher infection rates. Specific assessment of the infection rates for lumbar discectomy and TLIF was possible however, and did demonstrate that these minimally invasive approaches were associated with significantly lower rates of postoperative wound infection, especially deep wound infection, compared with traditional open approaches. This putative benefit of the minimally invasive approach, although often suggested, has not been previously well demonstrated.

The present study has several strengths, most notably the large number of cases that includes representation of both low- and high-complexity procedures. The contributors, although predominantly fellowship-trained spine surgeons, represent a broad range of experience levels, which enhances the generalizability of the data. Cases were submitted from multiple institutions, which helps to mitigate the effects that specific institutional factors and patient populations may have on the occurrence of infection. In addition, the database enables separate assessment of superficial and deep infection rates and stratifies cases based on patient age and primary *versus* revision status, allowing separate assessment of infection rates based on these parameters.

The present study also has limitations. Although the data were collected prospectively, the study design and analysis were performed retrospectively. There are no methods to determine completeness of data submission, nor the accuracy of the reporting. It is dependent upon the efforts of the participants. The database does not provide documentation of several factors that may be related to the occurrence of wound infection, including whether prophylactic antibiotics were administered, length of operative time, estimated blood loss, number of surgeons involved in the procedure, length of hospitalization, or patient comorbidities. In addition, there is no documentation of causative organisms, how the infection was managed, or outcomes. It is also not possible to assess for the association of infection occurrence based on the specific surgeon or institution.

CONCLUSIONS

Based on 108,419 spine surgery cases from the SRS M&M database, the overall rates of postoperative superficial and deep wound infections are 0.8% and 1.3%, respectively. For both adult and pediatric patients, the rates of postoperative wound infection were lowest for procedures performed for degenerative spine disease and highest for procedures performed for spine deformity. Procedures with spinal fusion or implants had a significantly higher rate of postoperative wound infection, likely reflective of the greater complexity and associated risk of cases that require fusion or the use of implants. Compared with a traditional open approach, use of a minimally invasive approach was associated with a lower rate of infection for lumbar discectomy and for TLIF.

➢ Key Points

- ❑ Based on 108,419 spine surgery cases from the Scoliosis Research Society Morbidity and Mortality database, the overall rates of postoperative superficial and deep wound infections are 0.8% and 1.3%, respectively.
- ❑ For both adult and pediatric patients, the rates of postoperative wound infection were lowest for procedures performed for degenerative spine disease and highest for procedures performed for spinal deformity.
- ❑ Revision cases had a significantly higher rate of infection compared with primary cases (3.3% *vs.* 2.0%; $P < .001$).
- ❑ Procedures with spinal fusion or implants had a significantly higher rate of postoperative wound infection, likely reflective of the greater complexity and associated risk of cases that require fusion or the use of implants.
- ❑ Compared with a traditional open approach, use of a minimally invasive approach was associated with a lower rate of infection for lumbar discectomy (0.4% *vs.* 1.1%; $P < 0.001$) and for transforaminal lumbar interbody fusion (1.3% *vs.* 2.9%; $P = 0.005$).

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CLINICAL CASE SERIES

Morbidity and Mortality in the Surgical Treatment of Six Hundred Five Pediatric Patients With Isthmic or Dysplastic Spondylolisthesis

Kai-Ming G. Fu, MD, PhD,* Justin S. Smith, MD, PhD,* David W. Polly, Jr., MD,† Joseph H. Perra, MD,‡ Charles A. Sansur, MD,§ Sigurd H. Berven, MD,¶ Paul A. Broadstone, MD,|| Theodore J. Choma, MD,** Michael J. Goytan, MD,†† Hilali H. Noordeen, MD,‡‡§§ D. Raymond Knapp, Jr., MD,¶¶ Robert A. Hart, MD,|||| William F. Donaldson III, MD,*** Oheneba Boachie-Adjei, MD,††† and Christopher I. Shaffrey, MD*

Study Design. Retrospective analysis of prospectively collected database.

Objective. To analyze the rate of complications, including neurologic deficits, associated with operative treatment of pediatric isthmic and dysplastic spondylolisthesis.

Summary of Background Data. Pediatric isthmic and dysplastic spondylolisthesis are relatively uncommon disorders. Several prior studies have suggested a high rate of complication associated with operative intervention. However, most of these studies were performed with sufficiently small sample sizes such that the presence of one complication could significantly affect the overall rate. The Scoliosis Research Society (SRS) prospectively collects morbidity and mortality (M&M) data from its members. This multicentered, multisurgeon database permits analysis of the surgical treatment of this relatively rare condition on an aggregate scale and provides surgeons with useful information for preoperative counseling.

Methods. Patients who underwent surgical treatment for isthmic or dysplastic spondylolisthesis from 2004 to 2007 were identified from the SRS M&M database. Inclusion criteria for analysis included age ≤21 and a primary diagnosis of isthmic or dysplastic spondylolisthesis.

Results. Of 25,432 pediatric cases reported, there were a total of 605 (2.4%) cases of pediatric dysplastic (n = 62, 10%) and isthmic (n = 543, 90%) spondylolisthesis, with a mean age of 15 years (range, 4-21). Approximately 50% presented with neural element compression, and less than 1% of cases were revisions. Surgical procedures included fusions in 92%, osteotomies in 39%, and reductions in 38%. The overall complication rate was 10.4%. The most common complications included postoperative neurologic deficit (n = 31, 5%), dural tear (n = 8, 1.3%), and wound infection (n = 12, 2%). Perioperative deep venous thrombosis and pulmonary embolus were reported in 2 (0.3%) and 1 (0.2%) patients, respectively. There were no deaths in this series.

Conclusion. Pediatric isthmic and dysplastic spondylolisthesis are relatively uncommon disorders, representing only 2.4% of pediatric spine procedures in the present study. Even among experienced spine surgeons, surgical treatment of these spinal conditions is associated with a relatively high morbidity.

Key words: pediatric, isthmic, dysplastic, spondylolisthesis, reduction, morbidity. **Spine 2011;36:308–312**

Isthmic and dysplastic spondylolisthesis are the result of pars defects and dysplastic facets, respectively. Pars abnormalities are present in a sizable proportion of the pediatric population, with reports suggesting that 4.4% of children at age 6 have either unilateral or bilateral pars defects.[1] The natural history of most low grade cases is benign, with long-term observation reporting little radiographic or symptomatic progression.[1,2] Given this natural history, operative intervention for this condition remains a small proportion of most spinal surgeons' practice. However, when such operations need to be performed, *i.e.*, for high grade symptomatic patients, they can result in significant morbidity. Complications

From the *Department of Neurosurgery, University of Virginia Medical Center, Charlottesville, VA; †Departments of Orthopedic Surgery and Neurosurgery, University of Minnesota, Minneapolis, MN; ‡Twin Cities Spine Center, Minneapolis, MN; §Department of Neurosurgery, University of Maryland Medical Center, Baltimore, MD; ¶Department of Orthopedic Surgery, University of California—San Francisco, San Francisco, CA; ||Spine Surgery Associates, Chattanooga, TN; **Department of Orthopedic Surgery, University of Missouri; Columbia, MO; ††Health Sciences Centre, Winnipeg, Canada; ‡‡The Royal National Orthopedic Hospital, London, United Kingdom; §§The Great Ormond Street Children's Hospital, London, United Kingdom; ¶¶Arnold Palmer Children's Hospital, Orlando, FL; ||||Department of Orthopedic Surgery, Oregon Health and Science University, Portland, OR; ***Department of Orthopedic Surgery, University of Pittsburgh Physicians, Pittsburgh, PA; and †††Department of Orthopedic Surgery, Hospital for Special Surgery, New York, NY.

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Address correspondence and reprint requests to Christopher I. Shaffrey, MD, Department of Neurosurgery, University of Virginia, PO Box 800212, Charlottesville, VA 22908; E-mail: CIS8Z@hscmail. mcc.virginia.edu

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ranging from wound infections to neurologic deficits have been reported to occur frequently in both historic and modern series.[3–16] Reduction procedures appear to have a higher complication rate, with some reports indicating a higher incidence of neurologic complications with reduction in the pediatric population.[6,7,9,12,17] However, the case numbers reported in these series are small, usually less than 50, and most often only 10 to 15, likely due to the infrequency of surgical treatment of this condition. To obtain a larger population from which to ascertain benchmark complication rates, we queried the multisurgeon, multicenter Scoliosis Research Society (SRS) Morbidity and Mortality (M&M) database.

MATERIALS AND METHODS

Before application for active membership in the SRS, surgeons must complete a 5-year period of candidate membership. International membership status is also available for surgeons outside of the United States. Candidate members are required to prospectively collect and submit data on all spine cases performed, including all associated morbidity and mortality. Active and international members are also encouraged to submit their cases.[18] Data are gathered using a questionnaire that was developed by the SRS M&M Committee in the early 1990s and updated to a secure internet-based data entry form in 2001. The SRS has invested substantial resources in this database and emphasizes to its membership the importance of accurate and consistent reporting. All data are deidentified on entry into the database. This project was submitted to the Hospital for Special Surgery (New York, NY) Institutional Review Board (IRB) and was determined to be exempt from IRB approval based on the use of deidentified data (IRB number 29045).

In order to assess the incidence of complications associated with the surgical treatment of pediatric isthmic and dysplastic spondylolisthesis, all reported surgical cases with these diagnoses from 2004 through 2007 were extracted from the SRS database. Inclusion criteria for this study were age $\leq$21 and a primary diagnosis of isthmic or dysplastic spondylolisthesis. Excluded were patients who received operations for other indications, such as concomitant burst fracture.

For each case included in the present study, the following data were extracted: patient age, diagnosis, presence of preoperative neural compression, procedure type (reduction, osteotomy, instrumentation), electrophysiological monitoring parameters, and presence or absence of complications and/or mortality. Complications were additionally subclassified (*e.g.,* pulmonary embolus, deep venous thrombosis, *etc.*). Occurrence of, extent of (spinal cord, *cauda equina,* or nerve root), and recovery from (none, partial, or complete) new neurologic deficits were also available and extracted. Not consistently included in the database were information on spondylolisthesis grade, ASA grade, success of reduction, long-term follow-up, and objective outcome measures. Therefore, with the exception of Meyerding Grade for patients from 2007, analyses related to these parameters were not performed in the present study. Although presence of preoperative neural element compression was not specifically collected as part of the SRS M&M database reporting process, surgeons did report whether a direct decompression of the neural elements was performed as part of the surgical procedure. In the present study, the reported performance of a direct decompression was used as a surrogate for the presence of preoperative neural element compression.

Statistical analysis was performed using SPSS software (SPSS Inc., Chicago, IL). Statistical comparisons between subgroups were performed using Fisher exact test. A $P < 0.05$ was considered statistically significant. Comparisons of complication occurrence, neurologic deficit occurrence, preoperative neurologic element compression, and specific complications were made between those who underwent reduction procedures *versus* those who underwent *in situ* fusion procedures. Comparison of the presence of preoperative neurologic element compression with the occurrence of new neurologic deficits was also performed. In addition, comparisons of age and electrophysiological monitoring status with complications or new neurologic deficits were also performed.

RESULTS

Of the 25,432 pediatric spine cases reported during the 4 years from 2004 to 2007, 605 cases of isthmic or dysplastic spondylolisthesis were identified, representing 2.4% of the total. Of the 605 cases reported in the database, 63 were reported to have complications, resulting in a 10.4% overall complication rate. No deaths were reported. The most common complications are listed in Table 1 in both the aggregate and by the presence or absence of a reduction procedure. By far, the most common complication was a new postoperative neurologic deficit, occurring in 31 cases and accounting for approximately 50% of the complications. Of the 31 new neurologic deficits, 30 were nerve root injuries, and 1 was a *cauda equina* syndrome. Four (13%) were reported to have occurred intraoperatively, presumably determined from electromonitoring. Nineteen (61%) occurred acutely within 24 hours of surgery, and 8 (26%) were reported to have occurred in a delayed fashion of greater than 24 hours after surgery. Of the 31 patients with new neurologic deficit, 29 (94%) had recovery from these deficits, with 15 making complete and 14 making partial recoveries. One patient had no recovery, and 1 patient's recovery was not specified. Two revisions were performed to remove implants after deficits occurred, and both of these patients were reported to have made complete recoveries.

The majority of patients undergoing fusion procedures were instrumented (n = 518). There was no statistically significant difference in terms of complication rate (11% *vs.* 7.5%, $P = 0.3$) or rate of new neurologic deficit (5.8% *vs.* 1.2%, $P = 0.1$) between those instrumented and those not instrumented. In addition, there were no differences between the complication (10.6% *vs.* 9.1%, $P = 0.7$) and new neurologic deficit (5.3% *vs.* 4%, $P = 0.8$) rates in those patients over age 12 (n = 506) *versus* those under age 12 (n = 99).

A striking difference illustrated in Table 1 is the difference in complication rates between those patients undergoing a reduction procedure and those that did not. The overall complication rate is almost double in patients undergoing

TABLE 1. Complications in 605 Cases of Surgically Treated Pediatric Isthmic/Dysplastic Spondylolisthesis

Complication, No. (%)	All (n = 605)	Reduction (n = 229, 38%)	No Reduction (n = 376, 62%)	*P*
Wound infection				
Superficial	7 (1.2)	2 (0.9)	5 (1.3)	0.72
Deep	5 (0.8)	1 (0.4)	4 (1.1)	0.66
Neurological injury	31 (5.1)	23 (10.0)	8 (2.1)	0.001
Implant related	2 (0.3)	1 (0.4)	1 (0.3)	0.9
Dural tear	8 (1.3)	5 (2.2)	3 (0.8)	0.16
Death	0 (0)	0 (0)	0 (0)	NA
Pulmonary embolus	1 (0.2)	0 (0)	1 (0.3)	1
DVT	2 (0.3)	0 (0)	2 (0.5)	0.53
Other	7 (1.2)	1 (0.4)	6 (1.6)	0.07
Visual acuity change	0 (0)	0 (0)	0 (0)	NA
Total complications (%)	63 (10.4)	33 (14.4)	30 (7.9)	0.01

reduction ($P = 0.01$). Most of this difference can be attributed to the difference in the rate of new neurologic deficits. Neurologic deficits developed in 10% (23/229) of reduced patients *versus* only 2% (8/376) of patients without reduction ($P = 0.001$). Electrophysiological monitoring did not necessarily mitigate the rate of new neurologic deficits, as significantly more patients who had electromonitoring (29/422 or 6.9%) developed deficits than those who were not monitored (2/183 or 1%) ($P = 0.02$), although this is likely related to the use of such monitoring with more complex cases. In patients undergoing reduction, there was no significant difference in the development of neurologic deficit between those who were monitored (22/189) and those who were not (1/40) ($P = 0.09$).

Specific electromonitoring methods were recorded in the database. Most procedures that were monitored employed multiple methods. Of those patients monitored with EMG, 73% were also monitored with SSEP and 50% were also monitored with MEP. Of those who had SSEP monitoring, 63% also underwent MEP monitoring. Determining the neurologic deficit rate for a specific modality is therefore not readily possible from the database. In 8 of the monitored cases, abnormalities were detected, of which 7 employed EMG. Of these 8 cases, 5 resulted in new neurologic deficit, whereas the remaining 3 did not have a new deficit. Whether or not the lack of deficits in these 3 cases was due to a change in the procedure secondary to monitoring findings can not be ascertained from the information in the database.

Presence of preoperative neurologic element compression that warranted direct decompression was present in 49% (296) of the patients. Patients undergoing reduction procedures were significantly more likely to have had compression compared with those undergoing *in situ* fusion procedures (75% *vs.* 33%; $P = 0.001$). Also, of note, patients with preoperative compression were more likely to develop new neurologic deficits (9.5% *vs.* 1%; $P = 0.001$). Beginning in 2007 Meyerding grades were recorded and an analysis could be performed based on grade for 127 of the patients. Patients with high grade spondylolisthesis (Grades 3–5) were more likely than patients with low grade spondylolisthesis (grades 1–2) to require decompression (66% *vs.* 40%, $P = 0.004$), undergo reduction (76% *vs.* 32%, $P = 0.001$), and suffer new postoperative neurologic deficit (11.3% *vs.* 1.4%, $P = 0.021$). Few of the patients were treated with minimally invasive procedures (43/605). While none of these patients were reported to have developed new neurologic deficits, the difference did not reach statistical significance ($P = 0.16$).

DISCUSSION

The surgical treatment of pediatric isthmic and dysplastic spondylolisthesis has been associated with a high rate of surgical complications (Table 2) .[4,7,9,10,17,19] This study, which reports the data from the SRS M&M database, provides morbidity data on 605 pediatric isthmic/dysplastic spondylolisthesis cases. The overall rate of major complications of 10.4% is comparable to previous studies. These prior studies report variable rates ranging from 5% to 40%.[3,4,7–10,12,14–16,19,20] However, most of these studies are sufficiently small that the presence of a single complication can greatly affect the overall complication rate. The SRS M&M database provides for a larger population as it is a multicenter and multisurgeon self reported prospective database. A recent report has supported its utility for conditions such as adolescent idiopathic scoliosis.[18]

Another variable found in most of the previous studies was the reporting of pseudarthrosis as a complication. These rates ranged from 0% to 19%.[2,3,7,8,10,19] The SRS M&M database does not specifically report pseudarthrosis as a complication as it is not designed for reporting long-term outcomes. Therefore this report does not provide data on this complication,

TABLE 2. Summary of Prior Reports on the Incidence of Complications With Surgical Treatment of Pediatric Isthmic and Dysplastic Spondylolisthesis

Study	No. Patients	Age Group	Procedure	Comp Rate Excluding Pseudoarthrosis	Neuro Deficit Rate
Boxall *et al*[9]	39	Children and adolescents	Reduction or in situ fusion	10%	5.1%
Stanton *et al*[21]	20	Children and adolescents	*In situ* fusion	5%	5%
Dick and Schnebel[10]	15	Adolescents and young adults	Reduction	40%	27%
Lindholm *et al*[3]	57	Children and adolescents	Reduction	7%	1.7%
Schoenecker *et al*[15]	189	Children and adolescents	*In situ* fusion	NA	6.3%
Poussa *et al*[4]	22	Adolescents	Reduction or *in situ* fusion	18%	4.5%
Hu *et al*[19]	16	Adolescents and young adults	Reduction	25%	25%
Muschik *et al*[12]	59	Children and adolescents	Reduction and *in situ* fusion	12%	3.4%
Molinari *et al*[7]	32	Children and adolescents	Reduction or *in situ* fusion	22%	11%
Roca *et al*[20]	14	Adolescents and young adults	*In situ* fusion	14%	14%
Shufflebarger *et al*[16]	18	Adolescents	Reduction	27%	0%
Ruf *et al*[14]	27	Adolescents and young adults	Reduction	26%	22%
Helenius *et al*[8]	70	Children and adolescents	*In situ* fusion	10%	5.7%

and the overall complication rate should be viewed accordingly. Specifically, as previous studies suggest a lower pseudarthrosis rate for reduction procedures,[6] the overall difference in complications between *in situ* fusion and reduction may decrease if pseudarthrosis could be included.

Past studies have suggested that pediatric patients undergoing reduction are more likely to suffer neurologic deficits.[4,7] The data from the SRS M&M database supports this, as patients undergoing reduction had a 5 times higher rate of developing new neurologic deficits. However, it should be noted that patients undergoing reduction were more likely to have had preoperative neurologic compression. One of the shortcomings of this database is its lack of data on specific preoperative parameters, including degree of slip. If one takes the reporting of preoperative neurologic element compression as a proxy for increased degree of spondylolisthesis, then it suggests that those patients undergoing reduction were more likely to have a higher grade of spondylolisthesis. It should also be noted that patients with preoperative neurologic element compression were more likely to develop new neurologic deficits after surgery. This begs the question of whether or not patients undergoing reduction were more prone to deficit due to the nature of their deformities and not necessarily because of the procedure performed. Recent studies with advanced reduction techniques have suggested that reduction can be performed safely with no or only minor transient neurologic deficits.[14,16] However, our analysis of complication rates based on the subset in 2007 in which Meyerding grades were reported suggests that many patients with high grade slips, underwent reduction, and suffered a significant rate of new neurologic deficit after surgery.

The present study has several strengths. This is the largest study to date to evaluate the incidences of complications in the surgical treatment of isthmic/dysplastic pediatric spondylolisthesis by a large group of spinal deformity surgeons. Furthermore, since all of these cases were submitted during a recent 4-year period, they likely reflect a relatively current standard of care.

There are several limitations of this study. The SRS database relies on the accurate submission of data by members. Thus, it is possible that not all complications have been thoroughly documented, which could result in underestimation of complication rates. No method currently exists to evaluate the completeness and accuracy of the data, instead relying on good faith of those surgeons reporting. It is likely that major complications and neurologic complications are more accurately reported, however, this cannot be verified.[18] In addition, although data are reported prospectively, this is a retrospective study and as such is subject to the weaknesses inherent to such investigations including confounding variables and reporting bias.

➢ Key Points

- ❑ The surgical treatment of pediatric isthmic and dysplastic spondylolisthesis is associated with a relatively high rate of morbidity.
- ❑ Patients undergoing reduction were more likely to suffer a new neurologic deficit, although most were transient.
- ❑ Patients with higher grades of spondylolisthesis were more likely to suffer a new neurologic deficit with surgical treatment.

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Complication Rates of Three Common Spine Procedures and Rates of Thromboembolism Following Spine Surgery Based on 108,419 Procedures

A Report From the Scoliosis Research Society Morbidity and Mortality Committee

Justin S. Smith, MD, PhD,* Kai-Ming G. Fu, MD, PhD,* David W. Polly, Jr., MD,† Charles A. Sansur, MD, MHSc,‡ Sigurd H. Berven, MD,§ Paul A. Broadstone, MD,¶ Theodore J. Choma, MD,‖ Michael J. Goytan, MD,** Hilali H. Noordeen, MD,†† Dennis Raymond Knapp, Jr., MD,‡‡ Robert A. Hart, MD,§§ William F. Donaldson III, MD,¶¶ Joseph H. Perra, MD,‖‖ Oheneba Boachie-Adjei, MD,*** and Christopher I. Shaffrey, MD*

Study Design. Retrospective review of a prospectively collected database.

Objective. The Scoliosis Research Society (SRS) collects morbidity and mortality (M and M) data from its members. Our objectives were to assess complication rates for 3 common spine procedures, compare these results with prior literature as a means of validating the database, and to assess rates of pulmonary embolism (PE) and deep venous thrombosis (DVT) in all cases reported to the SRS over 4 years.

Summary of Background Data. Few modern series document complication rates of spinal surgery as routinely practiced across academic and community settings. Those available are typically based on relatively low numbers of procedures or confined to single-surgeon experiences.

Methods. The SRS M and M database was queried for lumbar microdiscectomy (LD), anterior cervical discectomy and fusion (ACDF), and lumbar stenosis decompression (LSD) cases from 2004 to 2007. Revisions were excluded. The database was also queried for occurrence of clinically evident PE and DVT in all cases from 2004 to 2007.

Results. A total of 9692 LDs, 6735 ACDFs, and 10,329 LSDs were identified, with overall complication rates of 3.6%, 2.4%, and 7.0%, respectively. These rates are comparable to previously published smaller series. For assessment of PE and DVT, 108,419 cases were identified and rates were calculated per 1000 cases based on diagnosis, age group, and implant use. Overall rates of PE, death due to PE, and DVT were 1.38, 0.34, and 1.18, respectively. Among 82,082 adults, the rate of PE ranged from 0.47 for LD to 12.4 for metastatic tumor. Similar variations were noted for DVT and deaths due to PE.

Conclusion. Overall major complication rates for LD, ACDF, and LSD based on the SRS M and M database are comparable to those in previously reported smaller series, supporting the validity of this database for study of other less common spinal disorders. In addition, our data provide general benchmarks of clinically evident PE and DVT rates as a basis for ongoing efforts to improve care.

Key words: spine surgery, complications, lumbar discectomy, lumbar stenosis, cervical fusion, pulmonary embolism, thromboembolism. **Spine 2010;35:2140–2149**

From the *Department of Neurosurgery, University of Virginia Medical Center, Charlottesville, VA; †Department of Orthopedic Surgery, University of Minnesota, Minneapolis, MN; ‡Department of Neurosurgery, University of Maryland, Baltimore, MD; §Department of Orthopedic Surgery, University of California–San Francisco, San Francisco, CA; ¶Spine Surgery Associates, Chattanooga, TN; ‖Department of Orthopedic Surgery, University of Missouri, Columbia, MO; **Health Sciences Centre, Winnipeg, MB, Canada; ††The Royal National Orthopedic Hospital, the Great Ormond Street Children's Hospital, London, United Kingdom; ‡‡Orlando Health, Orlando, FL; §§Department of Orthopedic Surgery, Oregon Health & Science University, Portland, OR; ¶¶Department of Orthopedic Surgery, University of Pittsburgh Physicians, Pittsburgh, PA; ‖‖Twin Cities Spine Center, Minneapolis, MN; and ***Hospital for Special Surgery, New York, NY.

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This project was submitted to the Hospital for Special Surgery (New York, NY) Institutional Review Board (IRB) and was determined to be exempt from IRB approval based on the use of de-identified data (IRB number 29045).
Address correspondence and reprint requests to Christopher I. Shaffrey, MD, Department of Neurosurgery, University of Virginia, PO Box 800212, Charlottesville, VA 22908; E-mail: CIS8Z@hscmail.mcc.virginia.edu

All surgical procedures have inherent risks of complications. One of the most effective mechanisms by which the safety of patient care is improved is through assessing these complications. The Scoliosis Research Society (SRS) is a group of predominantly fellowship-trained spine surgeons dedicated to the study of spinal deformity. As part of its mission, the SRS collects surgical case data from its members, including morbidity and mortality (M and M). Three reports have documented complications associated with spine procedures based on the SRS M and M database.[1–3]

Lumbar microdiscectomy (LD), anterior cervical discectomy and fusion (ACDF), and lumbar stenosis decompression (LSD) are among the most common spine surgical procedures performed.[4] Several reports have documented the rates of complications associated with LD,[4–17] ACDF,[18–32] and LSD.[12,33–39] Advances in techniques have substantially decreased the rates of compli-

cations associated with these procedures.[5] However, there are few modern series that document complication rates of spinal surgery as routinely practiced across academic and community settings, and those that are available are typically based on relatively low numbers of procedures or confined to single-surgeon experiences.

Deep venous thrombosis (DVT) and pulmonary embolism (PE) are significant potential complications of spinal surgery. A limited number of reports have documented rates of these events in relatively small populations of patients.[40–54] A majority of these prior reports have focused on high-risk populations, including patients with trauma[44] or patients undergoing major reconstructive spinal surgery.[41,45–47,49,51,53]

Our objectives in the present study were 3-fold. First, we sought to assess the rates of complications associated with the surgical treatment of 3 common spine procedures in a large population of patients. Our second objective was to compare these rates with prior reports as a means of validating the SRS M and M database for the study of less common spine procedures and complications. Our third objective was to use all cases reported to the database over 4 consecutive years to assess rates of clinically evident PE and DVT following spinal surgery.

■ Materials and Methods

Patient Population

Before application for active membership in the SRS, surgeons must complete 5 years of candidate membership. International membership status is also available for surgeons outside of the United States. Candidate members are required to collect and submit data on all spine cases performed, including all associated M and M. Active members are also encouraged to submit their cases. Deidentified data are collected using a secure internet-based data entry form. The SRS has invested substantial resources in this database and emphasizes to its membership the importance of accurate and consistent reporting. In addition, data submission includes a process in which members formally attest that submitted data are true and complete.

The SRS M and M database was queried for all reported LD, ACDF, and LSD cases from 2004 to 2007. Revision cases were excluded. LD cases including fusion and ACDF cases including corpectomy or posterior fusion were excluded. Only adult patients (≥21-years old) were included for the assessment of LSD. For neurologic complications, degree of recovery (complete, partial, or none) was recorded. New acute neurologic deficits were defined as those that developed intraoperatively or within 24 hours of surgery. Delayed new neurologic deficits were defined as those that developed after 24 hours following surgery.

The SRS M and M database was also queried for reports of DVT and PE in all cases from 2004 to 2007, including both primary and revision cases. Rates of DVT and PE, as well as deaths due to PE, were calculated per 1000 cases.

Rates of complications derived from the present study were compared with previously published reports. These reports were identified through a structured literature review. The English literature available through PubMed was searched using the terms "spine surgery" and "complications." The 12 reports from no earlier than 1970 that included at least 100 cases of LD, ACDF, and/or LSD and had specific documentation of complication rates were selected for comparison with the corresponding cases in the present series. Additional search strategies included "pulmonary embolism" and "spine surgery," as well as "deep venous thrombosis" and "spine surgery." The 15 reports from no earlier than 1990 that included specific documentation of DVT and/or PE rates associated with spinal surgery were selected for comparison with the present series.

This project was submitted to the Hospital for Special Surgery (New York, NY) Institutional Review Board (IRB) and was determined to be exempt from IRB approval based on use of deidentified data (IRB 29045).

Statistical Analyses

Frequency distributions and summary statistics were calculated for all clinical and demographic data. For categorical variables, cross-tabulations were generated and Fisher exact tests were used to compare distributions. All statistical analyses were 2-sided. $P < 0.05$ was considered significant.

■ Results

Patient Population

A total of 108,419 surgical cases were reported to the SRS M and M database from 2004 to 2007, including 9692 LD, 6735 ACDF, and 10,329 LSD procedures that met inclusion criteria. Patient age, whether a minimal access approach was used, and membership status of the submitting surgeon are summarized in Table 1.

Rates of Complications

Lumbar Discectomy. Among 9692 cases of LD, there were 346 complications reported, resulting in an overall complication rate of 3.6% (Table 2). There was a nonsignificant trend toward increased complications in patients >50 years old compared with patients ≤50 years old ($P = 0.06$). The rate of dural tear was significantly higher among patients >50-years old (2.1%), *versus* those ≤50-years old (1.4%) ($P = 0.02$).

New neurologic deficits were reported in 28 patients. One was related to spinal cord injury, and this patient had partial recovery. One case of *cauda equina* syndrome was reported, and this patient also had partial recovery. The remaining 26 were nerve root injuries, with 5 (19%) having complete, 19 (73%) having partial, and 2 (8%) having no recovery. Five delayed new neurologic deficits were reported. All of these occurred at the level of the nerve root, and 2 (40%) had complete and 3 (60%) had partial recovery.

Neuromonitoring was used for 895 (9%) of the LD procedures, including EMG monitoring in 577 (6%), SSEP monitoring in 765 (8%), and MEP monitoring in 83 (<1%). Of the 28 cases complicated by a new neurologic deficit, none used neuromonitoring.

The overall rates of complications among candidate, active, and international members were 4.1%, 3.4%, and 3.7%, respectively. Although the overall rate of complications was greater among candidate members compared with active members, this difference did not

Table 1. Summary Characteristics of 108,419 Cases of Spinal Surgery Submitted to the Scoliosis Research Society Morbidity and Mortality Database From 2004 Through 2007

	Total	Lumbar Micodiscectomy	ACDF	Lumbar Stenosis Decompression
No. cases (%)	108,419 (100)	9692 (8.9)	6735 (6.2)	10,329 (9.5)
Patient age				
Mean	47 yr	43 yr	49 yr	63 yr
Range	1 mo–97 yr	10–92 yr	15–89 yr	21–96 yr
Minimal access approach, n (%)				
Yes	14,301 (13)	3619 (37)	—	1260 (12)
No	94,115 (87)	6073 (63)	—	9069 (88)
Not recorded	3 (<1)	0	—	0
Membership status, n (%)				
Active	76,798 (71)	6956 (72)	5115 (76)	7755 (75)
Candidate	25,004 (23)	2042 (21)	1512 (22)	1777 (17)
International	6533 (6)	694 (7)	108 (2)	797 (8)
Not specified	143 (<0.1)	0	0	0

ACDF indicates anterior cervical discectomy and fusion.

reach statistical significance ($P = 0.07$). Candidate and active members did not significantly differ with regard to rates of dural tear (1.8% and 1.5%, respectively, $P = 0.2$), wound infection (0.8% for each group, $P = 0.6$), or new neurologic deficit (0.4% and 0.3%, respectively, $P = 0.2$).

The rate of complications for procedures performed using an open approach was modestly higher than for procedures performed using a minimal access approach (3.9% *vs.* 3.1%, $P = 0.05$). Further assessment demonstrated no significant differences in the rates of dural tear or new neurologic deficit between open and minimal access cases ($P = 0.3$ and $P = 0.2$, respectively). Procedures performed through a minimal access approach had a significantly lower rate of wound infection compared with procedures performed through a traditional open approach (0.4% *vs.* 1.1%, $P < 0.001$) (Figure 1). This difference was most pronounced for deep wound infections, for which the rate was 0.5% for open approaches and 0.08% for minimal access approaches ($P < 0.001$).

Table 2. Complications in Cases of First-Time Lumbar Microdiscectomy, Anterior Cervical Discectomy and Fusion, and Lumbar Stenosis Decompression Procedures From the Year 2004 to 2007

	Procedure		
Complication, Number (%)	Lumbar Micodiscectomy (n = 9692)	ACDF (n = 6735)	Lumbar Stenosis Decompression (n = 10,329)
Dural tear	156 (1.6)	11 (0.2)	321 (3.1)
Wound infection			
Superficial	45 (0.5)	13 (0.2)	93 (1.0)
Deep	34 (0.4)	9 (0.1)	111 (1.1)
Other	57 (0.6)	24 (0.4)	9 (0.01)
Acute neurologic	28 (0.3)	19 (0.3)	62 (0.6)
Wound hematoma	11 (0.1)	19 (0.3)	51 (0.5)
Delayed neurologic	5 (0.05)	3 (0.04)	0
Cardiorespiratory	3 (0.03)	3 (0.04)	7 (0.01)
Pulmonary (not PE)	2 (0.02)	6 (0.09)	13 (0.01)
Pulmonary embolus	2 (0.02)	3 (0.04)	9 (0.01)
Nonfatal hematologic	2 (0.02)	2 (0.03)	0
DVT	1 (0.01)	1 (0.01)	8 (0.01)
Death	0	4 (0.06)	13 (0.01)
Brachial plexus injury	0	2 (0.03)	0
Sepsis	0	1 (0.01)	1 (0.001)
Visual acuity change	0	0	0
Implant related	NA	17 (0.3)	21 (0.2)
Dysphagia	NA	14 (0.2)	NA
Recurrent laryngeal nerve injury	NA	8 (0.1)	NA
Total complications	346	159	719
Percent complications*	3.6%	2.4%	7.0%

*Percent complications = 100 × (total no. complications)/(no. patients).
ACDF indicates anterior cervical discectomy and fusion; PE, pulmonary embolism; DVT, deep venous thrombosis.

Anterior Cervical Discectomy and Fusion. Among 6735 cases of ACDF, there were 159 complications reported, resulting in an overall complication rate of 2.4% (Table 2). There were 4 deaths (approximately 1:1500), 3 due to PE and 1 due to "cardiopulmonary collapse." The overall complication rate was significantly higher for patients >50-years old (3.1%) compared with patients ≥50-years old (1.9%, $P = 0.03$).

The overall rates of complications among candidate, active, and international members were 2.5%, 2.3%, and 3.7%, respectively. The overall rate of complications

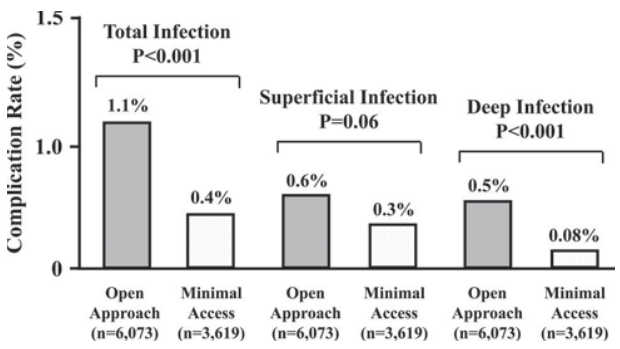

Figure 1. Rates of wound infection associated with primary lumbar discectomy, comparison of open approach with a minimal access approach.

Table 3. Thromboembolic Complications Reported for 108,419 Spine Procedures From the Year 2004 to 2007*

	n	PE Rate (per 1000 Cases)	PE Deaths (per 1000 Cases)	DVT Rate (per 1000 Cases)
Adult (≥21 yr old)				
Degenerative disease	47,389	0.89	0.19	0.80
Cervical	11,763	0.68	0.34	0.34
Thoracic	552	3.62	1.81	5.43
Lumbar	34,731	0.92	0.12	0.89
Lumbar discectomy	12,694	0.47	0.08	0.39
Lumbar stenosis	12,270	1.30	0.24	0.90
Fracture	6025	2.82	1.66	1.83
Kyphosis	2012	7.46	1.49	5.96
Spondylolisthesis	10,529	1.52	0.19	1.52
Scoliosis	5801	4.31	0.86	4.31
Other	10,326	2.32	0.58	1.55
Metastatic tumor	726	12.4	4.13	8.26
Total adult cases	82,082	1.69	0.43	1.44
Pediatric (<21 yr old)				
Degenerative disease	20	0.00	0.00	0.00
Fracture	623	1.61	0.00	1.61
Kyphosis	1555	0.64	0.64	2.57
Spondylolisthesis	827	1.21	0.00	2.42
Scoliosis	20,424	0.39	0.05	0.15
Other	1983	0.00	0.00	0.00
Total pediatric cases	25,432	0.43	0.08	0.39
Implants				
Yes	74,114	1.75	0.42	1.59
No	34,305	0.58	0.17	0.29
Primary *vs.* revision				
Adult primary	67,967	1.69	0.41	1.22
Adult revision	14,114	1.91	0.50	2.48
Pediatric primary	23,116	0.48	0.09	0.39
Pediatric revision	2316	0.00	0.00	0.43
All cases	108,419	1.38	0.34	1.18

*Patient age and/or diagnostic category not available for 905 cases.

between candidate and active members did not differ significantly ($P = 0.6$).

A total of 22 new neurologic deficits were reported, including 19 acute and 3 delayed. Acute injuries included 13 nerve root injuries, 3 incomplete spinal cord injuries, 1 Horner's syndrome, and 1 cerebellar stroke. Of the nerve root injuries, 4 had partial and 9 had complete recovery. All 3 of the incomplete spinal cord injuries had complete recovery. The median nerve injury was reported to have partial recovery. The outcomes of the cerebellar stroke and the Horner syndrome were not reported. The 3 delayed neurologic injuries included 2 nerve root injuries, 1 with partial and 1 with complete recovery, and an incomplete spinal cord injury that had partial recovery.

Neuromonitoring was used for 3110 (46%) of the ACDF procedures, including EMG monitoring in 1856 (28%), SSEP monitoring in 2606 (39%), and MEP monitoring in 956 (14%). Of the 4 cases complicated by spinal cord injury, only one used neuromonitoring (SSEP monitoring) and no intraoperative abnormalities were reported based on this monitoring. Of the 15 cases complicated by nerve root injuries, neuromonitoring was used for 6 cases, including combined EMG, SSEP, and MEP monitoring in 4 cases, combined EMG and SSEP monitoring in 1 case, and unspecified neuromonitoring methods in 1 case. No intraoperative abnormalities were reported based on these monitoring methods in cases complicated by nerve root injuries.

Lumbar Stenosis Decompression. Among 10,329 cases of LSD, there were 719 complications reported, resulting in an overall complication rate of 7.0% (Table 2). Further analyses of the complications associated with this series of LSD have been reported previously.[55]

Thromboembolic Events. Of the total 108,419 cases reported from 2004 to 2007, rates of PE, death due to PE, and DVT were 1.38, 0.34, and 1.18 per 1000 cases, respectively (Table 3). Among the 82,082 adult patients, the rate of PE varied based on diagnosis, ranging from 0.47 per 1000 cases for LD to 12.4 per 1000 cases for metastatic tumor. Similar variations were noted for the rate of deaths due to PE and the rate of DVT (Table 3). Among pediatric patients, the rates of thromboembolic events were substantially less than for adults (Table 3).

The rates of PE and DVT were significantly greater for cases that included implants (1.75 and 1.59 per 1000 cases, respectively), compared with cases not including implants (0.58 and 0.29 per 1000 cases, respectively; $P < 0.001$ for each comparison) (Table 3). There was a trend toward a higher rate of death due to PE in cases including

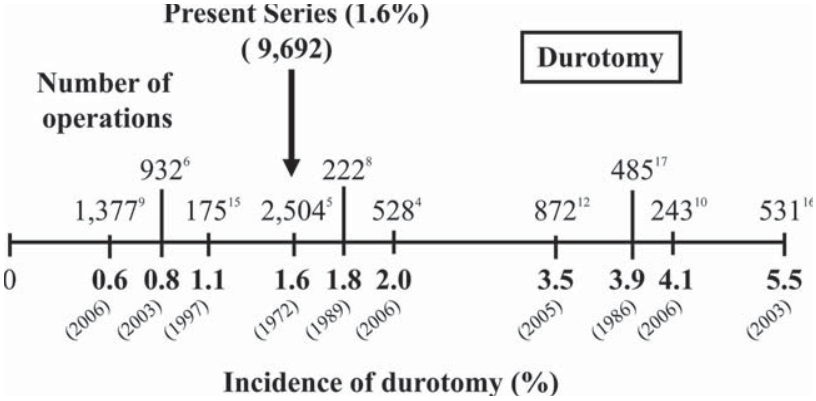

Figure 2. Comparison of the rates of dural tear associated with lumbar discectomy in the present series with rates from previously reported series. Numbers above the horizontal line indicate the number of procedures in each study, and numbers below the horizontal line indicate the percent of dural tears reported in each study.

implants (0.42 per 1000 cases) compared with cases not including implants (0.17 per 1000 cases; $P = 0.05$). Importantly, these data do not necessarily suggest a causation link between thromboembolic events and implants, but rather likely reflect a greater complexity and associated risk of cases that require implants.

Among adult cases, the rates of PE and death due to PE did not differ significantly between primary cases (1.69 and 0.41 per 1000 cases, respectively) compared with revision cases (1.91 and 0.50 per 1000 cases, respectively; $P = 0.6$ and $P = 0.7$, respectively) (Table 3). The rate of DVT for revision cases (2.48 per 1000 cases) was significantly greater than that of primary cases (1.22 per 1000 cases; $P = 0.001$). Among pediatric cases, the rates of PE, death due to PE, and DVT did not differ significantly between primary cases compared with revision cases (Table 3).

Comparison of Complication Rates With Prior Studies

Lumbar Discectomy. In an effort to assess the validity of the SRS M and M database as a resource to study less common spine procedures and complications, we compared the rates of the most common complications and the rate of mortality from the present series with those reported for similar series. For LD, the rate of dural tear ranged from 0.6% to 5.5% in previously reported series[4–6,8–10,12,15–17] (Figure 2). The majority of studies, especially those with greater numbers of patients, favored a rate of dural tear <2%. Each of these studies predominantly consisted of patients who underwent primary LD, except for the report by Best and Sasso,[9] in which 18% of the patients underwent LD for recurrent herniation. Despite the substantial inclusion of revision cases, the rate of durotomy was only 0.6%. The 1.6% rate of dural tear in the present series is consistent with these prior reports (Figure 2).

A majority of previous LD series report a rate of wound infection ≤1.6% (Figure 3).[5,6,9,10,13,15–17] Two series, published in 1986 and 1972,[5,17] report substantially higher infection rates (3.5% and 3.8%, respectively), however, these appear to be outliers relative to more recent studies. The report by Ramirez and Thisted[13] including 28,395 patients, was based on a national health care database and reports a rate of wound infection of 0.3%. The 0.8% rate of wound infection in the present series is comparable to recent reports (Figure 3).

New neurologic deficit is an uncommon event in LD, with recent series reporting rates of 0% to 0.6% (Figure 4).[5,6,8–10,13,15–17] The largest of these series,[13] reported a rate of 0.4%. Notably, the series of 1377 procedures from Best and Sasso, in which 18% of case were performed for recurrent herniation, reported a 0% rate of new neurologic deficit.[9] The 0.3% rate of new neurologic deficit in the present series is comparable to prior reports (Figure 4).

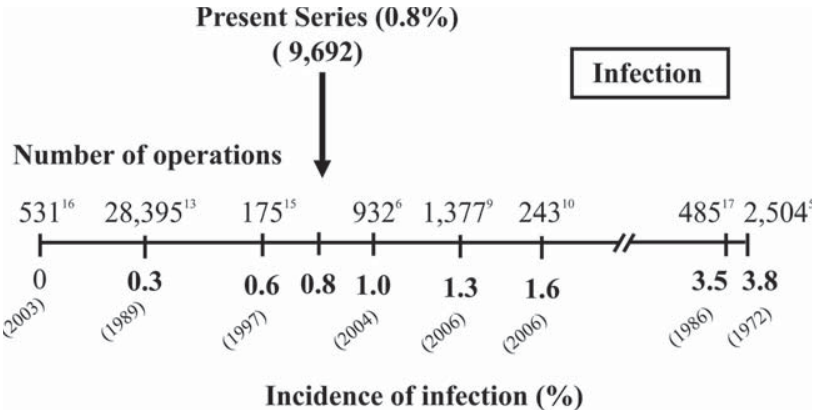

Figure 3. Comparison of the rate of wound infection associated with lumbar discectomy in the present series with rates from previously reported series. Numbers above the horizontal line indicate the number of procedures in each study, and numbers below the horizontal line indicate the percent of wound infections reported in each study.

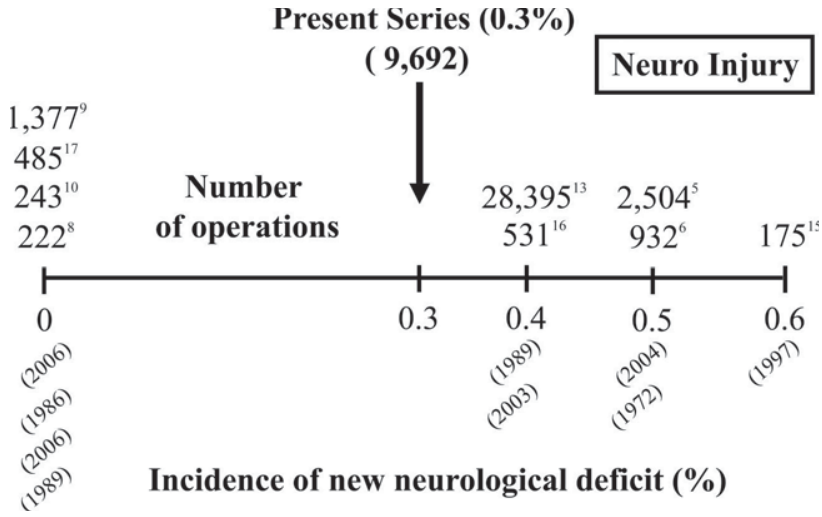

Figure 4. Comparison of the rate of new neurologic deficit associated with lumbar discectomy in the present series with rates from previously reported series. Numbers above the horizontal line indicate the number of procedures in each study, and numbers below the horizontal line indicate the percent of new neurologic deficits reported in each study.

Death associated with LD is a rare event in modern series, with reported rates ranging from 0% to 0.2% (Figure 5).[4–10,14–17] A substantial number of series lack any cases of mortality. The 0% rate of mortality in the present series is consistent with prior recent reports.

Anterior Cervical Discectomy and Fusion. Previous reports of overall complication rates for ACDF vary broadly: 0% (n = 42 and n = 251),[20,23] 1.8% (n = 109),[31] 10.8% (n = 56),[30] 11% (n = 90),[27] 11.7% (n = 103),[29] 14.2% (n = 450),[19] 19.3% (n = 1015),[26] 19.9% (n = 348),[21] 31.2%,[18] and 36.6% (n = 46).[28] Many factors may account for this wide variation, including how carefully one assesses for postoperative dysphagia and hoarseness, and the threshold at which one considers these to be complications. Dysphagia and hoarseness typically account for the majority,[18,26–28] or in some cases nearly all,[21,29,30] of the complications in the series with the greatest overall complication rates. In addition, earlier reports were typically performed on small populations of patients in which the complication rate can be significantly affected by a single incident. Furthermore, some series focus on more complex patient groups, for example, 3-level ACDFs,[28] which can substantially effect the complication rates. The overall rate of complications in the present series, 2.4%, is within the range of prior reports, but on the lesser end of this range, likely due to the considerably lower rates of dysphagia and hoarseness/recurrent laryngeal nerve palsy reported in the present series.

Dysphagia was reported in 0.2% of patients in the present series. Prior series have reported this rate to vary broadly: 0%,[23] 0.9%,[31] 5.4%,[30] 5.6%,[18] 5.8%,[29] 6.7%,[27] 9.5%,[26] 16.1%,[21] and 24%.[28] The reported rate of hoarseness/recurrent laryngeal nerve palsy also varies widely: 0%,[23,27,28] 0.9%,[31] 2.3%,[21] 3.1%,[26] 3.9%,[29] 5.4%,[30] and 7.9%.[18] Recurrent laryngeal nerve palsy was reported in 0.1% of cases in the present series.

The rate of wound infection in the present series, 0.3%, is comparable to prior series, in which it ranges from 0%[23,27,29–31] to 1.6%.[19] The rate of new neurologic deficit in the present series, 0.3%, is also compara-

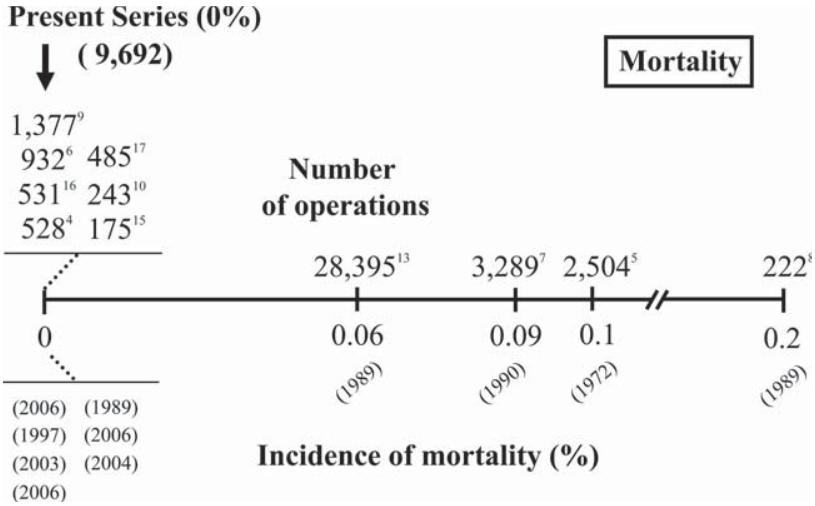

Figure 5. Comparison of the rate of mortality associated with lumbar discectomy in the present series with rates from previously reported series. Numbers above the horizontal line indicate the number of procedures in each study, and numbers below the horizontal line indicate the percent of deaths reported in each study.

ble to previous reports, in which it ranges from 0%[21,23,27,29–31] to 0.9%.[18] Wound hematoma was reported in 0.3% of cases in the present series and has been previously reported in 0%[20,23,27,29–31] to 5.6%[26] of cases. Death occurred in 0.06% (approximately 1 per 1500 cases) in the present series and has been previously reported to occur in 0%[18–21,23,27–31] to 0.1%[26] of cases.

Lumbar Stenosis Decompression. Few prior reports detail the M and M encountered with LSD.[33,35–37,39] The overall complication rate in the present series, 7.0%, is comparable to those of prior reports: 6% (n = 50),[39] 9.8% (n = 133),[35] 14.2% (n = 127),[36] 22% (n = 124),[37] and 12.2% (n = 471,215).[33] Durotomy was reported in 3.1% of the cases in the present series, which is comparable to prior reports: 2%,[34,39] 2.4%,[37] 2.6%,[12] 3.1%,[38] 6.8%,[35] and 11.8%.[36] The occurrence of wound infection in the present series, 2.1%, was also within the range of prior reports: 0%,[39] 0.3%,[33] 0.8%,[36,37] and 2.3%.[35] Three prior reports lacked any cases complicated by new neurologic deficit,[35,37,39] and 2 reports included neurologic complications with rates 0.7%[33] and 1.6%.[36] The neurologic complication rate in the present series, 0.6%, is comparable to these prior reports. Wound hematoma complicated 0.5% of the cases in the present series, which is comparable to the rates previously reported: 0%,[35,37] 2%,[39] 3.1%,[36] and 5.2%.[33] Most prior series lack any cases complicated by death.[35–37,39] The series from Li *et al*[33] reported a rate of death of 0.17% (approximately 2.5:1500), which is comparable to the present series (approximately 1:1500).

Thromboembolic Events. The reported rates of PE with spinal surgery range from 0% to 7.6%, and the reported rates of DVT range from 0.27% to 31% (Table 4). A majority of prior reports have focused on major spinal procedures[41,45–51,53] or specific high-risk patients.[42,44] In general, the reported rates of thromboembolic events are higher in studies that included routine screening methods (Table 4). For example, Piasecki *et al* reported 66 patients who underwent major spinal reconstructive surgery for adult deformity and identified DVTs in 9.1% and PEs in 7.6% of patients based on routine use of screening methods for both DVT and PE.[47] In contrast, Platzer *et al* reported 978 trauma patients who underwent spinal surgery and identified, in this high-risk population, symptomatic DVTs in 1.7% and PEs in 0.9% without routine use of screening methods.[44] It is unlikely, although there is no way of knowing for certain, that routine screening methods were used for the majority of cases in the present series, resulting in rates of DVT and PE that reflect clinically symptomatic events. In addition, in the present series, utilization of prophylaxis likely varied, not only among the surgeons who contributed cases but also based on patient age, diagnosis, and complexity of procedure.

■ Discussion

This study provides the rate of complications in the operative treatment of 3 common spine procedures based on a large number of cases performed predominantly by fellowship-trained spine surgeons. We have demonstrated that the rates of specific complications in the present series, especially major complications, are comparable to those in recent reports of smaller series. Collectively, these findings support the validity of the SRS M and M database as a resource to study other less common spinal disorders. Furthermore, we have used all of the cases submitted over a recent 4-year period to assess rates of PE, death due to PE, and DVT, and stratified these results based on diagnosis, patient age, whether implants were used, and whether the surgery was primary or revision.

The SRS M and M database is the compilation of submissions of SRS members around the world. The majority currently reside within North America. For the years that the data were collected and reviewed for this report, candidate members had a mandatory requirement to report their M and M. Active members were strongly encouraged to report their results. There is no currently existent method to determine the completeness of data submission, nor the accuracy of the reporting. It is dependent on the efforts of the participants. As such there are limitations to the data integrity, as there is with any large dataset.

It is the authors' opinion that major complications, such as new neurologic deficit, are probably most reliably reported within the database. These typically occur acutely and have the full attention of the treating surgeon. So, we suspect that recall bias in the reporting of major complications is limited but cannot prove or disprove this opinion. Importantly, the rates of DVT and PE reported herein likely reflect only those cases in which these events were symptomatically evident.

While it is possible that this dataset may have the bias of selective reporting, we believe that it does represent a cross section of practicing clinicians. Although there is the possibility that it may have the bias of reporting from younger surgeons in their candidate period and could represent somewhat of a worst case analysis, there are very senior surgeons reporting as well.

What is this database useful for? Rare conditions that have relatively uncommon events can only be examined in large datasets. While we wish we had a mechanism to achieve higher quality datasets, these have proven to be quite expensive ($13 Million NIH funding for the Spine Patient Outcomes Research Trial study, which enrolled perhaps 2000 patients with common conditions).[4,10] It would also be preferable to have outcomes data, but this carries additional expense and response burden. So, the question becomes what incremental value is gained from the incremental investment. No funding agency has paid for the SRS M and M dataset to date. It has been a goodwill volitional contribution from the members of

Table 4. Summary of Prior Reports of the Rates of Pulmonary Embolism (PE) and Deep Venous Thrombosis (DVT) in Spinal Surgery*

Author	Patient Population	No. Cases	Rates of PE/DVT	Prophylaxis	Comments
West and Anderson[50]	Major adult spinal surgery	41	DVT 14% PE 0%	Compression stockings	All patients screened with LE Doppler
Ferree[52]	Lumbar laminectomy/ laminotomy	60	DVT 5% PE 0%	Compression stockings	All patients screened with LE Doppler
Smith, *et al*[51]	Major reconstructive spinal surgery	317	DVT 0.6% PE 0.3% PE death 0.3%	Compression stockings and pneumatic compression	Prospective; 126 of the patients screened with LE Doppler
Rokito *et al*[41]	Major reconstructive spinal surgery	329	DVT 0.3% PE 0%	Compression stockings alone, with pneumatic compression, or with warfarin	Prospective; 110 of the patients screened with LE Doppler
Wood *et al*[49]	Major spinal surgery	136	DVT 0.7% PE 0.7% PE death 0%	Randomized to pneumatic thigh-high or foot wraps	Prospective, randomized; all patients screened with LE Doppler
Dearborn *et al*[53]	Major thoracolumbar spinal surgery	116	DVT 0.9% PE 2.6% PE death 0%	Compression stockings and pneumatic compression	Prospective; all patients screened with LE Doppler and 73 screened for PE with lung perfusion scan
Oda *et al*[40]	Posterior spinal surgery	110	DVT 15.5% PE 0%	None	All patients screened with LE Doppler
Epstein[43]	Anterior cervical spinal surgery with or without posterior cervical fusion	200	DVT 4% PE 1.5% PE death 0%	Compression stockings and pneumatic compression	Prospective; all patients screened with LE Doppler
Leon *et al*[42]	High-risk spinal surgery patients	74	DVT 31% PE 1.4% PE death 0%	Preoperative IVC filters placed in all patients	Prospective; all patients screened with LE Doppler and one-third underwent chest/pelvis CT
Platzer *et al*[44]	Trauma patients undergoing spinal surgery	978	DVT 1.7% PE 0.9% PE death 0.6%	792 LMWH; 153 compression stockings; 21 pneumatic compression; 12 no prophylaxis	No routine LE Doppler or CT imaging; rates reflect clinically symptomatic events
Cho *et al*[45]	Posterior fusion and instrumentation for degenerative scoliosis	47	DVT 0% PE 2.1% PE death 2.1%	Not specified	Whether routine screening performed not indicated
Piasecki *et al*[47]	Combined anterior/posterior reconstruction for adult spinal deformity	66	DVT 9.1% PE 7.6% PE death 0%	Mechanical prophylaxis with foot pumps	Prospective; all patients screened with LE Doppler, MRV, and chest CT
Pateder *et al*[46]	Adult spinal deformity surgery	361	PE 2.4% PE death 0%	Compression stockings and pneumatic compression; either warfarin or LMWH starting on postoperative day 1	No routine LE Doppler or CT imaging; rates reflect clinically symptomatic events
Schizas *et al*[48]	Vast majority were spinal fusion cases	270	PE 2.2% PE death 0%	Compression stockings and LMWH	Prospective; no routine LE Doppler or CT imaging; rates reflect clinically symptomatic events
Nicol *et al*[54]	Lumbar spine surgery	1111	DVT 0.27% PE 0%	No prophylaxis, aspirin, or LMWH	No routine LE Doppler or CT imaging; rates reflect clinically symptomatic events
Present study	All spine surgery	108,419	DVT 0.1% PE 0.14% PE death 0.03%	Mixed	Mixed

LE indicates lower extremity; DVT, deep venous thrombosis; PE, pulmonary embolus; LMWH, low-molecular-weight heparin; CT, computed tomography; MRV, magnetic resonance venogram; IVC, inferior *vena cava*.

the SRS. The total dataset has >108,000 cases enrolled for the 4-year period assessed in the present study. Presuming that each case required a 10-minute effort by the surgeon, this dataset represents a contribution of 18,000 man-hours. If a conservative estimate of the time value of the surgeons is made at $250 per hour, this dataset represents a $4.5 million dollar investment. To gain higher quality data of this scale would cost substantially more. So, while there are limitations in the dataset, the authors feel that it is a useful estimate of general information about current practice and complication rates for acute complications. At a minimum it is useful in discerning new trends, and for informing clinicians and patients about these particular risks.

■ Conclusion

The overall major complication rates for first-time LD, ACDF, and LSD based on the SRS M and M database are similar to those in previously reported smaller series. These data not only provide surgeons with potentially useful information for preoperative patient counseling but also serve to validate the SRS M and M database as a resource to study other less common spinal procedures and rare complications. Our data also provide general

benchmarks of clinically evident PE and DVT rates as a basis for ongoing efforts to improve safety of care.

■ Key Points

- The overall complication rates of 9692 lumbar discectomies, 6735 anterior cervical discectomies and fusions, and 10,329 LSDs reported to the Scoliosis Research Society M and M database were 3.6%, 2.4%, and 7.0%, respectively.
- The overall major complication rates for first-time LD, ACDF, and LSD based on the SRS M and M database are similar to those in previously reported smaller series, supporting the validity of this database as a resource to study other less common spinal disorders and complications.
- Based on 108,419 spinal surgery cases reported to the SRS M and M database, overall rates of clinically evident PE, death due to PE, and clinically evident DVT were 1.38, 0.34, and 1.18 per 1000 cases, respectively. Among 82,082 adults, the rate of PE ranged from 0.47 for LD to 12.4 for metastatic tumor.

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Incidence of Unintended Durotomy in Spine Surgery Based on 108 478 Cases

Brian J. Williams, MD*
Charles A. Sansur, MD*
Justin S. Smith, MD, PhD*
Sigurd H. Berven, MD†
Paul A. Broadstone, MD‡
Theodore J. Choma, MD§
Michael J. Goytan, MD¶
Hilali H. Noordeen, MD‖
D. Raymond Knapp, Jr, MD#
Robert A. Hart, MD**
Reinhard D. Zeller, MD††
William F. Donaldson, III, MD‡‡
David W. Polly, Jr, MD§§
Joseph H. Perra, MD¶¶
Oheneba Boachie-Adjei, MD‖‖
Christopher I. Shaffrey, MD#

*University of Virginia Medical Center, Charlottesville, Virginia; †Department of Orthopedic Surgery, University of California - San Francisco, San Francisco, California; ‡Spine Surgery Associates, PC, Chattanooga, Tennessee; §University of Missouri Orthopedic Surgery, Columbia, Missouri; ¶Health Sciences Centre, Winnipeg, Manitoba, Canada; ‖The Royal National Orthopedic Hospital, London, United Kingdom; #Arnold Palmer Children's Hospital, Orlando, Florida; **Orthopaedics Department, Oregon Health & Science University, Portland, Oregon; ††Division of Orthopedic Surgery, Hospital for Sick Children, Toronto, Ontario, Canada; ‡‡Orthopedic Surgery, University of Pittsburgh Physicians, Pittsburgh, Pennsylvania; §§Department of Orthopedic Surgery and Neurosurgery, University of Minnesota, Minneapolis, Minnesota; ¶¶Twin Cities Spine Center, Minneapolis, Minnesota; ‖‖Hospital for Special Surgery, New York, New York

Correspondence:
Christopher I. Shaffrey, MD,
University of Virginia Medical Center,
Department of Neurosurgery,
PO Box 800212,
Charlottesville, VA 22908.
E-mail: cis8z@hscmail.mcc.virgina.edu

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BACKGROUND: Unintended durotomy is a common complication of spinal surgery. However, the incidences reported in the literature vary widely and are based primarily on relatively small case numbers from a single surgeon or institution.

OBJECTIVE: To provide spine surgeons with a reliable incidence of unintended durotomy in spinal surgery and to assess various factors that may influence the risk of durotomy.

METHODS: We assessed 108 478 surgical cases prospectively submitted by members of the Scoliosis Research Society to a deidentified database from 2004 to 2007.

RESULTS: Unintended durotomy occurred in 1.6% (1745 of 108 478) of all cases. The incidence of unintended durotomy ranged from 1.1% to 1.9% on the basis of preoperative diagnosis, with the highest incidence among patients treated for kyphosis (1.9%) or spondylolisthesis (1.9%) and the lowest incidence among patients treated for scoliosis (1.1%). The most common indication for spine surgery was degenerative spinal disorder, and among these patients, there was a lower incidence of durotomy for cervical (1.0%) vs thoracic (2.2%; $P = .01$) or lumbar (2.1%, $P < .001$) cases. Scoliosis procedures were further characterized by etiology, with the highest incidence of durotomy in the degenerative subgroup (2.2% vs 1.1%; $P < .001$). Durotomy was more common in revision compared with primary surgery (2.2% vs 1.5%; $P < .001$) and was significantly more common among elderly (> 80 years of age) patients (2.2% vs 1.6%; $P = .006$). There was a significant association between unintended durotomy and development of a new neurological deficit ($P < .001$).

CONCLUSION: Unintended durotomy occurred in at least 1.6% of spinal surgeries, even among experienced surgeons. Our data provide general benchmarks of durotomy rates and serve as a basis for ongoing efforts to improve safety of care.

KEY WORDS: Complications, Durotomy, Scoliosis, Spine surgery, Spondylolisthesis

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Unintended durotomy is a common complication of spinal surgery. The incidences reported in the literature vary widely and are based primarily on relatively small case numbers from a single surgeon or institution.[1-28] Reported incidences of durotomy vary, depending on the type and complexity of procedure: lumbar decompression, 8.5% to 14%[7,14,23]; diskectomy, 0.2% to 5.9%[3-5,7,9,11,16,22,23,25]; reoperation, 8.1% to 17.4%[4,5,7,11,19,23,29]; and instrumentation, 0% to 35%.[1,8,20,21,28] Several factors have been associated with durotomy, including surgeon experience,[27] revision spinal surgery,[2,4,5,7,13,19,23] previous irradiation,[30] and age.[2,14] Typically, durotomy is treated primarily during surgery or managed postoperatively with flat bed rest that may include subarachnoid drain placement; however, despite these measures, some patients require surgical revision.[3,7,31-37] Complications resulting from dural tears include cerebrospinal fluid leak or fistula[6,35,38,39]; meningitis,[39,40]

ABBREVIATION: SRS, Scoliosis Research Society

arachnoiditis, or spinal epidural abscess[39-44]; psuedomeningocele[37,38,45-48]; intracranial or intraspinal hemorrhage[49-51]; and headache.[44,45,47,52]

The Scoliosis Research Society (SRS) routinely collects morbidity and mortality data from its members, which provide a wealth of data from experienced spine surgeons. The purpose of this study is to provide spine surgeons with a reliable incidence of unintended durotomy in spinal surgery and to assess various factors that may influence the risk of durotomy.

MATERIALS AND METHODS

The SRS morbidity and mortality database is the compilation of submissions of SRS members around the world, the majority of whom reside in North America. For the years the data were collected and reviewed for this report, candidate members had a mandatory requirement to report their morbidity and mortality results. Full active members were strongly encouraged to report their results.

To assess the incidence of unintended durotomy, all reported surgical cases from 2004 through 2007 were extracted from the SRS database. All data had previously been deidentified of any information regarding the patient, surgeon, or institution. This project was submitted to the Hospital for Special Surgery (New York, New York) Institutional Review Board and was deemed to be exempt from Institutional Review Board approval on the basis of the use of deidentified data (Institutional Review Board No. 29045).

In addition to evaluating whether an unintended durotomy occurred, other parameters analyzed included patient age, diagnosis, whether the surgery was primary or revision, presence or absence of preoperative neural compression, whether and how fusion was performed, whether instrumentation was used, whether surgery was performed using a minimally invasive approach, membership status of the surgeon (active, candidate, international), and whether an associated neurological complication occurred.

The overall incidence of unintended durotomy was tabulated and stratified further on the basis of clinical and surgical factors. Statistical analysis of these data was performed with SPSS for Windows version 15.0 (SPSS Inc, Chicago, Illinois). The 2-proportion Z test (2 tailed with unequal variances) was used when a Clopper Pearson distribution was approximated by a normal distribution. If a normal distribution did not approximate the data accurately, then the Fisher exact test was used. For analysis of 2 means, the 2-sample unpooled t test was used. Unless otherwise specified, a 2-proportion Z test was used. Statistical significance was based on a value of $P < .05$.

RESULTS

A total of 108 478 surgical cases were reported to the SRS database from 2004 through 2008, including 76 798 (71%) from active members, 25 004 (23%) from candidate members, 6533 (6%) from international members, and 143 (0.1%) lacking designation of membership status. The overall mean patient age was 47 years (median, 48 years; range, 1 month-97 years), and the most common preoperative diagnoses included degenerative spinal disorder (44% of cases), scoliosis (24% of cases), and spondylolisthesis (11% of cases; Table 1).

The overall incidence of unintended durotomy was 1.6% (1745 of 108 478). Based on primary diagnosis, the incidence of unintended durotomy ranged from 1.1% among patients treated for scoliosis to 1.9% among patients treated for kyphosis or spondylolisthesis (Table 1).

Degenerative spinal disorders were further characterized by subtype and spinal level (Table 2). The incidence of durotomy based on degenerative disease subtype ranged from 1.0% among

TABLE 1. Incidence of Unintended Durotomy Based on Primary Diagnosis[a]

Primary Diagnosis	Cases, n	Durotomies, n	Incidence of Durotomy, %
Kyphosis	3599	68	1.9
Spondylolisthesis	11 421	214	1.9[a]
Degenerative spinal disorder	47 399	852	1.8[a]
Other	12 456	216	1.7
Not recorded	475	7	1.5
Fracture	6704	92	1.4
Scoliosis	26 424	296	1.1[a]
Total	108 478	1745	1.6

[a]Spondylolisthesis and degenerative spinal disorder had significantly higher rates of durotomy compared with the overall rate ($P = .026$ and $P < .001$, respectively), and scoliosis had a significantly lower rate ($P < .001$).

TABLE 2. Incidence of Unintended Durotomy Among Patients With a Primary Diagnosis of Degenerative Spinal Disease, Stratified Based on Subtype of Degenerative Disease and Spinal Location

	Durotomies, n	Cases, n	Incidence of Durotomy, %
Subtype of degenerative disease			
Not recorded	17	614	2.8
Degenerative disk disease	234	9256	2.5[a]
Spondylotic radiculopathy	58	2861	2.0
Spinal stenosis	273	16 036	1.7
Disk herniation	264	18 008	1.5[a]
Lumbar postlaminectomy syndrome	6	624	1.0
Total	852	47 399	1.8
Degenerative spinal disorder level			
Thoracic	12	552	2.2[b]
Lumbar	721	34 731	2.1[b]
Not recorded	7	334	2.1
Cervical	112	11 782	1.0
Total	852	47 399	1.8

[a]Degenerative disk disease had a significantly higher rate of durotomy compared with the degenerative disease group ($P < .001$), and disk herniation had a significantly lower rate ($P < .001$).

[b]Cervical vs thoracic location had a significantly lower rate of durotomy ($P = .01$, Fisher exact test), as well as cervical vs lumbar ($P < .001$).

patients treated for lumbar postlaminectomy syndrome to 2.5% among patients treated for degenerative disk disease. The most common subgroups of degenerative spinal disease were disk herniation (n = 18 008) and spinal stenosis (n = 16 036), which had durotomy incidences of 1.5% and 1.7%, respectively. Among procedures performed for degenerative disease, cervical procedures were found to have a less frequent association with durotomy (1.0%) compared with thoracic (2.2%; $P = .01$) and lumbar (2.1%; $P < .001$) procedures (Table 2).

Scoliosis cases were also characterized by etiology of the deformity. The most frequent type of scoliosis in this series was idiopathic (n = 14 296), followed by neuromuscular (n = 5191) and degenerative (n = 2577; Table 3). The incidence of unintended durotomy ranged from 0.9% to 2.2% among patients treated for idiopathic and degenerative scoliosis, respectively. The incidence of durotomy was significantly higher for degenerative scoliosis cases compared with scoliosis cases that were not degenerative ($P = .001$).

The mean age of patients with durotomy was 56 years (median, 55; range, 1 month to 95 years), which was significantly older than patients who did not have durotomy ($P < .001$, Student t test). When stratified by patient age, the incidence of durotomy varied from a minimum of 1.1% (43 of 3791) among patients 0 to 9 years of age to a maximum of 2.2% (81 of 3602) in those > 80 years of age (Table 4).

Multiple surgical and radiographic parameters were assessed for association with unintended durotomy occurrence (Table 5). The incidence of durotomy did not differ significantly based on whether the procedure did or did not include spinal fusion (1.6% and 1.7%, respectively; $P = .3$). Among the 72 691 cases that included fusion, the incidence of unintended durotomy was significantly higher for posterior lumbar interbody fusions (2.2% vs 1.6%; $P < .001$) and for posterolateral fusions (2.0% vs 1.6%; $P = .001$). Anterior-only fusions had a significantly lower incidence of durotomy (1.0%; $P < .001$). Furthermore, the use of instrumentation did not result in more durotomies; there were more durotomies in the group without instrumentation (1.5% with vs 1.8% without instrumentation; $P = .002$; Table 5).

TABLE 3. Incidence of Unintended Durotomy Based on Scoliosis Subtype

Scoliosis Subtype	Cases, n	Durotomies, n	Incidence of Durotomy, %
Degenerative	2577	57	2.2[a]
Congenital	2198	31	1.4
Other	1959	21	1.1
Neuromuscular	5191	53	1.0
Not recorded	203	2	1.0
Idiopathic	14 296	132	0.9[a]
Total	26 424	296	1.1

[a]Incidence of durotomy for degenerative scoliosis cases was significantly greater compared with all scoliosis cases ($P < .001$) and significantly lower for idiopathic scoliosis cases ($P = .001$).

TABLE 4. Incidence of Unintended Durotomy in Spine Surgery Based on 108 478 Cases, Stratified by Patient Age

Patient Age, y	Cases, n	Durotomies, n	Incidence of Durotomy, %
0-9	3791	43	1.1[a]
10-19	16 162	282	1.7
20-39	17 209	265	1.5
40-59	38 769	560	1.4
60-79	28 029	503	1.8
>80	3602	81	2.2[a]
Age not recorded	916	11	1.2
Total	108 478	1745	1.6

[a]The 0- to 9-year-old group had a significantly lower incidence of durotomy compared with the overall rate ($P = .005$), and the > 80-year-old group had a significantly higher rate ($P = .006$).

Revision surgery was associated with a significantly greater (almost 1.5 times) incidence of unintended durotomy (2.2%) compared with primary surgery (1.5%; $P < .001$; Table 5). There was no difference in the incidence of durotomy between open (1.6%) and minimally invasive (1.6%; $P = .9$) techniques. In addition, the incidence of durotomy did not differ significantly between cases with or without preoperative neural compression (1.6% and 1.7%, respectively; $P = .06$; Table 5). Furthermore, the incidence of durotomy did not differ depending on physician experience, with active members presumably having more and candidate members presumably having less experience (1.6% and 1.5% incidence, respectively; $P = .9$; Table 5).

Of the 1745 patients with an unintended durotomy, 78 (4.5%) had a postoperative neurological deficit compared with a postoperative neurological deficit rate of 1.6% for patients who did not have an unintended durotomy ($P < .001$). Of the 78 neurological deficits in patients with incidental durotomies, 37 (47%) occurred within 24 hours of surgery, 22 (28%) occurred > 24 hours after surgery, 18 (23%) occurred intraoperatively, and the development of 1 (1%) was not recorded. There were 55 nerve root injuries. Of these, 17 patients (31%) had complete resolution of their symptoms, 35 (64%) had partial resolution, 1 (2%) experienced no recovery, and 2 (4%) were not recorded. There were 7 cauda equina injuries; subsequently, 4 patients (57%) had complete resolution of their symptoms, 2 (29%) had partial resolution, and 1 (14%) was not documented. There were 13 incomplete spinal cord injuries, of which 5 (38%) had complete and 8 (62%) had partial resolution of symptoms. There was 1 complete cord injury that did not improve.

DISCUSSION

Unintended durotomy is one of the most common complications of spinal surgery, with an incidence ranging from 0.2% to 17%

TABLE 5. Summary of Surgical and Radiographic Characteristics Assessed for Association With Durotomy[a]

	Cases, n	Durotomies, n	Incidence of Durotomy, %	*P*
Spinal fusion performed				
Yes	72 691	1147	1.6	.3
No	35 782	598	1.7	
Not recorded	5	0	0	
Type of fusion[b]				
PLIF	14 896	332	2.2	<.001
Posterolateral	9991	200	2.0	<.001
TLIF	7089	128	1.8	0.1
Anterior-posterior	8156	125	1.5	0.7
Interlaminer/facet	16 192	213	1.3	<.001
Anterior only	15346	146	1.0	<.001
Not recorded	1021	3	0.3	
Total	72 691	1147	1.6	
Instrumentation				
Yes	74 121	1130	1.5	.002
No	34 313	615	1.8	
Not recorded	44	0	0	
Revision surgery				
Yes	16 502	355	2.2	<.001
No	91 918	1390	1.5	
Minimally invasive approach[c]				
Yes	14 301	227	1.6	.8
No	94 177	1518	1.6	
Preoperative neural compression				
Yes	66 457	1032	1.6	.06
No	41 936	713	1.7	
Member status				
Active member	76 798	1224	1.6	.4
Candidate member	25 004	378	1.5	

[a]PLIF, posterior lumbar interbody fusion; TLIF, transforaminal lumbar interbody fusion.
[b]Note that posterior lumbar interbody fusion and posterolateral fusion are associated with significantly greater incidences of durotomy, whereas anterior-only and interlaminer surgery is associated with a significantly lower incidence of durotomy.
[c]The 227 durotomies encountered with minimally invasive procedures were with the following procedures: kyphoplasty/vertebroplasty (n = 8), mini-open (n = 139), other minimally incisional (n = 78), and thoracoscopic (n = 2).

(Table 6).[1-28] The overall incidence of durotomy in our series, 1.6%, is consistent with previously published literature. The rate of durotomy for diskectomy including all levels, cervical, thoracic, and lumbar, was 1.5%. This is consistent with previous literature for diskectomy, which is composed primarily of lumbar diskectomy with rates ranging from 0.2% to 5.9%.[3-5,7,9,11,16,22,23,25]

We confirm that revision cases have a significantly higher incidence of durotomy, which has been reported previously.[2,4,7,11] We also show that older patients have a higher incidence of durotomy.[2] However, in this study, the relationship does not appear to be linear and may represent an effect caused by confounding variables such as the indication for surgery and the type of procedure performed. Certainly, in an older cohort of patients, degenerative spinal disease predominates, whereas in a younger sample, scoliosis and trauma are more common. These variations in pathology alter the goal of surgery and the type of procedure performed. All these variables could affect the incidence of durotomy in our study. Furthermore, we demonstrate that durotomy in this series has a statistically significant association with the development of new postoperative neurological deficit, which usually occurred within the first 24 hours after surgery. Recovery from these injuries was either partial or complete in the vast majority of cases. The relationship may seem intuitive in that one primary mode of neural injury is direct trauma, which may involve penetration of the dura to reach the neural elements. However, we must iterate that this study cannot demonstrate a causative relationship between these 2 events but merely an association. It is equally likely that the association is due to other factors not evaluated. Other sequelae related to dural tears, not assessed in the present series, are rare but may be severe, including meningitis or pseudomeningocele.[37-40,45-47]

The overall incidence of durotomy with scoliosis surgery in the present series was 1.1%, with a range of incidences depending on the etiology of scoliosis. In a prior report of the SRS regarding adolescent idiopathic scoliosis, the overall incidence of durotomy was 0.18% (12 of 6339), whereas our incidence in a similar group of idiopathic scoliosis was 0.9%. The higher incidence may be due to variations in patient population or procedure characteristics[12] or may simply reflect greater diligence in reporting.

Surprisingly, in the present series, surgery without instrumentation had a higher incidence of durotomy than did surgery with instrumentation. Although instrumentation may be more technically demanding, the higher rate of durotomy in uninstrumented patients could be attributed to a greater focus on decompression, which could impose a greater risk of durotomy.

Among the instrumented cases, the rate of durotomy was significantly greater with posterior lumbar interbody fusion compared with transforaminal lumbar interbody fusion. This may be due to the more aggressive retraction of the thecal sac required to access the disk space in a posterior lumbar interbody fusion compared with the transforaminal lumbar interbody fusion approach, which is based on a more lateral approach with less thecal sac retraction.[18,28,53]

In the present study, the experience of the surgeon as indicated by member status did not result in a higher incidence of durotomy for the less experienced surgeons. Wiese et al[27] compared the incidence of durotomy between surgeons who had performed 50 to 100 and those who performed > 500 microdisketomies and demonstrated a higher incidence of durotomy in the former group (Table 6). Although not specifically assessed, the vast majority of candidate members of the SRS are fellowship-trained spine surgeons dedicated to the treatment of complex spinal conditions. This may contrast with the less experienced group described in the study of Wiese et al.

The present study has several strengths. This is the largest study to date to evaluate unintended durotomy in a broad range of spine surgeries in a group of dedicated spine surgeons. Furthermore, because all of these cases were submitted during

TABLE 6. Summary of Prior Reports of the Incidence of Durotomy in Spine Surgery[a]

Author	Patient Population	Cases, n	Durotomy, % (n/N)	Comments
Mayfield et al, 1975[34]	Laminectomy	1408	0.3	This is the CSF fistula incidence requiring reoperation
Oppel et al, 1977[55]	Lumbar disk herniation	3038	5.9 (179/3038)	
Jones et al, 1989[4]	Lumbar spine surgery	450	3.7 (17/450)	4 Dural tears were in revision cases
Bertalanffy et al, 1989[9]	Anterior cervical diskectomy	450	0.2 (1/450)	
Stolke et al ,1989[5]	Lumbar disk surgery	481	Microdiskectomy, 1.8 (222); macrodiskectomy, 5.3 (190); revision, 17.4 (69)	
Waisman and Schweppe, 1991[6]	Lumbar spine surgery	151	5.3 (8/151)	Only CSF leaks reported, not durotomy
West et al ,1991[8]	Transpedicular variable screw plate fixation	124	5.4 (7/124)	
Davne et al, 1992[1]	Lumbar transpedicular instrumentation	486 Patients, 553 procedures	Not reported	Occurred "occasionally"; all were repaired primarily.
Wang et al ,1998[7]	Lumbar spine surgery	641	14 (88/641)	2 Persistent CSF leaks with 1 requiring revision; 45 durotomies were in revision cases
Cammisa et al, 2000[11]	All spine surgery	2144	Overall, 3.1 (66/2144): cervical (2 of 422), thoracic (0/7), and lumbosacral (64/1715)	
Rosenberg and Mummaneni, 2001[21]	TLIF at L4-5 and L5-S1	22	4.5 (1/22)	
Wiese et al, 2004[27]	Primary lumbar microscopic disk surgery	Retrospective, 1872; prospective, 90	Experienced (> 500 cases): retrospective, 0.8 (932); prospective, 0 (45); not experienced (50-100 cases): retrospective, 7.3 (930); prospective, 4 (45)	Retrospective arm of study with 1872 patients and 90 prospective controlled patients
Saxler et al, 2005[22]	Lumbar disk surgery	1280	3 (41/1280)	Durotomy patients were more likely to have surgery again, increased back pain, and longer time to return to work
Tafazal and Sell, 2005[23]	Lumbar spine surgery	1549	Primary disk, 3.5 (31/872); revision, 13.2 (14/106); spinal stenosis, 8.5 (48/571)	
Khan et al, 2006[19]	Degenerative lumbar spine surgery	3183	Primary, 7.6 (153/2024); revision, 15.9 (185/1159)	6 Required reoperation
Epstein, 2007[14]	Lumbar multilevel laminectomy with noninstrumented fusion in geriatric patients	110	9 (10/110)	Predictive factors included age (74 vs 69 y), ossification of ligamentum flavum, and presence of synovial cyst
Fountas et al, 2007[16]	Primary ACDF for degenerative disk disease or cervical spondylosis	1015	0.5 (5/1015)	
Than et al, 2007[24]	1-Level PLIF	61	Minimally invasive, 0 (0/32); open, 0 (0/29)	

(Continues)

TABLE 6. *Continued*

Author	Patient Population	Cases, n	Durotomy, % (n/N)	Comments
Wang et al, 2007[25]	Cervical spine surgery for degenerative disease	932 009	0.13	Incidence of durotomy was not reported; this is the CSF leak or fistula rate
Rodriguez-Olaverri et al, 2008[20]	Lumbosacral spondylolisthesis	40	TLIF, 35 (7/20); pedicular transvertebral screw fixation, 5 (1/20)	
Yan et al, 2008[28]	Comparison of PLIF and TLIF	187	PLIF, 0 (0/91); TLIF, 0 (0/96)	
This study, 2009	All spine surgery	108 478	1.6	

[a]ACDF, anterior cervical diskectomy and fusion; CSF, cerebrospinal fluid; PLIF, posterior lumbar interbody fusion; TLIF, transforaminal lumbar interbody fusion.

a recent 4-year period, they likely reflect a relatively current standard of care.

Nevertheless, several limitations of this study remain. There currently is no method to determine the completeness of data submission and the accuracy of reporting; they are dependent on the efforts of the participants. Thus, there are limitations to the data integrity, as there is with any large data set. All members of the SRS are concerned about morbidity and mortality and look to the SRS to discern new trends and to identify risks and risk factors for significant complications. There have been 2 previously published articles using the SRS morbidity and mortality database. Because complications like new neurological deficits from surgery for adolescent idiopathic scoliosis are rare, it takes large data sets to capture these events. A similarly rare complication, the occurrence of acute ischemic optic neuropathy associated with prone spine procedures, was also evaluated through the SRS.[12,54]

Although the reported incidence of durotomy likely represents a lower estimate than what would be shown in a prospective clinical trial, it is our opinion that durotomy is probably reliably reported within the database. It typically occurs acutely and has the full attention of the treating surgeon when it occurs. So we suspect that recall bias in the reporting is limited, but we cannot prove or disprove this opinion.

Although it is possible that this data set may have the bias of selective reporting, we believe that it represents a cross section of practicing clinicians. Although there is the possibility that it may have the bias of having greater reporting from younger surgeons in their candidate period and could represent somewhat of a worst-case analysis, there are very senior surgeons reporting as well. In addition, although data are reported prospectively, this is a retrospective study and thus is subject to the weaknesses inherent to such investigations, including confounding variables and reporting bias.

CONCLUSION

Durotomy occurs in at least 1.6% of spinal surgeries, even among experienced spine surgeons. The incidence varies with age, pathology, surgical approach, and operative variables. Although new neurological deficit associated with durotomy occurred in the minority of cases, the present series demonstrates a statistically significant association between these 2 complications. Our data provide general benchmarks of durotomy rates based on preoperative diagnosis and surgical and clinical parameters and serve as a basis for ongoing efforts to improve safety of care.

Disclosure

The authors have no personal financial or institutional interest in any of the drugs, materials, or devices described in this article.

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COMMENT

This group of authors has mined the incredibly large database of spine surgeries compiled by the Scoliosis Research Society. The authors correctly point out that the major weakness of this study, despite its impressively large numbers, is that data reporting was largely voluntary and there was no way to independently validate the completeness or accuracy of data submitted. It would have been interesting to know the number of surgeons whose data were included and the range of durotomy incidence reported by individual surgeons. The database did not specify how many of these durotomies were detected at the time of surgery or whether some became evident later as low-pressure headaches, cerebrospinal fluid leaks, pseudomeningocele, etc.

Most experienced spine surgeons will not be surprised that they report higher frequencies of durotomy in degenerative spine disease, lumbar surgery, revision surgery, and interbody vs lateral spine fusions or that the greater frequency of durotomy in very elderly patients likely was due to the greater frequency of degenerative disease. I was personally somewhat surprised that the incidence was not higher in patients with lumbar spinal stenosis. Interestingly, although a total of 16 502 cases were performed for "revision surgery," only 624 of these cases were performed for lumbar postlaminectomy syndrome ($<$ 4% of the revisions).

This study reports that 4.5% of patients with unintended durotomy developed a postoperative neurological deficit, 3 times greater than the frequency in patients without durotomy. Only 2 of the 75 durotomy patients with deficits available for follow-up failed to improve, but 61% of those with deficits recovered only partially, and the patient with "complete cord injury" did not recover. Unfortunately, their database

did not include whether there were cases with meningitis or pseudomeningocele, both of which are definitely potential complications of durotomy and by themselves may cause new postoperative deficits, or whether the extent or severity of the durotomy influenced the frequency of associated neurological deficits. The incidence of aggravated pain, with or without deficit, was not stated. Likewise, the mechanism of injury that caused the durotomy was not stated but undoubtedly influenced these bad results.

If we can accept the accuracy of the data presented, the incidence of unintended durotomy is commendably low in this series. Granted that most of these surgeons are experienced spine surgeons, most having undergone spine fellowships, these data should serve as a benchmark for surgeons less experienced and less dedicated to spine surgery. I would add, however, a comment that I have frequently made to my residents, "Creating an unintended durotomy without injuring neurological tissue—especially in an elderly patient with adherent lumbar dura thinned by spinal stenosis—is not really a complication, but failing to recognize that you have created a durotomy and not repairing it is much more problematic."

Harold Wilkinson
Boston, Massachusetts

Fourth and Sixth Cranial Nerve Injury After Halo Traction in Children: A Report of Two Cases

Emma Pinches, BSc (Hons),[a,b] Dominic Thompson, FRCS,[c] Hilali Noordeen, FRCS,[d] Alki Liasis, PhD,[a,b] and Ken K. Nischal, FRCOphth[a,b]

Background: Spinal traction is the application of a longitudinal force to the spinal column as a means of stabilizing a damaged or abnormal spine. Although not well documented in the ophthalmic literature, complications include cranial nerve palsies, with the sixth nerve being most commonly affected. Fourth nerve palsies have not previously been reported to our knowledge. We present 2 cases of combined fourth and sixth palsies after cervical traction. ***Methods:*** Retrospectively, we reviewed the ophthalmic findings in 2 children with diplopia after spinal traction. ***Results:*** Case 1 suffered a traumatic rotatory atlantoaxial subluxation and underwent halo traction. Case 2 required traction to correct a scoliosis secondary to osteogenesis imperfecta. In both cases, sixth nerve palsies were apparent soon after traction. Careful orthoptic examination revealed additional fourth nerve involvement. After 3 months, both cases showed partial resolution of the cranial nerve injuries. ***Conclusions:*** Cranial nerve injury may occur with spinal traction. Fourth nerve palsy may be underreported because of masking by a coinciding sixth nerve palsy. (J AAPOS 2004;8:580-585)

Spinal traction is the application of a longitudinal force to the axis of the spinal column as a means of stabilizing or altering the relative positions of damaged parts of the spine. Such spinal damage may be the result of a developmental defect or may result from trauma. A distractive (upward) force is applied to the skull while the rest of the body is held in place. To ensure success, it is important that the device is firmly attached to the skull. Historically, this was achieved using a variety of straps, hooks, pins and tongs, each with its own particular complications, ranging from pressure sores to penetration of the skull.

In halo traction, a ring is attached directly to the skull via four pins and weights are gradually added until the desired distractive force is achieved.[1] The main complications of halo traction relate primarily to the skull pins and include superior migration of the halo to an inappropriate position, skin necrosis, and infection of the pin sites. However, more serious neurological injury, such as cranial nerve palsy, also may occur.[2-8]

We report 2 cases of combined fourth and sixth nerve palsies that follow a period of cervical spinal halo traction in children. To the best of our knowledge, fourth nerve palsy has not previously been reported as a complication of spinal traction.

CASE REPORTS

Case 1

A 9-year-old girl was struck from behind while playing on a slide and sustained a traumatic rotatory atlantoaxial subluxation (Figure 1). Consequently, she developed a fixed abnormal head posture of face turn to the left and head tilt to the right. Her parents did not notice any abnormality in the position or movement of the eyes at this time, and she did not complain of diplopia. A 6-month period of conservative management in her native country yielded no improvement. She was transferred to our center, where manipulation under anesthetic was performed, and she was placed in halo traction with a body jacket. This reduced the subluxation satisfactorily (Figure 2).

Immediately after surgery, the child reported diplopia and was referred for an ophthalmic opinion. On examination, vision was found to be 6/6 in either eye, color vision was full, pupils were normal, and visual fields were full to confrontation. A small amount of anisometropia was found on cycloplegic refraction (RE +0.25 LE +0.75), but the fundi and media were unremarkable. Examination of ocular motility revealed bilateral sixth nerve paresis and marked bilateral fourth nerve palsies, left greater than right. The Bielchowsky

From the Departments of Ophthalmology,[a] Great Ormond Street Hospital for Children, London, United Kingdom, Neurosurgery,[c] Great Ormond Street Hospital for Children, London, United Kingdom, and Orthopaedics,[d] Great Ormond Street Hospital for Children, London, United Kingdom, and Department of Visual Sciences,[b] Institute of Child Health, University College London, London, United Kingdom.
Submitted August 6, 2003.
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Reprint requests: Ken K. Nischal, FRCOphth, Department of Ophthalmology, Great Ormond Street Hospital for Children, Great Ormond Street, London WC1N 3JH, United Kindgom,e-mail: kkn@btinternet.com.

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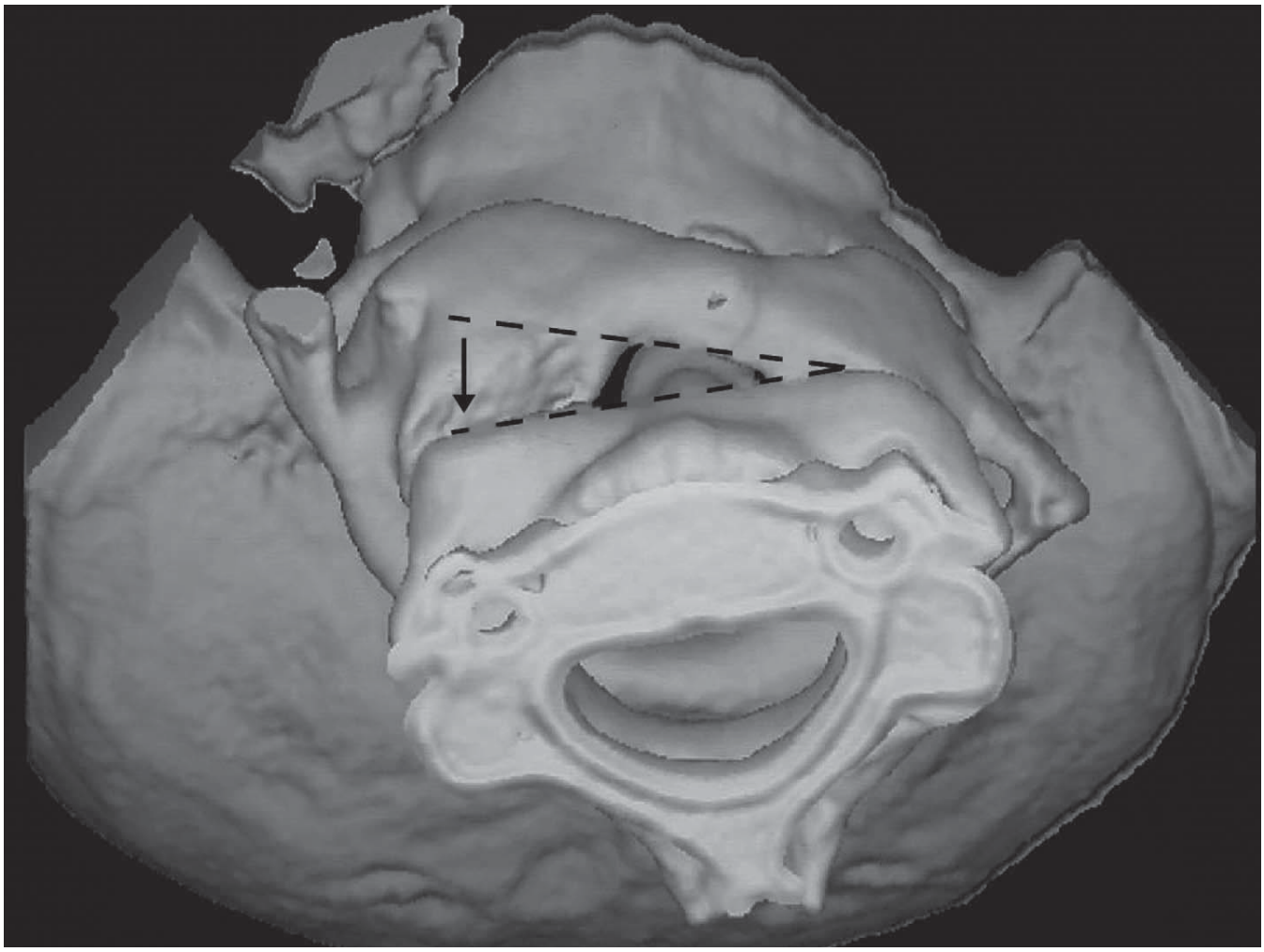

FIG 1. Inferior view of CT 3-dimensional reconstruction of the traumatic rotatory atlantoaxial subluxation in Case 1. The separation of the inferior articular facet of the atlas from the superior articular facet of the axis can clearly be seen (arrow).

head tilt test was performed despite the halo body jacket and found to be positive on head tilt left. To exclude brainstem pathology, a magnetic resonance imaging scan was performed and was found to be normal. Figure 3 shows the eye movements 1 week later, when the child was well enough to attend the audiovisual department. Partial resolution of the fourth nerve palsies had already occurred, but underactions of the superior oblique muscles (greater in the left eye) were still evident.

The halo body jacket was removed 3 months later and replaced with a cervical collar. The child's head posture was now normal. The bilateral sixth nerve palsies had almost resolved, and the patient was binocular in the primary position. However, partial bilateral fourth nerve palsies were still present.

Case 2

A 15-year-old girl with osteogenesis imperfecta developed a secondary scoliosis. Her orthopedic surgeon felt that her condition was likely to progress and therefore surgical intervention was recommended. An anterior discectomy was performed, followed by application of a Stryker traction frame. A week later, posterior spinal fusion of T6–T12 was conducted.

Two weeks after the traction was commenced, the patient complained of horizontal, vertical, and torsional diplopia. On direct questioning, she clearly recalled the onset of the diplopia upon regaining consciousness after application of the traction but failed to mention it until 2 weeks later. The neurology team noted a complete right sixth nerve palsy and, to exclude brainstem pathology, a computed tomography (CT) scan of the brain was performed. This revealed basilar invagination (documented on a previous magnetic resonance imaging scan) but no neural compression or hydrocephalus.

Examination by an ophthalmologist revealed vision of 6/6 in either eye, full color vision and normal fundi. Examination of ocular motility confirmed the complete

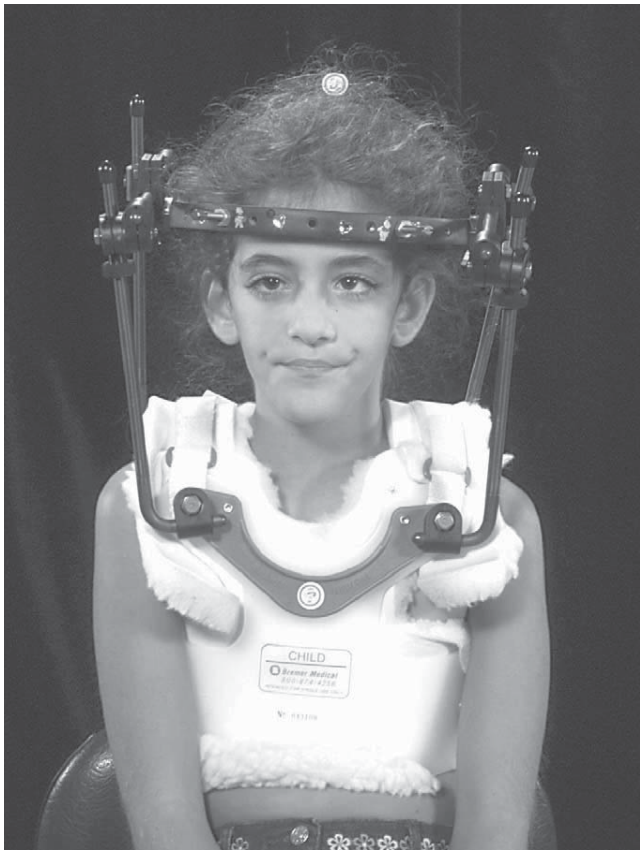

FIG 2. Case 1 wearing a halo body jacket to stabilize the head posture achieved by manipulation of the vertebrae and a period of halo traction. Note the left esotropia and hypertropia caused by bilateral sixth nerve paresis with fourth nerve involvement.

right sixth nerve palsy but also revealed an upshoot of the right eye on left gaze, with an underaction of the right eye on depression in left gaze, consistent with right fourth nerve paresis (Figure 4). Excyclotorsion of the fundus of the right eye was also noted. The Bielchowsky head tilt test was not possible due to the severe scoliosis. In addition, the assessment was limited due to pain and, consequently, torsion was not measured formally. Nevertheless, there was no elevation deficit or ptosis to suggest a superior rectus paresis to account for the torsion, and the CT scan revealed normal positioning of the extraocular muscles. Therefore, we conclude this was indeed a right fourth nerve paresis.

A month later, the palsies had partially resolved, and a Fresnel prism was fitted to relieve residual diplopia. By 2-month follow-up, there had been further resolution and the patient was free of symptoms.

DISCUSSION

Cranial nerve lesions are well-recognized complications of spinal traction, with a reported incidence of 0.07%.[5] Palsies of the abducens, glossopharyngeal, vagus, and hypoglossal nerves have all been reported after traction in the neurosurgical and orthopedic literature.[2-8] However, to the best of our knowledge, trochlear nerve palsy of this etiology has not previously been reported.

Although not well documented in the ophthalmic literature, the most commonly affected cranial nerve is the sixth nerve.[8] Despite the undoubtedly important concerns over the general condition of the patient, which required traction in the first place, the esotropia resulting from the VI nerve dysfunction may cause troublesome diplopia.

The pathophysiological mechanism behind cranial nerve injury during spinal traction is not well understood.[2,6,8] Possibilities include neuropraxia injury, ischemia secondary to edema, direct bony compression, or a combination of these factors.

Neuropraxia is characterized by a focal conduction block at the site of the lesion, with normal neuronal conduction distally.[9] This may occur during halo traction due to stretching or kinking of the nerve, or a change in its position[2,6,7,10] and generally resolves rapidly once the distraction force is removed. Wilkins and MacEwan[8] report that the sixth nerve is most vulnerable to kinking as it passes over the bony ridge of the petrosphenoidal junction, in the cavernous sinus.

An alternative explanation is that the traction may cause edema, which compromises the nutrient blood vessel supply of either the nerve itself or its nucleus. This may lead to localized ischemia and reduced nerve function.[6,7,11] Again, when the causative force is removed and the ischemia subsides, there may be resolution of the neurological deficit. Ginsberg and Bassett[3] described a case of hypoglossal nerve palsy after halo traction that they treated with corticosteroids to reduce the possibility of nerve compression due to edema. However, they concede that the significance of this treatment in the resolution of the palsy is unclear.

Cases of atlantoaxial and atlanto-occipital rotatory subluxation are unusual[12] but are well documented in the pediatric neurosurgical literature. Traumatic atlanto-occipital dislocation is a similar but often fatal injury, which also is rare.[13] In a previously documented case of traumatic atlanto-occipital dislocation, the most probable mechanism of cranial nerve injury was reported to be neuropraxia caused by traction of the midbrain.[11] Therefore, this may be the most plausible explanation for the injury sustained by Case 1.

Osteogenesis imperfecta is an inherited condition characterized by bone fragility attributed to low bone mass, abnormal organization of bone tissue, and anomalous bone size and shape.[14] The condition is classified into 4 types on the basis of clinical and genetic features. Case 2 had the most severe form of osteogenesis imperfecta (type III), and the abnormal spine may have mechanically compromised the blood supply to the midbrain during the traction. In addition to any edema, this may have caused localized ischemia and, consequently, reduced nerve function.

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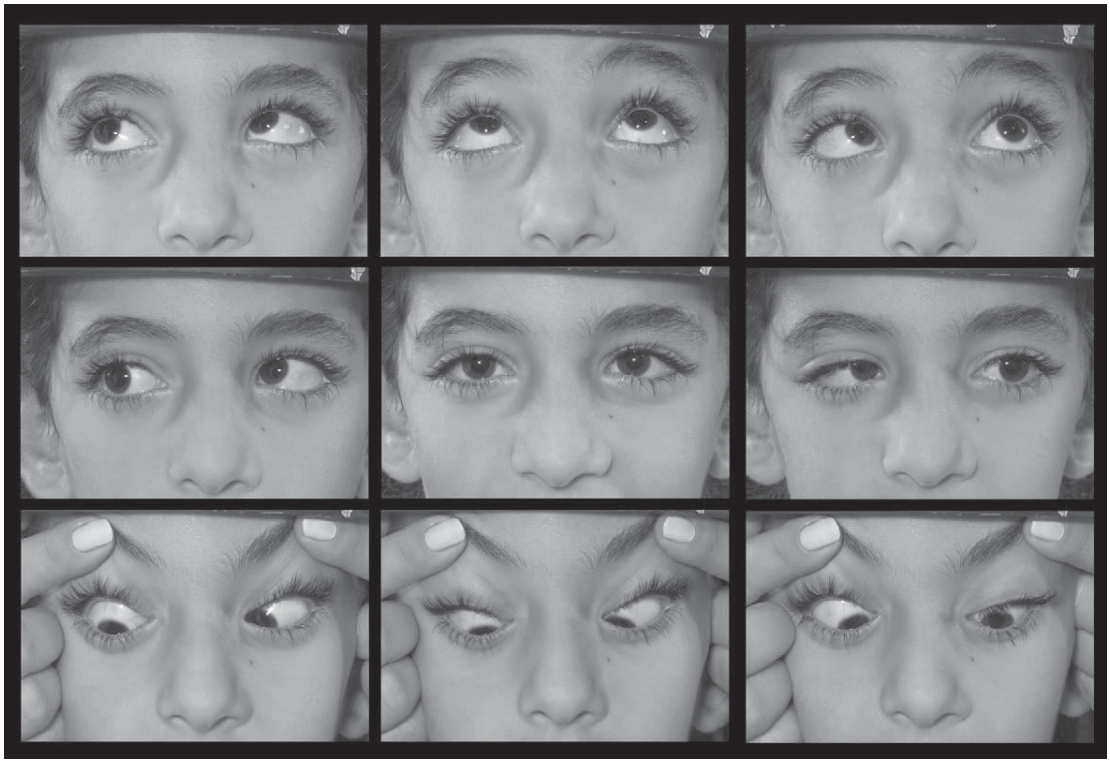

FIG 3. Ocular movements of Case 1 at 1 week after traction showing bilateral sixth and fourth nerve palsies.

Many authors state that nerve palsies that occur after traction are transient in nature and usually resolve fully within 6-10 weeks.[3-6,8] Other authors have reported that these lesions may be permanent.[11,15] In studies of isolated traumatic sixth nerve palsies, recovery rates vary from 27% to 84% in unilateral cases, and from 12% to 38% in bilateral cases.[16,17] This variance, especially in unilateral cases, may reflect heterogeneity in the pathomechanism of injury. Non-recovery of acute traumatic sixth nerve palsy has been shown to be associated independently with complete sixth nerve palsy (inability to abduct past the midline at presentation) and with bilaterality.[18]

The nerve palsies of the children described in this report have improved but have not fully resolved after 12 months of follow-up. This may be attributed to the bilateral nature of the first case and the complete sixth nerve palsy of the second case. Alternatively, the theory of cranial nerve compromise at the level of the nerve nucleus may account for the incomplete resolution of these palsies.

Traumatic atlantoaxial dislocation results in spasm of the neck muscles, which acts as a protective mechanism by immobilizing the neck.[11] This often leads to a rigid abnormal head posture, as seen in Case 1. It has been documented that under a paralytic anesthetic this spasm of the neck muscles is lost.[11] Consequently, traction at this time may induce neuropraxia injury. In Case 1, during the initial 6 months of conservative management for the neck injury, the child's parents are certain that her eyes were straight and she was visually asymptomatic. Immediately after application of the traction device, she developed an obvious esotropia and complained of diplopia. This strongly indicates that the cranial nerve palsies occurred as a result of the traction and not the initial trauma. We are confident, therefore, that the abnormal head posture seen in this case was a consequence of the traumatic rotatory atlantoaxial subluxation, and was not caused by a pre-existing fourth nerve palsy acquired at the time of the initial injury.

An extensive search of the literature has revealed no previous reports of fourth nerve palsy after traction. This may be the result of masking of fourth nerve involvement in many instances by coinciding sixth nerve palsy. It is often difficult to detect fourth nerve paresis in the presence of esotropia by examining ocular movements alone. Diagnosis may be hampered further in children when a Hess chart is not possible, either because of age or general health after surgery. Recent traction and residual scoliosis may also make the Bielchowsky head tilt test difficult to perform. Additionally, in young children, questions regarding subjective torsion may be inconclusive.

In the presence of a large angle esotropia, the superior oblique acts primarily as a depressor and the secondary action of intorsion may not be seen. However, in smaller angles of esotropia, as in the 2 cases reported here, the affected eye should be observed closely for signs of intorsion as the eye moves into the field of action of the

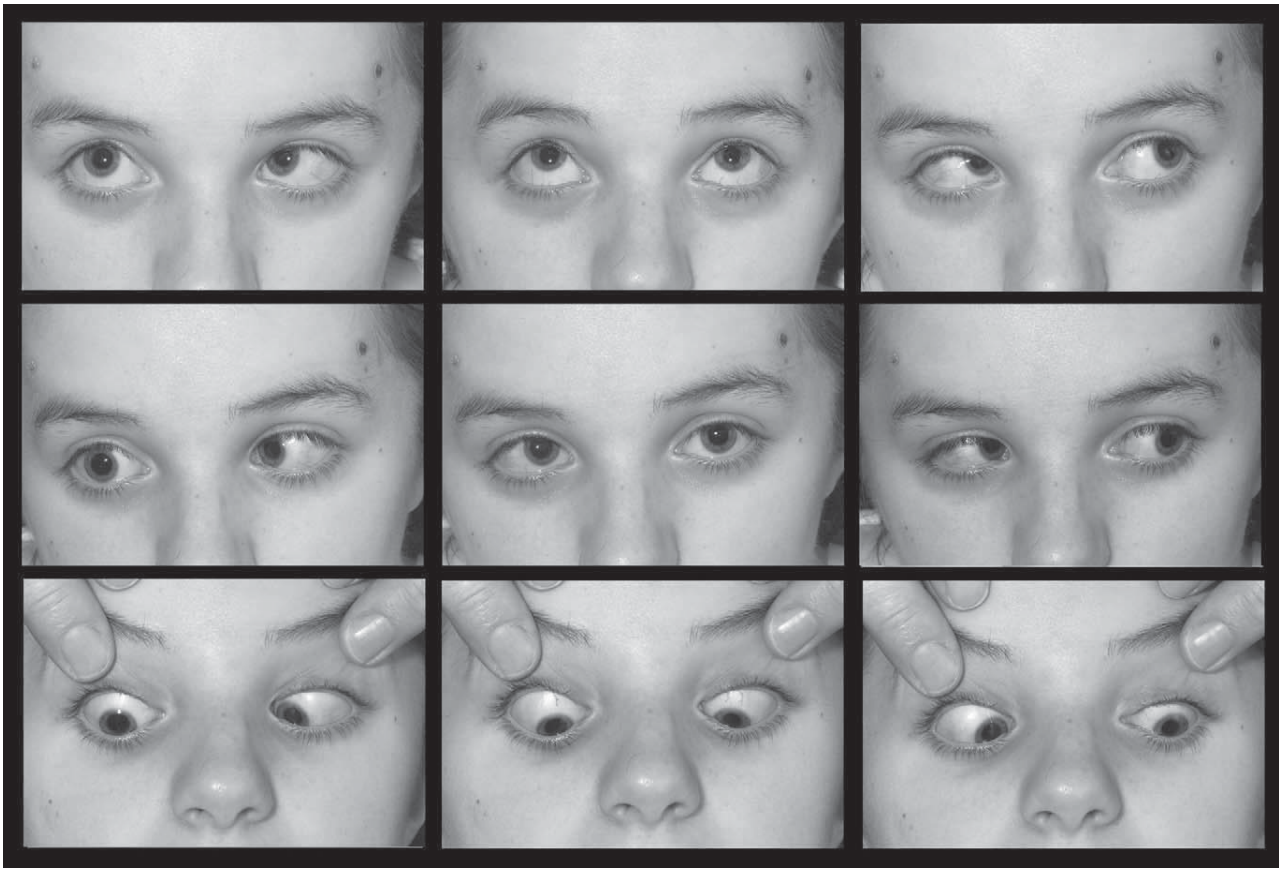

FIG 4. Ocular movements of Case 2 at 3 weeks after traction showing right sixth and right fourth nerve palsies.

superior oblique. The relative positions of conjunctival vessels often make useful landmarks when looking for torsion. Absence of intorsion of the globe in this position of gaze suggests fourth nerve involvement. Detection of a fourth nerve palsy may also be aided by viewing the fundus of the suspected eye for signs of excyclotorsion, as described in Case 2.

Although techniques are available to monitor extraocular muscle function using intraoperative electromyography,[19,20] the incidence of cranial nerve injury after spinal traction is so low (0.7 cases per 1,000[5]) that the expense of electromyogram monitoring of nerve function during application of halo traction cannot be justified.

In conclusion, cranial nerve palsies may occur as complications of spinal traction. The most frequently reported nerve palsy involving ocular motility is that of the sixth nerve. We describe 2 cases of combined sixth and fourth nerve palsies after traction in children. This may be the first report of fourth nerve dysfunction after halo traction. Careful assessment of ocular motility for absence of incyclotorsion in contralateral depression and examination of the fundus for signs of excyclotorsion may reveal fourth nerve palsies which are commonly masked by the large angle esotropia caused by an concurrent sixth nerve palsy. This may explain why fourth nerve palsies of this etiology have not, to the best of our knowledge, previously been reported in the literature.

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An Eye on the Arts – The Arts on the Eye

"Please, don't talk," said the man. "I can see your light now."

"Your personal light," he said, realizing that she didn't know what he was talking about.

Her personal light. Well, how wrong could he be, that innocent painter, who obviously hadn't learned much about life in his thirty-odd years. But then, as everyone knows, women mature more quickly than men, and although Maria might not spend sleepless nights pondering her particular philosophical problems, she knew one thing: she did not have what that painter called "light" and which she took to mean "a special glow." She was just like everyone else, she endured her loneliness in silence, tried to justify everything she did, pretended to be strong when she was feeling weak or weak when she was feeling strong, she had renounced love and taken up a dangerous profession, but now, as that work was coming to an end, she had plans for the future and regrets about the past, and someone like that doesn't have a "special glow." That must just be his way of keeping her quiet and still and happy to be there, playing the fool.

Personal light, indeed. He could have said something else, like "you've got a lovely profile."

How does light enter a house? Through the open windows. How does light enter a person? Through the open door of love. And her door was definitely shut. He must be a terrible painter; he didn't understand anything.

"I've finished," he said and started collecting up his things.

Maria didn't move. She felt like asking if she could see the painting, but that might seem rude, as if she didn't trust what he had done. Curiosity, however, got the better of her; she asked and he concurred. He had painted only her face; it looked like her, but if, one day, she had seen that painting, not knowing who the model was, she would have said that it was someone much stronger, someone full of a "light" she didn't see reflected in the mirror.

—Paulo Coelho (from *Eleven Minutes*)

Chapter 10

CONCLUSION

Advances in Surgical Techniques for EOSD

Early-onset spinal deformity (EOSD) continues to be an enigma to healthcare professionals. The pathophysiology of thoracic insufficiency syndrome (TIS) and the importance of pulmonary function and maturation are only recently being understood comprehensively. Genetic studies are underway to alter or regulate the gene expression responsible for causing scoliosis. These studies are still in their infancy, and they are currently restricted to animal experiments. A better understanding of the natural history in early-onset scoliosis secondary to neuromuscular disorders (EOS-NMD) will influence and guide treatment options. The advent of magnet driven growing rods (MdGR) has been a boon, especially to growing children, and I was the first in the world to report an improvement in pulmonary function with a marked reduction in chest infections in six children, which was presented at the Scoliosis Research Society (SRS) 2013 annual meeting as a podium presentation and published in Spine (P'hila 1976) this year.

MdGRs appear to be a promising option for EOSD, as an early definitive spinal fusion is unacceptable with a disproportionate upper segment: lower segment ratio and a premature arrest of pulmonary maturation. However, the current first generation MdGRs corrects only the spinal deformity, and do not address any chest cage deformation (for example, the '*parasol*' deformity of spinal muscular atrophy [SMA]). I am currently working on innovative surgical construct options, and exploring avenues to address both chest cage and spinal deformity correction with the newer second generation MdGRs.

In summary, the treatment of EOSD has evolved by leaps and bounds over the past decade, from mere observation and bracing, to complex surgeries to prolong life, enhance caregiver satisfaction, and optimise outcomes. My surgical practice has also evolved from conventional growing rods (CGRs) to MdGRs. Highly satisfied parents and caregivers continue to inspire me to pursue excellence in research, and test newer and more innovative surgical options to improve quality of life.

Advances in Surgical Techniques for EOSD

The magnet driven growing rods (MdGRs) have recently been approved for early-onset spinal deformities (EOSD) in North America by the United States Food and Drug Administration (USFDA). It is considered to be a *game changer* amongst distraction based devices in surgical management of EOSD. My published papers (nos. 6 and 7) cited earlier in chapter no. 5 (*Innovations and Recent Advances in Early-onset Scoliosis*) was a part of meta-analysis undertaken by the National Institute of Clinical Excellence (NICE) which concluded that MdGRs were:-

- Associated with a statistically significant (*p=0.03-.004*) lower incidence of infection rate than conventional growing rods (CGRs)
- More cost-effective in children who had three or more years of remaining growth potential in comparison to CGRs.
- The improvement in Cobb angle and height gain (T_1-S_1 length) were similar between the two devices (i.e. MdGRs vs. CGRs).

Despite tremendous advances in technology and understanding about EOSD over the past five years, adequate scope exists for further improvement. Better understanding of the natural history of various sub-etiologies of EOSD and its effect on pulmonary function is desired. The acceptable standard of care for EOSD is not an early spinal fusion but growth guided surgeries either by rib or spine based distraction devices. Despite NICE's recommendation of advocating the use of MdGRs for children with more than three years of remaining growth potential, many syndromic and congenital spinal deformities have bone age that lags behind chronological age. Comprehensive evaluation and individualised decision-making practicing fiduciary relationship with parents and care-givers is desired to optimise outcomes in such vulnerable children. More good quality multi-centric studies with high level of evidence (LoE) are desired to draft grades of recommendations (GoR) to guide the surgical community in our commitment to evidence-based practice of paediatric spinal deformity surgery.

Advances in Surgical Techniques for EOSD

My vision in pursuit of excellence would be to undertake:

- A long-term (i.e. until skeletal maturity or definitive spinal fusion whichever is later) Level of Evidence (LoE) II therapeutic study (prospective comparative clinical study) between MdGRs and CGRs to establish if either of them is superior over the other.
- A long-term LoE I prognostic study evaluating the pulmonary function before and after MdGR insertion esp. in neuromuscular, syndromic and congenital sub-etiologies of EOSD.
- A LoE I prognostic study comparing IQ, cognitive performance, scholastic grades and aptitude before and after growing rods insertion (i.e. MdGRs and CGRs) in early-onset idiopathic scoliosis.
- A LoE I economic / decision-making modeling analysis study to establish the cost-utility of CGRs vs. MdGRs.

Appendix - I

RNOH, Stanmore - Current Research Update

Magnet Driven Growing Rods (MdGRs)

as on April 2014

Advances in Surgical Techniques for EOSD

I have now operated and inserted magnet driven growing rods (MdGRs) on at least one hundred children with early-onset spinal deformity (EOSD) of all etiologies (i.e. idiopathic, congenital, neuromuscular and syndromic). My pioneering work has been extensively cited in literature by other investigators. Twenty five patients operated at Royal National Orthopaedic Hospital, Stanmore had completed minimum follow-up of one year by March 2014. The summary file of their clinico-radiological results depicting improvement in Cobb angle and height gain (T_1-S_1 length) is illustrated in next page. The same study has been accepted as a podium presentation at Société Internationale de Chirurgie Orthopédique et de Traumatologie (SICOT) Triennial Orthopaedic world congress later this year (Nov 2014) in Rio de Janeiro, Brazil. Seventeen patients in this study group had completed a minimum follow-up of 2 years and an abstract reporting their clinic-radiological results with complications has been submitted for the American Academy of Orthopaedic Surgeons (AAOS) annual congress 2015.

My research on MdGRs was also instrumental for the National Institute of Clinical Excellence (NICE) to produce the position statement and medical technology guidance document (no.18) on *The MAGEC system for spinal lengthening in children with Scoliosis*. The relevant NICE documents are incorporated as appendix III. The United States Food and Drug Administration, (USFDA - i.e. NICE equivalent of UK) has also approved MdGRs for EOSD. My collaborative work with other multi-centric international group of surgeons from USA, Europe and Hong Kong was contributory in getting the FDA approval.

I have trained other spinal surgeons nationally (in the UK) and internationally (Europe, North America, Middle-east, Far-east and Indian sub-continent) by teaching them how to insert MdGRs, imparting practical tips and sharing my knowledge. I am an invited speaker at the Scoliosis Research Society (SRS) annual meeting 2014 at Alaska, USA wherein I will be delivering a master class lecture on *MAGEC rods for early-onset scoliosis* to global audience. I also regularly lecture on *MdGRs for EOSD* as an invited speaker at global scientific meetings.

Clinico-radiological parameters of all 25 patients with early-onset scoliosis treated by MdGR insertions with minimum follow-up of one year

Patient Number	Diagnosis	Age at Sx	Follow-up duration	No of Rods	Cobb angle First visit	Cobb angle Imm pre-op	Cobb angle Imm post-op	Cobb angle latest f/u	T1-S1 Lgt First visit	T1-S1 Lgt Imm pre-op	T1-S1 Lgt Imm post-op	T1-S1 Lgt latest f/u	T1-S1 Lgt Gain
1	Congenital scolisois	12.49	4.25	1	50	58	40	28	351	359	368	392	24
2	Neurofibromatosis 1	6.49	4.16	1	67	76	46	41	303	313	354	378	24
3	Juvenile idiopathic	11.25	4.16	1	27	65	30	30	360	368	375	435	60
4	Ehler-danlos syndrome	5.11	4.16	1	63	95	70	82	217	242	287	245	-42
5	William syndrome	10.67	4.08	2	60	71	48	45	267	304	328	342	14
6	SMA - II	5.43	3.92	2	55	60	40	32	300	310	335	360	15
7	Congenital insensitivity to pain	10.91	3.92	2	64	50	41	50	256	317	330	343	13
8	Juvenile idiopathic	12.28	3.92	2	46	60	37	51	346	377	373	400	27
9	SMA - II	6.99	3.92	2	67	87	50	58	230	252	297	325	28
10	Cerebral palsy	10.59	3.00	2	44	65	52	65	340	367	373	373	0
11	Noonan syndrome	4.72	3.00	1	32	50	12	6	205	215	237	255	18
12	Prader-Wili syndrome	10.00	2.83	2	59	74	43	60	303	306	375	380	5
13	Prader-Wili syndrome	11.80	2.67	2	67	70	35	27	330	340	370	410	40
14	Autistic spectrum disorder	12.15	2.58	2	56	90	30	32	350	340	415	440	25
15	Juvenile idiopathic	10.95	2.25	2	55	57	25	25	395	400	433	487	54
16	Soto's syndrome	3.59	2.08	2	50	67	30	27	245	262	300	318	18
17	Cerebral palsy	8.71	2.08	2	52	85	50	48	270	260	302	318	16
18	VATER syndrome	8.50	1.92	1	58	76	77	60	270	282	290	305	15
19	Cerebral palsy	12.03	1.92	2	70	75	60	60	280	300	330	360	30
20	Congenital scolisois	10.36	1.83	1	70	85	64	60	243	270	335	340	5
21	Juvenile idiopathic	10.63	1.58	2	42	57	47	50	345	400	417	435	18
22	Rett syndrome	11.17	1.50	2	68	76	50	52	370	376	385	410	25
23	Anderson-Tawil syndrome	12.09	1.42	2	42	70	52	65	295	325	357	366	9
24	Cerebral palsy	11.07	1.16	2	20	44	13	15	262	350	373	382	9
25	Cerebral palsy	12.29	1.08	2	40	65	48	42	265	280	295	307	12
Mean		**9.69**	**2.77**		**52.96**	**69.12**	**43.6**	**44.45**	**295.92**	**316.6**	**345.36**	**364.24**	**18.88**

Appendix - II

Surgical Distraction Device Patent

PCT/GB 01/01668

INVESTOR IN PEOPLE

The Patent Office
Concept House
Cardiff Road
Newport
South Wales
NP10 8QQ

RECD 11 MAY 2001
WIPO PCT

I, the undersigned, being an officer duly authorised in accordance with Section 74(1) and (4) of the Deregulation & Contracting Out Act 1994, to sign and issue certificates on behalf of the Comptroller-General, hereby certify that annexed hereto is a true copy of the documents as originally filed in connection with the patent application identified therein.

In accordance with the Patents (Companies Re-registration) Rules 1982, if a company named in this certificate and any accompanying documents has re-registered under the Companies Act 1980 with the same name as that with which it was registered immediately before re-registration save for the substitution as, or inclusion as, the last part of the name of the words "public limited company" or their equivalents in Welsh, references to the name of the company in this certificate and any accompanying documents shall be treated as references to the name with which it is so re-registered.

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Signed P Mahoney

Dated 3 May 2001

PRIORITY DOCUMENT

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Patents Act 1977
(Rule 16)

The Patent Office

1/77

THE PATENT OFFICE 13 APR 2000 RECEIVED BY HAND

Request for grant of a patent

(See the notes on the back of this form. You can also get an explanatory leaflet from the Patent Office to help you fill in this form)

The Patent Office

Cardiff Road
Newport
Gwent NP9 1RH

1. Your reference

DCW/VSW

13 APR 2000

17APR00 E529454-7 D00016
P01/7700 0.00-0009107.4

2. Patent application number
(The Patent Office will fill in this par

0009107.4

3. Full name, address and postcode of the or of each applicant *(underline all surnames)*

University College London
Gower Street
London WC1E 4BT

Patents ADP number *(if you know it)*

If the applicant is a corporate body, give the country/state of its incorporation

7785200 2

4. Title of the invention

"SURGICAL DISTRACTION DEVICE"

5. Name of your agent *(if you have one)*

"Address for service" in the United Kingdom to which all correspondence should be sent *(including the postcode)*

Brookes & Martin

High Holborn House
52/54 High Holborn
London WC1V 6SE

Patents ADP number *(if you know it)*

471001

6. If you are declaring priority from one or more earlier patent applications, give the country and the date of filing of the or of each of these earlier applications and *(if you know it)* the or each application number

Country	Priority application number *(if you know it)*	Date of filing *(day / month / year)*

7. If this application is divided or otherwise derived from an earlier UK application, give the number and the filing date of the earlier application

Number of earlier application	Date of filing *(day / month / year)*

8. Is a statement of inventorship and of right to grant of a patent required in support of this request? *(Answer 'Yes' if:*

a) any applicant named in part 3 is not an inventor, or
b) there is an inventor who is not named as an applicant, or
c) any named applicant is a corporate body.
See note (d))

yes

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9. Enter the number of sheets for any of the following items you are filing with this form. Do not count copies of the same document

Continuation sheets of this form	
Description	7
Claim(s)	1
Abstract	
Drawing(s)	2 + 2

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Priority documents	
Translations of priority documents	
Statement of inventorship and right to grant of a patent *(Patents Form 7/77)*	
Request for preliminary examination and search *(Patents Form 9/77)*	1
Request for substantive examination *(Patents Form 10/77)*	
Any other documents *(please specify)*	

11. I/We request the grant of a patent on the basis of this application.

Signature Brookes & Martin Date 13 April 2000
BROOKES & MARTIN

12. Name and daytime telephone number of person to contact in the United Kingdom

0171 242 9631 - David C. Woodcraft

Warning

After an application for a patent has been filed, the Comptroller of the Patent Office will consider whether publication or communication of the invention should be prohibited or restricted under Section 22 of the Patents Act 1977. You will be informed if it is necessary to prohibit or restrict your invention in this way. Furthermore, if you live in the United Kingdom, Section 23 of the Patents Act 1977 stops you from applying for a patent abroad without first getting written permission from the Patent Office unless an application has been filed at least 6 weeks beforehand in the United Kingdom for a patent for the same invention and either no direction prohibiting publication or communication has been given, or any such direction has been revoked.

Notes

a) *If you need help to fill in this form or you have any questions, please contact the Patent Office on 0645 500505.*

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f) *For details of the fee and ways to pay please contact the Patent Office.*

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(71) Applicant(s):
UNIVERSITY COLLEGE LONDON [GB/GB]; Gower Street London WC1E 4BT (GB) *(for all designated states except US)*
BLUNN, Gordon [GB/GB]; 224 North End Bassingbourn Royston North Hertfordshire SG8 5NY (GB) *(for US only)*
PERRY, John [GB/GB]; 27 King Charles Road Shenley Hertfordshire WD7 9MZ (GB) *(for US only)*
NOORDEEN, Hilali [GB/GB]; 42 Addison Road London W14 8JH (GB) *(for US only)*
MESWANIA, Jay [GB/GB]; 65 Tollgate Road Colney Heath St Albans Hertfordshire AL4 0PX (GB) *(for US only)*
COBB, Justin [GB/GB]; 16 Provost Road London NW3 4ST (GB) *(for US only)*

(72) Inventor(s):
BLUNN, Gordon; 224 North End Bassingbourn Royston North Hertfordshire SG8 5NY (GB)
PERRY, John; 27 King Charles Road Shenley Hertfordshire WD7 9MZ (GB)
NOORDEEN, Hilali; 42 Addison Road London W14 8JH (GB)
MESWANIA, Jay; 65 Tollgate Road Colney Heath St Albans Hertfordshire AL4 0PX (GB)
COBB, Justin; 16 Provost Road London NW3 4ST (GB)

(74) Agent(s):
WOODCRAFT, David, Charles; Brookes Batchellor 102-108 Clerkenwell Road London EC1M 5SA (GB)

(54) Title (EN): SURGICAL DISTRACTION DEVICE

(54) Title (FR): DISPOSITIF CHIRURGICAL DE DISTRACTION

(57) Abstract:
(EN): A surgical distraction device is provided for applying extending or tensioning force non-invasively to a patient's skeleton or to an implant which comprises anchoring means for attaching first and second components of the device to a bone or to adjoining bones, said components being connected by a linkage of an extendible length, a magnet connected to the linkage via a reduction gearbox and actuating means located externally of the patient for generating a moving or varying electro-magnetic field, thereby causing the magnet to rotate and the linkage to be extended.

(FR): L'invention concerne un dispositif chirurgical de distraction destiné à appliquer une force de tension ou d'extension, de manière non invasive, sur le squelette d'un patient ou sur un implant qui comprend des moyens d'ancrage pour attacher les premier et deuxième constituants du dispositif à un os ou à des os contigus, ces constituants étant reliés par une liaison de longueur extensible, un aimant étant relié à la liaison au moyen d'une boîte de réduction et de moyens actionneurs situés extérieurement au patient et destiné à générer un champ électromagnétique mobile ou variable, faisant ainsi tourner l'aimant et provoquant l'extension de la liaison.

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European Patent Office (EPO) : AT, BE, CH, CY, DE, DK, ES, FI, FR, GB, GR, IE, IT, LU, MC, NL, PT, SE, TR
African Intellectual Property Organization (OAPI) : BF, BJ, CF, CG, CI, CM, GA, GN, GW, ML, MR, NE, SN, TD, TG
African Regional Intellectual Property Organization (ARIPO) : GH, GM, KE, LS, MW, MZ, SD, SL, SZ, TZ, UG, ZW
Eurasian Patent Organization (EAPO) : AM, AZ, BY, KG, KZ, MD, RU, TJ, TM

PATENT COOPERATION TREATY

PCT

INTERNATIONAL PRELIMINARY EXAMINATION REPORT

(PCT Article 36 and Rule 70)

Applicant's or agent's file reference	FOR FURTHER ACTION	See Notification of Transmittal of International Preliminary Examination Report (Form PCT/IPEA/416)
EEB/AKW		
International application No.	International filing date *(day/month/year)*	Priority date *(day/month/year)*
PCT/GB 01/ 01668	12/04/2001	13/04/2000
International Patent Classification (IPC) or national classification and IPC		
A61B17/72		
Applicant		
UNIVERSITY COLLEGE LONDON et al.		

1. This international preliminary examination report has been prepared by this International Preliminary Examining Authority and is transmitted to the applicant according to Article 36.

2. This **REPORT** consists of a total of 2 sheets, including this cover sheet.

   ☐ This report is also accompanied by ANNEXES, i.e., sheets of the description, claims and/or drawings which have been amended and are the basis for this report and/or sheets containing rectifications made before this Authority (see Rule 70.16 and Section 607 of the Administrative Instructions under the PCT).

   These annexes consists of a total of _____ sheets.

3. This report contains indications relating to the following items:

   I ☒ Basis of the report

   II ☐ Priority

   III ☐ Non-establishment of opinion with regard to novelty, inventive step and industrial applicability

   IV ☐ Lack of unity of invention

   V ☒ Reasoned statement under Article 35(2) with regard to novelty, inventive step or industrial applicability; citations and explanations supporting such statement

   VI ☐ Certain documents cited

   VII ☐ Certain defects in the international application

   VIII ☐ Certain observations on the international application

Date of submission of the demand	Date of completion of this report
13/10/2001	20/03/2002
Name and mailing address of the IPEA/	Authorized officer
European Patent Office D-80298 Munich Tel. (+49-89) 2399-0, Tx: 523656 epmu d Fax: (+49-89) 2399-4465	DEPRUN M Tel. +49-89-2399-2828

INTERNATIONAL PRELIMINARY EXAMINATION REPORT

International application No PCT/ GB 01/ 01668

I. Basis of the report

The basis of this international preliminary examination is the application as originally filed.

V. Reasoned statement under Rule 66.2(a)(ii) with regard to novelty, inventive step or industrial applicability

In light of the documents cited in the international search report, it is considered that the invention as defined in at least some of the claims does not appear to meet the criteria mentioned in Article 33(1) PCT, i.e. does not appear to be novel and/or to involve an inventive step (see international search report, in particular the documents cited X and/or Y and corresponding claim references).

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(71) Applicant *(for all designated States except US)*: **UNIVERSITY COLLEGE LONDON** [GB/GB]; Gower Street, London WC1E 4BT (GB).

(72) Inventors; and
(75) Inventors/Applicants *(for US only)*: **BLUNN, Gordon** [GB/GB]; 224 North End, Bassingbourn, Royston, North Hertfordshire SG8 5NY (GB). **PERRY, John** [GB/GB]; 27 King Charles Road, Shenley, Hertfordshire WD7 9MZ (GB). **NOORDEEN, Hilali** [GB/GB]; 42 Addison Road, London W14 8JH (GB). **MESWANIA, Jay** [GB/GB]; 65 Tollgate Road, Colney Heath, St Albans, Hertfordshire AL4 0PX (GB). **COBB, Justin** [GB/GB]; 16 Provost Road, London NW3 4ST (GB).

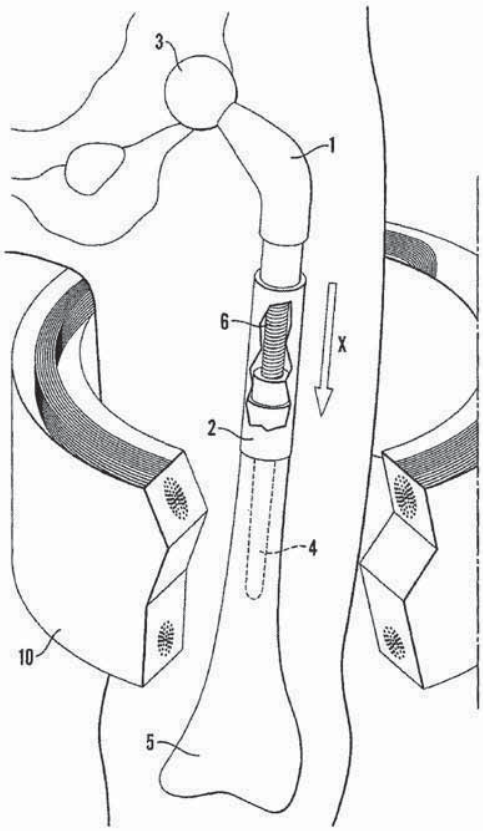
(74) Agent: **WOODCRAFT, David, Charles**; Brookes Batchellor, 102-108 Clerkenwell Road, London EC1M 5SA (GB).

(81) Designated States *(national)*: AE, AG, AL, AM, AT, AU, AZ, BA, BB, BG, BR, BY, BZ, CA, CH, CN, CO, CR, CU, CZ, DE, DK, DM, DZ, EE, ES, FI, GB, GD, GE, GH, GM, HR, HU, ID, IL, IN, IS, JP, KE, KG, KP, KR, KZ, LC, LK, LR, LS, LT, LU, LV, MA, MD, MG, MK, MN, MW, MX,

[Continued on next page]

WO 01/78614 A1

(54) Title: SURGICAL DISTRACTION DEVICE



(57) Abstract: A surgical distraction device is provided for applying extending or tensioning force non-invasively to a patient's skeleton or to an implant which comprises anchoring means for attaching first and second components of the device to a bone or to adjoining bones, said components being connected by a linkage of an extendible length, a magnet connected to the linkage via a reduction gearbox and actuating means located externally of the patient for generating a moving or varying electro-magnetic field, thereby causing the magnet to rotate and the linkage to be extended.

WO 01/78614 A1

MZ, NO, NZ, PL, PT, RO, RU, SD, SE, SG, SI, SK, SL, TJ, TM, TR, TT, TZ, UA, UG, US, UZ, VN, YU, ZA, ZW.

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For two-letter codes and other abbreviations, refer to the "Guidance Notes on Codes and Abbreviations" appearing at the beginning of each regular issue of the PCT Gazette.

SURGICAL DISTRACTION DEVICE

This invention relates to surgical distraction devices and in one form provides a surgical distraction device for exerting an extending or tensioning force to bones and/or implants within a patient's skeleton.

In the treatment of malignant bone cancer it is often necessary to surgically resect part of a bone and to replace it with a prosthesis. Where the patient is a child or adolescent, the skeleton continues to grow and in order to accommodate this the prosthesis requires replacement at intervals. In an effort to reduce the number of revision operations, procedures have been developed for extending the prosthesis *in situ*. Methods of achieving this include the provision of telescopic parts in the prosthesis which are extended by introducing spacers of approximate size between the parts. Devices which have been used for this purpose are described in the paper by Schindler et al, the Journal of Bone and Joint Surgery, Volume 79b, No. 6, November 1997, pages 927 to 937.

While the system described in the above paper is preferable to repetitive revision operations, it still requires surgical access to a part of the prosthesis at intervals which can be frequent, depending on the growth rate of the patient. Since all operations carry finite risks, including the risk of introducing an infection, it is one object of this invention to provide a device which can be used to extend a prosthesis non-invasively.

According to one aspect of the present invention there is provided a surgical distraction device for applying an extending or tensioning force non-invasively to a

patient's skeleton which comprises anchoring means for attaching first and second components of the device to a bone or to adjoining bones, said components being connected by a linkage of variable length, a magnet connected to the linkage via a reduction gearbox and means located externally of the patient for generating a moving or varying electro-magnetic field, thereby causing the magnet to rotate and the linkage to be varied in length.

In practice, the first and second components and its associated linkage, together with the magnet, are surgically introduced and anchored to the bone or bones, for example when fitting a prosthesis or implant.

At intervals when the device requires extension or tensioning, a moving electric field is established around the patient's body in the region of the prosthesis or implant, thereby causing the two components to be pressed apart. Normally, the magnetic field is arranged to rotate in the direction of desired movement of the magnet connected to the linkage.

According to a second aspect of the invention there is provided an extendable endoprosthetic replacement device which comprises first and second components which are connected by a linkage of extendable length, the first component comprising a component of a limb joint and the second component having means for attachment to a resected long bone, a magnet connected to the linkage and means adapted for location externally of the patient for generating a moving electromagnetic field, thereby causing the magnet to rotate and the linkage to be extended.

The distraction device in accordance with the invention has substantial advantages over existing practices such as described in the paper cited above, since it avoids the trauma of further surgical operations, reducing the risk of infection and the introduction of further scar tissue as a result of the operations. Moreover, the device can be extended or tensioned over a longer period than might be practical in the course of a surgical operation. The device can also be extended more gradually and more frequently, which induces an expansion or tensile force, which keeps pace with growth and stimulates biological growth of tissues.

The provision of a reduction gearbox between the magnet and the relatively movable components of the distraction device carries with it important advantages. First it enables a uniform extension force to be applied to the extensible components. Secondly it enables sufficient force to be applied to overcome resistance to extension and to resist any tendency for the device to contract in length when the bone is loaded.

Although a major use of the device in accordance with the invention is the periodic extension of prostheses for the reasons described in the paper cited above, there are a number of other surgical purposes for which the device is of value. One of these is in distraction osteo-genesis. This treatment involves tensioning of bone in order to stimulate its growth. The conventional procedure involves provision of an external frame, e.g. around a limb and pins are anchored to the bone at a spaced distance. Tensioning rods are then fitted between the pins in order to apply a constant tensioning force to the bone. These devices are extremely cumbersome and awkward for the patient, often making it difficult or impossible for the patient to walk. In

addition, there is a considerable risk of infection caused by the use of transcutaneous pins. With the device of the present invention, it can be surgically introduced and the components anchored to the bone and tensioning can be applied either continuously or at intervals by placing the means for generating the rotating electro-magnetic field around the implanted device.

The device also has application for curing curvature of the spine (scoliosis). In this procedure, the components of the device will be anchored to appropriate vertebrae or to a rib and selected vertebrae. The distraction force is applied using an externally generated electro-magnetic field to overcome the resistance of the tissue of the limb, together with the friction of the linkage. Use of the device is not limited to applying distraction forces to bone but could, in a modified form, be used for attachment to tendons and thereby used, e.g. for treating cerebral palsy.

According to a further aspect, therefore, there is provided a surgical distraction device for correcting curvature of the spine which comprises first and second extension rods, each having attachment means at one end for fixing to a respective vertebra, a linkage for linking the rods so that the ends carrying attachment means are relatively moveable, said linkage being connected to a drive mechanism comprising a rotatable magnet and a reduction gearbox and actuating means for location externally of the patient for generating a moving or varying electric/magnetic field, thereby causing the magnet to rotate and the attachment means on one rod to move relatively to the attachment means on the other rod.

One embodiment of the present invention will now be described with reference to the accompanying drawings, in which:-

Figure 1 is a perspective view of the distraction device fitted to a femur and forming part of a femoral prosthesis;

Figure 2 is an enlarged, diagrammatic view showing the linkage between the two relatively extendible parts of the prosthesis;

Figure 2a is a view similar to Figure 2 but showing the linkage in more detail;

Figure 3 is an exploded view of the gearbox;

Figure 4 illustrates an alternative method of fixing the prosthesis to a bone; and

Figure 5 is a perspective view of another embodiment of a distraction device for correcting curvature of the spine.

Referring to the drawings, the distraction device comprises two components, a first component (1) forming part of a femoral hip prosthesis including a ball joint (3), forming an artificial hip joint and a relatively moveable component (2) carrying a spigot (4) for anchorage to the re-sected femur (5). The components (1) and (2) are linked together by a linkage comprising an elongated screw (6) and a nut (7). Relative movement of the linkage axially in the direction of the arrow X is achieved by a rotatable magnet (8) connected to a gearbox (9). Magnet (8) is connected through the gearbox (9) to the spindle screw (6) and rotation of the magnet (8) causes rotation of the screw and hence elongation of the device.

Rotation of the magnet is achieved by placing an electro-magnetic coil (10) around the limb and generating a moving electrical field. The magnet is designed so

that the moving electric field causes the magnet to rotate in the desired direction, to cause extension of the device, the magnet and the electro-magnetic coil together constituting an electrical motor. In general the electromagnetic coil takes the form of a ring of coils which may be incorporated or embedded in a magnetically susceptible material, e.g. soft iron, the ring being dimensioned to fit around the limb which contains the extendable distraction device.

The gearbox is a high reduction ratio gearbox which may have a reduction ratio in the order of several hundred, e.g. about 500. This ratio, the speed of rotation of the magnet and the pitch of the screw is selected so that, preferably, one turn of the rotor will advance the screw by a fraction of a millimetre, e.g. 0.001 to 0.01 mm, typically about 0.005 mm. The patient can remain within the electro-magnetic coil for an extended period, e.g. one to two hours, depending upon the amount of extension or length of distracting force required.

In a procedure for lengthening a prosthesis, one would typically wish to extend the prosthesis by 5 to 6 mm at each stage. The distraction force should be such that it is capable of overcoming friction within the linkage and the resistance provided by soft tissue around the prosthesis.

Figure 3 shows details of the gearbox. The gearbox incorporates two planetary gears and is sealed within a casing to exclude body fluids. The parts shown are all manufactured from stainless steel with the exception of the part (12) which is manufactured from titanium alloy and the part (26) which is manufactured from a cobalt chrome alloy.

The gearbox and magnet assembly are designed to be housed within a chamber (201) at the lower end of the linkage assembly (see Figure 2a).

Reference numeral (11) shows an axial spindle on which the driving magnet rotates. The lower end of the spindle (11) is formed with a pointed portion so that the spindle is guided in a recess in the end cap (202 - see Figure 2a). Part (12) is a housing for containing the permanent magnet (8). The permanent magnet is preferably manufactured by a sintering process, has an annular shape and is fitted into an interlocking annular capsule (12) to ensure that the magnet will not rotate within the capsule.

After welding into the capsule (12) the magnet is magnetized with a field direction parallel to the diameter of the capsule (12). Part (13) is the input pinion to the first stage of the gearbox. The pinion is bored to be a free running fit on the spindle (1) and has a plane boss that is interlocked into the capsule (12). The components (14) and (17) are fitted together as a spigot and socket to form a planetary carrier assembly and planet gears (16) are fitted into the carrier assembly. Each gear stage has three identical planet gears which are fitted into the carrier assembly and each planetary gear is machined with a small integral stub shaft at each end. These stub shafts locate in holes in each half of the carrier assembly.

Components (15) and (22) are fixed internal gears for the first and second reduction stages respectively. These gears each mesh with approximately half of the face width of a set of planetary gears and are fixed into the body of a gearbox. The remaining half of the planetary gears is meshed with rotating internal gear (18). A

reduction of the number of teeth in the rotating internal gear (18) compared with the number of teeth in the fixed internal gear (15) provides speed reduction for the stage. Higher number of reductions in the number of teeth provide a greater reduction using the minimum number of gears and space. Components (18), (19) and (20) comprise an assembly which transfers the output from the first stage of the gearing to the input of the second. The assembly comprises a gear web part (19) which is welded to both the rotating internal gear of the first stage of gearing and the input pinion of the second stage.

Parts (21), (22), (23), (24) and (25) comprise the fixed internal gear and planetary gear and carrier assembly for the second stage. These parts are the same as parts (14), (15), (16), (17) and (18) of the first stage of gearing.

Part (25) is the rotating internal gear of the second stage of the gearing and this is welded to part (26) which is the output shaft from the gearbox. This passes through O-ring seals (203) to drive the screw (6) shown in Figures 1, 2 and 2a.

Part (26) is a spigot fit into a socket machined into part (25) so that this assembly is self-jigging when set up for welding. This is the same arrangement as in the case of parts (18) and (19).

Part (26) is shown with a short spigot that locates in a bore in one side of the carrier (24). The purpose of this spigot is to ensure that the planet carrier is accurately centred in the mechanism and a similar spigot is provided on part (19) to centre the first stage planet carrier.

The upper face of part (26) carries an output shaft (27) which terminates in a suitably shaped drive projection engaging in a lead screw (6). Screw (6) is threadably received within an inner shaft (7). Outer shaft (2) surrounds inner shaft (7) so that inner shaft is extended out of shaft (2) when lead screw (6) is rotated by the magnet (8) driving through the reduction gearbox. Inner shaft (7) is prevented from rotating by means of a key (204) which is fixed into a corresponding aperture in the outer shaft (7). Key (204) has a projecting portion which extends into a longitudinal slot (205) formed in the inner shaft (7).

The device may include an integral or separate sensor which counts the number of revolutions of the magnet (8) so that the degree of extension of the device can be monitored and/or recorded.

Figure 4 illustrates an alternative method of anchoring one component of the device to a long bone, which has been found to result in improved stability and bone integration. As can be seen, the tubular body (2) of the prosthesis is fixed to a resected bone (5) by means of two or more plate-like strips (401) extending from the tubular body. The plate-like strips are apertured to receive unicortical screws (402) for fixing the plate-like strips to the outer surface of the bone. The strips were formed with elongated holes (403) to encourage bone ingrowth and coated with a highly crystalline hydroxyapatite coating to stimulate integration with the bone.

Figure 5 shows a distraction device suitable for treatment of curvature of the spine. In this embodiment, threaded rods (501) and (502) are provided with attachment means (503), such as plates or blocks carrying screws or hooks.

Depending on the curvature of the spine, the threaded rods (501) may be straight or curved. The attachment means may include a universal bearing or joint connection to the threaded rods so that the attachment means can be rotated or swivelled to take account of the curvature or deformation of the spine, and to enable firm attachments to be made to appropriate vertebrae.

The two threaded rods are joined by a central spacer or block (504). The block (504) may be stabilised by attachment to the spine, e.g. using screws or hooks. Block (504) includes a drive mechanism comprising a rotatable magnet and reduction gearbox as described in Figures 1 to 3. However, the final drive component comprises a cog carried by the output shaft. The cog is linked to two worm drives which are turned by the cog. In one arrangement, the worms are arranged in series such that they are driven in opposite directions. In another arrangement, the worms may be arranged to be driven in the same direction. Single worm or multiple worm drives of two or more worms may also be used to link the cog to the threaded rods. Each worm is cylindrical and has a central tapping. A corresponding threaded rod (502) is received within the worm and the threads on the rod match the tapping on the inside of the worm. As the worm is turned by the cog, the threaded rod is also turned and is moved in a direction lengthwise of the rod. The direction of movement of the rod depends on the sense of the cooperating threads and on the arrangement of the worms and gears which transmit the rotation of the magnet. In a similar way to the arrangement described in connection with Figures 1 to 4, a rotating electrical field is generated by means of an external annular coil around the patients' spinal region.

CLAIMS:-

1. A surgical distraction device for applying an extending or tensioning force non-invasively to a patient's skeleton or to an implant which comprises anchoring means for attaching first and second components of the device to a bone or to adjoining bones, said components being connected by a linkage of an extendible length, a magnet connected to the linkage via a reduction gearbox and actuating means located externally of the patient for generating a moving or varying electro-magnetic field, thereby causing the magnet to rotate and the linkage to be extended.

2. A device according to claim 1 wherein the actuating means is adapted to generate a rotating magnetic field which rotates in the desired direction of rotation of the magnet.

3. A device according to claim 1 or 2 wherein the linkage comprises a screw and nut and the magnet is connected to the screw or nut via the gearbox to cause relative rotation, thereby extending the device.

4. A device according to any one of the preceding claims wherein there is a difference in the number of teeth between gears, thereby providing a reduction in the number of gears and space.

5. A device according to any one of the preceding claims wherein the gearbox has a gear ratio of 100:1 to 1000:1.

6. A device according to any one of the preceding claims wherein the two components are components of a prosthesis for fixing to a long bone.

7. A device according to claim 6 wherein the long bone is a femur.

8. An extendable endoprosthetic replacement device which comprises first and second components which are connected by a linkage of extendable length, the first component comprising a component of a limb joint and the second component having means for attachment to a resected long bone, a magnet connected to the linkage and means adapted for location externally of the patient for generating a moving electromagnetic field, thereby causing the magnet to rotate and the linkage to be extended.

9. A device according to claim 8 wherein the magnet is connected to the linkage via a reduction gearbox.

10. A device according to claim 8 or 9 wherein the first component comprises a component of a hip or knee joint.

11. A device according to any one of claims 8 to 10 wherein the second component comprises a spigot adapted for fixing into the intermediary canal of a long bone.

12. A surgical distraction device for correcting curvature of the spine which comprises first and second extension rods, each having attachment means at one end for fixing to a respective vertebra, a linkage for linking the rods so that the ends carrying attachment means are relatively moveable, said linkage being connected to a drive mechanism comprising a rotatable magnet and a reduction gearbox and actuating means for location externally of the patient for generating a moving or varying electric/magnetic field, thereby causing the magnet to rotate and the attachment means on one rod to move relatively to the attachment means on the other rod.

13. A device according to any one of the preceding claims wherein the actuating means is arranged to generate a rotating field which rotates in the desired direction of rotation of the magnet.

14. A device according to claim 12 or 13 wherein the linkage comprises a drive cog which engages with a worm encircling a rod so that movement of the worm causes the rod to extend lengthwise thereof.

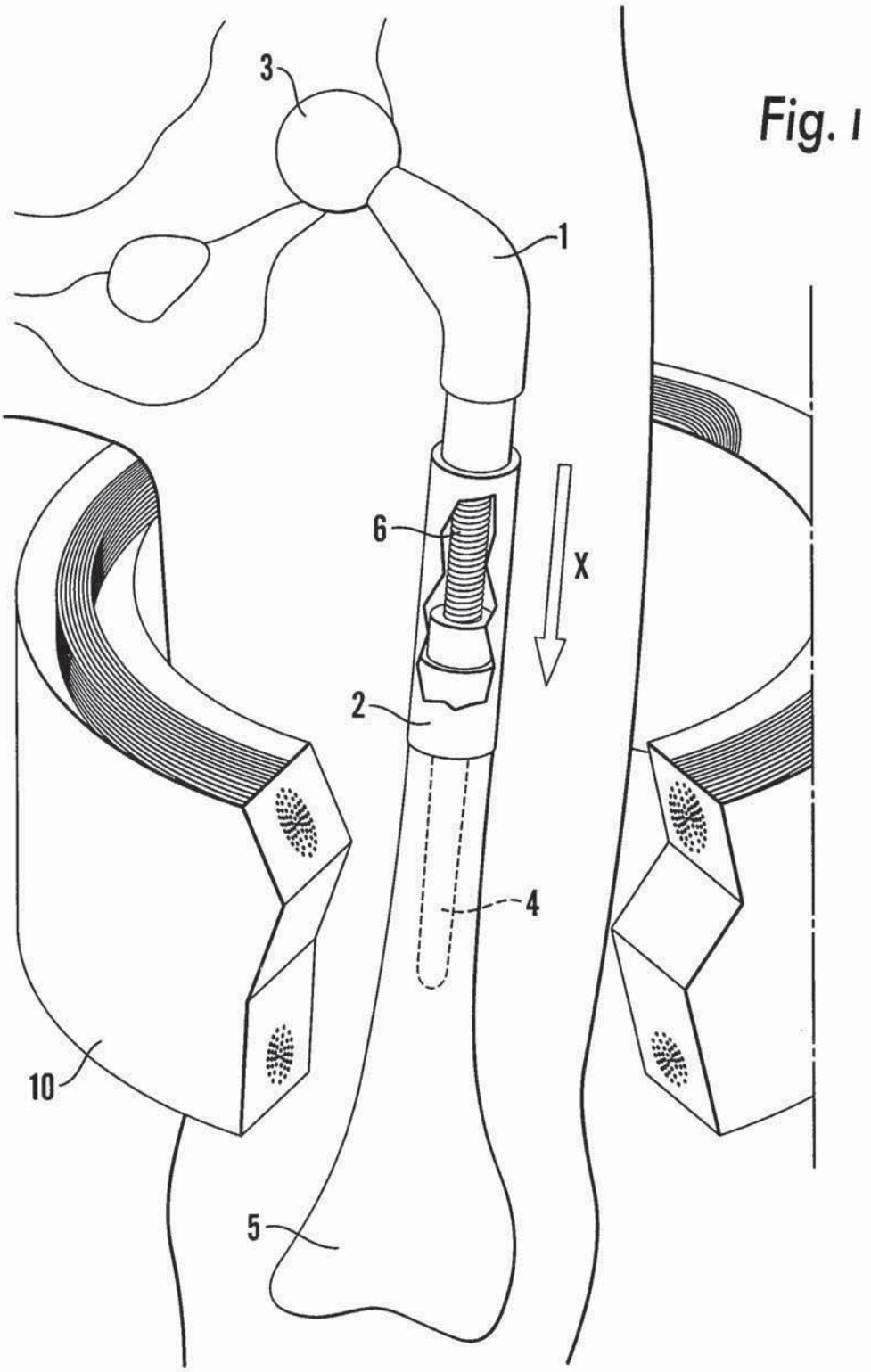
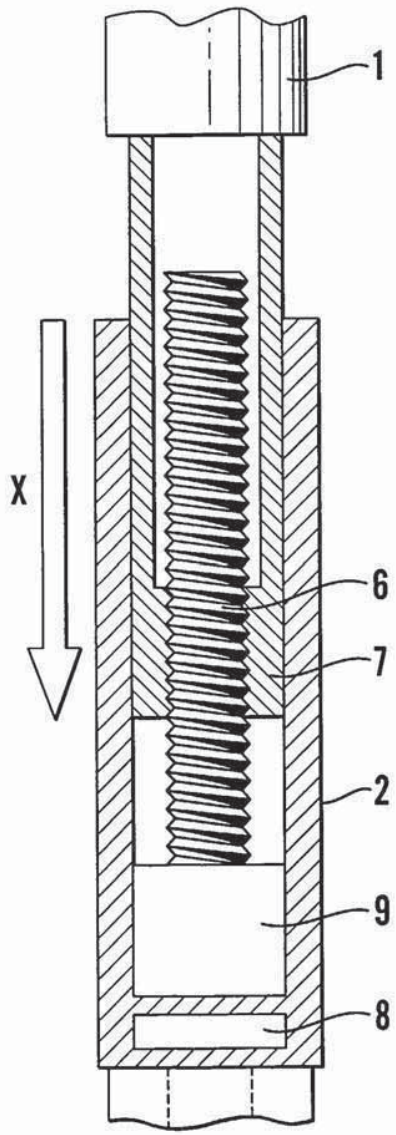


Fig. 1

Fig.2



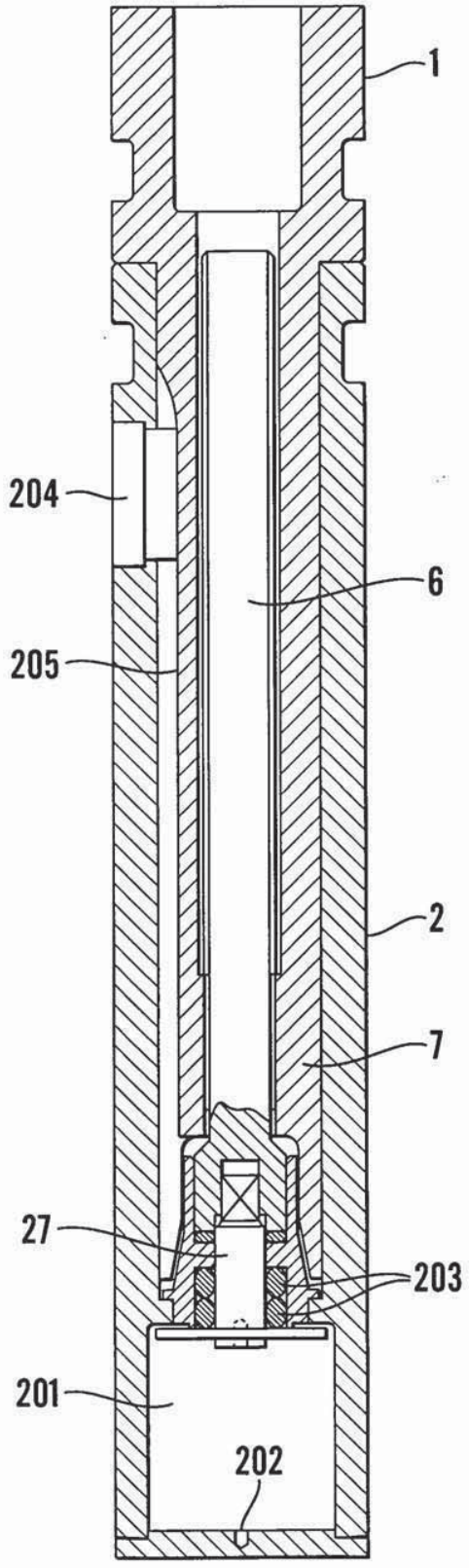


Fig.2a

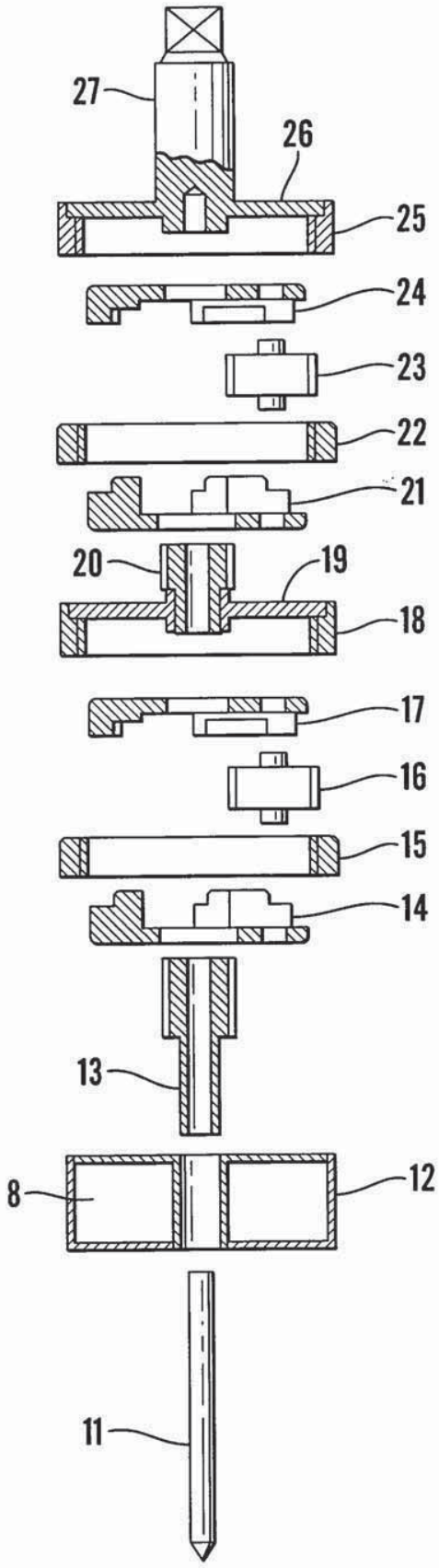


Fig.3

Fig.4

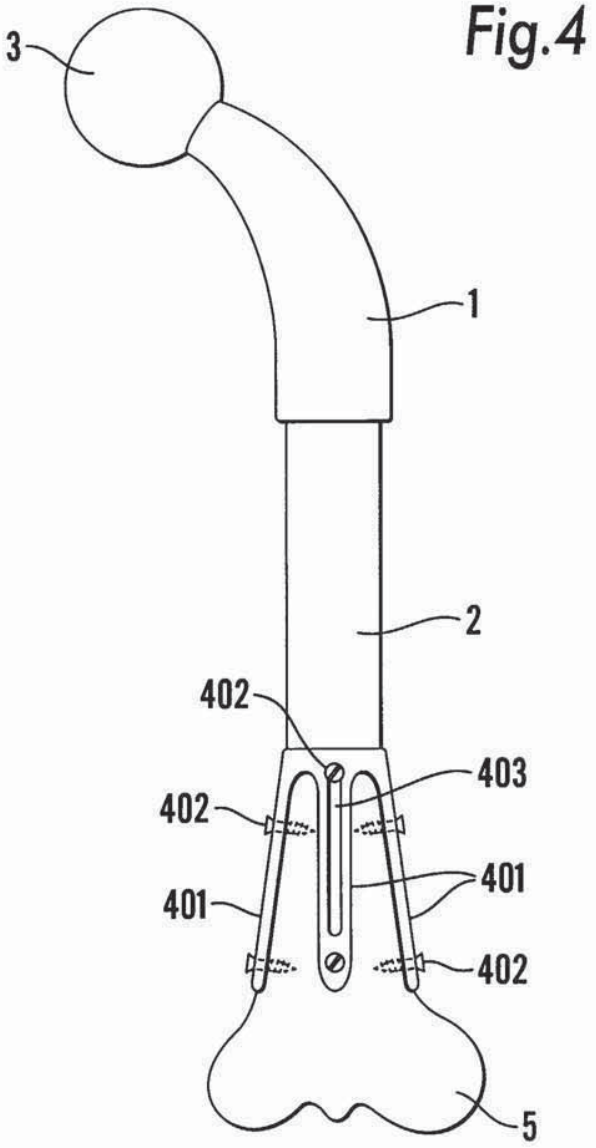
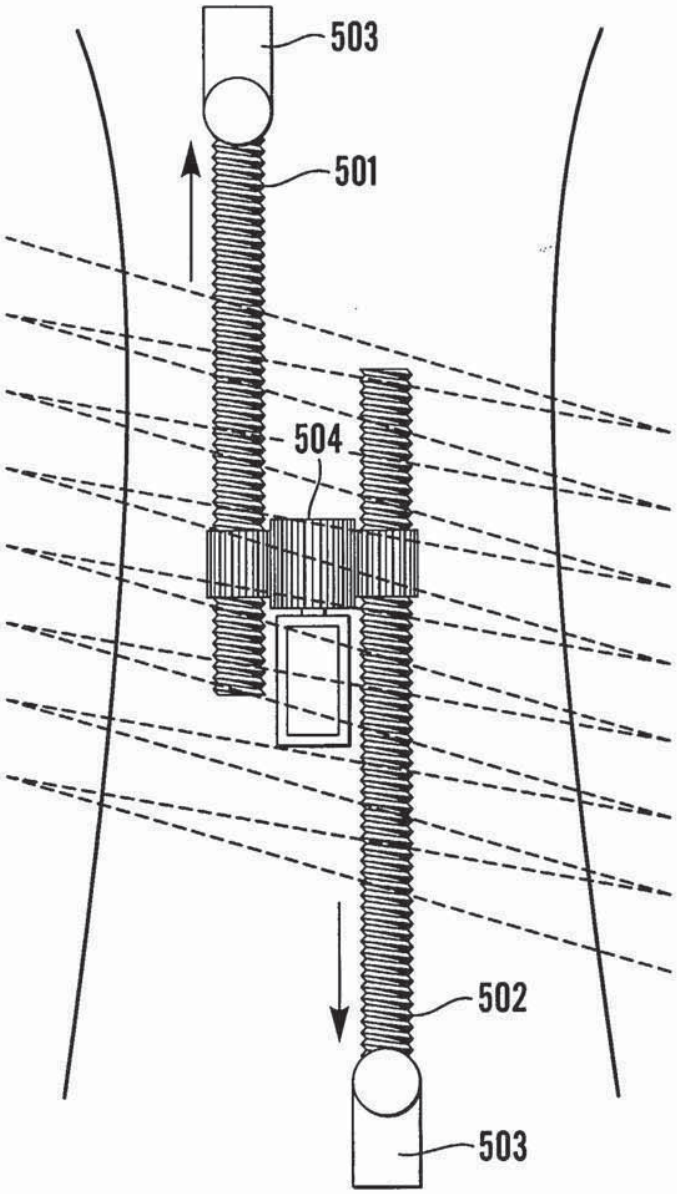


Fig.5



INTERNATIONAL SEARCH REPORT

Inter ıal Application No
PCT/GB 01/01668

A. CLASSIFICATION OF SUBJECT MATTER
IPC 7 A61B17/72 A61F2/36

According to International Patent Classification (IPC) or to both national classification and IPC

B. FIELDS SEARCHED

Minimum documentation searched (classification system followed by classification symbols)
IPC 7 A61B A61F

Documentation searched other than minimum documentation to the extent that such documents are included in the fields searched

Electronic data base consulted during the international search (name of data base and, where practical, search terms used)
EPO-Internal

C. DOCUMENTS CONSIDERED TO BE RELEVANT

Category °	Citation of document, with indication, where appropriate, of the relevant passages	Relevant to claim No.
X	US 5 704 939 A (JUSTIN DANIEL F) 6 January 1998 (1998-01-06)	8
Y	the whole document	1-7,9-14
X	WO 99 51160 A (NOAR JOSEPH ;YOUNG DAVID (GB); EVANS ROBERT (GB); HARRIS IVOR REX) 14 October 1999 (1999-10-14)	8
A	abstract; figure 1	1,12
Y	DE 197 00 225 A (BETZ AUGUSTIN PROF DR ;BUTSCH MICHAEL DR (DE)) 9 July 1998 (1998-07-09)	1-7,9-11
A	abstract; figure 1	8
Y	DE 24 37 752 A (SCHULZE WILHELM) 26 February 1976 (1976-02-26)	12,13
A	the whole document	1,8

-/--

[X] Further documents are listed in the continuation of box C.

[X] Patent family members are listed in annex.

° Special categories of cited documents :

"A" document defining the general state of the art which is not considered to be of particular relevance

"E" earlier document but published on or after the international filing date

"L" document which may throw doubts on priority claim(s) or which is cited to establish the publication date of another citation or other special reason (as specified)

"O" document referring to an oral disclosure, use, exhibition or other means

"P" document published prior to the international filing date but later than the priority date claimed

"T" later document published after the international filing date or priority date and not in conflict with the application but cited to understand the principle or theory underlying the invention

"X" document of particular relevance; the claimed invention cannot be considered novel or cannot be considered to involve an inventive step when the document is taken alone

"Y" document of particular relevance; the claimed invention cannot be considered to involve an inventive step when the document is combined with one or more other such documents, such combination being obvious to a person skilled in the art.

"&" document member of the same patent family

Date of the actual completion of the international search	Date of mailing of the international search report
29 June 2001	05/07/2001

Name and mailing address of the ISA	Authorized officer
European Patent Office, P.B. 5818 Patentlaan 2 NL – 2280 HV Rijswijk Tel. (+31–70) 340–2040, Tx. 31 651 epo nl, Fax: (+31–70) 340–3016	Hansen, S

Form PCT/ISA/210 (second sheet) (July 1992)

INTERNATIONAL SEARCH REPORT

Inter nal Application No
PCT/GB 01/01668

C.(Continuation) DOCUMENTS CONSIDERED TO BE RELEVANT

Category °	Citation of document, with indication,where appropriate, of the relevant passages	Relevant to claim No.
Y	US 5 364 396 A (ROBINSON RANDOLPH C ET AL) 15 November 1994 (1994-11-15)	14
A	abstract; figures 4,6	1,8,12
A	EP 0 548 535 A (HOWMEDICA GMBH) 30 June 1993 (1993-06-30) abstract; figure 1	1,8,12
A	US 4 931 055 A (BUMPUS JOHN ET AL) 5 June 1990 (1990-06-05) abstract; figures 1,3	1,8,12

Appendix - III

National Institute of Clinical Excellence (NICE) position statement on Magnet driven Growing Rods (MdGRs) for Early-Onset Scoliosis

NATIONAL INSTITUTE FOR HEALTH AND CARE EXCELLENCE

Medical technology guidance

SCOPE

MAGEC system for spinal lengthening in children with early onset scoliosis

1 Technology

1.1 Description of the technology

MAGEC system (Ellipse Technologies, Inc.) is used to treat early onset scoliosis (a side-to-side curve diagnosed before the age of 10 years) in children aged between 2 and 11 years. The technology is intended to be used in place of current growth rod systems that require repeated invasive surgical procedures. MAGEC system comprises a sterile implant (this can be one or two rods, the size of which depends on body weight - 4.5mm for children weighing ≤27kg, 5.5mm for children weighting ≤36kg and 6.35mm for larger children) and an external remote controller together with a manual distractor (to check the implant is functional prior to implantation) and wand locator (to locate the internal magnet). MAGEC system is inserted surgically in the same way as conventional growth rod systems. A portion of the MAGEC implant contains a distraction element – the 'actuator', which includes an internal cylindrical rare earth magnet, 9–10.5 mm in diameter and 90mm long. The magnet is rotated non-invasively by an external remote controller which, in turn, causes the rod to lengthen and increase distraction of the spine (on average every 3 months, depending on surgeon preference). This external remote control is portable and uses permanent magnets to modify the length of the implant. An indication of the amount of distraction/retraction is immediately visible on the controller's display module. Repeated lengthening of the rod is intended to be carried out in an outpatient setting without the use of sedation or pain relief

1.2 Regulatory status

MAGEC system received a CE mark in October 2009 to be used as a Spinal Bracing and Distraction System Implant. The CE mark also includes the External Adjustment Device (remote control).

1.3 Claimed benefits

The benefits to patients claimed by the sponsor relate to the avoidance of repeated surgical procedures which lead to:

- Reduced incidence of surgical complications including anaesthetic risk, infections and delayed recovery
- Reduced psychological trauma to the child and family
- Improved quality of life due to reduced time away from school (patient) and work (parent)

The benefits to the healthcare system claimed by the sponsor are:

- The avoidance of costs associated with repeated surgical interventions including theatre time, consumables, in-hospital stay and treatment of complications
- Reduced costs to society from parents taking time off work and the child taking time away from school

1.4 Relevant diseases and conditions

Scoliosis is a lateral, side-to-side deformity of the spine. MAGEC system is intended for use in early onset scoliosis specifically in patients in whom a distraction procedure is indicated.

In the UK, scoliosis affects 3 to 4 children out of every 1,000 and can develop at any time during childhood and adolescence (NHS Choices 2011). At the younger end of the spectrum, boys are affected slightly more than girls (Scoliosis Association, UK). Early onset scoliosis may be associated with congenital vertebral anomalies, bone dysplasia, connective tissue disorders, neuromuscular disorders or idiopathic spinal deformities. Lung development, pulmonary function, spine length and spinal mobility are at risk. In young

children, growth and lung function are affected. Scoliosis also impacts on the appearance of a patient's back. Treatment of early onset scoliosis must focus on both the spine and the development of the chest to improve the child's long-term quality of life.

1.5 Current management

In 90 percent of cases of childhood scoliosis, treatment is not required since the condition corrects itself with age (NHS Choices, 2011). Most of the remaining 10 percent of children with scoliosis can be successfully treated using a back brace to prevent the spinal deformity progressing. Approximately 1 in 350 children with scoliosis require surgery to correct the curvature of their spine.

There are four main types of treatment for early onset scoliosis, namely casting, bracing, insertion of growth rods and spinal fusion. Treatment selection depends on the age and development of the child as well as the type of curvature of the spine. Casting can be used to guide a child's spine into a normal position during growth. This can be done by applying an external brace, made from a combination of plaster-of-Paris and modern casting material, which is worn permanently by the child. Alternatively the patient may be put in a brace. This system rarely corrects scoliosis but allows growth before a more permanent treatment is offered.

Surgery, using distraction-based procedures, can be performed if the curvature continues despite bracing and casting. This involves surgically inserting growth rods (single or dual), which are attached to the spine or ribs above and below the curve. The rods can be placed from the level of the shoulder blade on the spine and extend down to the ribs, spine or pelvis. If a conventional growth rod is used the child returns about every six months to undergo a surgical procedure for rod lengthening. Once the spine has finishing growing a final spinal fusion operation is undertaken and the rods are removed.

2 Reasons for developing guidance on MAGEC system for Early-onset Scoliosis

The Committee considered that MAGEC system may provide significant benefit for children with early onset scoliosis in whom surgical intervention is indicated for spinal distraction.

The Committee considered that the main potential benefit of MAGEC system was the avoidance of repeat surgical procedures to lengthen the rods, thereby reducing anaesthetic, operative and post-operative risk (including infection).

The Committee was advised that lengthening of MAGEC system could take place in an outpatient setting with no sedation or pain relief. They considered that in conjunction with the avoidance of surgical procedures, use of MAGEC system could have a positive psychosocial impact on both the young children and their families.

The Committee was advised that MAGEC system has the potential to result in comparable lengthening to that achieved with conventional spinal rods and that more frequent lengthenings could be undertaken approximately every 3 months, as opposed to every 6 months with conventional rods. The potential impact of increasing the regularity of lengthening on the outcome of the distraction procedure should be evaluated and the number of imaging procedures associated with the increased number of distraction procedures should also be considered.

3 Statement of the decision problem

	Scope issued by NICE
Population	Children with early onset scoliosis in whom a surgical distraction procedure is indicated.
Intervention	MAGEC system
Comparator(s)	• Spine-based surgical distraction procedure with conventional growth rods
Outcomes	The outcome measures to consider include: • Total number of surgical procedures and anaesthetics • Total number of outpatient attendances and procedures • Recovery time • Total length of stay • Rate of distraction procedure success • Infection rates and other surgical complication rates • Total number of imaging procedures • Quality of life • Device failure • Device and radiation exposure-related adverse events
Cost analysis	Comparator(s): • Spine-based surgical distraction procedure with conventional growth rods Costs will be considered from an NHS and personal social services perspective. The time horizon for the cost analysis will be sufficiently long to reflect any differences in costs and consequences between the technologies being compared. The costs associated with failure of MAGEC system leading to replacement with conventional growth rods should be included. Sensitivity analysis will be undertaken to address uncertainties in the model parameters, which will include the scenario in which single and dual growth rods are needed. This analysis should include analysis of differing regularity of MAGEC distraction procedures, every 6 weeks and every 3 months.
Subgroups to be considered	None identified
Special considerations, including issues related to equality	Although MAGEC system is described for treatment of children with early onset scoliosis, who are between the ages of 2 and 11, the system could be used in children outside this age range providing they weigh more than 11.4kg and have a BMI <25. Patients with contraindications could include patients with pacemakers or who require an MRI scan during the period of implantation.

4 Related NICE guidance

4.1 Published

The EOS 2D/3D imaging system. NICE Diagnostics Guidance DG1 (2011). Available from http://guidance.nice.org.uk/DG1.

4.2 Under development

None identified.

4.3 Suspended or terminated

None identified.

5 External organisations

5.1 Professional organisations

5.1.1 Professional organisations contacted for expert advice

At the selection stage, the following societies were contacted for expert clinical and technical advice:

- British Association of Spine Surgeons
- British Orthopaedic Association
- British Scoliosis Society
- British Society for Children's Orthopaedic Surgery

5.1.2 Professional organisations invited to comment on the scope

The following societies have been alerted to the availability of the scope for comment:

- British Association of Spine Surgeons
- British Orthopaedic Association
- British Scoliosis Society
- British Society for Children's Orthopaedic Surgery

- Royal College of Paediatrics and Child Health

5.2 Patient organisations

At the selection stage, NICE's Patient and Public Involvement Programme contacted the following organisations for patient commentary and alerted them to the availability of the scope for comment:

- Action for Kids
- Action for Sick Children
- Action Medical Research
- Arthritis and Musculoskeletal Alliance (ARMA)
- Back Care
- Brainwave
- Breakthrough UK
- British Syringomyelia Chiari Society
- Child Growth Foundation (CGF)
- Disability Rights UK
- Fighting Back UK
- Muscular Dystrophy Campaign
- National Childbirth Trust (NCT)
- National Children's Bureau (NCB)
- Neurofibromatosis Association
- Newlife Foundation for Disabled Children
- Rare Disease UK
- Restricted Growth Association (RGA)
- Royal College of Paediatrics and Child Health (RCPCH)
- Scoliosis Association (UK)
- Scope
- Specialised Healthcare Alliance
- Spinal Injuries Association
- WellChild

NICE National Institute for Health and Care Excellence

Putting NICE guidance into practice

Costing statement - The MAGEC system for spinal lengthening in children with scoliosis Implementing the NICE guidance (MTG18)

Published: June 2014

NHS England commissions complex spinal surgery services for children from specialist paediatric spinal surgery centres.

The Committee for the topic was advised that the population of children for whom the MAGEC system would be considered is small, with an estimated 120 children per year in England who may be treated using growth rods. Because of this, it is unlikely that the guidance will result in a significant change in resource use in the NHS.

The External Assessment Centre estimated the insertion costs of MAGEC rods to be £27,400, with an annual lengthening cost of £900. In contrast, conventional growth rods are estimated to cost £15,300 for insertion and £5400 for annual lengthening.

The additional insertion cost of £12,100 for the MAGEC system has a payback period of less than 3 years. Anticipated savings per child after 6 years are estimated to be around £12,000.

NICE National Institute for Health and Care Excellence

The MAGEC system for spinal lengthening in children with scoliosis

Issued: June 2014

NICE medical technology guidance 18

guidance.nice.org.uk/mtg18

Contents

1 Recommendations

NICE medical technologies guidance addresses specific technologies notified to NICE by sponsors. The 'case for adoption' is based on the claimed advantages of introducing the specific technology compared with current management of the condition. This case is reviewed against the evidence submitted and expert advice. If the case for adopting the technology is supported, then the technology has been found to offer advantages to patients and the NHS. The specific recommendations on individual technologies are not intended to limit use of other relevant technologies which may offer similar advantages.

1.1 The case for adopting the MAGEC system for spinal lengthening in children with scoliosis is supported by the evidence. Using the MAGEC system would avoid repeated surgical procedures for growth rod lengthening. This could reduce complications and have other physical and psychological benefits for affected children and their families.

1.2 The MAGEC system should be considered for use in children with scoliosis aged 2 years and over who need surgery to correct their spinal curvature, for example when conservative methods such as bracing or casting have failed.

1.3 Findings from cost modelling estimate that using the MAGEC system is cost saving compared with conventional growth rods from about 3 years after first insertion. The estimated cost saving per child after 6 years is around £12,077. The cost savings remained robust in sensitivity analyses. Further savings could be made by avoiding the need for spinal cord monitoring, which is sometimes used during conventional growth rod lengthening but is not needed when lengthening the MAGEC growth rods.

2 The technology

Description of the technology

2.1 The MAGEC spinal bracing and distraction system (Ellipse Technologies Inc.) is used in children aged 2 years or over to brace the spine during growth and minimise the progression of scoliosis. The technology is intended to be used in place of current growth rod systems that need repeated invasive surgical procedures. The MAGEC growth rods are usually removed and replaced by a spinal fixation system to fuse the spine when skeletal maturity is reached.

2.2 The MAGEC system comprises 1 or 2 sterile titanium implantable growth rods and an external remote control for non-invasive lengthening. The diameter of the rods used depends on the child's body weight (4.5 mm for children weighing up to 27 kg, 5.5 mm for children weighing up to 36 kg). The choice of whether to insert 1 or 2 rods is made by the surgeon and depends mainly on the child's size. A portion of each MAGEC rod contains a proprietary distraction element, the 'actuator', which includes an internal cylindrical rare earth magnet. The system also includes a manual distractor (to check the implant is functional before implantation) and a wand locator (to locate the internal magnet).

2.3 The MAGEC system received a CE mark in 2009 for the growth rods and external remote control. A keeper plate was added to the actuator in 2010 to prevent the internal magnet rotating. If this happened when the rod was placed under large amounts of stress, it could cause the rod to slip with some loss of distraction. Following the examination of rod breakages, the design was further revised in 2012 to strengthen the titanium rods by using continuous wave, rather than pulse, laser welding.

2.4 The MAGEC rods are inserted surgically in the same way as conventional growth rods. Distractions (lengthenings) are carried out typically every 3 months, depending on clinical assessment and the surgeon's judgement as to the optimal length of time between distractions for the individual child. The distractions are performed in an outpatient setting without the need for anaesthesia, sedation or pain relief. The magnet within the actuator is

connected to a lead screw and is rotated non-invasively by the external remote control, causing the rod to lengthen and increase the distraction of the spine. The external remote control is portable and uses permanent magnets to rotate the magnet within the rod. The control's display module gives a real-time indication of the amount of distraction or retraction gained. Verification of the new rod length can be done using X-ray or ultrasound if needed.

2.5 The cost of the MAGEC system stated in the sponsor's submission is £13,500 to £14,000 for a set of dual rods, with an additional £450 to £500 for the hooks and screws (supplied by another company) to attach the rods to the spine. These costs are exclusive of VAT. The external remote control is loaned by the sponsor at no cost, other than that of sending it to the treatment centre.

2.6 The claimed benefits of the MAGEC system in the case for adoption presented by the sponsor are:

- The avoidance of repeated surgical procedures, leading to:
  - A reduction in the incidence of surgical complications, including anaesthetic risk, infections and delayed recovery.
  - A reduction in psychological trauma to the child and family.
  - Improved quality of life because of reduced time away from school (child) and work (parent).
- The avoidance of costs associated with repeated surgical interventions, including theatre time, consumables, in-hospital stay and treatment of complications.
- A reduction in costs to society associated with the parent or carer taking time off work and the child being away from school.

Current management

2.7 In many cases of childhood scoliosis, interventional treatment is not needed because the condition corrects itself as the child grows. For those children needing treatment, there are 4 main types, namely casting, bracing, insertion of growth rods and spinal fusion. The type of treatment chosen depends on

the age and development of the child as well as the type of curvature of the spine. Casting involves use of an external cast, made from a combination of plaster-of-Paris and modern casting material, which is applied to help guide a child's spine into a normal position during growth. The cast is worn permanently by the child and is replaced frequently as the child grows. Bracing involves use of a rigid or flexible brace. Bracing rarely corrects scoliosis, but can prevent curve progression and allow the spine to grow before a more permanent treatment is offered.

2.8 Around 120 children with scoliosis per year in England need surgical intervention to implant growth rods to correct the curvature of their spine. This type of surgery is performed if the spinal curvature progresses despite bracing and casting. It involves inserting growth rods (single or dual), which are attached to the spine or ribs above and below the curve using hooks and pedicle screws. The rods can be placed around the cervical or thoracic part of the spine, extending down to the ribs, lumbar spine or pelvis. The initial insertion procedure for the MAGEC system is similar to that for conventional growth rods and usually involves staying in hospital for 5–14 days and several weeks away from school and usual daily activities. If a conventional growth rod is used, the child returns about every 6 months to have a surgical procedure for rod lengthening. The procedure involves manually lengthening the growth rods via small incisions in the back under general anaesthesia. This can be done as a day case procedure but may often involve an overnight stay in hospital.

3 Clinical evidence

Summary of clinical evidence

3.1 Full details of all clinical outcomes considered by the Committee are available in the assessment report overview.

3.2 The key clinical outcomes for the MAGEC system presented in the decision problem were:

- the total number of surgical procedures and anaesthetics
- the total number of outpatient attendances and procedures
- recovery time
- total length of stay
- rate of distraction procedure success
- infection rates and other surgical complication rates
- total number of imaging procedures
- quality of life
- device failure
- device and radiation exposure-related adverse events.

3.3 The sponsor addressed 5 of the outcome measures defined in the decision problem, and added additional outcomes including Cobb angle (a measure of spinal curvature based on a spinal radiograph), thoracic and total spine height. Outcomes relating to pulmonary function were also reported. The sponsor stated that these outcomes were reported in clinical studies, whereas several outcomes in the scope, such as quality of life, recovery time and total length of stay were not and could, therefore, not be addressed.

3.4 The sponsor identified 4 published and 6 unpublished studies relevant to the MAGEC system. The sponsor excluded 2 of the unpublished studies because they were records of ongoing clinical trials and had not reported any findings. The remaining 8 papers were included in the sponsor's summary of clinical evidence (Akbarnia et al. 2012, 2013a, 2013b; Cheung et al. 2012; Dannawi et al. 2013; Ellipse Technologies Inc. 2013; Richards and Nnadi 2013; Yoon et al. 2013). The study by Akbarnia et al. (2012) was excluded from further consideration because it was an animal study. The unpublished study by Richards and Nnadi (2013) was excluded because it was a cost analysis, but this report was included as economic evidence elsewhere in the submission. The studies by Akbarnia et al. (2013b) and Yoon et al. (2013) were excluded from the sponsor's evidence synthesis because they described patients already included in the studies by Ellipse (2013) and Dannawi et al. (2013) respectively. In summary, 6 studies formed the basis of the sponsor's clinical evidence presentation, of which 4 were included in the evidence synthesis. The External Assessment Centre considered that the studies presented by the sponsor were in keeping with the scope and were appropriate for inclusion.

3.5 Akbarnia et al. (2013a) reported preliminary findings from a prospective, observational multicentre study involving 14 children with early-onset scoliosis who received the MAGEC system rods. The mean age of the participating children was 8 years 10 months, with a range of 3–12 years. Single-rod surgery was carried out in 5 children and dual-rod surgery in 9 children. All children were given a brace for 3–6 months after surgery. The mean follow-up time was 10 months and the mean number of distractions per child was 4.9. The mean pre-operative Cobb angle was 60°. Post-operatively, the mean Cobb angle was 34° initially and 31° at the latest follow-up. Mean pre-operative thoracic spine height was 178 mm, increasing to 198 mm after surgery and 208 mm at the latest follow-up. Mean pre-operative total spine height increased from 292 mm pre-operatively to 322 mm post-operatively and 338 mm at final follow-up. The changes in total spine height were all reported to be statistically significant ($p<0.05$). No significant differences in the Cobb angle correction were found between children receiving single or dual rods. Complications included superficial infection in 1 child with a single rod, a prominent implant in 1 child with dual rods, and partial loss of initial spine height in 3 children with single rods immediately after surgery. Partial loss of

distraction (a reduction in rod length) was observed after 11 out of 68 distractions but was subsequently regained.

3.6 Cheung et al. (2012) reported a prospective case series, reviewing outcomes for 2 children with scoliosis receiving MAGEC system rods, followed up for 24 months. A single rod was used for a 5-year-old child and dual rods for a 12-year-old child. Both children wore a brace for 3 months post-operatively. Distractions were carried out monthly in an outpatient setting, with an average lengthening of 1.5–2 mm. X-rays were used to measure the Cobb angle, spine height and amount of distraction. Pain was assessed using a visual analogue scale to produce a pain score and the children also completed a quality-of-life questionnaire for scoliosis (SRS-30). The child with a single rod had a change in Cobb angle from 74° pre-operatively to 19° immediately post-surgery and 26° after 24 months. In the child with dual rods, the Cobb angle changed from 60° pre-operatively to 31° after surgery and remained at 31° after 24 months. The authors reported the mean change in length of the instrumented segment of the spine after each distraction to be 1.9 mm±0.4 mm. Mean thoracic spine height increased from 199 mm at baseline to 203 mm post-operatively and subsequently to 229 mm after 24 months. Mean total spine height increased from 314 mm at baseline to 331 mm after surgery and 360 mm after 24 months. The authors reported that the mean monthly increases in spine height were greater than those predicted by standard growth charts. The pain score was 0 (no pain) pre-operatively and at each stage of follow-up. Superficial infection occurred in 1 child. Loss of distraction occurred after 1 of 43 procedures. This related to an excess bending moment in the rod causing slippage in the magnetic section, which led the sponsor to make an improvement in the rod's design (described in section 2.3).

3.7 Dannawi et al. (2013) reported a prospective case series involving 34 children with early-onset scoliosis receiving the MAGEC system rods in a UK hospital. Children were included if they had progression of curvature greater than 10° over 6 months and a Cobb angle greater than 40°. The mean age of the children was 8 years, ranging from 5 to 12 years. Mean follow-up time was 15 months, with a minimum of 12 months. Single rods were used for 12 children and dual rods for 22 children. Each child received at

least 3 distractions and the mean number of distractions per child was 4.8 (range 3–6). Surgery with conventional growth rods had already been carried out in 2 children before conversion to the MAGEC system rods at their parents' request (to potentially reduce the number of surgical procedures for their children). These children had Cobb angles of 75° and 80° at the time of conversion. Distractions were carried out approximately every 3 months (mean 87 days) in an outpatient setting with the aim of achieving 4.5 mm growth following each distraction. Cobb angle and total spine height were measured pre- and post-operatively. The mean pre-operative Cobb angle was 69°. This decreased to a mean of 47° post-operatively and to 41° at final reported follow-up. The mean pre-operative total spine height was 304 mm: this increased to 335 mm immediately after surgery and to 348 mm at final review. The differences between Cobb angle measurements pre- and post-operatively and after follow-up in the single (n=12) and dual rod (n=22) groups were found to be statistically significant within each group ($p<0.001$). Similarly, the differences in the mean total spine height pre- and post-operatively and after follow-up in each group were statistically significant ($p<0.001$). Superficial infection occurred in 2 children, and there were 2 rod breakages needing revision. Loss of distraction occurred in 2 children, which was later rectified. This contributed to the subsequent revision of the rod design, as did the study by Cheung et al. (2012).

3.8 The unpublished study by Ellipse (2013) was a retrospective review including 54 children with early-onset spinal deformity associated with thoracic insufficiency (inability of the thorax to support normal respiration or lung growth) receiving MAGEC system rods. Children were included from 15 centres in 8 countries. Of these, 30 children had MAGEC rods inserted as their first (de novo) surgery and 24 had had previous spinal surgery to insert conventional growth rods. The sponsor's submission included a study report describing outcomes for 14 of these children. The sponsor supplied further data on all 54 children but the External Assessment Centre only included data on the 30 de novo patients from the original submission because it considered that revision surgery outcomes may not be directly comparable with those for initial surgery. The mean age of the 30 included children was 7 years, ranging from 2 to 10 years. Out of the 30 children having de novo surgery, 9 received a single rod and 21 received dual rods. Mean follow-up time for the children

having de novo surgery was 21 months. At baseline the Cobb angle was measured in 28 children and total spine height was measured in 27 children. Thoracic spine height and space available for lungs (SAL) were also measured and each measurement was repeated post-operatively and at follow-up. The mean Cobb angle changed from 64° at baseline to 35° post-operatively and then to 43° at final follow-up. The mean total spine height was 264 mm pre-operatively, increasing to 308 mm post-operatively and 312 mm at final follow-up. The mean thoracic spine height increased from 164 mm pre-operatively to 192 mm post-operatively and 194 mm at final follow-up. These differences were found to be statistically significant ($p<0.001$). The change in SAL was reported for 5 children who had a baseline SAL of less than or equal to 90%. For these 5 children, mean SAL increased by 27% after 24 months.

3.9 Akbarnia et al. (2013b) reported on a retrospective matched case series involving children with early-onset scoliosis: 12 treated with the MAGEC system rods and 12 treated with conventional growth rods, in 5 centres worldwide. The 12 children in the MAGEC system group were included in the Ellipse (2013) study and were therefore not included in the sponsor's evidence synthesis. This study is summarised here because it is the only study that compared the MAGEC system against conventional growth rods. The children were matched by disease type, sex (when possible), age, degree of curvature and receipt of single or dual rods. Their mean age was 6.8 years in the MAGEC group and 6.6 years in the conventional growth rod group. The mean follow-up time differed between the 2 groups, being 2.5 years in the MAGEC group and 4.1 years in the conventional growth rod group. In the MAGEC group, the Cobb angle changed from a pre-operative (baseline) mean of 59° to 32° immediately post-operatively and 38° at the latest follow-up. In the conventional growth rod group, the mean Cobb angle changed from 60° at baseline to 31° post-operatively and 41° at the latest follow-up. The mean increase in spine height post-operatively was statistically significantly smaller in the MAGEC group compared with the conventional growth rod group (18 mm compared with 41 mm; $p=0.04$). The mean change in spine height from baseline to final follow-up between the 2 groups was also statistically significantly smaller in the MAGEC group compared with the conventional growth rod group (38 mm compared with 77 mm respectively; $p=0.01$). Mean annual total spine growth was less in the MAGEC group (7 mm per year)

compared with the conventional growth rod group (11 mm per year), but this difference was not statistically significant. The authors suggested that the initial surgery could have contributed to the difference in increase in total spine height from baseline to final follow-up, which could be a result of differences in surgical technique across the participating sites. Children in the MAGEC group needed 4 revision operations compared with 17 for those in the conventional growth rod group. In the MAGEC group, 14 complications occurred, of which 10 were implant-related; in the conventional growth rod group, 23 complications occurred, of which 15 were implant-related. Children in the MAGEC system group had fewer surgical site infections (1 compared with 3).

3.10 Yoon et al. (2013) reported on a retrospective case series evaluating pulmonary function in 6 children with early-onset scoliosis secondary to neuromuscular disease treated with the MAGEC system in a UK hospital. The mean age at the time of surgery was 7.5 years and mean follow-up was 2.5 years. Pulmonary function tests were carried out before and after the insertion of the MAGEC rods and at each distraction. The authors reported that the severity of lung function deficit for 2 of the children treated meant that they would not have been able to receive conventional growth rods, but no further details of the children's condition were provided. Other outcomes measured were Cobb angle, amount of rod lengthening and total spine height, but these results were included in the study by Dannawi et al. (2013). Dual rods were implanted in 5 of the 6 children. An average of 7 distractions per child was carried out during the study period. Forced vital capacity (FVC) increased from 27% to 41% of predicted value[‡] (p=0.028) and forced expiratory volume in 1 second (FEV1) increased from 27% to 45% of predicted value (p=0.027). Coronal and sagittal Cobb angle, thoracic kyphosis and spine height all showed improvement and the authors reported a moderate positive trend between the amount of distraction and the observed improvement in FVC.

3.11 The sponsor carried out meta- and qualitative analyses of the clinical evidence, including 4 studies of the MAGEC system (Akbarnia et al. 2013a; Cheung et al. 2012; Dannawi et al. 2013 and Ellipse 2013) and 1 study (Bess et al. 2010) which evaluated complication rates for 140 children treated with conventional growth rods. The conventional growth rod arm of the study by Akbarnia et al. (2013b) was also included. The outcomes considered in the meta-analysis

were Cobb angle, and thoracic and total spine heights. Qualitative analysis was carried out on infection rates, number of operations, distraction rates and device failure.

3.12 In addition to the adverse events reported in the clinical evidence, the sponsor described 12 adverse events in its submission that had been reported to regulatory agencies. These 12 events related to broken or malfunctioning rods and resulted in 4 revision operations. The External Assessment Centre sought clinical expert opinion on the relative safety of the MAGEC system compared with conventional growth rods; the experts did not describe any significant safety concerns.

3.13 The External Assessment Centre carried out revised meta- and quantitative analyses, in order to consider a broader range of evidence on conventional growth rods. The External Assessment Centre carried out a systematic literature review including data from 15 studies on conventional growth rods, along with the conventional growth rod arm of the Akbarnia et al. (2013b) study. The analyses included the same 4 MAGEC system studies used by the sponsor.

3.14 The External Assessment Centre noted several differences between the populations in the MAGEC system studies and the conventional growth rod studies at baseline, including a lower mean Cobb angle (65.7° and 72.4° respectively), a greater mean total spine height (288 mm and 258 mm respectively) and a difference in mean age at the time of rod insertion (8.0 years and 6.4 years respectively). The External Assessment Centre considered that these differences were likely to have influenced the potential change in each outcome over time. For example, greater height at baseline may limit the potential for growth during follow-up. Similarly, a less severe Cobb angle may limit the potential for improvement in Cobb angle measurement. Inclusion criteria in the conventional growth rod studies varied, with some using Cobb angle and others using age or progression of disease. Dual rods were used in approximately 64% of children involved in the study. Mean duration of follow-up was 4.3 years, which is longer than that of any of the MAGEC system studies (mean 2.5 years). However, the External Assessment Centre considered the studies to be sufficiently homogenous in

terms of population, intervention, setting, study design and outcomes to be included in a meta-analysis.

3.15 The conventional growth rod studies were considered in 2 groups, depending on the duration of follow-up: less than 38 months or 38 months and over. The period of 38 months was selected after considering the mean and range of follow-up times. The aim was to allow a more direct comparison with the shorter follow-up in the MAGEC system studies. The outcomes considered were Cobb angle, total spine height and infection rate. Heterogeneity was measured for each outcome and results from fixed and random effects models were presented. All 4 MAGEC system studies were included in the meta-analysis for each outcome, but conventional growth rod studies were only included if they provided usable data for the particular outcome. Quantitative analysis was conducted for the number of surgical procedures (per child and per year), rate of distraction and rate of device failure.

3.16 Findings from the External Assessment Centre's revised meta-analysis and quantitative analysis are presented in tables 1 and 2. The mean change in Cobb angle from baseline was 27° for the MAGEC system studies and 32° for the conventional growth rod longer follow-up studies, with low heterogeneity between studies. The External Assessment Centre considered that the figures for these 2 groups were unlikely to be comparable because of the difference in follow-up time. The shorter follow-up study reported a mean change in Cobb angle of 37°, but this showed variation from the other conventional growth rod studies. The MAGEC studies showed a mean change in total spine height of 4.55 cm with very low heterogeneity. Heterogeneity was very high between the conventional growth rod studies, indicating clinical or methodological diversity across the studies and so limiting the usefulness of the results. There were also differences in age range between the studies. The mean change in total spine height was 6.43 cm for the longer follow-up group and 10.76 cm for the shorter follow-up group. The MAGEC studies showed moderate heterogeneity and a mean infection rate per patient of 3–4%. The infection rates in the conventional rod groups varied from 3–17%, but these rates were complicated by limited reporting in the included studies, with uncertainty around the type of infection included.

3.17 Results of the External Assessment Centre's revised quantitative analysis showed that the number of surgical procedures per child was less in the MAGEC system group (1.2) than in both the shorter and longer follow-up conventional growth rod groups (4.3 and 5.8 respectively). Calculated annual procedure rates per child (including initial surgery) were 0.9 for the MAGEC system group and 1.5 and 1.1 for the shorter and longer follow-up conventional growth rod groups respectively. The mean number of distractions per child was higher in the MAGEC system group than in the conventional growth rod groups, at 6.2 compared with 4.2 (shorter follow-up) and 4.6 (longer follow-up). The only type of device failure reported was rod breakage. The mean rates of device failure per child were lower in the MAGEC system group than in either conventional growth rod group (MAGEC 6%, shorter follow-up conventional growth rods 13%, longer follow-up conventional growth rods 31%). The annualised rates were around 4.5% in the MAGEC system and shorter follow-up conventional growth rod groups and 7.2% in the longer follow-up conventional growth rod group. The External Assessment Centre noted that these results may reflect the more frequent use of single rods rather than dual rods in the past, which are known to have a higher risk of failure. It noted that results may also have been influenced by advances in rod design, mainly relating to the use of stainless steel rods in the past compared with the stronger titanium rods currently used.

Table 1 Results of the External Assessment Centre's revised meta-analysis

	Number of studies included	Heterogeneity I^{2*}	Fixed effects model or single study (mean, lcl–ucl)	Random effects model (mean, lcl–ucl)
Cobb angle (º)				
MAGEC system	4	44.89	27.16 (24.41–29.92)	27.17 (23.12–31.22)
CGR shorter follow-up	1	N/A (1 study)	37.03 (27.26–46.80)	–
CGR longer follow-up	4	34.83	32.14 (28.91–35.36	32.90 (28.61–37.18)

Change in total spine height (cm)				
MAGEC system	4	0	4.55 (3.98–5.11)	4.55 (3.98–5.11)
CGR shorter follow-up	2	92.33	12.29 (11.16–13.43)	10.76 (5.53–15.98)
CGR longer follow-up	3	96.5	4.25 (3.77–4.72)	6.43 (2.70–10.15)
Infection rates (% per patient)				
MAGEC system	4	61.68	0.03 (0.00–0.08)	0.04 (0.00–0.15)
CGR shorter follow-up	2	83.75	0.03 (0.00–0.08)	0.03 (0.00–0.25)
CGR shorter follow-up (Zhao et al. [2012] study only)	1	N/A (1 study)	0.12 (0.03–0.31)	–
CGR longer follow-up	8	57.33	0.14 (0.11–0.16)	0.15 (0.11–0.20)
CGR longer follow-up (without Kabirian [2012a] study)	7	38.74	0.16 (0.13–0.20)	0.17 (0.13–0.21)

*Values for measuring heterogeneity between studies: <50% is low, 50–70% is moderate and >70% is high

CGR, conventional growth rods; lcl, lower confidence interval; ucl, upper confidence interval

Table 2 Results of the External Assessment Centre's revised quantitative analysis

	Number of patients	Total number of outcomes	Mean per patient	Mean per patient per year	Average interval
Surgical procedures					

MAGEC	80	95	1.2	–	–
CGR shorter follow-up	78	336	4.3	–	–
CGR longer follow-up	264	1523	5.8	–	–
Distractions					
MAGEC	80	496	6.2	4.5	2.3
CGR shorter follow-up	30	125	4.2	1.4	8.6
CGR longer follow-up	555	2557	4.6	1.1	9.2
Device failure			**% per patient**		
MAGEC	80	5	6.3%	–	–
CGR shorter follow-up	103	13	12.6%	–	–
CGR longer follow-up	808	254	31.4%	–	–
CGR, conventional growth rods					

Committee considerations

3.18 The Committee considered that the clinical evidence was limited because it was restricted to small observational studies, and so was insufficient in quantity or quality to establish clinical superiority of the MAGEC system compared with conventional growth rod systems for lengthening the spine and reducing spinal curvature in children with scoliosis. However, it judged that the evidence provided by the various studies, taken together with clinical expert advice, was sufficient to demonstrate the clinical non-inferiority of the MAGEC system with the conventional growth rod systems.

3.19 The Committee was advised that the population of children with scoliosis is heterogeneous, because scoliosis has multiple causes and children may have different comorbidities and magnitudes of curvature. The Committee considered that this was an important confounding factor in the clinical studies. The Committee also noted that the differences in baseline characteristics such as age, Cobb angle and spine height between the children treated with the MAGEC system and conventional growth rods in the available clinical studies could have affected the results. The Committee was advised that the age at which growth rods are first inserted will affect the growth potential of the child's spine, with younger children having more growth potential than older children.

3.20 The Committee was advised that the MAGEC growth rods are functionally similar to conventional rods and are attached to the spine in the same way. It was also advised by experts that the initial reduction in Cobb angle at the time of insertion of growth rods should be similar for the 2 systems. The Committee noted that the External Assessment Centre's review of the clinical evidence indicated that the device failure rate of the MAGEC system rods was at least equivalent to that of conventional growth rods.

3.21 The Committee was advised that Cobb angle and spine height are often difficult to measure and are subject to variation in interpretation when viewing images of the spine. Nevertheless, experts stated that these measures are currently the most reliable indicators of changes in spinal curvature and growth of the spine for children with scoliosis.

3.22 The Committee judged that, because of the uncertainty around the clinical evidence and the available measures, a system for capturing data on individual cases – such as a register – would be useful to expand the evidence base for the use of growth rods, including the MAGEC system. Experts stated that a British Spine Registry was set up by the British Association of Spinal Surgeons in 2012, and it is hoped that this will include data on children with scoliosis. The Committee thought that this would be very valuable in view of the clinical data uncertainties.

3.23 The Committee considered that the MAGEC system had significant potential to improve quality of life for children needing surgical treatment for scoliosis and

for their families or carers. In particular, it noted the benefits associated with avoiding the repeated surgical procedures for lengthening conventional growth rods. These included outcomes such as less pain, less time in hospital and less time away from usual activities, as well as a reduced risk of infection and less scarring. In addition, the Committee noted the potential for less psychological distress, mainly fear and anxiety about the lengthening procedures. The Committee heard patient expert contributions describing the positive attitude of children going to hospital for distraction procedures with the MAGEC system, compared with the distress they experienced when attending for surgical lengthening of conventional growth rods. The Committee was told that grouping outpatient appointments together for children with the MAGEC system rods can not only enhance efficiency (because the external remote control can be used for a number of children in a single clinic), but has the additional benefit of allowing the children to interact, reducing their sense of isolation.

3.24 The Committee recognised that the MAGEC system offers the possibility of more frequent and gradual distraction of the spine than conventional growth rods. The Committee heard clinical expert advice that the optimal frequency of distractions and the amount of lengthening done at each appointment remains uncertain and that practice is evolving: specifically, the frequency of distractions may increase in future. The Committee was clear that any increase in the frequency of distractions would need to take into account the increased radiation exposure from multiple imaging procedures (see section 3.27).

3.25 The Committee noted that the MAGEC system may offer particular advantages for some children who are at high risk from repeated surgical procedures, by removing the need for those procedures and the associated risk of complications. The Committee was also advised that the MAGEC system may provide an option for children for whom treatment with growth rods would otherwise be considered unsuitable because of the risks from repeated surgical lengthening procedures. This may offer benefits to those children (in terms of their quality of life) and to the healthcare system (in terms of less need for treatment of associated conditions, such as chest infections). However, this group was outside the scope of this evaluation.

3.26 The Committee heard clinical expert advice that the MAGEC system may be unsuitable for use in children with severe kyphotic curves, because a flat section of spine is needed for distraction with the MAGEC system. The Committee considered that the decision to use the MAGEC system or conventional rods would depend on the individual child's condition and the wishes of the child and their family, after careful discussion of the available options.

3.27 The Committee considered the potential impact of additional imaging associated with more frequent distractions. The Committee was advised that X-rays were not always used after each distraction and that ultrasound may be an option for more frequent distractions. The Committee was also advised that the use of magnetic resonance imaging (MRI) is currently contraindicated for children with MAGEC system rods in place, but that research is in progress to determine whether any level of MRI can be carried out safely and effectively in these children. The Committee considered that further evidence should be developed about the relative merits of X-rays, MRI and other imaging techniques such as ultrasound in children being treated with the MAGEC system.

[1] The test result is often shown as a percentage of the 'predicted values' for patients of similar characteristics. A result close to 100% is considered to be normal, although results over 80% are often also considered normal.

4 NHS considerations

System impact

4.1 The sponsor claimed that the MAGEC system reduces the number of surgical procedures needed to treat children with scoliosis compared with conventional growth rods, because rod lengthenings and distractions are carried out non-invasively in an outpatient setting. This reduces resource use such as operating theatre and clinical time, as well as reducing the risk of complications needing hospital treatment, such as side effects from anaesthesia or infection.

Committee considerations

4.2 Having reviewed the clinical evidence and expert advice, the Committee was satisfied that using the MAGEC system could reduce the number of surgical procedures for each child by avoiding the need for operations to lengthen conventional growth rods. This would also reduce associated NHS resource use.

4.3 The Committee was advised that the population of children for whom the MAGEC system would be considered is small, but that the potential for releasing operating theatre and clinical time and shortening hospital stays by avoiding repeated surgical procedures could be substantial. The Committee also noted that resource use for treating complications associated with surgery, such as infection, could be reduced.

4.4 The Committee was advised that MAGEC growth rod lengthenings are often carried out in a single clinic, specially arranged so that a number of children can be treated. This allows more effective use of resources through reduced courier costs for the external remote control.

4.5 The Committee was advised that the MAGEC system may reduce the need for spinal cord monitoring and its associated costs. Spinal cord monitoring is used to assess neurological function during insertion of MAGEC or conventional growth rods, and in some children during conventional growth rod lengthening.

However, it is not needed during lengthening of the MAGEC system rods because the child is awake and any changes in function can be observed.

5 Cost considerations

Cost evidence

5.1 The sponsor presented 1 unpublished economic study set in an English hospital as evidence for the evaluation (Richards and Nnadi 2013), which was submitted to the Committee as academic-in-confidence. The External Assessment Centre judged that the study was relevant to the scope of the evaluation and found no additional relevant studies.

5.2 The sponsor submitted a de novo cost analysis comparing the cost consequences of using the MAGEC system with 1 type of conventional growth rod (Expedium 4.5 Spine System, Depuy Synthes). Costs were modelled from an NHS and personal social services perspective. The population included in the model was children aged 2 to 11 years with severe early-onset scoliosis. The model adopted a cost-minimisation approach based on an assumption of equivalent clinical efficacy between the MAGEC system and conventional growth rods. The model included the cumulative costs associated with initial surgery, device failure and rod lengthenings over a 6-year time horizon.

5.3 The model included 2 clinical parameters: device failure rate and frequency of lengthening. The sponsor used a device failure rate of 0% for both the MAGEC and conventional growth rods in its base-case analysis, based on clinical evidence. Frequency of lengthening was assumed in the model to be 3-monthly for the MAGEC system. Conventional growth rods were assumed to be lengthened every 6 months.

5.4 The sponsor took most of the resource use figures for its model from the study by Richards and Nnadi (2013). The cost of device failure in the sponsor's model was estimated to be around £11,000 less than the cost of initial insertion for both the MAGEC system and conventional growth rods, based on complete replacement of the rods. This cost included device costs and surgical time only and did not take into account other costs incurred such as pre- or post-operative care. A cost per lengthening was also included, based on an average of 2 figures taken from an NHS trust and from Richards and Nnadi (2013) for the MAGEC system and from Richards and Nnadi (2013) only for conventional

growth rods. These costs are currently academic-in-confidence and cannot be reported here.

5.5 The sponsor carried out sensitivity analyses by varying the device failure rate. The device failure rate was 0% in the base-case analysis and 8.8% and 17.2% in 2 scenario analyses. All device failures were assumed to occur in the first month only. A third sensitivity analysis was included, using a device failure rate of 8.8% with an additional failure rate for conventional growth rods of 176% per child at month 13, based on a study by Yang et al. (2011). The External Assessment Centre noted that this figure was calculated based on the total number of rod fractures in children experiencing any rod fractures (86 fractures in 49 children), and not the study population as a whole (327 children), which would give an overall rod fracture rate of 15%.

5.6 The results of the sponsor's base case suggested that the MAGEC system was cost saving at 6 years, with a break-even point 39 months after initial insertion. From the sponsor's model, the cost saving for the MAGEC system compared with conventional growth rods at 6 years was £9946. Results of the sensitivity analysis indicated that the model was robust and the MAGEC system remained cost saving over the 6-year time horizon. The month of break-even varied from 28 to 45 and the cost savings ranged from £8109 to £12,984.

5.7 The External Assessment Centre considered that the sponsor's model used an appropriate treatment pathway and captured key aspects of treatment. However, it noted a weakness of the model was the assumption of clinical equivalence between the MAGEC system and conventional growth rods. The External Assessment Centre considered that the available evidence did not support this assumption.

5.8 The External Assessment Centre also considered that many of the inputs to the sponsor's model were incorrect. In some instances it judged that the sponsor had taken the wrong cost from Richards and Nnadi (2013), and in others that the Personal Social Services Research Unit (2012) unit costs of health and social care would have been more accurate. Sensitivity analyses were only carried out on 1 parameter in the sponsor's model and no adverse

events other than complete device failure in the first month were considered. The External Assessment Centre considered that the costs for insertion should also be used when device failure occurred. The External Assessment Centre also noted that no discounting was applied in the sponsor's model.

5.9 The External Assessment Centre revised the sponsor's model to address some of the limitations identified:

- A monthly device failure rate was applied throughout the 6-year time horizon, taken from clinical evidence.
- Surgical site infections were also included in the model. The infection rate was taken from clinical evidence and included at month 0 for the MAGEC system and as a monthly rate for conventional growth rods to account for lengthening procedures.
- The difference in the proportion of dual and single rods was introduced into the model for both systems, based on the proportions used in the clinical evidence. Dual rods were assumed to be used in 65% of children with scoliosis and costs were weighted accordingly.
- A proportional cost for distraction was included at each month to allow for wider sensitivity analysis. Distraction was assumed to take place every 3 months for MAGEC and every 6 months for conventional growth rods.
- The costs for initial rod insertion and for complete device failure were assumed to be the same. This was calculated as £27,431 for the MAGEC system and £15,270 for conventional growth rods.
- Costs were discounted by 3.5% in line with the NICE medical technologies evaluation methods guide.

5.10 Results of the base case in the revised model showed that the MAGEC system generated cost savings of £12,077 after 6 years compared with conventional growth rods. The findings showed that the MAGEC system would become cost saving at month 35 after insertion.

5.11 The External Assessment Centre carried out extensive 1- and 2-way sensitivity analyses on most of the model inputs. This included varying: the costs and

months between distractions, cost and rate of device failure, costs of insertion and device failure for each system, and costs and rates of infection.

5.12 The results of the 1-way sensitivity analysis showed that the cost difference was most sensitive to the cost of and interval between distractions for conventional growth rods. Varying the cost of conventional growth rod distraction to its lowest value, £1133, led to the MAGEC system incurring costs after 6 years. Varying the months between distractions indicated that the MAGEC system would incur costs if the interval between conventional growth rod distractions was 10.2 months or more. Expert advice obtained by the External Assessment Centre was that conventional growth rod lengthening generally occurs at 6-monthly intervals in the NHS and so this would make the MAGEC system likely to be cost saving. In all other scenarios examined, the MAGEC system remained cost saving.

5.13 In the 2-way sensitivity analyses, varying the cost of conventional growth rod lengthening to the lower range of values and increasing the months between distractions caused the MAGEC system to become cost-incurring. This is consistent with results from the 1-way sensitivity analysis. According to the model, if conventional growth rod distractions were carried out every 6 months, the cost of distraction surgery would have to be below £1579 per episode before the MAGEC system incurred costs. Based on clinical advice and cost data, the External Assessment Centre judged that the average cost of distraction surgery was unlikely to be this low in practice.

5.14 Lowering the insertion cost of conventional growth rods and raising it for the MAGEC system caused the MAGEC system to become cost-incurring. However, the External Assessment Centre obtained expert advice which indicated that, in practice, the resource use for initial insertion of MAGEC system rods was likely to be equivalent to that of conventional growth rods. Therefore, it was unlikely that the cost of inserting the MAGEC system would be so much more than the cost of inserting conventional growth rods that it would become cost-incurring. The External Assessment Centre judged it unlikely that the cheapest available conventional growth rods would be used in all instances, also based on clinical expert advice.

5.15 The External Assessment Centre also varied the time horizon for the model in light of advice from 2 clinical experts who stated that the overall treatment time would vary depending on the child's age at the start of treatment and the time to achieve spinal maturity. Furthermore, rods may need replacement if the child's spinal growth exceeds the maximum lengthening capacity of the rods. The External Assessment Centre carried out analysis to explore the impact of replacing rods at different time periods between 3.5 and 5 years after initial insertion. Both systems were assumed to be replaced in the same month. Rod replacement meant that the MAGEC system would become cost saving at around 68 months (5.5–6 years) after initial insertion. If the duration of treatment was longer, the cumulative cost savings would increase over time. The findings also suggested that the MAGEC system would generate cost savings if used to replace conventional growth rods in children with more than 35 months of growth potential remaining. The External Assessment Centre judged that based on clinical and economic evidence and clinical expert advice, the typical length of treatment using growth rods is likely to be 35 months or longer. This indicates that the MAGEC system would be cost saving when compared with conventional growth rods, as in the base case.

5.16 The External Assessment Centre concluded that many of the uncertainties around the model inputs had been addressed in the sensitivity analysis and that the results of the base case were robust. The External Assessment Centre also noted that the results from the revised model were similar to the sponsor's base case despite revisions to many of the inputs. However, the External Assessment Centre acknowledged that the model assumed clinical equivalence between the MAGEC system and conventional growth rods when the available clinical evidence could not inform this assumption. It also noted that many of the model costs were taken from 1 unpublished study (Richards and Nnadi 2013), which may not be generalisable because this study was set in a single centre.

Committee considerations

5.17 The Committee noted that the revisions to the model prepared by the External Assessment Centre relied on costs from 1 unpublished study. However, the Committee considered that the extensive sensitivity analyses conducted by the External Assessment Centre and the expert advice it sought, addressed many

of the uncertainties in the economic evidence. The Committee judged the findings from these revisions to be sufficiently robust to allow a decision to be made and concluded that cost savings were likely to be realised in practice.

5.18 The Committee heard expert advice that the frequency of distractions using the MAGEC system may increase in future as clinical use of the product develops. The External Assessment Centre therefore recalculated the findings from the revised model using a 6-week distraction interval and concluded that even with more frequent distractions, the MAGEC system would still generate cost savings of more than £7000 over the 6-year time horizon.

5.19 The Committee considered the impact of the potential reduction in spinal cord monitoring during rod lengthening procedures (see section 4.5), which had not been considered in the sponsor's model. The External Assessment Centre investigated the use of spinal cord monitoring in conventional rod lengthenings and found variation in practice, with some centres using it routinely and others not using it at all. It also found that the modality of spinal cord monitoring varied. Through a review of the available literature, the External Assessment Centre identified a range of spinal cord monitoring costs. It subsequently incorporated these costs in the revised model for all conventional growth rod lengthenings. It found that not using spinal cord monitoring for MAGEC system rod lengthenings would generate cost savings of £13,170 per child over the 6-year time horizon if spinal cord monitoring cost £120 per lengthening procedure (the lowest cost identified), or £15,327 per child if it cost £343 per lengthening procedure (the highest cost identified). The Committee concluded that the base-case saving of around £12,077 per child after 6 years was likely to be conservative because it did not include the impact of these costs.

5.20 The Committee noted that it may not be cost saving to use the MAGEC system in older children with less than 35 months' growth potential, because the system is estimated to become cost saving only after about 35 months. The Committee judged that use of the system in older children would need careful patient selection, with consideration of individual circumstances and benefits.

6 Conclusions

6.1 The Committee concluded that the available clinical evidence, together with expert clinical advice and technical information, showed that the MAGEC system is an effective treatment for children with scoliosis for whom surgery is considered necessary. The Committee also concluded that the MAGEC system is likely to provide benefits compared with the use of conventional growth rods. These benefits would arise from avoiding repeated surgical procedures for growth rod lengthening, which would reduce pain and psychological distress, shorten hospital stays, and result in less time away from usual activities for children with scoliosis and their families or carers. In addition, the risk of complications such as infection would be reduced.

6.2 The Committee accepted the revised model and sensitivity analyses and concluded that the MAGEC system could generate substantial cost savings after about 3 years when compared with conventional growth rod treatment.

Sir Andrew Dillon

Chief Executive

June 2014

7 Committee members and NICE lead team

Medical Technologies Advisory Committee members

The Medical Technologies Advisory Committee is a standing advisory committee of NICE. A list of the Committee members who took part in the discussions for this guidance appears below.

Committee members are asked to declare any interests in the technology to be evaluated. If it is considered there is a conflict of interest, the member is excluded from participating further in that evaluation.

The minutes of each Medical Technologies Advisory Committee meeting, which include the names of the members who attended and their declarations of interests, are posted on the NICE website.

Professor Bruce Campbell (Chair)
Consultant Vascular Surgeon, Exeter

Dr Peter Groves (Vice Chair)
Consultant Cardiologist, Cardiff and Vale NHS Trust

Professor Dilly Anumba
Chair of Obstetrics and Gynaecology/Honorary Consultant Obstetrician and Gynaecologist, University of Sheffield

Ms Susan Bennett
Lay member

Dr Keith Blanshard
Consultant Interventional Radiologist, University Hospitals of Leicester NHS Trust

Mr Matthew Campbell-Hill
Lay member

Professor Daniel Clark
Head of Clinical Engineering, Nottingham University Hospitals NHS Trust

Professor Tony Freemont
Professor of Osteoarticular Pathology, University of Manchester

Professor Peter Gaines
Consultant Vascular Interventional Radiologist, Sheffield, Vascular Institute and Sheffield Hallam University

Professor Shaheen Hamdy
Professor of Neurogastroenterology, University of Manchester

Dr Jerry Hutchinson
Independent Medical Technology Adviser

Dr Cynthia Iglesias
Health Economist, University of York

Professor Mohammad Ilyas
Professor of Pathology, University of Nottingham

Dr Greg Irving
General Practitioner and Clinical Lecturer, University of Cambridge

Dr Eva Kaltenthaler
Reader in Health Technology Assessment, ScHARR, University of Sheffield

Dr Paul Knox
Reader in Vision Science, University of Liverpool

Mrs Jacqui Nettleton
Programme Director, Commissioning, Western Sussex Hospitals NHS Trust

Professor Brian J Pollard
Professor of Anaesthesia, University of Manchester. Consultant Anaesthetist, Central Manchester University Hospitals

Mr Brian Selman
Managing Director, Selman and Co

Professor Wendy Tindale
Scientific Director, Sheffield Teaching Hospitals NHS Foundation Trust

Professor Allan Wailoo
Professor of Health Economics, School of Health and Related Research (ScHARR), University of Sheffield

Mr John Wilkinson
Director of Devices, Medicines and Healthcare Products Regulatory Agency

Dr Janelle Yorke
Lecturer and Researcher in Nursing, University of Manchester

NICE lead team

Each medical technology assessment is assigned a lead team of a NICE technical analyst and technical adviser, an expert adviser, a technical expert, a patient expert, a non-expert member of the Medical Technologies Advisory Committee and a representative of the External Assessment Centre.

Joanne Higgins
Technical Analyst

Bernice Dillon
Technical Adviser

Jeremy Fairbank
Lead Expert Adviser

Neil Davidson
Lead Expert Adviser

Jane Clarke
Patient Expert

Eva Kaltenthaler
Non-Expert MTAC Member

Joyce Craig
External Assessment Centre Representative

Michelle Jenks
External Assessment Centre Representative

8 Sources of evidence considered by the Committee

The External Assessment Centre report for this assessment was prepared by Newcastle upon Tyne Hospitals and York Health Economics Consortium:

- Craig J, Jenks M, Willits I et al. MAGEC system for spinal lengthening in children with early onset scoliosis, November 2013

Submissions from the following sponsor:

- Ellipse Technologies Inc.

The following individuals gave their expert personal view on the MAGEC system by providing their expert comments on the draft scope and assessment report.

- Mr Sashin Ahuja, ratified by the British Scoliosis Society – clinical expert
- Mr Colin Bruce, nominated by the British Society for Children's Orthopaedic Surgery - clinical expert
- Jane Clarke, nominated by the Scoliosis Association UK – patient expert
- Mr Neil Davidson, ratified by the British Scoliosis Society – clinical expert
- Professor Jeremy Fairbank, ratified by the Royal College of Surgeons of England - clinical expert
- Mr John Hutchinson, ratified by the British Scoliosis Society – clinical expert
- Mr Peter Milner, ratified by the British Orthopaedic Association – clinical expert
- Mr Colin Nnadi, ratified by the British Scoliosis Society – clinical expert (draft scope only)
- Mr Hilali Nordeen, ratified by the British Scoliosis Society – clinical expert

The following individuals gave their expert personal view on the MAGEC system in writing by completing a patient questionnaire or expert adviser questionnaire provided to the Committee.

- Mr Sashin Ahuja, ratified by the British Scoliosis Society – clinical expert

- Mr Colin Bruce, nominated by the British Society for Children's Orthopaedic Surgery - clinical expert
- Jane Clarke, nominated by the Scoliosis Association UK – patient expert
- Mr Neil Davidson, ratified by the British Scoliosis Society – clinical expert
- Professor Jeremy Fairbank, ratified by the Royal College of Surgeons of England - clinical expert
- Mr John Hutchinson, ratified by the British Scoliosis Society – clinical expert
- Mr Peter Milner, ratified by the British Orthopaedic Association – clinical expert
- Mr Colin Nnadi, ratified by the British Scoliosis Society – clinical expert
- Mr Hilali Nordeen, ratified by the British Scoliosis Society – clinical expert

About this guidance

This guidance was developed using the NICE medical technologies guidance process.

It has been incorporated into the NICE pathway on musculoskeletal conditions, along with other related guidance and products.

We have produced a summary of this guidance for the public. Tools to help you put the guidance into practice and information about the evidence it is based on are also available.

Related NICE guidance

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