

What is the incidence of late detected developmental dysplasia of the hip (DDH) in England?

A 26-year national study of children diagnosed over 1 year of age

Broadhurst C (MMedSc)¹, Rhodes A (MRCS)², Harper P (MMedSc)¹, Perry DC (FRCS)^{3,4}, Clarke NMP (FRCS)⁵, Aarvold A (FRCS)⁶

¹ Final year medical student, Faculty of Medicine, University of Southampton, University Road, Southampton SO17 1BJ, UK

² Academic Clinical Fellow, Orthopaedic Surgery, University Hospital Southampton, Tremona Road, Southampton SO16 6YD, UK

³ Associate Professor, Oxford Trauma, NDORMS, University of Oxford, Oxford,

⁴ Honorary Consultant Paediatric Orthopaedic Surgeon, Alder Hey Children's Hospital, Liverpool, L12 2AP.

⁵ Emeritus Professor of Orthopaedic Surgery, University of Southampton, University Road, Southampton SO17 1BJ

⁶ Honorary Associate Professor and Consultant Paediatric Orthopaedic Surgeon, Southampton Children's Hospital, University Hospital Southampton, Tremona Road, Southampton SO16 6YD, UK

Correspondence to:

Alexander Aarvold

Email: alexaarvold@doctors.org.uk

Address: Southampton Children's Hospital, University Hospital Southampton, Tremona Road, Southampton SO16 6YD, UK.

Contributors

Contributors CB, DCP, NMPC and AA were responsible for designing the methodology for the study. CB and PH completed the whole cohort data analysis. CB, PH and AR created the figures used in the results. CP and AR wrote the first draft of the manuscript, which was edited, contributed to and approved by all authors (CB, AR, PH, DCP, NMPC and AA). AA and NMPC managed the overall process.

Funding

This project was funded by the Southampton Children's Hospital Paediatric Orthopaedic Research Fund. This article presents independent research supported by a National Institute for Health Research (NIHR) clinician scientist fellowship (to Daniel C Perry; grant number NIHR/CS/2014/14/012). All authors carried out this research independently of the funding bodies. The findings and conclusions in this report are those of the authors and do not necessarily represent the views of the UK Department of Health.

Abstract

Aim

To establish the incidence of DDH diagnosed after one-year of age in England, stratified by age, sex, year and region of diagnosis

Methods

A descriptive observational study was performed by linking primary and secondary care information from two independent national databases of routinely collected data: the UK Clinical Practice Research Datalink (CPRD) and Hospital Episode Statistics (HES). The study examined all children from 01/01/1990 to 01/01/2016 who had a new first diagnostic code for DDH aged between one and eight years old.

Results

The incidence of late-diagnosed DDH was 1.28 cases per 1000 live births. Within the study population, 754 children were identified with a diagnosis of DDH after one-year of age. Of all late diagnoses, 536 (71.1%) were detected between 1-2 years of age. The ratio of females to males was 4.2:1. Distribution was evenly spread throughout England.

Conclusion

The incidence of late-diagnosed DDH has not been reduced from that reported forty years ago, prior to the introduction of the national selective screening programme for DDH.

Clinical relevance

- Provides a detailed up to date analysis, against which previous decades' and future incidence rates may be compared
- Adds to the ongoing unresolved debate regarding DDH screening in the UK

Introduction

Developmental dysplasia of the hip (DDH) is a significant public health issue, representing the single largest cause for total hip arthroplasty in young adults.^{1,2} If detected early in infancy the problem is often treated by a removable splint that is worn for 2-3 months. Diagnoses made in infants older than a few months of age are associated with increased rates of surgery, longer hospital stays, increased healthcare costs and long-term complications.³⁻⁶ The later the diagnosis, the more invasive the surgical intervention, ultimately involving surgery to the joint, the pelvis and the femur. Long-term prognosis is related to the time of treatment, with earlier treated hips having more favourable outcomes.⁷

In England, selective screening has formed part of the NHS Newborn & Infant Physical Examination (NIPE) programme since 1986, guided by the Standing Medical Advisory Committee (SMAC).⁸ Despite these screening programmes, late-diagnosed DDH and the clinical presentation of dislocated hips in children over one year of age remains a persistent reason for referral to paediatric orthopaedic units.⁹ Many of these children do not have risk factors identified by the selective screening programme. Litigation regarding missed and misdiagnosed DDH cases is increasing; representing the third most frequent cause of litigation within paediatric orthopaedics.¹⁰ In contrast, other countries including Austria and Germany undertake 'universal screening' by performing an ultrasound scan (USS) of the hips of all newborns. In such, the incidence of late-diagnosed DDH is negligible, with a low requirement for surgery and fewer hospital admissions related to DDH.^{4,11-13} However, universal USS screening brings significant costs, resource demands, and over-treatment, fuelling a debate regarding universal versus selective screening – which has been unresolved for thirty years since the introduction of ultrasound screening methods. The estimated incidence of late DDH diagnosis in European literature is 0.34 – 2.4/1000 live births,¹⁴⁻¹⁷ however there is no consensus related to how 'late' is defined. There are no UK population based studies concerning the incidence of late-diagnosed DDH. Historic UK studies prior to the introduction of the selective screening programme report an incidence of 0.88 per 1000 births.¹⁸

This study aimed to establish the incidence of DDH diagnosed after one-year of age in England, stratified by age, sex, year and region of diagnosis. The results may be used to inform the ongoing debate regarding hip screening in the UK.

Methods

This is a descriptive observational study, using data from two national UK databases: the Clinical Practice Research Datalink (CPRD) and Hospital Episode Statistics (HES). Ethical approval for the use of CPRD and HES data was obtained from the Medicines and Healthcare Products Regulatory Agency (MHRA) Independent Scientific Advisory Committee (ISAC) on the 20th December 2016.

Data sources

The Clinical Practice Research Datalink (CPRD) provides an anonymised UK-based database funded by the National Institute for Health Research (NIHR) and the Medicines and Healthcare Products Regulatory Agency (MHRA). The CPRD GOLD database contains the longitudinal medical records from 714 primary care providers throughout the UK. Established in 1987, the database now includes active records for over 2.45 million individuals (3.5% of the UK population), and is known to be representative of the national population with respect to age, sex and regional distribution. This database has been validated and proven to provide results that are consistent with other data sources in the UK.¹⁹ It is widely considered the “Gold Standard” database for epidemiological research. Hospital Episode Statistics (HES) provides linked inpatient data on all admissions to NHS secondary care centres in England. All NHS healthcare providers in England, including acute hospital trusts, primary care trusts and mental health trusts have contributed data to HES since 1997.

Case identification and validation

CPRD GOLD was independently and systematically searched by one investigator to generate a list of diagnostic codes pertaining to be indicative of DDH. This list was validated by two further investigators (Appendix A). Cases were extracted from the CPRD GOLD database if they had at least one indicative diagnostic code for DDH, from 1st January 1990 to 1st January 2016, that appeared only after their first birthday.

Cases were extracted from CPRD only if they satisfied all of the following criteria:

- (a) Age above or equal to one year old, and up to eight years old, on the day of the first diagnostic record for DDH.
- (b) Defined as ‘acceptable’ by CPRD; patients with continuous follow-up and good quality data recorded to ensure the validity of that patient’s record (Appendix D).
- (c) The general practice where the patient was registered had at least one year of prior up-to-standard (UTS) data within their computerised record (Appendix D).

The study population was restricted to patients who were eligible for linkage between CPRD GOLD and HES to improve the diagnostic validation within our study population. Eligibility criteria for linkage with HES require the patient to reside in England, and to have consented to the linkage scheme (within General Practices who have opted in to linkage). As such, whilst CPRD GOLD covers the whole UK, this study is restricted to England. All identified cases underwent an internal validation process, achieved by examining diagnostic and procedural codes two years either side of the DDH diagnosis (as defined by the CPRD diagnosis date) within the HES datasets, and examining codes two years following the diagnostic event within CPRD GOLD. The relevant HES diagnostic and procedural codes are represented by International Statistical Classification of Diseases and Related Health Problems (ICD-10) codes and Office of Population Censuses and Surveys (OPCS) codes respectively (Appendix B). The strength of the validation was classified using a validation algorithm, which was developed by three researchers (Appendix C). All cases extracted from CPRD were systematically reviewed according to this algorithm, divided between two researchers.

Exclusions from the data extracted from CPRD:

Prior to dataset delivery, cases were excluded by CPRD if they had specific codes indicative of co-existing neuromuscular disease (Appendix A). During the validation process, exclusion codes were specifically searched for in order to exclude any patients where there was an additional diagnostic code suggestive of either; neuromuscular disease that may precipitate secondary hip dislocation (i.e. a 'teratologic' DDH) or syndromes. In addition, patients were excluded if there was evidence of traumatic hip dislocation without any additional support diagnosing DDH (Appendices A & B).

Case identification process

A total of 14,756,663 'research acceptable' patients were included within the CPRD GOLD database at the time of data extraction. From this, there were 3,635,163 children aged one year or over within England, up to eight years old, between the study period 01/01/1990 – 01/01/2016. Amongst this group, 835 children were identified by CPRD who had a first diagnostic code for DDH after one year of age, and who met all of the CPRD requirements. The validation process excluded a further 81 patients for neuromuscular disease / syndromes / trauma (Appendices A & B) (Figure 1). Table 2 details the number of patients within each validation criteria.

Statistical analysis

Denominators were obtained from CPRD to calculate the annual incidence of disease amongst 1 - 8 year olds stratified by age, sex, year and region. The external validity of the dataset was assessed by comparing the incidence of cases to those published previously. Descriptive statistics including frequencies, percentages and range were applied to the demographic data. Overall and study specific (age, sex, year and region) incidence rates were calculated using specifically calculated 'at risk' populations. Poisson confidence intervals were calculated for rate estimations. Statistical analyses were undertaken using IBM SPSS Statistics (version 24, Armonk, NY: IBM Corp) and Microsoft Excel (version 15.16, Microsoft, Redmond, Washington).

Results

Baseline demographics

Of the 754 patients identified within the study population as having DDH detected between 1 and 8 years of age, 608 (80.6%) were female and 146 (19.4%) were male, giving a female: male ratio of 4.2:1. The majority of patients (n=536, 71.1%) had a first diagnostic record between the ages of one and two years (range 1 - 8 years). Ethnicity was available for 720 (95.5%) patients within HES. The majority of whom were White (n=603, 83.8%), reflecting the population mix in the 2011 UK census (White population of 86.0%).²⁰

Incidence of late diagnoses

The overall incidence of late diagnosed DDH equates to 1.28 cases per 1000 live births, detected during the childhood years of one to eight years old.

The peak incidence of late DDH diagnosis was found to be between the ages of 1 – 2 years; with an incidence of 0.88 (95% CI 0.81, 0.96) cases per 1000 (Table 1). Thereafter the incidence falls, with the lowest between the ages of 7-8 years of age (0.02/1000) (Figure 2). When accounting for an individual's cumulative risk up to 8 years of age, incidence totals 1.28/1000 (Table 1). There were no clear geographical patterns of variation across England (Figure 3, Figure 4). The incidence fell between 1990-1994, and remained static thereafter (1994-2016) (Figure 5).

Discussion

This is the first study to investigate the incidence of late presenting DDH using linked primary and secondary care records, enabling a validated national analysis. To our knowledge this is the largest population study on DDH to date, made feasible by the use of national datasets. A considerable ongoing burden of late-diagnosed DDH was identified, with an incidence similar to that reported prior to the introduction of the selective screening programmes.

The overall incidence of late diagnosis within England is 1.28 per 1000 live births. This is a greater incidence than previously reported 40 years ago in Southampton (1965-1978)²¹ and Bristol (1970-1979)¹⁸, which demonstrated incidences of 0.47 and 0.39 cases per 1000 1-5 year olds, respectively.

Unsurprisingly, the age category of 1 – 2 years showed the highest incidence of late diagnosis. This is consistent with the most common clinical presentation, whereby classically an abnormal gait is noticed after the child has commenced walking. When children up to 3 years of age are included in this group, 86.3% of all late diagnoses are accounted for. This study identified an incidence of late diagnosed DDH in 1 – 2 year olds of 0.88 (95% CI 0.81, 0.96) cases per 1000. This is greater than the reported incidence (0.29/1000) in this age group from the 1970-84 cohort in Avon, England.¹⁸ This finding exacts searching questions regarding the impact, if any, of selective screening on the prevention of late diagnoses.

The early dataset included only a limited number of GP practices, which is reflected by the wide confidence intervals during this period. Whilst the incidence between 1990-1994 appeared higher, the wide confidence intervals during this period means that natural variation within the dataset could account for this.

Whilst this study represents the largest cohort examined to date, there are limitations inherent in the use of routinely collected data. The validity of the study relies on the accuracy of the primary codes. The authors are aware of the wide diagnostic criteria, ranging from an unstable hip to a dislocated hip. However, after the age of one year there is intervention of a similar magnitude required, regardless of whether the hip is dislocated or dysplastic. Unfortunately, NIPE criteria (including breech presentation and family history) are inconsistently coded within the CPRD dataset, so were not able to be explored as risk factors. In addition, the nature of coding within CPRD means we are unable to identify the nature of surgical procedures, which we recognise as a limitation within this study. The major limiting factor is corresponding codes existing in both HES and CPRD in 56.5% of cases. However, even considering only the codes recorded in both CPRD and HES (i.e. the cases with greatest internal validity), the rate of late detected DDH would still be of a similar level to that recorded prior to the introduction of selective hip screening. The index date used was that recorded within CPRD. Delays in communication to the General Practitioner may have introduced delay in the record of the 'index date', however all practices were up-to-standard chosen for the high research-quality of their data.

CPRD age cohorts are limited to annual increments. It was therefore not possible to include those children presenting late but less than 12 months' of age. 'Late' diagnosis may be variably defined, but by not incorporating 6-11 month old infants, the 'late' figure in this manuscript may be an under-estimation. If age was defined by month within the CPRD dataset, or in six monthly intervals, a more accurate picture of later diagnosis may have been provided.

In conclusion, the UK approach of selective screening appears to have had little impact on incidence rates of late diagnosed DDH. Thirty-five years on from published frustrations at the apparent failures of neonatal screening to make a substantial impact, DDH appears to remain a “still uncontrolled disease”.²¹ Urgent research and debate is required to identify the optimal means to screen for DDH, and to identify whether implementing screening at a national level will effectively reduce the societal burden of this disease.

Declaration of interests

All authors confirm no conflicts of interest

Data sharing statement

Patient data collected and analysed for this study, aside from that included in Appendices A-D will not be made available to others.

References

1. **Engesaeter I, Lehmann T, Laborie LB, Lie SA, Rosendahl K, Engesaeter LB.** Total hip replacement in young adults with hip dysplasia: Age at diagnosis, previous treatment, quality of life, and validation of diagnoses reported to the Norwegian Arthroplasty Register between 1987 and 2007. *Acta Orthopaedica*. 2011;82(2):149–154.
2. **Furnes O, Lie SA, Espehaug B, Vollset SE, Engesaeter LB, Havelin LI.** Hip disease and the prognosis of total hip replacements. A review of 53,698 primary total hip replacements reported to the Norwegian Arthroplasty Register 1987-99. *J Bone Joint Surg Br*. 2001;83(4):579–86.
3. **McAllister DA, Morling JR, Fischbacher CM, Reidy M, Murray A, Wood R.** Enhanced detection services for developmental dysplasia of the hip in Scottish children, 1997-2013. *Arch Dis Child*. 2018;0:1–6.
4. **Thallinger C, Pospischill R, Ganger R, Radler C, Krall C, Grill F.** Long-term results of a nationwide general ultrasound screening system for developmental disorders of the hip: the Austrian hip screening program. *J Child Orthop*. 2014;8(1):3–10.
5. **Studer K, Williams N, Antoniou G, et al.** Increase in late diagnosed developmental dysplasia of the hip in South Australia: risk factors, proposed solutions. *Med J Aust*. 2016;204(6):240.
6. **R. Biedermann, J. Riccabona, J.M. Giesinger, A. Brunner, M. Liebensteiner, J. Wansch, D. Dammerer, M. Nogler.** Results of universal ultrasound screening for developmental dysplasia of the hip: A Prospective Follow-Up of 28 091 Consecutive Infants. *The Bone and Joint Journal*. 2018;100-B(10)
7. **Sewell MD, Rosendahl K, Eastwood DM.** Developmental dysplasia of the hip. *BMJ*. 2009;339:b4454.
8. **Standing Medical Advisory Committee, Standing Nursing and Midwifery Advisory Committee Working Party for the Secretaries of State for Social Services and Wales.** Screening for the detection of congenital dislocation of the hip. *Archives of Disease in Childhood*. 1986;61(9):921–926.
9. **Talbot C, Adam J, Paton R.** Late presentation of developmental dysplasia of the hip: a 15-year observational study. *The Bone and Joint Journal*. 2017;99-B(9).
10. **Atrey A, Nicolaou N, Katchburian M, Norman-Taylor F.** A review of reported litigation against English health trusts for the treatment of children in orthopaedics: present trends and suggestions to reduce mistakes. *J Child Orthop*. 2010;4(5):471–6.

11. **von Kries R, Ihme N, Oberle D, et al.** Effect of ultrasound screening on the rate of first operative procedures for developmental hip dysplasia in Germany. *The Lancet*. 2003;362(9399):1883–7.
12. **Kohler G, Hell AK.** Experiences in diagnosis and treatment of hip dislocation and dysplasia in populations screened by the ultrasound method of Graf. *Swiss Med Wkly*. 2003;133(35-36):484–87.
13. **Ihme N, Altenhofen L, von Kries R, Niethard FU.** Hip ultrasound screening in Germany. Results and comparison with other screening procedures. *Orthopade*. 2008;37(6):541–6, 548–9.
14. **Clarke NM, Reading IC, Corbin C, Taylor CC, Bochmann T.** Twenty years experience of selective secondary ultrasound screening for congenital dislocation of the hip. *Arch Dis Child*. 2012;97(5):423–9.
15. **Bjerkreim I, Johansen J.** Late diagnosed congenital dislocation of the hip. *Acta Orthop Scand*. 1987;58(5):504–6.
16. **Heikkila E, Ryoppy S, Louhimo I.** Late diagnosis in congenital dislocation of the hip. *Acta Orthop Scand*. 1984;55(3):256–60.
17. **Kamath S, Mehdi A, Wilson N, Duncan R.** The lack of evidence of the effect of selective ultrasound screening on the incidence of late developmental dysplasia of the hip in the Greater Glasgow Region. *J Pediatr Orthop B*. 2007;16(3):189–91.
18. **Dunn PM, Evans RE, Thearle MJ, Griffiths HE, Witherow PJ.** Congenital dislocation of the hip: early and late diagnosis and management compared. *Arch Dis Child*. 1985;60(5):407–14.
19. **Khan NF, Harrison SE, Rose PW.** Validity of diagnostic coding within the General Practice Research Database: a systematic review. *Br J Gen Pract*. 2010;60(572):e128–36.
20. **Office for National Statistics (ONS).** Ethnicity and National Identity in England and Wales: 2011 - licensed under the Open Government Licence v.3.0. <https://www.ons.gov.uk/>. Accessed 5/10/2018.
21. **Catford JC, Bennet GC, Wilkinson JA.** Congenital hip dislocation: an increasing and still uncontrolled disability? *Br Med J (Clin Res Ed)*. 1982;285(6354):1527–30.

Figures and Tables

Figure 1: Study population flow diagram summarising the case identification process, producing a total study population of 754 children with a new diagnosis of DDH between 1 and 8 years of age.

Table 1: Incidence of late diagnosed DDH in England by years of age, with specific population numbers and 95% confidence intervals listed per year group.

Table 2: The distribution of data validity for the total study population of 754 patients.

Figure 2: The annual age distribution at diagnosis. Presented by incidence per 1000 live births, with 95% confidence intervals.

Figure 3: The annual regional variation in late diagnosed DDH incidence per 1000 live births, with 95% confidence intervals.

Figure 4: The annual incidence of late diagnosed DDH per 1000 live births, listed by region (95% CI).

Figure 5: The variation in late diagnosed DDH by year of diagnosis. Presented by incidence per 1000 live births, with 95% confidence intervals.

Appendices A-D

Appendix A – CPRD GOLD codes

Included CPRD GOLD codes

| Medical code | Description |
|-------------------------|---|
| Diagnostic codes | |
| 423 | Dislocation or subluxation of hip |
| 933 | Congenital dislocation of hip |
| 2889 | Congenital hip dysplasia |
| 3691 | Dysplastic hip |
| 4804 | H/O: Cong. Dislocation – hip |
| 9073 | Dislocation of hip NOS |
| 9239 | Congenital dislocation and subluxation of the hip |
| 9299 | Pemberton osteotomy for congenital deformity of hip |
| 14938 | Congenital subluxation of hip |
| 19123 | Developmental dysplasia of the hip |
| 19216 | Wearing Pavlik harness |
| 21334 | Closed reduction of dislocation of hip |
| 24917 | Unstable hip |
| 27301 | Congenital dislocation of hip NOS |
| 30363 | Open reduction of congenital dislocation of hip |
| 32418 | Congenital acetabular dysplasia |
| 33576 | Congenital dislocation of hip NOS |
| 34019 | Unilateral congenital subluxation of hip |
| 34246 | Unilateral unstable hip |
| 34285 | Bilateral dysplastic hip |
| 38373 | Bilateral congenital dislocation of hip |
| 38772 | Unilateral dysplastic hip |
| 45970 | Unilateral congenital dislocation of hip |
| 50468 | Bilateral unstable hip |
| 55872 | Ferguson open reduction of congenital deformity of hip |
| 61870 | Bilateral congenital subluxation of hip |
| 62586 | Congenital subluxation of hip |
| 63645 | Unstable hip |
| 69116 | Ludloff open reduction of congenital deformity of hip |
| 96461 | Adams correction of congenital dislocation of hip |
| 97269 | Congenital instability of hip joint |
| 99310 | Congenital dislocation one hip with subluxation other hip |
| 101676 | Intraarticular soft tissue procedure correct congenital deformity hip |
| Supportive codes | |

| | |
|-------|---|
| 24670 | Correction of congenital deformity of hip |
| 7170 | Seen in orthopaedic clinic |
| 18783 | Seen in child orthopaedic clinic |
| 19702 | Splinting of dislocated hip |
| 1423 | Clicking hip |
| 8356 | Congenital clicking hip |
| 4460 | Hip stiff |
| 5570 | Clicking joint |
| 853 | Irritable hip |
| 6824 | Snapping hip |
| 6454 | Clicky hips congenital |
| 7806 | O/E hip joint abnormal |
| 8356 | Congenital clicking hip |

Excluded CPRD GOLD codes – excluded by CPRD

| Medical code | Description |
|---------------------|--|
| 1543 | Down's syndrome – trisomy 21 |
| 2069 | Congenital cerebral palsy |
| 4451 | Ehlers-Danlos syndrome |
| 5512 | Cerebral palsy with spastic diplegia |
| 5560 | Infantile cerebral palsy |
| 10759 | Down's syndrome NOS |
| 10956 | Prader - Willi syndrome |
| 12666 | Other infantile cerebral palsy NOS |
| 15530 | Congenital spastic cerebral palsy |
| 15846 | Klinefelter's syndrome |
| 16956 | Cerebral palsy, not congenital or infantile, acute |
| 16977 | Athetoid cerebral palsy |
| 18415 | Trisomy 21 |
| 21548 | Ataxic infantile cerebral palsy |
| 23320 | Arthrogryposis multiplex congenita. |
| 25306 | Angelman syndrome |
| 25570 | Spastic cerebral palsy |
| 27280 | Prader-Willi syndrome |
| 28306 | Congenital cerebral palsy NOS |
| 28335 | Ehlers-Danlos syndrome type III |
| 31418 | Arthrogryposis |
| 32010 | Trisomy 21, mosaicism |
| 34498 | Suspected Downs syndrome |
| 38479 | Arthrogryposis multiplex congenital |
| 39538 | Distal arthrogryposis syndrome |
| 41461 | Prader-Willi syndrome |
| 42701 | Trisomy 21, meiotic nondisjunction |
| 49967 | Dyskinetic cerebral palsy |
| 52659 | Ataxic diplegic cerebral palsy |
| 53178 | Other congenital cerebral palsy |
| 53755 | [X]Cerebral palsy and other paralytic syndromes |

| | |
|--------|---|
| 53984 | Ehlers-Danlos syndrome type VI |
| 54490 | Klinefelter's phenotype, karyotype |
| 56545 | Klinefelter's syndrome, XXXY |
| 59439 | Klinefelter's syndrome NOS |
| 60165 | Other arthrogryposis syndromes |
| 61499 | Trisomy 21, translocation |
| 61627 | Trisomy 21 NOS |
| 62414 | Klinefelter's syndrome, XXXXY |
| 63549 | Ehlers-Danlos syndrome type I |
| 64871 | Neuromuscular dislocation of the hip |
| 67854 | Klinefelter's syndrome, male with more than two X chromosomes |
| 68109 | Klinefelter's syndrome, XY/XXY mosaic |
| 70415 | Ehlers-Danlos syndrome type IV |
| 72787 | Ehlers-Danlos syndrome type II |
| 73943 | Athetoid cerebral palsy |
| 90520 | [X]Other infantile cerebral palsy |
| 91262 | Klinefelter's syndrome, XXYY |
| 95643 | Ehlers-Danlos syndrome type VII |
| 96257 | Klinefelter's syndrome, male with 46XX karyotype |
| 97059 | Angelman's syndrome |
| 98263 | Ehlers-Danlos syndrome type VIII |
| 98617 | Angelman syndrome |
| 101309 | Partial trisomy 21 in Down's syndrome |
| 104498 | Cerebral palsy |
| 104580 | Spastic quadriplegic cerebral palsy |
| 104654 | Cerebral palsy NOS August 2012 |
| 104775 | Spastic diplegic cerebral palsy |
| 104782 | Other cerebral palsy |
| 104828 | Flaccid infantile cerebral palsy |
| 105133 | Spastic hemiplegic cerebral palsy November 2012 |
| 107551 | Choreoathetoid cerebral palsy |
| 107844 | Gross Motor Function Classification System Cerebral Palsy |
| 107919 | Trisomy 21, mitotic nondisjunction |
| 108609 | Gross Motor Function Classification System CP level finding |

Excluded CPRD GOLD codes – further exclusions made

| Medical code | Description |
|---------------------|---|
| 4543 | Liveborn with birth asphyxia NOS |
| 8021 | Birth asphyxia |
| 8070 | H/O: birth asphyxia |
| 15082 | Severe birth asphyxia - apgar score less than 4 at 1 minute |
| 21694 | [D]Asphyxia |
| 24457 | Asphyxia by regurgitated food |
| 27409 | Asphyxiation or strangulation NOS |
| 35685 | Asphyxiation and strangulation |
| 37494 | Asphyxia by food |
| 41296 | Intrauterine hypoxia and birth asphyxia |

| | |
|-------|--|
| 43595 | Birth trauma, asphyxia and hypoxia |
| 46121 | Asphyxia due to foreign body in larynx |
| 46424 | Mild to moderate birth asphyxia - apgar score 4-7 at 1 min |
| 49662 | O/E - collapse – asphyxia |
| 49827 | Asphyxia by bone in food |
| 54994 | Blue asphyxia |
| 55879 | Birth trauma, asphyxia or hypoxia NOS |
| 56646 | Asphyxia by seed in food |
| 56681 | Self-asphyxiation |
| 62372 | Accidental mechanical asphyxia NOS |
| 65935 | Asphyxiating thoracic dysplasia |
| 66818 | Other specified birth trauma, asphyxia or hypoxia |
| 93787 | White asphyxia |
| 3584 | Hydrocephalus |
| 4675 | Acquired communicating hydrocephalus |
| 5306 | Spina bifida with hydrocephalus, unspecified |
| 5431 | Spina bifida without mention of hydrocephalus |
| 9611 | Congenital hydrocephalus |
| 10288 | Normal pressure hydrocephalus |
| 15388 | Acquired obstructive hydrocephalus |
| 28353 | Congenital hydrocephalus NOS |
| 37509 | Drainage of hydrocephalus of fetus to facilitate delivery |
| 42497 | Unspecified spina bifida with hydrocephalus |
| 45734 | Infantile posthaemorrhagic hydrocephalus |
| 46790 | Spina bifida with hydrocephalus |
| 47288 | Spina bifida with hydrocephalus - open NOS |
| 48609 | Spina bifida without hydrocephalus – closed |
| 50565 | Thoracic spina bifida with hydrocephalus |
| 50976 | Low pressure hydrocephalus |
| 52683 | Myelocoele with hydrocephalus |
| 53471 | Rachischisis with hydrocephalus |
| 53773 | [X]Other hydrocephalus |
| 57113 | Lumbar spina bifida with hydrocephalus |
| 57243 | Thoracic spina bifida with hydrocephalus – open |
| 60623 | Sacral spina bifida with hydrocephalus – open |
| 64717 | Spina bifida with hydrocephalus NOS |
| 65246 | Spina bifida without hydrocephalus - closed NOS |
| 65936 | Lumbar spina bifida without hydrocephalus – closed |
| 68221 | Cervical spina bifida without mention of hydrocephalus |
| 69370 | Unspecified spina bifida without hydrocephalus – closed |
| 69397 | Lumbar spina bifida without hydrocephalus – open |
| 70569 | Communicating hydrocephalus - acquired NOS |
| 70923 | Sacral spina bifida without hydrocephalus – closed |
| 71525 | Sacral spina bifida without hydrocephalus – open |
| 72018 | Cervical spina bifida without hydrocephalus – closed |
| 72928 | Sacral spina bifida with hydrocephalus – closed |
| 73085 | Other specified spina bifida with hydrocephalus |
| 73608 | Spina bifida without hydrocephalus - open NOS |

| | |
|--------|--|
| 90482 | Thoracolumbar spina bifida with hydrocephalus – closed |
| 93902 | Spina bifida with hydrocephalus – closed |
| 95018 | Unspecified spina bifida without hydrocephalus NOS |
| 95478 | Thoracic spina bifida without hydrocephalus – open |
| 96407 | Lumbar spina bifida without mention of hydrocephalus |
| 96709 | Thoracic spina bifida without mention of hydrocephalus |
| 97663 | Hydrocephalus with atresia of foramina of Magendie+Luschka |
| 98280 | Spina bifida without mention of hydrocephalus NOS |
| 98298 | Spina bifida with hydrocephalus NOS |
| 98811 | Spina bifida with hydrocephalus – open |
| 99281 | Spina bifida without hydrocephalus – open |
| 99894 | Other specified spina bifida without hydrocephalus |
| 100673 | [X]Other congenital hydrocephalus |
| 101013 | Spina bifida without hydrocephalus, site unspecified |
| 102628 | Spina bifida with hydrocephalus of late onset |
| 103284 | Lumbar spina bifida with hydrocephalus – open |
| 104943 | Fissured spine with hydrocephalus |
| 105767 | Lumbar spina bifida with hydrocephalus – closed |
| 106579 | [X]Unspecified spina bifida with hydrocephalus |
| 107207 | X-linked hydrocephalus |
| 107917 | Congenital hydrocephalus due to toxoplasmosis |
| 109243 | Cervical spina bifida with hydrocephalus |
| 111641 | Other specified congenital hydrocephalus |
| 16118 | Dystrophia myotonica (Steinert's disease) |
| 17990 | Myotonic disorders |
| 35137 | Other specified myotonic disorder |
| 44867 | Myotonic disorder NOS |
| 105696 | Myotonic chondrodysplasia |
| 1632 | Myopathy or muscular dystrophy NOS |
| 17392 | Proximal myopathy |
| 18307 | Mitochondrial myopathy, not elsewhere classified |
| 21022 | Myopathy due to Sjogren's disease |
| 25429 | Myotubular myopathy |
| 53868 | [X]Mitochondrial myopathy, not elsewhere classified |
| 63333 | Benign congenital myopathy |
| 93228 | [X]Paraneoplastic neuromyopathy and neuropathy |
| 103772 | Congenital myopathy |
| 108795 | [X]Myopathy in other diseases classified elsewhere |
| 5393 | Duchenne muscular dystrophy |
| 5964 | Muscular dystrophy |
| 7470 | Duchenne Aran muscular atrophy |
| 21425 | Hereditary progressive muscular dystrophy NOS |
| 22174 | Congenital hereditary muscular dystrophy NOS |
| 28210 | Other limb-girdle muscular dystrophy |
| 32749 | Becker muscular dystrophy |
| 34985 | Emery-Dreifuss muscular dystrophy |
| 48036 | Erb's muscular dystrophy |
| 64690 | Congenital hereditary muscular dystrophy |

| | |
|--------|---|
| 68118 | Hereditary progressive muscular dystrophy |
| 71128 | Other specified hereditary progressive muscular dystrophy |
| 91544 | Pelvic muscular dystrophy |
| 6599 | Muscular dystrophies and other myopathies |
| 32016 | Other myopathies and muscular dystrophies |
| 97444 | [X]Other specified myopathies |
| 110910 | [X]Other specified brain damage due to birth injury |
| 5118 | Anoxic brain damage |
| 37459 | Anoxic brain damage complication |
| 40868 | Birth brain damage NOS |
| 14917 | Chondrodysplasia NOS |
| 18193 | Chondrodysplasia punctate |
| 22756 | Chondrodysplasia |
| 33811 | Metaphyseal chondrodysplasia |
| 58035 | Chondrodysplasia calcificans congenital |
| 63146 | Chondrodysplasia calcificans congenital |
| 68427 | Osteochondrodysplasia |
| 73905 | Chondrodysplasia, unspecified |
| 3336 | Spina bifida occulta |
| 3947 | Spina bifida |
| 9298 | Repair of spina bifida |
| 21802 | Spina bifida NOS |
| 50672 | Other specified repair of spina bifida |
| 56362 | Spina bifida with stenosis of aqueduct of Sylvius |
| 59218 | Dandy - Walker syndrome with spina bifida |
| 62742 | Repair of spina bifida NOS |
| 63513 | Closed spina bifida with Arnold-Chiari malformation |
| 98265 | Insertion of Halber valve for spina bifida |
| 1749 | Hemiplegia |
| 2019 | Infantile hemiplegia NOS |
| 3293 | Right hemiplegia |
| 8492 | Hemiplegia NOS |
| 8933 | Left hemiplegia |
| 20122 | Spastic hemiplegia |
| 22135 | O/E – hemiplegia |
| 27966 | Congenital hemiplegia |
| 39085 | Flaccid hemiplegia |
| 3063 | Paraplegia |
| 3514 | Hereditary spastic paraplegia |
| 9375 | Spastic paraplegia |
| 36133 | O/E – paraplegia |
| 37160 | Congenital paraplegia |
| 46175 | Flaccid paraplegia |
| 58576 | Tropical spastic paraplegia |
| 59494 | Massive muscular calcification associated with paraplegia |
| 99040 | Paraplegia - congenital |
| 16033 | Monoplegia of lower limb |
| 19038 | Trisomy 13 NOS |

| | |
|--------|--|
| 33642 | Edward's syndrome - trisomy 18 |
| 35665 | Patau's syndrome - trisomy 13 |
| 37591 | Trisomy 7 |
| 37702 | Whole chromosome trisomy syndromes |
| 43565 | Trisomy 13, mosaicism |
| 45512 | Whole chromosome trisomy, mosaicism |
| 46133 | Trisomy 13, translocation |
| 46787 | TRISOMY 18 NOS |
| 65509 | Trisomy 9 |
| 67234 | Trisomy 18, mosaicism |
| 69476 | Trisomy 8 |
| 70198 | Trisomy 12 |
| 71815 | Major partial trisomy |
| 72139 | Trisomy 13, meiotic nondisjunction |
| 93133 | Trisomy 18, translocation |
| 99674 | Minor partial trisomy |
| 100024 | Trisomy 22 |
| 100174 | Partial trisomy syndrome NOS |
| 101732 | Other specified whole chromosome trisomy syndrome |
| 101982 | Whole chromosome trisomy syndrome NOS |
| 102102 | Partial trisomy 13 in Patau's syndrome |
| 103536 | 15q partial trisomy syndrome |
| 103873 | Trisomy 18, meiotic nondisjunction |
| 106114 | Trisomy 9p syndrome |
| 107029 | 10q partial trisomy syndrome |
| 107119 | Trisomy of autosomes NEC NOS |
| 107162 | Partial trisomy 18 in Edward's syndrome |
| 107670 | Whole chromosome trisomy, meiotic nondisjunction |
| 109223 | Trisomy 9 Mosaic Syndrome |
| 110833 | Trisomy 10 |
| 54377 | Trisomies of autosomes NEC |
| 3802 | Unspecified encephalopathy |
| 3979 | Hypertensive encephalopathy |
| 4501 | Wernicke's encephalopathy |
| 5644 | Anoxic - ischaemic encephalopathy |
| 11107 | Wernicke's encephalopathy |
| 37906 | Early infant epileptic encephalopathy wth suppression bursts |
| 41744 | Toxic encephalopathy |
| 45602 | Myoclonic encephalopathy |
| 57183 | Mitochond encephalopathy, lact acidosis & strokelike episode |
| 99684 | Cerebral degeneration due to multifocal leukoencephalopathy |
| 104239 | Hypoxic ischaemic encephalopathy of newborn |
| 106012 | Perinatal hypoxic - ischaemic encephalopathy |
| 4158 | Osteogenesis imperfecta |
| 53933 | Osteogenesis imperfecta NOS |
| 58635 | Osteogenesis imperfecta type I |
| 69436 | Osteogenesis imperfecta type III |
| 97751 | Osteogenesis imperfecta type IV |

| | |
|--------|---|
| 98662 | Osteogenesis imperfecta type II |
| 103017 | Osteogenesis imperfecta - unclassifiable |
| 69613 | Mucopolysaccharidosis type III |
| 34387 | Multiple epiphyseal dysplasia |
| 101702 | Multiple epiphyseal dysplasia NOS |
| 36694 | Intrauterine hypoxia |
| 41296 | Intrauterine hypoxia and birth asphyxia |
| 2006 | Dystonia, unspecified |
| 11881 | Idiopathic torsion dystonia |
| 25777 | Other specified symptomatic torsion dystonia |
| 27655 | Drug-induced dystonia |
| 27967 | Symptomatic torsion dystonia |
| 29022 | Torsion dystonias other involuntary movements drugs Band 1 |
| 50078 | Fragments of torsion dystonia |
| 51777 | Paroxysmal dystonia |
| 52917 | [X]Dystonia, unspecified |
| 62081 | Symptomatic torsion dystonia NOS |
| 62243 | Idiopathic familial dystonia |
| 65734 | [X]Other dystonia |
| 71249 | Fragments of torsion dystonia NOS |
| 94690 | Myoclonic dystonia |
| 54085 | Post-traumatic hydrocephalus, unspecified |
| 100957 | [X]Post-traumatic hydrocephalus, unspecified |
| 103462 | [X]Hydrocephalus in neoplastic disease classified elsewhere |
| 28262 | Inflammatory myopathy, not elsewhere classified |
| 111255 | [X]Inflammatory myopathy, not elsewhere classified |
| 32916 | Toxic myopathy |
| 36079 | Drug-induced myopathy |
| 39725 | Myopathy due to endocrine disease NOS |
| 44708 | Myopathy due to endocrine disease EC |
| 44925 | Myopathy in metabolic diseases |
| 48167 | Myopathy due to thyrotoxicosis |
| 49482 | Myopathy due to malignant disease |
| 51416 | Myopathy due to myxoedema |
| 52519 | Myopathy due to sarcoidosis |
| 55601 | Myopathy due to scleroderma |
| 57638 | Symptomatic inflammatory myopathy in disease NOS |
| 57888 | Myopathy due to polyarteritis nodosa |
| 60690 | Myopathy due to Cushing's syndrome |
| 62297 | Myopathy due to amyloid |
| 63541 | Symptomatic inflammatory myopathy in disease EC |
| 69198 | Myopathy due to Addison's disease |
| 108072 | Myopathy due to disseminated lupus erythematosus |
| 31742 | Alcoholic myopathy |
| 22638 | Brain damage – traumatic |
| 8846 | Suspect fetal spina bifida |
| 4921 | O/E – monoplegia |
| 29657 | [D]Transient monoplegia NOS |

| | |
|----------------------------------|--|
| 33925 | Congenital monoplegia |
| 45795 | Monoplegia unspecified |
| 19062 | Partial trisomy syndromes |
| 22411 | Encephalopathy – hepatic |
| 36748 | Alcoholic encephalopathy |
| 46157 | Influenza with encephalopathy |
| 47802 | Bilirubin encephalopathy |
| 49541 | Progressive multifocal leukoencephalopathy |
| 52673 | Progressive multifocal leukoencephalopathy |
| 68194 | Binswanger's encephalopathy |
| 97528 | Activity management for myalgic encephalopathy |
| 104407 | Referral for myalgic encephalopathy activity management |
| 36671 | Facioscapulohumeral muscular dystrophy |
| 66726 | Distal (Gower's) muscular dystrophy |
| 9179 | Spinal muscular atrophy |
| 57632 | Spinal muscular atrophy NOS |
| 66575 | Adult spinal muscular atrophy |
| 70572 | Unspecified spinal muscular atrophy |
| 95615 | Infantile spinal muscular atrophy |
| 101222 | Juvenile spinal muscular atrophy |
| Trauma codes searched for | |
| 4047 | Primary closed reduction of traumatic joint dislocation NOS |
| 14847 | Primary open reduction of traumatic dislocation of joint NOS |
| 34503 | Other dislocation or subluxation due to birth trauma |

Appendix B – HES codes

Included ICD-10 Codes

| ICD code | Description |
|-------------------------|--|
| Diagnostic codes | |
| Q65.0 | Congenital dislocation of hip, unilateral |
| Q65.1 | Congenital dislocation of hip, bilateral |
| Q65.2 | Congenital dislocation of hip, unspecified |
| Q65.3 | Congenital subluxation of hip, unilateral |
| Q65.4 | Congenital subluxation of hip, bilateral |
| Q65.5 | Congenital subluxation of hip, unspecified |
| Q65.6 | Unstable hip |
| Supportive codes | |
| M24.4 | Recurrent dislocation and subluxation of joint |
| R29.4 | Clicking hip |

Excluded ICD-10 codes

| ICD code | Description |
|-----------------|-------------------------------------|
| G80 | Cerebral palsy |
| G80.0 | Spastic quadriplegic cerebral palsy |
| G80.1 | Spastic diplegic cerebral palsy |
| G80.2 | Spastic hemiplegic cerebral palsy |

| | |
|-------|---|
| G80.3 | Dyskinetic cerebral palsy |
| G80.4 | Ataxic cerebral palsy |
| G80.8 | Other cerebral palsy |
| G80.9 | Cerebral palsy, unspecified |
| Q74.3 | Arthrogryposis multiplex congenita |
| Q79.6 | Ehlers-Danlos syndrome |
| Q90 | Down syndrome |
| Q90.0 | Trisomy 21, meiotic nondisjunction |
| Q90.1 | Trisomy 21, mosaicism (mitotic nondisjunction) |
| Q90.2 | Trisomy 21, translocation |
| Q90.9 | Down syndrome, unspecified |
| Q98.0 | Klinefelter syndrome karyotype 47,XXY |
| Q98.1 | Klinefelter syndrome, male with more than two X chromosomes |
| Q98.2 | Klinefelter syndrome, male with 46,XX karyotype |
| Q98.3 | Other male with 46,XX karyotype |
| Q98.4 | Klinefelter syndrome, unspecified |
| Q98.5 | Karyotype 47,XX |
| Q82.4 | Ectodermal dysplasia (anhidrotic) |
| Q71.8 | Other reduction defects of upper limbs |
| Q72.8 | Other reduction defects of lower limb(s) |
| Q99.9 | Chromosomal abnormality, unspecified |
| Q91.6 | Trisomy 13, translocation |
| G71.1 | Myotonic disorders |
| G81.9 | Hemiplegia, unspecified |
| Q92.9 | Trisomy and partial trisomy of autosomes, unspecified |
| G82.4 | Spastic tetraplegia |
| G93.1 | Anoxic brain damage, not elsewhere classified |
| Q77.3 | Chondrodysplasia punctata |

Included OPCS codes

| OPCS code | Description |
|------------------|--|
| X22.1 | Open reduction of congenital deformity of hip |
| X22.2 | Primary osteotomy of pelvis for correction of congenital deformity of hip |
| X22.3 | Secondary arthroplasty of hip for correction of congenital deformity of hip |
| X22.4 | Intra-articular soft tissue procedures for correction of congenital deformity of hip |
| X22.5 | Extra-articular procedures for correction of congenital deformity of hip |
| X22.8 | Other specified correction of congenital deformity of hip |
| X22.9 | Unspecified correction of congenital deformity of hip |

Excluded OPCS codes

One patient was excluded for having evidence of a traumatic hip dislocation without any additional support diagnosing a congenital hip dysplasia.

| |
|----------------------------------|
| Trauma codes searched for |
|----------------------------------|

| | |
|--------------|---|
| W65.1 | Primary open reduction of fracture dislocation of joint and skeletal traction HFQ |
| W65.2 | Primary open reduction of traumatic dislocation of joint and skeletal traction NEC |
| W65.3 | Primary open reduction of fracture dislocation of joint NEC |
| W65.4 | Primary open reduction of fracture dislocation of joint and internal fixation NEC |
| W65.5 | Primary open reduction of fracture dislocation of joint and combined internal and external fixation |
| W65.8 | Other specified primary open reduction of traumatic dislocation of joint |
| W65.9 | Unspecified primary open reduction of traumatic dislocation of joint |
| W66.1 | Primary closed reduction of fracture dislocation of joint and skeletal traction HFQ |
| W66.2 | Primary closed reduction of traumatic dislocation of joint and skeletal traction NEC |
| W66.3 | Primary manipulative closed reduction of fracture dislocation of joint NEC |
| W66.4 | Primary closed reduction of fracture dislocation of joint and internal fixation |
| W66.8 | Other specified primary closed reduction of traumatic dislocation of joint |
| W66.9 | Unspecified primary closed reduction of traumatic dislocation of joint |
| W67.1 | Secondary open reduction of fracture dislocation of joint and skeletal traction HFQ |
| W67.2 | Secondary open reduction of traumatic dislocation of joint and skeletal traction NEC |
| W67.3 | Secondary open reduction of fracture dislocation of joint NEC |
| W67.4 | Secondary open reduction of traumatic dislocation of joint NEC |
| W67.5 | Remanipulation of fracture dislocation of joint |
| W67.6 | Remanipulation of traumatic dislocation of joint |
| W67.7 | Secondary open reduction of fracture dislocation of joint and internal fixation NEC |
| W67.8 | Other specified secondary reduction of traumatic dislocation of joint |
| W67.9 | Unspecified secondary reduction of traumatic dislocation of joint |

Appendix C – Validation algorithm for codes between the CPRD and HES datasets

- 1) (Diagnostic DDH code within CPRD) and (diagnostic ICD DDH code within HES or specific procedural code pertaining to an OPCS code within HES)
- 2) (Diagnostic code within CPRD) and (a similar code or codes which may be erroneously used in the ICD HES dataset (‘*recurrent dislocation and subluxation of joint*’, ‘*clicking hip*’)).
- 3) (Diagnostic code within CPRD) and (evidence in the HES dataset of a hospital admission within six months of the index date).
- 4) (Diagnostic code within CPRD), without any related codes in the HES dataset, but with either: (a) evidence of diagnostic code followed by regular (>3 visits) orthopaedic follow-up hospital attendances, (b) multiple entries (2 or more) of a diagnostic code representing DDH, (c) Other supportive evidence within CPRD GOLD as detailed in *Appendix A*.

- 5) Diagnostic code within CPRD, without additional supportive evidence (as defined above) to ratify diagnosis.

Appendix D - CPRD ‘acceptable’

The following information is copied directly from the CPRD data specification.

Patients are labelled as ‘acceptable’ for use in research by a process that identifies and excludes patients with non-continuous follow up or patients with poor data recording that raises suspicion as to the validity of the that patients record. Patient data is checked, for the following issues:

- *An empty or invalid first registration date*
- *An empty or invalid current registration date*
- *Absence of a record for a year of birth*
- *A first registration date prior to their birth year*
- *A current registration date prior to their birth year*
- *A transferred out reason with no transferred out date*
- *A transferred out date with no transferred out reason*
- *A transferred out date prior to their first registration date*
- *A transferred out date prior to their current registration date*
- *A current registration date prior to their first registration date*
- *A gender other than Female/Male/Indeterminate*
- *An age of greater than 115 at end of follow up*
- *Recorded health care episodes in years prior to birth year*
- *All recorded health care episodes have empty or invalid event dates*
- *Registration status of temporary patients*

If any of these conditions are true then the patient is labelled unacceptable, and is not recommended for use in research.

‘UTS’ refers to practices defined by CPRD to have continuous high quality data sufficient to be used for research.