CRANIOFACIAL SOCIETY ANNUAL MEETING, BIRMINGHAM APRIL 2013

Abstracts and Posters

Thursday Abstracts / **Friday Abstracts** / **Posters**

Friday April 19th
10.30-11.50
Arnold Huddart Medal

Radical Muscle Dissection In Cleft Palate Repair – What About The Lesser Palatine Nerves?

J. Mortimer, F.V. Mehendale & S. Parson

Aim: The lesser palatine nerves (LPNs) are overlooked in radical muscle dissection techniques for cleft palate repair. As well as supplying sensation, taste and glandular secretion to the velum, reports suggest they carry facial nerve motor fibres to levator veli palatini (LVP). This study explores LPN anatomy with reference to a Sommerlad dissection.

Methods: LPNs and LVP were dissected under magnification in 3 cadavers. Sihler nerve staining was carried out for the first time in the velopharyngeal region. A literature review was also undertaken to identify trends in the reported inconsistencies of LVP innervation.

Results: LPN branches ran oral to the tensor aponeurosis, terminating within the glandular layer oral to the muscular velum. No LPN fibres entered LVP. The main LVP nerve emerged from the lateral pharyngeal wall. Evidence refuting LPN facial nerve motor contribution to LVP seems only found in non-primate studies.

Conclusions: Radical muscle dissection severs LPNs, which carry fibres of sensation, taste and glandular secretion to the velum. Primates may have evolved facial nerve motor relays between the facial speech muscles and LVP via LPNs that would also be lost. Post-cleft palate repair outcome studies of non-motor velar function should be undertaken, possibly informing modifications in technique and timing of surgery.

Prospective subjective evaluation of cleft rhinoplasty using a validated questionnaire

A.R. Sawyer & M. Cadier

Aim
We evaluated patient subjective satisfaction and quality of life following cleft rhinoplasty.

Methods
Subjective assessment was performed using a validated rhinoplasty outcomes evaluation questionnaire (Alsarraf 2000). This instrument is composed of six

questions that capture three quality-of-life domains: physical, mental/emotional, and social. 22 patients completed prospective patient evaluation forms preoperatively and 3-6 months post operatively. In addition the forms were completed by patients having non-cleft rhinoplasty and by subjects who were not considering rhinoplasty.

Results
Average age was 28 (range 18-59) years. There was a significant subjective improvement in the total evaluation scores (from 7 +/- 3 to 19 +/3; $p<0.01$), and in specific scores for nasal aesthetical appearance (from 0.35 +/-0.6 to 3.2+/-0.3; $p<0.01$). No significant change was seen in breathing capacity (from 2.7+/-0.3 to 3.2+/-0.2; $p=0.29$). Specifically patients complained of the cosmetic appearance of their nose rather than the functional problems. All patients said they would undergo the procedure again.

Conclusion
Our results demonstrate a high patient satisfaction following cleft rhinoplasty, particularly with regard for the cosmetic appearance, quality of life and is similar to those for non-cleft rhinoplasty. We would recommend the use of this simple and quick validated outcome tool in all rhinoplasty patients.

When Is Hearing Loss Truly Hearing Loss? The Effect of Different Published Audiological Guidelines on the 'Epidemiology' of Hearing Loss in Cleft Children
C. Russell, A. Irvine, C. Napier, H. Sadiq, A. Ray, M. Devlin & D. Wynne

Little information exists on prevalence of hearing impairment in cleft populations. Furthermore lack of consistency in thresholds describing hearing impairs scientific discussion. This study describes 'epidemiological ' differences in hearing impairment in cleft patients dependant on diagnostic thresholds used.

335 patients underwent audiological assessment between 2006-2011. Data were analysed for differences in prevalence and severity of hearing impairment according to published thresholds (American Speech Language and Hearing Association, British Society of Audiologist, World Health Organisation, Clinical Standards Advisory Group Cleft and GG Browning Thresholds)

The prevalence ($p<0.001$) and severity ($p<0.001$) of hearing impairment in the cohort examined varies significantly dependent upon published thresholds used. Prevalence by sex also varies significantly for all thresholds apart from GG Brownings ($p<0.02$). Full age and sex specific data will be presented for all thresholds.

Hearing impairment in cleft is common no matter the threshold employed. This study highlights the need for threshold concordance to facilitate scientific discourse. For UK practice it is important to highlight differences in 'prevalence'/'severity' of hearing loss described by BSA and CSAG thresholds. When hearing is so integral to speech outcomes it is concerning that CSAG interpretation of hearing loss identifies significantly fewer children as having hearing impairment.($p<0.001$).

A Pan-Regional Operative Proforma for Cleft Palate Repair
K. Moar, V. Beale, F.V. Mehendale

Aim: To design an Operative and Review Proforma for Cleft Palate Repair to facilitate audit with consistent and comparable data collection within a Pan-Regional Cleft Audit Group.

NORCLEFT is the quad-centre audit group for the Cleft Services in Northern England and Scotland. Following a retrospective review of fistula rate for primary palate repair, the Surgical Special Interest Group (SIG) proposed a prospective audit.

Method: A proforma was designed for each audit point to aid consistent data collection:

- Palate repair
- 1 week review Cleft Nurse Specialist
- 6 week review Surgeon (MDT clinic)
- 3 year review Surgeon

Feedback was obtained by email, phone and direct conversations with members of the NORCLEFT Surgical and Nursing SIGs until a consensus opinion was reached. The proforma was trialled and further modifications made.

Results: The draft operative proforma for palate repair is based on a modified Sommerlad protocol. The review proforma collects a variety of data but mainly relates to fistula occurrence, incorporating the Pittsburgh Classification1 of fistula position.

Conclusion: The NORCLEFT audit network facilitates collaborative working such that, despite significant local variations in methods used for operative and review data collection, it is possible to achieve consensus in prospective data collection.

Reference

- Smith DM, Vecchione L, Jiang S et al. The Pittsburgh Fistula Classification System: A Standardized Scheme for the Description of Palatal Fistulas. *CPCJ*2007 44(6) 590-4

Does the age of the patient or the surgical procedure chosen affect secondary speech surgery outcome? – a retrospective cohort study over the past decade
C. Wallner, J. Smith, A. Harding- Bell, T. Ahmad & P.N. Hall

Aims: To investigate whether cleft type, surgical procedure or patient age influenced the need for additional speech surgery.
Methods: Patients undergoing surgery for speech (Cleft and Non-Cleft VPD) between 2000-2011 were identified from the Cleft.Net.East database. 323 correctional speech procedures were performed on 262 patients.
Results: 45/262 patients (17%) needed more than one surgical procedure for speech. The initial diagnosis (all cleft types and non-cleft VPD), was not a factor in

those needing additional procedures.
Adults (>16 years) were 1.5 times more likely to need further procedures than those under 16 (p=0.33). For the whole cohort (including non-cleft VPD), those whose first operation was a buccal flap were approximately 5 times less likely to undergo further procedures (odds ratio 5.34 (p=0.02; 95% CI: 1.25-22.96). This result remained unchanged when non-cleft VPD cases were excluded from the analysis (odds ratio 4.86 (p=0.04; 95% CI: 1.1-21.6).
Final speech outcome based on clinical GOSSPASS records and surgical history were analysed for 48 patients with cleft palate only. In the 'buccal flap 1st' group 61.5% of VPD was eliminated, compared with 37.5% treated with other procedures. Factors affecting these results will be reviewed.
Conclusion: In our centre buccal flaps result in better functional outcome (p=0.07).

Novel genes downstream of *FGFR2* implicated in the pathogenesis of coronal suture synostosis in a Crouzon mouse model
S. Kumar, E. Peskett, J.A. Britto & E. Pauws

Introduction: The FGFR2Cys342Tyr mutation causes heterozygote murine coronal synostosis, analogous to human Crouzon syndrome, whilst the homozygote displays cleft palate. Morphological differences between wild-type (Wt) and *Fgfr2C342Y/+* (Mut) calvaria were found after embryonic day E17.5 suggesting enrichment for genes responsible for the coronal suture fusion.

Methods: Coronal sutures were micro-dissected from E17.5 Wt/Mut mouse calvaria and gene expression profiles were examined by microarray analysis, qPCR, and *in situ* hybridisation (E15.5 to E18.5). RNAi knockdown of *Fgfr2c* was performed in the murine MC3T3 osteoblast and effects on these genes were analysed.

Results: 51 genes were differentially expressed by at least 1.5-fold ($p<0.05$); some of which are osteogenic regulators. *Dpt*, *Osr1*, *Nov*, *Dlk1* and *Kera* are downregulated up to 3.5-fold in the Mut fusing suture, implying functional significance. Complementary spatio-temporal expression is observed in dura and periosteum. *In vitro* studies in MC3T3 cells identified transcriptional changes in a subset of these genes after *Fgfr2c* knockdown($p<0.05$).

Conclusions: Novel genes, implicated in the pathogenesis of murine coronal synostosis, are identified and related to FGFR signaling. These molecular pathways may identify key targets for genetic diagnosis and pharmacotherapy in syndromic craniosynostosis. Amplification of these pathways in the homozygote investigates molecular mechanisms in FGFR-related cleft palatogenesis.

Should We Give Routine Post-Operative Intravenous Fluids After Cleft Surgery?
O. Onyekwelu, J. Seaward & V. Beale

Aim: In 2012, the James Lind Alliance, together with CFSGB&I and CLAPA, set priorities for unanswered questions in cleft management. One of these priorities included post-operative fluid management.

The authors' post-operative regimen does not include intravenous fluids, unless the child fails to achieve adequate oral intake by the first evening post-operatively. This study evaluated whether this is appropriate and safe practice.

Methods: All patients undergoing cleft-related surgery by a single surgeon in a single centre during August 2011-12 were included. Patient age, weight and surgery type were recorded together with fluid requirement, length of stay and any returns to theatre or readmissions.

Results: Of the 79 patients included, none required readmission or return to theatre, and the mean length of stay was 1.9 days. 19 (24%) patients required intravenous fluids but these tended to be the older children in the group (p-value 0.034).
In the youngest patients undergoing primary lip repair, only 1of 20 required intravenous fluids.

Conclusions: This study demonstrates that, especially in the younger patients, omitting intravenous fluids as a post-operative routine is associated with a shorter length of stay without an increased complication rate. The authors advocate early post-operative feeding and the return to physiological fluid balance.

Cleft oro-nasal fistulae: A cause of dental decay?
H. Richards, A.V. Bommel, V. Clark & B. Richard

Introduction: Oro-nasal fistula is an acknowledged complication of cleft palate repair. We investigate if there is a correlation between fistula and dental decay.

Methods : A retrospective review of a single surgeon's palate repairs over nine years, from 2003 – 2011. Data collected: age, sex, LAHSAL, fistula, date of repair and recorded number of decayed, missing or filled primary teeth (dmft) at five years old.

Results: Of 261 palate repairs in the first year of life, 24.1% had a fistula. At age five, children with a fistula had dmft scores ranging from 0-13 with a median of 1, those without ranged from 0-20 and had a median of 0. Dental decay was found in 40% of children with fistulae and 20% of those without. Chi squared test showed this to be significant, p value = 0.036.
It was shown that a decay score (d) of greater than 2 correlated to a higher fistula rate. (Odds ratio 5.68, p = 0.005).

Conclusion : The presence of an oro-nasal fistula correlates with higher dmft scores.
We propose food clearance in children with fistulae is prolonged and that nasal mucus increases retention of sugary food in the mouth, causing higher rates of dental decay.

Friday 18th April 2013
13.35 – 14.10 Free papers

Illuminating Or Alarming; Early Home Oximetry Screening For Sleep Breathing Disorders in Cleft Patients?
C. Russell, J. Pettigrew, N. Gibson, A. Crawford, S. Wallace, M. Delvin , A. Ray & D. Wynne

RCPCH guidelines suggest early sleep investigation (by 4 weeks) in PRS/Stickler patients. Previously reported pre-operative sleep investigations demonstrate significant overlap between PRS/non-syndromic CP+/-L patients. This study aims to investigate whether early sleep investigation adds to general cleft patient management.

Between January and December 2012 a consecutive series of 42 patients were offered home oximetry screening for Sleep Breathing Disorder (SBD). After exclusions (declined/in-patient studies, NICU admissions, microform cleft deformity) 28 patient studies were available for review (early study cohort (<6wks,n=14)/late study cohort (>6 wks, n=14)).

Early study results are consistent with severe SBD in most patients (93%) (desaturation index = 24(11.6-56.6) dips/hr (median(range)). In contrast, the late study cohort demonstrated significantly lower incidence of severe SDB (20%) (desaturation index = 11.6(6.7-17.6) dips/hr), (Chi squared analysis $p<0.001$). Initial and follow-up studies demonstrate improvement but not normalisation of cleft patient SBD during infancy.

Significant difference between early/late home oximetry cohorts, and temporal improvements in early cohort data are consistent with previous normative data demonstrating transient hypoxias as part of normal infant development. A pragmatic algorithm for the investigation/management of sleep breathing disorders in cleft children will be presented that avoids unnecessary resource burden as well as minimising clinician and parental anxiety!

Airway and Feeding Management of babies with Pierre Robin Sequence in Great Britain and Ireland
C. Theopold & G. Thorburn

Aim: To determine variations in regional management for babies presenting with Pierre Robin Sequence.

Methodology: A questionnaire distributed amongst all cleft units in Great Britain and Ireland.
Results: 12 completed questionnaires were returned. In 75% of centres, referrals are made to the cleft team and initially assessed by cleft CNSs. 70% of units do not classify PRS, and no algorithm for admission exists in 88% of units. Routine and downloadable O2 saturation monitoring are mainstream investigations, with polysomnography and blood gases playing less of a role than anticipated.Only 1 unit routinely uses prone positioning, with side-to-side positioning predominating. NPAs are routinely used (88% also at home). Lip adhesion, tracheostomy and mandibular distraction are widely reported in literature but rarely used in UK.
Feeding assessments are mainly led by Cleft CNSs and 75% of units follow formal

protocols. All units attempt feeding with NPAs in situ but also try positional changes, modified teats and different milks as indicated.

Discussion: This study provides an overview of the diversity in management amongst cleft units for babies with PRS, mainly related to respiratory investigations and management. Adoption of a national standardised classification and care pathway could allow better comparison of treatment options and outcomes.

Airway Management and Early Oral Feeding in Babies with Pierre Robin Sequence
A. Wilkinson, L. Paton, F.V. Mehendale, D. Urquhart, S. Cunningham & O. Duncan

Aim/Background: Many patients with Pierre Robin Sequence(PRS) receive prolonged naso-gastric feeding(NGF). A systematic review of feeding outcomes found limited data on oral feeding in PRS. Clinical experience suggested that early airway optimisation and use of the Haberman bottle improved feeding in PRS. This study aims to 1) audit effectiveness of multidisciplinary team(MDT) management in establishing full oral feeding 2) identify predictive factors

Methods: Prospective data collection on 52 consecutive babies referred with isolated cleft palates from January 2007-December 2009
(Lothian,Fife,Borders,Highlands)
Exclusions-2 babies with unsafe swallows
24/50 were diagnosed with PRS following MDT assessment, including sleep studies
Airway management –positioning alone(lateral/prone) 12/24, Nasopharyngeal Airway(NPA) 12/24
PRS severity was graded clinically into borderline(12), moderate(6) and severe(6).
The Haberman feeder was used in all moderate/severe PRS.

Results: All moderate/severe PRS required NPAs
Mean time to total oral feeding-
Severe PRS- 23 days(5-126).
Moderate PRS- 4 days(0-11)

Conclusions: These findings are in contrast to studies that report long-term NGF in PRS.
This study is limited by its lack of a control group, but historical comparisons show a decrease in the time to establishment of full oral feeding. Variables that have changed include increased specialist respiratory input, sleep studies, early NPA use and use of the Haberman feeder.

Regional experience with management of infants with Pierre Robin Sequence using common guidelines and home-based monitoring
S. Kansra, B. Davies, J. Neil-Dwyer, K. Latter & D. Thomas

Introduction: Pierre Robin Sequence (PRS) is a congenital condition which predisposes to problems including airway obstruction and feeding difficulties. Nasopharyngeal airway (NPA) insertion is effective for managing airway

obstruction and historically patients have required prolonged and/or multiple hospital admissions.

Aims: We present our network management of PRS using a common guideline. This incorporates training for carers and professionals enabling NPA management at home with feeding support and respiratory monitoring including home oximetry.

Methods: Retrospective review of medical records between 2010 and 2012.

Results: 23 children with PRS were managed with NPA between 2010-2012. Mean duration of hospitalisation was 56 days (median 31, range 11-195). 6 children still require NPA. The remaining 17 children stopped daytime NPA use after a mean 128 days; night use was stopped after 148 days. One child required a tracheostomy. For two children non-invasive ventilation at home was necessitated post NPA.

Conclusion: This data is comparable to a recently published case series. Remarkably we demonstrate similar success in managing PRS at home. Adopting a common guideline across our region has allowed management of children with PRS and NPA at local hospital units and facilitated early discharge. Specialist nurse and carer involvement has minimised hospitalisations.

Friday 18th April 2013
15.00 – 15.40 Free papers

Parent Experiences of Robin Sequence: A multi-centre perspective
K. Latter

Aims: In the absence of literature surrounding parental experiences of the management of infants with PRS the focus of the study was to explore experiences, views and feelings of the care and support that parents have received. This is to contribute to identifying gaps in care provision for future planning and implementation of care based on expressed needs and perspectives and provides a basis on which to inform practice for holistic management.

Methods: The study was undertaken utilizing a hermeneutic phenomenological approach. Twenty-five participants were recruited from five UK cleft centres and open ended interviews were conducted within the home. Thematic content analysis allowed the interpretation of commonalities linking together the various experiences within the phenomena under investigation.

Results: The findings of the study revealed the three key themes of impact, coping and communication. The parent accounts highlighted differences between care provision, delivery and management of the condition between the five centres and how these impacted on the families involved. The key messages were for better

communication and information provision. The findings of the study bridge the gap in the knowledge of how PRS impacts on families and provides valuable insights for cleft teams in meeting the needs of this group.

Laryngoscopy In Dysphonic Cleft Children: An Intervention Both Possible And Necessary
N. McNeil, C. Russell, L. Crampin, L. Campbell, W. Cohen, M. Delvin , A. Ray & D. Wynne

Speech disorders in CP/CLP are well researched. Voice disorders receive less attention. Recent studies suggest they are a significant issue in CP/CLP. However, little information exists on associated laryngeal pathology. This study aims to investigate/describe laryngeal pathology associated with dysphonia in CLP children.

Retrospective case-note review of all cleft patients attending specialist paediatric voice clinic was performed. Patient demographics, cleft diagnosis/interventions, video-fluoroscopy and audiology results, socio-economic status and birth order were collated. Data were analysed in relation to laryngoscopic findings.

Seven CP/CLP patients (4.5-11.5 yrs, median 5.5) were reviewed (10% of referrals) between January 2010 and May 2012. Three females (100%) and two males (50%) had vocal cord nodules. One male(14%) had adenoidal pad hypertrophy. Six (86%) patients had clinical velopharyngeal insufficiency. Although hearing had been mutable from birth, only 2 (patients 29%) had a current hearing issue. Interestingly, 4 (57%) patients are from the lowest 10% of socioeconomic scale, all patients had at least one older sibling and the majority (n=5) were CP patients.

Clinical dysphonia in cleft children is associated with significant rates of vocal cord pathology. Long-term significance of such findings remains undetermined. This initial study provides insight into potential causal associations that warrant further discussion and investigation.

Living with OME in Cleft palate patients: Experiences of children and parents - a qualitative study
S. Tierney, K. O'Brien, N. Harman, R.K. Sharma, C. Madden, P. Callery on behalf of the mOMEnt Study Management Group

Background: Approximately 90% of children with Cleft Palate (CP) develop otitis media with effusion (OME) due to impaired Eustachian tube function. The impact this has on everyday life has not been examined in-depth previously.

Aim: To explore experiences of OME from the perspective of children and parents in terms of social development, school and family life.

Design: Qualitative research.
Sample: 37 families with a child aged 0-11 years with CP and history of OME.
Data collection: Semi-structured interviews with mothers and/or fathers and children

aged 6-11 years.
Analysis: Framework analysis was applied to interview transcripts.

Results: Engaging in the world was a key theme to emerge from data. Parents were concerned about the child's emotional functioning (e.g. becoming withdrawn or frustrated in social settings) and educational performance. Children expressed difficulties associated with activities (swimming, cinema) due to hearing loss. Some relied on siblings to help them communicate with others.

Conclusion: What parents could perceive initially as a secondary complaint connected to CP appeared to have significant psychosocial repercussions for some interviewees. Discussion of OME, its course and likely treatments with parents might help them assist children to play a full role in their social environment.

An overview of the profiles and psychological issues of over 2000 patients with cleft lip and/or palate and their families seen by the South West Cleft service
J. Cadogan, G. Smith, T. Owen & R. Winter

Background : The main objective is to present a summary of the profiles of over 2000 patients extracted from a secure database administered by the psychology service, with a particular emphasis on psychological issues across the lifespan.

Results: Specifically, a total of 325 (16%) of patients were identified as being affected by 54 different syndromes. Pierre Robin sequence and 22q11 deletion syndrome accounted for 106 (5%) and 92 (4%) of the total cohort respectively. Approximately 25% of the cohort were seen by the psychology service for additional support. The most prevalent psychological concerns were incidences of teasing, social anxiety and feeling self-conscious. Family relationships were the greatest concern for parents of children aged 0-6months and speech was found to be the greatest concern for families where the children were between 6 months and 4 years. Learning difficulties were the most prevalent concern for patients with submucous cleft palate. The content of 1390 consultations with members of the psychology team involving 470 families was explored in order to establish the incidence of concerns across the lifespan and in relation to cleft type.

Conclusion: In the current financial climate it is important to be able to identify when and why interventions such as psychological support are most likely to be required in order to ensure that outcomes are optimal.