This monthly Current Awareness Bulletin is produced by the Healthcare Library to provide Salisbury NHS Foundation Trust staff with a range of resources to support practice. It includes recently published guidelines and research articles, news and policy items, and details of new library resources.

OpenAthens
To access journal articles that are available in full text you will need to have a username and password for OpenAthens. To register for an OpenAthens account click here.

For further information or support Salisbury staff can contact the Healthcare Library, SDH Central, Salisbury District Hospital, Salisbury, Wiltshire SP2 8BJ. 01722 429054 or 01722 336262 ext 4430, Library.office@salisbury.nhs.uk, or visit the library website at www.library.salisbury.nhs.uk

Cochrane Systematic Reviews
Issue 2, February 2015
New Reviews

As required versus fixed schedule analgesic administration for postoperative pain in children

Cleft Palate-Craniofacial Journal – Latest Issue
Cleft Palate-Craniofacial Journal
ISSN: 1055-6656 Latest issue available from Allen Press in Journals@Ovid (Athens Authorization)

Journal Articles
Please click on the blue links (where available) to access full text. You may need an OpenAthens username and password. To register for an OpenAthens account click here. If you have any difficulty accessing the full text articles, or if you would like us to obtain any of the articles for you, please contact the Healthcare Library.

Titles highlighted in green may be of particular interest to Speech and Language Therapists
Titles highlighted in orange may be of particular interest to Clinical Psychologists

Table of Contents
1. A Clinical and Molecular Analysis of Branchio-Oculo-Facial Syndrome Patients in Russia Revealed New Mutations in TFAP2A.
3. Assessment of morbidity following insertion of fixed preoperative orthopedic appliance in infants with complete cleft lip and palate.
4. Association between maternal MTHFR polymorphisms and nonsyndromic cleft lip with or without cleft palate in offspring, a meta-analysis based on 15 case-control studies.
5. Association of single-nucleotide polymorphisms in the IRF6 gene with non-syndromic cleft lip with or without cleft palate in the Xinjiang Uyghur population.
6. Atypical expanded-spectrum hemifacial microsomia: a case report
7. Baraitser-Winter cerebrofrontofacial syndrome: delineation of the spectrum in 42 cases
9. Comparative evaluation of salivary electrolytes in cleft lip and palate and healthy children
10. Comparative validity and reproducibility study of various landmark-oriented reference planes in 3-dimensional computed tomographic analysis for patients receiving orthognathic surgery
12. Converting a nasoendotracheal tube to orotracheal, following fibreoptic intubation under general anaesthesia in a paediatric patient with complete cleft palate
13. Craniofacial malformations and the orthodontist.
15. Distraction osteogenesis and orthognathic surgery for a patient with unilateral cleft lip and palate.
16. Double-loop technique using titanium molybdenum alloy wire for fabrication of nasal stents in nasoalveolar molding therapy for cleft lip and palate patients
17. Early and late operation of cleft lip and intelligence quotient and psychosocial development in 3-7 years.
18. Ectrodactyly ectodermal dysplasia cleft lip/palate (EEC) syndrome in a family
19. Expanding the genetic and phenotypic spectrum of popliteal pterygium disorders.
20. Extent of palatal lengthening after cleft palate repair as a contributing factor to the speech outcome
21. Fine Mapping of 6q23.1 Identifies TULP4 as Contributing to Clefts.
24. Hypohidrotic ectodermal dysplasia with ankylosis of temporomandibular joint and cleft palate: A rare presentation.
25. Impact of primary palatoplasty on the maxillomandibular sagittal relationship in patients with unilateral cleft lip and palate: A systematic review and meta-analysis
26. Long term speech outcomes following late cleft palate repair using the modified Furlow technique.
27. Longitudinal study of growth of children with unilateral cleft lip and palate: 2 to 10 years of age.
28. Measurement of health-related and oral health-related quality of life among individuals with nonsyndromic orofacial clefts: a systematic review and meta-analysis
30. Modification of the dingman mouth gag for better visibility and access in the management of cleft palate.
33. Older adults' experiences of living with cleft lip and palate: a qualitative study exploring aging and appearance.
34. Optimization of dental status improves long-term outcome after alveolar bone grafting in unilateral cleft lip and palate.
35. Preliminary evidence of an interaction between the CRISPLD2 gene and non-syndromic cleft lip with or without cleft palate (nsCL/P) in Xinjiang Uyghur population, China
36. Relationship between reduced folate carrier gene polymorphism and non-syndromic cleft lip and palate in Indian population
37. Risk factors involved in orofacial cleft predisposition-review
38. Risks of congenital malformations in offspring exposed to valproic acid in utero: A systematic review and cumulative meta-analysis
39. SLC26A2 disease spectrum in Sweden - high frequency of recessive multiple epiphyseal dysplasia (rMED)
40. The Association between Folate Pathway Genes and Cleft Lip With or Without Cleft Palate in a Chinese Population.
41. The burden of selected congenital anomalies amenable to surgery in low and middle-income regions: cleft lip and palate, congenital heart anomalies and neural tube defects.
42. The double unilimb z-plasty technique for whistler deformity repair in unilateral cleft lip patients: an anthropometric study.
43. The role of the folic acid to the prevention of orofacial cleft: an epidemiological study
44. Three-dimensional evaluation of nasopharyngeal airways of unilateral cleft lip and palate patients
45. Unusual foreign body in the nasal cavity of an adult with repaired cleft lip and palate.
46. Upper triangular flap method for primary repairs of incomplete unilateral cleft lip patients: minor to two-thirds way defects

1. Title: A Clinical and Molecular Analysis of Branchio-Oculo-Facial Syndrome Patients in Russia Revealed New Mutations in TFAP2A.
The mean change in FLACC measurements was observed approximately 2 hours postoperatively. By 3 hours, evaluation. The median respiratory rate and heart rate remained elevated throughout the study period. The highest mean change in FLACC measurements was observed approximately 2 hours postoperatively. By 3 hours, heart rate returned to the median baseline level.

The median systolic blood pressure increased postoperatively and remained elevated throughout the study period. The highest mean change in FLACC measurements was observed approximately 2 hours postoperatively. By 3 hours.
postoperatively, the scores decreased. CONCLUSIONS: Although there was a large individual variability, the FLACC scores became reduced after 3 hours following surgical insertion of the DMA and the ECPR appliance.

Publication type: journal article
Source: CINAHL

4.Title: Association between maternal MTHFR polymorphisms and nonsyndromic cleft lip with or without cleft palate in offspring, a meta-analysis based on 15 case-control studies
Citation: International Journal of Fertility and Sterility, January 2015, vol./is. 8/4(463-480), 2008-076X;2008-0778 (01 Jan 2015)
Language: English
Abstract: Background: The methylenetetrahydrofolate reductase (MTHFR) is thought to be involved in the development of nonsyndromic cleft lip with or without cleft palate (NSCL/P). However, conflicting results have been obtained when evaluating the association between maternal MTHFR C677T and A1298C polymorphisms and the risk of NSCL/P. In light of this gap, a meta-analysis of all eligible case-control studies was conducted in the present study. Materials and Methods: A total of 15 case-control studies were ultimately identified after a comprehensive literature search and Hardy-Weinberg equilibrium (HWE) examination. Cochrane’s Q test and index of heterogeneity (I²) indicated no obvious heterogeneity among studies. Results: Fixed or random-effects models were used to calculate the pooled odds ratios (ORs). The results showed that the TT genotype in mothers increased the likelihood of having NSCL/P offspring 1.25 times (95% CI: 1.047-1.494) more than the CC homozygotes. Mean-while, maternal TT genotype increased the risk of producing NSCL/P offspring in recessive model (OR=1.325, 95% CI: 1.124-1.562). However, the CT heterozygote and the CT+TT dominant models had no association with NSCL/P offspring compared with the CC wild-type homozygote model. Subgroup analyses based on ethnicity indicated that maternal TT genotype increased the likelihood of having NSCL/P offspring in Whites (OR=1.308, 95% CI: 1.059-1.617) and Asians (OR=1.726, 95% CI: 1.090-2.733) in recessive model. Also, subgroup analyses based on source of control showed that mothers with the 677TT genotype had a significantly increased susceptibility of having NSCL/P children in hospital based population (HB) when compared with CC homozygotes (OR=1.248, 95% CI: 1.024-1.520) and under the recessive model (OR=1.324, 95% CI: 1.104-1.588). Furthermore, maternal A1298C polymorphism had no significant association with producing NSCL/P offspring (dominant model OR=0.952, 95% CI: 0.816-1.111, recessive model OR=0.766, 95% CI: 0.567-1.036). Conclusion: MTHFR C677T polymorphism is associated with the risk of generating NSCL/P offspring, and being a 677TT homozygote is a risk factor. MTHFR A1298C polymorphism was not associated with generating NSCL/P offspring. However, further work should be performed to confirm these findings.
Publication type: Journal: Article
Source: EMBASE
Full text: Available International Journal of Fertility and Sterility at International Journal of Fertility and Sterility

5.Title: Association of single-nucleotide polymorphisms in the IRF6 gene with non-syndromic cleft lip with or without cleft palate in the Xinjiang Uyghur population.
Citation: British Journal of Oral & Maxillofacial Surgery, March 2015, vol./is. 53/3(268-74), 0266-4356;1532-1940 (2015 Mar)
Author(s): Mijiti A, Ling W, Guli, Moming A
Language: English
Abstract: Our main aim was to investigate the association between the interferon regulatory factor (IRF6) gene and non-syndromic cleft lip and palate (nsCLP) in the Xinjiang Uyghur population. Twelve single nucleotide polymorphisms (SNP) were screened in a group of 100 patients with nsCLP and in a control group of 60 unaffected subjects by next generation sequencing using a MiSeq Benchtop Sequencer (Illumina). Our case-control association analysis showed that the SNP marker rs7545538 differed significantly in genotype (codominant model; CC compared with CG compared with GG; p=0.038) and allele frequencies (odds ratio (OR)=1.89, 95% CI 1.18-3.03, p=0.007) between patients with nsCLP and controls. Analysis of the recessive model of inheritance showed that distribution of the recessive model of rs7545538 (GG compared with CC+GC) was significantly higher in patients with nsCLP than in controls (OR=2.5, 95% CI 1.13-5.37, p=0.021) and had a borderline association with an increased risk of nsCLP (OR=2.5, 95% CI 1.13-5.37, p=0.021). Markers rs2235377 and rs2235371 also differed significantly in dominant and over-dominant models of inheritance (p=0.037) while increased G allele frequency was seen in SNP rs2235373 (p=0.03). A haplotype analysis showed four common haplotypes in Block 1: CCGGT>CCGAT>CACAT>TAGAC (in frequency). The 5-marker combination haplotype CCGGT was significantly more common in patients with nsCLP than in controls (p=0.032). In Block 2, the overall distribution of the haplotypes TAC and TAG predicted by the three SNP differed significantly between the patients with nsCLP and control subjects (p=0.009 and 0.003, respectively). Our
results showed that genetic polymorphism of the IRF6 gene is associated with increased risk of nsCLP in a Xinjiang Uyghur population. Copyright © 2014 The British Association of Oral and Maxillofacial Surgeons. Published by Elsevier Ltd. All rights reserved.

**Title:** Atypical expanded-spectrum hemifacial microsomia: a case report.

**Citation:** Cleft Palate-Craniofacial Journal, March 2015, vol./is. 52/2(240-5), 1055-6565;1545-1569 (2015 Mar)

**Author(s):** Magge KT, Kim J, Rogers GF, Oh AK

**Language:** English

**Abstract:** Objective: To present the case report of a patient with expanded hemifacial microsomia (HFM) that illustrates the complex interactions of these embryogenic factors and to review current theories and mechanism regarding the etiopathogenesis of HFM. Design and Method: We present the case of an African American girl who was born at full term by cesarean section and transferred to our institution for systemic malformations. Her craniofacial findings include holoprosencephaly, cleft lip and palate, low set and posteriorly rotated ears, flat midfacial features, micrognathia, left HFM with grade 1 microtia, hypoplastic mandible, and a small preauricular pharyngeal arch remnant. Systemic anomalies included ectopic kidney, atrial-septal defect, bilateral hip dysplasia, bilateral humeroradial fusion, bilateral club feet, and bilateral low-set thumbs with the right side also being triphalangeal. Genetic evaluation did not identify a molecular diagnosis or other known syndrome. Conclusions: Although vasculogenic disruption of the stapedial artery during early fetal gestation has been implicated in the etiology of HFM, the grouping and bilateral findings seen in our patient argue against this relatively simple and localized phenomenon. Instead, such diverse and widespread anomalies in the setting of expanded spectrum HFM seem to support the theory of a disorder in blastogenesis as the cause of HFM.

**Publication type:** Journal Article

**Source:** MEDLINE

**Full text:** Available The Cleft palate-craniofacial journal : official publication of the American Cleft Palate-Craniofacial Association at Cleft Palate-Craniofacial Journal

**Title:** Baraitser-Winter cerebrofrontofacial syndrome: delineation of the spectrum in 42 cases.

**Citation:** European Journal of Human Genetics, March 2015, vol./is. 23/3(292-301), 1018-4813;1476-5438 (2015 Mar)


**Language:** English

**Abstract:** Baraitser-Winter, Fryns-Aftimos and cerebrofrontofacial syndrome types 1 and 3 have recently been associated with heterozygous gain-of-function mutations in one of the two ubiquitous cytoplasmic actin-encoding genes ACTB and ACTG1 that encode beta- and -actins. We present detailed phenotypic descriptions and neuroimaging on 36 patients analyzed by our group and six cases from the literature with a molecularly proven actinopathy (9 ACTG1 and 33 ACTB). The major clinical anomalies are striking dysmorphic facial features with hypertelorism, broad nose with large tip and prominent root, congenital non-myopathic ptosis, ridged metopic suture and arched eyebrows. Iris or retinal coloboma is present in many cases, as is sensorineural deafness. Cleft lip and palate, hallux duplex, congenital heart defects and renal tract anomalies are seen in some cases. Microcephaly may develop with time. Nearly all patients with ACTG1 mutations, and around 60% of those with ACTB mutations have some degree of pachygyria with anteroposterior severity gradient, rarely lissencephaly or neuronal heterotopia. Reduction of shoulder girdle muscle bulk and progressive joint stiffness is common. Early muscular involvement, occasionally with congenital arthrogryposis, may be present. Progressive, severe dystonia was seen in one family. Intellectual disability and epilepsy are variable in severity and largely correlate with CNS anomalies. One patient developed acute lymphocytic leukemia, and another a cutaneous lymphoma, indicating that actinopathies may be cancer-predisposing disorders. Considering the multifaceted role of actins in cell physiology, we hypothesize that some clinical manifestations may be partially mutation specific. Baraitser-Winter cerebrofrontofacial syndrome is our suggested designation for this clinical entity.

**Publication type:** Journal Article

**Source:** MEDLINE
8. Title: Behavioral assessment of auditory processing disorder in children with non-syndromic cleft lip and/or palate.
Citation: International Journal of Pediatric Otorhinolaryngology, March 2015, vol./is. 79/3(349-55), 0165-5876;1872-8464 (2015 Mar)
Author(s): Ma X, McPherson B, Ma L
Language: English
Abstract: OBJECTIVE: Peripheral hearing disorders have been frequently described in children with non-syndromic cleft lip and/or palate (NSCL/P). However, auditory processing problems are rarely considered for children with NSCL/P despite their poor academic performance in general compared to their craniofacially normal peers. This study aimed to compare auditory processing skills, using behavioral assessment techniques, in school age children with and without NSCL/P. METHODS: One hundred and forty one Mandarin-speaking children with NSCL/P aged from 6.00 to 15.67 years, and 60 age-matched, craniofacially normal children, were recruited. Standard hearing health tests were conducted to evaluate peripheral hearing. Behavioral auditory processing assessment included adaptive tests of temporal resolution (ATTR), and the Mandarin pediatric lexical tone and disyllabic-word picture identification test in noise (MAPPID-N). RESULTS: Age effects were found in children with cleft disorder but not in the control group for gap detection thresholds with ATTR narrow band noise in the across-channel stimuli condition, with a significant difference in test performance between the 6 to 8 year group and 12 to 15 year group of children with NSCL/P. For MAPPID-N, the bilateral cleft lip and palate subgroup showed significantly poorer SNR-50% scores than the control group in the condition where speech was spatially separated from noise. Also, the cleft palate participants showed a significantly smaller spatial separation advantage for speech recognition in noise compared to the control group children. CONCLUSION: ATTR gap detection test results indicated that maturation for temporal resolution abilities was not achieved in children with NSCL/P until approximately 8 years of age compared to approximately 6 years for craniofacially normal children. For speech recognition in noisy environments, poorer abilities to use timing and intensity cues were found in children with cleft palate and children with bilateral cleft lip and palate compared to craniofacially normal children. Consequently, it is worthwhile to consider the potential for auditory processing disorder in when assessing the auditory status of children with NSCL/P. Copyright &© 2014 Elsevier Ireland Ltd. All rights reserved.
Publication type: Journal Article
Source: MEDLINE

9. Title: Comparative evaluation of salivary electrolytes in cleft lip and palate and healthy children
Citation: Research Journal of Pharmaceutical, Biological and Chemical Sciences, 2015, vol./is. 6/1(860-864), 0975-8585 (2015)
Author(s): Deshpande R.R., Sabhlok S., Patil V., Singh S., Chhabra R.S., Mutha M., Dasgupta S., Ankit K.
Language: English
Abstract: Saliva, a modern age biomarker for defects and pathological diseases, was used in our study to compare the levels of electrolytes in healthy individuals and the individuals having congenital defect. Saliva being a non-invasive method for determining the status of health can be the most widely used tool for the diagnosis of such cases. Materials & Method: Group A - Mixed dentition children free from any congenital defects. Group B - Mixed dentition children with cleft lip and palate. Method of saliva collection- The saliva was allowed to drool into the funnel held to the lower lip. Methods of laboratory analysis- Diluted saliva sample were subjected to inductively coupled plasma emission spectroscopy. The electrolytes were detected by liquid chromatography coupled with mass spectrometry (LC-MS) which were sodium, potassium, calcium and chlorine. Result and conclusion - It was observed that the level of potassium showed a linear increase in patients with cleft lip and palate. Thus this study helps us correlate the salivary electrolytic values between cleft lip and palate and healthy children in mixed dentition age group and showcases the importance of utilizing saliva as a biomarker.
Publication type: Journal: Article
Source: EMBASE

10. Title: Comparative validity and reproducibility study of various landmark-oriented reference planes in 3-dimensional computed tomographic analysis for patients receiving orthognathic surgery
Citation: PLoS ONE, February 2015, vol./is. 10/2, 1932-6203 (10 Feb 2015)
Author(s): Lin H.-H., Chuang Y.-F., Weng J.-L., Lo L.-J.
Language: English
Abstract: Background Three-dimensional computed tomographic imaging has become popular in clinical evaluation,
11. Title: Comparison of risk indicators of dental caries in children with and without cleft lip and palate deformities.

Citation: Contemporary Clinical Dentistry, January 2015, vol./is. 6/1(58-62), 0976-237X;0976-2361 (2015 Jan-Mar)

Author(s): Shashni R, Goyal A, Gauka K, Utreja AK, Ray P, Jena AK

Language: English

Abstract: OBJECTIVE: To test the hypothesis that there are no differences in various risk factors of dental caries among children with cleft lip and palate when compared to non-cleft high caries risk and non-cleft caries free children.

DESIGN: Seventy-three children in the age range of 4-9 years comprised three groups; Group-I (n = 23, children with cleft lip and palate), Group-II (n = 25, non-cleft high caries risk children) and Group-III (n = 25, non-cleft caries free children). Various risk factors for dental caries like type of oral hygiene practice, sugar exposures/day, developmental defects of enamel, caries activity, salivary streptococci mutans levels and lactobacilli levels were evaluated and compared among the three groups of children.

RESULTS: The mean deft score among Group-II children was significantly more (P < 0.01) as compared to the Group-I children. The mean deft + DMFT score among Group-I and Group-II children was comparable (P = 0.149). Developmental enamel defects were more among Group-I children as compared to Group-II and Group-III children (P < 0.01). Hypoplasia of the maxillary anterior teeth was more common among Group-I children as compared to Group-II (P < 0.05) and Group-III children (P < 0.001). The association between hypoplastic teeth and dental caries was significant (P < 0.05). The salivary acidogenic potential as evaluated by Snyder test was comparable among Group-I and Group-II children. The salivary streptococcus mutans levels in Group-I and Group-II children were higher when compared to lactobacillus counts.

CONCLUSION: The risk factors of dental caries among children with cleft lip and palate were more as compared to non-cleft high caries risk and non-cleft caries free children.

Publication type: Journal Article
Source: MEDLINE
Full text: Available ProQuest at Contemporary Clinical Dentistry

12. Title: Converting a nasoendotracheal tube to orotracheal, following fibreoptic intubation under general anaesthesia in a paediatric patient with complete cleft palate

Citation: Indian Journal of Anaesthesia, 2015, vol./is. 59/2(131-132), 0019-5049 (2015)

Author(s): Rajan S., Paul J., Kumar L.

Language: English

Publication type: Journal: Letter
Source: EMBASE
Full text: Available Journal of Ayurveda and Integrative Medicine at Indian Journal of Anaesthesia
13. **Title:** Craniofacial malformations and the orthodontist.

**Citation:** British Dental Journal, February 2015, vol./is. 218(3)(129-41), 0007-0610;1476-5373 (2015 Feb 16)

**Author(s):** Akram A, McKnight MM, Bellardie H, Beale V, Evans RD

**Language:** English

**Abstract:** This review article presents an overview of craniofacial malformations and the role of the orthodontist in their management. The first part of this article focuses on cleft lip and palate, followed by more complex deformities including craniosynostosis and craniofacial microsomia. The main features of these anomalies are discussed as well as the clinical problems seen in this group of patients. The emphasis is on the role of the orthodontist in the multidisciplinary management of these cases.

**Publication type:** Journal Article

**Source:** MEDLINE

14. **Title:** Curvilinear transformation of z-shaped upper lip scar by diamond-shaped excision in secondary cleft lip deformities: a photogrammetric evaluation.

**Citation:** Cleft Palate-Craniofacial Journal, March 2015, vol./is. 52(2)(143-51), 1055-6656;1545-1569 (2015 Mar)

**Author(s):** Han K, Jeong H, Choi TH, Kim JH, Son D

**Language:** English

**Abstract:** Purpose : The visible Z-shaped upper lip scar that occurs after the Tennison and Randall triangular flap technique remains a cleft stigma. Herein, we present our curvilinear transformation technique for the Z-shaped upper lip scar by diamond-shaped excision and evaluate the results using photogrammetric analyses. Patients and Methods : From 1997 to 2006, 23 patients with secondary cleft lip deformity with the visible Z-shaped upper lip scar underwent correction with the technique. The scar was excised in the diamond shape above the muscle. After curvilinear closure, the elongated length of the upper lip was excised just below the nostril sill, as the measured Cupid’s bow height discrepancy. The result was assessed by the authors’ standardized photogrammetry technique. Results : There was a statistically significant decrease between the preoperative central limb of the Z-shaped scar and the width of the postoperative curvilinear upper lip scar. The pre- and postoperative Cupid’s bow height differences were not statistically significant. Conclusions : The curvilinear transformation of the Z-shaped scar is an efficient procedure that provides (1) a significant decrease in the width of an upper lip scar to make it less conspicuous, (2) incorporation of the scar into the philtral column, (3) a biconcave natural philtral column shape in frontal view, and (4) formation of the natural concave philtral contour in profile view.

**Publication type:** Journal Article

**Source:** MEDLINE

Full text: Available **The Cleft palate-craniofacial journal:** official publication of the American Cleft Palate-Craniofacial Association at **Cleft Palate-Craniofacial Journal**

15. **Title:** Distraction osteogenesis and orthognathic surgery for a patient with unilateral cleft lip and palate.

**Citation:** American Journal of Orthodontics & Dentofacial Orthopedics, March 2015, vol./is. 147(3)(381-93), 0889-5406;1097-6752 (2015 Mar)

**Author(s):** Kim JH, Lee IH, Lee SM, Yang BE, Park IY

**Language:** English

**Abstract:** Maxillary deficiency is a common feature in patients with repaired cleft lip and palate. Orthognathic surgery has been the conventional approach for the management of cleft-related maxillary hypoplasia. However, for patients with a severe maxillary deficiency, orthognathic surgery alone has many disadvantages, such as high relapse rates of 25% to 40%, instability, limited amount of advancement, and a highly invasive surgical technique. As an alternative treatment method, distraction osteogenesis has been used successfully in the distraction of the mandible, the maxilla, the entire midface, and the orbits as well as the cranial bones, with stable outcomes. The type of distraction device, either external or internal, can be chosen based on the surgical goals set for the patient. In this study, we report on the use of a rigid external distraction device for maxillary advancement in a 22-year-old woman with a repaired unilateral cleft lip and palate and severe maxillary hypoplasia. After the distraction osteogenesis, 2-jaw surgery was performed to correct the maxillary yaw deviation and the mandibular prognathism. Copyright ©2015 American Association of Orthodontists. Published by Elsevier Inc. All rights reserved.

**Publication type:** Journal Article

**Source:** MEDLINE

16. **Title:** Double-loop technique using titanium molybdenum alloy wire for fabrication of nasal stents in
nasal alveolar molding therapy for cleft lip and palate patients.

Citation: Cleft Palate-Craniofacial Journal, March 2015, vol./is. 52/2(246-9), 1055-6656;1545-1569 (2015 Mar)

Author(s): Nagraj N, Nagarjuna M, Desai AK, Gandedkar N, Jayade B, Gopalakrishnan K

Language: English

Abstract: This article presents a technique for fabricating a modified nasoalveolar molding appliance using 0.032-inch titanium-molybdenum alloy wire for nasal stents incorporating a double loop. The nasal stents are included in the acrylic molding plate at the time the appliance is inserted. The acrylic retention button used in a conventional appliance is replaced by a simple retention hook fabricated using titanium-molybdenum alloy wire. This technique is an effective alternative to a conventional appliance, and it simplifies the appliance-modification process during follow-up visits.

Publication type: Journal Article

Source: MEDLINE

Full text: Available The Cleft palate-craniofacial journal : official publication of the American Cleft Palate-Craniofacial Association at Cleft Palate-Craniofacial Journal

17. Title: Early and late operation of cleft lip and intelligence quotient and psychosocial development in 3-7years.

Citation: Early Human Development, February 2015, vol./is. 91/2(149-52), 0378-3782;1872-6232 (2015 Feb)


Language: English

Abstract: BACKGROUND: Early and late operations of the cleft lip represent exposure to general anesthesia during the first year of life. The early exposure to the anesthetics may influence long term neurological outcome. Timing of the operation may also influence the quality of life as babies with early repair might be accepted better by their families. AIMS: The aim of the study was to compare outcomes between two groups of patients operated on for the cleft lip in the first year of life. STUDY DESIGN: Observational cohort study. SUBJECTS: Early repair group included patients operated on in the first eight days of life and late repair group those operated on between 3 and 10 months. OUTCOME MEASURES: Intelligence quotient (IQ) and psychosocial development of children who were operated on for cleft lip were compared at the age of 3-7 years. RESULTS: No differences were found between early (n=15) and late (n=17) repair group in terms of IQ. In both IQ was within the normal range: 100.00 (SD 13.867), 98.76 (SD 10.109), respectively. Significantly better results in physical functioning (P=0.042) and self-esteem (P=0.014) concepts in early repair group were found. CONCLUSIONS: We compared outcomes of two groups of patients operated on for cleft lip in the first year of life. The earlier anesthesia did not show a negative impact on intelligence quotient in 3-7 years compared to later anesthesia. The earlier repair of the cleft lip showed a significant positive impact on psychosocial development in 2 out of 13 concepts tested. Copyright © 2015 Elsevier Ireland Ltd. All rights reserved.

Publication type: Journal Article

Source: MEDLINE

18. Title: Ectrodactyly ectodermal dysplasia cleft lip/palate (EEC) syndrome in a family

Citation: International Journal on Disability and Human Development, February 2015, vol./is. 14/1(97-100), 2191-1231;2191-0367 (01 Feb 2015)

Author(s): Gaur A.K., Sharma R., Mhambre A.S., Mishra S.

Language: English

Abstract: The combination of ectrodactyly, ectodermal dysplasia, and cleft lip/palate is known as EEC syndrome. This disorder is characterized by deformities of fingers and toes (ectrodactyly), anomalies of hair, teeth, nails, nasolacrimal ducts, sweat glands, etc. (ectodermal dysplasia), and cleft lip/palate. Other less common findings include abnormalities of eyes and genitourinary anomalies, etc. In the patient, having ectrodactyly hand function depends on the type and severity of deformities. The function can be improved to a variable extent by surgery or assistive devices, aids, or appliances. We present a case of a 17-year-old with EEC that had familial presentation as the mother of the patient was also affected. The boy had hand function, which was enough for routine work, however, he wanted improvement in his hands to do fine work. His mother had almost normal hand function. The hand function of the boy improved with aids.

Publication type: Journal: Article

Source: EMBASE

19. Title: Expanding the genetic and phenotypic spectrum of popliteal pterygium disorders.

Citation: American Journal of Medical Genetics. Part A, March 2015, vol./is. 167/3(545-52), 1552-4825;1552-4833 (2015 Mar)
20. Title: Extent of palatal lengthening after cleft palate repair as a contributing factor to the speech outcome.

Citation: Annals of Plastic Surgery, March 2015, vol./is. 74/3(330-2), 0148-7043;1536-3708 (2015 Mar)

Author(s): Bae YC, Choi SJ, Lee JW, Seo HJ

Language: English

Abstract: Operative techniques in performing cleft palate repair have gradually evolved to achieve better speech ability with its main focus on palatal lengthening and accurate approximation of the velar musculature. The authors doubted whether the extent of palatal lengthening would be directly proportional to the speech outcome. Patients with incomplete cleft palates who went into surgery before 18 months of age were intended for this study. Cases with associated syndromes, mental retardation, hearing loss, or presence of postoperative complications were excluded from the analysis. Palatal length was measured by the authors' devised method before and immediately after the cleft palate repair. Postoperative speech outcome was evaluated around 4 years by a definite pronunciation scoring system. Statistical analysis was carried out between the extent of palatal lengthening and the postoperative pronunciation score by Spearman correlation coefficient method. However, the authors could not find any significant correlation. Although the need for additional research on other variables affecting speech outcome is unequivocal, we carefully conclude that other intraoperative constituents such as accurate reapproximation of the velar musculature should be emphasized more in cleft palate repair rather than palatal lengthening itself.

Publication type: Journal Article

Source: MEDLINE

Full text: Available Ovid at Annals of Plastic Surgery

21. Title: Fine Mapping of 6q23.1 Identifies TULP4 as Contributing to Clefts.

Citation: Cleft Palate-Craniofacial Journal, March 2015, vol./is. 52/2(128-34), 1055-6656;1545-1569 (2015 Mar)

Author(s): Vieira AR, de Carvalho FM, Johnson L, DeVos L, Swailes AL, Weber ML, Deely K

Language: English

Abstract: Objective: The aim of this work was to fine-map the region 6q23.1, which obtained suggestive linkage signal (logarithm of the odds [LOD] score = 2.22 under a recessive model) to cleft lip with or without cleft palate (CL+/P) in our previous genome-wide linkage scan to identify possible genetic variants that may contribute to CL+/P. Design: We used densely spaced markers spanning the entire 6q23.1 region to test for association with CL+/P in a family cohort sample. Setting: Clinical information and DNA samples were obtained from families in the Philippines at their homes or primary health care clinics. Participants: The study sample consisted of 477 subjects (224 females and 253 males), segregating isolated CL+/P, from 72 living in the same area in the Philippines. Main Outcome Measure: Overtransmission of alleles to persons born with CL+/P. Results: We found statistical evidence of association between a marker of TULP4 (rs651333) with CL+/P (P = .00007). Conclusions: Our results further support the linkage results for the chromosome 6q region and reveal a novel candidate gene for CL+/P.

Publication type: Journal Article

Source: MEDLINE
22. Title: Genome-wide Chromatin Mapping Defines AP2alpha in the Etiology of Craniofacial Disorders. 
Citation: Cleft Palate-Craniofacial Journal, March 2015, vol./is. 52/2(135-42), 1055-6656;1545-1569 (2015 Mar)  
Author(s): Enkhmandakh B, Bayarsaihan D  
Language: English  
Abstract: Objective: The aim of this study is to identify direct AP2alpha target genes implicated in craniofacial morphogenesis. Design: AP2alpha, a product of the TFAP2A gene, is a master regulator of neural crest differentiation and development. AP2alpha is expressed in ectoderm and in migrating cranial neural crest (NC) cells that provide patterning information during orofacial development and generate most of the skull bones and the cranial ganglia. Mutations in TFAP2A cause branchio-ocular-facial syndrome characterized by dysmorphic facial features including cleft or pseudocleft lip/palate. We hypothesize that AP2alpha primes a distinctive group of genes associated with NC development. Human promoter ChiP-chip arrays were used to define chromatin regions bound by AP2alpha in neural crest progenitors differentiated from human embryonic stem cells. Results: High-confidence AP2alpha-binding peaks were detected in the regulatory regions of many target genes involved in the development of facial tissues including MSX1, IRF6, TBX22, and MAFA. In addition, we uncovered multiple single-nucleotide polymorphisms (SNPs) disrupting a conserved AP2alpha consensus sequence. Conclusions: Knowledge of noncoding SNPs in the genomic loci occupied by AP2alpha provides an insight into the regulatory mechanisms underlying craniofacial development.  
Publication type: Journal Article  
Source: MEDLINE  
Full text: Available The Cleft palate-craniofacial journal : official publication of the American Cleft Palate-Craniofacial Association at Cleft Palate-Craniofacial Journal

23. Title: Hearing outcomes in patients with cleft lip/palate.  
Citation: Cleft Palate-Craniofacial Journal, March 2015, vol./is. 52/2(e23-31), 1055-6656;1545-1569 (2015 Mar)  
Author(s): Skuladottir H, Sivertsen A, Assmus J, Remme AR, Dahlen M, Vindenes H  
Language: English  
Abstract: Objective: Children with cleft lip and palate or cleft palate only have a high incidence of conductive hearing loss from otitis media with effusion. Studies demonstrating longitudinal results are lacking. This study was undertaken to investigate long-term longitudinal hearing outcomes of children with cleft lip and/or cleft palate and cleft palate only. Design: Retrospective chart review. Setting: Clinical charts of patients born with cleft lip and palate or cleft palate only in 1985 to 1994 who were referred to the cleft team in Bergen, Norway. Study findings include 15 years of follow-up. Participants: The study population consisted of 317 children of whom 159 had nonsyndromic cleft lip and palate and 158 had nonsyndromic cleft palate. Main Outcome Measures: Pure tone average calculated from pure tone audiometry at ages 4, 6, and 15 years. Results: The median pure tone average significantly improved with increasing age. For the cleft lip and palate group, the median pure tone average at ages 4, 6, and 15 years was 16 dB hearing level (HL), 13 dB HL, and 9 dB HL, respectively (P < .001). In the cleft palate group the median pure tone average at ages 4, 6, and 15 years was 15 dB HL, 12 dB HL, and 9 dB HL, respectively (P < .001). There was no significant difference in the hearing levels between the two groups. Patients who had surgical closure of the palate at age 18 months had a significantly better pure tone average outcome at age 15 compared with patients who had surgery at 12 months. Conclusions: Hearing improves significantly from childhood to adolescence in patients with cleft lip and palate and cleft palate only.  
Publication type: Journal Article  
Source: MEDLINE  
Full text: Available The Cleft palate-craniofacial journal : official publication of the American Cleft Palate-Craniofacial Association at Cleft Palate-Craniofacial Journal

24. Title: Hypohidrotic ectodermal dysplasia with ankylosis of temporomandibular joint and cleft palate: A rare presentation.  
Citation: Contemporary Clinical Dentistry, January 2015, vol./is. 6/1(110-2), 0976-237X;0976-2361 (2015 Jan-Mar)  
Author(s): Goyal M, Pradhan G, Gupta S, Kapoor S  
Language: English  
Abstract: The ectodermal dysplasias are a heterogeneous group of diseases, which have one or more anomalies of the hair, teeth, nails, and sweat glands. Hypohidrotic ectodermal dysplasia (HED) is the most common type and is usually transmitted as an X-linked recessive trait. It is characterized by classical triad of hypotrichosis,
anhidrosis/hypohidrosis, and hypodontia/anodontia. Here, we describe an Indian boy affected with HED and rare features including ankylosis of temporomandibular joint and cleft palate.

**Publication type:** Journal Article  
**Source:** MEDLINE  
**Full text:** Available ProQuest at Contemporary Clinical Dentistry

### 25. Title: Impact of primary palatoplasty on the maxillomandibular sagittal relationship in patients with unilateral cleft lip and palate: A systematic review and meta-analysis

**Citation:** International Journal of Oral and Maxillofacial Surgery, January 2015, vol./is. 44/1(50-56), 0901-5027;1399-0020 (01 Jan 2015)  
**Author(s):** Bichara L.M., Araujo R.C., Flores-Mir C., Normando D.  
**Language:** English  
**Abstract:** The study objective was to evaluate, through a meta-analysis, the impact of primary palatoplasty on the sagittal maxillary and mandibular relationship among patients with complete unilateral cleft lip and palate (UCLP). Electronic database and hand searches were performed. Controlled clinical trials involving non-syndromic UCLP patients were included. Selected papers had to include a group of patients undergoing lip and palate repair and a group undergoing lip repair only. Data heterogeneity was demonstrated and individual means, standard deviations, and sample sizes were collected and summarized using a random effects model meta-analysis. Although six articles were selected for the systematic review, only four were included in the meta-analysis due to large discrepancies in the standard surgical protocol. Only one variable assessing the intermaxillary relationship (A point-nasion-B point; ANB), maxillary position (sella-nasion-A point; SNA), and mandibular position (sella-nasion-B point; SNB) was common among the selected studies. No significant differences in SNA and SNB were indentified between patients undergoing lip surgery alone and those undergoing lip and palate surgery. Evaluation of ANB showed a small statistical standard mean difference of 0.36 degree. Impaired maxillary sagittal growth, observed in patients with UCLP, appears to be a basic consequence of lip surgical repair. Additional changes to the maxilla and mandible produced by palatal repair are minor. Methodologically rigorous controlled studies are needed to provide a stronger evidence-based basis for the surgical management of patients with UCLP.

**Publication type:** Journal: Review  
**Source:** EMBASE

### 26. Title: Long term speech outcomes following late cleft palate repair using the modified Furlow technique.

**Citation:** International Journal of Pediatric Otorhinolaryngology, December 2014, vol./is. 78/12(2275-80), 0165-5876;1872-8464 (2014 Dec)  
**Author(s):** Pasick CM, Shay PL, Stransky CA, Solot CB, Cohen MA, Jackson OA  
**Language:** English  
**Abstract:** OBJECTIVES: Published reports and previous studies from our institution have reported worse overall speech results, including significantly higher rates of persistent articulation errors, in patients undergoing palatoplasty at age >18 months. This study further investigates the effects of late repair on long term speech outcomes.METHODS: A retrospective review was performed of non-syndromic patients undergoing primary palatoplasty at age >18 months between 1980 and 2006 at our institution. Longitudinal speech results were compared based on reason for late repair and age at repair.RESULTS: Forty-one patients were greater than 18 months of age at the time of palatoplasty, and 24 fit criteria for longitudinal data analysis. There was a statistically significant improvement in nasality scores at Time Point 1 for international adoptees compared to the non-adopted population (p=0.04). Patients with submucosal clefts were found to have significantly less severe nasal emission scores at Time Point 1 compared to those with overt clefts (p=0.04), but not at Time Point 2. There were no significant differences between scores if repair was performed between 18 and 36 months or >36 months, nor any difference in incidence of articulation errors between subgroups of patients with late repair at either Time Point.CONCLUSION: Our experience demonstrates that cleft palate repair after 18 months of age is associated with a significantly increased incidence of articulation errors associated with VPI, irrespective of reason for late repair, highlighting the persistence of learned compensatory behaviors in speech and the importance of proceeding with early repair. Copyright © 2014 Elsevier Ireland Ltd. All rights reserved.

**Publication type:** Journal Article  
**Source:** MEDLINE

### 27. Title: Longitudinal study of growth of children with unilateral cleft lip and palate: 2 to 10 years of age.

**Citation:** Cleft Palate-Craniofacial Journal, March 2015, vol./is. 52/2(192-7), 1055-6656;1545-1569 (2015 Mar)  
**Author(s):** Marques IL, Nakashji J, Borgo HC, Martinelli AP, de Souza L, Dutka Jde C, Williams WN, Pegoraro-Krook MI
Abstract: Objective: To study the growth of children with complete unilateral cleft lip and palate (UCLP) from 2 to 10 years of age and to assess whether growth varied from that of children without UCLP (typical children). Design: Physical growth was one of the outcome measures of a National Institutes of Health-sponsored longitudinal, prospective clinical trial conducted by the University of Florida and the University of Sao Paulo. Setting: Hospital of Rehabilitation of Craniofacial Anomalies, University of Sao Paulo (HRAC-USP), Bauru, Brazil. Main Outcome Measures: Height and weight were prospectively measured for 360 healthy children with UCLP who were nonsyndromic, belonged to median socioeconomic status, and received health care at HRAC-USP. To compare growth of children with UCLP to that of typical children, growth curves for UCLP were developed and compared with World Health Organization curves for 2006 and 2007, which were used as reference for typical children. Third-degree polynomials were used to explain the relationship of length and weight with age. Confidence limits of 95% were used for the mean curve using the statistic Z ~ N (0,1). Results: Children with UCLP from 2 to 10 years old presented height and weight growth curves similar to those of typical children for both genders. Conclusion: Children with UCLP from 2 to 10 years old presented physical growth similar to that of typical children.

Publication type: Journal Article
Source: MEDLINE
Full text: Available The Cleft palate-craniofacial journal : official publication of the American Cleft Palate-Craniofacial Association at Cleft Palate-Craniofacial Journal

28. Title: Measurement of health-related and oral health-related quality of life among individuals with nonsyndromic orofacial clefts: a systematic review and meta-analysis.
Citation: Cleft Palate-Craniofacial Journal, March 2015, vol./is. 52/2(157-72), 1055-6656;1545-1569 (2015 Mar)
Author(s): Queiroz Herkrath AP, Herkrath FJ, Rebelo MA, Vettore MV

Abstract: Objective: To compare health-related quality of life and oral health-related quality of life between nonsyndromic individuals with and without cleft lip and/or cleft palate and to identify the most affected quality of life dimensions in individuals with cleft lip and/or palate. Design: Systematic review and meta-analysis were conducted. Of the 314 identified citations, 23 articles were submitted to quality assessment. Data from nine studies on health-related quality of life and six on oral health-related quality of life were extracted for meta-analysis. Main Outcome Measures: Pooled mean differences of health-related quality of life between adults with and without cleft lip and/or palate, pooled means of health-related quality of life dimensions of children and adults with cleft lip and/or palate and oral health-related quality of life dimensions of children and adolescents with cleft lip and/or palate with a 95% confidence interval were calculated. Results: Quality assessment revealed methodological differences between studies. Lack of subgroup stratification and absence of control for confounders were the main limitations. Heterogeneity was detected on the comparison of oral health-related quality of life and health-related quality of life between children with and without cleft lip and/or palate, and oral health-related quality of life between adolescents with and without cleft lip and/or palate. A random-effect model showed a significant difference on health-related quality of life between adults with and without cleft lip and/or palate (mean difference = 0.10; 95% confidence interval, 0.16 to 0.05). Psychological health (mean, 78.9; 95% confidence interval, 70.1 to 87.7) and vitality (mean, 68.1; 95% confidence interval, 48.0 to 88.1) were the most affected health-related quality of life dimensions in children and adults with cleft lip and/or palate, respectively. Means of health-related quality of life dimensions in children and adults with cleft lip and/or palate and oral health-related quality of life in children and adolescents with cleft lip and/or palate varied yet did not differ in indirect comparisons. Conclusion: The presence of cleft lip and/or palate negatively affected the health-related quality of life of adults, mainly on psychosocial dimensions.

Publication type: Journal Article
Source: MEDLINE
Full text: Available The Cleft palate-craniofacial journal : official publication of the American Cleft Palate-Craniofacial Association at Cleft Palate-Craniofacial Journal

29. Title: Methods of Identifying and Managing the Difficult Airway in the Pediatric Population.
Citation: AANA Journal, 01 February 2015, vol./is. 83/1(35-41), 00946354
Author(s): Belanger, Joshua, Kossick, Mark
Language: English
Abstract: The goal of this literature review is to provide the anesthesia practitioner with the skill set to detect and prepare for a difficult pediatric airway. The authors have reviewed and compiled information on some of the most common conditions that can predispose pediatric patients to a difficulty airway, such as macroglossia, mandibular...
hypoplasia, micrognathia, cervical instability, limited cervical movement, maxillary and midfacial hypoplasia, and cleft palate. This article provides an overview of preoperative assessment techniques, normal pediatric airway anatomy, and respiratory physiology. An emphasis is placed on some common syndromes and their related anatomical abnormalities that can compromise the airway, as well as anesthetic approaches recommended to successfully secure a potentially difficult airway.

Publication type: journal article
Source: CINAHL
Full text: Available AANA journal at AANA Journal
Full text: Available AANA journal at AANA Journal

30. Title: Modification of the dingman mouth gag for better visibility and access in the management of cleft palate.
Citation: Cleft Palate-Craniofacial Journal, March 2015, vol./is. 52/2(250-3), 1055-6656;1545-1569 (2015 Mar)
Author(s): Rao LP, Peter S
Language: English
Abstract: Palatal and pharyngeal surgeries often require wide visibility and access. Various mouth gags and retractors have been devised and many modifications suggested to optimize these surgeries. The Dingman mouth gag, one of the commonly used retractors, offers a lot of advantages in terms of good mouth opening, tongue retraction, self-retaining cheek retractors, and anchorage for sutures, but it has a main limitation in that it allows only limited visibility of the anterior palate and alveolus. Hence, a modification of the Dingman mouth gag is presented for better visibility of and accessibility to the anterior palate.
Publication type: Journal Article
Source: MEDLINE
Full text: Available The Cleft palate-craniofacial journal : official publication of the American Cleft Palate-Craniofacial Association at Cleft Palate-Craniofacial Journal

31. Title: MuSK: a new target for lethal fetal akinesia deformation sequence (FADS).
Citation: Journal of Medical Genetics, March 2015, vol./is. 52/3(195-202), 0022-2593;1468-6244 (2015 Mar)
Language: English
Abstract: BACKGROUND: Fetal akinesia deformation sequence syndrome (FADS, OMIM 208150) is characterised by decreased fetal movement (fetal akinesia) as well as intrauterine growth restriction, arthrogryposis, and developmental anomalies (eg, cystic hygroma, pulmonary hypoplasia, cleft palate, and cryptorchidism). Mutations in components of the acetylcholine receptor (AChR) pathway have previously been associated with FADS.METHODS AND RESULTS: We report on a family with recurrent fetal loss, where the parents had five affected fetuses/children with FADS and one healthy child. The fetuses displayed no fetal movements from the gestational age of 17 weeks, extended knee joints, flexed hips and elbows, and clenched hands. Whole exome sequencing of one affected fetus and the parents was performed. A novel homozygous frameshift mutation was identified in muscle, skeletal receptor tyrosine kinase (MuSK), c.40dupA, which segregated with FADS in the family. Haplotype analysis revealed a conserved haplotype block suggesting a founder mutation. MuSK (muscle-specific tyrosine kinase receptor), a component of the AChR pathway, is a main regulator of neuromuscular junction formation and maintenance. Missense mutations in MuSK have previously been reported to cause congenital myasthenic syndrome (CMS) associated with AChR deficiency.CONCLUSIONS: To our knowledge, this is the first report showing that a mutation in MuSK is associated with FADS. The results support previous findings that CMS and/or FADS are caused by complete or severe functional disruption of components located in the AChR pathway. We propose that whereas milder mutations of MuSK will cause a CMS phenotype, a complete loss is lethal and will cause FADS.Copyright Published by the BMJ Publishing Group Limited. For permission to use (where not already granted under a licence) please go to http://group.bmj.com/group/rights-licensing/permissions.
Publication type: Journal Article
Source: MEDLINE

32. Title: Nasal changes after orthognathic surgery for patients with prognathism and Class III malocclusion: Analysis using three-dimensional photogrammetry
Citation: Journal of the Formosan Medical Association, February 2015, vol./is. 114/2(112-123), 0929-6646;1876-0821 (01 Feb 2015)
Author(s): Worasakwutiphong S., Chuang Y.-F., Chang H.-W., Lin H.-H., Lin P.-J., Lo L.-J.
Language: English
Abstract: Background/Purpose: Orthognathic surgery alters the position of maxilla and mandible, and consequently changes the nasal shape. The nasal change remains a concern to Asian patients. The aim of this study was to measure the nasal changes using a novel three-dimensional photographic imaging method. Methods: A total of 38 patients with Class III malocclusion and prognathism were enrolled. All patients underwent two-jaw surgery with the standard technique. A nasal alar cinching suture was included at the end of procedure. Facial landmarks and nasal morphology were defined and measured from pre- and postoperative three-dimensional photographic images. Intra-rater errors on landmark identification were controlled. Patient’s reports of perceptual nasal changes were recorded. Results: The average width of the alar base and subalar remained similar after surgery. Alar width was increased by 0.74 mm. Nasal height and length remained the same. Nasolabial angle increased significantly. The area of nostril show revealed a significant increase and was correlated with a decrease of columella inclination. Nasal tip projection decreased significantly, by 1.99 mm. Preoperative nasal morphology was different between patients with and without cleft lip/palate, but most nasal changes were concordant. In the self-perception, 37% of patients reported improved nasal appearance, 58% reported no change, and 5% were not satisfied with the nasal changes. Conclusion: After the surgery, characteristic nasal changes occurred with an increase of nasolabial angle and nostril show, but a preserved nasal width. The majority of patients did not perceive adverse nasal changes.

Publication type: Journal: Article
Source: EMBASE

33. Title: Older adults’ experiences of living with cleft lip and palate: a qualitative study exploring aging and appearance.

Citation: Cleft Palate-Craniofacial Journal, March 2015, vol./is. 52/2(e32-40), 1055-6656;1545-1569 (2015 Mar)
Author(s): Hamlet C, Harcourt D
Language: English
Abstract: Objective: To explore older adults’ experiences of living with cleft lip and/or palate (CL/P), focusing on aging and appearance. Design: An exploratory-descriptive qualitative study. Participants: Individual semi-structured interviews (five via telephone, one face-to-face) conducted with six adults between the ages of 57 and 82 years. Results: Interview transcripts were analyzed using interpretative phenomenological analysis, which resulted in five themes: cleft across the life span, keeping up appearances, being one of a kind, resilience and protection, and cleft in an ever-changing society. A CL/P had an ongoing impact on participants’ lives, although its relevance shifted over time and some aspects of life (e.g., romantic relationships, decisions about having children of their own) were particularly affected. Participants seemed at ease living with CL/P as an older adult and considered it an important aspect of their identity, yet they still described feeling isolated at times and had little contact with other people with a cleft. They felt that health care could be more considerate to the needs of older people with a cleft, particularly around dentistry and information provision. Participants thought societal attitudes toward visible differences had changed over the years, but not necessarily for the better. A paradox was evident between reports of being noticed by others because of their cleft and simultaneously feeling invisible or ignored because of their age. Conclusions: These findings have implications for provision of care for older adults with a CL/P and for younger people with a CL/P who will be the older generation of the future.

Publication type: Journal Article
Source: MEDLINE
Full text: Available The Cleft palate-craniofacial journal : official publication of the American Cleft Palate-Craniofacial Association at Cleft Palate-Craniofacial Journal

34. Title: Optimization of dental status improves long-term outcome after alveolar bone grafting in unilateral cleft lip and palate.

Citation: Cleft Palate-Craniofacial Journal, March 2015, vol./is. 52/2(210-8), 1055-6656;1545-1569 (2015 Mar)
Author(s): Jabbari F, Skoog V, Reiser E, Hakelius M, Nowinski D
Language: English
Abstract: Objective: To evaluate the importance of dental status for long-term outcome after alveolar bone grafting in patients with unilateral cleft lip and palate. Design: Retrospective longitudinal study. Setting: Cleft lip and palate-craniofacial center, Uppsala University Hospital, Sweden. Patients: A total of 67 consecutive patients with unilateral complete cleft lip and palate. Interventions: Secondary alveolar bone grafting, prior to the eruption of the permanent canine, was performed at the average age of 10.0 years (range, 8.5 to 12.0 years). Main Outcome Measures: Alveolar bone height was evaluated with the modified Bergland index at 1 and 10 years after surgery. Results: Of the patients, 97% had modified Bergland index grade I and the remaining 3% had modified Bergland index grade II at 1 year after surgery. At 10 years’ follow-up, 43% showed modified Bergland index grade I; 55%, modified Bergland index grade II; and 2% (one patient), modified Bergland index grade III. The degree of dental
anomalies in the cleft area, such as enamel hypoplasia, incisor rotation, incisor inclination, canine inclination, and oral hygiene registered preoperatively, all correlated negatively to the modified Bergland index at 10 years after surgery. Enamel hypoplasia (p = 0.70195, P < .0001), followed by canine inclination (p = 0.55429, P < .0001), showed the strongest correlation to reduced bone height in the cleft area. Conclusions: In patients with unilateral cleft lip and palate, excellent results from secondary alveolar bone grafting in terms of bone height in the alveolar cleft tend to decrease with time. This seems to be correlated with factors that might to some extent be treated preoperatively through adequate planning and execution of the orthodontic treatment.

**Publication type:** Journal Article  
**Source:** MEDLINE  
**Full text:** Available The Cleft palate-craniofacial journal : official publication of the American Cleft Palate-Craniofacial Association at Cleft Palate-Craniofacial Journal

**35. Title:** Preliminary evidence of an interaction between the CRISPLD2 gene and non-syndromic cleft lip with or without cleft palate (nsCL/P) in Xinjiang Uyghur population, China  
**Citation:** International Journal of Pediatric Otorhinolaryngology, 2015, vol./is. 79/2(94-100), 0165-5876;1872-8464 (2015)  
**Author(s):** Mijiti A., Ling W., Maimaiti A., Tuerdi M., Tuercun J., Moming A.  
**Language:** English  
**Abstract:** Background Non-syndromic cleft lip with or without cleft palate (nsCL/P) is a common birth defect results from the genetic factors alone or interactions with environmental changes. Single nucleotide polymorphisms (SNPs) of CRISPLD2 gene have been found to be an etiologic factor in the development of nsCL/P. However, few studies to date focused on the association of genetic variation of CRISPLD2 gene with nsCL/P, and the results are conflicting based on the different study population. The main purpose of the present study was to investigate the association between the CRISPLD2 gene and nsCL/P in the Xinjiang Uyghur population. Methods Eighteen SNPs were screened in a group of 200 patients with nsCL/P and in a control group consisting of 180 unaffected individuals by next generation sequencing using MiSeq Benchtop Sequencer (Illumina). Results Our case-control association analysis showed that the SNP marker rs1546124 showed statistically significant differences in genotype (CC vs. CG vs. GG P = 0.004) and allele frequencies (49% vs. 37.8% OR = 1.58; 95% CI = 1.19-2.1, P = 0.002) between nsCL/P and controls. Under the recessive model of inheritance, the GG homozygotes had an OR of 2.4 (95% CI = 1.37-4.18; P = 0.002), and the result of significance was maintained even after multiple testing correction. Haplotype combinations of CACC were significantly more frequent in the nsCL/P patients than in controls (P = 0.037). Finally, the MDR analysis identified the two-SNP model including rs1546124 and rs4782675 as best combination of possibly interactive polymorphisms (P < 0.001). Conclusion Our results demonstrate that genetic polymorphism of CRISPLD2 gene is associated with an increased risk of nsCL/P in a Xinjiang Uyghur population.  
**Publication type:** Journal: Article  
**Source:** EMBASE

**36. Title:** Relationship between reduced folate carrier gene polymorphism and non-syndromic cleft lip and palate in Indian population.  
**Citation:** Journal of Maternal-Fetal & Neonatal Medicine, February 2015, vol./is. 28/3(329-32), 1476-4954;1476-4954 (2015 Feb)  
**Author(s):** Lakkakula B, Murthy J, Gurramkonda VB  
**Language:** English  
**Abstract:** UNLABELLED: Abstract Objective: Folate metabolism involves absorption, transport, modifications and interconversions of folates. The reduced folate carrier does not participate directly in folate metabolism but plays a major role in intracellular transport of metabolically active 5-methyltetrahydrofolate and maintains the intracellular concentrations of folate. The purpose of this study was to identify the prevalence of reduced folate carrier 1 (RFC1) A80G polymorphism and to further delineate its association with non-syndromic cleft lip and palate (NSCLP) in a south Indian population. METHODS: In the present case-control study, we studied RFC1 gene A80G polymorphism to evaluate its impact on NSCLP risk in south Indian population. Blood samples of 142 cases with NSCLP and 141 controls were collected and genotyped using PCR-RFLP. RESULTS: The genotype distribution in the control group followed Hardy-Weinberg equilibrium (p=0.633). The G allele frequency of cases was 64.8% (184/284) and was significantly lower than that found in the control group 56.4% (160/282). The genotype distributions between NSCLP cases and controls was not significantly different (p=0.131). The allelic model significantly increased the risk of NSCLP (G versus A; OR=1.40; 95% CI: 1.00-1.97; p=0.050). In subgroup analysis, the A80G variant showed significant association for the CLP group in dominant and allelic models. CONCLUSIONS: Altogether, our findings support the hypothesis that RFC1 A80G variant may contribute to NSCLP susceptibility in a south Indian population.
37. Title: Risk factors involved in orofacial cleft predisposition-review
Citation: Open Medicine, January 2015, vol./is. 10/1(163-175) (01 Jan 2015)
Author(s): Kawalec A., Nelke K., Pawlas K., Gerber H.
Language: English
Abstract: Clefts that occur in children are a special topic. Avoiding risk factors, and also an early diagnosis of cleft possibility can result in minimizing or avoiding them. If on the other hand when clefts occur they require a long-term, multistage specialized treatment. Etiology of clefts seems to be related to many factors. Factors such as genetic, environmental, geographic and even race factors are important. Identification of risk factors can lead to prevention and prophylactic behaviors in order to minimize its occurrence. Exposure to environmental factors at home and work that lead to cleft predisposition should not be disregarded. It seems that before planning a family it would be wise to consult with doctors of different specializations, especially in high-risk families with cleft history in order to analyze previous lifestyle. Clefts are very common in hereditary facial malformations and are causing a lot of other irregularities in the head and neck region. In this paper after a brief papers review authors present socio-geographic, environmental and also work place related factors that are influencing pregnant women condition and should be taken under serious consideration.

38. Title: Risks of congenital malformations in offspring exposed to valproic acid in utero: A systematic review and cumulative meta-analysis
Citation: Clinical Pharmacology and Therapeutics, February 2015, vol./is. 97/(S99-S100), 0009-9236 (February 2015)
Author(s): Kobayashi T., Tanoshima M., Tanoshima R., Beyene J., Koren G., Ito S.
Language: English
Abstract: BACKGROUND: Valproic acid (VPA) has been suspected to adversely affect fetal development. The recent surge of published data prompted us to systematically address: 1) time profiles of emergence of the VPA teratogenicity signals over the last 30 years; and 2) expand risk estimates of specific congenital malformations (CMs) associated with VPA which have not been addressed to date. METHODS: A systematic literature search was conducted on Medline, Embase classics plus Embase, and Cochrane Central Register of Controlled Trials between 1947 and May 2014. Cumulative and conventional meta-analyses were performed. Pooled relative risk (RR) and 95% confidence intervals of VPA monotherapy-associated risks of combined and specific major CMs, compared to other antiepileptic drugs (AEDs), were calculated. RESULTS: We identified 59 cohort studies. Cumulative meta-analyses showed the increased risk of combined major CMs associated with VPA exposure in utero has been statistically significant since 1990. Signals of significant risks of specific major CMs (neural tube defects, congenital heart defects, cleft lip/palate, genitourinary anomalies and musculoskeletal anomalies) all emerged between 1992 and 2006. Conventional meta-analyses showed RR of VPA-associated major CMs were 2 to 7 fold in combined and each of the specific CMs, compared to patients with other AED exposure. Subgroup analyses suggested lamotrigine conferred the lowest risk of major CMs among the 5 common AEDs. CONCLUSION: The significant risk signal of each of the major specific CMs associated with VPA has become apparent as early as 22 years ago. Our analysis confirms for the first time that VPA causes congenital heart defects, cleft lip/palate, genitourinary and musculoskeletal anomalies. (Table Presented).

39. Title: SLC26A2 disease spectrum in Sweden - high frequency of recessive multiple epiphyseal dysplasia (rMED)
Citation: Clinical Genetics, March 2015, vol./is. 87/3(273-278), 0009-9163;1399-0004 (01 Mar 2015)
Author(s): Makitie O., Geiberger S., Horemuzova E., Hagenas L., Mostrom E., Nordenskjold M., Grigelioniene G., Nordgren A.
Language: English
Abstract: Diastrophic dysplasia (DTD) is an autosomal recessive skeletal dysplasia caused by SLC26A2 mutations. Clinical features include short stature, joint contractures, spinal deformities, and cleft palate. SLC26A2 mutations also result in other skeletal dysplasias, including the milder recessive multiple epiphyseal dysplasia (rMED). DTD is overrepresented in Finland and we speculated that this may have influenced the prevalence and spectrum of SLC26A2-related skeletal conditions also in Sweden. We reviewed the patient registry at Department of Clinical Genetics, Karolinska University Hospital, Stockholm to identify subjects with SLC26A2 mutations. Seven patients
from six families were identified; clinical data were available for six patients. All but one patient had one or two copies of the Finnish SLC26A2 founder mutation IVS1+2T>C. Arg279Trp mutation was present in compound heterozygous form in five patients with phenotypes consistent with rMED. Their heights ranged from 2.6 to 1.4 standard deviation units below normal mean and radiographic features included generalised epiphyseal dysplasia and double-layered patellae. Two rMED patients had hypoplastic C2 and cervical kyphosis, a severe manifestation previously described only in DTD. Our study confirms a high prevalence of rMED in Sweden and expands the phenotypic manifestations of rMED.

Publication type: Journal: Article
Source: EMBASE

40. Title: The Association between Folate Pathway Genes and Cleft Lip With or Without Cleft Palate in a Chinese Population.
Citation: Biomedical & Environmental Sciences, February 2015, vol./is. 28/2(136-9), 0895-3988;0895-3988 (2015 Feb)
Author(s): Jin LL, Chen EJ, Hou W, Liu XH, Hu Y
Language: English
Abstract: NSCL/P is a common congenital defect and gene-environmental factors involve in this disorder. Periconceptional intake of folate may reduce the risk of NSCL/P. The present study investigated three SNPs (rs1801198, rs955516, and rs3733890) in three folate pathway genes, including TCN2, MTR, and BHMT among 481 patients and 558 healthy subjects. Rs955516 showed allelic association with NSCL/P. More patients carry rs955516 AA and rs3733890 AA genotypes. The gene-gene interaction test showed trans-phase combination effects for MTR and BHMT genes. Our study suggests that the interaction of MTR and BHMT genes play a vital role in the pathogenesis of NSCL/P in Chinese population. Copyright &©x9; 2015 The Editorial Board of Biomedical and Environmental Sciences. Published by China CDC. All rights reserved.
Publication type: Journal Article
Source: MEDLINE
Full text: Available Biomedical and environmental sciences : BES at Biomedical and Environmental Sciences

41. Title: The burden of selected congenital anomalies amenable to surgery in low and middle-income regions: cleft lip and palate, congenital heart anomalies and neural tube defects.
Citation: Archives of Disease in Childhood, March 2015, vol./is. 100/3(233-8), 0003-9888;1468-2044 (2015 Mar)
Author(s): Higashi H, Barendregt JJ, Kassebaum NJ, Weiser TG, Bickler SW, Vos T
Language: English
Abstract: OBJECTIVE: To quantify the burden of selected congenital anomalies in low and middle-income countries (LMICs) that could be reduced should surgical programmes cover the entire population with access to quality care.DESIGN: Burden of disease and epidemiological modelling.SETTING: LMICs from all global regions.POPULATION: All prevalent cases of selected congenital anomalies at birth in 2010.MAIN OUTCOME MEASURES: Disability-adjusted life years (DALYs).INTERVENTIONS AND METHODS: Surgical programmes for three congenital conditions were analysed: clefts (lip and palate); congenital heart anomalies; and neural tube defects. Data from the Global Burden of Disease 2010 Study were used to estimate the combination of fatal burden that could be addressed by surgical care and the additional long-term non-fatal burden associated with increased survival.RESULTS: Of the estimated 21.6 million DALYs caused by these three conditions in LMICs, 12.4 million DALYs (57%) are potentially addressable by surgical care among the population born with such conditions. Neural tube defects have the largest potential with 76% of burden amenable by surgery, followed by clefts (59%) and congenital heart anomalies (49%). Sub-Saharan Africa and South Asia have the greatest proportion of surgically addressable burden for clefts (68%), North Africa and Middle East for congenital heart anomalies (73%), and South Asia for neural tube defects (81%).CONCLUSIONS: There is an important and neglected role surgical programmes can play in reducing the burden of congenital anomalies in LMICs. Copyright Published by the BMJ Publishing Group Limited. For permission to use (where not already granted under a licence) please go to http://group.bmj.com/group/rights-licensing/permissions.
Publication type: Journal Article
Source: MEDLINE
Full text: Available ARCHIVES OF DISEASE IN CHILDHOOD at Archives of disease in childhood
Full text: Available ARCHIVES OF DISEASE IN CHILDHOOD at Salisbury District Hospital Healthcare Library

42. Title: The double unilimb z-plasty technique for whistler deformity repair in unilateral cleft lip patients: an anthropometric study.
Citation: Annals of Plastic Surgery, March 2015, vol./is. 74/3(324-9), 0148-7043;1536-3708 (2015 Mar)
OBJECTIVE: The purpose of this study was to evaluate the symmetry in lip and vermilion height after using the double unilimb Z-plasty method for whistler deformity repair.

DESIGN: This is a retrospective audit of 1 surgeon's outcome of 52 consecutive performed whistler deformity repairs.

SETTING: Data from the Outreach Surgical Center Program, Lima, Peru, were used.

PATIENTS: Since 2009, 52 adult patients with lip deformity related to unsatisfactory unilateral cleft lip repair were operated on using the double unilimb Z-plasty. All these patients met the study criterion of having anthropometric measurements performed at least 1 year postoperatively.

MAIN OUTCOME MEASURES: Data collection of lip and vermilion height was performed at the right and left side of the lip, immediately before the surgery (preoperative) and at least 1 year postoperatively. The lip measurements were obtained using calipers. ANALYSIS: The matched pair t test analyses were performed when the assumptions required were met. When the normality assumption was not met, the Wilcoxon signed rank test, a nonparametric test, was used to assess the statistical significance of differences between the cleft side and the noncleft side.

RESULTS: The study found no statistically significant differences between the right and left side in lip height (P = 0.51) and vermilion height (P = 0.57) after lip repair using the double unilimb Z-plasty technique measured at least 1 year postoperatively.

CONCLUSIONS: The findings suggest that the double unilimb Z-plasty technique is a good alternative to address the whistler deformity related to the unilateral cleft lip primary repair. This is a simple method, easy to perform by surgeons, for whistler deformity management in unilateral cleft lip patients.

Publication type: Journal Article
Source: MEDLINE
Full text: Available Ovid at Annals of Plastic Surgery
airways revealed that the NV and PV airways showed no difference between the cleft and noncleft subjects. It is interesting to postulate that the reported nasal abnormalities of the cleft patients do not produce a significant measurable effect on the overall volume, at least as shown in our study sample. In addition, the nasopharyngeal airways of patients are not larger than those of age-matched controls, at least at the ages that were sampled, after cleft palate repair.LEVEL OF EVIDENCE: 3b. Laryngoscope, 125:736-739, 2015.Copyright &© 2014 The American Laryngological, Rhinological and Otological Society, Inc.

**Publication type:** Journal Article

**Source:** MEDLINE

45. **Title:** Unusual foreign body in the nasal cavity of an adult with repaired cleft lip and palate.

**Citation:** Cleft Palate-Craniofacial Journal, March 2015, vol./is. 52/2(219-22), 1055-6656;1545-1569 (2015 Mar)

**Author(s):** Ravikumar N, GunaShekhar M, Prasad SR, Lalitha N, Raju PR, Natesh YA

**Language:** English

**Abstract:** Intranasal foreign bodies arising from dental clinical practice, especially in patients with cleft lip and palate (CLP) occur rarely and are very scarce in the literature. This article reports an unusual case of a dental impression material presenting as a foreign body in the nasal cavity of an adult with repaired CLP who presented for dental prosthetic rehabilitation. To our knowledge, this is only the second report presenting nasal foreign body in a cleft patient arising due to a dental impression procedure.

**Publication type:** Journal Article

**Source:** MEDLINE

**Full text:** Available The Cleft palate-craniofacial journal : official publication of the American Cleft Palate-Craniofacial Association at Cleft Palate-Craniofacial Journal

46. **Title:** Upper triangular flap method for primary repairs of incomplete unilateral cleft lip patients: minor to two-thirds way defects.

**Citation:** Annals of Plastic Surgery, March 2015, vol./is. 74/3(318-23), 0148-7043;1536-3708 (2015 Mar)

**Author(s):** Koh KS, Oh TS, Song JW

**Language:** English

**Abstract:** Incomplete unilateral cleft lips show a wide range of deformities, ranging from microform to near-complete clefts. Because there are different amounts and qualities of tissue present on the cleft and non-cleft sides, surgical approaches should make distinctions based on the remnant tissue. A new procedure using an upper triangular flap that combines characteristics of both rotation advancement and straight line repair was applied and the surgical results were reviewed. Between June 2007 and April 2011, 28 patients with minor to two-thirds way unilateral cleft lips [minor (n = 12), one-third (n = 2), halfway (n = 11), and two-thirds way (n = 3)] were subjected to the upper triangular flap method. The patients ranged in age from 62 days to 6 years (mean, 9 months). The average follow-up period was 25 months (range, 12-60 months). The repairs were successful in all 28 patients without complications. The scar was acceptable because it ran along the vertical philtral columns. During the follow-up period, long lip deformities and Cupid bow drooping were not observed in any of the patients. However, misalignment of the white skin roll was observed due to insufficient rotation at the cleft side in 1 patient. The repairs of minor to two-thirds way unilateral cleft lips using the upper triangular flap method allowed for a symmetric Cupid bow and philtrum. Moreover, this method allowed for satisfactory nostril sill reconstruction with acceptable scarring. The upper triangular flap method is recommended as an alternative to conventional methods for repair of minor to two-thirds way incomplete unilateral cleft lips.

**Publication type:** Journal Article

**Source:** MEDLINE

**Full text:** Available Ovid at Annals of Plastic Surgery

**Full text:** Available Ovid at Annals of Plastic Surgery

---

**Disclaimer and Feedback**

This current awareness bulletin contains a selection of information which is not intended to be exhaustive, and although library staff have made every effort to link only to reputable and reliable websites, the information contained in this bulletin has not been critically appraised by library staff. It is therefore the responsibility of the reader to appraise this information for accuracy and relevance.

This bulletin was produced by Caroline Thomas, Librarian, Salisbury NHS Foundation Trust Healthcare Library. If you have any comments to make about this bulletin please contact Caroline.Thomas@salisbury.nhs.uk.