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### Cochrane Systematic Reviews

**New Reviews – January 2015**

**Antibiotic prophylaxis for preventing infectious complications in orthognathic surgery**

### 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

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**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**

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1. Title: A bibliometric analysis of the 50 most cited papers in cleft lip and palate.
Citation: Journal of Plastic Surgery and Hand Surgery, February 2015, vol./is. 49/1(52-8), 2000-6764;2000-6764 (2015 Feb)
Author(s): Mahon NA, Joyce CW
Language: English
Abstract: Abstract Citation analysis is an established bibliometric method which catalogues papers according to the number of times they have been referenced. It is believed that the total number of citations an article receives reflects its importance among its peers. Never before has a bibliometric analysis been performed in the area of Cleft Lip and Palate. Our citation analysis creates a comprehensive list of the 50 most influential papers in this field. Journals specializing in Cleft Palate, Craniofacial, Plastic Surgery, Maxillofacial Surgery, Aesthetics and Radiology were searched to establish which articles most enriched the specialty over the past 70 years. The results show an interesting collection of papers which reveal developing trends in surgical techniques. These landmark papers mould and influence management and decision-making today.
Publication type: Journal Article
Source: MEDLINE

2.Title: A polymorphic marker associated with non-syndromic cleft lip with or without cleft palate in a population in Heilongjiang Province, northern China.
Citation: Archives of Oral Biology, February 2015, vol./is. 60/2(357-61), 0003-9969;1879-1506 (2015 Feb)
Author(s): Mi N, Hao Y, Jiao X, Zheng X, Shi J, Chen Y
Language: English
Abstract: OBJECTIVE: To further assess the roles of the ABCA4 (ATP-binding cassette, sub-family A, member 4) gene in NSCL/P, we investigated two tag SNPs in ABCA4 (rs481931 and rs560426) in a northern Chinese Han population where the prevalence of NSCL/P is high.MATERIALS AND METHODS: The two SNPs were examined for association with NSCL/P in 344 patients and 324 healthy controls. Peripheral blood samples were acquired at study enrollment, and DNA samples were extracted. SNPs were genotyped using a mini-sequencing (SNAPSHOT) method.RESULTS: We observed a significant correlation between the ABCA4 SNP rs560426 and NSCL/P (p=0.0041) but no evidence of association between rs481931 and NSCL/P. The G/G genotype at the rs560426 SNP in ABCA4 gene had an odd ratios of 2.39 (95%CI: 1.38-4.14, p=0.0041) compared with the A/A genotype, and a similar significance was noted in the CL/P subgroup (ORA/G=1.60, 95%CI: 1.09-2.34, ORG/G=2.96, 95%CI: 1.63-5.37 and ORG/G+A/G=1.80, 95%CI: 1.25-2.60, p=0.0007).CONCLUSIONS: Our study provides further evidence regarding the roles of genetic markers in ABCA4 in NSCL/P development in this northern Chinese Han population. G allele of rs560426 may be a risk factor for developing NSCL/P. Copyright 2014 Elsevier Ltd. All rights reserved.
Publication type: Journal Article
Source: MEDLINE

3.Title: A retrospective evaluation of the aesthetics of the nasolabial complex after unilateral cleft lip repair using the Tennison-Randall technique: A study of 44 cases treated in a single cleft center.
Citation: Journal of Cranio-Maxillo-Facial Surgery, December 2014, vol./is. 42/8(1679-83), 1010-5182;1878-4119 (2014 Dec)
Author(s): Iliopoulos C, Mitsimponas K, Lazaridou D, Neukam FW, Stelzle F
Language: English
Abstract: INTRODUCTION: Among numerous techniques that have been described for lip repair, the Tennison-Randall method has gained popularity over time and is preferred by many surgeons due to the predictability of the outcome. This study aims to evaluate the esthetic outcome reached in the nasolabial region following primary lip repair with the use of this method.MATERIALS AND METHODS: Forty-four patients with unilateral cleft lip (with or without alveolar cleft) were assessed retrospectively through a photographic evaluation by two clinicians with regard to the aesthetics of the lip and nose separately as anatomical subunits as well as of the nasolabial region as an anatomical complex. The collected data were statistically analyzed with regard to the cleft subtype and the performance of corrective surgeries for the lip and/or the nose.RESULTS: The method was associated with good results, especially when it comes to the appearance of the nose as an anatomical subunit, as well as of the nasolabial region as a complex, regarding cleft lip patients without an alveolar cleft.CONCLUSION: The Tennison-Randall technique proved to be a very satisfying method in terms of the esthetic long-term outcome in our patient collective. Copyright 2014 European Association for Cranio-Maxillo-Facial Surgery. Published by Elsevier Ltd. All rights reserved.
Publication type: Journal Article
Source: MEDLINE

4.Title: Analysis of genetic regulation and cytokine expressions of distraction osteogenesis reconstruction for cleft palate.
Citation: Journal of Craniofacial Surgery, November 2014, vol./is. 25/6(2231-6), 1049-2275;1536-3732 (2014 Nov)
Author(s): Bai N, Liu Y, Chen G, Zhu Z
Language: English

Abstract: Because cleft palate (CP) is one of the most common congenital deformities, surgeons have tried for a long time to achieve an ideal reconstruction of the palatal bone defects and restoration of muscle attachments. In this study, a new CP model on rhesus was established and corrected by an approach of distraction osteogenesis (DO), and then quantitative studies of regulation of osteogenesis genes and expression of alkaline phosphatase (ALP), insulin-like growth factor-I (IGF-I), osteopontin (OPN), and osteocalcin (OC) in different phases of new bone formation were performed. The CP models (23 animals) were established surgically. In the experimental group (21 animals), the tissue defects were repaired by means of DO at the rhythm of 0.4 mm twice per day. The specimens were retrieved in 1, 2, 4, 6, 8, 12, and 24 weeks (3 animals each) after completion of distraction. The IGF-I, ALP, OPN, and OC messenger RNA (mRNA) were detected by real-time reverse transcription polymerase chain reaction, and their proteins were then analyzed by enzyme-linked immunosorbent assay tests. The results are compared with those of the experimental control and empty control groups (2 animals each). In the distraction gap, the mRNA and protein expression levels of IGF-I and ALP were both highly upregulated and reaching apex in the early phase of new bone formation. Otherwise, the mRNA and protein expressions of OPN and OC demonstrated high level during intermediate and later remodeling stages. These results suggest that the reconstruction of CP bone defect by means of DO could get definitely intramembranous new bone formation and eventually quite normal bone structure via consecutive remodeling in situ.

Publication type: Journal Article
Source: MEDLINE

5.Title: Are interventions for accelerating orthodontic tooth movement effective?.
Citation: Evidence-Based Dentistry, December 2014, vol./is. 15/4(116-7), 1462-0049;1476-5446 (2014 Dec)
Author(s): Abdallah MN, Flores-Mir C
Language: English

Abstract: Data sourcesPubmed, Embase, Sciences Citation Index, Cochrane Central Register of Controlled Trials (CENTRAL) and grey literature database of SIGLE were searched from January 1, 1990 to August 20, 2011 with no language restrictions.Study selectionRandomised controlled trials (RCTs) or quasi-RCTs in which the participants were healthy and received additional interventions to conventional orthodontic treatment for accelerating tooth movements were included. Subjects with defects in oral and maxillofacial regions (ie, cleft lip/palate), dental pathologies and medical conditions were excluded.Data extraction and synthesisStudies were selected by two independent reviewers and disagreements were resolved by discussion with a third reviewer. The primary outcomes included accumulative moved distance (AMD) or movement rate (MR) and time required to move the tooth to its destination. Secondary outcomes were pain improvement, anchorage loss, periodontal health, orthodontic caries, pulp vitality and root resorption. The reviewers performed statistical pooling, where possible, according to a priori criteria on the basis of comparability of patient type, treatments and outcomes measured and risk of bias. The reviewers tested for heterogeneity, publication bias and sensitivity. A quality assessment test was conducted to evaluate the method used to measure AMD.ResultsThe authors selected seven RCTs and two quasi-RCTs, which included a total of 101 patients with an age range of 12-26.3 years. Eight studies compared four intervention methods to no intervention group (control group). From them, four studies assessed low laser therapy (LLL), two evaluated corticotomy (CC), one assessed electrical current therapy (EC) and one evaluated pulsed electromagnetic field (PEF). Another study compared dentoalveolar distraction (DAD) vs periodontal distraction (PDD).Quality assessment scores showed that only two studies were of high quality, five studies were of medium quality, while two studies were of low quality. All studies, except one, compared left and right sides of the same participant (split mouth design), and measured the AMD. The method for measuring AMD was reliable in three studies, relatively reliable in one study and unreliable in four studies. The authors only performed pooled AMD mean meta-analysis for the LLL studies. The meta-analysis showed pooled mean AMD of 0.32 (95% confidence interval (CI), 20.04, 0.68), 0.76 (95% CI, 20.14, 1.65), and 0.73 (95% CI, 20.68, 2.14) for one month, two months and three months, respectively. Two LLL studies showed no differences regarding periodontal health and two LLL studies showed no differences in root resorption between LLL intervention and control groups. Compared to control group, one study reported that CS had significantly higher MR and another study showed that CS exhibited larger AMD for one month, two months, three months and four months. Two studies revealed that CS did not show any difference in the periodontal health status. One study reported the EC showed significantly larger AMD for one month, whereas another study reported that PEF induced larger AMD for five + 0.6 months. DAD showed faster MR and less anchorage loss compared to PDD. Teeth remained vital in both DAD and PDD interventions and one out of six cases presented root resorption in the PDD group.ConclusionsAmong the five interventions corticotomy is effective and safe to accelerate orthodontic tooth movement, low-level laser therapy was unable to accelerate orthodontic tooth movement. The level of evidence
does not support whether electrical current and pulsed electromagnetic fields are effective in accelerating orthodontic tooth movement and dentoalveolar or periodontal distraction is promising in accelerating orthodontic tooth movement.

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

6. **Title:** Association between NOGGIN and SPRY2 polymorphisms and nonsyndromic cleft lip with or without cleft palate.  
**Citation:** American Journal of Medical Genetics. Part A, January 2015, vol./is. 167/1(137-41), 1552-4825;1552-4833 (2015 Jan)  
**Author(s):** Song T, Shi J, Guo Q, Lv K, Jiao X, Hu T, Sun X, Fu S  
**Language:** English  
**Abstract:** Nonsyndromic cleft lip with or without cleft palate (NSCLP) is a common congenital malformation with a worldwide prevalence rate of 0.4-2.0% among live births, depending on race and ethnic background. Single-nucleotide polymorphisms (SNPs) of genes may contribute to NSCLP risk, although the risk factors and pathogenesis of NSCLP remain unknown. The objective of this study was to investigate association of SNPs of noggin (NOG) and sprouty homolog 2 (SPRY2) with NSCLP risk. A total of 188 NSCLP patients and 228 healthy controls from northern China were recruited for genotyping of these SNPs using the SNaP shot method. The frequency of the NOG rs227731 genotype was significantly lower among NSCLP cases than among controls. Logistic regression analysis showed rs227731 CC genotype was associated with decreased NSCLP susceptibility (OR=0.31, 95% CI=0.12-0.80) compared to the AA homozygote. However, no association between SPRY2, SNPs, and NSCLP risk were observed in this cohort of patients. In conclusion, NOG rs227731 genotype was associated with decreased NSCLP risk in a Northern Chinese population.  
2014 Wiley Periodicals, Inc.  
2014 Wiley Periodicals, Inc.

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

7. **Title:** Bony defect of palate and vomer in submucous cleft palate patients.  
**Citation:** International Journal of Oral & Maxillofacial Surgery, January 2015, vol./is. 44/1(63-6), 0901-5027;1399-0020 (2015 Jan)  
**Author(s):** Ren S, Ma L, Zhou X, Sun Z  
**Language:** English  
**Abstract:** The aim of this study was to visualize bony defects of the palate and vomer in submucous cleft palate patients (SMCP) by three-dimensional (3D) computed tomography (CT) reconstruction and to classify the range of bony defects. Forty-eight consecutive non-operated SMCP patients were included. Diagnosis was based on the presence of at least one of three classical signs of SMCP: bifid uvula, a translucent zone in the midline of the soft palate, and a palpable 'V' notch on the posterior border of the bony palate. Patients were imaged using spiral CT. 3D reconstruction models were created of the palate and vomer. The sagittal extent of the bony cleft in SMCP was classified into four types: type I, no V-shaped hard palate cleft (8.3%); type II, cleft involving the partial palate (43.8%); type III, cleft involving the complete palate and extending to the incisive foramen (43.8%); type IV, cleft involving the complete palate and the alveolar bone (4.2%). The extent of the vomer defect was classified into three types: type A, vomer completely fused with the palate (8.3%); type B, vomer partially fused with the palate (43.8%); type C, vomer not fused with the palate up to the incisive foramen (47.9%). Significant variability in hard palate defects in SMCP is the rule rather than the exception. The association of velopharyngeal insufficiency with anatomical malformations may be complex. Copyright 2014 International Association of Oral and Maxillofacial Surgeons. Published by Elsevier Ltd. All rights reserved.  
**Publication type:** Journal Article  
**Source:** MEDLINE

8. **Title:** Botulinum toxin to improve results in cleft lip repair: a double-blinded, randomized, vehicle-controlled clinical trial.  
**Citation:** PLoS ONE [Electronic Resource], 2014, vol./is. 9/12(e115690), 1932-6203;1932-6203 (2014)  
**Author(s):** Chang CS, Wallace CG, Hsiao YC, Chang CJ, Chen PK  
**Language:** English  
**Abstract:** BACKGROUND: Most patients with facial scarring would value even a slight improvement in scar quality. Botulinum toxin A is widely used to alleviate facial dynamic rhytides but is also believed to improve scar quality by reducing wound tension during healing. The main objective was to assess the effect of Botulinum toxin on scars resultant from standardized upper lip wounds. METHODS: In this double-blinded, randomized, vehicle-controlled,
Prospective clinical trial, 60 consecutive consenting adults undergoing cleft lip scar revision (CLSR) surgery between July 2010 and March 2012 were randomized to receive botulinum toxin A (n = 30) or vehicle (normal saline; n = 30) injections into the subjacent orbicularis oris muscle immediately after wound closure. Scars were independently assessed at 6-months follow-up in blinded fashion using: Vancouver Scar Scale (VSS), Visual Analogue Scale (VAS) and photographic plus ultrasound measurements of scar widths. RESULTS: 58 patients completed the trial. All scar assessment modalities revealed statistically significantly better scars in the experimental than the vehicle-control group. CONCLUSION: Quality of surgical upper lip scars, which are oriented perpendicular to the direction of pull of the underlying orbicularis oris muscle, is significantly improved by its temporary paralysis during wound healing. TRIAL REGISTRATION: ClinicalTrials.gov NCT01429402.

Publication type: Journal Article
Source: MEDLINE
Full text: Available ProQuest at PLoS ONE
Full text: Available ProQuest at PLoS One

Citation: Dento-Maxillo-Facial Radiology, 2015, vol./is. 44/1(20140282), 0250-832X;0250-832X (2015)
Author(s): Kapila SD, Nervina JM
Language: English
Abstract: Since its introduction into dentistry in 1998, CBCT has become increasingly utilized for orthodontic diagnosis, treatment planning and research. The utilization of CBCT for these purposes has been facilitated by the relative advantages of three-dimensional (3D) over two-dimensional radiography. Despite many suggested indications of CBCT, scientific evidence that its utilization improves diagnosis and treatment plans or outcomes has only recently begun to emerge for some of these applications. This article provides a comprehensive and current review of key studies on the applications of CBCT in orthodontic therapy and for research to decipher treatment outcomes and 3D craniofacial anatomy. The current diagnostic and treatment planning indications for CBCT include impacted teeth, cleft lip and palate and skeletal discrepancies requiring surgical intervention. The use of CBCT in these and other situations such as root resorption, supernumerary teeth, temporomandibular joint (TMJ) pathology, asymmetries and alveolar boundary conditions should be justified on the basis of the merits relative to risks of imaging. CBCT has also been used to assess 3D craniofacial anatomy in health and disease and of treatment outcomes including that of root morphology and angulation; alveolar boundary conditions; maxillary transverse dimensions and maxillary expansion; airway morphology, vertical malocclusion and obstructive sleep apnoea; TMJ morphology and pathology contributing to malocclusion; and temporary anchorage devices. Finally, this article utilizes findings of these studies and current voids in knowledge to provide ideas for future research that could be beneficial for further optimizing the use of CBCT in research and the clinical practice of orthodontics.
Publication type: Journal Article
Source: MEDLINE

10. Title: Cephalometric analysis of craniofacial morphology and growth in unrepaired isolated cleft palate patients.
Citation: Journal of Cranio-Maxillo-Facial Surgery, December 2014, vol./is. 42/8(1853-60), 1010-5182;1878-4119 (2014 Dec)
Author(s): Xu Y, Yang C, Schreuder WH, Shi J, Shi B, Zheng Q, Wang Y
Language: English
Abstract: OBJECTIVE: The aim of this study is to analyze the craniofacial morphology in patients with unrepaired isolated cleft palate (UICP) at childhood, adolescence and adulthood, in order to assess the influence of nonsurgical factors on the craniofacial growth in these patients.MATERIAL AND METHODS: Lateral and posteroanterior cephalograms of 106 non-syndromic UICP patients and 102 normal matched controls were obtained and analyzed. Patients and controls were divided into three subgroups: children (<5 years), adolescents (12-14 years), and adults (>18 years).RESULTS: UICP patients in childhood showed a shortened cranial basal length; reduced bony nasopharyngeal height; short maxillary depth and height with a posterior positioned maxilla and an increased width of the nasal cavity, maxilla and orbit; and a shortened mandibular length and height. UICP patients in adulthood showed a normal nasopharyngeal and mandibular morphology. However, the patients in this subgroup still showed a shortened cranial basal length, and short maxillary depth and anterior height with increased width of the nasal cavity, maxilla and orbit.CONCLUSIONS: Craniofacial morphology and growth in patients with UICP were significantly affected by nonsurgical factors. Growth of the cranial base and upper face were absolutely reduced, while growth of the bony nasopharynx and mandible were only postponed. Copyright 2014 European Association for Cranio-Maxillo-Facial Surgery. Published by Elsevier Ltd. All rights reserved.
11. Title: Cephalometric evaluation of upper lip symmetry after functional unilateral cleft lip repair with the Kernahan and Bauer technique and primary cleft rhinoplasty
Citation: Israel Medical Association Journal, November 2014, vol./is. 16/11(693-696), 1565-1088 (01 Nov 2014)
Author(s): Margulis A., Alperson E., Billig A.
Language: English
Abstract: Background: Cleft lip repair with the Millard technique has undergone many modifications throughout the years, yet analysis of the successes of these various methods is still lacking. Objectives: To make a quantitative evaluation of the outcomes obtained after unilateral cleft lip surgical repair using the Kernahan and Bauer technique with primary rhinoplasty. Methods: Five anatomical parameters for evaluating upper lip and nostril symmetry were compared between the cleft and the normal side at least 1 year post-surgery in 23 children who underwent unilateral cleft lip repair with this particular technique. Results: Surgical success (defined as a 10% or less deviation between the cleft and contralateral side) was achieved for four of the five parameters: distance between oral commissure and peak of cupid's bow, nasal sill width, distance between peak and lowest point of Cupid's bow, and vertical distance between the highest point of the philtral column and lowest point of the upper lip. Surgical success was not achieved for the last parameter: length of philtral column. Conclusions: Unilateral cleft lip repair using the Kernahan and Bauer technique with primary cleft rhinoplasty is mostly successful when aiming to achieve symmetry between the cleft and the normal side of the upper lip. Success was elusive in achieving symmetry between the philtral columns despite an overall average difference of only 1.2 mm.

12. Title: Changing strategy and implementation of a new treatment protocol for cleft palate surgery in "Maria Sklodowska Curie" (MSC) Children’s Hospital, Bucharest, Romania.
Citation: Journal of Plastic Surgery and Hand Surgery, December 2014, vol./is. 48/6(356-61), 2000-6764;2000-6764 (2014 Dec)
Author(s): Spataru R, Mark H
Language: English
Abstract: In "Maria Sklodowska Curie" (MSC) Children's Hospital, Bucharest, Romania, cleft palate repair has been performed according to von Langenbeck since 1984. The speech was good in most patients but wide clefts had a high percentage of fistulas, abnormal speech due to short length and limited mobility of the soft palate. In 2009, the protocol was changed to Gothenburg Delayed Hard Palate Closure, (DHPC) technique. The present evaluation was performed to study the implementation of this technique. One hundred and sixty-eight patients with cleft palate were admitted, 89 isolated cleft palate (ICP), 53 unilateral (UCLP) and 26 bilateral (BCLP). In these, 228 surgical interventions were performed. Soft Palate Repair (SPR) and Hard Palate Repair (HPR) were performed with the DHPC procedure. The transfer to this technique was successfully performed in three steps: one team visit to Gothenburg by a surgeon from MSC and two visits by surgeons from Gothenburg to the MSC. Patients with SPR and HPR were operated on without major complications and there were no differences in results between Gothenburg surgeons and MSC surgeons. The interventions with SPR and HPR technique were proven to be easy to teach and learn and successfully performed without major complications. For cleft patients at MSC hospital it has meant earlier surgery, less re-operations and complications. This report shows a successful change of strategy for palatal repair with improved outcome regarding surgery. In future, speech and growth will be followed on a regular basis and will be compared with results from the Gothenburg Cleft Team.

13. Title: Cleft palate: A clinical review.
Citation: Birth Defects Research. Part C, Embryo Today: Reviews, December 2014, vol./is. 102/4(333-42), 1542-975X;1542-9768 (2014 Dec)
Author(s): Shkoukani MA, Lawrence LA, Liebertz DJ, Svider PF
Language: English
Abstract: Orofacial clefts, including cleft palates (CP), are one of the most common birth defects. CP have a multiplicity of effects on the individual and society in terms of economic costs, loss of productivity, psychosocial effects, and increased morbidity and mortality at all stages of life. Embryological development of the palate is well delineated, with developments in the last decade regarding the biomolecular processes involved. Etiology is
complex, involving a number of genetic and environmental factors. Various techniques can be employed for the repair of CP, depending on whether the cleft is of the primary or secondary palate, the width of the cleft, whether lengthening of the palate is necessary, and with regard to concerns of disruption of midfacial growth. All surgical techniques have the goals of restoring functional speech, swallowing, and aesthetics. A multidisciplinary team is necessary for the long-term pre- and postoperative care of CP patients to handle complications, associated anomalies, and to optimize function and quality of life. Birth Defects Research (Part C) 102:333-342, 2014. 2014 Wiley Periodicals, Inc. 2014 Wiley Periodicals, Inc.

**Publication type:** Journal Article

**Source:** MEDLINE

14. Title: Clinical and genetic study on 356 Brazilian patients with a distinct phenotype of cleft lip and palate without alveolar ridge involvement.

**Citation:** Journal of Cranio-Maxillo-Facial Surgery, December 2014, vol./is. 42/8(1952-7), 1010-5182;1878-4119 (2014 Dec)

**Author(s):** Alvarez CW, Guion-Almeida ML, Richieri-Costa A

**Language:** English

**Abstract:** Oral clefts include cleft lip (CL), cleft lip with cleft palate (CLP) and cleft palate (CP), with wide variations in clinical presentation and degree of severity. We described a sample of individuals with CL and CP without alveolar arch involvement (CL + CP) to verify if the characteristics of this group are distinct from those with CL with or without CP (CL/P) described in literature. The sample was composed of 356 patients with CL + CP, registered at HRCA-USP, Bauru-SP-Brazil. The following characteristics were investigated: sex ratio, parental age at the time of conception, parental consanguinity, familial recurrence, laterality of the cleft and associated anomalies. A subgroup of 30 individuals with microforms of CL and CP were taken from the sample and compared with the remaining cases. Statistical differences were found between this CL + CP sample and the literature data for groups with CL/P regarding laterality, sex ratio, consanguinity, familial recurrence, and the presence of associated anomalies. The microform sample showed a statistical difference in paternal age. In most evaluated aspects, this sample presents similar characteristics to the consulted literature data for CL/P; as do the group of microform cleft cases when compared with the remaining CL + CP sample in this study. Microforms of cleft can represent a target group for investigation into the embryogenetic mechanisms of oral clefts and their phenotypic variability. Copyright 2014 European Association for Cranio-Maxillo-Facial Surgery. Published by Elsevier Ltd. All rights reserved.

**Publication type:** Journal Article

**Source:** MEDLINE

15. Title: Comparative Effectiveness Studies Examining Patient-Reported Outcomes among Children with Cleft Lip and/or Palate: A Systematic Review.

**Citation:** Plastic & Reconstructive Surgery, January 2015, vol./is. 135/1(198-211), 0007-1226;1529-4242 (2015 Jan)

**Author(s):** Ranganathan K, Vercrer CJ, Warschauysky SA, MacEachern MP, Buchman SR, Waljee JF

**Language:** English

**Abstract:** BACKGROUND: Health care policy makers are increasingly encouraging comparative effectiveness research. Little is known regarding comparative studies among children with cleft lip and/or palate. Cleft lip and/or palate profoundly influences self-perception and social functioning, and patient-reported outcomes provide a unique perspective on the success of reconstruction. The purpose of this study was to systematically review the literature regarding patient-reported outcomes among patients with cleft lip and/or palate.METHODS: The authors reviewed articles from MEDLINE, Embase, and Psycinfo that examined the use of patient-reported outcome instruments for cleft lip and/or palate. Studies of patients with cleft lip and/or palate across any age that described the use of patient-completed measures in patient and control populations were included. A research librarian confirmed the search, and two independent, blinded reviewers performed full-text review.RESULTS: The authors identified 1979 articles and selected 30 for inclusion. Forty-two different assessment tools were used to analyze factors such as self-esteem, behavior, and social support. The Strengths and Difficulties Questionnaire was most commonly used (n = 7), followed by the Childhood Experience Questionnaire (n = 5), and the Satisfaction with Appearance survey (n = 4). Barriers to analysis included lack of standardization of survey administration, effect of publication bias, and variations in patient populations between individual studies.CONCLUSIONS: Comparative studies of patient-reported outcomes among patients with cleft lip and/or palate are infrequent. Many instruments exist to measure patient-reported outcomes in this population, but no specific standard exists. Identifying efficient and targeted forms of instrument selection and administration will enhance comparative studies among children with cleft lip and/or palate.CLINICAL QUESTION/LEVEL OF EVIDENCE: Diagnostic, III.

**Publication type:** Journal Article
16. Title: Comparison of Two Models of Surgical Care for Patients with Cleft Lip and Palate in Resource-challenged Settings.

Citation: World Journal of Surgery, January 2015, vol./is. 39/1(47-53), 0364-2313;1432-2323 (2015 Jan)

Author(s): Rossell-Perry P, Segura E, Salas-Bustierna L, Cotrina-Rabanal O

Language: English

Abstract: BACKGROUND: The Peruvian health system is limited in providing specialized care for patients with clefts because there are an insufficient number of hospitals and few specially trained doctors in rural areas of the country. The most common model of care in these areas is the surgical mission wherein experienced cleft surgeons perform surgeries and teach local doctors. The purpose of this research was to identify the differences in outcome between the surgical mission trip and the referral center model of care provided by the same team. METHODS: A retrospective analysis (2002-2012) was performed on data from surgical outcomes provided by the Outreach Surgical Center Lima that utilized both models of care (surgical mission and referral center). A total of 935 procedures were performed in 680 patients with clefts who were treated by the Outreach Surgical Center Program Lima since 2002. Patients in both groups were identified from our records (medical records and screening-day registries). All patients underwent a physical examination, had photographs taken, and any unfavorable results and complications were documented. Comparison of categorical variables (including outcomes) between care models was performed using Pearson's chi (2) test or Fisher's exact test when appropriate. In all cases a two-tailed test was performed and the p value for rejecting the null hypothesis (no difference or no association) was set at 0.05. RESULTS: We found significant differences between the two models of care with respect to unilateral cleft lip and cleft palate dehiscence (p = 0.02 and p = 0.04, respectively), palate postoperative hemorrhage (p < 0.01), and palatal fistula (p < 0.01) outcomes. DISCUSSION: Differences in observed surgical outcomes between the two models might be attributed to the surgeon's performance and/or the patient's age, and these factors are also considered with respect to the model of care. Limitations in long-term medical evaluation at each site should be identified and strategies to improve surgical outcomes must be developed to ensure that patients served by surgical missions obtain the same results achieved at a referral center. LEVEL OF EVIDENCE: Therapeutic III.

Publication type: Journal Article

Source: MEDLINE

17. Title: Complete trisomy 9 with unusual phenotypic associations: Dandy-Walker malformation, cleft lip and cleft palate, cardiovascular abnormalities

Citation: Taiwanese Journal of Obstetrics and Gynecology, December 2014, vol./is. 53/4(592-597), 1028-4559;1875-6263 (01 Dec 2014)

Author(s): Tonni G., Lituania M., Chitayat D., Bonasoni M.P., Keating S., Thompson M., Shannon P.

Language: English

Abstract: Objective: Trisomy 9 is a rare chromosomal abnormality usually associated with first-trimester miscarriage; few fetuses survive until the second trimester. We report two new cases of complete trisomy 9 that both present unusual phenotypic associations, and we analyze the genetic pathway involved in this chromosomal abnormality. Case report: The first fetus investigated showed Dandy-Walker malformation, cleft lip, and cleft palate at the second trimester scan. Cardiovascular abnormalities were characterized by a right-sided, U-shaped aortic arch associated with a ventricular septal defect (VSD). Symmetrical intrauterine growth restriction and multicystic dysplastic kidney disease were associated findings. The second fetus showed a dysmorphic face, bilateral cleft lip, hypoplastic corpus callosum, and a Dandy-Walker malformation. Postmortem examination revealed cardiovascular abnormalities such as persistent left superior vena cava draining into the coronary sinus, membranous ventricular septal defect, overriding aorta, pulmonary valve with two cusps and three sinuses, and the origin of the left subclavian artery distal to the junction of ductus arteriosus and aortic arch. Conclusion: Complete trisomy 9 may result in a wide spectrum of congenital abnormalities, and the presented case series contributes further details on the phenotype of this rare aneuploidy.

Publication type: Journal: Article

Source: EMBASE

18. Title: Comprehensive orthodontic treatment of adult patient with cleft lip and palate.

Citation: Case Reports in Dentistry, 2014, vol./is. 2014/(795342), 2090-6447 (2014)

Author(s): Leiva Villagra N, Munoz Domon M, Veliz Mendez S
The aim of the paper is to present full orthodontic treatment of an operated cleft lip adult patient. Case Report. An 18-year-old patient consulted for severe crowded teeth. He comes from a poor family. At that time he already had four operations (velum, palate, lip, and myringotomy). Treatment included maxillary expansion, tooth extraction, and fixed orthodontic, as well as kinesiology and speech therapy treatment. A multidisciplinary approach allowed us to achieve successfully an excellent result for this patient and gave him a harmonic smile and an optimal function without orthognathic surgery. Two years after treatment, occlusion remains stable.

Publication type: Journal Article
Source: MEDLINE
Full text: Available ProQuest at Case Reports in Dentistry

19. Title: Comprehensive treatment approach for bilateral cleft lip and palate in an adult with premaxillary osteotomy, tooth autotransplantation, and 2-jaw surgery.
Citation: American Journal of Orthodontics & Dentofacial Orthopedics, January 2015, vol./is. 147/1(114-26), 0889-5406;1097-6752 (2015 Jan)
Author(s): Kokai S, Fukuyama E, Sato Y, Hsu JC, Takahashi Y, Harada K, Ono T
Language: English
Abstract: We report the successful treatment of a woman aged 25 years 3 months with bilateral cleft lip and palate. She had a protruded premaxilla, collapsed posterior segments, wide alveolar defects with oronasal fistulae, a congenital missing tooth, and severe facial asymmetry with a transverse occlusal cant. The comprehensive treatment approach included (1) premaxillary osteotomy combined with alveolar bone grafting to reposition the premaxilla and minimize the wide alveolar defects, (2) autotransplantation of a tooth with complete root formation to the grafted bone region to restore the missing tooth without a prosthesis such as a dental implant or bridge, and (3) 2-jaw surgery to improve facial asymmetry. The premaxillary osteotomy was managed orthodontically, in combination with bone grafting. The results suggest that surgical orthodontic treatment with tooth autotransplantation might be useful to improve the occlusion and facial esthetics without prosthetics. Copyright 2015 American Association of Orthodontists. Published by Elsevier Inc. All rights reserved.
Publication type: Journal Article
Source: MEDLINE

20. Title: Dental arch relationships assessed by GOSLON Yardstick in children with clefts in Northern Finland.
Citation: European Journal of Paediatric Dentistry, December 2014, vol./is. 15/4(389-91), 1591-996X;1591-996X (2014 Dec)
Author(s): Harila V, Ylikontiola LP, Sandor GK
Language: English
Abstract: AIM: Our purpose was to evaluate the dental arch relationships using the GOSLON Yardstick in children with cleft lip and or cleft palate in Northern Finland. MATERIALS AND METHODS: The subjects consisted of 62 Finnish patients (36 girls and 26 boys) with clefts born between 1995-2005 in the Northern Ostrobothnia Hospital District of Finland. There were 36 patients with cleft palate, 9 with unilateral cleft lip and palate, 6 with cleft in soft palate, 5 with bilateral cleft lip and palate, 2 with cleft lip and 4 with submucous clefts. The study casts were obtained at the mean age of 6.3 years (5.8-7.8 years) and the cases were selected randomly. The dental arch relationships were assessed by the GOSLON Yardstick method by one calibrated researcher. RESULTS: After the assessment, 77.1% of cases were allocated to categories 1 and 2 (excellent and good), 10.4% category 3 (fair), and 12.5% categories 4 and 5 (poor and very poor). Patients with cleft palate had good prognosis in 84.6% of the cases. Of the patients with soft palate clefts and unilateral cleft lip and palate, 66.7% were allocated to categories 1 and 2. Bilateral cleft lip and palate patients had the poorest prognosis. Patients with submucous cleft and cleft lip had all good prognosis. CONCLUSION: The GOSLON Yardstick is a useful method for assessing dental arch relationships and treatment prognosis not only in cleft lip and palate patients, but also in cleft palate patients.
Publication type: Journal Article
Source: MEDLINE

21. Title: DLX1 and MMP3 contribute to oral clefts with and without positive family history of cancer.
Citation: Archives of Oral Biology, February 2015, vol./is. 60/2(223-8), 0003-9969;1879-1506 (2015 Feb)
Author(s): Saboia TM, Reis MF, Martins AM, Romanos HF, Tannure PN, Granjeiro JM, Vieira AR, Antunes LS, Kuchler EC, Costa MC
Language: English
Abstract: OBJECTIVE: It has been suggested that oral clefts and cancer share a common genetic background. This
study aimed to investigate the epidemiological and molecular association between oral clefts and cancer.

METHODS: One hundred forty-eight nuclear families with oral clefts and 162 subjects with no birth defect were recruited. Data on self-reported family history of cancer among first, second, and third degree relatives of each patient were collected via a structured questionnaire. We also investigated the association between polymorphisms in the genes AXIN2, BMP2, BMP4, BMP7, DLX1, DLX2, and MMP3 and oral cleft with and without history of cancer. Markers in these genes were genotyped using real time PCR. Chi-square and t-test were used to assess the differences among self-reported family history of cancer between oral cleft and non-cleft individuals. The transmission disequilibrium test (TDT) was used to analyze the distortion of the inheritance of alleles from parents to their affected offspring.

RESULTS: Families with oral clefts had an increased risk of having a family history of cancer (p=0.01; odds ratio=1.79; 95% confidence interval, 1.07-1.87). TDT results showed an association between DLX1 and cleft lip and palate, in which the A allele was undertransmitted (p=0.022). For MMP3, G was undertransmitted among affected progeny (p=0.019) in cleft palate subgroup.

CONCLUSION: Oral clefts were associated with positive self-reported family history of cancer and with variants in DLX1 and MMP3. The association between oral clefts and cancer raises interesting possibilities to identify risk markers for cancer. Copyright 2014 Elsevier Ltd. All rights reserved.

Publication type: Journal Article
Source: MEDLINE

22. Title: Effective retropulsion and centralization of the severely malpositioned premaxilla in patients with bilateral cleft lip and palate: A novel modified presurgical nasoalveolar molding device with retraction screw.

Citation: Journal of Cranio-Maxillo-Facial Surgery, December 2014, vol./is. 42/8(1903-8), 1010-5182;1878-4119 (2014 Dec)

Author(s): Li W, Liao L, Dai J, Zhong Y, Ren L, Liu Y

Language: English

Abstract: PURPOSE: A novel, modified presurgical nasoalveolar molding (MPNAM) device with retraction screw was designed and used in patients with bilateral complete cleft lip and palate (BCCLP) to rapidly retract and centralize the protuberant and malpositioned premaxilla and correct the nasolabial and palatal deformities. The orthopedic effects and possible complications were evaluated.

PATIENTS AND METHODS: Nine patients with BCCLP who met the inclusion criteria were selected. After the maxillary model was obtained, the new MPNAM device with retraction screw was designed and worn until cheilorrhaphy. Changes in local deformities and complications were observed continuously, and the orthopedic effect was evaluated.

RESULTS: All patients quickly adapted to the MPNAM appliance, and the treatment was finished after 5-8 return visits. The columella was significantly prolonged, the nasal tip was elevated, and the collapsed nasal dome was obviously improved. Simultaneously, the premaxilla was rapidly retracted and rotated, and gradually centralized; the clefts were gradually reduced and closed, and a nearly normal dental arch was formed. Although there were some complications, the orthopedic treatment was continued until cheiloplasty.

CONCLUSIONS: The MPNAM device with retraction screw can simultaneously correct nasolabial and palatal deformities and also rapidly retract and centralize the premaxilla. Copyright 2014 European Association for Cranio-Maxillo-Facial Surgery. Published by Elsevier Ltd. All rights reserved.

Publication type: Journal Article
Source: MEDLINE

23. Title: Esthetic evaluation of the facial profile in rehabilitated adults with complete bilateral cleft lip and palate.

Citation: Journal of Oral & Maxillofacial Surgery, January 2015, vol./is. 73/1(169.e1-6), 0278-2391;1531-5053 (2015 Jan)

Author(s): Ferrari Junior FM, Ayub PV, Capelozza Filho L, Pereira Lauris JR, Garib DG

Language: English

Abstract: PURPOSE: To assess the facial esthetics of patients with complete bilateral cleft lip and palate, and to compare the judgment of raters related and unrelated to cleft care.

MATERIALS AND METHODS: The sample comprised 23 adult patients (7 women and 16 men) with a mean age of 26.1 years, rehabilitated at a single center. Standardized photographs of the right and left facial profile were taken of each patient and subjectively evaluated by 25 examiners: 5 orthodontists and 5 plastic surgeons with expertise in oral cleft rehabilitation, 5 orthodontists and 5 plastic surgeons without expertise in oral cleft rehabilitation, and 5 laypersons. The facial profiles were classified into 3 categories: esthetically unpleasant, esthetically acceptable, and esthetically pleasant. Intraexaminer and interexaminer agreements were evaluated with the Spearman correlation coefficient and Kendall coefficient of concordance. The differences between rater categories were analyzed using the Student-Newman-Keuls test (with P < .05 indicating a statistically significant difference).

RESULTS: Most of the sample was classified as esthetically acceptable. Orthodontists and plastic surgeons related to oral cleft rehabilitation gave the best scores to the facial profiles, followed by layperson examiners and by orthodontists and plastic surgeons unrelated to oral cleft
rehabilitation. The middle third of the face, the nose, and the upper lip were frequently pointed out as contributors
to the esthetic impairment.CONCLUSIONS: The facial profile of rehabilitated adult patients with complete bilateral
cleft lip and palate was considered esthetically acceptable because of morphologic limitations in the structures
affected by the cleft. Laypersons and professionals unrelated to oral cleft rehabilitation seem to be more critical
regarding facial esthetics than professionals involved with cleft rehabilitation. Copyright 2015. Published by Elsevier
Inc.

Publication type: Journal Article
Source: MEDLINE

24. Title: Functional and aesthetic outcome after surgery on cleft lip nasal deformities [German] Funktionelle und
asthetische Ergebnisse nach Spaltinasenoperationen
Citation: Laryngo- Rhino- Otologie, 2014, vol./is. 93/11(778-784), 0935-8943;1438-8685 (2014)
Author(s): Scheithauer M., Veit J., Rotter N., Sommer F., Lindemann J., Hoffmann T.K., Nordmann M.
Language: German
Abstract: Objective: Patients with cleft lip nasal deformities suffer not only from aesthetic stigmatization but also
from functional problems. Many different techniques to operate on this deformity have been described. Methods:
24 patients (16 male and 8 female, aged 20-56 years) have been enrolled in this study. A subjective value of the
postoperative result of aesthetics and function was measured by a visual analogue scale ranging from -5 (significant
aggravation) to +5 (significant improvement) including 0 (no change). Additionally the primary nasal symptoms (PNS)
of the SNOT-20 GAV were collected. Results: 10 primary surgeries and 14 revisions were performed with 20 open and
4 endonasal approaches. In 11 cases a composite graft from the concha was used. 75% of the patients looked up to
both features aesthetic and function favorably. The mean evaluation was +3.4 for aesthetics and +2.6 for function.
91% of the patients who received a composite graft looked up to the result favorably. The mean PNS were
14. Conclusions: Altogether good results especially concerning the subjective aesthetics can be achieved by precise
preoperative evaluation and the described operative algorithm. In our sample of patients better results could be
accomplished by using composite grafts. Nevertheless it seems to be more difficult to improve the function of cleft
lip nasal deformities than the aesthetics. More innovations and objective measurements are needed to come up to
the still unsolved problems of the challenging surgery of cleft lip nasal deformities.

Publication type: Journal: Article
Source: EMBASE

25. Title: Incidence of palatal fistula formation after primary palatoplasty in northern Finland.
Citation: Oral Surgery, Oral Medicine, Oral Pathology and Oral Radiology, December 2014, vol./is. 118/6(632-6),
2212-4411 (2014 Dec)
Author(s): Lithovius RH, Ylikontiola LP, Sandor GK
Language: English
Abstract: OBJECTIVE: The purpose of this retrospective study was to determine the incidence of palatal fistulas after
primary cleft palate repair. STUDY DESIGN: The study included 136 patients who were treated at the Oulu University
Hospital cleft lip and palate center between 1998 and 2011. All patients were treated by the same surgeons with 1-
stage palatoplasty closing the hard and soft palate concurrently. RESULTS: The overall frequency of postoperative
fistula was 9.6% of patients. Patients with cleft lip and palate (20.0%) were more likely to develop postoperative
palatal fistulas than patients with cleft palate (6.6%). Surgical technique and cleft severity were not significant
factors for the development of palatal fistulas. CONCLUSIONS: The majority of patients undergoing primary palatal
repair do not develop palatal fistulas. Copyright 2014 Elsevier Inc. All rights reserved.

Publication type: Journal Article
Source: MEDLINE

26. Title: Incidence of positive screening for obstructive sleep apnea in patients with isolated cleft lip and/or
palate.
Citation: Canadian Journal of Plastic Surgery, 2014, vol./is. 22/4(259-63), 1195-2199;1195-2199 (2014)
Author(s): Silvestre J, Tahiri Y, Paliga JT, Taylor JA
Language: English
Abstract: OBJECTIVE: To determine the incidence of obstructive sleep apnea (OSA) in children with isolated cleft lip
and/or palate (CL/P). METHODS: The present prospective study was performed at a pediatric tertiary care centre.
Consecutive patients evaluated at the cleft clinic from January 2011 to August 2013 were identified. Patients’
families prospectively completed the Pediatric Sleep Questionnaire (PSQ), a validated tool used to predict moderate
to severe OSA. Patients with CL/P and an underlying syndrome or other craniofacial diagnosis were excluded. A
positive OSA screen was recorded if the ratio of positive to total responses was >0.33. Risk factors associated with a positive screen were identified using the Student's t or ANOVA test.RESULTS: A total of 867 patients completed the PSQ, 489 of whom with isolated CL/P met inclusion criteria. The mean age was 8.4 years. The overall incidence of positive screening was 14.7%. The most commonly reported symptoms among positive screeners were 'fidgets with hands or feet' (73.6%), 'interrupts others' (69.4%) and 'mouth breather during the day' (69.4%). The most sensitive items were 'stops breathing during the night' and 'trouble breathing during sleep', with positive predictive values of 0.78 and 0.67, respectively. Sex, body mass index, ancestry and cleft type were not significantly associated with increased risk for positive screening.CONCLUSION: One in seven children with isolated CL/P screened positively for OSA according to the PSQ. This finding highlights the potential importance of routine screening in this at-risk group. Publisher: Abstract available from the publisher. Language: French

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

**Full text:** Available National Library of Medicine at [Canadian Journal of Plastic Surgery, The](#)

**27. Title:** Influence of different palate repair protocols on facial growth in unilateral complete cleft lip and palate.  
**Citation:** Journal of Cranio-Maxillo-Facial Surgery, January 2015, vol./is. 43/1(43-7), 1010-5182;1878-4119 (2015 Jan)  
**Author(s):** Xu X, Kwon HJ, Shi B, Zheng Q, Yin H, Li C  
**Language:** English

**Abstract:** OBJECTIVE: To address the question of whether one- or two-stage palatal treatment protocol has fewer detrimental effects on craniofacial growth in patients aged 5 years with unilateral complete cleft lip and palate.MATERIALS AND METHODS: Forty patients with non-syndromic unilateral complete cleft lip and palate (UCCLPs) who had received primary cleft lip repair at age 6-12 months and cleft palate repair at age 18-30 months were selected in this study. Eighteen UCCLP patients who received two-stage palate repair were selected as group 1, and 22 UCCLP patients who received one-stage palate repair were selected as group 2. The control group consisted of 20 patients with unilateral incomplete cleft lip (UICL patients) whose age and gender matched with UCCLP patients. A one-sample Kolmogorov-Smirnov test was used to analyze the nature of data distribution. Bonferroni test and Kruskal-Wallis H tests were used for multiple comparisons.RESULTS: Both case groups showed reduced maxillary sagittal length (ANS-PMP, A-PM, p < 0.05) and retraction of the maxilla (S-Ptm, p < 0.05), A point and ANS point (Ba-N-A, Ba-N-ANS, p < 0.05). Patients treated with two-stage palate repair had a reduced posterior maxillary vertical height (R-PMP, p < 0.05).CONCLUSIONS: Our results indicated that maxillary sagittal length and position could be perturbed by both one- and two-stage palate repair. Vomer flap repair inhibited maxilla vertical growth. Delayed hard palate repair showed less detrimental effects on maxillary growth compared to early hard palate repair in UCCLP patients aged 5 years. Copyright 2014 European Association for Cranio-Maxillo-Facial Surgery. Published by Elsevier Ltd. All rights reserved.  
**Publication type:** Journal Article  
**Source:** MEDLINE

**28. Title:** Intellectual disability secondary to a 16p13 duplication in a 1;16 translocation. Extended phenotype in a four-generation family.  
**Citation:** American Journal of Medical Genetics. Part A, January 2015, vol./is. 167/1(128-36), 1552-4825;1552-4833 (2015 Jan)  
**Author(s):** Mohamed AM, Kamel A, Mahmoud W, Abdelraouf E, Meguid N  
**Language:** English

**Abstract:** We describe a large family from the Gaza Strip presented with multiple congenital anomalies. The proband was presented with intellectual disability and multiple congenital anomalies including cleft palate, low-set ears, everted upper lip, diaphragmatic hernia, and arthrogryposis. Pedigree analysis showed 19 affected patients over five generations, only 6 were alive and 11 individuals were obligate carriers. The proband had an apparently normal karyotype, although FISH studies showed a derivative chromosome 1 with duplication of 16p13.3 and deletion of the 1p subtelomere. Her father however had a balanced translocation. The seven affected patients had a similar phenotype, one of them died before genetic testing was carried out and the living six patients had the same unbalanced translocation. Array CGH revealed an 8.8Mb duplication in 16p13 and 200,338bp deletion in 1p36.3. Accordingly, intellectual disability, hypertelorism, cupped ears, everted upper lip, and limb anomalies were presenting clinical features of the 16p13 duplication syndrome while deep set eyes were perhaps related to the 1p terminal deletion. Prevention of recurrent intellectual disability in this family can be achieved through carrier detection and prenatal genetic diagnosis. 2014 Wiley Periodicals, Inc. 2014 Wiley Periodicals, Inc.  
**Publication type:** Journal Article
29. Title: Is a polymorphism in 10q25 associated with non-syndromic cleft lip with or without cleft palate? A meta-analysis based on limited evidence.

Citation: British Journal of Oral & Maxillofacial Surgery, January 2015, vol./is. 53/1(8-12), 0266-4356;1532-1940 (2015 Jan)

Author(s): Li C, Li Z, Zeng X, Guo Z

Language: English

Abstract: Non-syndromic cleft lip with or without cleft palate (NSCL/P) is one of the most common defects in the human facial structure. A polymorphism, rs7078160 in 10q25, has been reported to be involved in susceptibility to the condition but results remain conflicting. To address this we did a meta-analysis of 7 eligible studies. Two authors independently collected information from the papers, and fixed effects or random effects models were used to calculate the combined estimates of risk. The pooled results showed that rs7078160 on 10q25, the minor allele A, had a higher risk of NSCLP than the major allele G (OR: 1.32, 95% CI: 1.18 to 1.47). Overall, the results showed that the 10q25 rs7078160 polymorphism was associated with a risk of NSCL/P. Copyright 2014 The British Association of Oral and Maxillofacial Surgeons. Published by Elsevier Ltd. All rights reserved.

Publication type: Journal Article

Source: MEDLINE

30. Title: Isolated keratinized gingiva incision in alveolar cleft bone grafts improves qualitative outcomes: A single surgeon's 23 year experience.

Citation: Journal of Cranio-Maxillo-Facial Surgery, December 2014, vol./is. 42/8(1692-7), 1010-5182;1878-4119 (2014 Dec)

Author(s): Lopez-Cedrun JL, Gonzalez-Landa G, Figueroa A

Language: English

Abstract: BACKGROUND: Few publications have described the flap design of the secondary cleft alveoloplasty. In this article we describe a modified technique of the classical flap design with the purpose of minimizing injury to the dental papillae and periodontium of the involved dentition. We report our long-term experience, specifically with regards to oronasal fistulae recurrence, wound healing and graft exposure and loss. METHODS: All the patients were operated on using the same technique by a single surgeon. A total of 148 clefts have been operated with this approach, involving 117 patients with complete cleft lip and palate with a follow-up between 12 and 240 months. RESULTS: The most important finding in this study is the excellent wound healing observed in almost all patients. Only three patients (2%) suffered a dehiscence with oronasal fistulae recurrence and bone loss. Another patient lost the graft without fistula recurrence. Minor dehiscence with partial bone loss occurred in 4 patients (2.7%). These patients did not need surgical closure and only superficial exposed bone particles were lost without compromising the clinical outcome. CONCLUSIONS: Our modification presents a flap design that is easy to elevate and mobilize, without disturbing the buccal sulcus or the gingival inter-dental papillae. Copyright 2014 European Association for Cranio-Maxillo-Facial Surgery. Published by Elsevier Ltd. All rights reserved.

Publication type: Journal Article

Source: MEDLINE

31. Title: Lateral incisor agenesis predicts maxillary hypoplasia and Le Fort I advancement surgery in cleft patients.

Citation: Plastic & Reconstructive Surgery, January 2015, vol./is. 135/1(142e-8e), 0007-1226;1529-4242 (2015 Jan)

Author(s): Lai LH, Hui BK, Nguyen PD, Yee KS, Martz MG, Bradley JP, Lee JC

Language: English

Abstract: BACKGROUND: Severe maxillary hypoplasia in cleft patients is caused by a combination of pathogenic and iatrogenic factors. In this work, the authors investigated anatomical deficiencies in dentition for predicting Le Fort I maxillary advancement surgery for severe maxillary hypoplasia in cleft patients. METHODS: Cleft lip-cleft palate and cleft palate patients older than 14 years of age were reviewed for demographics, dental anomalies, and Le Fort I advancement. Chi-square tests, t tests, and multivariate logistic regression analyses were performed to delineate the contribution of quantity and position of dental agenesis to maxillary advancement surgery. RESULTS: In the 114 patients reviewed (mean age, 19.2 years), 64.0 percent were male patients, 71.9 percent had dental agenesis, and 59.6 percent required Le Fort I advancement. In patients who did not exhibit dental agenesis, 18.8 percent required Le Fort I advancement compared with 74.4 percent of patients with dental agenesis (p < 0.0001). Le Fort I advancement surgery was increased to 76.3 percent when dental agenesis was at the lateral incisor position (p < 0.0001) and 86.4 percent when patients were missing two or more teeth (p < 0.0001). Both sella-to-nasion-to-A point angle (p = 0.003) and A point-to-nasion-to-B point angle (p = 0.04) measurements were decreased in patients...
missing dentition at the lateral incisor position. Adjusting for multiple missing teeth and orthodontic compensations, multivariate logistic regression analyses demonstrated that lateral incisor agenesis is an independent predictor for Le Fort I advancement surgery (OR, 4.4; 95 percent CI, 1.42 to 13.64; p = 0.01). CONCLUSIONS: Lateral incisor agenesis correlated to maxillary hypoplasia and independently predicted the need for Le Fort I advancement in cleft patients, potentially as an anatomical readout of intrinsic growth deficiency. CLINICAL QUESTION/LEVEL OF EVIDENCE: Risk, III.

**Publication type:** Journal Article

**Source:** MEDLINE

**Full text:** Available *Journal of reconstructive microsurgery* at Plastic and Reconstructive Surgery

**Full text:** Available *Journal of reconstructive microsurgery* at Plastic and Reconstructive Surgery

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32. **Title:** Long-term follow-up of large maxillary advancements with distraction osteogenesis in growing and non-growing cleft lip and palate patients.

**Citation:** Journal of Plastic, Reconstructive & Aesthetic Surgery: JPRAS, January 2015, vol./is. 68/1(79-86), 1748-6815;1878-0539 (2015 Jan)

**Author(s):** Meazzini MC, Basile V, Mazzoleni F, Bozzetti A, Brusati R

**Language:** English

**Abstract:** BACKGROUND: Maxillary distraction osteogenesis (DO) in cleft lip and palate patients has been described by several authors, but most studies have a relatively short follow-up and do not clearly separate growing patients from non-growing patients. METHOD: The records of 22 consecutive patients affected by cleft lip and palate, who underwent Le Fort I osteotomy and maxillary distraction with a rigid external distractor (RED), were reviewed. The sample was subdivided into a growing and a non-growing group. All patients had pre-DO cephalometric records, immediately post DO, 12 months post DO and long-term records with a long-term follow-up of >5 years (range 5-13 years). As a control sample for the growing group, cleft children with a negative overjet not subjected to distraction or any protraction treatment during growth were followed up until the completion of growth. RESULTS: The average maxillary advancement in the growing group was 22.2 ± 5.5 mm (range: 15-32 mm); in the non-growing group, it was 17.7 ± 6.6 mm (range: 6-25 mm). Excellent post-surgical stability was recorded in the adult sample. On the other hand, growing children had an average 16% relapse in the first year post DO and an additional 26% relapse in the long-term follow-up. CONCLUSIONS: This study seems to point out that early Le Fort I DO allows for the correction of very severe deformities. It is followed by a relatively high amount of true skeletal relapse in children with cleft lip and palate. Prognosis should be discussed in depth with the family and true aesthetic and psychological needs assessed. Copyright 2014 British Association of Plastic, Reconstructive and Aesthetic Surgeons. Published by Elsevier Ltd. All rights reserved.

**Publication type:** Journal Article

**Source:** MEDLINE

**Full text:** Available *JOURNAL OF PLASTIC, RECONSTRUCTIVE & AESTHETIC SURGERY (formerly BRITISH JOURNAL OF PLASTIC SURGERY)* at Journal of Plastic, Reconstructive and Aesthetic Surgery

**Full text:** Available *JOURNAL OF PLASTIC, RECONSTRUCTIVE & AESTHETIC SURGERY (formerly BRITISH JOURNAL OF PLASTIC SURGERY)* at Salisbury District Hospital Healthcare Library

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33. **Title:** Loss of Tbx1 induces bone phenotypes similar to cleidocranial dysplasia.

**Citation:** Human Molecular Genetics, January 2015, vol./is. 24/2(424-35), 0964-6906;1460-2083 (2015 Jan 15)

**Author(s):** Funato N, Nakamura M, Richardson JA, Srivastava D, Yanagisawa H

**Language:** English

**Abstract:** T-box transcription factor, TBX1, is the major candidate gene for 22q11.2 deletion syndrome (DiGeorge/Velo-cardio-facial syndrome) characterized by facial defects, thymus hypoplasia, cardiovascular anomalies and cleft palates. Here, we report that the loss of Tbx1 in mouse (Tbx1(-/-)) results in skeletal abnormalities similar to those of cleidocranial dysplasia (CCD) in humans, which is an autosomal-dominant skeletal disease caused by mutations in RUNX2. Tbx1(-/-) mice display short stature, absence of hyoid bone, failed closure of fontanelle, bifid xiphoid process and hypoplasia of clavicle and zygomatic arch. A cell-type-specific deletion of Tbx1 in osteochondro-progenitor (Tbx1(OPKO)) or mesodermal (Tbx1(MKO)) lineage partially recapitulates the Tbx1(-/-) bone phenotypes. Although Tbx1 expression has not been previously reported in neural crest, inactivation of Tbx1 in the neural crest lineage (Tbx1(NCKO)) leads to an absence of the body of hyoid bone and postnatal lethality, indicating an unanticipated role of Tbx1 in neural crest development. Indeed, Tbx1 is expressed in the neural crest-derived hyoid bone primordium, in addition to mesoderm-derived osteochondral progenitors. Ablation of Tbx1 affected Runx2 expression in calvarial bones and overexpression of Tbx1 induced Runx2 expression in vitro. Taken together, our current studies reveal that Tbx1 is required for mesoderm- and neural crest-derived osteoblast differentiation and normal skeletal development. TBX1 mutation could lead to CCD-like bone phenotypes in human. The Author 2014. Published by
34. Title: Median cleft of the upper lip associated with a mass: A rare case.
Citation: Journal of Cranio-Maxillo-Facial Surgery, December 2014, vol./is. 42/8(1557-61), 1010-5182;1878-4119 (2014 Dec)
Author(s): Jian XC, Zheng L, Xu P, Liu DY
Language: English
Abstract: Median cleft lip is a midline vertical cleft through the upper lip. This is a very rare anomaly described in the literature. Median cleft lip is caused by the failure of fusion of the medial nasal prominences. In this case report, a 4-month-old boy with a median cleft associated with a mass of the upper lip is presented. The patient has no other anomalies of the nose or alveolus. The patient has normotelorism. A Z-plasty technique was used on the skin of the base of the columella. A vertical excision of the cleft with muscle approximation was performed on the white roll and the wet-dry border of either side of the defect of the upper lip. Postoperatively, the patient had a satisfactory result. The incisive scars were not visible. Cupid's bow was appropriately aligned, and the height of the upper lip was equal on both sides. Copyright 2014. Published by Elsevier Ltd.

35. Title: Occipital meningoencephalocele with cleft lip, cleft palate and limb abnormalities- A case report
Citation: Journal of Clinical and Diagnostic Research, December 2014, vol./is. 8/12(AD03), 2249-782X;0973-709X (05 Dec 2014)
Author(s): Ganapathy A., Sadeesh T., Hydrinaswer M., Rao S.
Language: English
Abstract: A 21-week-old still born female fetus with occipital encephalocele, cleft lip and cleft palate was received from the Department of Obstetrics and Gynecology, Mahatma Gandhi Medical College and Research Institute, Pondicherry and was studied in detail. It was born to Primigravida, of a second degree consanguineous marriage, with unremarkable family history. The biometric measurements were noted which corresponded to the age of the fetus. Further the fetus was embalmed and dissected. On examination an encephalocele of 2.7x1.5 cm was seen in the occipital region with a midline defect in the occipital bone and herniated brain tissue. Other anomalies observed were right unilateral cleft lip, right cleft palate, and bilateral syndactyly of the lower limbs and associated Congenital Talipes Equino Varus of the right foot. Other internal organs were developed appropriate for the age of the fetus.

36. Title: Oculoauriculovertebral spectrum with a full range of severe clinical manifestations - Case report.
Citation: Journal of Cranio-Maxillo-Facial Surgery, December 2014, vol./is. 42/8(2064-8), 1010-5182;1878-4119 (2014 Dec)
Author(s): Gawrych E, Janiszewska-Olszowska J, Chojnacka H
Language: English
Abstract: Oculoauriculovertebral spectrum (OAVS) is a rare congenital malformation consisting of epibulbar dermoids, lid colobomas, auricular deformities, hypoplasia of the soft and bony tissues of the face, associated oral deformities and vertebral anomalies. This report presents a child with a choroid of the right eye, coloboma of the upper eyelid, epibulbar dermoid of the left eye, mandibular hypoplasia, facial asymmetry, bilateral complete cleft lip and palate, hypoplasia of the left alar cartilage, appendage of the left nose, butterfly vertebral defects of Th-1, Th-2 and abnormality of rib XI. Widened sulci of frontal and parietal lobes, bilateral white matter density decrease and calcifications of falx cerebelli were noted. Axial hypotony and delayed psycho-motor development were apparent. This rare case presents a range of severe clinical manifestations of oculoauriculovertebral spectrum. Despite a normal cervical vertebral column, tracheostenosis was present. It caused difficulties in tracheal intubation, creating the need for a tracheostomy, and death after a failed attempt at decannulation. This case indicates that in patients with clinical manifestations including cerebral anomalies, a risk of respiratory insufficiency should be always taken under consideration, when planning surgery. Copyright 2011. Published by Elsevier Ltd.

37. Title: Recurrence rate of repaired hard palate oronasal fistula with conchal cartilage graft
EMBASE

Citation: Journal of Research in Medical Sciences, 2014, vol./is. 19/10(956-960), 1735-1995;1735-7136 (2014)
Author(s): Abdali H., Hadilou M., Feizi A., Omranifard M., Ardakani M.R., Emami A.
Language: English
Abstract: Background: After cleft palate repair, oronasal fistula (ONF) formation is one of the considerable and troublesome complications. Conchal cartilage graft is one option that can be used in recurrent fistula correction. The aim of the current study is investigating the recurrence rate of the hard palate ONF or ONF at the junction of hard and soft palate after utilizing conchal cartilage graft and comparing this rate with other methods.
Methods: In this observational prospective study, 29 patients suffering from ONF with small, medium and large sizes who were referring to Alzahra university hospital, Isfahan, Iran and Fateme Zahra university hospital, Tehran, Iran between November 2011 and November 2012 were enrolled. All patients had midline cleft palate, 29.6% of them had cleft lip too that was repaired previously. All patients were followed-up for 2 years (every 2 months) after repair.
Results: The mean (range) age of studied samples was 10.7 (2-23) years. 16 patients (55.7%) were female, and reminders were male. During 2 years followup, we detected recurrence of ONF in 6 patients (20.68%) and the success rate was 79.32%. The recurrence rate, after applying the current approach, among who experienced the several times of recurrence was significantly higher than among those who experienced first time of recurrence (33.3% vs. 7.1%; P < 0.001). The mean [+SD] age of failed and successfully repaired patients were 11.3 (+4.5) and 8.4 (+5.25) years, respectively (P > 0.1).
Conclusion: Using of conchal cartilage graft for recurrent ONF with <1 cm was safe and efficacious, in ONF >1 cm conchal cartilage graft can be used as a primary method and if recurrence occurred chooses other complex procedure.
Publication type: Journal: Article
Source: EMBASE
Full text: Available ProQuest at Journal of Research in Medical Sciences

38. Title: Regional brain morphometric characteristics of nonsyndromic cleft lip and palate
Citation: Developmental Neuroscience, November 2014, vol./is. 36/6(490-498), 0378-5866;1421-9859 (19 Nov 2014)
Author(s): Adamson C.L., Anderson V.A., Nopoulos P., Seal M.L., Da Costa A.C.
Language: English
Abstract: Nonsyndromic cleft lip and palate (NSCLP) encompasses a group of orofacial abnormalities. Emerging evidence has revealed the presence of structural brain abnormalities in affected individuals. Previous studies have performed structure-based volumetric analysis of the brain assessing gross lobular subdivisions of the cerebral cortex and white matter which may have only vague relationships to the functional subregions implicated in behavioral and cognitive deficits observed in NSCLP patients. High-resolution magnetic resonance imaging structural data were acquired to provide a detailed characterization of the brain with respect to both regional cortical volume and thickness in 26 children with NSCLP and 26 age- and demographically matched typically developing children. Children with NSCLP exhibited abnormally large cerebral cortex grey matter volumes with decreased volumes of subcortical grey matter and cerebral white matter structures. Hemisphere-specific patterns of cortical volume and thickness abnormalities were identified. This study is the first to examine cortical thickness abnormalities in NSCLP. Overall, these findings suggest that the brains of children with NSCLP are less mature than those of their age-matched peers. Gender-specific comparisons reveal that NSCLP females were more immature compared to their typically developing peers compared to NSCLP males.
Publication type: Journal: Article
Source: EMBASE

39. Title: Reliable critical sized defect rodent model for cleft palate research.
Citation: Journal of Cranio-Maxillo-Facial Surgery, December 2014, vol./is. 42/8(1840-6), 1010-5182;1878-4119 (2014 Dec)
Author(s): Mostafa NZ, Doschak MR, Major PW, Talwar R
Language: English
Abstract: BACKGROUND: Suitable animal models are necessary to test the efficacy of new bone grafting therapies in cleft palate surgery. Rodent models of cleft palate are available but have limitations. This study compared and modified mid-palate cleft (MPC) and alveolar cleft (AC) models to determine the most reliable and reproducible model for bone grafting studies. METHODS: Published MPC model (9 x 5 x 3 mm(3)) lacked sufficient information for tested rats. Our initial studies utilizing AC model (7 x 4 x 3 mm(3)) in 8 and 16 weeks old Sprague Dawley (SD) rats revealed injury to adjacent structures. After comparing anteroposterior and transverse maxillary dimensions in 16 weeks old SD and Wistar rats, virtual planning was performed to modify MPC and AC defects dimensions, taking the adjacent structures into consideration. Modified MPC (7 x 2.5 x 1 mm(3)) and AC (5 x 2.5 x 1 mm(3)) defects were employed in 16 weeks old Wistar rats and healing was monitored by micro-computed tomography and
INTRODUCTION: Cleft palate is a relatively common deformity with various techniques described for its repair. Most techniques address the hard palate portion of the cleft with bilateral mucoperiosteal flaps transposed to the midline. This results in superimposed, linear closure layers directly over the cleft and may predispose the repair to oronasal fistula formation. This report details an alternative technique of flap rotation with an outcome analysis.

METHODS: A retrospective chart analysis was performed of all patients having undergone primary palatoplasty for cleft palate. Demographics and cleft Veau type were recorded. Postoperative speech outcomes were

RESULTS: Maxillary dimensions in SD and Wistar rats were not significantly different. Preoperative virtual planning enhanced postoperative surgical outcomes. Bone healing occurred at defect margin leaving central bone void confirming the critical size nature of the modified MPC and AC defects.

CONCLUSIONS: Presented modifications for MPC and AC models created clinically relevant and reproducible defects.

Publication type: Journal Article
Source: MEDLINE

40. Title: Role of OVCA1/DPH1 in craniofacial abnormalities of Miller-Dieker syndrome
Citation: Human Molecular Genetics, November 2014, vol./is. 23/21(5579-5596), 0964-6906;1460-2083 (01 Nov 2014)
Author(s): Yu Y.-R., You L.-R., Yan Y.-T., Chen C.-M.
Language: English
Abstract: OVCA1/DPH1 (OVCA1) encodes a component of the diphthamide biosynthesis pathway and is located on chromosome 17p13.3. Deletions in this region are associated with Miller-Dieker syndrome (MDS). Ovca1/Dph1 (Ovca1)-null mice exhibit multiple developmental defects, including cleft palate, growth restriction and perinatal lethality, suggesting a role in the craniofacial abnormalities associated with MDS. Conditional ablation of Ovca1 in neural crest cells, but not in cranial paraxial mesoderm, also results in cleft palate and shortened lower jaw phenotypes, similar to Ovca1-null embryos. Expression of transgenic myc-tagged Ovca1 in craniofacial structures can partially rescue the cleft palate and shortened mandible of Ovca1-null embryos. Interestingly, Ovca1-null mutants are resistant to conditional expression of diphtheria toxin subunit A in both neural crest cell and paraxial mesoderm derivatives. However, Ovca1-dependent diphthamide biosynthesis is essential for neural crest cell- and paraxial mesoderm-derived craniofacial development but is dispensable for paraxial mesoderm-derived craniofacial structures in mammals. These findings suggest that Ovca1 deficiency in the neural crest contributes to the craniofacial abnormalities in patients with MDS. Also, our findings provide new insights into the molecular and cellular mechanisms that lead to the craniofacial defects of MDS.

Publication type: Journal: Article
Source: EMBASE

41. Title: Strain-dependent effects of transforming growth factor-beta1 and 2 during mouse secondary palate development
Citation: Reproductive Toxicology, January 2015, vol./is. 50/(129-133), 0890-6238;1873-1708 (January 01, 2015)
Author(s): Jin J.-Z., Ding J.
Language: English
Abstract: Cleft palate is a common birth defect affecting 1 in 700 births. Transforming growth factor-betas (TGF-betas) are important signaling molecules, and their functions in murine palate development have received great attention. TGF-beta3 is expressed exclusively in palatal epithelial cells and mediates epithelial fusion, whereas the importance of TGF-beta1 and 2 in palate have not yet been demonstrated in vivo, since inactivation of Tgf-beta1 or Tgf-beta2 genes in mice did not reveal significant palate defects. We hypothesized that TGF-beta1 and TGF-beta2 can compensate each other during palate formation. To test this, we generated Tgf-beta1 and Tgf-beta2 compound mutant mice and found that approximately 40% of [Tgf-beta1<sup>+/-</sup>; Tgf-beta2<sup>+/-</sup>] compound mutant embryos display cleft palate on C57 background. In addition, 26% of Tgf-beta2<sup>+/-</sup> embryos on 129 background, but not in C57 or Black Swiss, displayed cleft palate. TGF-beta1 and 2 functions are required for murine palate development in strain-dependent manner.

Publication type: Journal: Article
Source: EMBASE

42. Title: Transverse mucoperiosteal flap inset by rotation for cleft palate repair: technique and outcomes.
Citation: Annals of Plastic Surgery, 2014, vol./is. 72/6(S90-3), 0148-7043;1536-3708 (2014)
Author(s): Black JS, Gampper TJ
Language: English
Abstract: INTRODUCTION: Cleft palate is a relatively common deformity with various techniques described for its repair. Most techniques address the hard palate portion of the cleft with bilateral mucoperiosteal flaps transposed to the midline. This results in superimposed, linear closure layers directly over the cleft and may predispose the repair to oronasal fistula formation. This report details an alternative technique of flap rotation with an outcome analysis.

METHODS: A retrospective chart analysis was performed of all patients having undergone primary palatoplasty for cleft palate. Demographics and cleft Veau type were recorded. Postoperative speech outcomes were
assessed by standardized speech evaluation performed by 2 speech language pathologists. The presence and location of oronasal fistulae was assessed and recorded by the surgeon and speech language pathologists in follow-up evaluations. RESULTS: The study revealed an overall incidence of velopharyngeal insufficiency of 5.7% using this surgical technique. It also revealed a fistula rate of 8.6%. Secondary surgery has been successful in those patients in which it was indicated. Eleven (31%) patients were diagnosed with Robin sequence. CONCLUSIONS: This technique demonstrates excellent early outcomes in a difficult subset of cleft patients including a high proportion of those with Pierre Robin sequence. The technique addresses the inherent disadvantages to a linear closure over the bony cleft. The variability in its design provides the surgeon another option for correction of this deformity.

**Publication type:** Journal Article

**Source:** MEDLINE

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

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43. **Title:** Unmet burden of cleft lip and palate in rural gujarat, India: a population-based study.

**Citation:** World Journal of Surgery, January 2015, vol./is. 39/1(41-6), 0364-2313;1432-2323 (2015 Jan)

**Author(s):** Khajanchi MU, Shah H, Thakkar P, Gerdin M, Roy N

**Language:** English

**Abstract:** BACKGROUND: The burden of cleft lip and palate (CLP) in the developing world is being tackled by local hospitals and international surgical missions. However, the unmet surgical burden of these conditions is not known, because there are few population-based studies. We conducted this study to find the incidence and prevalence of cleft lip (CL), cleft palate (CP), and CLP and also estimate the unmet burden of these conditions. METHODS: Four blocks comprising of half a million people in the Patan district of Gujarat were chosen as the study areas. This study was conducted over a period of 3 months in 2009. Patients with CL, CP, and CLP were identified by community health workers using snowball sampling method. Data collected included demographics, type of cleft, operated or not, and place of operation. Disability adjusted life years (DALY) was calculated to measure the unmet burden of this disease. RESULTS: The most common among the three conditions was CL (69.4%). Overall, cleft abnormalities were more common in males (61%). The overall incidence and prevalence of cleft deformity was 0.73 per 1,000 live births and 0.1 per 1,000 people respectively. The unmet burden of surgical disease of these four blocks was 230 to 494 DALYs. CONCLUSIONS: The incidence of CL with or without palate was found to be 0.7 per 1,000 live births. The large number of unoperated cases (backlog) of cleft deformities suggests a big burden of unmet need in rural India.

**Publication type:** Journal Article

**Source:** MEDLINE

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44. **Title:** Whole exome sequencing identifies a POLR1D mutation segregating in a father and two daughters with findings of Klippel-Feil and Treacher Collins syndromes.

**Citation:** American Journal of Medical Genetics. Part A, January 2015, vol./is. 167/1(95-102), 1552-4825;1552-4833 (2015 Jan)


**Language:** English

**Abstract:** We report on a father and his two daughters diagnosed with Klippel-Feil syndrome (KFS) but with craniofacial differences (zygomatic and mandibular hypoplasia and cleft palate) and external ear abnormalities suggestive of Treacher Collins syndrome (TCS). The diagnosis of KFS was favored, given that the neck anomalies were the predominant manifestations, and that the diagnosis predated later recognition of the association between spinal segmentation abnormalities and TCS. Genetic heterogeneity and the rarity of large families with KFS have limited the ability to identify mutations by traditional methods. Whole exome sequencing identified a nonsynonymous mutation in POLR1D (subunit of RNA polymerase I and II): exon2:c.T332C:p.L111P. Mutations in POLR1D are present in about 5% of individuals diagnosed with TCS. We propose that this mutation is causal in this family, suggesting a pathogenetic link between KFS and TCS. 2014 Wiley Periodicals, Inc. 2014 Wiley Periodicals, Inc.

**Publication type:** Journal Article

**Source:** MEDLINE

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45. **Title:** Will the right Robin patient rise, please? Definitions and criteria during management of Robin sequence patients in the Netherlands and Belgium.

**Citation:** Journal of Cranio-Maxillo-Facial Surgery, January 2015, vol./is. 43/1(92-6), 1010-5182;1878-4119 (2015 Jan)

**Author(s):** Basart H, Kruisinga FH, Breugem CC, Don Griot JP, Hennekam RC, Van der Horst CM

**Language:** English
Abstract: BACKGROUND: Robin Sequence (RS) is characterized by micrognathia and upper airway obstruction (UAO), with or without cleft palate, causing respiratory and feeding problems. Management options are: positioning; nasopharyngeal airway (NPA); tongue-lip adhesion (TLA); mandibular distraction (MDO); and tracheostomy. Controversy exists in literature regarding RS definition and management. Here we describe definitions, management strategies and criteria in opting for management strategies, used by Dutch and Belgian cleft teams. METHODS: A specifically designed questionnaire was sent to members of all 16 Dutch and Belgian cleft teams. RESULTS: 14 cleft teams returned 35 questionnaires. All used micrognathia as definition criterion, 93.4% cleft palate, 51.5% glossoptosis and 45.7% UAO. Six different RS definitions were used; even within a single team >1 definition was used. All teams used different management strategies: all used positioning, 10 NPA, 6 TLA, 7 MDO, 8 tracheostomy, 5 refer patients with invasive treatment indication. Criteria in opting management modalities were: O2-saturation (89.3%), clinical presentation (86.2%), growth and feeding problems (69.0%), polysomnography (62.1%), and differed within teams. CONCLUSION: The Dutch and Belgian cleft teams use variable RS definitions, different management modalities and criteria in choosing management strategies. A single, strict definition and evidence-based management guidelines should be formulated for optimal patient care. Copyright 2014 European Association for Cranio-Maxillo-Facial Surgery. Published by Elsevier Ltd. All rights reserved.

Publication type: Journal Article
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