Review Article

Cleft Lip and Palate: A Review

Nitish Bhat[1], Kalpna Thakur[2], Nandini Bhardwaj[3], Hemwati Nandan[4], Ankit Rawat[5], Anjali Lathwal[6]

[1]Department of Oral Pathology, Indira Gandhi Government Dental College Jammu, J&K India; [2]Department of Oral Pathology and Microbiology, HP Government Dental College and Hospital, Shimla, India; [3]Department of Oral Pathology, Microbiology and Forensic Odontology, HIDS, Poanta Sahib, India; [4]Department of Dentistry, All India Institute of Medical Sciences, Rishikesh, Uttarakhand, India; [5]Prosthodontics and Implantology, Pauri, Uttarakhand, India; [6]Department of Prosthodontics, Crown and Bridge, Sudha Rustagi College of Dental Sciences and Research, Faridabad, India

Corresponding author:
Dr Nitish Bhat, Department of Oral Pathology, Indira Gandhi Government Dental College Jammu, J&K India,
Tel: +6005947413,
E-mail: nitishbhat04@gmail.com

Abstract

Clefts of the lip and palate are the congenital anomalies which may range from a small furrow in the lip to a complete separation of the two maxillary processes. They possess a significant social and financial burden to the individuals and their families. Numerous theories about the etiopathogenesis have been put forward from the past and numerous authors have classified them in different forms for easy understanding. Here in this review we have tried to compile the information from the past literature to present a brief update on various aspects of this entity.

Keywords: Cleft lip; Cleft palate; Orofacial clefts

Introduction

Considered as one of the most common birth defects, clefts of the lip and palate are commonly encountered congenital anomalies that often result in severe functional deficits of speech, mastication and deglutition. [1]They are known to cause significant medical, psychological, social, and financial implications on the affected individuals and families [2] Clefts may be variable in character ranging from furrows in the skin and mucosa to extensive cleavages involving muscle/bone and represent a developmental defect, usually of the upper lip, characterized by a wedge shaped defect resulting from the failure of two parts of the lip to fuse into a single structure. [3]

Cleft palate represents the non-fusion of the palatal shelves and a combination of cleft lip and palate is the most commonly seen clefting deformity. [1] Cleft lip, with or without cleft palate, occurs more frequently than cleft palate alone and is the most common of the significant orofacial anomalies. [4] The frequency of occurrence of cleft lip, with or without cleft palate, has been computed on a global scale and is estimated to be 1 in every 800 new-born babies. [5]Here we present a review on various aspects including the etiopathogenesis, incidence, clinical manifestations, classifications and management of cleft lip and palate.

Literature Review

Etiopathogenesis of cleft lip & palate

Failure of fusion of maxillary & nasal process seems to be the main reason which contributes to clefting. This leads to unilateral or bilateral cleft of the palate & results in clefting of secondary palate because of non-fusion of two palatal shelves. A variety of factors involving genetic, anatomical, teratogens & certain other factors contributes to the complex etiology for clefting.

Anatomical factors

It is postulated in the Tongue Obstruction Hypothesis that during mandibular development anatomical obstruction occurs. [6] There is interposition of the tongue in the space created between the ascending shelves when there is compression of the chin against the sternum. These results in U shaped palatal deficiency & have tissue deformation with a normal growth potential. Ectomesenchyml differences & cellular phenomenon affects this palatal deficiency. [7]

Genetic factors

In cases of cleft lip or cleft palate, a defect in the developing foetus results from many gene locus contributing to its risk which includes SKI/MTHFR 1p36, RARA 17q21, BCL3 19q13, PVRL1 11q23, TGFB3 14q24, TGFB2 1q41, TGFA 2p13, SX1 4p16, 4q31, 6p23 & GABRB3 15q11. [8]These genes interact with each other & with the environment which is explained by "Multifactorial Threshold Hypothesis". This multifactorial inheritance theory determines the degree of abnormality threshold. The risk of occurrence & transmission of isolated cleft lip and palate among family members of infected individual is explained by multifactorial or polygenic inheritance. Occurrence of cleft lip at sixth to seventh week in utero results due to non-penetration of mesodermal cells in the epithelial groove between medial and lateral process which itself fails to form. Maldevelopment of lips and palate results from disrupted normal facial growth patterns including facial processes deficiencies. [1]

Teratogens

There is limited contribution & association of teratogens & clefting. The collaboration of different teratogens acting on a single mechanism is controlled by very less number of genes.

How to Cite this Article: Bhat N, et al. Cleft Lip and Palate: A Review. Ann Med Health Sci Res. 2020;10: 927-930.

Teratogens like retinoic acid which are used in the treatment of acne and psoriasis act through biochemical defect, hormonal defect & extrinsic biochemical interference. These interfere with cell differentiation or migration & have direct effect on morphogenesis of face. [9]

Other factors

Many environmental factors contribute to the pathogenesis of cleft lip and palate. Deficiency or excess of Vitamin A or riboflavin i.e., nutritional factors, enlarged tongue resulting in mechanical obstruction, ischemia, conditions like physiologic, traumatic & emotional stress & infections postulates an accessory role in the pathogenesis of cleft lip and palate. [3]Maternal alcohol consumption & cigarette smoking are also associated with syndromic & non- syndromic clefts. Smoking mothers doubles the frequency of development of cleft as compared to non-smoking mothers. Anticonvulsants like phenytoin increase the risk of cleft formation tenfold. Along with this, folic acid deficiency also contributes to cleft development. [8] The 3.4 times greater risk of Orofacial clefting also results from increased consumption of maternal corticosteroids. The effects of vitamin A deficiency and excess, Riboflavin deficiency & cortisone administration are also seen in experimental animals including rats & rabbits with a resultant exponential increase in cleft palate incidence. Also, non- conclusively proven stress induced hydrocortisone secretion in humans also results in cleft palate formation. [3]

Clinical Manifestations

- Cleft lip with or without palate is more common in males than in females whereas isolated cleft palate is more common in females.
- Babies with cleft lip face difficulty when they try to make a contact between upper and lower lips. [10]
- One of the first manifestations associated with cleft palate is difficulty with feeding. [11]
- Speech is often characterized by air emission from the nose and has a hyper nasal quality. [1]
- Patients with cleft lip usually have cosmetic problems and problems for production of labial sounds. [12]
- Sounds most frequently misarticulated include [11]
- S - (63%)
- Z - (61%)
- D- (48%)
- Ch- (44%)
- P- (11%)
- B- (9%)
- Clefts of the soft palate, including sub mucosal clefts, are often associated with velopharyngeal incompetence or Eustachian tube dysfunction. 1Hearing disorders are prevalent among individuals with orofacial clefts. These disorders are a result of chronic otitis media with effusion due to eustachian tube dysfunction. [11]
- The defect may involve the hard and soft palates or the soft palate alone. The minimal manifestation of Cleft palate is a cleft or bifid uvula. The prevalence of cleft uvula is much higher than that of Cleft Palate, with a frequency of 1 in every 80 white individuals. [8]
- Dental problems: Abnormalities in the position of teeth, delay of eruption of permanent teeth and delay of formation of permanent teeth. [13,14] The incidence of congenitally missing teeth is high, especially among deciduous and permanent maxillary lateral incisors adjacent to the alveolar cleft. The prevalence of hypodontia increases directly with the severity of the cleft. Complete unilateral and bilateral alveolar clefts are often associated with supernumerary teeth as well, usually the maxillary lateral incisors. Tooth formation is often delayed, and enamel hypoplasia, microdontia or macrodontia, and fused teeth are often seen. [1]
- The clefting anterior to the incisive foramen (i.e., Lip and alveolus) is also defined as a cleft of primary palate.
- The clefting posterior to the incisive foramen is defined as a cleft of secondary palate.
- Clefts can be divided into non-syndromic and syndromic forms. [5]
- Syndromic forms of clefts include those cases that have additional birth defects like lip pits or other malformations.
- Non-syndromic clefts are those cases wherein the affected individual has no other physical or developmental anomalies and no recognized maternal environmental exposures.

Classification

There is no universally accepted classification of clefts, although the most commonly used is the Veau's classification, which was described in 1931. [15] Many others classifications by different authors also exist. [Table 1]. [5,15-17]

Discussion and Management

Management of cleft lip and palate includes both non-surgical and surgical procedures with surgical procedures being a definitive treatment for the same. Cleft palate repair is a challenging procedure because of the delicate tissue handling required and the small confines of the infant oral cavity. [18] Management of cleft lip and palate therefore requires a multidisciplinary team collaboration committed to managing the patient from birth to maturity including a number of specialists (reconstructive surgeon, otolaryngologists, orthodontist, Speech and language therapist, audiologists, geneticists, psychiatrists, maxillofacial surgeons, Paediatricians, Ear, nose and throat surgeon and prosthodontists. [14,19]

Table 1: Various classification schemes for cleft lip and palate.

Veau's classification for cleft lip and palate	Veau's classification for cleft lip
Class I - Isolated soft palate cleft	**Class I-** A Class I- A unilateral notching of the vermillion not extending into the lip
Class II - Hard/soft cleft palate	**Class II-** A unilateral notching of the vermillion, with the cleft extending into the lip but not including the floor of nose
Class III - Unilateral cleft lip and palate	**Class III-** A unilateral notching of the vermillion border of the lip extending into the floor of the nose.
Class IV- Bilateral cleft of the lip and palate	**Class IV-** Any bilateral clefting of the lip, whether incomplete notching or complete clefting

Davis and Ritchie classification	International confederation of plastic and reconstructive surgery classification
Each of the following subgroups is further subdivided into the extent of the cleft (1/3, 1/2, etc):	This system uses an embryonic framework to divide clefts into 4 groups, with further subdivisions to denote unilateral or bilateral cases.
Group I: Clefts anterior to the alveolus (unilateral, median, or bilateral Cleft Lip)	**Group I:** Defects of the lip or alveolus
Group II: Post alveolar clefts (Cleft Palate alone, soft palate alone, soft palate and hard palate, or sub mucous cleft).	**Group II:** Clefts of the secondary palate (hard palate, soft palate, or both)
	Group III: Any combination of clefts involving the primary and secondary palates.

Kernahan and Stark symbolic classification

This system provides a graphic classification scheme

using a Y-configuration, which can be divided into 9 areas:

Areas 1 and 4: Lip
Areas 2 and 5: Alveolus
Areas 3 and 6: Palate between the alveolus and the incisive foramen
Areas 7 and 8: Hard palate
Area 9: Soft palate.

Non-surgical treatment involves the use of prosthodontics devices to correct velopharyngeal incompetence but they were rarely used as the prosthesis had to be readjusted every 2 weeks until growth was finished. Also the prosthesis posed many problems such as irritation to mucosa, difficulty in cleaning etc. [11] On the other hand, infants with cleft lip and palate face challenges when feeding they are incapable of sucking either their mother's nipple or from a bottle. For that reason, feeding devices such as nipples, cross curt nipples and longer nipple can be successfully assist the infant when feeding. [10,14] Some babies may not have the energy to suck from a teat, and here a cup and spoon method may be helpful. [18]

Surgical procedures are often debated considering the age of the patients and many techniques are often employed for the same, one of which is Millard technique lip repair performed at 3 months of age and palate repair at 12 months of age. The other is Malek protocol [20] in which soft palate repair is performed at 3 months of age and lip. Hard and soft palate repair is performed at the age range of 9–18 months. [18]

Pediatricians used to strictly follow a rule of 'three 10s' as a necessary requirement for identifying the child's status as suitable for surgery (i.e., 10 lb, 10 mg/L of hemoglobin, and age 10 weeks). [12,21]

During the Primary dentition period "Lip revision and closure of palatal fistula" are considered to support the speech development. [22] During this stage the orthodontic treatment part is usually limited to correction of mild to moderate posterior cross bite and anterior cross bites [23].

Conclusion

During the mixed dentition phase consideration of alveolar bone graft begins to correct defects of the maxillary arch. 9–11 years is considered as an ideal age for "bone grafting". Orthodontic treatment is not generally commenced until age 9 or 10 years when, if necessary, the maxillary segments are expanded to correct the transverse relationship using palatal expansion appliances, these include upper removable appliance, quad helix, rapid maxillary expansion, bonded "fan" appliance and others. It is during the permanent dentition the definitive orthodontic treatment must be commenced.

Competing Interests

The authors declare that they have no competing interests.

References

1. Regezi JA, Sciubba JJ, Jordan RCK. In Oral Pathology: Clinical Pathologic Correlations, 5th ed. Saunders 2008.
2. El-Kehdy J, Abbas O, Rubeiz N. A review of Parry-Romberg syndrome. J Am Acad Dermatol 2012;67:769-784.
3. Sapp JP, Eversole LR, wysocki GP. In Contemporary Oral and maxillofacial Pathology. 2nd ed. Mosby
4. Precious DS, Goodday RH, Morrison AD, Davis BR. Cleft Lip and Palate: A Review for Dentists. J Can Dent Assoc 2001;67:668-673.

5. Rajendran R, Shivapathasundram B. Developmental Disturbances of Oral and Paraoral Structures. In Shafer's textbook of oral pathology. 7th ed. India: Elsevier; 2012.
6. Melnick J. Cleft lip with or without cleft palate: Etiology and pathogenesis. J Calif Dent Assoc.1986;14:92-98.
7. Poswillo D. The etiology and surgery of cleft palate with micrognathia. Ann Roy Coll Surg. 1968;43:61-68
8. Neville BW, Damm DD, Allen CM, Bouquot JE. In Oral And Maxillofacial Pathology. 3rd ed. Saunders
9. Amaratunga NA. A study of etiologic factors for cleft lip and palate in Sri Lanka. J Oral Maxillofac.1989;47:7-10.
10. Duarte G, Ramos R, Cardoso M. Feeding methods for children with cleft lip and/or palate: a systematic review. Braz J Otorhinolaryngol, 2016;82:602-609.
11. Kosowski TR, Weathers WM, Wolfswinkel EM, Ridgway EB. Cleft Palate. Semin Plast Surg 2012;26:164-169.
12. Kati FA. Cleft Lip And Palate: Review Article. World j pharm med res.. 2018;4:155-163
13. Mosahebi A, Kangesu L. Cleft lip and palate. Surgery (Oxford), 2006;24:33-37.
14. Beumer J, Marunick M, Esposito S. Maxillofacial Rehabilitation: Prosthodontic and Surgical Management of Cancer –Related, Acquired, and Congenital Defects of the Head and Neck, 2011.
15. Muhamad AH, Azzaldeen A, Watted N. Cleft lip and palate; A comprehensive review. Int J App Basic Med Res. 2014;4:338-355.
16. Strohecker B, Lahey D. Mandibular elongation by bone distraction: treatment for mandibular hypoplasia with Robin sequence, Plast Surg Nurs. 1997;17:8-10.
17. Zreaqat MH, Hassan R, Hanoun A. Cleft lip and palate management from birth to adulthood: An overview. Insights into Various Aspects of Oral Healt. 99-121.
18. Van Aalst JA, Kolappa KK, Sadove M. MOC-PSSM CME article: Nonsyndromic cleft palate. Plast Reconstr Surg 2008;121:1-14
19. da Silva FO, de Castro MM, de Andrade A, De Souza Freitas A, Bishara S. Upper dental arch morphology of adult unoperated complete bilateral cleft lip and palate. Am J Orthod Dentofac Orthop. 1998;114:154-161.
20. Allam E, Stone C. Cleft Lip and Palate: Etiology, Epidemiology, Preventive and Intervention Strategies. Anat Physiol 2014;4:150.
21. Paliobei V, Psifidis A, Anagnostopoulos D. Hearing and speech assessment of cleft palate patients after palatal closure. Long-term results. Int J Pediatr Otorhinolaryngol. 2005;69:1373-1381.
22. Figueiredo D, Cardinal L, Bartolomeo F, Palomo M, Horta C, Andrade I, et al. Effects of rapid maxillary expansion in cleft patients resulting from the use of two different expanders. Dental Press J Orthod. 2016;21:82-90.
23. Garib DG, Garcia LC, Pereira V, Lauris RC, Yen S. A rapid maxillary expander with differential opening. J Clin Orthod. 2014;48:430-435.