CIBTech Journal of Surgery ISSN: 2319-3875 (Online)
An Online International Journal Available at http://www.cibtech.org/cjs.htm
2013 Vol. 2 (1) January-April, pp.45-51/Banerjee and Dhakar
Review Article

EPIDEMIOLOGY-CLINICAL PROFILE OF CLEFT LIP AND PALATE AMONG CHILDREN IN INDIA AND ITS SURGICAL CONSIDERATION

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ABSTRACT
Cleft lip and palate being most common is congenital deformity and also present with many syndrome condition. Child with CL/P face difficulty in early breast feeding due to improper lip seal and nasal regurgitation, thus compromising nutrition of new born. Patient with oro-facial cleft deformity needs to be treated at right time and at right age to achieve functional and esthetic wellbeing. The treatment process is complex, multidisciplinary and interdisciplinary approach. India lack in proper reporting and registry of child with cleft and also fail to provide full professional team of experts in common center. Thus is its difficult to get true incidence and prevalence of congenital birth defects as well as provide centralize treatment facilities. Etiology of CL/P multiple and it may due to association of various environmental factors. India being vast in diversity in culture, ethnicity and dynamic genetic pool, there is variation in occurrence of congenital birth defect in different states of India. Post-surgical early complications of cleft lip and palate repair are respiratory obstruction, wound infection, bleeding, and dehiscence. Thus through understanding of defect and proper surgical intervention from team of expertise is needed. Early diagnosis during pregnancy by ultrasonography and stoppage of drugs at time of pregnancy can reduce the risk of child birth with oro-facial cleft. India needs more efforts and research in field of congenital birth defects.

Key Words: *Cleft Lip/Palate, Patients Care with CL/P, Epidemiology*

INTRODUCTION
Cleft lip and palate (CL/P) is one of the most common congenital deformities of craniofacial malformation leads to various dental anomalies in early childhood. There is possibility that child may suffers from either cleft lip or cleft palate or both simultaneously at same time during birth. A cleft lip is an opening or split in the upper lip whereas cleft palate is an opening or split the roof of the mouth i.e palate or maxillary bone (Fig. 1).

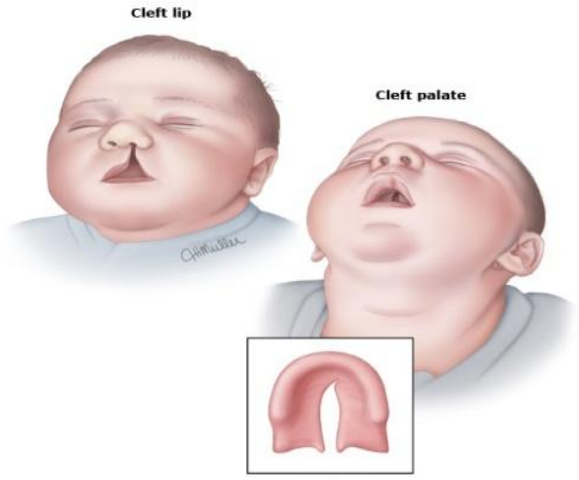

Figure 1: Cleft lip and palate
(Source: *Haug et al., 2012**)***

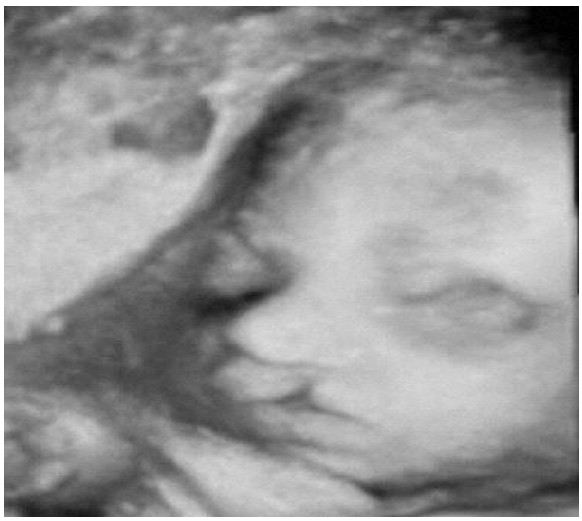

Figure 2: 3D-ultrasoundofcomplete
(*Source:* *Haug et al., 2012**)***

Child with this anomaly not only suffers for poor dental development but also deprived from breast feeding due to improper oral seal, swallowing and nasal regurgitation, other associated problems are hearing difficulties due to abnormalities in the palatal musculature, and speech difficulties due to nasal escape and articulation problems (Haug *et al.,* 2012**)**. Though oro-facial cleft defects can be surgically repaired in early childhood, but post-surgical residual deformity is due to scarring and abnormal facial growth and development results in continuing functional and psychosocial problems. India have diversity and varied population which provide huge diverse population in terms of genetics, social-demography as well as cultural difference but lack in proper reporting of cleft lip and palate registry and thus many children with this defect is not treated in their early childhood. Thus, clefts have a prolonged, adverse influence on the health and social integration of affected individuals. National epidemiological data on cleft lip and palate (CL/P) is not available thus true incidence and prevalence to estimated cleft lip and palate in India is difficult (Mossey *et al.,* 2002). Few scattered data from various studied reported in different states of country is present thus there is wide variation of incidence of cleft lip and palate ranging from 0.25-2.29 per 1000 live birth is reported. To treat the case of cleft lip and palate interdisciplinary team is required to meet the standard of treatment apart from surgical intervention. Thus centralization of therapy is needed in country to fulfill the demand considering the burden of problem in different population of states in India.

Epidemiology and Etiology of Problem Statement

Prevalence of this congenital CL/P deformity is reported as 0.5-2 per 1000 live births depending on the country's population. Cleft lip and palate is more frequent in Asian countries (2.1 in 1000), as compared to African and American countries (Canfield *et al.,* 2006). Incidence of CL/P is more in White Americans than compare to Black Americans. Cleft lip occurs more common in male than female whereas cleft palate occurs more commonly in female than in male reason for this is that fusion of the palatine shelves 1 week later in girls than in boys is thought thus it could be one of factor contributing in higher frequency of cleft palate in girls (Jensen *et al.,* 1988). Indian being the second most populous country of world with population of 1.21 billion, it is estimated that 24.5 million births per year and the birth prevalence of clefts is somewhere between 27,000 and 33,000 clefts per year. In India prevalence of cleft lip is estimated as 9.1 per 10,000 depending upon various epidemiological factors such as ethnicity, geography location and socio-demographic parameter.

Various epidemiological studies show that, if one parent affected with a cleft has a 3.2% chance of having a child with cleft lip and palate and a 6.8% chance of having a child with isolated cleft palate (Grosen *et al.,* 2010). Presence of a cleft in one parent and in one sibling is associated with a 15.8% chance that the next child will have a cleft lip or palate, and a 14.9% chance that the next child will have a cleft palate (Christensen *et al.,* 1996). In case where parents with one is child affected with a cleft have a 4.4% chance of having another child with a cleft lip and palate and a 2.5% chance of having a child with isolated cleft palate (Table 1).

Untreated clefts of the lip and palate is consider as significant health care problem in India leading to aesthetic loss as well as psychological trauma in early childhood to adulthood. Etiology of clefts is multi factorial with both genetic and environmental factors associated with it. Through various studies it have been found that environmental risk factors such as maternal exposure to tobacco smoke, alcohol, poor nutrition, viral infection, medications, and teratogens in the workplace and at home in early pregnancy contribute in etiology of this defect (Shi *et al.,* 2008). Certain types of anti-epileptic drugs have also been reported to increase the risk. Genetic factors also make strong hold for occurrence of defect. Recently it is found that gene (Transcription factor) IRF6, the gene implicated in Van der Woude syndrome (VDWS) has been shown to play a strong role in the isolated form of cleft deformities. Presence of cleft lip and palate is more common monozygotic twin pairs than in di-zygotic pairs (Sivertsen *et al.,* 2008). Folic acid deficiency also contributes to a range of birth defects and mostly associations are with neural tube defects and cardiac malformations but some evidence through literature search suggest it is also associated with

CP but more research is needed in this field. Maternal obesity has been associated several anomalies, including orofacial clefts which also show statistical significance (Rooji *et al.*, 2003).

Table 1: Frequency of oral clefts in relatives based on the proband's phenotype (Source: Haug *et al.*, 2012**)**

Relative	CL (%)	CL/P (%)	CP (%)
Sibling	2.5	3.9	3.3
Half sibling	1.0	0.5	1.0
Parent	2.5	2.5	2.1
Offspring	3.5	4.1	4.2
Niece/nephew	0.9	0.8	1.1
Aunt/uncle	0.6	1.1	0.6
First cousin	0.3	0.5	0.4

Objective

To do through review of epidemiological studies and journal articles related to cleft lip and palate among children in India and provide authentic review article and quality data to the readers.

Methodology

All the articled indexed in authentic journals and search system of Manipal "Health Science" library are taken into consideration. Search mainly focused on epidemiological studies done on cleft lip and palate among children India and developed country to understand actual scenario of problem statement in India and comparing it with developed nations. Search was mainly done to understand the clinical condition along with, best early childhood intervention used. Articles selected which fit into criteria of reviewing up to year 2012 as well as contribution by famous and renounce author in this field were mainly focused. Articles and books were taken into account for surgical aspect of the disease thus care of patient with CL/P and surgical procedure are review from authentic standard books, articles from library source and Muportal search. Articles were mainly searched from internet by using Muportal Intranet surfing from Proquest data base, Pubmed, BMJ, Medline data, UpToDate data base used aiming to achieve reliable source. Period of review was one months.

Classification Of Cleft Lip And Palate

Embryologic development of the lip and palate play major role in classification cleft systems. Cleft lip is classified as unilateral or bilateral and classified as complete or incomplete. A complete cleft involves the entire vertical thickness of the upper lip and is associated with an alveolar cleft palatine bone of maxillary process. An incomplete cleft lip involves only a portion of the vertical height of the lip with a variable segment of continuity across the cleft region. Unilateral clefts of the lip should be distinguished as right or the left side is involved (Tolarová *et al.*, 1998**)**. Palatal clefts also are described as being unilateral or bilateral, and their extent may be classified as complete or incomplete. In addition to this cleft palates are also classified according to their location relative to the incisive foramen in anterior part of maxillary foramen. Clefts of the primary palate occur anterior to the incisive foramen and clefts of the secondary palate involve the segment posterior to the incisive foramen. A unilateral cleft of the secondary palate is defined by a cleft in which the palatal process of the maxilla on one side is fused with the nasal septum. A bilateral complete cleft of the secondary palate has no point of fusion between the maxilla and the nasal septum. A complete cleft of the entire palate involves both the primary and secondary palate, including one or both sides of the premaxilla/alveolar arch of maxillary bone and frequently involve a cleft lip. An isolated cleft palate usually involves the secondary palate only and has varying degrees of severity. The least severe incomplete cleft is the sub-mucous cleft palate (SMCP) in which the underlying palatal musculature is deficient and inappropriately attached (Fig. 3).

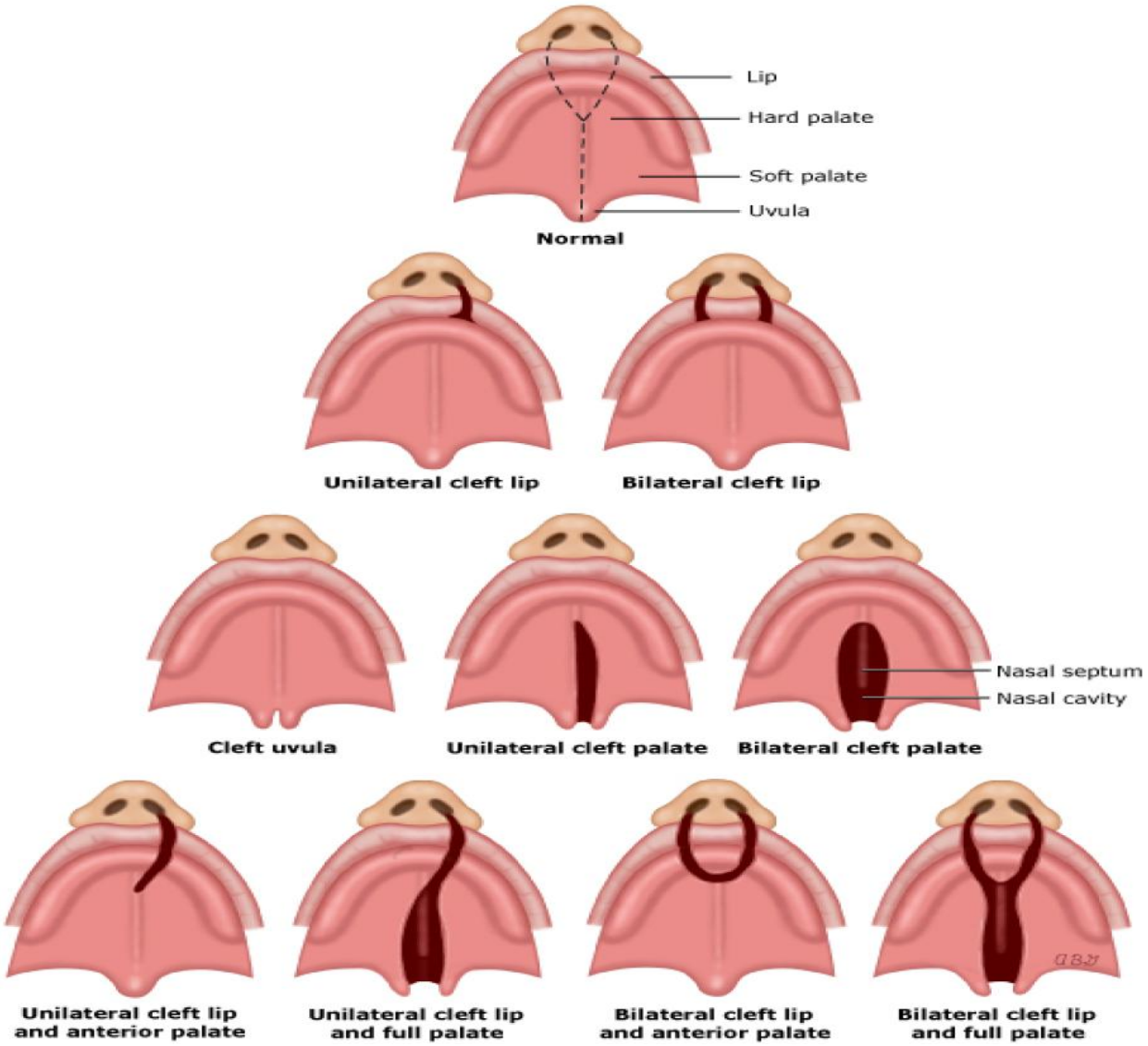

Figure 3: Classification of cleft lip and palate
(Source: *Haug et al., 2012 www.uptodate.com* ***)***

Presence of A Syndrome And CL/P

Orofacial clefting is found to be associated with more than 300 syndromes. In DiGeorge syndrome patients have CP, conotruncal, cardiac defects, thymic hypoplasia, and velopharyngeal webs and it occurs due to deletion of chromosome 22q11. In case of Oral-facial-digital syndrome patient have (CL/P). This is one of the few X-linked dominant chromosomal syndromes hyperplastic frenula, cleft tongue, CL/P and digital anomalies and is due to mutations within a specific gene on the X chromosome. Treacher-Collins syndrome is an autosomal dominant disorder with characteristic facial features including downward slanting palpebral fissures, micrognathia, dysplastic ears, deafness and Cleft Palate but mental development is normal in this case. Stickler syndrome patient have hereditary arthro-ophthalmopathy or Stickler syndrome is an autosomal dominant disorder characterized by flat facies, myopia and spondyloepiphyseal dysplasia. Clefts of the hard and/or soft palate and uvula may occur. This syndrome should be considered in infants with CP, especially when there is a family history of CP (Barrow *et al.*, 2002).

Diagnosis and Management of Cl/P

Prenatal Diagnosis and Obstetrics Management

Ultrasonography is use for in prenatal and antepartum diagnosis of cleft lip and palate. Prenatal ultrasonography routinely used method and consider as standard practice. Along with prenatal ultrasonography amniocentesis for karyotype should be done as orafacial cleft is associated many syndromes due high rate of chromosomal defects. In many cases orofacial clefts with absence of congenital abnormalities are unlikely to be associated with a chromosomal abnormality which is difficult to diagnose by prenatal ultrasonography diagnosis. To achieve more accurate diagnosis 2D ultrasonography is combined with 3D ultrasonography (Fig. 2). Diagnosis is done as early as18 weeks and accuracy of diagnosis improves with the age of the foetus. Itis estimated that 12% of these foetuses will have other anomalies. In few circumstances most families find antenatal counselling helpful in planning for care of the child with an orofacial cleft (Haug *et al.,* 2012).

Postnatal Care and Management of Child with Cl/P

To provide care to patient with CL/P, multidisciplinary approach and team of experts to provide interdisciplinary care to child to maintain physical, psychological, social, mental and nutritive wellbeing of the child with oro-facial cleft.

Table 2: Age wise multidisciplinary approach to care patients with cleft lip and cleft palate
*(**Source:** Flint et al., 2010)*

Age Range	Intervention
Prenatal	Refer to Cleft Palate Team. Medical diagnosis. Genetic counselling. Address psychosocial issues.
Neonatal (0-1 month)	Same as above. Provide feeding instructions. Monitor growth. Hearing screening.
1-4 months	Monitor feeding and growth. Repair cleft lip. Monitor ears and hearing.
5-15 months	Monitor feeding, growth, and development. Repair cleft palate. Monitor ears and hearing; consider ear tubes. Instructions in oral hygiene.
16-24 months	Assess speech and language development Monitor ears and hearing; ear tubes if indicated Monitor development
2-5 years	Monitor speech and language development; manage velopharyngeal insufficiency Monitor ears and hearing; ear tubes if indicated Assess development and psychosocial needs Consider lip/nose revision before child reaches school age
6-11 years	Monitor speech and language; manage. Velopharyngeal insufficiency. Orthodontic evaluation and treatment. Alveolar bone graft. Monitor school and psychosocial needs.
12-21 years	Monitor school and psychosocial needs Orthodontics and restorative dentistry Genetic counseling Rhinoplasty (if needed) Orthognathic surgery (if needed)

As per America Cleft Palate-Craniofacial Association special team includes-reconstructive surgeon, otolaryngologist, plastic surgeon, oral maxillofacial surgeon, speech-language pathologist, orthodontist, paediatric or prosthodontics, dentist developmental or general paediatrician genetics counsellor audiologist nurse social worker psychologist. To achieve best results one should ensure that the appropriate interventions are carried out at the appropriate time to in achieving functional and aesthetic results (Table 2).

Surgical Consideration

Time to perform surgeries varies among surgeons. Surgeon perform lip repair within 48 hours of the child's birth in institutional delivery case to ensure utilization of hospital stay and facility as well as reduce stigma in society by allowing parents to leave the hospital with a healthy-appearing new-born. While some surgeon prefer surgery to allow for more tissue availability for the repair, thus providing more time for parent-child bonding, and more time for the parents to gain a better understanding and acceptance of the infant's congenital deformity (Show *et al.,* 2002) . Most popular in CL/P surgery is the "RULE OF 10S" applied surgery which is consider best time to perform surgical intervention - infant is at least 10 weeks old, weighs 10 pounds, and has a haemoglobin level of 10 g/dL, thereby surgeries are performed during the child's second to third month of life for health wellbeing among children (Tolorovo *et al.,* 1971).

Various surgical techniques use as-Unilateral and bilateral lip Repair, Milliard Technique is used in which surgical procedure like advancement flap, rotation advancement flap and rotation flap. For cleft palate repair (palatoplasty) basic surgical techniques include primary veloplasty (Schweckendiekpalatoplasty), bipedicled flap palatoplasty (von Langenbeck), V-Y pushback palatoplasty, unipedicled two-flap palatoplasty, and the double-opposing Z-plasty (Furlow) palatoplasty (Tolorovo *et al.,* 1998).

CONCLUSION

Cleft lip and palate is congenital birth deformity which needs to be treated at time and right age. It has complex approach and need team of expertise to provide better development and functional efficacy. Global burden vary as per geographic location, race, ethnicity, genetic factors, maternal health, drug use and environmental factor associated with it. In India lack reporting of congenital deformities due to lack of access to health care, awareness, social stigma, poverty, literacy thus true incidence and prevalence of cases not available. Need for National registry of birth defects is necessary state wise as well as considering vast diversity of ethnicity. Though India has initiative to take up more research in to address the etiology and care of orofacial cleft and planned proper multidisciplinary approach to tackle this problem. WHO have also made collaborative initiative with 6 Asian countries among this countries India through National research body like Indian Council of medical research have made initiative to provide more data base in genetic disorders and birth defects among population are focused and intensive recording through NHFS team in India. Presently there is need to aware and educate Indian people regarding slow and steady increase in genetic and congenital birth defects and National approach to deal with congenital birth defects should be provided in government health care centers.

ACKNOWLEDGEMENT

We are thankful to Manipal University Library "Health Science" for providing best search system and conducting awareness program and encouraging us in doing authentic search of indexed articles from data base of UpToDate, Muportal, Proquest etc. We pay our gratitude to library staffs for providing us with full support and helping us with books and journal search.

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