

Cleft lip and palate

Orofacial clefts (cleft Lip (CL), cleft lip and palate (CLP), cleft palate (CP) alone, as well as median, Lateral (transversal), Oblique facial clefts) are among the most common congenital anomalies. The incidence of orofacial clefting varies among racial backgrounds. Worldwide prevalence of cleft lip and palate is 1 per 700 live births.^{1,2} People of African descent have the lowest incidence of 0.5 per 1000 live births, followed by whites (1 per 1000 live births), and Asians (1.3 per 1000 per live births). Lip and cleft palate are among the most common birth defects, they most commonly occur as isolated birth defects but are also associated with many inherited genetic condition or syndromes. A child with a cleft lip with or without cleft palate has an approximately thirty percent chance of having an associated syndrome; interestingly, a child with an isolated cleft palate has a fifty percent incidence of an associated syndrome.^{2,3}

With rapidly advancing knowledge in medical genetics and with new DNA diagnostic Technologies, more cleft lip and palate anomalies are diagnosed prenatally and more cortical clefts identified as syndrome. Genetic and environmental factors have both been associated with clefting. There are some syndromes in which clefting can be passed down in an autosomal dominant fashion, such as van der woude syndrome, where presence of the cleft palate is an autosomal dominant trait. Use of anticonvulsants, maternal smoking alcohol use, Zinc and folate deficiencies have all been associated with cleft Lip & Palate.⁴ Patients with cleft lip & palate have constant feeding difficulty as they are not able to suck because of the defect. Risk of aspiration is always there leading to aspiration pneumonias. Cosmetic issues are constantly causing mental stress to the patients. Timing of repair is between the ages of 3 – 6 months for cleft lip and 9 – 12 months for palate. Depending on the severity of the deformity, various forms of presurgical orthopedics can be used to prepare the child for lip surgery. In general, the goals of these techniques are to improve the alignment of the alveolar segments, decrease the size of the soft tissue cleft, and to improve the symmetry of the nose. Some centers also use a technique termed nasoalveolar molding (NAM) to address the three major components of the cleft deformity NAM addresses the slumping of the nasal alar cartilage, helps realigning the alveolar ridges, and brings the soft tissue of the lips into closer proximity.⁵⁻⁶

There are multiple surgical techniques for cleft lip repair out of which three are predominant; Millard rotation advancement technique, Modified Millard rotation advancement technique and triangular flap techniques.⁷ Bilateral cleft lip repairs are especially challenging because of a central lack of soft tissue, and the anterior displacement of the premaxilla, which functionally increases the transverse width of cleft defect. Realistically, patients with bilateral cleft lips will ultimately require revisional surgeries to correct the secondary stigmata of the repair, which include a shortened columella, blunted nasal tip, widened nasal ala, and a widened philtrum. The choice of techniques for palate repair depends on the type of cleft. Recent surveys show that the most commonly used techniques are the Bardach two-flap palatoplasty and the furlowpalatoplasty. The veau-Wqard-kilner pushback and the von langenback techniques, although less common, are also used. The common denominators for all of these techniques are repair in three layers; nasal mucosa, muscle layer of the soft palate, and the oral mucosa, and anatomic repositioning of the soft palate musculature. Bilateral cleft palate repair is similar in principle in that a three-layer repair is achieved.⁸

Patients with orofacial clefts require multidisciplinary care that is provided by plastic surgeons, otolaryngologists, dentists, orthodontists, oral surgeons, geneticists, audiologists, and speech and language pathologists. This team approach yields more comprehensive and coordinated care, which benefits the patient.⁸

The incidence of cleft lip and palate has been increased in Pakistan over the years. Leading causes are poverty, cousin marriages, illiteracy, folate deficiency and lack of antenatal visits. Incidence is

high in hilly areas, south Punjab and Sindh. Almost 1200 cases of cleft lip and palate have been operated in Rahim Yar Khan with collaboration of an international NGO SMILE TRAIN and department of pediatric surgery providing free surgical facility.

What is needed in our society is that a health education campaign be started at national level with focus on risk factors, prenatal diagnosis and treatment options available, so that prevention, early diagnosis, treatment and rehabilitation goals may be achieved.

References:

1. Waitzman NJ, Romano PS, Scheffler RM. Estimates of the economic costs of birth defects. *Inquiry*. 1994;188-205.
2. Fogh-Andersen P. Inheritance of harelip and cleft palate. Busck, Copenhagen.
3. Tolarova MM, Cervenka J. Classification and birth prevalence of orofacial clefts. *Amer J Med genet*. 1998;75:126-137.
4. Tessier P. Anatomical Classification facial, cranio-facial and latero-facial clefts. *J Maxillofac Surg*. Jun 1976;4(2):69-92.
5. Tolarova M. Significance of cleft Microforms. In: karfik V, ed. *Burian's Laboratory of Congenital anomalies*. 1999:120
6. Millard DR. cleft lip. In: McCarthy JG, May JW, Littler JW, eds. *Plastic Surgery* Philadelphia, Pa: WB Saunders Co; 1990.
7. Datana S, Bhalla A, Kumar P, et al. Comparative evaluation of prevalence of upper cervical vertebrae anomalies in cleft lip/palate patients: a retrospective study. *Int J clinpediatr Dent*. Sep-Dec 2014;7(3): 168-71.
8. Michalski AM, Richardson SD, Browne ML, et al. Sex ratios among infants with birth defects, National Birth Defects Prevention Study 1997-2009. *Am J Med Genet A*. 2015; 167(5):1071-81

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