CONGENITAL MALFORMATIONS ASSOCIATED WITH CLEFT LIP AND PALATE

1ABDUL RASHID, BDS, MDS
2JAVERIA ASIF CHEEMA, BDS, FCPS
3M SUMAIR FAROOQ, BDS, MPhil
4MUHAMMAD AZEEM, BDS

ABSTRACT

Clefts of the lip and palate generally represent a heterogeneous group of disorders affecting the lips and oral cavity. Cleft palate affects almost every function of the face except vision. Effects on speech, hearing, appearance, and psychology can lead to long lasting adverse outcomes for health and social integration. Typically, children with these disorders need multidisciplinary care from birth to adulthood and have higher morbidity and mortality throughout life than do unaffected individuals.

Objectives of this study were to describe congenital malformations associated with cleft lip and palate and to describe the ratio of cleft palate in males and females. It was an observational & comparative study conducted at the Department of Oral and Maxillofacial Surgery of the Children Hospital & The Institute of Child Health, Lahore from January 2013 to December 2013. 120 patients without specifica- tion of gender were included in the study. Data were collected through Questionnaires during 6 months. Results showed that out of 120 patients, 65 % were males while 35% were females, 61.7% have unilateral and 38.3% have bilateral cleft. In patients with unilateral cleft, left side was more affected than right. Parents of 63.3% patients have cousin marriage. About 20% patients have family history of cleft palate. Infants with oral clefts often have significantly high risk of other associated congenital defects along with congenital anomalies, so various malformations especially cardiac, central nervous system, ocular and facial defects may need to be considered in infants with facial clefts. This study also concludes that cleft palate is more common in male than female pediatric population.

Key Words: Cleft lip, Cleft palate, Associated congenital malformations.

INTRODUCTION

Cleft lip (cheiloschisis) and cleft palate (palatoschisis), which can also occur together as cleft lip and palate are variations of clefting congenital deformity caused by abnormal facial development during gestation.1 A cleft is a fissure or opening that is the non-fusion of the body’s natural structures that form before birth. Cleft palate affects almost every function of the face except vision. Commonly affected parts of the face other than palate are eyes, ears, nose, cheeks, and forehead. The group of orofacial cleft anomalies is heterogeneous.2

Cleft lip is formed at the top of the lip as either a small gap or an indentation in the lip (partial or incomplete cleft) or it continues into the nose (complete cleft). It is due to the failure of fusion of the maxillary and medial nasal processes (formation of the primary palate). A mild form of a cleft lip is a microform cleft.3 A microform cleft can appear as small as a little dent in the red part of the lip or look like a scar from the lip up to the nostril. In some cases muscle tissue in the lip underneath the scar is affected and might require reconstructive surgery.4

Cleft palate is an opening in the roof of the mouth (the palate) due to a failure of the palatal shelves to come fully together from either side of the mouth and fuse during embryonic development. Among some babies, both the front and back parts of the palate are open whereas in others only part of the palate is open. It may involve the full length of the palate including soft palate as well. A cleft palate is categorized according to whether it affects the hard palate, the soft palate, or both. These patients showed statistically significantly smaller values for intercanine alveolar widths and larger values for intermolar dental and alveolar widths compared with the normal occlusion group.5 Significant
three-dimensional tooth size asymmetries were found in CLP subjects.³

There are various classifications of cleft palate, most common are Kernahan and Stark classification and Veau classification. According to Kernahan and Stark the incisive foramen is the dividing line between the primary and secondary palate.³ In Veau the palatal defects have been assigned different classes i.e., Class I Defects of the soft palate only. Class II: Defects involving the hard and soft palates (not extending anterior to the incisive foramen). Class III: Defects involving the palate through to the alveolus, Class IV: Complete bilateral clefts.

In most cases, the cause of cleft lip and cleft palate is unknown. Most scientists believe clefts are due to a combination of genetic and environmental factors.³ There appears to be a greater chance of clefting in a newborn if a positive family history exists. Genetic factors contributing to cleft lip and cleft palate formation have also been identified for some syndromic cases.⁹ Environmental influences may also cause, or interact with genetics to produce orofacial clefting e.g. maternal hypoxia, alcohol abuse or some forms of hypertension treatment.¹⁰

Problems associated with Cleft Palate patients include eating problems because with a separation or opening in the palate, food and liquids can pass from the mouth back through the nose. Ear infections/hearing loss since they are more prone to fluid build-up in the middle ear and if untreated, ear infections can cause hearing loss.¹¹ Speech problems because the voice may take on a nasal sound, and the speech may be difficult to understand.¹² Dental problems like missing, extra, malformed, or displaced teeth and specifically alveolar ridge defect which can displace, tip, or rotate permanent teeth, may prevent permanent teeth from appearing, and can prevent the alveolar ridge formation. These problems can usually be addressed through surgery.

Although the preventive and restorative dental care of children with clefts are the same as for other children however, these children require close monitoring due to some special dental problems. This monitoring includes early dental care like proper cleaning with a small, soft-bristled toothbrush, fluoride treatment and good nutrition. The first dental visit to be scheduled at about one year of age or even earlier and routine dental care can begin around 3 years of age. Orthodontic care includes a first orthodontic appointment before any dentition to assess facial growth especially jaw development, assessment of a child's short and long-term dental needs after eruption of deciduous dentition and orthodontic treatment can be applied to align the teeth at permanent dentition. Prosthodontic care like to make a dental bridge for missing teeth, to make special appliances called "speech bulbs" or "palatal lifts" to help close the nose from the mouth so that speech sounds more normal.

Treatment usually begins in infancy and often continues through early adulthood. Most common treatment protocol currently used in most cleft centers is: Newborn - Diagnostic examination, general counseling of parents, feeding instructions, palatal obturator (if necessary). Age 3 months - Repair of cleft lip (and placement of ventilation tubes). Age 6 months - Presurgical orthodontics, if necessary; first speech evaluation. Age 9 months - Speech therapy begins. Age 9-12 months - Repair of cleft palate (placement of ventilation tubes if not done at the time of cleft lip repair) Age 1-7 years - Orthodontic treatment. Age 7-8 years - Alveolar bone graft Older than 8 years - Orthodontic treatment continues.¹³

A multidisciplinary approach is usually involved in the management of these children due to a number of oral and medical problems associated with cleft palate.¹⁴ Members of a cleft lip and palate team typically include: Plasticsurgeon, Otolaryngologist, Oral and Maxillofacial surgeon, Orthodontist, General dentist, Prosthodontist, Speech pathologist and Speech therapist, Audiologist, Nurse coordinator, Social worker/psychologist and Geneticist. Although treatment for a cleft lip and/or cleft palate may extend over several years and require several surgeries depending upon the involvement, most children affected by this condition can achieve normal esthetic and functions subsequently.

The purpose of this study was to describe congenital malformations associated with cleft lip and palate, and to determine the percentage of cleft lip and palate in males and females.

**METHODOLOGY**

It was an observational and comparative study conducted at the Department of Oral and Maxillofacial Surgery of the Children Hospital and The Institute of Child Health, Lahore from January 2013 to December 2013. 120 patients without specification of gender were included in the study. Data were collected through Questionnaires and by using SPSS 16 the data were managed and analyzed. Categorical variable were expressed in the form of frequency table and percentages.

Inclusion Criteria Patients within 1-3 months of age, having Congenital cleft palate, and both genders Exclusion Criteria All syndromic children including Pier Robin syndrome, Patients above 3 months of age.

**RESULTS**

120 patients of cleft palate, 78(65%) were males and 42(35%) were females. The patients were divided in to three categories; unilateral cleft lip with palate,
bilateral cleft lip with palate and isolated cleft palate. Of these 76 patients (63.33%) had unilateral cleft lip with palate, 26 patients (21.66%) had bilateral cleft lip with palate and 18 patients (15%) had isolated cleft palate. The number of patients with associated anomalies was 18. 10 (55.55%) were males and 8 (44.44%) were females, who had total 34 congenital malformations. Among these patients with congenital malformations, 8 patients (44.44%) had unilateral cleft lip with palate, 6 patients (33.33%) had isolated cleft palate and 4 patients (22.22%) had bilateral cleft lip with palate. The commonest anomaly was the facial anomaly with the total of 10 cases (29.41%) followed by ocular anomalies 8 cases (23.52%), central nervous system anomalies 6 cases (17.64%), cardiovascular anomalies 6 cases (17.64%), auricular anomalies 4 (11.76%) (Table 1 & 2). Among family history 80% patients had no history of cleft while 20% patients had history of cleft palate. Parents having cousin marriage will have more chance that their babies have cleft palate e.g. in this study the parents of 63.3% patients had cousin marriages. In ratio of rest of babies 83.7% parents of the patients had other siblings normal while 16.3% parents were having cleft anomaly in other babies as well. The mothers of 10% patients had history of medication and radiation exposure during pregnancy.

**DISCUSSION**

The purpose of this study was to describe the congenital malformations associated with cleft lip and palate and to find out the percentage of cleft palate in male and female pediatric patients. The available data and studies on associated malformations in children with cleft, are quite few and inadequate, which led us to undertake this study. Some of these malformations and anomalies if undetected can prove potentially life threatening, particularly while administering medical and surgical care to the patient with cleft deformity. An international study conducted by Martelli, Cruz, Barros, Silveira, Swerts and Martelli Júnior shows same results regarding congenital anomalies and also proves that cleft palate is more common in males than female peadiatric population. Some other studies regarding this scenario were done by Eshete, Gravenm, Topstad and Befikadu (2011). They studied the incidence of cleft lip and palate. Their study shows that cleft palate is more in females than males. It might be due to some environmental and racial factors. But that study does not nullify present study.

The highest rate of associated malformations was found with unilateral cleft lip with palate (44.44%)
followed by isolated cleft palate (33.33%) and bilateral cleft lip and palate (22.22%). The presence of facial dysmorphic features along with ocular anomalies in a cleft lip or palate child should raise suspicion of an associated malformation. The anomalies were subdivided according to anatomical sites, the most commonly affected group being the facial region. Genetic counseling and studies are important in each case of cleft in which associated anomalies are present. There are also case reports of anomaly of cervical vertebrae found on orthodontic examination of 8-year-old boy with cleft lip and palate diagnosed with Klippel-Feil syndrome. Recent study concluded that echocardiographic screening should be considered in facial clefts infants with nonspecific signs of cardiovascular disease.

In craniofacial region, Unilateral cleft lip and palate patients with congenitally missing permanent teeth have a unique craniofacial morphology with a reduced vertical dimension. There are also studies that the presence of Simonart's band is associated with a higher frequency of maxillary lateral incisor development in the maxillary process.

Substantial observation, documentation and analysis of clinical data is essential for further progress and studies in medical profession. The present study attempts to do the same and provide a platform for advanced researches on chromosomal, genetic, familial or any other study related to the cleft patients, which will guide and help many clinicians to provide comprehensive care for the benefit of these unfortunate patients and their families.

CONCLUSION

Infants with oral clefts often have significantly high risk of other associated congenital defects along with congenital anomalies, so a routine screening for various malformations especially cardiac, central nervous system, ocular and facial defects may need to be considered in infants with facial clefts. The significantly high risk of associated anomalies in children with clefts need to be highlighted and disseminated to the health professionals involved in looking after children with cleft. This study also concludes that cleft palate is more common in males than female’s pediatric population.

REFERENCES

18 Ting-Ting Wu, Craniofacial characteristics in unilateral complete cleft lip and palate patients with congenitally missing teeth AJO-DO: 144; (3): 2013; 381-90.