Prevalence of Cleft Lip and Palate in Cousin Marriages

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Background: Birth defects are one of the leading causes of paediatric disability and mortality in developing as well as developed countries. The purpose of this study is to report our 3 months experience regarding the prevalence of cleft lip and palate in cousin marriages. Methods: A retrospective review of 50 patients who presented with cleft lip or cleft lip/palate during 3 months, from 9th March to 8th June, 2005 was carried out.. Results: Among the risk factors cousin marriages is a most frequent risk factor. Cleft lip is more common than cleft lip/palate and cleft lip alone is a different entity than cleft lip and palate combined.

Key words: Cleft lip, Cleft palate, Consanguinity.

A descriptive study of 50 cases was conducted at the Department of Plastic and Reconstructive Surgery, Mayo Hospital, Lahore from March 2005 to June 2005 to evaluate the prevalence of cleft lip and/or cleft palate in cousin marriages.

42 patients were male and 8 were female. Maternal age ranged from 22 to 29 years. 38 patients were with cleft lip alone and 12 patients were with combined cleft lip and palate.34 patients had a defect on left side, while 8 patients had a defect on right side. 8 patients were suffering from bilateral defect. Cousin marriage was the commonest risk factor, present in 39 patients whereas other risk factors included maternal smoking, epilepsy and obesity. X-ray chest and abdominal ultrasound were done to find the associated defects. Surgery was undertaken in all cases, at 3 months for cleft lip and 9 months for repair of cleft palate.

Patients and methods:

50 consecutive patients with cleft lip or combined cleft lip and palate were admitted in Department in Plastic and Reconstructive Surgery, Mayo Hospital, Lahore, during 3 months from 9th March to 8th June 2005.

A thorough clinical examination was carried out in all patients. Investigations like blood and urine complete examination, blood sugar, blood urea, serum creatinine were carried out. X-ray chest and abdominal ultrasound was also carried out to rule out other associated congenital defects. Surgery was carried out in all patients aim was to achieve a good functional and cosmetic results. Surgical procedures included repair of cleft lip and palate at the age of 3 months and 9 months respectively.

Results:

In this study of 50 cases, 42 patients were male (84%) and 8 patients were female (16%) (Fig. 1). Male to female ratio was 5:1. The age ranged from 6 months to 15 years with a mean age was 7.75 years. Maternal age

ranged from 22 to 29 years with a mean age was 25.5 years.



Fig 1. Sex distribution of patients

Among 50 cases, 38 patients were with the cleft lip alone (76%) and 12 patients were with the combined cleft lip and palate (24%) (Table 1). 34 patients (68%) had a left sided defect while 8 patients had a right sided defect (16%). Bilateral defects were seen in 8 cases (16%).

Table 1. Clinical presentation

Presentation	=n	%age
Cleft lip alone	38	76
Combined cleft lip and palate	12	27

Cousin marriage was the commonest risk factor present in 39 patients (78%). Mothers of 5 patients had a history of smoking (10%). Mothers of 4 patients had a history of epilepsy (8%), while in 2 patients the mothers were obese (4%). Surgery was undertaken in all cases (Table 2).

Table 2. Risk factors

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Risk factors	n=	%age
Cousin marriage	39	78
Smoking	5	10
Disease e.g. Epilepsy	4	8
Obesity	2	4

Discussion:

Birth defects are one of the leading causes of paedriatic disability and mortality in developed and developing countries. Mothers having affected babies also have an association with advanced maternal age and higher rates of previous abortions¹. Orofacial defects are common congenital anomalies which causes a social and moral depression to the patients and their parents. In this study, 42 patients are male (84%) and 8 are female(16%). This male preponderance is also seen in the study of Banskey R² and Baily³. According to presentation,34 patients(68%)present with left sided defect whereas 8 patients (16%) have right sided defect. Bilateral defects are seen in 8 patients(16%). 38 patients present with cleft lip alone (76%) while 12 patients have combined defect of cleft lip and palate (27%). The reverse is shown in the study of Banskey R.

While the cleft of lip, alveolus, soft and hard palate are most common amongst the facial abnormalities (Fig. 2) with the incidence of cleft lip and palate of 1 in 650 live birth, they frequently occur as isolated deformity but can be associated with other conditions. Cleft palate can affect feeding, weight gain, middle ear function, speech and facial growth. Interviews conducted in rural India regarding causation, the vast majority 84% ascribed the defect to "God's will" and 10% to "sins" committed in the past lives. Only 1 parent acknowledged the influence of genetics. Marriage prospects were the main concern, more so for girls than boys⁴.



Fig 2. Cleft lip

Maternal smoking is a known risk factor for orofacial clefts. Maternal smoking during pregnancy increases risk of clefts among fetuses lacking enzymes involved in the detoxification of tobacco derived chemicals. 5 patients (10%) are with the history of maternal smoking. Triple risk is shown in the study of Lammer EJ⁵.

Risk is increased in those children, the mothers of those having influenza, sinusitis and bronchitis. Epilepsy and angina pectoris also show a higher prevalence⁶. Epilepsy is present in 4 patients (8%). Higher incidence is also shown by Metneki J⁷. 2 patients have a history of maternal obesity (4%). Surgery is undertaken in special plastic surgery unit for good cosmetic and functional results⁸.

In this study, the cousin marriage is the commonest risk factor in 39 patients (78%) with cleft lip and/or cleft palate. The same association is also found in the study of Harville EW⁹. The risk of cleft lip only and not of cleft palate is increased for twins and infants, whose parents are first cousins.

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In conclusion, cleft lip with or without cleft palate is more common in males. It shows a peak incidence in 3rd decade of maternal life. Incidence of cleft lip alone is more common especially on left side than combined defect of cleft lip and palate. Cousin marriage is the single most common factor related to this congenital defect. The other less common factors are maternal smoking, epilepsy and obesity.

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