Cardiac Malformations in Children with Cleft Lip and Palate

Zubia Masood, Muhammad Ashraf Ganatra, Hasina Changazi, Asad Awan

ABSTRACT
Objective: Children with cleft lip and palate suffered from increase frequency of other congenital malformations including cardiac anomalies. Cardiac anomalies carries a high risk of aesthetic morbidity and mortality. This study was done to find out associated cardiac malformation in patients of cleft lip and palate presented to plastic surgery department of Civil Hospital, Karachi.

Patients and Methods: All children with cleft lip and palate who presented to Civil Hospital, Karachi were prospectively enrolled in the study during a period of six months from March 2007 to August 2007. All children underwent a thorough clinical examination and an echocardiogram as part of the study protocol. Data was analyzed with the help of SPSS version 10.

Results: Total 55 patients were presented during study period which included 33 (60%) males and 22 (40%) females. Out of them, 19 (34.5%) patients have cleft lip alone, 14 (25.5%) had cleft palate alone and 22 (40.0%) had both cleft lip and palate. Out of 22 patients with complete clefts, 17 (77.3%) were unilateral and 5 (22.7%) were bilateral. Associated cardiac malformations were found in 11 (20%) of patients which included 8 (72.7%) males and 3 (27.3%) females. Most common associated cardiac malformation was atrial septal defect in 5 (45.4%) patients followed by ventricular septal defect in 3 (27.3%). Consanguinity was present in 9 (81.8%) patients with associated malformation and 20 (45.5%) patients without associated cardiac malformation.

Conclusion: Our results showed 20% frequency of cardiac anomalies in patients suffering from cleft palate. The high frequency of cardiac malformations in patients with cleft lip and palate should be highlighted to all the health professional involved in the management of these patients.

Keywords: Cleft lip, Cleft palate, Cardiac anomalies, Congenital malformation

Introduction
Cleft lip and palate occurs relatively commonly with an incidences ranging from 1.2–2/1000 births depending on the population. In a large population-based study of 4,433 children born with orofacial cleft, the birth prevalence of nonsyndromic Cleft lip and palate was 0.77 per 1,000 births and prevalence of nonsyndromic Cleft palate was 0.31 per 1,000 births. In that study, the risk of Cleft lip and palate was slightly lower among the offspring of non-US-born Chinese women compared to US-born Chinese women and slightly higher among non-US-born Filipinos relative to their US-born counterparts. For Cleft palate, lower prevalences were observed among blacks and Hispanics than among whites. The risk of Cleft palate was higher among non-US-born Filipinos compared to US-born Filipinos. These prevalence differences may reflect change in both environmental and genetic factors affecting risk for development of orofacial cleft.
Cleft lip is a feature in 171 syndromes. Sixty four are autosomal recessive, 35 are autosomal dominant and 6 are X-linked recessive. Non-syndromic cleft lip-palate is a heterogeneous disease entity with candidate clefting loci on chromosomes 1, 2, 4, 6, 11, 14, 17 and 19. Ethnicity also plays an important role. Clefts are more common in certain ethnic groups. It is highest in Oriental group, medium in Caucasian and lowest in Blacks. In Pakistan, no definite study has been done to identify particular ethnic group harbouring this abnormality.

It is a well known fact the children with cleft lip and palate suffered from increase frequency of other congenital malformations. Different authors reported different type and frequency of malformations. Extremities were found out to the most common type of malformation in some studies, while head and neck region was the most commonly effected region reported by Shprintzen. Frequency of other congenital anomalies also varies widely with reported range of 7.5 to 63.4% in different studies. Shafi, et al. reported congenital cardiac defects to be the most common associated malformation in children suffering from cleft lip and palate in Pakistan. Cardiac malformations are one of those defects which carry a high risk of aesthetic morbidity and mortality. This hospital based study was done to find out the associated cardiac malformations in patients presented with cleft lip and palate at Civil Hospital, Karachi.

Patients And Methods
All children with cleft lip and palate who presented to Civil Hospital, Karachi were prospectively enrolled in the study during a period of six months from March 2007 to August 2007. Civil Hospital, Karachi (CHK) is a 1670-bed tertiary care teaching hospital in the public sector that imparts both undergraduate and postgraduate teaching and training. It is one of the teaching hospitals affiliated with Dow University of Health Sciences (DUHS). CHK attracts patients not only from Karachi but also from the rural areas of Sindh and Balochistan provinces.

All children underwent a thorough clinical examination and an echocardiogram as part of the study protocol. A specially designed proforma was used to collect data, which included type of cleft, associated cardiac malformations, consanguinity and family history of cleft, apart from demographic details. Data was analyzed with the help of SPSS version 10. Chi square test was used to find out significant difference between categorical data.

Results
Total 55 patients were presented during study period which included 33 (60%) males and 22 (40%) females. Majority of patient's age at presentation was 12 months (mode) with range from 4 months to 11 years. Consanguinity was present in 15 (27.3%) patients while family history of cleft was present in 10 (18.2%) patients. Out of them, 19 (34.5%) patients have cleft lip alone, 14 (25.5%) had cleft palate alone and 22 (40.0%) had both cleft lip and palate. Out of 22 patients with complete clefts, 17 (77.3%) were unilateral and 5 (22.7%) were bilateral. Associated cardiac malformations were found in 11 (20%) of patients which included 8 (72.7%) males and 3 (27.3%) females. Out of them, 2 (18.2%) patients had cleft palate alone and 9 (81.8%) had both cleft lip and palate and none had cleft lip alone.
Of the patients with complete clefts, 7 (77.8%) were unilateral and 2 (22.2%) were bilateral. Most common associated cardiac malformation was atrial septal defect in 5 (45.4%) patients followed by ventricular septal defect in 3 (27.3%). All cardiac malformations is shown in Table-I.

Table-I: Associated congenital malformations in patients with clefts (n=11)

<table>
<thead>
<tr>
<th>Cardiac Malformation</th>
<th>n (%)</th>
</tr>
</thead>
<tbody>
<tr>
<td>Atrial Septal Defect</td>
<td>5 (45.4)</td>
</tr>
<tr>
<td>Ventricular Septal Defect</td>
<td>3 (27.3)</td>
</tr>
<tr>
<td>Valvular disease</td>
<td></td>
</tr>
<tr>
<td>Mitral Stenosis (1)</td>
<td></td>
</tr>
<tr>
<td>Aortic Stenosis (1)</td>
<td></td>
</tr>
<tr>
<td>Pulmonary Stenosis (1)</td>
<td>3 (27.3)</td>
</tr>
</tbody>
</table>

Consanguinity was present in 9 (81.8%) patients with associated malformation and 20 (45.5%) patients without associated cardiac malformation (p-value<0.05). Family history of cleft was present in 2 (25.0%) patients with associated malformation and 8 (26.7%) patients without associated cardiac malformation (p-value=1.0) (Table-II).

Table-II: Consanguinity and family history among cleft patients

<table>
<thead>
<tr>
<th>Risk factors</th>
<th>Cardiac malformation</th>
<th>P-value*</th>
</tr>
</thead>
<tbody>
<tr>
<td></td>
<td>Yes</td>
<td>No</td>
</tr>
<tr>
<td>Patients with consanguinity</td>
<td>9</td>
<td>20</td>
</tr>
<tr>
<td>Patients without consanguinity</td>
<td>2</td>
<td>24</td>
</tr>
<tr>
<td>Family history of cleft present</td>
<td>2</td>
<td>8</td>
</tr>
<tr>
<td>Family history of cleft absent</td>
<td>9</td>
<td>36</td>
</tr>
</tbody>
</table>

* Chi square test was used to find significance between the groups

Discussion

Diagnosis of cardiac anomalies in the newborn is complicated by the physiologic changes that occur in the postnatal period. Even with a detailed cardiac antenatal scan and clinical examination up to 30% of cases can be missed. As many as half of these missed cases can be severe, presenting with heart failure or sudden death.10,13

There are very few studies from our country about associated cardiac defects in cleft patients.10 Our results showed 20% prevalence of cardiac anomalies in patients suffering from cleft lip and palate. Other study from Pakistan10 showed 15% frequency of associated cardiac defects. Stoll10 and Milerad10 reported congenital heart disease to be five times and 16 times more common than general population in French and Swedish population respectively. However both of these studies used clinical examination alone to used as diagnostic protocol.

We have found atrial septal defect (ASD) to be most common malformation. Shafi et al.10 also reported ASD to be the most common defect followed by Ventricular septal defect (VSD) and patent ductus arteriosus (PDA). Other showed VSD to be the most common heart anomaly.1,16 Contrary to this, we did not encounter any patients with PDA in our series. We fail to understand this observation.

Consanguinity is an important risk factor for many congenital anomalies including cleft lip and palate. This trend is very well prevalent in our society. Genetic factors are passed to the next generation, thus creating an increased risk for such anomaly in offspring. We found significant relationship of consanguinity with associated cardiac defects as also observed by Shafi, et al.9 Stoll11 also reported significant association consanguinity and birth defects while some other authors showed no difference.15
Undiagnosed cardiac defects may cause problem in anaesthesia during surgical management of cleft lip and palate. The preoperative assessor of a child with a cleft defect must maintain a high index of suspicion of associated congenital heart disease even in the face of unremarkable scans.

**Conclusion**
The high frequency of cardiac malformations in patients with cleft lip and palate should be highlighted to all the health professional involved in the management of these patients. A routine screening for cardiac defects especially in neonatal period with the help of echocardiography should be protocol at every institute.

**References:**


