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RESEARCH ARTICLE

Congenital Heart Diseases in the Cleft Lip Palate Patients and its Perioperative Implications: An Observational Study In Rural Central Maharashtra, India

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ABSTRACT

Introduction: Congenital heart disease (CHD) is a common birth defect that occurs in approximately 1% of live births worldwide. Cleft lip and palate (CLP), another common birth defect, affecting approximately 1 in 600- 800 live births in India. Several studies have reported an association between CHD and CLP, however, there is limited data on the prevalence and types of CHD in patients with CLP in rural areas of developing countries, including India. This study aimed to investigate the prevalence and types of CHD in patients with CLP at a rural hospital in central Maharashtra, India.

Methods: This retrospective study included medical records of patients with isolated Cleft lip, isolated cleft palate and patients with cleft lip and palate, between January 2018 and December 2020. The study was conducted at a rural hospital in central Maharashtra, India. The patients' records were examined for any heart disease. Patients who had a diagnosis of CHD were identified through medical records and echocardiography reports. The prevalence and types of CHD were compared between the two groups using descriptive statistics and chi-square tests.

Results: A total of 291 patients were included in our study. 169 were males and 122 were females. Congenital heart diseases were found in 12.37% (n=36) patients. 5 Cleft lip (13%), 12 Cleft lip and palate (48%), 19 Isolated cleft palate (39%) were diagnosed with Congenital heart diseases. The types of Congenital Heart Diseases reported in our study were atrial septal defect, Ventricular septal defect, Patent ductus arteriosus, Pulmonic Stenosis and Tetralogy of Fallot.

Conclusion: The most common types of CHD in both groups were ASD and VSD, but the prevalence of these defects was significantly higher in the CLP group. These findings have important implications for the multidisciplinary Perioperative management of patients with CLP, as they may require close monitoring for CHD and appropriate intervention if necessary, especially in resource-limited settings.

Keywords: congenital heart disease, cleft lip and palate, isolated cleft palate, cleft lip, non-syndromic clefts.

Introduction:

Cleft lip, Cleft lip with cleft palate and isolated cleft palate, are one of the most common craniofacial abnormalities. They affect the feeding, speech, hearing, dental health, appearance and never the least, psychosocial health¹⁻⁴. Such children are often abandoned from the society, and this is very common in rural parts of India⁵.

Congenital heart diseases are structural abnormalities of the heart and/ or great vessels which presents at birth^{6,7}. CHD is one of the most common birth defects, affecting approximately 1% of live births worldwide⁸⁻¹⁰. CHD are further classified into cyanotic & acyanotic diseases^{11,12}. Cyanotic CHDs are Tetralogy of Fallot, tricuspid atresia, transposition of great vessels, total anomalous pulmonary venous return, and truncus arteriosus¹³. Acyanotic CHDs are Patent ductus arteriosus, atrial septal defect, ventricular septal defect with left to right shunt, pulmonary stenosis. Cleft lip and palate, another common birth defect, affecting approximately 1 in 600- 800 live births in India ^{5,9,11}. Overall incidence of cleft lip and palate is approximately 1.42 in 1000 and isolated cleft palate occurs approximately in 1 in 2000 live births¹¹.

The co-occurrence of Congenital Heart Diseases and Cleft of Lip and Palate has been reported, with studies suggesting that patients with CLP have an increased risk of CHD^{1,2,14}. However, there is limited research on the prevalence of CHD in patients with CLP particularly in rural areas of developing countries. This paper aims to discuss the congenital heart disease in patients with cleft lip and/or palate, and their implications in providing safe and comprehensive cleft care in rural parts of central Maharashtra, India.

Materials & Methods:

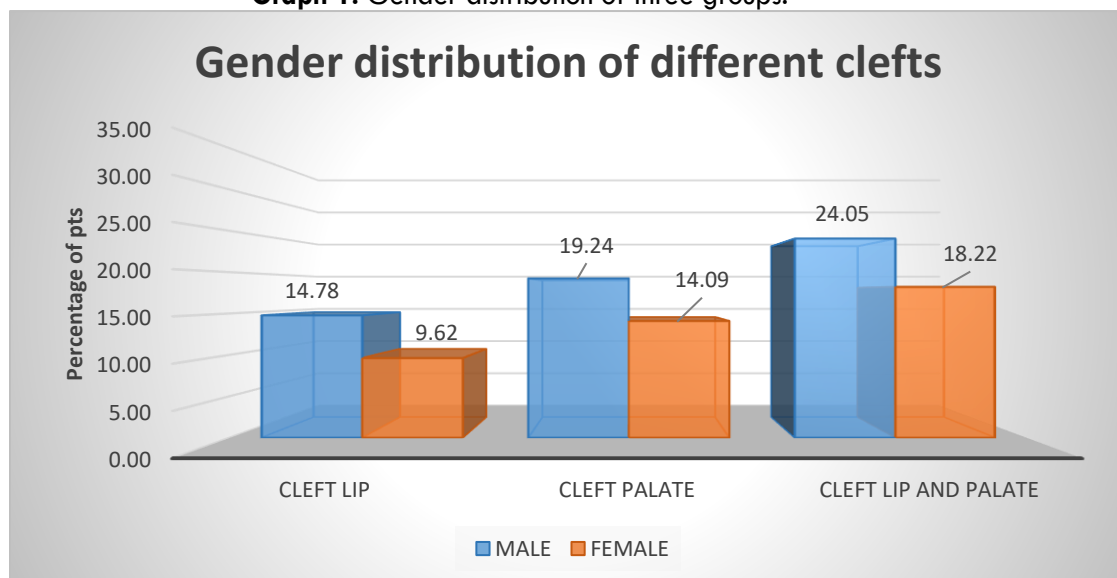
This retrospective study included medical records of patients with isolated Cleft lip, isolated cleft palate and patients with cleft lip and palate, between January 2018 and December 2020. The study was conducted at a rural tertiary care hospital in central Maharashtra, India with the approval of institutional ethics committee. The patients' records were examined for any heart disease. Patients who had a diagnosis of CHD were identified through medical records and echocardiography reports. Patients with incomplete medical records & syndromic cleft patients were excluded from the study. The prevalence and types of CHD were compared between the groups. Three groups were made as per the anatomical classification viz. Cleft lip, cleft palate & cleft lip palate both. The cleft lip group & cleft lip with palate group were subdivided into unilateral & bilateral group. Isolated cleft palate group was subdivided into Cleft of Hard & soft palate & cleft of soft palate only. Statistical analysis was done using SPSS 20 & chi square test was applied for statistical significance. $p < 0.05$ was considered statistically significant.

Results:

Demography of the sample:

A total of 291 patients were included in our study. 169 were males and 122 were females. There was a male preponderance in the individual cleft lip and cleft palate and also in patients with cleft of lip and palate (graph1). The incidence of cleft lip was 33.33%, cleft palate was 24.40% while those with cleft of lip and palate was 42.27%.

Graph 1: Gender distribution of three groups.



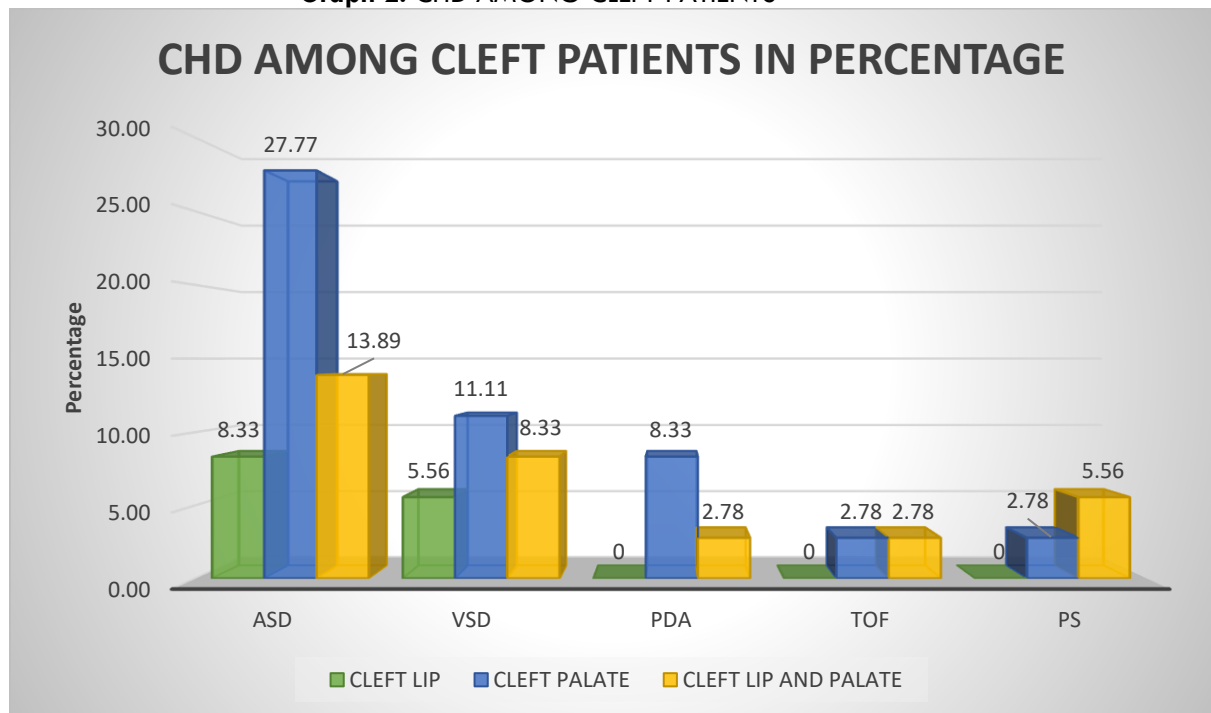
DISTRIBUTION OF CLEFT PATIENTS:

Of the cleft lip patients, 78.87% (n=56) were unilateral cleft lip and 21.13% (n=15) were bilateral cleft lip. Cleft soft palate only were identified in 35.05% (n=34) cleft palate patients and 64.95% (n=63) patients had cleft of hard and soft palate both. There were 123 patients who were diagnosed with cleft lip & palate, out of which 57.72% (n=71) as unilateral defects of lip and palate and 42.28% (n=52) were having bilateral cleft of lip and palate.

Congenital heart diseases were found in 12.37% (n=36) patients. 5 Cleft lip (13%), 12 Cleft lip and palate (48%), 19 Isolated cleft palate (39%) were diagnosed with Congenital heart diseases.

The types of Congenital Heart Diseases reported in our study were ASD (Atrial septal defect) (50%), VSD (ventricular septal defect) (25%), PDA (patent ductus arteriosus) (11.11%), PS (Pulmonic Stenosis) (8.33%) and TOF (tetralogy of fallot) (5.56%) (Graph 2).

Graph 2: CHD AMONG CLEFT PATIENTS



When comparing the prevalence of specific types of CHD between the two groups, there was no statistically significant difference in the prevalence of TOF, PS or PDA. However, the prevalence of ASD was higher in the CP group (27.77%) compared to the isolated cleft lip group (8.33%), and cleft lip and palate (13.89%).

Discussion:

Cleft lip refers to a fissure in the upper lip and may be incomplete or complete (through the nasal cavity), unilateral, or bilateral. Cleft palate is a gap in the hard and/or soft palate creating an oro-nasal communication. Children born with syndromic or non-syndromic oro-facial clefts have a significantly higher incidences of congenital heart diseases¹⁵. Various heart lesions have been associated with the cleft of lip or palate or both lip and palate in Indian population. Many authors have studied the prevalence of various congenital

defects in patients presenting with oro-facial cleft in developed countries but there are very limited studies done in rural population of developing countries¹⁵.

We have studied the prevalence of cleft lip and palate, or both in the Rural Parts of Central Maharashtra from January 2018 to December 2020.

Of the total 291 patients' medical records, we observed congenital heart lesion in 36 patients, corresponding around 12.37% of the total sample size. This was similar to the incidence found by Kasatwar et al in eastern part of Maharashtra, India, where the incidence found was 15%¹⁶. However, Al Hammad et al had found higher incidence of congenital heart disease in cleft patients, which was around 50%¹⁵.

The incidence of CHD was more in cleft palate patients (19.59%), with almost equal distribution in cleft of soft palate or that with soft

and hard palate. Atrial septal defect was found to be the predominant lesion in these patients. Bilateral cleft of lip (13.34%) had more frequency of congenital heart disease as compared to the

unilateral cleft lip (5.36%). Patients with unilateral or bilateral cleft of lip and palate had similar incidences of CHD. (Table 1)

Table1: Prevalence of CHD in cleft patients.

TYPE OF CLEFT	NUMBER OF PTS	CASES OF CHD	%	MALE (%)	FEMALE (%)	P value
CLEFT LIP UNILATERAL	56 (78.87%)	3	5.36	2(66.67)	1(33.33)	
CLEFT LIP BILATERAL	15 (21.13%)	2	13.34	0(0)	2(100)	
TOTAL 1	71	5	7.04	2(40)	3(60)	(p=0.16,NS)
CLEFT OF HARD AND SOFT PALATE	63 (64.95%)	12	19.05	9(75)	3(25)	
CLEFT OF SOFT PALATE ONLY	34 (35.05%)	7	20.59	4(57.14)	3(42.86)	
TOTAL 2	97	19	19.59	13(68.42)	6(31.58)	(p=0.021, S)
CLEFT LIP AND PALATE UNILATERAL	71 (57.72%)	7	9.86	5(71.43)	2(28.57)	
CLEFT LIP AND PALATE BIILATERAL	52 (42.28%)	5	9.62	1(20)	4(80)	
TOTAL 3	123	12	9.76	6(50)	6(50)	(p=1.03 NS)
GRAND TOTAL	291	36	12.37	21(58.33)	15(41.67)	

Study conducted by Aljohar et al have found incidences as close to 38% of CHD in patients with malformations in cleft patients¹⁷. In a similar study conducted by Nancy Geis et al, they found more incidences of CHD in Bilateral Cleft of Lip, Palate and alveolus³. The number of CHD patients were evenly distributed among the various types of cleft patients in the study undertaken by Barbara E. Otaigbe¹⁸. Similar studies conducted by Ting Sun et al, found that incidences of CHD was higher in Cleft palate (20%) as compared to cleft lip (3.1%) or cleft of lip and palate (16.3%)¹⁹. Barbosa et al found that about 9.5% of cleft lip and palate patients had congenital heart disease⁸. From a study conducted in Pakistan by Shafi et al, the authors found higher incidences of CHD (54%) in patients with cleft lip and palate²⁰.

The exact cause of the co-occurrence of CHD and CLP is not known. However, several risk factors have been proposed. One theory is that the co-occurrence of CHD and CLP is due to a shared genetic etiology. For example, some genetic syndromes such as 22q11.2 deletion syndrome are associated with both CHD and CLP. Another theory is that environmental factors such as maternal

smoking and alcohol consumption during pregnancy may increase the risk of both CHD and CLP²¹.

In our study, amongst all the congenital cardiac malformations, atrial septal defect (ASD) was amongst the most commonly associated cardiac anomaly, accounting for 50% in patients with cleft lip and palate (Graph 2). The result of this study is similar to that of Ziyad Al Hammad¹⁵, BL Akhiwu²², who found ASD to be the predominant congenital cardiac anomalies in cleft lip and palate cases. Ting sun et al, also noted that the ASD was higher in cleft palate than in cleft of isolated lip or cleft lip and palate¹⁹.

Congenital heart diseases significantly increase morbidity, mortality, and medical care cost in children. It is one of the leading causes of mortality in early childhood²³. Congenital cardiac disease accounts for about 10% of infant mortality rate in India²⁴. Increased incidence of CHD IN CLP is frustrating to the parents as well cleft care team as it puts additional monetary investments for multiple investigations, more visits to hospitals as well as sometimes delays the surgical management. Generally, first cleft surgery of the cleft child is primary cheiloplasty. It is done when age of the child is 4 months and weight is equal or more than

5.5kg. Palatoplasty is done when the child attains 10 months of age & weight is around 8.5kg. Congenital heart diseases patients many times lack to get paediatric & anaesthetic fitness for surgery under general anaesthesia at the proper age interval. This may lead to delayed physical growth, poor speech quality and late social acceptance.

Peri-operatively, cleft patients with CHDs are at increased risk of cardiac arrhythmias, stroke due to blood clots, congestive cardiac failure, pulmonary hypertension, infective endocarditis, upper respiratory tract infections²⁵. Thus, post-operatively ICU stay becomes mandatory thereby increasing the cost for total care.

Early diagnosis and management of CHD in patients with CLP is important for improving outcomes. Diagnosis of CHD typically involves a combination of medical history, physical examination, and diagnostic testing such as echocardiography. In patients with CLP, it is recommended that a baseline echocardiogram to be performed at the time of diagnosis to screen for CHD. If CHD is detected, referral to a pediatric cardiologist is necessary for further evaluation and management.

The management of CHD in patients with cleft depends on the type and severity of the defect. Some defects may require medical management with medications such as diuretics or beta-blockers, while others may require surgical intervention such as cardiac catheterization or defect closures and valve replacement surgeries. In patients with CHD and CLP, coordination of care between the pediatric cardiologist and cleft team is essential to ensure optimal outcomes.

Certain precautions need to be taken preoperatively in patients with CHDs associated with cleft lip and palate. Careful clinical examination, medical history, essential diagnostic evaluation and necessary laboratory investigations should be done to identify various other systemic abnormalities. Blood investigations including complete blood count, coagulation profile such as prothrombin time, activated partial thromboplastin time and International Normalised Ratio (INR) are mandatory as the patient may be on antiplatelet medications. C reactive protein and WBC count will determine the potential diagnosis of infections. Patient may be taking certain medications like Aspirin, Clopidogrel (dual antiplatelet therapy), warfarin, diuretics, antiarrhythmic, and antihypertensive. All the necessary medications

related to CHDs should be continued even during the surgical management of cleft patients. For simple and short time surgeries, low dose aspirin can be continued. However, for major complex surgeries prior cardiologist and anaesthetist opinion should always be done to avoid uncontrolled bleeding. CHD patient's on warfarin should be preoperatively monitored and should be shifted on heparins before surgery. All the patients with infective endocarditis associated with cleft lip and palate should mandatorily undergo antibiotic prophylaxis according to American Heart Association (AHA) guidelines to further reduce the magnitude of the postoperative risk.

Cardiac risk assessment should be done for all patients undergoing cleft repair surgery as a critical part of preoperative evaluation as they may have associated cardiac anomalies. According to European Society of Cardiology (ESC) and The European Society of Anaesthesiology (ESA) guidelines for non-cardiac surgeries, patients with stable cardiac disease can undergo low and intermediate risk surgeries which include head and neck surgeries^{16,23,26}. Patients with complicated CHDs have to be critically evaluated by the cardiologist to reduce postoperative complications^{4,8,24}. High risk cardiac patient's categorized as ASA III, IV and V are at a relatively higher operative risk and require superior degree of monitoring and care^{5,27}. In a high-risk group if the underlying cardiac anomaly is of great concern and if it is significant, it is advisable to treat the congenital cardiac disease first and then the cleft lip and palate surgeries to reduce further medical and surgical complications.

Conclusion:

In conclusion, this study provides evidence for a higher prevalence of CHD in isolated cleft palate patients at a rural hospital setting in central Maharashtra. Further research is needed to confirm this association and to determine the underlying mechanisms. The findings of this study have important clinical implications for the adequate perioperative risk assessment for surgery to provide patient safety in multi stage cleft care. This study also emphasizes the essential role of Pediatric cardiologist in the multidisciplinary cleft care team. Mandatory screening new born babies with cleft defect using noninvasive tools like 2D echocardiography may help in prompt recognition and improved care for the patients.

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