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Original Article

Prevalence of congenital cardiac anomalies in patients with cleft lip and palate – Its implications in surgical management

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ABSTRACT

Background: Cleft lip and palate is one of the most common congenital craniofacial deformities seen in children. Various congenital anomalies are reported in the literature to be associated with cleft lip and palate. Cardiac anomalies are one of the most common congenital disorders associated in cleft lip and palate patientsIt includes Cyanotic and acyanotic cardiac diseases likel fallot's tetralogy, transposition of greater vessels, atresia of tricuspid, total anomalous pulmonary venous return (TAPVR), truncus arteriosus, ebstein's anomaly, hypoplastic left heart syndrome and pulmonary atresia, patent ductus arteriosus, ventricular septal defect, atrial septal defect, pulmonary stenosis, aortic stenosis and coarctation of aorta.
Aim: To study the prevalence of congenital cardiac anomalies in cleft lip and palate patients.
Objectives: To study different types of congenital cardiac anomalies/defects in patients with cleft lip and palate and its implications in surgical management.
Materials and Methods: This is a retrospective study carried out for a period of one year. In the present study medical records of 200 patients with cleft lip and palate were evaluated and analyzed for presence of congenital cardiac anomalies.
Results: Out of 200 patients of cleft lip and palate, 30 patients (15%) were associated with congenital cardiac anomalies with male to female ratio of 1:1. Associated congenital cardiac anomalies were most frequently seen in unilateral cleft palate patients (21.05%) The most common cardiac anomaly was Ventricular septal defect (36.6%).

1. Introduction

Defective embryogenesis leading to structural and functional abnormalities of prenatal origin is referred to as congenital malformations. Congenital malformations may be caused by environmental factors, genetic and of unknown origin. The mode of the cause of such defects in most of the cases is uncertain. Various congenital abnormalities reported in the literature includes skeletal system disorders, nervous system(central) disorders, congenital cardiac diseases, respiratory system abnormalities, polydactyly, eyes and ear anomalies, limb anomalies, chromosomal disorders, talipes equinovarus, cleft of lip and palate, anencephaly, spina bifida, and many more.[1,2]

Congenital anomalies or malformations are often associated with certain types of secondary major defects. In an epidemiologic study by Vallino-Napoli,one third of the cleft patients were associated with other birth related defects in Victoria, Australia, and the prevalence rate of cleft lip and palate (CL/P) was higher than that of cleft palate(CP).[3] Associated deformities of cleft lip and palate patients with other congenital malformations was around 32.2% as reported by Beriaghi in USA and patients with CP (38.7%) were having more congenital anomalies than in patients with CL/P (26.4%).[4] In another study by Luijsterburg, 10% of patients with clefts had other anomalies of the craniofacial region and congenital defects involving the other systems were 13%.[5]

Congenital cardiovascular anomalies are the structural flaws in the heart predisposing the patients to various complications. According to the study done by Hoffman, congenital heart disease (CHD) had incidence of about 4 to 5 per 1000 live births.[6] Various studies in the literature has reported the relations of CHD with extra-cardiac malformations. Many of these patients may require noncardiac surgeries. This study was intended to determine the prevalence of congenital cardiac anomalies in patients with cleft

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lip and palate, to analyze their association and its surgical implications in the management of patients with cleft lip and palate.

2. Materials and method

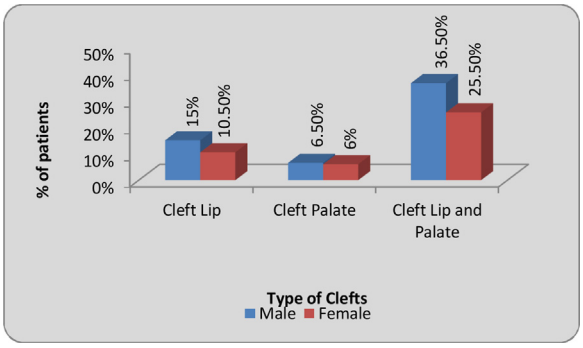
All the patients operated for cleft lip and palate between October 2015 to September 2016 in the Oral and Maxillofacial Surgical Centre at Acharya Vinoba Bhave Rural Hospital, Sawangi (Meghe), Wardha, India, were retrospectively analyzed for the presence of congenital cardiac anomalies (CCA's). The data of 200 patients was collected from Medical Record Department. All the patients showing features of CCA's were included in the study. The data of the discharged patients signifying clinical signs and symptoms like shortness of breath, palpitations, failure to thrive, digital clubbing, cyanosis, abnormal cardiac murmur, history of recurrent chest infections, abnormal chest x-ray and other diagnostic investigations including ECG & echocardiography were selected for the study.

3. Results

In the present study, the medical report of 200 patients of cleft lip and palate were analyzed of which 134 (67%) were males and 66 (33%) were females. The frequency, type and sex distribution of the patients with cleft lip and palate are highlighted in Graph 1. The occurrence of bilateral cleft lip and cleft palate was 32.5%(n = 63) the most frequent subtype, followed by unilateral cleft lip with palate 30.5%(n = 61), unilateral lip 20%(n = 40), unilateral cleft palate 9.5%(n = 19), bilateral cleft lip 5.5%(n = 11) and bilateral cleft palate3%(n = 6).

Congenital cardiac anomalies were associated with 30 patients (15%) of congenital clefts. Patient's with unilateral cleft palate (21.05%) were more frequently born with CCA's followed by those with unilateral (one side) cleft lip alveolus and palate(18.03%), bilateral(two side) cleft palate(16.67%),bilateral cleft lip alveolus and palate(15.87%),bilateral cleft lip (9.09%) and unilateral cleft lip (7.5%) in that order (Table 1). Various types of CCA's associated with Cleft lip and palate is summarised in Graph 2.

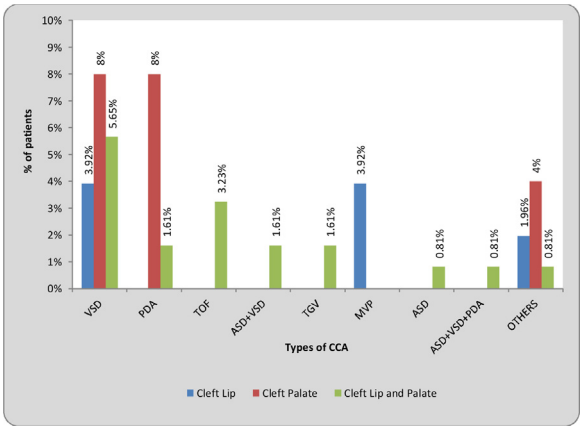
The different type of CCA reported in the study were VSD (ventricular septal defect)(36.67%), PDA(patent ductus arteriosus) (13.33%),TOF (tetralogy of fallot)(13.33%), TGV(transposition of greater vessels) (6.67%),MVP (mitral valve prolapse)(6.67%), ASD (Atrial septal defect) (3.33%), ASD associated with VSD(6.67%) and ASD + VSD + PDA (3.33%) and others which included two patients with rheumatic heart disease and mitral regurgitation and one patient with mitral regurgitation with associated aortic regurgitation (10%) Graph 3. Statistical analysis was done by using descriptive and inferential statistics using z-test for single proportions. The software used in the analysis was SPSS 17.0 and Graph Pad Prism 5.0 and $p < 0.05$ was considered as level of significance



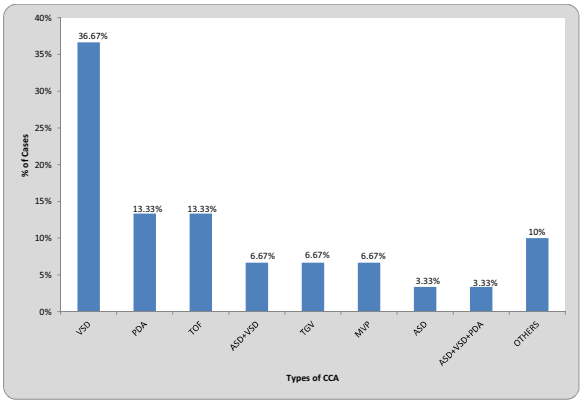
Graph 1. Showing Type, Frequency And Sex Distribution Of The Patients with Cleft Lip & Palate.

Table 1
Prevalence of CCA in Cleft Lip & Palate Patients.

Type of cleft	No of patients	Cases of CCA	%	Male (%)	Female (%)
Cleft Lip					
Unilateral	40	3(1.80,NS)	7.50	1(33.33%)	2(66.67%)
Bilateral	11	1(1.05,NS)	9.09	0(0%)	1(100%)
Total	51	4	7.84	1(25%)	3(75%)
Cleft Palate					
Unilateral	19	4(2.25,S)	21.05	2(50%)	2(50%)
Bilateral	6	1(1.10,NS)	16.67	1(100%)	0(0%)
Total	25	5	20.00	3(60%)	2(40%)
Both(CLAP)					
Unilateral	61	11(3.66,S)	18.03	9(81.82%)	2(18.18%)
Bilateral	63	10(3.45,S)	15.87	2(20%)	8(80%)
Total	124	21	16.94	11 (52.38%)	10(47.62%)
Grand Total	200	30	15.00	15(50%)	15(50%)



Graph 2. Prevalence of Different Types of CCA in Cleft Lip And Palate Patients.



Graph 3. Prevalence of different types of Congenital Cardiac Anomalies.

4. Discussion

Cleft and craniofacial malformations are the most common anomalies in head and neck region results due to an inborn error in structural morphogenesis .They may invariably occur with other congenital defects.[7] Congenital malformations are commonly seen in almost all parts of the world. The prevalence of cleft and craniofacial anomalies is relatively higher in central region of India.

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