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A Study of Associated Anomalies With Cleft Lip and Palate in Children

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ABSTRACT

Introduction: Cleft lip and Cleft palate also known as orofacial cleft is a common congenital conditions that include cleft lip, cleft palate and both together. They are type of birth defect, cause is mostly unknown.
Aim: Aim of this study to identify associated anomalies among children with oral clefts and to study the risk factors for associated anomalies in oral clefts.
Methods: Prospective observational study was done in pediatrics with cleft lip and cleft palate. Clinical examination and presence of associate anomalies were assessed with Echo cardiogram, USG abdomen. CT brain/ X-ray were done for required cases.
Results: 23 cases of orofacial cleft were associated with anomalies, musculoskeletal anomalies 9 cases, CNS anomalies 7 cases, CVS anomalies 6 cases. There is no risk factor for associated anomalies.
Conclusion: Orofacial cleft were most commonly associated with musculoskeletal, CNS and CVS anomalies. Early diagnosis with antenatal USG help to plan delivery at tertiary care and follow up.

KEYWORDS Cleft lip, Cleft palate, Orofacial cleft, risk factors

INTRODUCTION

Facial Clefts are one among the most common of congenital anomalies. It has been widely studied as a part of malformation or deformation. The etiology is as varied as the defect presentation itself. Both genetic and environmental factors may predispose to its occurrence, where the defect is brought out by environmental factors in genetically predisposed individual. Some instances are considered to be sporadic in occurrence where neither environmental nor genetic factors can be traced. Oral Clefts are associated with malformation.[1] Malformation signifies a morphological defect of an organ, part of organ or large region of body resulting in an intrinsic abnormal development process. A major malformation has serious medical, surgical or cosmetic consequences. Lead on to functional disability and may compromise normal life expectancy. The majority of birth defects almost 2/3rd are isolated involving a single organ or system. The common examples are oral clefts and congenital heart disease.[2] The cause of isolated anomalies is usually Multifactorial or polygenic. Less commonly congenital anomalies may affect several organs or body systems producing multiple congenital defects. The common pattern of multiple structural anomalies includes association, sequences, field defects and syndromes. A deformation is an alteration in shape or structure of a part that has differentiated normally. A minor anomaly is 10 cosmetic and can be arbitrarily differentiated. About 2-3% of newborn have major anomaly and minor anomaly is observed in 10% of new born babies. Children with 3 or more minor anomalies are like to have a dysmorphic syndrome. The incidence of cleft palate alone is about 1/2500 live births and incidence of cleft lip with palate is about 1.4/1000 live births. Along with facial clefts other congenital anomalies coexist. There is a wide range of syndrome with facial clefts and many other single anomalies not included in any syndromes. Knowledge of these associated anomalies especially the lethal ones will enable the pediatrician to identify and treated effectively thus reduces the mortality and morbidity.

AIM

Aim of this study to identify associated anomalies among children with oral clefts and to study the risk factors for associated anomalies in oral clefts.

MATERIALS AND METHODS

Prospective analytical study was done Institute of Social Pediatrics, Stanley Medical College Hospital, Chennai. Institutional Ethics Committee approval and informed consent from the children parents were obtained. Children in the age group up to 12 years with congenital oral clefts, with all degree of severity and occurring alone or a part of syndromes were included in study. All acquired cases of oral clefts were excluded. All children undergo clinical examination, age, gender, birth order, family history, consanguinity, antenatal risk factors and details examination for presence of associate anomalies. Echocardiogram, USG abdomen was done for all cases. CT brain/ X-ray done for whenever necessary.

RESULTS

The study sample consisted of 106 cases. Of these 55 cases were males and 51 cases were females. Among the total number of oral cleft cases 19 cases were cleft lip, 69 cases were lip with palate and 20 cases were cleft palate alone. Among the total number oral cleft cases (106) had associated anomalies. 14 male cases and 9 female cases were associated with anomalies.

Orofacial anomalies were the most common anomalies followed in sequence by musculoskeletal, CNS, CVS anomalies. Four recognized syndromes were present among the 23 cases with associated anomalies. Total number of oral cleft cases 106, among these Cleft lip cases were 19 (17.92%), Cleft lip with Palate cases were 69 (65.09%) and Cleft palate cases were 18 (16.98%).

TABLE 1 TYPE OF ORAL CLEFTS

Sl. No.	Types	Children (%)	95% CI
1.	Cleft Lip	19 (17.92)	11.15 – 26.57
2.	Cleft lip with Palate	69 (65.09)	55.23 – 74.09
3.	Cleft Palate	18 (16.98)	10.39 – 25.50

2 cases (8.7%) of cleft lip had anomalies and 16 cases (69.9%) of cleft lip with palate had anomalies and 5 cases (21.7%) of cleft palate had associated anomalies.

TABLE 2 ASSOCIATED ANOMALIES SYSTEMWISE (n=106)

Sl. No.	System	Children (%)	95% CI
1.	Oro facial	23 (21.7)	(82.2 – 100)
2.	Musculoskeletal	9 (8.5)	(20.5 – 61.2)
3.	CNS	7 (6.6)	(14.1 – 53.0)
4.	CVS	6 (5.66)	(11.1 – 48.7)
5.	Skin	3 (2.85)	(2.78 -33.58)
6.	Renal	1 (0.94)	(0.2 – 24)

The above table shows 23 cases (21.7%) had orofacial anomalies, 9 cases (8.5%) had musculoskeletal anomalies, 7 cases (6.76%) had CNS anomalies, 6 cases (5.66%) CVS anomalies, 3 cases (2.85%) had Skin anomalies and 1 case (0.94%) had renal anomaly.

Among 19 cleft lip cases had 1 CNS anomaly and 1 musculoskeletal anomaly. In 69 Cleft lip cases with palate cases 3 CNS anomalies, 4 CVS anomalies, 12 orofacial anomalies, 7 musculoskeletal anomalies, 1 renal anomaly and 1 skin anomaly were present. In 18 Cleft palate cases 3 CNS anomalies 2 CVS anomalies, 11 Orofacial anomalies, 1 musculoskeletal anomaly and 2 skin anomalies were noted. Total anomalies of cleft lip, cleft lip with palate and cleft palate were 2, 28, 18 respectively.

Associated orofacial anomalies with oral clefts Megalo cornea 1 (4.3%), Coloboma 1 (4.3%), Hypertelorism 2 (8.6%), Flattened nasal bridge 2 (8.6%), Beaked nose 1 (4.3), Low set ears 2 (8.2%), Micro ostia 3 (13%), Bifid Uvula 2 (8.6%), Hypoplastic Maxilla 2 (8.6%), Hypognathia 3(13%), Retrognathia 2 (8.6%)

TABLE 3 MUSCULO SKELETAL ANOMALIES

Sl. No.	Types	No. (n = 23)
1.	Hypoplastic Thumb	2
2.	Syndactyly	3
3.	Bilateral Hip Subluxation	1
4.	Bilateral proximal femur hypoplasia	1
5.	Rocker Bottom Foot	1
6.	CTEV	1

The above table shows associated musculoskeletal anomalies with oral clefts, Hypoplastic thump 2 (22.2%), Syndactyly 3 (33.3%). Bilateral hip subluxation 1(11.1%), Bilateral proximal femur hypoplasia 1 (11.1%), Rocker Bottom Foot 1 (11.1%) and CTEV 1 (11.1%).

TABLE 4 CNS AND CVS ANOMALIES

Sl. No.	Types	Numbers
1.	Cranioschisis	1
2.	Neural tube defects	4
3.	Facial Palsy	2
4.	ASD	1
5.	VSD	4
6.	Pulmonary Stenosis	1

The above tables shows CNS anomalies associated with oral cleft Cranioschisis 1 (14.3%), Neural tube defects 4 (57.2%), Facial palsy 2 (28.6%). The above table shows associated CVS anomalies with oral clefts cases were ASD 1 (16.7%), VSD 4 (66.7%), Pulmonary Stenosis 1 (16.7%).

TABLE 5 ANALYSIS OF RISK FACTORS FOR ORAL CLEFT WITH ASSOCIATED ANOMALIES

Sl. No.	Risk Factors	Without Anomalies	With Anomalies	Total
1.	Consanguinity	29	6	35
2.	Family H/o Oral Cleft	8	2	10
3.	Previous H/o abortion still birth	18	9	27
4.	Preterm	5	2	7
5.	Low birth weight	8	3	11
6.	Hyper emesis	2	2	4
7.	Infection	3	1	4
8.	Threatened abortion	0	1	1
9.	Medical Problems	6	2	8

The above table shows children with oral cleft with associated anomalies had a higher incidence of previous abortion, still birth. Antenatal risk factors were not associated with increase incidence of anomalies in oral cleft children.

DISCUSSION

In 106 cases, 55 cases were males and 51 cases were females. J. Womersly et al in their study reported that males predominated in cleft lip group and females predominated in cleft palate. Milerad et al in their study reported 21% had associate malformations, in our study we reported 21.7%. Rustemeyer et al in their study in their study reported CNS anomalies 16%, CVS 15%, musculoskeletal 8%, in our study we reported 39% musculoskeletal, 30% CNS, 26% CVS. P A Boyd et al in their study antenatal diagnosis with sonography reported high prevalence neural tube defect in cleft cases. Liang et al study shows overall prevalence in congenital heart disease 5.4%, Isolated, atrial septal defect and ventricular septal defect were two common congenital defect which presented defects 23% and 21% in oral cleft cases. In our study CVS anomalies shows ASD 4%, VSD 17%, Pulmonary stenosis 4%. Rajabian et al reported increased occurrence of associated malformation in oral cleft children born to consanguineous parentage. In our study consanguinity were not associated with increased incidence of associated anomalies. Drillen et al found and increased incidence of threatened abortion, hyperemesis in pregnancy associated with oral cleft. In our study hyperemesis and threatened abortion were not associated with increased incidence.

CONCLUSION

Orofacial cleft were most commonly associated with musculoskeletal, CNS and CVS anomalies. Early diagnosis with antenatal USG help to plan delivery at tertiary care and follow up. Family history, antenatal risk factors are not associated with increased incidence of associate anomalies in orofacial cleft.

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