

CLINICAL STUDY OF CLEFT LIP AND PALATE AND RARE FACIAL CLEFTS IN UPPER ASSAM AND ARUNACHAL PRADESH

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ABSTRACT

BACKGROUND

Facial clefts occur in varying degree of severity and variety of patterns. It is a nightmare which every parent wants to avoid. Facial clefts occur when the structures contributing to the formation of face namely- the frontonasal prominence, the maxillary prominence, the medial nasal prominence, the lateral nasal prominence and the mandibular prominences fail to fuse with one another. Many rare facial clefts cannot be explained by simple embryological concepts. These defects result in abnormal facial appearance, difficulties in feeding, defective teeth, defective speech, repeated infections and social stigma.

The study was carried out from January 2008 to June 2018, among the 1000 cases of facial clefts who attended the Smile Train Centre in Srishti Hospitals and Plastic SOPD of Assam Medical College Hospital, Dibrugarh, Assam.

MATERIALS AND METHODS

This descriptive study was carried out among the 1000 cases of facial clefts who attended the Smile Train Centre at Srishti Hospitals and PSOPD in AMCH, Dibrugarh, Assam, from January 2008 to June 2018. All the cases were evaluated on the basis of sex, religion, race, type of cleft, severity of the cleft, family history of facial clefts, birth order, consanguinity, maternal age, associated disease, radiation exposure and drug history during 1st trimester of pregnancy. Also, the socio-economic status, parents' literacy, source of information for treatment and parents' satisfaction after treatment was noted down.

RESULTS

It was found that isolated cleft lip (42.2%) occurred more commonly than isolated cleft palate (15.1%). Cleft lip was more common in males (55.7%) than in females (44.3%). 412 (41.2%) of the cases presented with both cleft lip and palate and among these 268 (65.04%) were females and 144 (34.95%) were males. Isolated cleft palate was found to be more common in females (71.5%) than in males (28.09%). All these cases underwent reconstructive surgery and left the hospital with a socially presentable face.

CONCLUSION

Cleft lip and palate and rear facial clefts are no longer considered a stigma, as they are surgically correctable.

KEY WORDS

Cleft Lip, Cleft Palate, Rare Facial Cleft, Tessier's Cleft, Transverse Facial Cleft.

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BACKGROUND

The face in a human embryo is formed by fusion of some facial prominences that appear in the 4th week of development. The frontonasal prominence forms the forehead, bridge of the nose, medial and lateral nasal prominences. The maxillary prominence forms the cheeks and lateral portion of upper lip. The medial nasal prominence forms the philtrum of upper lip, crest and tip of nose, while the lateral nasal prominence forms the ala of nose. The mandibular prominence forms the entire lower lip and jaw. Different types of cleft lip result if the medial nasal prominence fail to fuse with the maxillary prominences. Bilateral cleft lip occurs due to failure of both maxillary processes to fuse with the medial nasal prominence, which then appears as a separate flap. Failure of whole maxillary prominence to fuse with the lateral margin of the lateral nasal

prominence results in an oblique facial cleft. Cleft palate results due to failure of fusion of the palatine shelves with each other or with the anterior triangular primary palate. Cleft lip and cleft palate are one of the most common congenital deformities. The incidence of cleft lip is approximately 1/1000 births and it occurs more frequently in males (80%) than in females. The incidence of cleft palate is approximately 1/2500 births and occurs more often in females (67%) than in males.^(1,2) To have a child with grossly deformed face is certainly a horrifying experience for the entire family. For all such children, the problem goes beyond the obvious disfigurement of the face to repeated infections, social stigma, problems with speech, hearing and teeth formation.⁽³⁾ These children are often teased by their friends and they lose self-confidence. They suffer with emotional "burn out" in adolescence.⁽⁴⁾ Children with clefts often suffer from anaemia and malnutrition, mainly due to poverty and illiteracy on the part of the parents. In addition, due to lack of awareness many patients of cleft lip remain untreated or mistreated by quacks.⁽⁵⁾ All children born with a cleft lip or palate need thorough paediatric assessment to exclude other congenital abnormalities. Genetic counselling must be sought if a syndrome is suspected. Keeping in mind that no studies regarding the incidence of cleft lip and palate are published from this part of Assam, India. This study was undertaken to form a clinical picture of the patients specific to this region for the benefit of public and the treating doctors.

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MATERIALS AND METHODS

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The Aims and Objectives of this Study are-

1. To study the incidence of isolated cleft lip.
2. To study the incidence of isolated cleft palate.
3. To study the incidence of cleft lip with cleft palate.
4. To study the incidence of rare facial clefts.
5. To study the result of surgical correction.

RESULTS

The data collected were tabulated and analysed as follows-

Male	Female	Total
557 (55.7%)	443 (44.3%)	1000

Table 1. Showing Sex distribution of the Cases

Isolated Cleft Lip	Isolated Cleft Palate	Cleft Lip with Cleft Palate	Rare Facial Clefts	Total
422 (42.2%)	151 (15%)	412 (41.2%)	15 (1.5%)	1000

Table 2. Showing Types of Facial Clefts

Unilateral	Bilateral	Total
373 (88.39%)	49 (11.6%)	422

Table 3. Showing Side distribution of Cleft Lip (CL)

Right Sided	Left Sided	Total
135 (36.19%)	238 (63.80%)	373

Table 4. Showing Side distribution of Unilateral Cleft Lip

Right Side		Left Side		Total
Complete	Incomplete	Complete	Incomplete	
57	98	85	133	373

Table 5. Showing Completeness of Unilateral Cleft Lip

Sex	No. of Cases	Total
Male	276 (65.4%)	
Female	146 (34.59%)	422

Table 6. Showing Sex distribution of Cleft Lip

Sex	No. of Cases	Total
Male	144 (34.95%)	
Female	268 (65.04%)	412

Table 7. Showing Sex distribution of Cleft Lip with Cleft Palate

Sex	No. of Cases	Total
Male	43 (28.09%)	
Female	108 (71.52%)	151

Table 8. Showing Sex distribution of Isolated Cleft Palate

Complete	Incomplete	Total
412	151	563

Table 9. Showing Type of Cleft Palate



Photograph 1. Few Cases of Complete Cleft Lip and Palate and Incomplete Cleft Lip



Photograph 2. Few Post-Operative Cases of Cleft Lip

As for family history, in only one family both the parents had cleft lip and palate and now their two siblings have the same deformity. The age of the cases ranged from 3 months to 25 years. The maternal age ranged between 18 - 39 years.

Anaemia	613
Delayed milestone	42
Mental retardation	2
Respiratory illness	102
Ear infection	13
Underweight for age	160
Associated congenital anomaly	1

Table 10. Showing associated illness

Hb (gm%)	Number of Cases	Percentage
7-8	36	3.6%
8.1-9	232	23.2%
9.1-10	210	21%
10.1-11	135	13.5%
11.1-12	297	29.7%
12.1-13	90	9%

Table 11. Haemoglobin Level of the Patients

It was found that majority of the cases were suffering from anaemia. Approximately, 38% of the cases had haemoglobin level ranging between 11 to 13 gm%.



Photograph 3. Two Cases of Cleft Palate, Pre- and Post-Operative

Type	Number of Cases
Tessier cleft 3, bilateral	1
Tessier cleft 3 and 4	1
Tessier cleft 4	2
Tessier cleft 7	11

Table 12. Rare Facial Clefts

Thus, from the data summarised in Tables 1 - 11, it is seen that majority of the cleft patients were males. Most of the families said that they came to know about the treatment facilities from friends and also from advertisements on radio and television. Since majority of them were of lower socio-economic status, they were grateful that such free treatment was available at their doorstep. The study also shows that 0.64% of parents of children with facial clefts were absolutely illiterate, 37.77% of parents were undergraduates and 61.58% of parents were graduates. As for maternal antenatal history, 15% of women never took iron and folic acid during pregnancy, 41% of women took the same but irregularly and the rest 44% of women regularly supplemented their nutritional requirements as per advice of their doctors. The age of the mother ranged from 18 to 37 years in this study and 52 were consanguineous marriages.

Race or Tribe	No. of Cases	%
Tea-garden community	244	24.4%
Assamese (General)	155	15.5%
Ahom	230	23%
Mishing	89	8.9%
Kachari	59	5.9%
Bengali	48	4.8%
Bihari	29	2.9%
Nepali	31	3.1%
Arunachali	47	4.7%
Naga	32	3.2%
Others	36	3.6%

Table 13. Showing Ethnic distribution of Cleft Lip and Palate

As can be seen, facial clefts are more common among the Ahom community (21.89%) and the Tea-garden community (21.28%) of Assam as compared to others.

Type of Operation	Number of Cases
Rotation and Advancement	316
Triangular variant	439
Straight line repair and others	79

Table 14. Type of Surgery done for Cleft Lip

Type of Operation	Number of Cases
Veau repair	5
Straight line double stage repair	42
Others	2

Table 15. Type of Surgery for Bilateral Cleft Lip

Type of Operation	Number of Cases
Veau-Kilner-Wardill	72
Furlow's procedure	12
Delaire two-stage procedure	36
Langenbeck procedure	159
Push back procedure	284

Table 16. Surgery done for Cleft Palate Cases

Complications	Number of Cases
Wound dehiscence	6
Bleeding	2
Scar hypertrophy	40
Cupid's bow defect	10
Residual nasal deformity	228

Table 17. Complications of Cleft Lip Surgery

Complications	Number of Cases
Bleeding	4
Dehiscence	12
Fistula	20

Table 18. Complications of Cleft Palate Surgery

Operations for wound dehiscence, fistula and residual nasal deformity are not included in the present study.



Photograph 4. Few Cases of Rare Facial Clefts; Pre- and Immediate Post-Operative

DISCUSSION

Due to illiteracy facial clefts are still presumed to be a curse of God by various sections of the society, especially in remote areas. Review of studies shows that there are no trends for incidence of cleft lip and palate in different parts of the world. According to a WHO study on craniofacial anomalies carried out in 13 countries, the incidence varies from 0.22 to 1.67 per thousand live births.⁽⁶⁾

According to available literature, the incidence of cleft lip and palate exhibit ethnic variation. It is found to be highest in Asian or Native North American descents followed by the Caucasians. It is least among the Africans.⁽⁷⁾ In the present study, we found it to be more common in Ahom community and the Tea-garden community of Assam.

Fogh-Anderson⁽⁸⁾ reported that siblings of patients with cleft lip and palate have an increased incidence of cleft lip and palate, but not isolated cleft palate; conversely, siblings of patients with cleft palate have an increased frequency of cleft palate but not of cleft lip and palate. He also found that 30-40% of 703 patients with cleft lip or cleft palate in Denmark had near relative with similar deformities. But in the present study, only 0.12% of 781 cases had near relative with similar deformity.

As for sex ratio, there is an excess of cleft lip and palate in males. Female excess has been reported in isolated cleft palate.^(8,9,10) The findings of our study is also similar. Cleft lip occurred more in males (65.4%) than in females (34.59%). Cleft palate occurred in a higher frequency in females (71.52%) than in males (28.09%).

As noted by Fogh-Anderson⁽⁸⁾ and confirmed by many other studies,^(9,10,11,12,13) there is a left-sided preponderance of cleft lip. In this study also, cleft lip was more (63.80%) on the left side and less (36.19%) on the right side.

Patients of facial clefts are brought for surgery later than the ideal surgical time which may be due to ignorance, poverty or unawareness about the fact that such anomaly can be corrected by surgery. Without repair these children would have suffered from social isolation, feeding problem, abnormal speech and repeated infections.⁽¹⁴⁾ Many patients do not come to the hospital, because they cannot afford the treatment offered. May be this is the reason why the maximum age was 25 years in our study.

As stated earlier, we also enquired about the type of marriage and found that consanguineous marriage is not an uncommon practice in the state.

To correct the facial clefts, different standard operative procedures were followed.^(15,16,17,18,19,20,21) Majority of cleft

palate cases (284) were corrected by pushback procedure followed by Langenbeck procedure (159 cases). Among the bilateral cleft lip cases, 42 were corrected by Veau III and two stage repair procedures. Few complications in some cases were corrected subsequently.

CONCLUSION

Cleft lip and palate is no longer considered a stigma, as these deformities can be corrected by various surgical procedures. Most of these operations take about 45 minutes to one and a half hours' time. It is found that cleft lip and palate is more common in males and on the left side. However, isolated cleft palate is more common in females. Transverse facial cleft is the most common rare facial cleft in our study. Anaemia is the most commonly associated condition followed by underweight for age and respiratory illness. Similarly, maternal malnutrition and anaemia is commonly seen. Most of the cases belonged to lower socio-economic group. Out of 1000 cases, highest number of cases are from Tea-garden community (244) followed by Ahom community (230). Various operative procedures were performed for cleft lip and palate. All cases were done under general anaesthesia. Cleft lip and cleft palate operations were done in separate stages. Rotation and advancement and triangular flap procedures were most commonly done procedures for cleft lip, whereas pushback procedure and Langenbeck procedures were most commonly done for cleft palate. Two stage repair procedure was commonly done for bilateral cleft lip. Commonly encountered complications for cleft lip included scar hypertrophy and residual nasal deformity and for cleft palate; palatal fistula.

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