



A STUDY OF THE CLINICAL PROFILE OF PATIENTS HAVING CLEFT LIP AND PALATE

Dr. Jyoti Dvivedi^{1*}, Dr. Sanjay Dvivedi²

^{1*}Professor, Department of Physiology, Himalayan Institute of Medical Sciences, Swami Rama Himalayan University, Dehradun, Mobile: 9412968996, Email: jdvivedi@gmail.com

²Professor, Department of Surgery, Himalayan Institute of Medical Sciences, Swami Rama Himalayan University, Dehradun, Mobile: 9412961683, Email: sanjaydvivedi@srhu.edu.in

***Corresponding Author:** Dr. Jyoti Dvivedi

*Professor, Department of Physiology, Himalayan Institute of Medical Sciences, Swami Rama Himalayan University, Dehradun, Mobile: 9412968996, Email: jdvivedi@gmail.com

Abstract:

Cleft deformities are a challenge to the surgeons since very long. The present study was undertaken at a tertiary medical care centre in Dehradun, Uttarakhand keeping in mind the scarcity of such studies in this region. In this study 312 patients of cleft lip or cleft palate or both cleft lip and palate, were evaluated over a period of 12 months. The authors investigated the differences between age and sex with cleft status and family history of clefts, type of clefts, surgical procedures done on them; follow up in such children and patient/family satisfaction after treatment.

Maximum numbers of patients were in the age group of 1–10 years and the incidence of clefts was higher in males. Bilateral clefts were present in 19.26% cases where as 50.37% cases had left sided clefts, 30.37% cases had right sided clefts. In the study sample maximum numbers of patients of cleft lip were treated by Randall–Tennison’s technique in almost 70% cases, while almost all the cleft palate patients were operated by Pushback technique. 6 cases showed over or under correction of the lip, 5 cases showed contracted scars but more than 90% of the families were fully satisfied by the treatment and results. This study will provide baseline information on the status of cleft patients in this state of Uttarakhand for future reference to health workers.

Introduction

Cleft lip and cleft palate are the most common congenital deformities after clubfoot. Development of the Head and Face comprises one of the most complex embryonic developments, coordinated by a network of transcription factors and signaling molecules together with proteins conferring cell polarity and cell- cell interactions (1, 2).

The incidence of cleft lip and palate is 1 in 600 live births and 1:1000 live births of isolated cleft palate. The incidence increases in Oriental groups (1:500) and decreases in the black population (1:2000). The highest incidence for cleft lip and palate occurs in the Native American tribes of Montana. In India, it is estimated to occur once in every 781 live births (3, 4).

Cleft lip and palate predominate in males whereas cleft palate alone appears more common in females. In unilateral cleft lip, the deformity affects the left side in 60% of cases (5).

Cleft lip results from the failure of fusion of the median nasal, lateral nasal and maxillary processes on either or both side. Isolated cleft palate may be a result of disruption of one or more number of

mechanisms (3). It is thought that both genes and environmental factors, acting either independently or in combination, are responsible for facial clefting. While numerous non - genetic risk factors have been identified such as use of anti-epileptic's drugs, maternal alcohol intake or cigarette use during first trimester of pregnancy. A large majority of people have beliefs that exposure to solar eclipse, bad luck, and past birth karma are the causes for this birth defect (4, 6).

Cleft lip can be easily diagnosed by performing ultrasound in the second trimester of pregnancy when the position of the fetal face is located correctly. Usually, it is not possible to diagnose a cleft palate by an ultrasound; however, an experienced physician or technician may catch an atypical movement of the fetal tongue in a lateral view. In the case of a large cleft palate, the tongue moves up into an open space (cleft) in the roof of the oral cavity. Recently, 3-dimensional imaging has been introduced to prenatal ultrasound diagnostics of cleft anomalies and seems to be very promising for recognizing a cleft palate in a fetus (7).

Research has shown that attractive children are seen by others as brighter, as having more positive social behavior and receive more positive treatment than their less attractive cleft counterparts. These children suffer with problems of speech and hearing during early childhood, dental anomalies and malocclusion in the developing dentition, secondary facial deformities & emotional "Burn out" in adolescence. Patients were teased because of their clefts and felt their self-confidence was affected by the cleft condition. They were frequently teased about cleft-related features such as speech, teeth, and lip appearance. Parents also reported that their children were being teased because of their clefts and that their children's self-confidence was affected by the clefts (8, 9). The present study has been undertaken to study the clinical profile of patients having cleft lip and palate and to evaluate the outcome of surgical repair, keeping in mind the scarcity of such studies in this region.

Material and Methods

This work was carried out in the Department of Surgery (Plastic Surgery) at Himalayan Institute of Medical Sciences, Swami Ram Nagar, Dehradun over a period of 12 months. All cases of cleft lip and palate attending the Plastic Surgery outpatient department were included in this study.

The details of the subjects in terms of history, clinical features and investigations were recorded on an investigator-designed proforma (annexure I), after obtaining an informed consent from the patient.

International classification of cleft lip and palate was used for description purposes.

For each patient following routine was followed –

1. History taking and Clinical examination
2. Laboratory investigations
3. Pre anaesthetic check up
4. Preop- photographic record
5. Operation of lip and / or palate
6. Postop – photographic record
7. Follow-up

Clinical study

Relevant points in history and physical examination were recorded on special proforma of cleft lip and palate patients.

Unfit Patients

These were the patients who were suffering from cold, fever, low hemoglobin, associated congenital anomalies, underweight and found to be unfit on anesthesia grounds. These patients were prescribed appropriate treatment and called later for the surgery.

Photographs of all these patients were taken at initial presentation, at the time of surgery preoperatively and in follow up.

Following views were taken:

1. Front – full face
2. Basal view or worm's eye view
3. Left lateral profile view
4. Right lateral profile view

On being fit for anaesthesia and surgery patients were operated. Various techniques were employed for the cleft lip and palate cases.

Techniques for unilateral lip repair used in our study

1. Randall Tennison Repair
2. Millard's Repair
3. Straight Line Repair
4. Secondary Lip repair

Techniques for bilateral lip repair used in our study

1. Bilateral VEAU III repair
2. Bilateral Millard's repair
3. Secondary lip repair bilateral

Techniques for palate & palatal fistula repair

1. Pushback palatoplasty
2. Doyen's flap
3. Fistuloplasty

Follow up

Patients of cleft lip were generally discharged on the 2nd or 3rd postop day in case there was no complication. In case of complications patient were hospitalized till they were fit to be discharged. They were called for follow-up on 7th post-op day for suture removal and then monthly up to 3 months. On follow-up patients were evaluated for:

- Scar – normal or abnormal (hypertrophic or contracted)
- Persisting nasal deformity if any
- Any other complaints by the patient or attendants

Patients of cleft palate were discharge on the 3rd or 4th postop day in case there was no complication. In case of complications patient were hospitalized till they were fit to be discharged. Patients were called for follow-up after 2 to 3 weeks and then monthly upto 2 months and then 3 to 6 months interval. On follow-up they were evaluated for

- Any fistula
- Palatal movement
- Clinical speech assessment

Postoperative assessment and patient/family satisfaction was recorded on an investigator designed proforma, after obtaining an informed consent of the patient or guardian.

Subjective analysis of patients in terms of their satisfaction after surgery were also recorded.

The data collected was then tabulated and analyzed.

Observations

The study was conducted over a period of 1 year. It comprised of 312 patients of Cleft lip, cleft palate, cleft lip-palate and secondary cleft deformities who presented to the department of Plastic

Surgery, Himalayan Institute of Medical Sciences, Swami Ram Nagar, Dehradun. They were subjected to detailed history and thorough examination as per preformed proforma.

In this study, out of 312 patients the highest incidence was in the age group of 1- 10 years i.e., 183, 63, were in age group of 11-20 yrs. 46 in age group of 0-11 months.17 in age group of 21-30 yrs, while only 3 lie in the age group of 31-40 yrs.

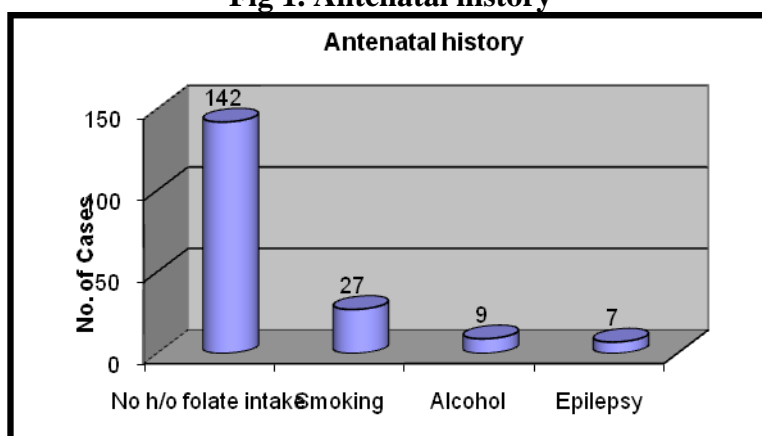
Majority of the cleft patients presenting for cleft treatment were males, male: female ratio in our study was 2:1

Table I : Sex distribution of cases

| Sex | No. of cases |
|---------|--------------|
| Males | 209 |
| Females | 103 |
| Total | 312 |

Out of the study sample of 312 patients, 142 mothers reported that they have not taken any folate supplement during gestation while 170 mothers reported that they have been taking folate tablets regularly through the government and non-government network of maternal and child health service, depicted in fig 1.

Fig 1. Antenatal history



Bilateral clefts were present in 52 (19.26%) cases where as 136 (50.37%) cases had left sided clefts, 82 (30.37%) cases had right sided clefts.

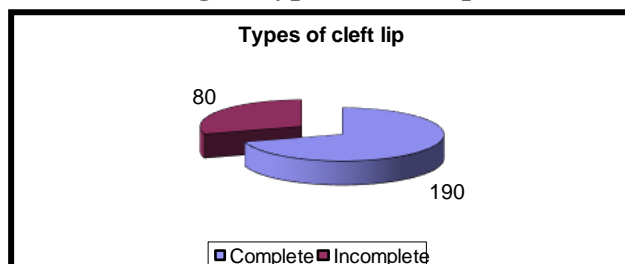
In the study done to see the distribution of the location of clefts; it emerged that clefts of the lip are the most common type of clefts seen in 163 (52.24%) cases, as seen in table II.

Table II: Location of clefts

| CLEFT LOCATION | No. Of Cases |
|----------------------|--------------|
| Cleft lip | 163 |
| Cleft palate | 42 |
| Cleft lip and palate | 107 |
| Total | 312 |

Present study shows complete cleft lip in 190 cases and 80 cases had incomplete type as shown in fig 2.

Fig 2. Types of cleft lip



Various techniques used for cleft repair are depicted in table III

Table III: Surgical techniques used for repair

| | No. of Cases |
|----------------------|--------------|
| Randall – Tennison | 177 |
| Millard's | 4 |
| Bilateral Veau III | 40 |
| Secondary lip repair | 39 |
| Straight line repair | 10 |
| Total | 270 |

In present study, in only 11 cases postoperative bleeding occurred of which, 7 had reactionary bleed, 4 had secondary bleed, dehiscence of repair and formation of fistulae was noted in 2 cases, 2 patients suffered from pulmonary edema at the time of reversal. The unfavourable results of cleft surgery are depicted in table IV

Table IV: Unfavorable results in cleft lip and palate treatment

| CLEFTS | Unfavorable Results | No. Of Cases |
|------------------|-----------------------------------|--------------|
| CLEFT LIP | ▪ Contracted scar | 5 |
| | ▪ Hypertrophied scar | 4 |
| | ▪ Mismatch of vermilion border | 2 |
| | ▪ Vermilion notching | 3 |
| | ▪ Over or under correction of lip | 6 |
| Cleft Palate | ▪ Short palate | 2 |
| | ▪ Poor mobility | 3 |
| | ▪ Unsatisfactory speech | 10 |
| | ▪ Fistula | |
| | ○ Anterior | 1 |
| ○ Posterior | 1 | |

Of 312 clefts repaired 291 (93.26%) cases enjoyed aesthetic and functional satisfaction while 21 case reported some short of dissatisfaction for reasons like scar problem, lengthy treatment and follow-up, projecting teeth or improper tissue alignment in cases of cleft lip repair and in terms of control of nasal regurgitation, speech and improve self esteem in cases of cleft palate repair.

Discussion

Cleft lip and cleft palate are among the most common birth defects. It is a failure of the immigration of neural crest cells resulting in the inability of mesodermal penetration of the soft tissues of the cleft region. Cleft deformities are a challenge to the surgeons since very long. Surgeons through ages have attempted to correct the abnormal anatomical arrangements of the cleft deformities and achieve a normal appearance.

A total of 312 cases were taken with age ranging from 3 month to 40 years. Maximum numbers of patients were in the age group of 1–10 years. The age groups under study in some of the previous studies were 1 day – 26 years (Orkar et al 2002), 10 months–11 years. (Song et al 2006). However, they did not specify the commonly affected age group. It is evident that majority of cleft patients report for treatment by 10 years of age (10, 11)

The sex wise analysis of the number of cleft cases reveals that two-thirds of them were males and one-third were females and such proportions were consistently noticed in the study by Orkar et al (2002). Zhou et al (2006) observed that male: female was 2:1. Study done by David et al shows similar results. (David A Nyberg 1995). It is assumed that many of female cleft patient are not brought to the hospital for treatment at all (6,10,12).

It was noted that the incidence of isolated cleft lip (52.24%) was much higher than isolated cleft palate (13.47%) or cleft lip associated with cleft palate (34.29%); while Richard and William (2000) found isolated cleft lip in 15%, cleft lip and palate in 45% and isolated cleft palate in 40% cases. Zhou et al (2006) similarly found in their study that unilateral clefts were more common than bilateral, with unilateral to bilateral ratio being 10.4: 1. They noticed that left sided defects were more common than right. A study conducted by Blanco- Davila (2003) also suggested that clefts of the left side occurred more often than right and boys were affected more commonly than girls (5,12,13).

Factors like smoking, alcohol intake and epilepsy have been found to be associated with cleft lip and palate. Although no direct link of these factors with cleft deformities is found. Pennell (2002) analyzed in his study that pregnancy in women with epilepsy is associated with increased obstetric risks and increased adverse neonatal outcomes, and the most common major malformations in children born to epileptic mothers are cleft lip/palate, heart defects and neural tube defects. Consistent and moderate association between maternal tobacco smoking during pregnancy and non-syndromic orofacial cleft in infants has been confirmed by Deacon S (2005) (14, 15)

Criteria of selection of a suitable candidate for lip surgery vary from centre to centre. In some parts of the world infants with clefts have been operated upon in local anaesthesia within 48 hours of birth. Lazarus et al (1998) retrospectively compared 5 techniques: Millard's rotation advancement repair, David's Z plasty, modified Z plasty, Tennison Randall triangular flap repair and Nakajima Yoshimure straight line repair. He noticed that the outcome following repair of unilateral cleft lip was similar in all five surgical methods (16). In our study it was noted that Randall Tennison's triangular flap technique gave the most satisfactory result in complete as well as incomplete clefts, narrow as well as wide, unilateral clefts of the lip. There was minimal contraction of broken-scar and the lack of normal-like philtral architecture was not significant. However, the few cases operated by Millard's technique suffered from scar contraction in our study. Adults with repaired cleft lip and palate often do not achieve fully normal aesthetic and functional status in spite of multiple surgical procedures performed during childhood and adolescence. These persons often have a visual blemish or a speech problem that continues to identify them as different (17).

Sameul and Philip (2006) evaluated 77 complete cleft lip patients, in view of need for revision surgery. 64% of their patients had residual nasal deformity, 10% had mismatch of vermilion border and 1% had hypertrophic scarring. The most common problem encountered in that series was some excess fullness of the free border of the lip and inadequate rotation of Cupid's bow. Our study also has similar findings (18).

Majority of children recover quickly from cleft palate repair and the incidence of complications is low in experienced hands. Airway obstruction and bleeding are the two most dangerous complications.

It is evident in our study that more than 90% families enjoyed satisfaction following cleft repair. Landsberg et al (2006) also assessed the satisfaction of cleft patients with the treatment result and facial appearance (19). In a similar study conducted by Sameul and Philip (2006), 85% patients were satisfied following the repair of cleft.

Conclusion

It needs to be emphasized to the general masses, that these deformities are correctable provided the patients get the right treatment at the right time. There is a need to spread awareness in the society about the factors which are responsible for cleft lip and palate so that the various myths can be dispelled, and the correctable causative factors can be taken care of. This study depicts a reference for future workers on clefts, who intend to take a state wise regional demographic study on cleft patients, on a much larger scale.

Bibliography

1. Philip Stamier, Gudrum E Moore. Genetics of cleft lip and palate: Syndromic genes contribute to the incidence of non-syndromic clefts. 2003; 12. Available from URL: www.highwirepress.com.
2. Olasoji HO, Ukiri OE, Yohaya A. Incidence and aetiology of oral clefts: a review. *Afr J Med Sci* 2005; 34:1-7.
3. Gregg T, Richardson A. Dental development, orthodontics, cleft lip and palate. In: David A Adams, Michael J Cinnamon, editor. *Scott - Brown's Otolaryngology*. 6th ed. London: Butterworth Heinemann; 1997; 6:1 – 35.
4. S. Riva Raju. In search of a smile: Study of children born with Cleft Lips and palates in India. Tata Institute of social sciences. Available from URL; www.smiletrain.com.
5. Richard PJ, William PS. Cleft lip and palate. Developmental abnormalities of the face, Palate, jaws and teeth. In: Russel RCG, Williams NS, Bulstrode CJK, editor. *Bailey & love's short practice of surgery*. 23rd ed. London: Arnold; 2000: 586-9.
6. David AN, Kimberly GS, Friederick NH, Dauna JK, Robert JK. Fetal cleft lip with or without cleft palate: US classification and correlation with outcome. *Radiology* 1995; 195: 677-84.
7. Gullino E, Serra M, Ansaldi C, Massobrio M, Pagliano M. Bilateral cleft lip and palate diagnosed sonographically at 11 weeks of pregnancy. *J Clin Ultrasound*. 2006; 34(8):398-401.
8. Orlagh H, Donald B, Peter H, Chris J. The psychosocial effects of cleft lip and palate-a systemic review. *European journal of Orthodontics* 2005; 27: 274-83.
9. Vasan N. Management of children with clefts of the lip or palate: an overview. *NZ Dent J* 1999; 95(419): 14-20.
10. Orkar KS, Ugwu BT, Momoh JT. Cleft lip and palate: the Jos experience. *East Afr Med J* 2002; 79(10): 510-3.
11. Song O, Shi B, Zheng Q. Study on repair approach of bilateral cleft lips and nose deformity. *Zhongguo Xiu Fu Chong Jian Wai Ke Za Zhi* 2006; 20(9): 899-901.
12. Zhou OJ, Shi B, Shi ZD, Zheng O, Wang Y. Survey of the patients with cleft lip and palate in China who were funded for surgery by the Smile Train Program from 2000 to 2002. *Chin Med J (Engl)* 2006; 119 (20): 1695-700.
13. Blanco-Davis, Feliciano. Incidence of cleft Lip and Palate in the northwest of Mexico: A 10-Year study. *Journal of Craniofacial Surgery*. 2003; 14 (4):533-547.
14. Deason S. Maternal smoking during pregnancy is associated with a higher risk of non-syndromic orofacial clefts in infants. *Evid Based Dent*. 2005; 6(2):43-4.
15. Haapanen ML, Rintala AE. Comparison of quality of speech after Veau – Wardill – Kilner pushback operation and the Cronin modification in the primary treatment of cleft palate. *Scand J Plast Surg Hand Surg*. 1993; 27(2): 113-8.
16. Lazarus DD, Hudson DA, Vanzyj JE, Fleming AN, Fernandes D. Repair of unilateral cleft lip: a comparison of five techniques. *Ann Plas Surg* 1998; 41(6): 587-94.
17. Richard P. Strauss, Hillary Broder. Interdisciplinary team care of cleft lip and palate: Social and psychological aspects. *Clinics in Plastic Surgery* 1985; 12 (4):543-51
18. Samuel MN, Philip KTC. Unilateral Cheiloplasty. In: Stephen J. Mathes editor. *Plastic Surgery*. 2nd ed. Philadelphia: Saunders Elsevier; 2006 P 165-215.
19. Landsberger P, Proff P, Dietze S, Hoffmann A, Kaduk W, Meyer FU, Mack F. Evaluation of patient satisfaction after therapy of unilateral clefts of lip, alveolus and palate. *J Craniomaxillofac Surg* 2006; 34(2): 31-3.