

Management of Commonly Encountered Secondary Cleft Deformities of Face-A Case Series

JACOB JOHN, ARJUN MADHU USHA, MUBARAK AZIZ, VINIT RENKAN ACHARY, HARI KRISHNAN

ABSTRACT

Orofacial clefts are one of the most common congenital deformities occurring due to disturbances during the embryological formation, development and growth of orofacial region. The treatment of cleft deformities in the early period of life is mandatory to address the aesthetic, functional and psychological problems affecting the child. A considerable number of surgical modalities for definitive correction of unilateral and bilateral cleft lip, nose, and eyelid deformities have been reported over the past half century. After the initial cleft repair, there is a long period of dramatic growth. This powerful variable of growth may ultimately distort the immediate surgical result. Hence, the correction of secondary deformities plays a very important part in the care of these patients. In this article we are enlisting some simple techniques to correct the commonly encountered secondary cleft deformities of face that gave acceptable results to the patients. These corrective surgeries not only improved the function and aesthetics but also increased the confidence level, satisfaction and overall quality of life of the patient.

Keywords: Bilateral cleft lip, Cleft of eye, Orofacial clefts

Clefts of face are one of the most common congenital deformities occurring due to disturbances in developmental process during intrauterine life. The global prevalence of orofacial clefts is one affected individual in every 600 new born babies [1]. It is necessary to treat the deformity at early stage of life as it aid in reducing the psychological problems created by cleft both in the child and parent and there by changing the plight of the patient and facilitate aesthetics and function.

A considerable number of surgical modalities for definitive correction of unilateral and bilateral cleft lip, nose, and eyelid deformities have been reported over the past half century. Generally, it is argued that a clear understanding of the associated complex anatomical and pathological abnormalities is required to obtain a desirable facial form. The cleft deformities of face can be considered area wise into lip, nose and eyelid. The abnormalities include all components of the face, including the facial skeleton, cartilage, muscle, skin, subcutaneous tissue, and mucosal lining. To obtain desirable and stable outcomes these deformities should be approached in each of the above components.

Secondary deformities of the clefts are the rule rather than the exception [2]. After the initial cleft repair, there is a long period of dramatic growth, this powerful variable of growth may ultimately distort the immediate surgical result. Hence, the correction of these secondary deformities are indeed mandatory for the care of these patients and the procedures are usually more complicated than the initial surgery [3,4]. The problems are so widely varied that the choice of an appropriate technique to correct them is challenging [2]. In this article the surgical correction of commonly encountered secondary cleft deformities of nose, lip and eyelid has been discussed, using simpler techniques.

CASE 1

Secondary Cleft Rhinoplasty

The most common secondary nasal deformities of cleft lip includes a retro displaced dome of the ipsilateral nasal tip, alar columellar web, hooding of the alar rim, short columella, nasal tip deformities and other deficiencies [5-7] .

Patient 1: A 22-year-old female patient reported to the OPD

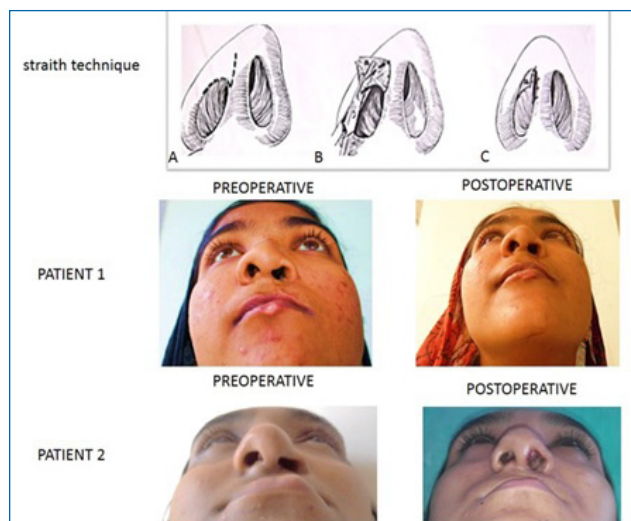
with a chief complaint of asymmetry of nose and depression of nose towards the left side. Patient gave a history of unilateral left complete cleft lip and palate, for which surgical correction of cleft lip was performed at the age of 10 weeks and for cleft palate at one year and nine months in a different center. Patient had no relevant medical history and family history, parents were of non-consanguineous marriage. Examination showed a linear vertical scar over the left side of the lip extending into the philtral column and base of the nose with lack of nasal support on left side with widening of ala and alar columellar web.

Patient 2: A 21-year-old female patient reported to the OPD with a chief complaint of depression of nose on the left side with asymmetry of nose. Patient was very much concerned about the esthetic deformity of the nose. Patient gave a history of unilateral left complete cleft and palate. Surgical correction of cleft lip was performed at the age of 10 weeks and for cleft palate one year and eight months. Patient had no relevant medical history and family history, parents were of non-consanguineous marriage. Examination showed a vertical scar over the left side of the lip extending into the philtral column. Lack of nasal support at the base of the nose was present. Widening of ala and alar columellar web was evident on the left side.

The correction of alar columellar web can be done by different techniques. We performed Straith technique of modified Z-plasty in both the cases for the correction of alar columellar web, because the procedure is simple with predictable and acceptable results, after obtaining written informed consent from the patients.

Technique:

a) Outline of the incision was made,



[Table/Fig-1]: Secondary cleft rhinoplasty- Straith technique of modified Z-plasty was performed in both the cases.

- Skin flap was raised exposing the cartilage, exposed alar cartilage was excised and the remaining mucosal flap was incised, rotated anteromedially, trimmed and sutured to the columella.
- The skin flap was rotated into the vestibule and sutured to the lateral wall.

Follow-up after six months for the first patient and after ten months for the second patient showed good aesthetic result with minimal relapse [Table/Fig-1].

CASE 2

Secondary Cleft Rhinoplasty

A female patient aged 26 years reported to the OPD with a chief complaint of depression of nose towards her left side and wide nasal opening on the left side. Patient gave a history of unilateral left complete cleft lip and palate, for which surgical correction of cleft lip was performed at the age of 11 weeks and for cleft palate one year and ten months. Patient had no relevant medical history and family history, parents were of non-consanguineous marriage. Examination showed a widening of alar base and alar columellar web. A scar over the left side of upper lip extending into the philtrum was observed. After obtaining a written informed consent from the patient, correction of nasal tip deformity was done by means of cartilage or bone augmentation by open rhinoplasty method as adequate nasal projection was not obtained by suspension and reposition of alar cartilage.

Technique:

- Incisions were marked. An inverted V mid columellar incision with bilateral marginal incision was made and the lower lateral cartilage and septal cartilages were separated.
- Bone graft from the anterior maxilla was harvested.
- The bone graft was inserted in a pocket between the lower lateral cartilages towards the columella of the nose. The graft provides support for the depressed ala and augments the nasal sill in addition to the effects on the nasal tip.



[Table/Fig-2]: Secondary cleft rhinoplasty-nasal tip deformity correction was done by means of cartilage or bone augmentation by open rhinoplasty method.

- d) Bone graft secured to the medial crura of both alar cartilages by non-absorbable sutures and closure is done.

Patient is on follow-up for the past two years, some resorption of the bone graft was observed [Table/Fig-2].

CASE 3

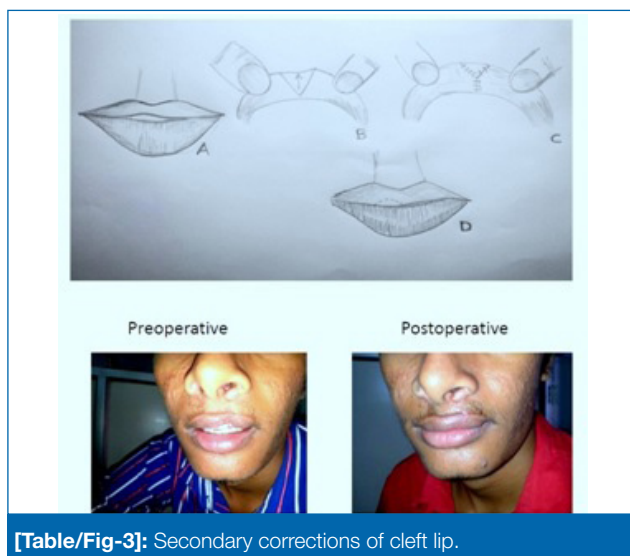
Secondary Corrections of Cleft Lip

The most common secondary deformity of the vermilion in unilateral cleft lip cases is the whistle deformity or the central notching defect on the lips and cupid's bow deficiency. A whistle deformity is defined as the central vermilion notching due to complex causes and appears as if the person is blowing a whistle [8]. Cupid's bow deficiency is the discrepancy between the heights from the central point of the base of columella to the two peaks of the cupid's bow.

A 21-year-old male patient reported to our OPD with a chief complaint of asymmetry of width of the upper lip on the left side. Patient gave a history of unilateral left complete cleft lip, for which surgical correction of cleft lip was performed at the age of 12 weeks in a different institute. Patient had no relevant medical history and family history, parents were of non-consanguineous marriage. Examination showed an incompetency present on the left lip with exposure of teeth. A linear scar was present on the left side of the upper lip which was extending up to the base of the ala. Correction of whistle deformity of lip was done by the following technique after obtaining a written informed consent from the patient.

Technique:

- A V-flap based in the vermilion-cutaneous border was designed.
- Undermining was done around 0.75 cm in all directions. The flap was advanced towards its base and repositioning



[Table/Fig-3]: Secondary corrections of cleft lip.

of orbicularis oris muscle was done depending on the amount of deficiency.

- Flap closed into a V-Y fashion. The technique results in vertical scar in the wet mucosa with over all lengthening of the central vermilion. Closure was done with interrupted absorbable suture.

Patient was followed-up for six months and the post-operative results showed good aesthetic correction [Table/Fig-3].

CASE 4

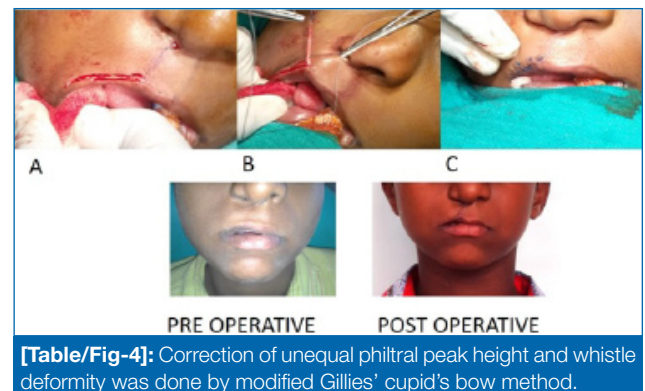
A male patient aged 11 years reported to our OPD with a chief complaint of deformity in the upper lip on the right side. He had esthetic concern regarding shape of the lip. Patient gave a history of unilateral left complete cleft lip, for which surgical correction was performed at 9th week of life, in a different institute. Patient had no relevant medical history and family history, parents were non consanguineous. On examination, a vertical scar was present in the philtral region.

Correction of unequal philtral peak height and whistle deformity was done by modified Gillies' cupid's bow operation.

Technique:

- Myocutaneous flap was designed above the deficient area.
- The flap was raised from the lateral upper margin of the lip, de-epithelialized and tunneled under the right philtrum and sutured in position, that equalizes the height of both philtrum.
- V-Y plasty was also done to correct the whistle deformity. Resultant scar on the lateral upper margin of the lip will become white roll which gives continuity to the white roll.

Post-operatively patient came for review for first the two weeks and failed to report back [Table/Fig-4].



[Table/Fig-4]: Correction of unequal philtral peak height and whistle deformity was done by modified Gillies' cupid's bow method.

CASE 5

Coloboma Correction

A 20-year-old male patient reported to the OPD with the complaint of deformity in the left lower eye lid with inability

to close the eye lid completely. Patient gave a history of Treacher-Collin's syndrome. Patient had no relevant medical history and family history, parents were non consanguineous. On examination coloboma of left lower eye lid was present. An eyelid coloboma is a full-thickness defect of the eyelid. Colobomas can occur as associated with congenital anomalies or due to trauma and it can be either a small notch or even a complete absence of eyelid. Upper eyelid is most commonly affected. An eyelid coloboma is an almost constant feature of Treacher-Collin's syndrome, in this syndrome the lower eye lid is mostly affected.

Coloboma correction in this case was achieved by Tessier Z-plasty and lateral canthopexy.

Technique:

- Outline for skin incision were marked .
- Marginal excision and incision for a z-plasty for cutaneous lengthening was given.
- Overlapping suture of the preseptal orbicularis muscle was given. Transposition and suture of the skin flaps with a lateral canthopexy was done.

Patient is on regular follow-up for the past one year [Table/ Fig-5].



[Table/Fig-5]: Coloboma correction achieved by Tessier Z-plasty and lateral canthopexy.

DISCUSSION

The development of face is a complex process, which involves the fusion of frontonasal process, maxillary, mandibular, primary and secondary palatal processes. Any defects during the fusion of these processes results in clefts. Various timings for primary lip and palate repair have been established but no single technique can restore the normal anatomical, functional and aesthetic values of face, thus requiring secondary surgeries. Generally for both unilateral and bilateral deformity of cleft lip, correction can be done at the time of initial cleft lip surgery itself [9]. A secondary surgery is necessary to modify the deformity of nasal shape and a large number of patient's desire complete septorhinoplasty in their early years [10].

One of the cleft lip nasal deformities is the alar-columellar

web deformity. Saylor KE, has proposed few principles for the correction of the nasal deformity due to cleft lip. They are-

- (1) In cases of severe early deformity, secondary correction should be done earlier.
- (2) Nasal deformity is addressed to improve the form, function and to reduce the psychological stress to the patient.
- (3) The skeletal base, the septum, the tip, and the alae should be corrected.
- (4) Bone grafting and cartilage augmentation may be required in few cases.
- (5) Definitive rhinoplasty can be done only at 14 years of age or older; and
- (6) Definitive rhinoplasty is a contraindication in patients with severe skeletal base asymmetry [11].

A reverse-U incision with a suture suspension of the repositioned cleft cartilage was given by Tajima and Maruyama. Nakajima T et al., introduced a new technique of Z-plasty in the lateral nasal vestibule to correct the alar-columellar web after the rotation of the mucochondrial flap [6]. A V-Y plasty of the nasal mucosa by the advancement of the lateral crus was described by Cronin and Denkler [7]. A bilateral reverse-U incision in combination with Cronin's method was proposed by Meada K et al., [12]. A bilateral reverse-U incision combined with a short banked forked flap was advocated by Nakajima and Yoshimura [6].

Secondary correction of the lip is required to restore normal lip symmetry, to improve functions, and to maintain proper lip seal [13,14].

Several techniques for secondary cleft lip repair have been described in literature. Abbe flap reconstruction technique, in which a full thickness flap of tissue from the lower lip is transferred to the upper lip. Donor site trauma is created and a mismatch is observed in texture and color of the upper lip. Another simple method is transposition of bilateral lateral vermilion border flaps into the midposterior line of the prolabium to correct the "whistling lip" deformity proposed by Matsuo K et al., [15]. The main drawback is the difference in color between the dry and wet mucosa and the development of additional scars in the vermilion. Patel and Hall used a dermis fat graft to augment the free border of the lip to correct the vermilion notch deficiency [16]. Disadvantage is the degree of resorption of the graft. A modification of Kapetansky's axial-based pendulum flap to correct the deficiencies of central lip was described by Grewal NS et al., in both unilateral and bilateral cleft lip patients [14]. Yin NB et al., advocated the "curved-line method" for secondary bilateral cleft lip deformities [17]. For the correction of the complex interrelated secondary deformities of the lip, the nose, and eyes, a careful and systematic evaluation of the deformity, an integrated treatment plan and a high quality surgical technique is required.

CONCLUSION

Secondary deformities are common in children born with clefts. They are both intrinsic to the malformation and iatrogenic. Correction of secondary deformities of cleft should be always advocated to improve the quality of life of primary corrected cleft patients. In this article some simple techniques of secondary cleft corrections were used that gave acceptable results to the patients. These corrective surgeries not only improved the function and aesthetics of the patient but also increased the confidence level, satisfaction and overall quality of life of the patient.

Ethical Clearance Statement

All procedures performed in studies involving human participants were in accordance with the ethical standards of the institutional and with the 1964 Helsinki declaration and its later amendments or comparable ethical standards.

REFERENCES

- [1] Mossey P, Little J .Addressing the challenges of cleft lip and palate research in India. *Indian J Plast Surg.* 2009;42Suppl:S9-S18.
- [2] Stal S and Hollier L. Correction of secondary cleft lip deformities. *Plastic and reconstructive surgery.* 2002;109:1672-81.
- [3] Klaus & Fanaroff. *Care of the high-risk neonate.* Saunders. 5th edition. 2001 USA. p. 32-34
- [4] Colin D. Rudolph. In *Rudolph's Pediatrics.* McGraw-Hill. 21st edition. 2002. USA. p. 956-62.
- [5] Tajima S, Maruyama M. Reverse-U incision for secondary repair of cleft lip nose. *Plast Reconstr Surg.* 1977;60:256-61.
- [6] Nakajima T, Yoshimura Y, Kami T. Refinement of the "reverse-U" incision for the repair of cleft lip nose deformity. *Br J Plast Surg.* 1986;39:345-51.
- [7] Cronin TD, Denkler KA. Correction of the unilateral cleft lip nose. *Plast Reconstr Surg.* 1988;82:419-32.
- [8] Choi WY, Yang JY, Kim GB, Han YJ. Surgical correction of whistle deformity using cross-muscle flap in secondary cleft lip. *Archives of Plastic Surgery.* 2012;39(5):470-476.
- [9] Christensen K. Cleft-twin sets in Finland 2001. *Cleft Palate Craniofac J.* 1996;33:530.
- [10] Ahamed AS and Ramadan MM. Correction of secondary deformities of cleft lip nose. *Suez Canaluniv Med J.* 2008;11:01-07.
- [11] Salyer KE. Primary correction of the unilateral cleft lip nose: A 15-year experience. *Plast Reconstr Surg.* 1986;77:558-66.
- [12] Maeda K, Ojimi H, Yoshida T. A new method of secondary correction of the bilateral cleft lip nose. *Br J Plast Surg.* 1987;40:52-60.
- [13] Choi WY, Yang JY, Kim GB, Han YJ. Surgical correction of whistle deformity using cross-muscle flap in secondary cleft lip. *Archives of Plastic Surgery.* 2012;39(5):470-76.
- [14] Grewal NS, Kawamoto HK, Kumar AR, Correa B, Desrosiers AE 3rd, Bradley JP. Correction of secondary cleft lip deformity: the whistle flap procedure. *PlastReconstr Surg.* 2009;124(5):1590-98.
- [15] Matsuo K, Fujiwara T, Hayashi R, Ishigaki Y, Hirose T. Bilateral lateral vermilion border transposition flaps to correct the "whistling lip" deformity. *Plast Reconstr Surg.* 1993;91:930-35.
- [16] Patel IA, Hall PN. Free dermis-fat graft to correct the whistle deformity in patients with cleft lip. *Br J Plast Surg.* 2004;57:160-64.
- [17] Yin NB, Gao F, Wang YQ, Song T, Li HD. Correction of secondary bilateral complete cleft lip and whistling deformities: the advance and rearrangement of the bilateral lip tissues. *Aesthetic Plast Surg.* 2011;35:750-55.

AUTHOR(S):

1. Dr. Jacob John
2. Dr. Arjun Madhu Usha
3. Dr. Mubarak Aziz
4. Dr. Vinit Renkan Achary
5. Dr. Hari Krishnan

PARTICULARS OF CONTRIBUTORS:

1. Professor, Department of Oral and Maxillofacial Surgery, Azeezia College of Dental Sciences and Research, Kollam, Kerala, India.
2. Postgraduate Student, Department of Oral and Maxillofacial Surgery, Azeezia College of Dental Sciences and Research, Kollam, Kerala, India.
3. Senior Lecturer, Department of Oral and Maxillofacial Surgery, Azeezia College of Dental Sciences and Research, Kollam, Kerala, India.

4. Postgraduate Student, Department of Oral and Maxillofacial Surgery, Azeezia College of Dental Sciences and Research, Kollam, Kerala, India.
5. Postgraduate Student, Department of Oral and Maxillofacial Surgery, Azeezia College of Dental Sciences and Research, Kollam, Kerala, India.

NAME, ADDRESS, E-MAIL ID OF THE CORRESPONDING AUTHOR:

Dr. Arjun Madhu Usha,
Thushara Miniestate Road, Meppukkada,
Malayinkil-695571, Trivandrum, Kerala, India.
E-mail: drarjunmu@gmail.com

FINANCIAL OR OTHER COMPETING INTERESTS:

None.

Date of Publishing: **Oct 01, 2017**