

Construction of the congenitally missing columella in midline clefts

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Summary

Background

Repair of the rare median cleft lip occurring with agenesis of the columella is a challenge as there is a major deficiency of skin and underlying structures.

Material and methods

Over a 23- year period, five children underwent construction of a columella. A new surgical technique was designed for this: an internal dorsal nasal flap was used to create a cutaneous-cartilaginous flap inside the nasal dome, and was then sutured inferiorly against a superior triangle of the repaired median cleft lip.

Results

Projective nasal growth could be seen in three of the five surgically constructed columellae in whom the medium-term results could be evaluated.

Conclusion

Five holoprosencephaly cases, all with a median cleft lip and agenesis of the columella, underwent a median cleft lip repair and the creation of a columella by means of an internal nasal dorsal skin flap procedure.

Introduction

Congenital agenesis of the columella is extremely rare and it seems as though only six cases have been reported in the literature (*Putman and Postlethwaite, 1994; Bilkay et al., 2004*). This very important aesthetic entity of the midface may be missing congenitally (*Conley et al., 1983; Mavili and Akyürek, 2000; Bilkay et al., 2004*), or due to vascular malformations (*Paletta and Van Norman, 1962; Vecchione, 1980*), tumour resection (*Kaplan, 1972; Saad and Barron, 1980; Nicolai, 1982; Conley et al., 1983; Bianchi et al., 1984; Baker and Swanson, 1985; Puterman et al., 1985; Earley and Chantarasak, 1989; Özkus et al., 1992; Neto et al., 1999; Okazaki and Ueda, 2003*), traumatic loss (*Paletta and Van Norman, 1962; Saad and Barron, 1980; Conley et al., 1983*), and acquired loss due to other causes (*Yanai et al., 1986; Güçer, 2002*) as well as by infectious disease (*Cardoso, 1958; Paletta and Van Norman, 1962; Vecchione, 1980; Nicolai, 1982*).

Midline defects, such as cleft lip and agenesis of the columella, also occur in combination with brain defects, which are often incompatible with life. A number of patients, who have survived after birth may present at a cleft lip and palate clinic. Most of these cases are suffering from holoprosencephaly (*Putman and Postlethwaite, 1994*). As the life expectancy of these patients is generally very limited, very few cases can be surgically repaired and followed up.

Numerous techniques have been developed to create a columella for alleviation of this severe blemish, normally in the way of local flaps: alar interpositional (*Bianchi et al., 1984*) or marginal flaps (*Saad and Barron, 1980*), internal nasal vestibular flaps (*Vecchione, 1980; Mavili and Akyürek, 2000*), nasolabial flaps (*Paletta and Van Norman, 1962; Nicolai, 1982; Conley et al., 1983; Yanai et al., 1986; Özkus et al., 1992*), nasal septal flap (*Puterman et al., 1985; Bilkay et al., 2004*), nasal malar flaps (*Kaplan, 1972*), transverse forked flap (*Earley and Chantarasak, 1989*) and extended Abbé flap (*Putman and Postlethwaite, 1994; Okazaki and Ueda, 2003*). Even extended flaps have been used: forehead flap (*Cardoso, 1958; Paletta and Van Norman, 1962; Baker and Swanson, 1985*), island-shaped depressor anguli flap (*Neto et al., 1999*), multi-stage pedicled cervical tube flap (*Paletta and Van Norman, 1962; Mavili and Akyürek, 2000*), multi-stage composite pedicled ear flap (*Güçer, 2002*) and hand flap (*Puterman et al., 1985*). Composite free flaps, such as an ear lobe graft (*Paletta and Van Norman, 1962*) have also been reported. Most of these surgical techniques were developed for loss of the columella, such as by tumour resections. They have seldom been used for the congenital agenesis of the columella (*Putman and Postlethwaite, 1994; Okazaki and Ueda, 2003*).

This report describes a single stage repair of the midline cleft lip with an internal nasal dorsal flap for the creation of a columella. There also was unexpected nasal growth of the nose.

Material and methods

Database

The database of this cleft lip and palate clinic was analysed for incidence of median cleft, as well as its presentation in various subdivisions of cleft lip (CL), cleft lip and alveolus (CLA) and cleft lip, alveolus and palate (CLAP). Furthermore, patients with median clefts and agenesis of the columella were also identified (Fig. 1).

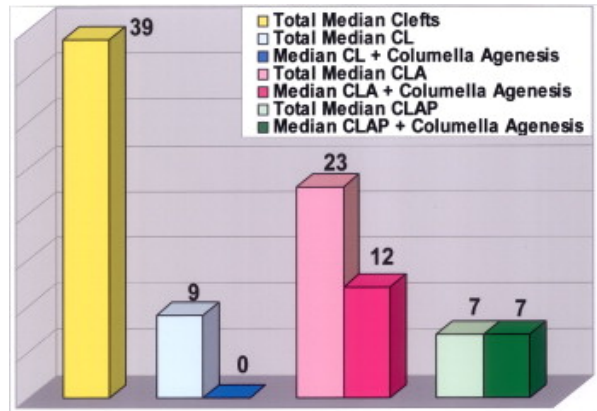


Fig. 1. Total median clefts as median cleft lip, median cleft lip and alveolus, or median cleft lip, alveolus and palate in combination with agenesis of the columella.

Surgical technique

The surgical technique consisted of creation of a dorsal skin flap from inside the nasal dome (Fig. 2a), which became the new columella and which was rotated and sutured in a v-y join inferiorly against the superior triangle of the repaired median cleft lip. The surgical incision for the internal nasal dorsal flap was made in the cutaneous and cartilaginous parts of the inside of the nasal dome. Small sections of the central or medial parts of the alar and lateral nasal cartilages were elevated together with the skin flap, which was pedicled at the nasal dome. The donor area, with its adjacent alar and lateral nasal cartilages of the nose, was then approximated and sutured in the anterior part of the nose (Fig. 2b). The inferior part of the newly created columella, designed as a fork flap, was sutured into the superior triangle of the repaired median cleft lip. Closure of the median cleft lip was performed by means of a straight-line closure, with a vermilion-skin triangle and an upper lip triangle. The lip and the columella were connected by means of a triangular flap (the sharp point was the superior part of the repaired median cleft lip) wedged between the forked columella flap (Fig. 2c). This enabled the creation of a columella within the lip repair.

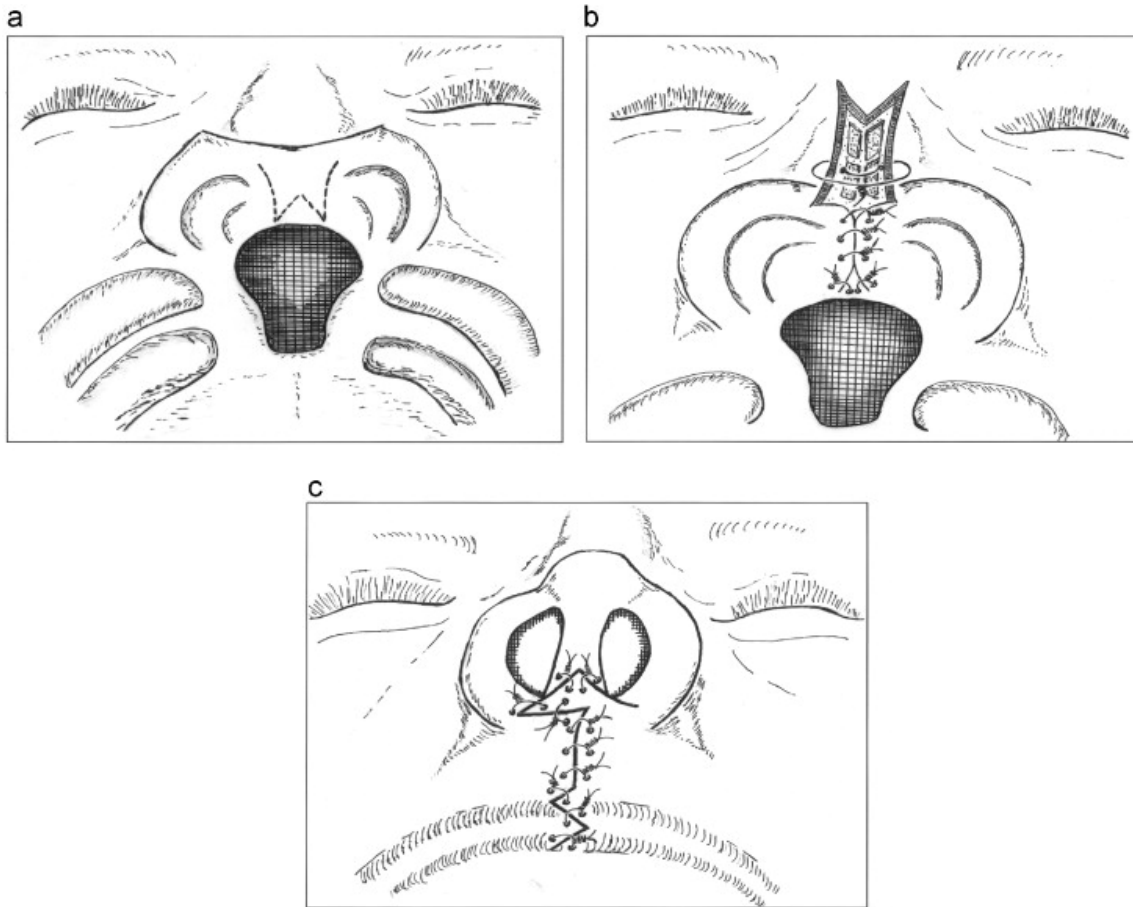


Fig. 2. (a) The outline of the intra-nasal dorsal flap for creation of the columella; (b) columella, as pedicled flap at the nasal dorsum; (c) the created columella and the median cleft lip closure.

Results

The analysis of the 2905 cleft lip and palate patients revealed that a total of 39 (2.2%) cases of the 1777 different cleft lip deformities, presented as a median cleft deformity.

The various subdivisions of this cleft accounted for: nine cleft lips (CL; 0.51%), 23 cleft lip and alveolus patients (CLA; 1.29%) and seven cleft lip, alveolus and palate cases (CLAP; 0.39%; Fig. 1). Nineteen of these 39 median cleft cases presented with additional agenesis of the columella, and all were diagnosed as holoprosencephaly. However, there is a variation in the appearance of a median cleft with agenesis of the columella (Fig. 3).

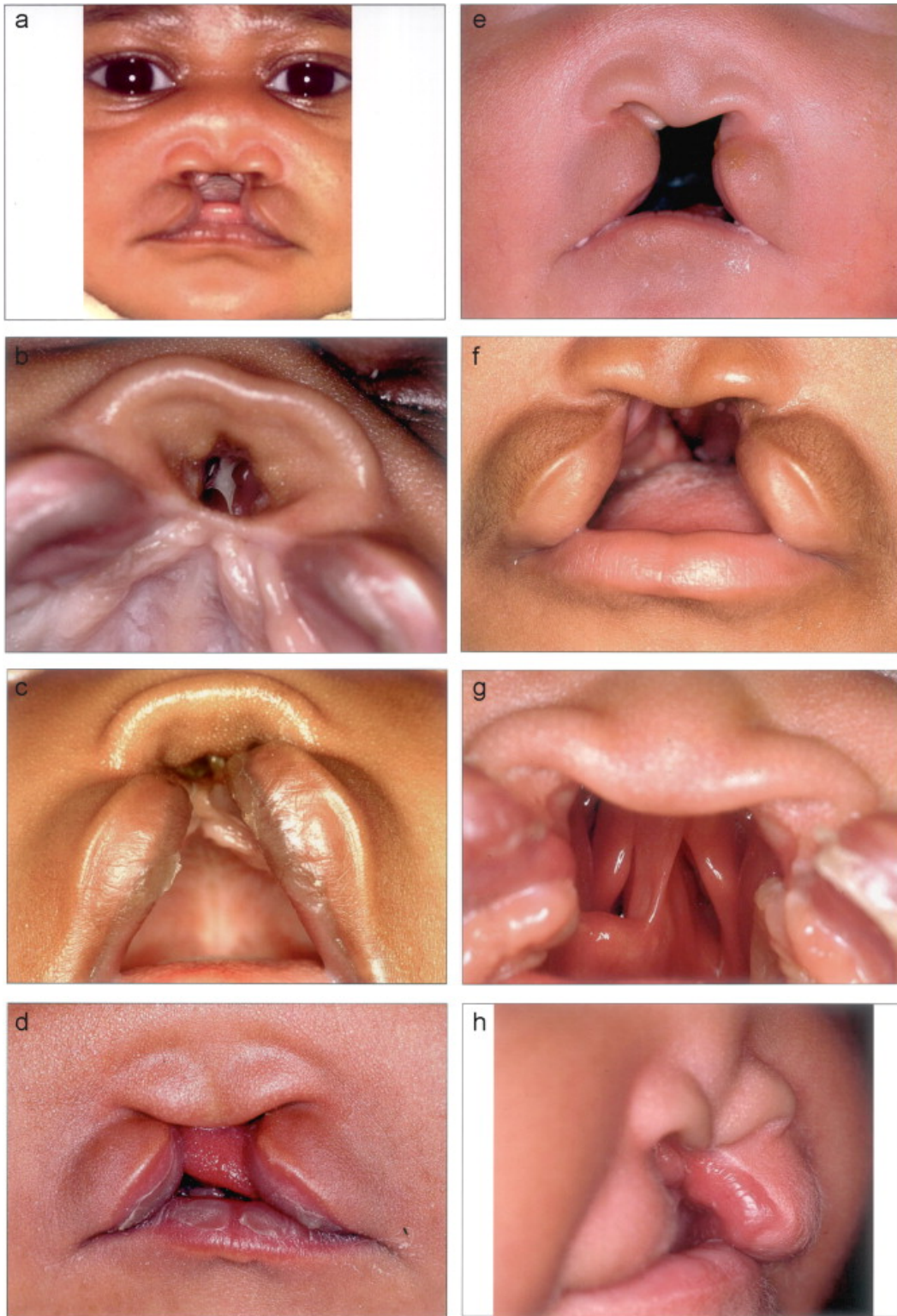


Fig. 3. (a–h) Eight variations in the appearance of a median cleft lip (CLA or CLAP) with an agenesis of the columella.

Five children underwent repair of the median cleft lip as described here, combined with the creation of a columella (Fig. 4). It was possible to evaluate the nasal growth in three of these five over a longer period, and it was a surprise to see that some kind of a projective growth of the nose and midface followed (Fig. 5).

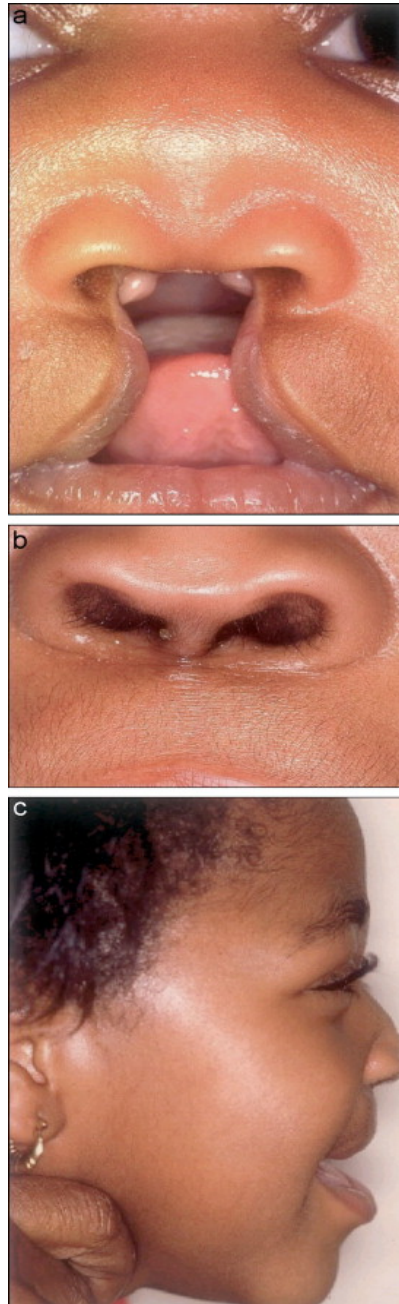


Fig. 4. (a–c) Pre-operative and long-term post-operative appearance of a median cleft lip (CLA) with agenesis of the columella.

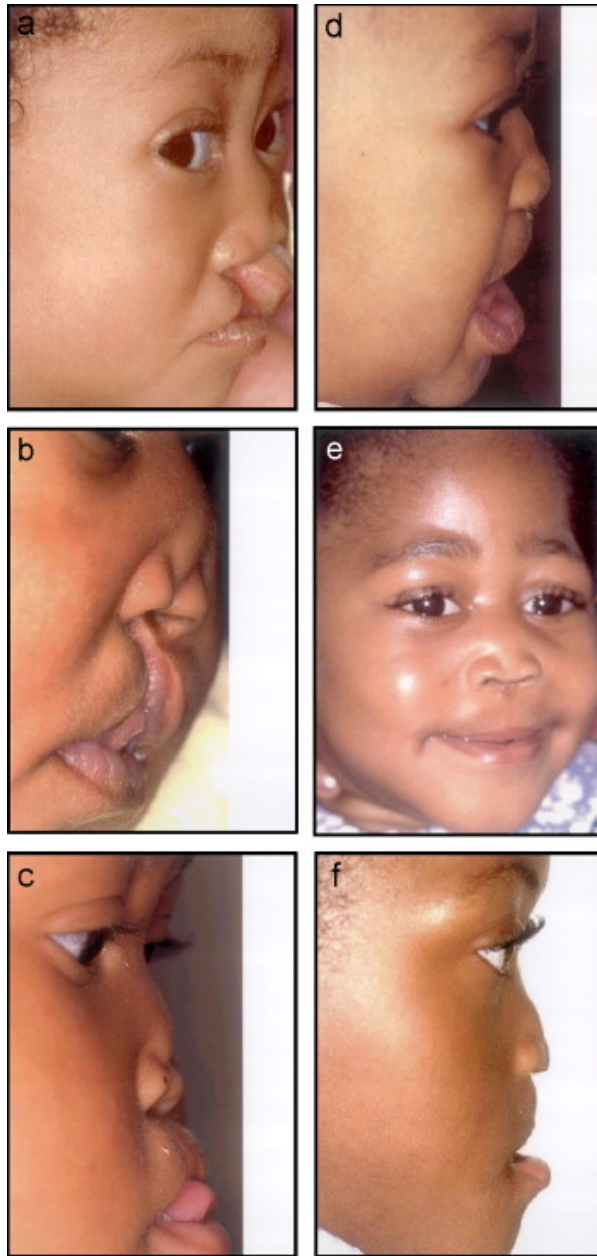


Fig. 5. (a–f) Three patients with pre-operative and post-operative appearances of midface and nose after creation of columella and closure of median cleft lip. Note projection of nasal tip in (d) and (f).

A single complication occurred in one of the five children: The newly created columella had to be disconnected from the superior part of the lip, i.e. at its insertion in the repaired median cleft lip due to post-operative breathing restriction. Breathing was immediately re-established. This mentally handicapped child had not learned how to breathe orally.

Discussion

The columella is an important aesthetic unit of the midface. Its absence is glaringly obvious. It is therefore quite surprising that up until 2004, congenital agenesis has apparently only been reported in six cases (*Bilkay et al., 2004*). Nineteen cases of congenital agenesis of the columella have been listed in just this database. All these patients have also been diagnosed as holoprosencephaly cases. The incidence of congenital columella agenesis in 2905 facial cleft deformities was therefore 0.65%. These cases presented with variations in their appearance. None of them had agenesis of the columella combined with just a median cleft lip. The majority presented with a cleft lip and alveolus (12 cases), or (the remaining seven cases) as a cleft lip, alveolus and palate. The alveolus was therefore involved in all cases as an agenesis of the premaxilla. The cartilaginous part of the nasal septum also showed agenesis. A 20th patient with holoprosencephaly (CLA), not counted under the 39 median clefts, had partial agenesis of the columella of the left side only with a missing premaxilla, a so-called pseudomedian cleft. Aggenesis of the prolabium, columella, nasal septum and premaxilla, is the term used to describe a deficiency of midline structures, and should not be confused with a bilateral cleft, where the midline structures are present, but are not fused to the lateral elements.

Surgical construction of a missing columella is aesthetically challenging. (*Bianchi et al., 1984*). In cases with a median or pseudomedian cleft lip, the deficiency of the upper lip, including the deficiency of the premaxilla and part of the nasal structures, challenges the surgeon to construct the most natural-looking nose/lip configuration. There is no particular procedure as the first choice for this type of repair in the literature. The difficulty of creating this subtle aesthetic keystone has stimulated many elaborate surgical techniques sometimes requiring multiple stages (*Vecchione, 1980*). The combination of median cleft lip and agenesis of the columella, has therefore mostly been repaired by means of an extended Abbé flap (*Putman and Postlethwaite, 1994; Okazaki and Ueda, 2003*).

Although the life expectancy of these holoprosencephaly patients is relatively limited (*Putman and Postlethwaite, 1994*), there are three reasons why they would be offered surgery: those children who stay alive and healthy for more than a year so they can withstand surgery more readily; those whose parents have been informed of the long-term prognosis and who still desire the external facial correction; and those where a church baptism or equivalent is planned, and the child will be presented to the wider community.

The internal nasal dorsal skin flap for creating a columella has several advantages. The main advantage is that the columella may be created at the same time that the cleft lip is closed. This is a relatively small procedure for a potentially compromised child usually suffering from holoprosencephaly. Any major reconstruction or multiple reconstructions could compromise the patient's survival during the peri-operative phase. The other advantage is that an excellent tissue texture and colour match at the recipient site is possible, with good support of the nasal dome, and no abnormal bulkiness (*Güçer, 2002*) of the columella created.

The disadvantages of this design are: the repair of the median cleft lip, as well as the creation of a columella, may lead to breathing problems as may be seen in any other

repaired cleft patient. There is no nasal septum that can be connected to the columella during the same procedure, so there is a hole instead of a septum dividing the two nasal cavities. The missing nasal septum therefore has to be reconstructed during a second procedure. Bilateral vestibular labial mucosal flaps (Bilkay et al., 2004), may be used for this purpose as soon as the patient's long-term life expectancy has been determined.

Some favourable projective growth of the nasal structure, as well as of the midface was observed, in those three cases who could be followed up.

Conclusion

It is a challenge to the surgeon to construct the most natural-looking labio-nasal junction in a patient born with a median cleft lip, combined with a deficient premaxilla and missing columella. An internal dorsal nasal skin flap may be used to create a columella. This has several advantages in that the columella is constructed during the same surgical procedure that is used to close the median cleft lip. It is also a relatively small procedure for a potentially compromised child who is often born with holoprosencephaly. In addition, the texture and colour of the donor site is an excellent match with the tissue of the recipient site.

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