The surgical repair of half-nose

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Summary
Absence of half-nose is an extremely rare congenital malformation, which has a devastating impact on the patient and the family. A review of indexed English-language literature found 91 cases of half-nose, including 50 patients with proboscis lateralis. Pathogenesis is not clear, and the reported cases have sporadically occurred. Many aspects must be considered when reconstructing a congenital half-nose, such as timing of surgery, type of tissue to be used and the need to reconstruct nasal airway. The aim of this article is to present personal experience in seven cases of half-nose reconstruction, in order to review the literature regarding to this rare entity, highlighting aspects of incidence, pathogenesis and surgical treatment. Nasal reconstruction was performed at ages of 5–7 years to minimise psychological trauma. Forehead skin demonstrated to be an excellent donor site to re-surface the nose. For the inner lining, contralateral cutaneous nasal flap was our preference. Concerning the nasal framework reconstruction, alar contour was restored using a cartilage graft from the lower portion of ear tragus and concha.

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Absence of half-nose is an extremely rare congenital malformation, which has a devastating impact on the patient and the family. Cleft nose, proboscis lateralis and nasal agenesis are rare; whereas absence of half-nose is also extremely rare. A review of indexed English-language literature found 91 cases of half-nose — 50 of them with proboscis lateralis. There were 31 males and 21 females (39 cases were not defined). With regards to laterality, 37 cases were on the right side and 30 cases on the left side. Twenty cases had isolated half-nose, without other associated anomalies, while 22 had eye and lacrimal system malformations; 22 had orbital malformations, which were associated with cleft lip and palate in nine patients; three other cases presented isolated cleft lip and/or palate and other unusual anomalies such as craniosynostosis,
Pathogenesis is not clear, and the reported cases in the literature have occurred sporadically. It has been suggested that lack of nasal development probably results from growth failure of medial and lateral nasal processes. Mazzola classified craniofacial malformations according to the area of commitment: upper face, midcephalic borderline and lower lateral region of the face. Nasal aplasia belongs to the first group and may be subdivided into three subgroups: arhinia, half-nose and half-nose with proboscis lateralis. Any congenital anomaly of the nose can also be accompanied by other facial and palatal anomalies. In 1993, Nicolaides et al. detected chromosomal abnormalities in 32% of cases with nasal hypoplasia, proboscis lateralis or single nostril.

Nasal development starts in the third week of gestation when the primordial structure first appears. Olfactory placode is the primary organiser of developing nose and is responsible for olfactory nerves formation. Nasal placodes become apparent in the fourth week of intrauterine life. These placodes soon sink below the surface to create nasal pits, which subsequently form the nostrils. Medial and lateral raised edges of the pits are called medial and lateral nasal placodes, respectively. Nasal alae are formed by fusion of lateral and medial nasal processes. Mesoderm becomes heaped up in the median plane to form nasal prominence. By means of evagination, placodes form nasal pits, which are widely spaced on anterolateral area of developing head. Nasal cavities are formed by extension of nasal pits, which are widely spaced on anterolateral area of developing head. Absence of nasal placodes probably leads to heminasal aplasia. Nasal lacrimal system develops into the nasal maxillary groove, but in half-nose cases it would be blind. Absence of nasal placodes probably leads to heminasal aplasia. Nasal lacrimal system develops into the nasal maxillary groove, but in half-nose cases it would be blind. During facial development, maxilla generally has a growth deficiency in both sagittal and vertical dimensions. Malocclusion can be explained by growth arrest of nasomaxillary complex.

In 1976, Tessier postulated that it could be one of the possible presentations of facial cleft no. 2, while Ortiz-Monasterio, in a large series of facial clefts, reported only five cases of half-nose and classified this malformation as a facial cleft no. 3. Other authors believe it is just a nasal displasia. The aim of this article is to present personal experience in half-nose reconstruction and review the literature with regards to this rare entity, highlighting aspects of incidence, pathogenesis and surgical treatment.

Clinical cases

Seven sporadic cases have been treated in our institutions. Patients presented with different forms of commitment (Table 1), and consequently, individualised surgical techniques have been used for nasal reconstruction.

Surgical technique

Half-nose reconstructions have been complex and multi-staged. Our standard technique of reconstruction consisted of three-stage procedures. First stage was usually performed between 4 and 6 years of age (Figure 1A). A foil template of normal heminose was drawn and transferred to the hypoplastic side. Distance from the midpoint of columellar base to alar base was determined in normal side and transferred to affected side. Based on these marks, width

<table>
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<tr>
<th>Case</th>
<th>Age</th>
<th>Gender</th>
<th>Side</th>
<th>PL</th>
<th>Craniofacial malformations</th>
<th>Other findings</th>
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<tr>
<td>1</td>
<td>6 y</td>
<td>Male</td>
<td>Left</td>
<td>No</td>
<td>• Hypertelorism;</td>
<td>Polycystic kidney;</td>
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<td></td>
<td></td>
<td>• Cleft orbital floor;</td>
<td>Hidden bifid spine (C2, C3C4);</td>
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<tr>
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<td></td>
<td></td>
<td></td>
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<td>• Lachrymal obstruction</td>
<td>Interatrial communication</td>
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<tr>
<td>2</td>
<td>5 y</td>
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<td>No</td>
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<td>Epispadia</td>
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<td>3</td>
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<td></td>
<td></td>
<td></td>
<td></td>
<td>• Microphthalmus</td>
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<td></td>
<td></td>
<td>• Nystagmus</td>
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<tr>
<td>4</td>
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<td></td>
<td></td>
<td>• Cleft lip and palate</td>
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<tr>
<td>5</td>
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<td>No</td>
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<td>No</td>
<td>• Right cleft lip</td>
<td></td>
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</table>

* Age of patient at the moment of first-stage surgical reconstruction.
* PL: Proboscis lateralis.
Figure 1  Patient 1. A. Preoperative frontal view. B. Schematic drawing of the nasal skin and forehead flaps. C. Incision of the nasal flap to the inner lining. D. Flap rotation to the contralateral side. E. Silicone catheter through the nasal hole. F. Forehead flap undermining. G. Final result.

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and length of a forehead flap was determined (Figure 1B and C). For patients without any nasal cavity, anterior piriform aperture was drilled out and an inferiorly based local skin flap was rotated to line internal nostril (Figure 1D and E). A silicone stent was used in the reconstructed nostril and pyriform aperture for at least 6 months. A cartilage graft from the lower portion of ear tragus and concha was used to provide nasal-tip support. A forehead flap was harvested, including frontalis muscle, along a cleavage plane, superficial to the periosteum (Figure 1F). At eyebrows level, a careful blunt dissection was performed to preserve the suprathroclear vessels. The flap was rotated 180° to cover the whole external defect of the nose (Figure 1G). Three weeks later, its pedicle was trimmed. Third-stage refinements consisted of flap thinning and nostril aperture revision.

In two cases we have used different approaches to nasal reconstruction. In case 2, we opted to reconstruct only the affected side using local mucosal and forehead flap (Figure 2). In case 6, we performed a forehead expansion prior to harvest of the frontal flap. Both of them did not demonstrate any advantage over our standard approach (Figures 3 and 4).

Discussion

We have classified half-nose malformation as a Tessier facial cleft no. 2, due to the absence of lower eyelid coloboma, medial canthus dystopia, lacrimal obstruction and hypertelorism. The affected nose may be hypoplastic or completely absent, and some cases may present unilateral proboscis or a tissue tag. In our series, four patients were male, and three female; four on the right side and three on the left; proboscis lateralis was not present and five cases of a non-functional eye globus were noted. Three of our cases had hypoplastic maxilla with class III malocclusion.

Many aspects must be considered when reconstructing a congenital half-nose, such as timing of surgery, tissues available to reconstruct the defect and the necessity to create an internal nasal passage. The most appropriate age to operate on the patients, and if nasal...
reconstruction will be definitive remains unanswered. Management of psychological aspects is very important in paediatric patients with nasal malformations. By about 5 years of age, children become self-conscious and develop self-image and self-esteem. Some surgeons have suggested reconstructing the nose at ages of 5–7 years in order to minimise psychological trauma. Guided by social and psychological reasons, we believe this age is the most appropriate moment to reconstruct the nose and avoid or minimise psychological problems. Two cases of this series arrived in early age, and we followed this protocol.

The goals of total nasal reconstruction include restoration of aesthetic and functional aspects, which remains a challenge for plastic surgeons. Basically, full-thickness nasal defects require reconstruction of the inner lining, internal support and external coverage. Burget and Menick have proposed nine subunits for nasal repair, while Giugliano divided the nose into three main areas: tip, dorsum and ala. All these units and subunits should be considered when reconstruction half-nose defects. There are few adequate options to reconstruct external coverage of the nose, including local flaps, forehead flaps, forearm free flaps and also tissues of the proboscis lateralis. We believe when proboscis lateralis is present, it is very useful for external nasal reconstruction as the skin colour and texture is more similar. When proboscis lateralis is absent or insufficient, forehead skin is an excellent donor site to resurface the nose.

Majority of the articles using forehead flaps concern adults. There are few reports mentioning its use in paediatric patients and usually with short-term follow-up. We agree with Giugliano et al. that have used tissue expansion only for cases with large nasal defect and proportionally small donor site. Forehead expansion creates a large capsule, which can contract in postoperative period, leading to a short nose on long-term result. The pedicle

Figure 3 Patient 3. A. Preoperative frontal view. B. Final result.

Figure 4 Patient 4. Preoperative frontal view.
may be designed contralateral or ipsilateral to the primary nasal defect. Historically, contralateral design has been preferred because it results in less torsion on the pedicle. A thinner flap may improve the pliability of the flap with better nasal contour. A third-stage procedure aiming to thin the flap and revise the scars should be performed about 6 months later when wound healing and scar contractures are definitive.

Local flaps such as contralateral cutaneous nasal flap, nasolabial flaps, nasal or septal mucosal flaps are feasible options to reconstruct the inner lining. Bhandari and others demonstrated the use of a nasolabial flap for nasal lining. Other authors have published the use of skin graft or oral mucosal flap. Nasal airway lined by split-skin graft, however, tends to be stenotic, needing splints or silicone stents to prevent it. We have used contralateral cutaneous nasal flap to reconstruct inner lining and a forehead flap to cover external defect. As illustrated in case 2, forehead flap can also be used for partial nasal reconstruction. Due to limitations in the final aesthetic result, we believe that a second-stage procedure to achieve a natural shading of the nostril in the reconstruction of a half nose. Br J Plast Surg 1998;51:131.

Reconstruction of congenital half-nose has additional challenges, as in the restoration of nasal airway. Its reconstruction in the same first stage is even more difficult and has scarcely been reported in the literature. Moreover, reconstructed airway tends to become stenotic, and measures must be taken to minimise tissue contraction. Nagase stated that nasal airway reconstruction might not be repaired in congenital hemi-nose, because these patients usually do not have problems in breathing when they have the contralateral nasal airway intact. The author reconstructs pyriform aperture in an intact.8 The author reconstructs pyriform aperture in a second-stage procedure to achieve a natural shading of the nostril floor, but not intending to have a second pervious airway.

Regarding the lacrimal system reconstruction, it should be ideally performed after complete nasal reconstruction, once it allows tracking lacrimal duct into the new created nasal cavity.

Congenital half-nose is an extremely rare malformation. Pathogenesis remains unclear, and reported cases have occurred sporadically. Literature review found 91 cases of half-nose, including 50 patients with proboscis lateralis. This article presented other seven cases submitted to nasal reconstruction. Most common form of reconstruction used contralateral nasal skin flap to restore inner lining, conchal—tragus cartilage graft to restorealar framework and forehead flap to cover the external nasal defect. Follow-up has demonstrated good results using these techniques for half-nose reconstruction.

Conflict of interests

There was no financial interest by any of the authors, as well as no product, device or drug was specifically tested in this study.

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