Journal of Plastic, Reconstructive & Aesthetic Surgery (2010) 63, 15—21

JPRAS
An International Journal of Surgical Reconstruction
www.JPRASurg.com

The surgical repair of half-nose

Renato da Silva Freitas [a,b,*], Nivaldo Alonso [c], Thais de Freitas Azzolini [b], Luciano Busato [a], André Ricardo Dall'Oglio Tolazzi [b], Gilvani Azor de Oliveira e Cruz [b], Dov Goldenberg [c]

[a] *Assistance Center for Cleft Lip and Palate (CAIF), Curitiba-PR, Brazil*
[b] *Division of Plastic and Reconstructive Surgery of Federal University of Parana, Curitiba-PR, Brazil*
[c] *Division of Plastic and Reconstructive Surgery of Sao Paulo University, Sao Paulo-SP, Brazil*

Received 3 January 2008; accepted 31 August 2008

KEYWORDS
Half-nose;
Heminasal aplasia;
Nasal reconstruction;
Nose;
Facial cleft

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.

Absence of half-nose is an extremely rare congenital malformation, which has a devastating impact on the patient and the family.[1—5] Cleft nose, proboscis lateralis and nasal agenesis are rare; whereas absence of half-nose is also extremely rare.[6] A review of indexed English-language literature found 91 cases of half-nose — 50 of them with proboscis lateralis.[7—30] 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,

* Corresponding author. Address: R. 25 de Janeiro, 2460, Condominio Anhangava (casa 23), Quatro Barras, Parana 83420-000, Brazil.
E-mail address: dr.renato.freitas@gmail.com (R. da Silva Freitas).

1748-6815/$ - see front matter © 2008 British Association of Plastic, Reconstructive and Aesthetic Surgeons. Published by Elsevier Ltd. All rights reserved.
doi:10.1016/j.bjps.2008.08.040

teratoma, encephalocoele and microtia. Reports of 22 cases did not mention associated deformities.

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[6] 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.[31,32] Any congenital anomaly of the nose can also be accompanied by other facial and palatal anomalies.[33] In 1993, Nicolaides et al. detected chromosomal abnormalities in 32% of cases with nasal hypoplasia, proboscis lateralis or single nostril.[32]

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.[12] 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.[34] 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.[16] Maxillary and lateral processes fuse to form the nostrils. Defective development of the lateral nasal process can result in alar rim anomaly.[35] Medial nasal processes fuse with each other, thereby forming the ridge, tip and columella of the nose, as well as the philtrum and medial part of the upper lip. Fusing with maxillary processes, medial nasal processes separate nasal cavities and mouth. Heminasal aplasia with or without proboscis lateralis seems to be the result of one group of developmental disorders.[16] 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 sagital and vertical dimensions. Malocclusion can be explained by growth arrest of nasomaxillary complex.[9]

In 1976, Tessier[36] postulated that it could be one of the possible presentations of facial cleft no. 2, while Ortiz-Monasterio,[27] 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.[6,31] 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 multistaged. 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 1 Epidemiological data and clinical findings of 7 cases with half-nose

Case	Age[a]	Gender	Side	PL[b]	Craniofacial malformations	Other findings
1	6 y	Male	Left	No	• Hypertelorism; • Lateral and inferior orbital displacement; • Cleft orbital floor; • Lachrymal obstruction	• Polycystic kidney; • Hidden bifid spine (C2, C3C4); • Interatrial communication
2	5 y	Male	Right	No	• Right hemifacial microsomia • Median cleft lip • Absence of lachrymal duct	• Epispadia
3	9 y	Male	Right	No	• Nasoethmoidal encephalocele • Microphtalmus • Nystagmus • Absence of lachrymal duct	• No
4	15 y	Female	Left	No	• Anophtalmus • Cleft lip and palate	• No
5	19 y	Male	Left	No	• Anophtalmus	• No
6	23 y	Female	Right	No	• Microphtalmus • Right cleft lip	• No
7	15	Female	Right	No	• Hypertelorism; • Lateral and inferior orbital displacement; • Lachrymal obstruction	• No

[a] Age of patient at the moment of first-stage surgical reconstruction.
[b] PL: Proboscis lateralis.