International Journal of Pediatric Otorhinolaryngology 86 (2016) 72–76

Contents lists available at ScienceDirect

International Journal of Pediatric Otorhinolaryngology

journal homepage: www.elsevier.com/locate/ijporl

Surgical correction of bifid nose

Ali Tawfik, Hossam Elsayed El-Sisi *, Ahmed Musaad Abd El-fattah

Otolaryngology Department, Faculty of Medicine, Mansoura University Hospital, Mansoura, Egypt

ARTICLE INFO

Article history:
Received 4 January 2016
Received in revised form 2 April 2016
Accepted 4 April 2016
Available online 20 April 2016

Keywords:
Bifid nose
Tessier No:0 cleft
Surgery

ABSTRACT

Objectives: Evaluation of the aesthetic outcome and functional aspect after surgical correction of bifid nose by combined Millard forked flap with external rhinoplasty
Background: Bifid nose is a rare congenital anomaly that results during facial development but the explicit mechanism is not clearly understood. Clinical findings are quite variable with a wide range of severity. Surgical correction still represents great challenge to facial plastic surgeons; extensive deformities in many cases, rarity of condition and paucity of publications are contributing factors.
Methods: Surgical correction of six patients with bifid nose by a combined Millard forked flap with external rhinoplasty
Results: The aesthetic and functional outcomes were acceptable for all patients and parents. There were no considerable postoperative complications.
Conclusions: This approach is highly effective for various grades of bifid nose. Early management is preferable to avoid psychological morbidity. Secondary rhinoplasty is usually needed for cosmetic refinement.

1. Introduction

Bifid nose is a rare congenital anomaly that results during the embryological development of the nose. In spite of being a rare incident (1.5 to 4.8 per 100,000 births), it is considered the most common craniofacial cleft and corresponds to Tessier 0 facial cleft [1]. Based on his experience, Tessier merged his clinical, radiological and surgical observations in more than 300 cases to establish a classification for craniofacial clefts. This classification remains the most widely accepted despite many modification trials, such as those adopted by Zhonghua et al and Mazzola [2].

Clinical presentation shows wide variations and different degrees of severity. Examination may reveal: flat, faintly grooved or deeply furrowed nasal tip, separated alar cartilages, short and wide columella, absent anterior nasal spine "ANS", thick, duplicated or absent nasal septum, separation of ascending maxillary processes and nasal bones causing flat dorsum. The nose generally looks short. The forehead may be wide with or without orbital hypertelorism. Airway is mostly not obstructed in spite of overt disfigurement [3,4].

Several anomalies may be associated with bifid nose. The most common are orbital hypertelorism, midline cleft lip or just a central vermilion notch. Less common associations are duplication of prolabium, coloboma of the eye, high arched palate and anophthalmia [3,5,6]. Association with intracranial anomalies had also been reported where the most severe facial abnormality corresponded to the more significant brain anomalies and developmental delay. Examples are frontonasal encephalocele, mental retardation, agenesis of corpus callosum and holoprosencephaly which is usually incompatible with life [7].

Surgical correction of bifid nose is challenging due to the rarity of cases, complexity of the present deformities and limited number of publications. We present six cases of bifid nose managed by combined Millard flap and external rhinoplasty approach. The surgical technique, outcome and complications are evaluated and presented. Written informed consents were obtained from the parents to publish the case details and photos of their children.

2. Materials and methods

Ethical approval for this work was obtained by ethical committee board in our department. Through the period between 2000 and 2015, 6 cases of bifid nose were surgically treated in our Otorhinolaryngology department. The age at treatment ranged from 5 months to 9 years at time of surgery. Baseline data, clinical findings and associated anomalies are presented in Table 1.

CT scans on the nose and facial bones and MRI of the brain were done for all patients. Radiology was used to assist clinical diagnosis, assess midline structures and to rule out the presence of associated intracranial anomalies.

For surgical correction, we used Millard forked flap combined with external rhinoplasty approach. The Millard flap is extended from the superior margin of the upper lip – not involving the white line – then connected to bilateral vestibular marginal incisions for

* Corresponding author at: Otolaryngology Department, Faculty of Medicine, Mansoura University Hospital, Elgomhoria St. P.O Box: 35516, Mansoura, Egypt. Tel.: +20502202776.
E-mail addresses: elsisihossam@yahoo.com; elsisihossam@gmail.com (H.E. El-Sisi).

http://dx.doi.org/10.1016/j.ijporl.2016.04.004
0165-5876/

Table 1
Baseline data, clinical findings and associated anomalies.

Case	Age	Sex	Septum	Ant nasal spine	Nasal dorsum	Lip	Columella	Inter- orbital space	Association	Neuro-logical assesment[a]	Family history[b]
1	5 y	M	Thickened	absent	Broad	Notched	Very wide	Wide	coloboma of nostril – bilateral bat ears	Free	Free
2	3 y	F	Doubled	absent	Flat	Notched	Very wide	Normal	No	Free	Free
3	5 m	F	Thickened	present	Broad	Normal	Short-wide	Wide	coloboma of nostril- coloboma left eye- left bat ear	Free	Free
4	9 y	F	Normal	present	Normal	Normal	Furrowed	Normal	Low set ear	Free	Free
5	4 y	M	Normal	absent	Broad	Notched	Wide	Wide	incomplete median cleft alveolas- small low set ears- skin tags-	Free	Free
6	3 y	M	Doubled	absent	Flat	Notched	Wide	Very wide	Incomplete median cleft alveolas – high palatal arches- coloboma of nostril.	Free	Free

[a] Neurological examination: detection of neurological or brain abnormalities by clinical examination and MRI findings.
[b] Family history include: prenatal maternal exposure to drugs, radiation, infections, or abnormal delivery, other family members affection.

decortication of external nose as in classic external rhinoplasty. Excision of surplus skin from nasal dorsum and upper lip was then done. Grafting of alar cartilage was done when appropriate. At skin closure, the Millard flap was used for lengthening and reconstruction of the abnormally short columella by suturing its limbs in midline. In one mild case, we only performed tip-plasty by open rhinoplasty approach. Secondary rhinoplasty surgery for cosmetic adjustment was done in three patients. Surgical procedures are demonstrated in Table 2.

3. Results

The aesthetic and functional outcomes were acceptable for all patients and parents. There were no major postoperative complications. Follow up period ranged from 6 months to 15 years. Surgical outcome, complications and follow up periods are represented in Table 3.

In one case there was postoperative wound infection; this was treated effectively by systemic antibiotics for one week. Another patient complained of mild unilateral nasal obstruction due to septal deviation which was managed during a secondary rhinoplasty operation. Mild bilateral vestibular stenosis was detected in one case without functional or aesthetic complaint.

4. Discussion

4.1. Pathophysiology

The morphological aspects of bifid nose may range from simple faint groove in the columella or nasal tip up to extensive cleft of all nasal structures resulting in double nose formation in severe cases [8]. In between these two extremities, variable degrees of widening, thickening and sometimes duplication of the skeletal tissue could be present. Agenesis or hypoplasia could also be detected.

The developmental origin is not clearly understood. The embryological formation of the nose dates to the 4th week of gestation and this anomaly results when there is failure of midline fusion between the two medial nasal processes of the frontonasal process. Oligohydramnios, amniotic bands, maternal metabolic imbalances and exposure to infection, radiations, drugs or chemicals were all proposed as risk factors. Associated affection of forehead, glabella, intra orbital region and prolabium in many cases is attributed to being derivatives of frontonasal process [9,10].

Most cases of bifid nose are sporadic but many authors suggested genetic element. Esser reported a family with multiple cases proposing autosomal domination with reduced penetration [10]. Boo-Chai described 3 cases in siblings assuming a genetic linkage, but he could not define a specific pattern of inheritance [11] while, Anyane-Yeboa et al demonstrated a family in which multiple members in different generations were affected with direct transmission to their offsprings, suggesting dominant inheritance with variable penetration [12].

4.2. Surgical treatment

4.2.1. Evolution of surgical treatment

Owing to rare incidence of bifid nose, a lot of surgical techniques are still not widely accepted. Consequently, there is no universal agreement about a certain procedure; rather, there are different techniques based on personal experience and preference. Throughout more than a century, many authors tried to find out the most suitable approach and corrective strategies to achieve the best possible cosmetic improvement.

The first publication about bifid nose correction was made by ROE in 1887. Fifty years later, Esser performed osteotomies then augmentation with bone graft in 11 patients. Joseph then described