

Heminasal Aplasia: Clinical Picture, Radiological Findings and Early “Temporary” Reconstruction with a Nasolabial Flap

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Abstract

Heminasal aplasia or hemi-arhinia is an extremely rare congenital malformation with the exact mechanism or etiology of its development still unknown. The rarity of this anomaly makes its reconstruction a surgical challenge with a diversity and controversy on the timing and technique of such reconstruction. We report a new case of hemi-arhinia which to the best of our knowledge is the seventieth reported case of congenital defect. The case was a female infant with absence of the lower 2/3 of the right side of the nose. The clinical and the CT scan manifestations of a case of right-sided hemi-arhinia together with the reconstructive plan of it are presented. We recommend early soft-tissue reconstruction using adjacent nasolabial flap without cartilage grafts to provide “temporary” correction of the disfigurement until the definitive reconstruction can be undertaken when the child grows up.

This case was presented and operated upon in Plastic Surgery Dep., Faculty of Medicine, Sohag University, Sohag, EGYPT 82524

INTRODUCTION AND REVIEW OF LITERATURES

Nasal hypoplasia ranging from underdevelopment or partial absence of parts to complete arhinia is the most frequently seen nasal anomalies [1]. Heminasal aplasia, hemi-arhinia or unilateral aplasia of the nose is a rare congenital malformation in which there is absence of half of the external nose together with a variable degree of abnormality in the internal anatomy of the nose as well as the adjacent facial structures. It imposes a major psychological burden to the parents and may have physiological impact on the child.

The nose develops from the mesodermal frontonasal process and the two nasal placodes. The frontonasal process appears in the third to fourth week of gestation together with two bilateral ectodermal thickenings known as nasal placodes that grow caudally. During the fifth week of gestation, a central invagination, called the nasal pit, divides each nasal placode into a medial and a lateral nasal process. The nasal pits extend posteriorly to form the nasal cavity, which is separated from the oral cavity by a thin nasobuccal membrane. The nasobuccal membrane eventually ruptures at

week 6 to form the posterior choanae. The epithelium around the forebrain thickens to become specialized olfactory sensory cells. The medial nasal processes from both sides fuse, forming the nasal septum and philtrum while the lateral processes develop into the external wall of the nose, the nasal bones, the upper lateral cartilages, the alae, and the lateral crura of the lower lateral cartilages. The failure of the development of nasal placodes probably lead to the congenital absence of nose [2]. Although the exact mechanism is unknown, several theories for the pathogenesis of arhinia were hypothesized. These theories include 1) failure of the medial and lateral nasal processes to grow, 2) premature fusion of the medial nasal processes, 3) lack of resorption of the nasal epithelial plug, and 4) abnormal migration of the neural crest cells [3,4]. Congenital arhinia may be in part induced by Chromosomal aberrations as some chromosomal change has been reported in several cases. The genetic analysis of five patients with complete arhinia identified a 19 Mb large deletion involving 3q11–q13 in one patient of them [5]. Another patient had and a translocation between chromosomes 3 and 12 [6]. In another report, one case was found to associate with inversion of chromosome 9 and another had mosaic of chromosome 9 [7].

Failure of the development of both nasal placodes results in complete nasal aplasia or arhinia while failure of one

placode leads to heminasal aplasia or hemi-arhinia [8]. Nasal anomalies rarely occur alone and are frequently associated with other coexistent craniofacial anomalies. They were classified into two major groups: 1. total arhinia, with absence of the nose and both olfactory nerves; 2. partial arhinia with presence of at least one nostril and one olfactory tract. Both groups can be seen without (a) or with (b) other craniofacial malformations [7]. Partial arhinia includes all the range from hypoplasia or absence of only an individual structure to a complete absence of the heminose. Few cases were reported with congenital absence of the columella, with the medial crura of the lower lateral cartilages and their soft-tissue covering were missing, while the remaining septum and other nasal structures were normal [9,10]. Isolated nasal bone agenesis or hypoplasia has also been reported [11,12].

Sixty-nine cases of hemi-arhinia have been reported, with the majority of these cases having an associated proboscis lateralis found on the same side of the missing half of the nose. In 1976, Mazzola reviewed 39 cases of them 27 cases were associated with proboscis lateralis [13]. In 1997, a review of 23 cases was published with the patients' data including gender, laterality and associated anomalies were presented [8]. In 2004, there was a report of five unrelated Brazilian patients [14] and another case reported in India [15]. In 2005, a new case from the USA was reported [16]. All reported cases are sporadic with no apparent gender or side preference.

THE CASE REPORT

HISTORY AND CLINICAL PICTURE

A female infant, presented to us when she was 5-days old. The infant was the outcome of normal vaginal delivery after full-term pregnancy. She was 3200 grams. The mother didn't take any medicines during pregnancy, no consanguinity between parents and no such abnormality was found in either of her parents' families. The infant was delivered in a primary health care unit in a village. No history of breathing or suckling problem was reported by the parents who were concerned only by the abnormally disfigured nose. On examination, there was absence of the lower 2/3 of the right side of the external nose, while the upper bony third was looking normal. There was a tiny opening to the right side of the columellar-lip junction, and a small coloboma of the upper eyelid at the junction between its medial and middle thirds (figure 1). The infant was breathing normally with no respiratory distress or cyanosis. Pediatric examination of the infant did not reveal any other abnormality; neither did the

abdominal ultrasonography nor the echocardiography. Blood picture and liver functions were within normal ranges. The anomaly was explained to the parents, and they were instructed to come back for follow up when the child is 3 month of age.

CT SCANNING

At the age of 3 months, the patient was admitted to the hospital and a CT scanning was performed (figure 2). CT findings were: 1) normal and symmetric nasal bones on both sides; 2) marked narrowing of the nasal cavity on the right side with the nasal septum markedly deviated towards this same side; 3) hypertrophic conchae in the left heminose and 4) no associated orbital abnormality. The small opening at the base of columella was successfully canulated and a contrast dye was injected into the nasopharynx confirming choanal patency.

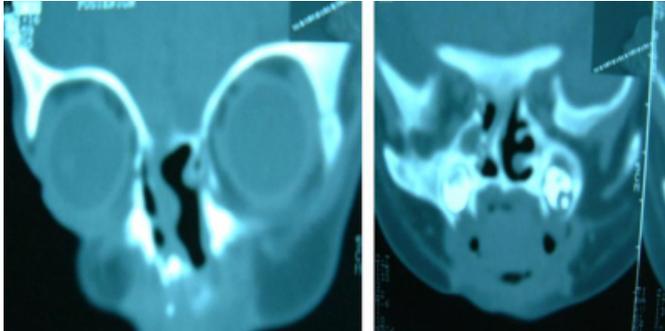
Figure 1

Figure 1: The preoperative pictures of the 5-days-old baby (left) and the 3-months-old child (right). Note the erythematous area in the medial third of the right upper eyelid accompanying the coloboma in the left picture while the right picture shows only the colobomatous notch of the eyelid.



Figure 2

Figure 2: The CT scan showing the inner anatomy of the nose with marked septal deviation to the right side, partial obliteration of the right heminasal cavity and hypertrophic conchae in the left heminose.



THE OPERATIVE RECONSTRUCTION

The parents of the patient were told that the reconstruction of the missing half of the nose would be undertaken when the child is around 15-years-old. However, they asked to have the right heminose reconstructed for their baby as early as possible even if the reconstruction would not be definitive and they accepted that the child might need one or more revisions. The operative plan was to reconstruct the absent heminose with a superiorly-based nasolabial flap (figure 3).

Figure 3

Figure 3: The pre-operative marking of the nasolabial flap.



An incision was made from a point inferomedial to the right inner canthus to a point on the nasolabial fold halfway

between the level of the nose and the level of the oral commissure. From this point, a cutback incision was made to the proposed site of the right alar-cheek junction. The superiorly-based nasolabial flap was then elevated; the triangular distal portion of it was thinned-out and turned-in with a few stitches providing the lining of the newly-constructed vestibule. A piece of split-thickness skin graft was used to line the depth of the nostril. A limited subcutaneous dissection in the cheek enabled the advancement of the cheek skin to close the secondary defect of the flap. The newly-constructed nostril was then packed with antibiotic-impregnated Vaseline gauze that was changed on the 5th postoperative day. The early postoperative result of the reconstructed right heminose was excellent (figure 4); however, on later follow up at 4 and 10 months after the operation, there was a noticeable upward retraction of the constructed nostril (figure 5).

Figure 4

Figure 4: Early postoperative results; the anteroposterior (left) and right lateral (right) views of the child at 5 days after surgery.



Figure 5

Figure 5: Follow-up photos at 4 months (left) and at 10 months (right) after surgery with evident upward retraction of the nostril.



DISCUSSION

To the best of our knowledge, this is the 70th case of hemi-arhinia reported worldwide. The previously reported cases of hemi-arhinia show strikingly similar features with most of them having concomitant ipsilateral proboscis lateralis [13]. In our case, the child was a female and the aplastic half of the nose was on the right side, the parents were not relatives and they declined any family history of similar anomaly. There was no proboscis and the only associated extranasal anomaly was the small upper eyelid coloboma.

The management of arhinia is a surgical challenge and because of its rarity, there is still a controversy concerning the timing as well as the technique of such reconstructive procedures. The ability of the child to breathe, the adequacy of the air passages and the cosmetic appearance are the main factors determining the timing of surgery. Early surgical intervention in childhood is thought to be better, both from the cosmetic and psychological point of view [17]. Onizuka et al. began their constructive surgery in a case of arhinia at the age of 6 months, and the reconstruction was complete at the age of 18 months [18]. They recommended that operative correction of arhinia to be undertaken as early as possible to achieve a better morphology as long as there is no cerebral anomaly. Muhlbauer et al. started their reconstruction of both the internal and external nose at the ages of 4 months and the reconstruction was completed at the age of 26 months [19]. On the other hand, many authors agree that surgical reconstruction of the nose and its internal cavities should be delayed at least until preschool years [7,20,21,22,23]. Others postpone the nasal reconstruction till the age of 15 years when facial development is nearly complete [24]. The nasolabial flap was not previously reported to be used in the reconstruction the missing nostril, nevertheless, some authors had used it as a lining of the forehead flap [15]. We believe that early surgical correction is better particularly in children with an isolated nasal anomaly.

In this case, no cartilage framework was incorporated during this procedure because the cartilaginous framework of the left heminose and the columella was strikingly normal, so I preferred to keep it undisturbed to avoid any adverse effects on its growth. In the same context, the forehead flap was not used in this “temporary” reconstruction of the missing heminose as it will be probably used in the final reconstruction when the growth of the nose and its osteocartilaginous skeleton is completed around the age of 14 years [25]. By that time, a cartilage graft will be

incorporated to provide the supporting skeleton for to the reconstructed heminose.

On follow-up visits of the child, there was evident retraction of the constructed heminose (figure 5), which is partly due to the lack of cartilaginous skeletal support and partly a consequence of the linear contraction of the wound. This contraction is planned to be corrected by Z-plasty that will transpose the constructed ala caudally to the same level of the other normal ala (figure 6).

Figure 6

Figure 6: The proposed Z-plasty for the correction of the upward retraction of the constructed heminose.



Even if there is no functional abnormality, early reconstruction of the missing heminose can provide major esthetic improvement. However, the parents have to know that the child will need at least one or two additional procedures to sustain satisfactory results and that the final surgical reconstruction will be concluded at the age of fourteen. One of the most important benefits of the early reconstruction is the normal or near-normal appearance of the child. This will obviate the psychological stress in the parents as well as the child when he/she gets enrolled in a kindergarten or a school. However, we recommend minimizing the surgical trauma as much as possible to avoid scarring of tissues even if the outcome of the procedure will not be optimal at this time.

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