Rhinoplasty: Congenital Deficiencies of the Alar Cartilage

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Abstract

Background: Congenital deficiencies of the alar cartilages are rare and often visible at birth but can occasionally present later.

Objectives: The authors review the anatomical development and discuss the incidence and treatment of congenital defects within the alar cartilages seen in rhinoplasty cases.

Methods: The charts of 869 consecutive patients who underwent open rhinoplasty were retrospectively reviewed, and 8 cases of congenital defects of the alar cartilage within the middle crura were identified. Intraoperative photographs were taken of the alar deformities, and each patient underwent surgical correction. To simplify analysis, a classification of the defects was developed. A division was a cleft in the continuity of the alar cartilage with the 2 ends separate. A gap was a true absence of cartilage ranging from 1 to 4 mm, which can be accurately assessed in unilateral cases. A segmental loss was a defect greater than 4 mm.

Results: The 8 cases of deformity could be classified as 4 divisions, 3 gaps, and 1 segmental loss. None of the patients had a history of prior nasal trauma or nasal surgery. Six patients were women and 2 patients were men. In all cases, adequate projection and stability were achieved with a columellar strut. Asymmetry was minimized through concealer or tip grafts. There were no complications.

Conclusions: Surgeons performing rhinoplasty surgery will encounter and should be prepared to deal with unexpected congenital defects of the alar cartilage. These defects within the middle crura will require stabilization with a columellar strut and, often, coverage with a concealer tip graft. We speculate that the cause of these defects is a disruption of the hedgehog signals that may arrest the condensation or block the differentiation of the underlying neural crest cells.

Level of Evidence: 4

Keywords

nasal tip, projection, medial crura, domal notch, congenital, alar cartilage, rhinoplasty

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Figure 1. Congenital deficiencies of the alar cartilages. (A, B) A division is a cleft in the continuity of the alar cartilage with the 2 ends separate and rounded off. (C, D) A gap is a true absence of cartilage ranging from 1 to 4 mm when compared with the contralateral side. (E, F) A segmental loss is a defect greater than 4 mm.
To simplify analysis, we classified the defects into 3 categories: division, gap, or segmental loss (Figure 1). A division was a cleft in the continuity of the alar cartilage with the 2 ends separate and rounded off. A gap was a true absence of cartilage ranging from 1 to 4 mm when compared with the contralateral side. A segmental loss was a defect greater than 4 mm. Surgical treatment became more complex as the extent of the deficiency increased. In all cases, some type of columellar strut was utilized to provide support to the crura. For divisions, a standard columellar strut was inserted and the division repaired in a similar way to a domal division or an interruption in the alar cartilage that can be encountered in a secondary rhinoplasty case. In most cases, a “concealer graft” of excised alar cartilage was placed to cover any irregularities or asymmetries.1 For gaps, the surgical treatment was determined more by overall tip deformity and especially the loss of projection and definition, which is particularly common in bilateral cases. A major septocolumellar strut was required to provide maximum support to the tip, and a rigid tip graft also was often required for definition (Figure 2).2 For a segmental loss, treatment depended largely on whether the deformity was unilateral or bilateral. In unilateral cases, the goal was to “match” the sides (Figure 3). The domal junction and columellar breakpoint were aligned on the 2 sides, which defined the segmental loss. An interposition graft of excised alar cartilage was then placed to bridge the defect, and a concealer graft was applied to smooth the area.

RESULTS

There were 8 cases of congenital deficiencies in 869 open primaries. Patients with these deficiencies ranged from 14 to 68 years of age (average, 31 years). Six patients were women and 2 were men. There were 4 divisions, 3 gaps, and 1 segmental loss. All defects occurred in the middle crura. Three cases were unilateral and 5 were bilateral. In the bilateral cases, 4 patients had the same type of defect on each side. A single patient had a division on 1 side and a gap on the opposite side; her defect was classified as a gap (Figure 2). Thus, there were 13 crura with the following defects: 7 divisions, 5 gaps, and 1 segmental loss. None of the patients had a history of prior nasal trauma or nasal surgery. To date, none of the patients has undergone revisional surgery. In all cases, adequate projection and stability were achieved with a columellar strut. Asymmetry was minimized through concealer or tip grafts. There were no complications.

Clinical results are shown in Figures 4 and 5.
DISCUSSION

Any analysis of the complex embryology of the nose must be preceded by a brief review of the anatomy and terminology of the alar cartilages. Traditionally, surgeons and anatomists have considered the alar cartilages to be divided into medial and lateral crura. In 1967, Denecke and Meyer described the alar cartilages as having 2 angulations (medial and lateral) that were 2 to 3mm apart, thus presaging the concept of a domal segment. In 1987, Sheen and Sheen introduced the concept of a middle crus as “the missing link in the analysis and surgical correction of aesthetic problems relating to the nasal base.” They assigned the medial crura to the columellar, the middle crura to the lobule, and the lateral crura to the alae. Importantly, they defined the junction of the medial and middle crura to be the columellar-lobular junction, which in turn led to the angle of rotation. Equally important, they believed that the middle crura was a primary determinant of tip projection, especially a short middle crura, resulting in a tip with inadequate projection.

With the ascendancy of the open approach and especially tip suturing techniques, the alar cartilages came under even greater scrutiny. Guyuron investigated the anatomy and clinical management of the medial crura. Tebbetts emphasized the columellar breakpoint and the linkage between the medial and middle crura. Daniel subdivided each of the 3 crura into 2 segments. The middle crura are subdivided into a lobular segment and a domal segment. The lobular segment begins at the columellar breakpoint and ends at the medial genu. The domal segment is defined by the medial and lateral genu, which is often expressed as a domal notch on the caudal border of the middle crura. In reviewing the intraoperative photographs of the congenital deficiencies in our patient series, it is evident that the divisions occur at the level of the medial genu, signaling a separation between the lobular and domal segments. Equally, gaps occur within the domal segment, extending from the medial genu toward the lateral genu. The only segmental loss in our series involved the entire middle crura from the columellar breakpoint to the lateral genu.

In reviewing the literature on congenital deficiencies of the alar cartilages, it is important to distinguish between visible defects obvious at birth (columellar absence, cleft notch of the alar rim) and those that occurred in our patients, which were much more occult. In a single previous case report of an isolated congenital absence of the lateral crura,
Figure 4. (A, C, E, G) This 40-year-old woman presented for rhinoplasty. There were no definitive preoperative signs of the underlying congenital malformations, but preoperatively, the surgeon predicted that her alar cartilages would be extremely small. This patient’s deformity is revealed clearly in the intraoperative photographs in Figure 2. She had an unusual 4-mm gap on the right side and a division on the left side. (B, D, F, H) One year after surgical correction (shown in Figure 2) consisting of (1) an open approach, (2) harvest of the septal cartilage, (3) incremental dorsal reduction (bone 1 mm, cartilage 8 mm), (4) no caudal septal excision, (5) low to high osteotomies, (6) insertion of a columellar strut, (7) repair of the congenital defects with 5-0 sutures, (8) tip-shaping sutures, and (9) application of a diamond-shape tip graft of thin septal cartilage. At 3 years follow-up, the patient reported being pleased with the overall result but wondered if her nose could be made smaller.
the primary complaint was nasal obstruction. In contrast, all 8 of our patients presented for cosmetic rhinoplasty and had tip or nostril asymmetry as the only suggestion of a possible underlying anatomical problem. Thus, the location (middle crura) and range of severity of these congenital defects in the middle crura represent a significant new finding with regard to congenital anomalies of the nose.

Embryology of the Nose

The prevalence of nasal anomalies ranges from 1 in 20,000 to 40,000 live births, which is significantly fewer than have been reported for facial clefting. In a review of 261 patients, Losee et al classified nasal anomalies into the following 4 categories: (1) hypoplasia and atrophy,
Figure 5. (A, C, E, G) This 51-year-old woman presented for rhinoplasty to “soften” the appearance of her nose. Her nostrils had a distinct asymmetry. There was no history of prior nasal surgery or trauma. Preoperatively, an obvious difference in the nostril apices was noted, which often signals an asymmetry of the middle crura, and meticulous dissection did reveal an absence of the entire middle crura on the right side. (B, D, F, H) Two and a half years after surgical correction (Figure 3) consisting of the following: (1) an open approach, (2) incremental dorsal reduction (1.5 mm bone, 5.0 mm cartilage), (3) harvest of the septal cartilage, (4) insertion of spreader grafts, (5) insertion of a columellar strut with careful alignment of the domal segments and columellar breakpoint, (6) placement of an interposition graft using excised cephalic lateral crura, and (7) coverage of the infralobule and domes with an add-on graft of excised alar cartilage. The patient reported being pleased and referred her daughter, who did not have an alar deficiency, for a rhinoplasty.
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(2) hyperplasia and duplications, (3) clefts, and (4) neoplasms and vascular anomalies. The underlying etiologies for most nasal anomalies are unknown, but at least a subset of them likely arise from disruptions during the embryonic period when the nasal structures are specified.

The nose is built upon an embryonic cartilaginous frame called the nasal capsule, which itself is derived from the fusion and growth of 2 facial prominences, the median nasal and lateral nasal prominences. The median nasal prominence gives rise to midline structures, including the vomer and nasal septum, the medial and inferior portions

Figure 5. (continued) (A, C, E, G) This 51-year-old woman presented for rhinoplasty to “soften” the appearance of her nose. Her nostrils had a distinct asymmetry. There was no history of prior nasal surgery or trauma. Preoperatively, an obvious difference in the nostril apices was noted, which often signals an asymmetry of the middle crura, and meticulous dissection did reveal an absence of the entire middle crura on the right side. (B, D, F, H) Two and a half years after surgical correction (Figure 3) consisting of the following: (1) an open approach, (2) incremental dorsal reduction (1.5 mm bone, 5.0 mm cartilage), (3) harvest of the septal cartilage, (4) insertion of spreader grafts, (5) insertion of a columellar strut with careful alignment of the domal segments and columellar breakpoint, (6) placement of an interposition graft using excised cephalic lateral crura, and (7) coverage of the infralobule and domes with an add-on graft of excised alar cartilage. The patient reported being pleased and referred her daughter, who did not have an alar deficiency, for a rhinoplasty.
of the nostril, the philtrum, and the primary palate. The lateral nasal prominences give rise to the alar bases and the paired lateral crura of the alar cartilages.

Both the median and lateral nasal prominences are filled with a unique population of cells called cranial neural crest (CNC) cells, which share many similarities with stem cells. Similar to stem cells, the CNC population is capable of extensive self-renewal. In addition, CNC cells can differentiate into a wide array of cell types, including glia, neurons, melanocytes, chondrocytes, osteoblasts, myofibroblasts, and the pericytes lining blood vessels. The CNC-derived nasal cartilage is anatomically composed of a dorsal component, called the ectethmoid, and a ventral component, called the mesethmoid. The ectethmoid comprises the olfactory system, including the lamina cribosa, the crista galli apophysis, the crura, and the conchae. The mesethmoid gives rise to the nasal septum and the vomer. The mesethmoid structures are responsible for determining the proximodistal size of the nose and the position of the premaxillary bone, whereas the ectethmoid is responsible for the position and size of the nasal bridge.

By clarifying the embryonic origins of the nasal capsule components, scientists have begun to understand the basis for some malformations of the nasal capsule, such as those that are characterized by abnormal mesethmoid development and normal ectethmoid development. An example of these pathologies include hypo- and hyperseptoethmoid symptoms such as a flat nose, short nose, very prominent nose, and even a median nasal cleft with cerebral anomalies. Jacobs reported the first case of a patient with an absent columella. In 1988, Lewin reported 3 isolated cases of an absent columella with otherwise normal nasal and lip anatomy. In all 3 patients, it appeared that the medial crura, overlying skin, and membranous septum were all absent. As described earlier, the median nasal prominence forms the columella as well as the nasal septum and middle of the upper lip. Lewin speculated that a teratogen could selectively arrest the formation of the full thickness of the columella and that a bifid columella could be a partial expression of this malformation. Recent experimental data demonstrate that if growth factor signaling is perturbed (either through gene manipulations or exposure to embryos, and perturbations at this stage can result in an arrest in chondrogenic differentiation. This can lead to incomplete cartilaginous structures such as the cases of divisions, gaps, and segmental losses we observed in this series. These malformations were likely related to disruptions in the differentiation process rather than in the migration of the CNC cells into the median and lateral nasal prominences since the majority of the neural crest–derived alar cartilages were present.

For simple divisions, surgical correction is done with sutures, similar to repairing a previous domal division encountered in a secondary rhinoplasty case. For more extensive gaps, bilateral deformities are more easily corrected than unilateral. Insertion of a columellar strut and a concealer tip graft offers a simple solution. With segmental losses, some type of replacement graft is probably necessary and is done with excised cephalic lateral crura. Surgeons performing rhinoplasty surgery should be prepared to recognize and reconstruct congenital deficiencies of the alar cartilage, which occur in approximately 1% of primary cases.

**CONCLUSIONS**

Before the seventh week of fetal development, when the “blueprint” of facial patterning has yet to be established, perturbations in the gene regulatory network responsible for nasal development can result in severe malformations or a complete absence of structures derived from the median and lateral nasal prominences. After the seventh week of development, CNC cells begin their differentiation into chondrocytes, and perturbations at this stage can result in an arrest in chondrogenic differentiation. This can lead to incomplete cartilaginous structures such as the cases of divisions, gaps, and segmental losses we observed in this series. These malformations were likely related to disruptions in the differentiation process rather than in the migration of the CNC cells into the median and lateral nasal prominences since the majority of the neural crest–derived alar cartilages were present.

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