The Bifid Nose: Early functional and aesthetic management of frontonasal dysplasia
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ABSTRACT

Objectives: 1) To review the normal midfacial embryologic development, and review the bifid nose anomaly as part of the midline facial cleft spectrum.
2) To review an esthetically acceptable, single staged nasal reconstruction, via extensive resection of the skin, cartilaginous, and bony nasal framework in our 7 months old female patient with a bifid nose.

Study Design: Case report.

Methods: The clinical presentation, physical examination, imaging features, surgical findings, and technique for repair were reviewed in the case of a pediatric patient with bifid nose. The literature on normal midfacial embryologic development was reviewed as was that of the spectrum of midfacial malformations with emphasis on the bifid nose. Permission was obtained from the patient’s parents to publish the findings.

Results: We describe a newborn female who was noted to have a widened nasal bridge by prenatal ultrasonograph, and a bifid nose at birth. Examination revealed a wide nasal bridge, duplication of the nasal tip, columnella, philtral columns and a submucosal cleft. No other genetic malformations were identified. CT scan of facial bones showed bilateral piriform aperture stenosis, three central incisors and duplication of the nasal septum. MRI revealed no intracranial anomalies. Via an external approach, skin, bone, and cartilaginous resection resulted in an esthetically acceptable nasal contour. Review of the literature identified only a few such reported cases.

Conclusion: The bifid nose poses challenges to the reconstructive surgeon. Successful outcomes depend on a thorough understanding of normal midface embryology, the bifid nose anatomy, proper patient evaluation, and precise surgical technique.

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INTRODUCTION

The bifid nose, also referred to as frontonasal dysplasia (FND), is a relatively rare congenital anomaly of unknown etiology. It typically occurs sporadically, however it can occur as part of a broader syndrome such as holoprosencephaly. Its severity may range from a simple groove at the nasal apex to a complete duplication of nasal structures. The degree of facial deformity usually parallels those of the brain, with the most dramatic malformations presenting with severe brain anomalies (encephalocele) and developmental delay. As a rare anomaly, few cases and their surgical management have been reported in the literature, however milder forms of FND are often not repaired until later in childhood. We present a case of a 7 months old girl with a duplication of the entire nasal osteocartilagenous framework. Within this report we review normal midfacial development and describe an esthetically acceptable, single-staged surgery that addressed symptoms of nasal obstruction associated with this malformation.

CASE REPORT

The Pediatric Otolaryngology service was consulted on a 1 day old female patient because of a “double nose”. The mother’s pregnancy was uneventful and the baby was delivered vaginally at term without complications. She was the daughter of healthy non-consanguineous parents, with no significant paternal or maternal family history. Antenatal ultrasonography had suggested an isolated broad nasal root. Clinical examination revealed a wide nasal dorsum, widely spaced eyes, a bifid nose, stenosis of the left nasal cavity, a midline cleft of the upper alveolar ridge, and duplication of the labial frenulum (Tessier 0 facial cleft) (FIGURE 1). There were no other anomalies. A computed tomography (CT) scan of the facial bones revealed the presence of 2 widely-spaced upper central incisors separated by a paramedian alveolar cleft, bifurcation of the bony-cartilaginous septum causing left > right piriform aperture stenosis, and a broad nasal root (FIGURE 2). An MRI of the brain revealed no intracranial abnormalities.

The patient underwent a single staged repair at 7 months of age due to symptoms of nasal obstruction (FIGURES 3 and 4). Access to the nasal framework was obtained via a dorsal vertical midline incision. Mucoperichondrial flaps were elevated on either side of the two cartilaginous septa. A dermoid was noted at the bony bifurcation. This was removed along with the midline bony plate, which was drilled down to the vomer. The cartilaginous septa were sutured together with 5.0 PDS to close the dead space. The soft tissue was separated from the piriform apertures and the skin and cartilage of the nose was closed through medial advancement and reapproximation. The duplication of the philtral subunit and puncatum of the dermoid was then addressed by resection of the redundant midline soft tissue. The lip was closed by reapproximation of the orbicularis oris, a modified Mohler cleft lip repair.

DISCUSSION

Frontonasal dysplasia (FND) is a rare congenital nasal anomaly. Most cases are sporadic and have multiple etiologic factors; however, familial cases and syndromic associations have been described. Deformities reported in FND include a broad nasion, a midline cleft in the nasal dorsum, unilateral or bilateral cleft of the alae nasi, notching or coloboma of the nostrils, S-shaped deformity of the nasal septum, midline defect of the frontal bone (cranial bifidum occulatum), and widow’s peak (V shape extension of the hairline onto the forehead). The cause of FND is still unknown. The most accepted etiology is a failure of nasal capsule development. Between the 4th and 6th week of embryogenesis, unknown factors arrest migration of the olfactory epithelium into the nasal capsule. The nasal capsule does not form properly and the primitive forebrain tissue fills the space between the two nasal ridges.

The time of arrest of nasal capsule development determines the severity of the deformity. When early, clefts are seen in conjunction with midline ocular signs and normal brain development. Later arrests result in various forms of holoprosencephaly. Classically FND is described as Tessier 0, the most common of the craniofacial clefts. Radiologic imaging, (CT or magnetic resonance imaging), is very important before performing surgery, as even mild FND cases may present with synchronous intracranial anomalies. Surgical procedures for FND have only been reported in a few studies. The best procedure and time of surgery have not been established. In cases of nasal airway obstruction, early intervention may be considered. Most published cases use a modified open rhinoplasty approach similar to that used in this case.

REFERENCES