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REVIEW ARTICLE

Pyriform aperture enlargement in all aspects

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Abstract

Background: The pyriform aperture comprises the central area of facial bone structure. It is formed by the free corners of the nasal bone and the frontal processes of the maxillae, which articulate with each other at the nasomaxillary suture lines. Congenital nasal pyriform aperture stenosis might be linked to various craniofacial problems. This review presents all aspects of pyriform aperture stenosis and enlargement.

Methods: A literature search was conducted. Pyriform aperture definition, nasal development, congenital nasal pyriform aperture stenosis and pyriform aperture enlargement were reviewed.

Results: One of the most common abnormalities is holoprosencephaly, which is a midline developmental deficiency that may also be present in combination with facial clefting. The aetiology of nasal pyriform aperture stenosis remains unclear. When diagnosed, the choice of treatment is between non-surgical and operative methods, depending on the seriousness of the problem. Provided the sufferer can maintain a secure air passage with the help of specialised medical procedures and respiratory tract adjuvants, operative therapy may be delayed.

Conclusion: The operative outcomes are extremely good, and the prognosis relies mainly on coexisting neural and endocrine problems. This paper evaluates the nasal pyriform aperture in detail.

Key words: Congenital Abnormalities; Facial Bones; Maxilla; Nose; Stenosis

Pyriform aperture definition

The pyriform aperture is the central entrance of the facial skeletal frame structured by the free corners of the nasal bone and the frontal processes of the maxillae; these articulate against each other at the nasomaxillary suture line.[1] Rohrich *et al*. revealed that the upper lateral cartilage articulates laterally along with the bony side of the pyriform aperture through the pyriform soft tissue.[2] The pyriform soft tissue is a fascial membrane layer that surrounds the entire bony edge within the pyriform aperture, remaining uninterrupted along with the periosteum of the nasal bones and maxilla. The pyriform ligament may act as a further support to protect against upper lateral cartilage collapse and lateral internal nasal valve reduction. The lateral internal nasal valve region is thus based on the caudolateral side of the upper lateral cartilage, the pyriform ligament and the frontal process of the maxilla, which form the bony edge of the pyriform aperture.[1]

The pyriform aperture is the mainly anterior, and narrowest, part of the bony component of the nose. The borders of the pyriform aperture are: superior – nasal bone; lateral – frontal (nasal) process of the maxilla; and inferior – pre-maxilla, and the anterior nasal spine of the maxilla.[3]

The anterior nasal aperture is a heart-shaped or pyriform area with a vertical long axis, and with its narrow end pointing upward. It is greatly contracted by the lateral and alar cartilages of the nasal passages. Its upper boundary is formed by the lower limits of the nasal bone tissue; sideways, it is bounded by the thin, sharp edges that distinguish the anterior aperture from the nasal floors of the maxillae. Its lower boundary is characterised by similar limits, where they contour medialward to join one another at the anterior nasal spine.[4]

Various widths of the pyriform aperture have been determined. The thickness of the pyriform aperture was 26.5 mm for Ashantis, compared to 21.6 mm for Austrians, 25.2 mm for American Indians and 23.4 mm for African Americans.[5] Ofodile determined that Austrians had the longest nose bones, and that Ashantis had the greatest pyriform aperture width (oval contours).[5] Karadag *et al*. analysed 80 Anatolian individuals, and revealed a mean nasal bone length of 30.6 mm in males and 29.01 mm in females.[6] The mean width of the pyriform aperture

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was 18.83 mm in males and 18.19 mm in females. It was determined that the mean length of the nasal bone was greater, and the width of the pyriform aperture reduced, in Anatolian people when compared to Koreans, Austrians and Germans.[6] According to an evaluation of different but related trials, the pyriform aperture dimensions were found to be narrowest in Anatolians, while Ashantis had the broadest dimensions. Pyriform aperture width was smaller in Iranian people than in Ashantis and American Indians, and relatively similar to that of Koreans and Germans. It was also broader than that of African Americans, Austrians and Anatolians. Ashantis had the shortest nasal bones.[5]

This review involved a search of Medline (PubMed), the search engine of Kirikkale University, ProQuest Central and Google for all relevant articles, published up until 15 April 2016, using the keywords 'pyriform aperture', 'nasal', 'congenital nasal pyriform aperture stenosis', 'nasal pyriform aperture stenosis', 'stenosis', 'facial', 'bone', 'treatment', 'holoprosencephaly', 'nasal pyriform aperture', 'nose development' and 'enlargement', alone or together. Pyriform aperture definition, nasal development, congenital nasal pyriform aperture stenosis and pyriform aperture enlargement were reviewed.

Nasal development

Nasal development proceeds during the fifth to eighth week of gestation. The initial pharyngeal arch generates well-known maxillary and mandibular prominences in the fifth gestational week.[7] Medial limbs of the horseshoe-like nose swellings unite at the midline to produce the middle portion of the nose and philtrum, and the incisor section of the maxilla and primary palate.[7] The lateral branches of the nasal protuberances constitute the alae of the nose.[7] Concurrently, the maxillary prominence joins the lateral part of the nose bulge to produce the lateral nasal structure and the pyriform aperture.[7–11]

Aetiology

The aetiology of nasal pyriform aperture stenosis is uncertain. The initial article that described congenital nasal pyriform aperture stenosis noted it as a separate condition.[10] There remains disagreement concerning the aetiology of congenital nasal pyriform aperture stenosis and its correlation with craniofacial anomalies. The most frequently revealed abnormality is holoprosencephaly,[12] which is a midline developmental deficiency that can present with facial clefting.[13]

Arlis and Ward demonstrated the existence of related midbrain abnormalities, including holoprosencephaly, pituitary hypofunction and craniofacial abnormalities, particularly of the upper central maxillary incisor.[14] A solitary maxillary central incisor was discovered in four of the six individuals in their study. Holoprosencephaly is a type of midline dysgenesis with regard to the embryonic prosencephalon and midline facial components. A relation between the single upper central maxillary mega-incisor and holoprosencephaly has previously been demonstrated.[11]

Congenital nasal pyriform aperture stenosis

Congenital nasal pyriform aperture stenosis is an uncommon but life-threatening nasal obstruction in neonates given that they are nasal breathers in the first six to eight weeks of life.[15] A bony blockage of the nasal hole, either unilaterally or bilaterally, frequently arises as a result of choanal atresia.[16] Congenital nasal pyriform aperture stenosis develops in about a quarter to a third of choanal atresia cases.[17] Other conditions that may result in nasal blockages include: meningocele, meningoencephalocele, dermoid and epidermoid cysts, sinonasal tumours, septal dislocation and/or haematoma, and nasal hypoplasia due to warfarin teratogenicity or chondrodysplasia.[18] These conditions should be included in the differential diagnosis of congenital nasal pyriform aperture stenosis.[12]

Structurally, the pyriform aperture consists of the nasal and maxillary bones. Hypothetically, congenital nasal pyriform aperture stenosis is the result of out-of-control expansion of the maxillary nasal process bone that develops throughout the initial four months of fetal progression.[15,18–20] Congenital nasal pyriform aperture stenosis can occur individually or in connection with various other abnormalities, including those of the central nervous system, hormonal system and craniofacial region. One of the most likely hypotheses is that congenital nasal pyriform aperture stenosis is a microform holoprosencephaly (malformation as a consequence of incomplete cleavage of the embryonic holoprosencephalon).[12,20]

Nasal pyriform aperture stenosis was first described by Douglas in 1952,[21] while congenital nasal pyriform aperture stenosis resulting in respiration deficiencies in newborns was first reported by Brown *et al.* in 1989.[9] Congenital nasal pyriform aperture stenosis is a condition seen in babies. Its hereditary basis and typical clinical appearance as infant respiration difficulty have been well-documented.[4,12] Infants are obligatory nose breathers; for that reason, nasal bony inlet stenosis can result in respiratory tract blockage. Newborns with inadequate breathing as a result of pyriform aperture stenosis exhibit numerous symptoms, including apnoeic events and cyclic cyanosis improved by crying. These clinical characteristics resemble those observed in sufferers with bilateral choanal atresia.[11]

Congenital nasal pyriform aperture stenosis may be suspected by the finding of narrow nasal inlet blocking with the initial use of a fibrescope or nasal tube. The distinction between congenital nasal pyriform aperture stenosis and choanal atresia lies in the area within this narrowing: promptly at the access of the nasal hole in congenital nasal pyriform aperture stenosis (1st centimetre) and more posterior in choanal atresia (about 3 cm).[22] The computed tomography (CT) scanning

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criteria for congenital nasal pyriform aperture stenosis comprise: (1) pyriform aperture width inferior to 11 mm in a term neonate, as reported by Belden *et al.*;[23] (2) triangular shape of the palate; (3) bony overgrowth of the nasal process of the maxilla; and (4) problems with the teeth, for example a solitary median mega-incisor.[23]

Nasal pyriform aperture stenosis is diagnosed according to the clinical characteristics associated with respiratory tract blockage, including tachypnoea, poor feeding, nasal congestion, episodes of apnoea or cyanosis, and resistance to the manoeuvring of a nasogastric tube in either nostril. Clinical analysis is then validated with craniofacial CT.[13]

Pyriform aperture enlargement

The dimensions of the pyriform aperture do not correlate with the requirement for surgery, and the mean circumference of the nasal inlet was found to be higher in patients who received surgical treatment. Although this appears contradictory, it emphasises an important aspect of the normal management of hereditary respiratory tract defects. Every baby is different in their capacity to recover from a blockage, and it is essential to evaluate and deal with the baby rather than respond according to specific aperture sizes.[24]

When diagnosed, such defects can be treated therapeutically with conservative or operative options, according to the seriousness of the signs and symptoms. Provided the affected patient has a reliable respiratory tract (as a result of adequate medical care and the use of air passage adjuvants), operative treatment may be delayed. The surgical outcomes are excellent, and the prognosis is mainly based on coexisting neural and/or hormonal challenges.[13] Van den Abbeele *et al.*, in their series of 20 patients, advised a primary non-surgical procedure using nasal steroids and topical decongestants for as long as 14 days prior to the consideration of surgery.[25] The requirements for operative therapy consist of sleep apnoea, growing impairments, lack of ability to wean from air passage assistance and/or inadequate non-surgical management.

Visvanathan and Wynne documented that the stenosed pyriform aperture can be increased by way of a sublabial procedure.[13] Here, the mucosa is elevated to expose the bony stenosis and, using a 2.8 mm diamond burr, excessive bone tissue is drilled away from the inferior inlet along the ground of the nose to the lateral process of the maxilla. Care is taken to avoid drilling inferiorly and posterolaterally, to prevent damage to the sockets of the teeth and the nasolacrimal duct, respectively. Once successfully widened, stents are positioned to prevent re-stenosis. In our unit, we have successfully used size 3.5 Portex® endotracheal tubes fashioned into nasal stents with a common anterior cuff.[26]

Operative treatment is commonly carried out with a sublabial procedure, to allow lateral drilling of the nasal process of the maxilla, followed by nasal stenting. Van den Abbeele *et al.* advised that stenting should not exceed 14 days, but two of the patients in their series endured more than 20 days of stenting without any elevation in risk and with effective outcomes.[25] Some authors have recommended balloon dilatation of the pyriform aperture using the normal plasticity of the facial bones and cartilage.[27] Surgical treatment provides good results. As the size of the nasal cavity follows the craniofacial development, congenital nasal pyriform aperture stenosis is a neonatal problem that tends to resolve with development, and it has a remarkable long-term respiration prognosis. No secondary recurrence has ever been described in the literature following initially adequate operative therapy.[22]

Gonik *et al.* reported 9 surgical treatment failures in 63 patients (14 per cent) who underwent surgery.[24] In their study, 'failure was determined by the requirement for further respiratory tract surgical treatments to bypass or further open the nasal airway'. Associated craniofacial dysmorphism was the main factor that limited the success of the surgical procedures (odds ratio = 9; $p = 0.013$).

Conclusion

Nasal pyriform aperture stenosis is suspected when there are signs of respiratory tract blockage, including tachypnoea, poor feeding, nasal congestion, apnoea or cyanosis episodes, and resistance to the manoeuvring of a nasogastric tube in either nostril. To make the diagnosis, craniofacial CT scans should be obtained. Good results can be achieved with surgical treatment. The most common operative treatment is a sublabial procedure that allows lateral drilling of the nasal process of the maxilla, followed by nasal stenting.

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