Choanal Atresia

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Introduction

Choanal atresia is one of the etiologies for congenital nasal airway obstruction. It is relatively uncommon with an estimated incidence of 1:5000 to 1:9000 live births [1]. It was first described in 1755 by Roederer [2]. Surgical techniques to correct choanal atresia have been first described as early as the 19th century, and by the mid 1920's four surgical techniques have been described: trans-nasal, trans-septal, trans-palatal and trans-maxillary approaches [3].

Anatomy, Embryology and Pathogenesis

Choanal atresia is the anatomical closure of the posterior nasal choanae which are formed by the sphenoid bone superiorly, vomer medially, medial pterygoid bones laterally and palatine bone inferiorly. Atresia is the complete closure of the choanae, while stenosis is partial narrowing of the choanae. Atresia is either mixed bony and membranous (70%) or pure bony (30%), with no pure membranous cases found [4] (**Figure 1**).

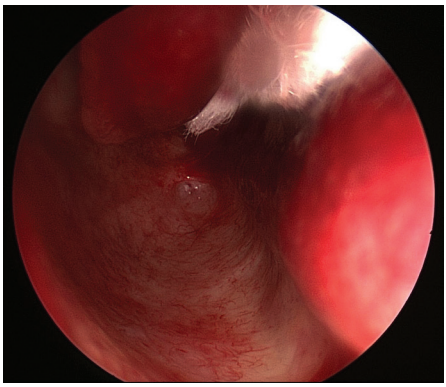

Figure 1. Endoscopic view through a zero degree rigid scope of Right nasal cavity with atresia seen.

Choanal atresia can be unilateral or bilateral with a ratio of 2:1. Same ratio applies to female-to-male distribution, and right to left sided atresia [5].

Many theories have been suggested for choanal atresia/stenosis formation[6]. The most widely accepted theory is persistence and incomplete canalization of the nasobuccal membrane. Other theories include persistence of the buccopharyngeal membrane from the foregut, abnormal persistence or location of mesoderm forming adhesions in the nasochoanal region, or misdirection of neural crest cell migration.

Choanal atresia can also be associated with syndromes, especially in cases of bilateral atresia. CHARGE (coloboma of the eye, heart defects, choanal atresia, developmental retardation, genitourinary anomalies, and ear anomalies) syndrome is the most commonly known, associated with mutations in the CHD7 gene (8q12.2)[7].

Clinical presentation

Clinical presentation is different in bilateral versus unilateral choanal atresia. Patients with associated syndromes also may have accompanying symptoms specific for other findings.

Bilateral choanal atresia usually presents with acute respiratory distress at birth. Newborns are obligate nasal breathers until 4-6 weeks of life. With both sides of their nose blocked, infants will have acute respiratory distress with cyclic cyanosis typically relieved with crying. They usually also have feeding difficulty because of their inability to breath while feeding.

Unilateral choanal atresia or stenosis usually present later in life. Chronic unilateral nasal obstruction is the most common presentation which can lead to chronic thick rhinorrhea and recurrent and chronic rhinosinusitis.

Diagnosis

For bilateral choanal atresia, diagnosis is usually made at birth. Inability to pass 6 or 8 fr catheter raises suspicion for atresia. Resistance to the catheter is usually encountered about 3-3.5 cm deep to the alar rim [5]. Nasal endoscopy using flexible nasopharyngoscope after proper nasal decongestion and suctioning allows for visualization of the area of atresia or stenosis.

Maxillofacial or sinus CT scan with thin 2-5 mm cuts allows definitive evaluation of the choanae, including type of atresia (bony versus membranous and bony), thickness of the atretic plate and relationship to skull base. This allows careful planning for repair. Choanal space less than 6 mm on CT scan indicates choanal stenosis [8].

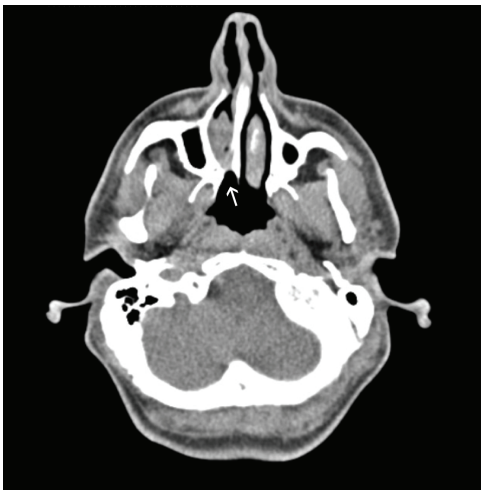

Figure 2. Axial CT scan nasal cavity showing choanal atresia on right side with wide open one on left

CT scan will also help in ruling out and differentiating other causes of nasal obstruction (**Figure 2**), including obstruction from nasal septal deviation, congenital piriform aperture stenosis, or nasolacrimal duct cysts.

Management

Management of the airway

Patients with bilateral choanal atresia usually present with respiratory distress at birth. They may often need emergent intubation. However, respiratory distress can be relieved with oral airway as well. McGovern nipple is frequently used. This is an intraoral nipple with a large opening by cutting its end off, secured in the mouth with ties around the infant's head [5]. If oral airway is not effective, intubation may be required. Tracheostomy is done when long term intubation is expected due to delay in atresia repair, or due to other syndromic anomalies. Patients with bilateral choanal atresia and CHARGE syndrome are more likely to fail repair [9] and more likely to require tracheostomy.

Patients with unilateral choanal atresia or choanal stenosis usually have no respiratory distress at birth. They may have snorting and labored breathing that is improved with crying.

Surgical management

Patients with bilateral choanal atresia require surgical repair early in life. Repair of unilateral atresia can be postponed until patient is older. Topical vasoconstrictive agents in the non-affected side allow that.

Different techniques have been described for surgical repair of choanal atresia. There is no consensus on the best technique. Many variables regarding the surgical approaches, use or non-use of stents, and duration of stenting, make accurate comparison between published studies limited. **Table 1**.

Table 1. Summary of data regarding surgical approach, use of stenting and Mitomycin C,

Surgical approach	Success rate	Complications
Transpalatal	up to 84%	distorted maxillary growth (cross bite, high palatal arch), oronasal fistula, velopharyngeal insufficiency, restenosis
Endoscopic nasal	67-88%	restenosis
Adjuvant treatment		
Stenting	variable	Alar injury, vestibular stenosis, columellar tear, and stent dislodgement or blockage.
Mitomycin C	no clear benefit	potentially carcinogenic, long term effect unknown

Transpalatal approach

This is one of the first approaches used for repair of choanal atresia, first described by Owens in 1965. Exposure is through a U-shaped mucosal flap based on the greater palatine vessels. The palatine bone distal to the greater palatine foramen, vomer, medial pterygoid plates and the atretic plate are drilled using a diamond bur drill.

This approach has high success rate [10], however, the rate of significant complications is high. Maxillary growth is affected, which lead to cross bite and high palatal arch. Other complications include wound breakdown and oronasal fistula formation, and velopharyngeal insufficiency.

Endoscopic transnasal approach

This approach has become the most widely used approach for repair of choanal atresia. It was first introduced by Stankiewicz in 1990 [11]. Different modifications of the original technique have been reported over the years.

Under endoscopic visualization, the bony posterior atretic plate is exposed after raising a laterally based mucosal flap. The atretic plate can be punctured with a urethral sound if thin, or drilled with microdrill. The opening is then enlarged by resecting the posterior septum (i.e. vomer) using backbiting forceps or powered instruments. Mucosal flap is then used to cover the exposed bone to prevent restenosis. Primary success rate using this technique has been reported between 67 and 88% [12]. A recent study by El-Anwar *et al* 2016 [13], 25 newborns with bilateral choanal atresia had endoscopic nasal repair without stenting. 18 patients (72%) had

a wide choana with adequate nasal breathing; Six patients (24%) had narrowing of the choana (<50%), still with adequate and satisfactory airway without feeding difficulties, and one patient (4%) developed restenosis (>50%) after 7 months, which necessitated repeated surgery.

Post-operative stenting to reduce restenosis is still controversial. It is widely used for variable post-operative duration. Recent studies have showed that restenosis rate is the same irrespective of stenting. A recent systematic review in 2016 [14] showed that success rates for bilateral choanal atresia repair were similar with and without the use of nasal stents. Thirteen studies (167 patients) included patients who were stented; the weighted pooled proportion of successful surgery was 65%. Mean duration of stenting ranged from 48 hours to 16 weeks. Six studies (42 patients) evaluated patients who were not stented; the weighted pooled proportion of successful surgery was 64%. This study also showed that the use of nasal stents may be associated with more complications, mainly alar injury, vestibular stenosis, columellar tear, and stent dislodgement or blockage.

Mitomycin C has also been used as an adjunct to surgical repair. It inhibits fibroblast proliferation and migration, and decrease scarring and restenosis. There is insufficient data to support the efficacy of Mitomycin C in choanal atresia.

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