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Choanal Atresia

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1. Background

Choanal atresia (CA) occurs in approximately one of every 5,000 to 9,000 live births. The earliest Western report was in 1755 in Germany by Roederer and the first documented repair was by Emmert in 1854 using a curved transnasal trocar [1]. Interestingly it is thought to occur more frequently on the right and more often in females [2-3].

Choanal atresia can be bony, membranous or mixed (bony and membranous). Most CA is bony-membranous (70%) with the remainder being bony and few, if any, membranous [4].

Choanal atresia can be unilateral or bilateral, but is more frequently unilateral (2:1 ratio) [5]. It can occur in isolation or as part of the CHARGE syndrome, first described by Hall in 1979 and composed of ocular Colobomas, Heart defects, choanal Atresia, Retarded growth and CNS defects, genitourinary hypoplasia, Ear anomalies [6]. Bilateral atresia is more commonly associated with CHARGE syndrome than unilateral atresia. The CHD7 gene has been implicated in the pathogenesis of CHARGE [7].

CA can also be associated with the Treacher Collins, Crouzon's, Pfeiffer or Apert's syndromes. Chromosomal anomalies are found in 6% of 444 children with choanal atresia [8]. Of the remaining 94% without chromosomal abnormalities, 47% had associated malformations. In their paper from 1964 Flake and Ferguson also note congenital heart disease, esophageal atresia, tracheo-esophageal fistula, meningocele and craniosynostosis among their 40 patients with CA [9]. More recently, in 2003, Samadi et al also found an association with heart disease (19%). In addition, they noted an association of CA with upper and lower airway diseases (32% and 23%, respectively) as well as gastrointestinal tract disorders (18%). Among those patients with bilateral CA, there were correlations with CHARGE, OSA, hematological problems, prematurity and failure to thrive [3].

2. Embryology / Etiology

Between the 35th and 38th days of embryologic development, a membrane of nasal and oral epithelium, separating the primitive mouth and nose, ruptures to form the choana. Eventually these primitive choana take on a more posterior and vertical location. Most believe CA results from persistence of the bucopharyngeal membrane. Other theories of CA development postulate that the disorder arises from medial outgrowth of the horizontal and vertical processes of the palatine bone, abnormal mesodermal or neural crest migration, persistence of the nasobuccal membrane of Hochstetter or mesodermal adhesions [1,10].

It is also possible that CA results from abnormalities in vitamin A metabolism, which leads to abnormalities in fibroblast growth factor (FGF) expression (similar to craniosynostosis) [11]. Barbero et al found patients with nonsyndromic CA occasionally (10/61) had mothers were who hyperthyroid. It is unclear whether

it is the hyperthyroidism itself or the medications (methimazole) that may be associated with CA, but thyrotropin causes alterations in FGF signaling [12].

3. Signs/Symptoms

Bilateral choanal atresia usually presents at birth with neonatal respiratory distress. The obstruction is generally cyclical ("cyclical cyanosis") with obstruction at rest and then relief when the baby takes a breath. Babies are obligate nasal breathers (until approximately 2-3 weeks old), but nasal obstruction is bypassed when they take a breath through their mouth such as with crying. It is often difficult for these children to feed; aspiration is not uncommon. In White's study of 30 children with Choanal Atresia associated with CHARGE, 60% (18 /30) had evidence of aspiration [13].

Unilateral choanal atresia often presents later (average 5-24 months), with unilateral rhinorrhea (usually thick mucous) or nasal obstruction or sometimes no symptoms at all. There have been reports of unilateral CA being recognized only after unsuccessful septoplasty in adults [2]. Unilateral atresia in association with other syndromic features may present earlier or be discovered on a neonatal exam.

Intrauterine ultrasound diagnosis is now possible though it can be difficult to distinguish this from severe septal deviation. Color flow Doppler with respirations can aid in the identification of CA as well [14].

4. Differential Diagnosis

The differential diagnosis should include, neonatal rhinitis, deviated or dislocated nasal septum, pyriform aperature stenosis, bilateral nasolacrimal duct cysts, nasal foreign body, turbinate hypertrophy, nasal mass such as polyp, dermoid, glioma or encephalocele or an occluding nasopharyngeal mass such as a Thornwaldts cyst or chordroma.

5. Physical Exam

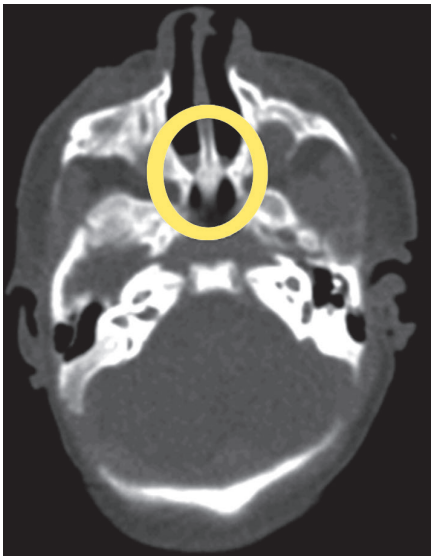
A full head and neck exam should be performed. Attention should be paid to any other congenital anomalies or syndromic features, such as those occurring with the CHARGE syndrome. In CA, a cotton wisp placed under the nare will not move due to a lack of airflow, and there is absence of fog when a mirror is placed under the nose. Ultimately the diagnosis is suggested when a number 6 French catheter is unable to be passed beyond approximately 32mm from the alar rim (the approximate location of the choana in a neonate) [15]. Methylene blue can be placed in the suspected atretic nare, and the oropharynx observed for any evidence of transit. Diagnosis is facilitated with a flexible fiberoptic nasopharyngolaryngoscope (NPL) to assess the nasal vaults. The benefit of the NPL is in permitting anatomic assessment of both endonasal cavities, ruling out alternative diagnoses, and perhaps in characterizing the bony versus membranous composition of the CA.

6. Imaging

Non-contrast enhanced computed tomography (CT) scan in axial and sagittal planes can be useful in evaluating CA. CT scan can help evaluate for other causes of symptoms and for operative planning. On axial CT scan the appearance of the nasal vaults may be described as a "champagne flute" (**Figure 1**)[16]. The pterygoid plates may be medialized, as can the lateral nasal wall, and the

vomer is often thickened. The nasopharynx may be narrowed, and a high arched palate seen. These other anatomical anomalies are present often in children with nonsyndromic CA[17]. There is often dependent mucus present as well on imaging, and for this reason it is helpful if the nares are suctioned prior to the CT scan because mucous can be difficult to distinguish from an atretic plate [4].

Figure 1: Bilateral choanal atresia. Computed tomography (CT) (head) without contrast. Axial cut. Atresia plate at posterior nasal vault (circle) demonstrating "champagne flute" appearance characteristic for choanal atresia. (*Reprinted from Int Journal of Pediatric Otorhinolaryngology; Shah UK, Daniero JJ, Clary MS, Depietro JJ, Johnston DR: Low birth weight neonatal choanal atresia repair using image guidance, 2011:75(10); 1337-40, with permission from Elsevier.)*



7. Management

Bilateral choanal atresia is an airway emergency. These patients may benefit from a McGovern nipple or oral airway in the short-term, but obtaining a definitive airway is of paramount importance including repair if possible, or sometimes intubation (or tracheotomy in the most desperate cases) in the interim. Placement of an oral airway will break the seal between the tongue and palate allowing air passage. Unilateral atresia generally warrants an elective repair.

8. Surgical Repair

There are many approaches used for repair of choanal atresia including transpalatal, transseptal, and transnasal endoscopic. In general, there is little agreement as to the best approach. While there is much published literature, it is generally of small series, often with conflicting results. Furthermore, in neonates, the anatomy is much smaller making some approaches more suited for the older children with unilateral atresia. Image guidance systems, particularly when there are other craniofacial anomalies, are often quite useful as an adjunct [16]. Instruments used include a range of endoscopes (0 degree to 120 degree), dilators such as ureteral sounds or balloons, otologic instruments, a drill, a micodebrider

and/or lasers. Generally the carbon dioxide (CO_2) or potassium-titynyl-phosphate (KTP) laser are used. A contact diode laser (810 nm) has also been reported [18].

Timing of repair depends first and foremost on whether it is unilateral atresia (which is generally elective, and often repaired at school age when the nares are larger allowing better instrumentation) or bilateral atresia (which is an airway emergency and often repaired urgently). One should keep in mind however, with unilateral atresia in the young infant, if the other side becomes obstructed due to an upper respiratory infection for example, an airway emergency can develop. Patient size and comorbidities also affect the timing of surgery.

The first technique used in repair of CA was a blind nasal puncture with a curved trocar. This not only carried a high incidence of restenosis but also put surrounding structures at risk such as the nasal septum, Eustachian tube orifice or even the cervical spine due to limited visualization [19]. This is particularly true in children with other craniofacial anomalies and altered anatomy. That being said, it can still be used in an emergency to establish a nasal airway, now with ureteral sounds or Fearon dilators. Subsequently a similar procedure was performed with a microscope which allowed some increased visualization, but still very high rates of restenosis.

The first report of a successful transpalatal repair was in 1931 by Blair [20]. It usually involves a curvilinear incision, though reports of vertical, horizontal and cruciate incisions also exist. Mucoperiosteal flaps are raised off the hard palate, with care not to injure the greater palatine neurovascular bundle, after which a portion of hard palate and vomer are removed. Some assert this approach takes longer, putting any unstable patient at increased risk. This approach also includes the risk of temporary or permanent palate dysfunction or palatal fistula, or even dental occlusion abnormalities, but it does offer excellent exposure, particularly for revision cases or children with skull base anomalies.

Today most repairs are performed transnasally, using endoscopes. A range of endoscopes are needed from 0 degree to 120 degree for adequate visualization of the atretic plate. A variation on transnasal repairs, *transseptal* repairs, consists of making a window/opening through the posterior septum, just anterior to the choana which can allow working through one nare and endoscopic visualization through the other nasal vault.

Some surgeons perform a combination of transnasal and transpalatal approaches with endoscopes and an assistant.

Topical steroid drops perioperatively and intraoperative application of mitomycin C may reduce scarring and granulation formation. Mitomycin C works by inhibiting fibroblast proliferation, having a direct cytotoxic effect, and promoting fibroblast apoptosis [21]. However, there is concern about its long term effects on tissue, which are still largely unknown (e.g. cancers). Some surgeons recommend a full course of postoperative antibiotics while others use none at all. Gastroesophageal reflux management may be important in preventing granulation tissue.

Post-operative stenting is also somewhat controversial. While some believe it improves patency of the choana, others feel that stenting creates a foreign body reaction and granulation tissue formation. Furthermore, stents can cause pressure

necrosis of nearby structures such as the ala or palate or airway distress if they become crusted and are not monitored carefully. There are also many options for stents including cut endotracheal tubes (usually 3.0 or 3.5) [22]. Theogaraj et al., advocate for a double barreled silicone Elastomer tube with a wire within its walls which prevents stent collapse [23]. Length of time of stenting can range from a few days up to 3 months [24]. In most patients, placement of a stent necessitates a return to the operating room for removal, which may be risky in medically complex patients. Advocates of stents argue that stenting permits fewer re-operations in the long-run.

Post-operatively, children often require significant home care and families should be instructed and trained on how to clean and suction the nares and/or stents. They should also be trained in CPR should that become necessary with acute nasal airway obstruction from crusting. Sometimes home nursing may be necessary.

9. Prognosis

A planned return to the OR is often required in patients with stents, but also those patients experiencing any new or increased symptoms should be re-evaluated in the office or operating room. Those with craniofacial syndromes or undergoing another airway procedure may deserve a "re-look" as well.

While both unilateral and bilateral choanal atresia often require multiple procedures for restenosis, bilateral, in general, require more. A total of 4.9 procedures were performed for bilateral CA versus 2.7 total procedures for unilateral CA in one study.[2] In general, following the procedure, the choanae should accommodate passage of a number 8 French catheter.

Pirsig reviewed more than 100 papers dealing with choanal atresia and different operative approaches and he found that the transnasal approach was successful in up to 80% of the cases [25]. Pirsig found that the transpalatal approach was successful in up to 84% of the cases [25].

10. Complications

The most common complications are scarring and restenosis along with granulation formation. Skull base injury is rare but more likely in children with craniofacial malformations. The effects of transpalatal or transseptal approaches on facial growth are largely unknown. While most studies suggest normal craniofacial growth after transnasal approaches for other procedures, there is concern for palatal abnormalities and subsequent altered occlusion with transpalatal approaches. Freng et al reported 52% of his patients with the transpalatal approach had stunting of palatal growth [26].

11. Future

Advances in intraoperative image guidance, drug eluding stents, and robotic surgical techniques offer more precise surgery and reduced complications. Increased intrauterine diagnosis may facilitate expectant planning for immediate airway protection of children born with CA and permit expedient surgical correction.

12. Conclusion

CA is a rare but life threatening disorder which requires expedient multidisciplinary management. Radiographic imaging, prenatal diagnosis, and surgical techniques continue to evolve.

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