Choanal atresia

Choanal atresia is a congenital narrowing of the nasal airway at the posterior choanae. Early diagnosis is important for prompt treatment as this may be protracted, involving multiple surgical procedures. This article discusses the presentation, assessment and management of the child with suspected choanal atresia.

Choanal atresia is an occlusion of the posterior nasal airway at the level of the posterior choanae. Based on a study in 1910 traditional teaching has suggested that around 90% of cases were bony and 10% were membranous. However, evidence suggests than bony occlusion makes up around 30% of cases while the remainder involve a mixed bony and membranous obstruction (Brown et al, 1986).

The incidence of choanal atresia is around 1 in 5000 to 1 in 8000 births (Schwartz and Savetsky, 1986; Stankiewicz, 1990). The female:male ratio is 2:1. Most cases are sporadic but there is a familial association in about 10% of cases. Of particular importance is the association with the CHARGE syndrome although there are also associations with other chromosomal anomalies including Down syndrome. There is some evidence that a patient with bilateral choanal atresia is more likely to present in combination with associated anomalies, e.g. craniosyntoses (Apert syndrome, Crouzon syndrome, Pfeiffer syndrome), than a child with unilateral choanal atresia (Burrow et al, 2009).

Aetiology

There are a number of embryological theories for choanal atresia including failure of neural crest cell migration and persistent of the buccopharyngeal membrane of the foregut but there is no conclusive evidence to support any of the theories (Hengerer et al, 2008).

Presentation

Bilateral choanal atresia presents with airway obstruction at birth because neonates are obligate nasal breathers for the first 6 weeks of life. The presentation includes airway obstruction and apnoea during quiet respiration. Paradoxical cyanosis may be encountered with oxygen saturations improving with crying, because neonates mouth breathe when crying. There may be thick tenacious secretions in the nasal airway. On examination, it is not possible to pass a 6F suction catheter into the nasopharynx from either nasal airway.

If possible, the nasal airway may be assessed with a paediatric 1.9 mm flexible nasendoscope to directly visualize the atresia. This should only be carried out once the airway has been secured and should not delay the immediate management of the patient.

Unilateral choanal atresia usually presents with a ipsilateral mucoid discharge. Presentation is usually delayed and often only identified in older children or even adults. In the older patient there is usually unilateral nasal obstruction.

The atresia may be diagnosed by passing a catheter as above or by flexible nasendoscopy if tolerated. A difference in nasal patency can also be demonstrated by misting caused by condensation on the surface of a laryngeal mirror held close to the airway. A lack of misting implies little or no airflow.

CHARGE syndrome

CHARGE syndrome is an autosomal dominant condition associated with a number of congenital anomalies including Coloboma (iris), Heart defects (e.g. tetralogy of Fallot, ventricular septal defect), Atresia of the choanae, Retardation growth and development, Genital hypoplasia, Ear abnormalities (e.g. Mondini dysplasia, absent or malformed ossicles). Between 7 and 29% patients with choanal atresia will have CHARGE syndrome (Corrales and Koltai, 2009). Other associations include cranial nerve palsies (e.g. facial palsy), cleft palate and trachea-oesophageal fistula. Molecular testing for mutations in the CHD7 gene confirms the diagnosis in most cases.

Differential diagnosis

There are a few rare conditions which may be misdiagnosed as choanal atresia because of their similar presentation in neonates.

Congenital nasal pyriform aperture stenosis

This is an uncommon cause of airway obstruction that presents in a similar fashion to bilateral choanal atresia. There is bony overgrowth of the medial aspect of the nasal process of the maxilla which causes significant nasal obstruction more anteriorly in the airway than choanal atresia. Examination of the nose by anterior rhinoscopy and/or flexible nasendoscopy may show an anterior bony obstruction of the nasal vestibule. Diagnosis can be difficult clinically but computed tomography imaging is diagnostic and delineates the site of obstruction. It may also show the classical association with a single central upper incisor.

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Nasolacrimal duct mucocoele (dacrocystocoele)

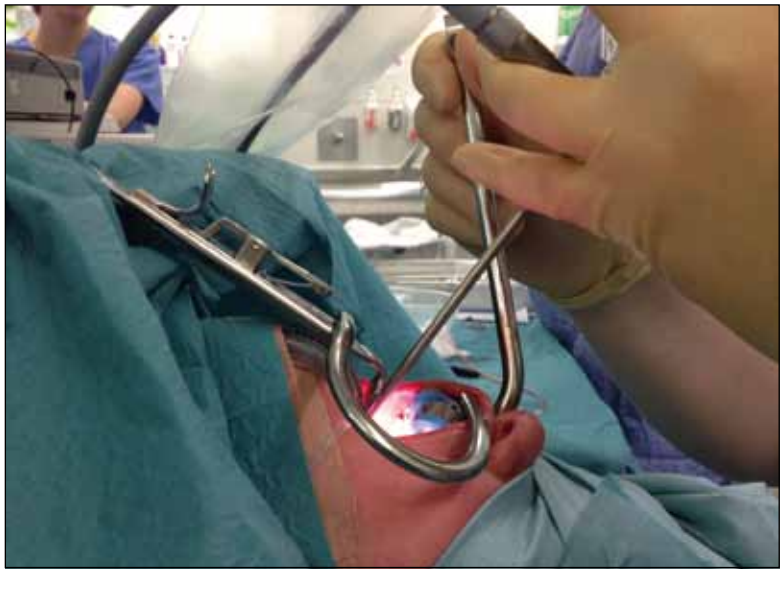
This is an uncommon cause of congenital nasolacrimal duct obstruction. Clinically it may be obvious if there is obstruction leading to a soft cystic mass around the medial canthus (just inferior and medial to the orbit). However, it may also present with a nasal cyst arising from the inferior meatus and causing nasal obstruction. Diagnosis is usually made by endoscopic examination but computed tomography is often required to rule out the diagnosis of meningo-encephalocoele.

Imaging for choanal atresia

An axial high resolution computed tomography choanae with contrast with 1–1.5 mm slices is the imaging of choice to make the diagnosis. Computed tomography imaging also provides useful information regarding the anatomy of the obstruction. In particular the nature and degree of bony involvement of the posterior vomer and lateral nasal wall can be assessed which is not possible with magnetic resonance imaging. It may also identify other diagnoses that can present in a similar fashion to choanal atresia, e.g. nasal pyriform aperture stenosis. The authors advocate including fine cut computed tomography temporal bone at the same time as the computed tomography choanae to identify any middle and inner ear abnormalities.

Figure 1. McGovern nipple is a large nipple or pacifier with the end cut off which acts as an oro-pharyngeal airway. It can be secured using tapes or ties.

Figure 2. Operative position with Boyle–Davis gag to open mouth, 120° endoscope directed into nasopharynx to visualize choanae and urethral sound inserted trans-nasally.



Other investigations

A diagnosis of choanal atresia in the neonate should always prompt investigation for any other components of CHARGE syndrome even if they are not clinically obvious at the time of diagnosis. A multidisciplinary team approach is recommended.

All patients should have an echocardiogram and renal ultrasound. Examination of the eyes by a paediatric ophthalmologist to evaluate the presence or extent of coloboma is important. The authors advocate review by a clinical geneticist with appropriate cytogenetic analysis and/or molecular genetic testing. There is a much reduced incidence of associated congenital anomalies in patients with unilateral choanal atresia compared with bilateral atresia.

If the patient presents as an older child or even adult, it is extremely unlikely that unilateral choanal atresia is present as a part of the CHARGE association. In these cases, a thorough history and clinical examination should suffice rather than more intensive investigation. Further investigations and/or genetic testing would only be indicated if there was clinical suspicion of pathology or anomalies.

Management

Bilateral choanal atresia

Bilateral choanal atresia is a medical emergency as neonates are obligate nasal breathers during the first 6 weeks of life. An oropharyngeal anaesthetic airway is the intervention of choice. If unavailable, a large intra-oral nipple can be modified by having its end cut off to provide an airway. This can be secured with ties around the back of the head and is known as a McGovern nipple (*Figure 1*). A small oro-gastric tube should be passed to allow feeding. If there is persistent respiratory compromise despite these initial measures, there should be a low threshold to secure the airway via oral intubation.

Surgery should be carried out as early as possible, ideally in the first week of life. Intranasal correction of the choanal atresia using endoscopic guidance is the treatment of choice (*Figures 2–5*).

The procedure is performed under general anaesthetic with the patient in the supine position. A Boyle–Davis gag is used to hold the mouth open and the soft palate is retracted using either a suction catheter in the contralateral nostril (unilateral cases) or a suture though the

base of the uvula (in bilateral cases). The choanae are visualized by passing a 120° endoscope into the nasopharynx via the mouth, which provides a retrograde view of the choanae. The atretic choanae can then be progressively dilated using urethral sounds. A choanal diamond burr drill can also be passed transnasally to open up the atretic area.

The risk of postoperative stenosis is higher in the neonate and traditionally in cases of bilateral choanal atresia many surgeons elect to leave stents in situ for several weeks. There is no significant evidence to conclusively advocate the use of stents and a number of studies cite higher stenosis rates and complications, e.g. granulation formation (Teissier et al, 2008; Zuckerman et al, 2008). Many centres use shorter periods of stenting or no stents at all.

There are numerous methods of opening up the atretic area including choanal dilators (*Figures 4–6*), cold steel instrumentation, choanal drill and laser. A recent Cochrane review suggested that there is no evidence for any significant advantage of any specific surgical technique (Cedin et al, 2012).

Traditionally a transpalatal approach was used, particularly in refractory cases later in life. Complications including heavy bleeding, fistula formation, high arched palate deformity and a high incidence of dental malocclusion have resulted in a significant reduction in the use of this technique (Freng, 1978; Keller and Kacker, 2000). *Figure 7* summarizes the management of bilateral choanal atresia.

Unilateral choanal atresia

Patients with unilateral choanal atresia are often undiagnosed until early childhood as they do not suffer from airway obstruction. In any case, there is no urgency for surgery and so intervention is usually delayed until at

Figure 3. Bilateral choanal atresia. View with 120° endoscope in the oropharynx giving a retrograde view of the abnormality.

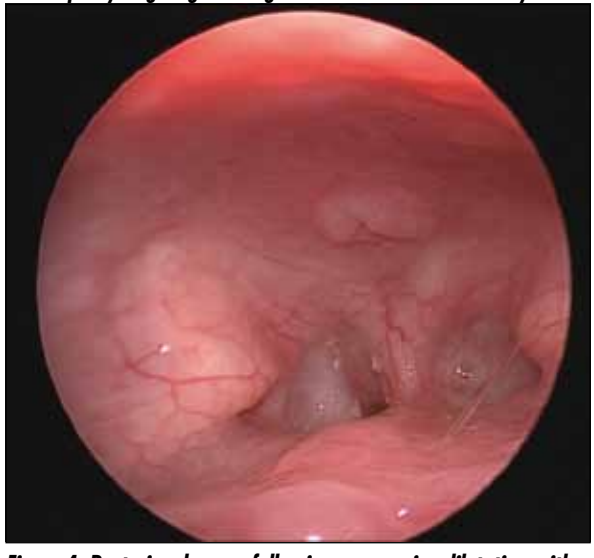


Figure 4. Posterior choanae following progressive dilatation with urethral sounds.

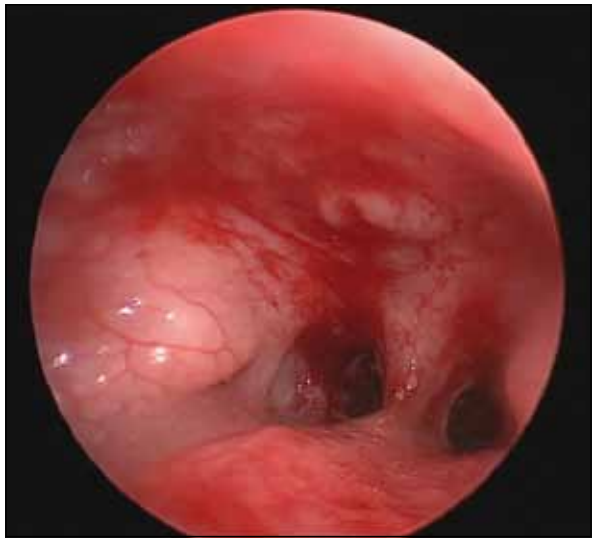


Figure 5. View with 120° endoscope in oropharynx shows metal urethral sound (dilator) passed through the nasal airway and through the right-sided choanal atresia.

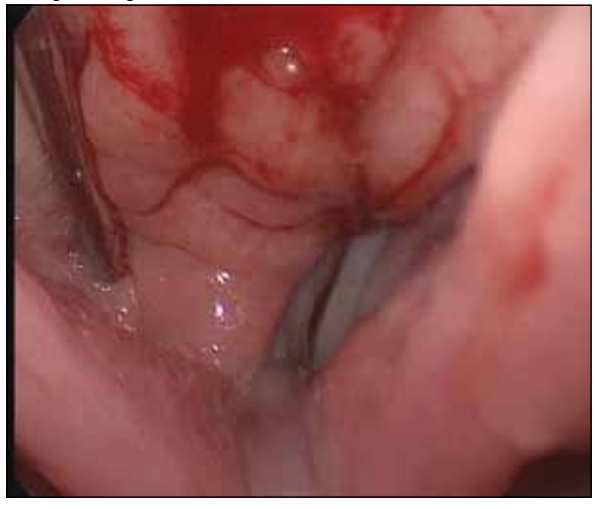


Figure 6. Appearance of unilateral right-sided choanal atresia following dilatation. A suction catheter can be seen in the left nasal airway which is used to lift the palate and aid surgery.

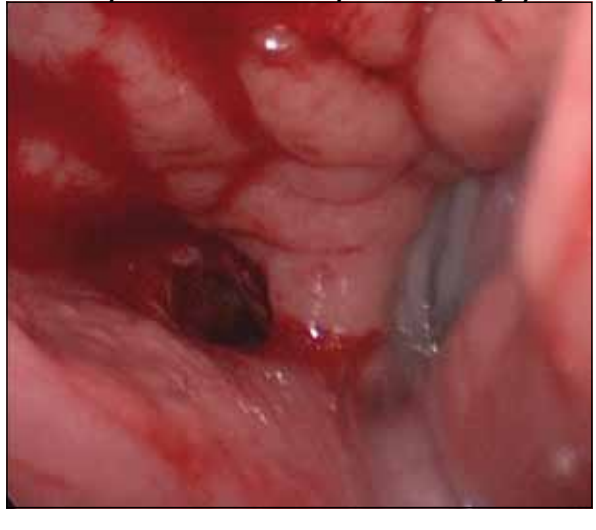


Figure 7. Flowchart summarizing the management of bilateral choanal atresia.

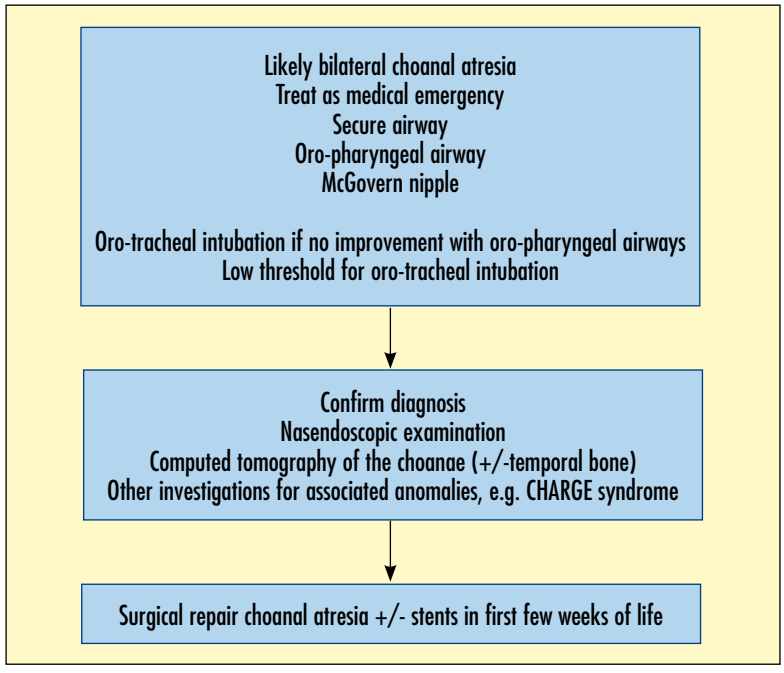
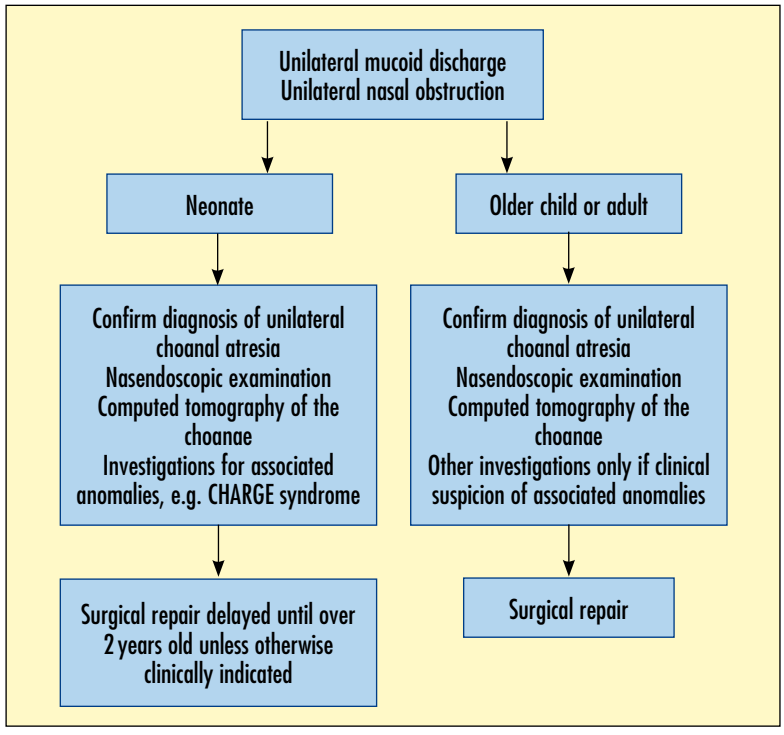


Figure 8. Flowchart summarizing the management of unilateral choanal atresia.



KEY POINTS

- Bilateral choanal atresia in neonates is an airway emergency and requires urgent securing of the airway.
- Management of choanal atresia requires a multidisciplinary team approach particularly to identify any associated anomalies.
- Unilateral choanal atresia may be missed and present in the older child with unilateral mucoid discharge or nasal obstruction.
- Transnasal endoscopic repair by a number of techniques is the surgical treatment of choice.

least 2 years of age unless it is causing significant respiratory compromise. For example, many patients with CHARGE syndrome have other associated levels of airway obstruction, e.g. laryngomalacia, hypopharyngeal collapse or subglottic stenosis, that may affect timing of choanal atresia surgery.

The endoscopic approach is the surgical intervention of choice with the same techniques that are used in bilateral choanal atresia surgery. It is not uncommon for patients to require multiple procedures as a result of restenosis. Male gender, bilateral disease, associated congenital anomalies, low birth weight and small stent size are potential risk factors for restenosis of choanal atresia (Elloy et al, 2008).

Figure 8 summarizes the management of unilateral choanal atresia.

Conclusions

Choanal atresia is a rare congenital abnormality of the nasal airway. Bilateral choanal atresia is a medical emergency and prompt diagnosis is vital for appropriate management. It is also important to be aware that management of choanal atresia requires a multidisciplinary team approach and many medical and surgical specialties may be involved in investigating other associated anomalies. BJHM

Conflict of interest: none.

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