

CHOANAL ATRESIA SURGERY

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Choanal atresia is defined as a blockage of the posterior nasal openings. It is a rare condition and has an incidence of 1/7000 live births. It can be unilateral or bilateral. The blockage is purely bony in 30% of cases; in the remaining 70% it is a mixed bony and membranous obstruction (*Figure 1*). Complete total bony obstruction is extremely rare.

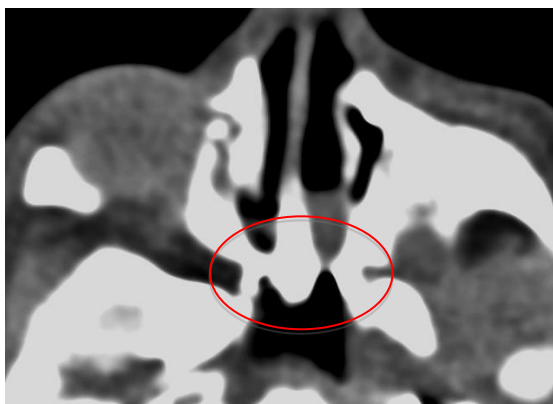


Figure 1: Axial CT demonstrating bilateral choanal atresia with left-sided mixed bony / membranous atresia and right-sided bony atresia

It is believed that membranous obstruction occurs due to failure of the buconasal membrane to rupture sometime between the 5th and 6th weeks of foetal development. Bony obstruction occurs due to a combination of

- A narrow nasal cavity
- Medial thickening of the medial pterygoid plate
- Lateral thickening of the vomer

Unilateral choanal atresia (*Figures 2, 3*) is most commonly an isolated problem. However 75% of bilateral cases (*Figure 4*) have associated syndromic or congenital abnormalities. These include CHARGE syndrome (coloboma, heart defects, choanal atresia, retardation of growth, genital abnor-

malities and ear abnormalities) and other syndromes that have associated craniofacial abnormalities such as Treacher Collins, Crouzon syndrome, velocardiofacial syndrome or craniosynostosis.

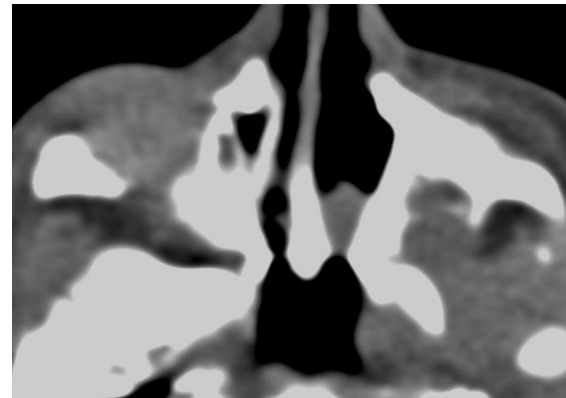


Figure 2: Axial CT scan demonstrating unilateral left-sided choanal atresia

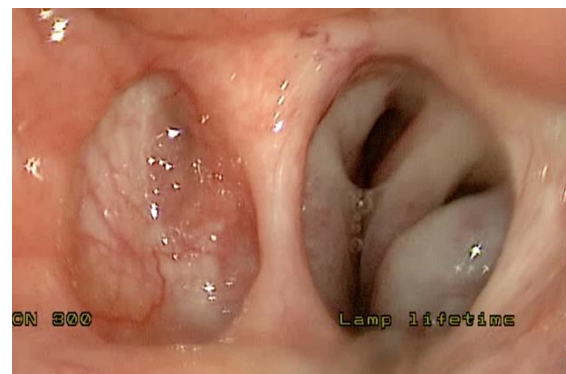


Figure 3: Retropalatal 70° endoscopic view demonstrating unilateral choanal atresia

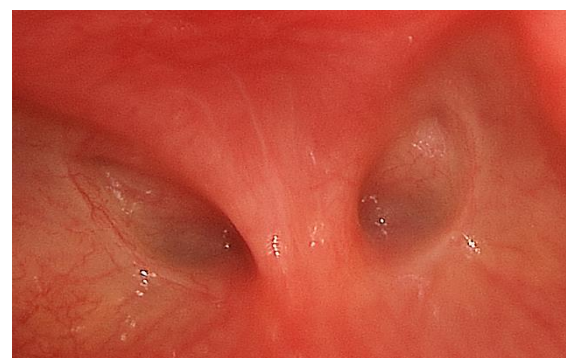


Figure 4: Retropalatal endoscopic view demonstrating bilateral choanal atresia

Differential diagnosis

The differential diagnosis of choanal atresia includes

- Nasal pyriform aperture stenosis (*Figure 5*)
- Deviated nasal septum
- Dislocated nasal septum
- Septal haematoma
- Neonatal rhinitis
- Mucosal swelling
- Turbinate hypertrophy
- Encephalocele
- Nasal glioma
- Intranasal dermoid
- Chordoma
- Teratoma

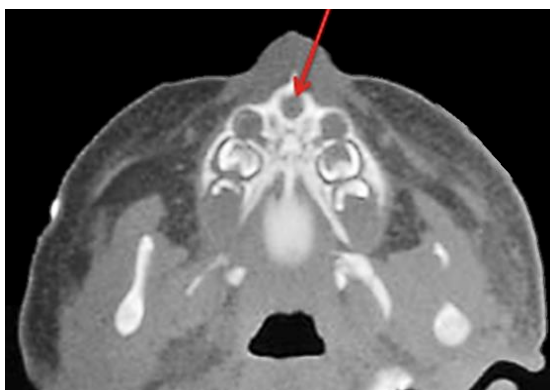
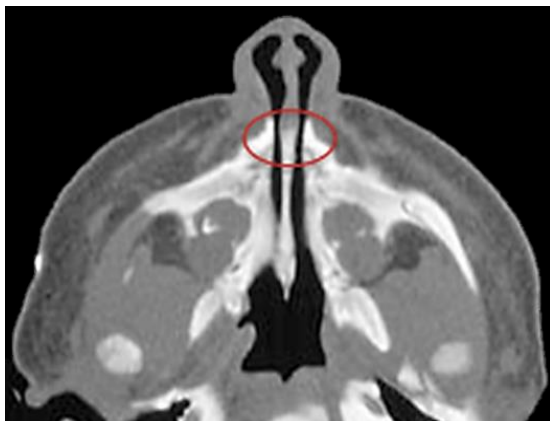


Figure 5: Axial CT scan in a patient with pyriform aperture stenosis and with a single central incisor (Red arrow)

Presentation

Unilateral choanal atresia may not present until later in life with complaints of a unilateral blocked nose or profuse unilateral nasal discharge (*Figure 6*).



Figure 6: Previously undiagnosed membranous choanal stenosis in an adult

On the contrary, **bilateral choanal atresia presents as an acute emergency at birth** as respiratory distress because newborns are obligate nasal breathers. A common feature of bilateral atresia is *cyclical cyanosis* whereby a newborn has respiratory distress with cyanotic episodes that improve on crying, and worsen on feeding.

Clinical evaluation

1. Two simple tests can be used to **demonstrate airflow** through the nose to exclude choanal atresia
 - Movement of a thin piece of cotton wool held directly underneath the nares during inspiration and expiration
 - A shiny metal tongue depressor held under the nares will reveal nasal misting in patients with patent nasal airways

2. **Failure to pass an 8F (or smaller) suction catheter** into the nasal cavity more than 5.5cm from the alar rim can suggest the diagnosis
3. **Direct visualisation** with a flexible nasendoscope after nasal suctioning will definitively diagnose the condition (Figure 7)

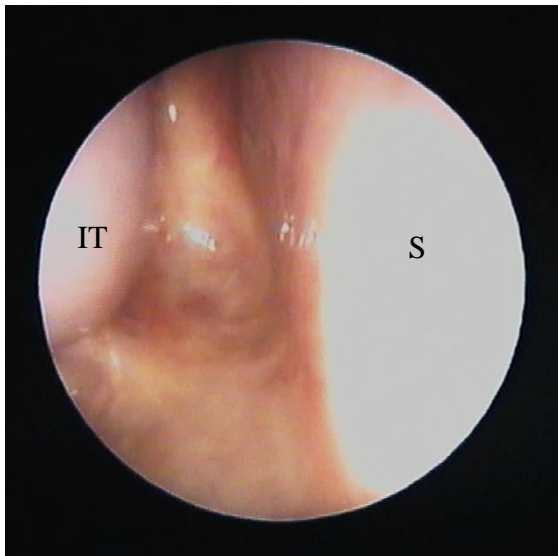


Figure 6: Transnasal endoscopic view of previously undiagnosed (R) unilateral choanal atresia in an adult (S = Septum; IT = inferior turbinate)

Radiological evaluation

The diagnosis can be confirmed with a **lateral skull X-ray** after filling the nose with **radioopaque dye** and demonstrating hold-up of the dye in the posterior nasal cavity (Figure 7).

The atresia can be subclassified by **CT scanning** into bony or mixed bony-membranous types (Figure 1). It is important to apply nasal decongestant (0.5% xylocetazaline) 30 minutes prior to the CT scan and to suction the nose immediately before the CT scan to remove all nasal secretions to enable accurate radiological diagnosis.

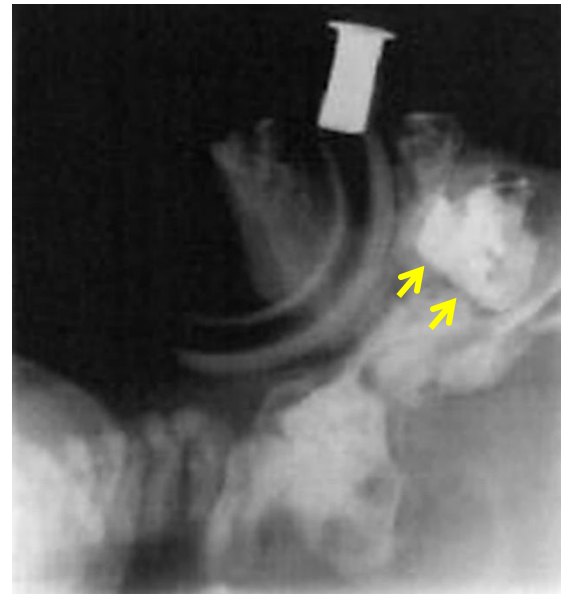


Figure 7: Lateral X-ray after filling the nose with radio-opaque dye demonstrates hold-up in the posterior nasal cavity (yellow arrows). Note oropharyngeal airway to maintain the airway

Immediate Emergency Management

An **oral airway** is generally adequate to temporarily secure an airway in a newborn with bilateral choanal atresia and respiratory distress (Figure 8).



Figure 8: Oral airway secured with tape

A **McGovern nipple** or a standard feeding **bottle teat or dummy with the tip cut off** (Figure 9) can also be used to facilitate oral breathing. In the short-term this has the effect of stenting the oral cavity open

to allow mouth breathing; however it is not a long-term solution. Alternative options include a **Guedel oropharyngeal airway** (Figure 10). If an oral airway is not effective one has to proceed to **endotracheal intubation**.



Figure 9: Standard dummy with the tip cut off



Figure 10: Guedel oropharyngeal airway

Feeding should be established via orogastric tube until surgical correction of the atresia is performed.

The patient should ideally be managed in a **neonatal or paediatric intensive care unit** with involvement of neonatologists, geneticists, paediatric ENT surgery and other specialties as required.

Timing of surgery

Surgery is urgently required for bilateral disease and symptomatic unilateral disease. Definitive surgery can often be delayed in patients with asymptomatic unilateral atresia. However, syndromic babies can have respiratory complications from unilateral atresia that require earlier surgery.

Preoperative Workup

All patients should undergo a full workup and evaluation once the airway has been secured, including for possible syndromic associations. The minimum investigations beyond the initial **CT scan** include **ultra-sonography of the cardiac and renal systems** as well as **ophthalmological** and **audiological** assessment.

Genetic testing may be useful for patients with features of CHARGE syndrome; 67% are due to a recognised mutation in the chromodomain helicase DNA-binding protein-7 (CHD7) gene. The remainder are related to mutations in unrelated genes or other cytogenetic anomalies such as chromosome deletions.

Surgical management

The key to a successful outcome, regardless of surgical approach, is to create a large, 3-dimensional opening by a combination of (Figure 11)

1. Puncturing the membranous obstruction
2. Widening the medial bony thickening of the medial pterygoid plate
3. Thinning the thickened vomer

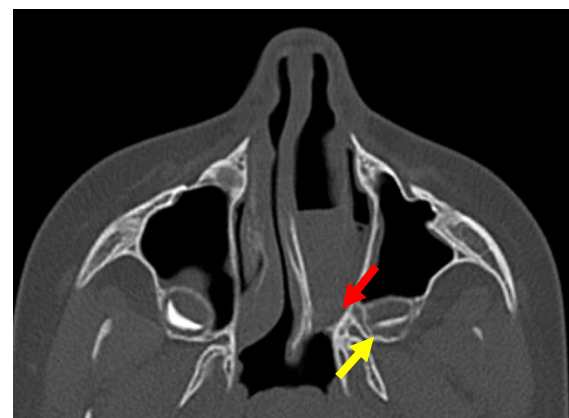


Figure 11: CT demonstrates need to thin the thickened vomer (red arrow) and widen the medialised medial pterygoid plate (yellow arrow)

Traditionally the atresia was approached by transnasal / retropalatal, transseptal or transpalatal routes, but the last two are rarely used today. Transnasal punctures alone have high rates of restenosis.

Multiple procedures are often required to achieve a permanent opening in the posterior nares.

Retropalatal endoscopic approach

This endoscopic technique is currently favoured by most paediatric otolaryngologists. The benefit of the retropalatal approach is that the newborn's nasal cavity is usually too small to accommodate both a 0° endoscope and a drill.

Transnasal instrumentation

Visualisation is achieved with a retropalatal 120° Hopkins rod (Figure 12).



Figure 12: 120 degree rigid endoscope

Additional equipment includes curved urethral sounds and powered drills (Figures 13, 14).



Figures 13a, b: Urethral sounds used for initial perforation of the atresia



Figure 14: Powered drill (Choanal atresia bur)

Surgical steps

- Surgery is done under general anaesthesia with a downwards-facing Rae tube



Figure 15: Rae tube

- Tape the Rae tube to the lower lip in the midline as for a tonsillectomy
- Lay the patient supine with a shoulder roll in place
- Decongest the nose using 0.5% xylocaine
- A cleft palate / tonsil gag is used to open the mouth and placed in suspension (Figure 16)



Figure 16: Cleft palate / tonsil gag

- Retract the soft palate
 - With unilateral atresia, insert a nasogastric tube on the **unaffected** side and pull it out of the mouth
 - With bilateral choanal atresia, place a suture in the soft palate to retract the palate until such time as a nasogastric tube can be inserted
- Insert the 120-degree rigid endoscope through the mouth and inspect the nasopharynx and the choanal atresia
- Insert a urethral sound into the nose and puncture the atretic plate under direct view with the 120 degree scope (*Figure 17*)

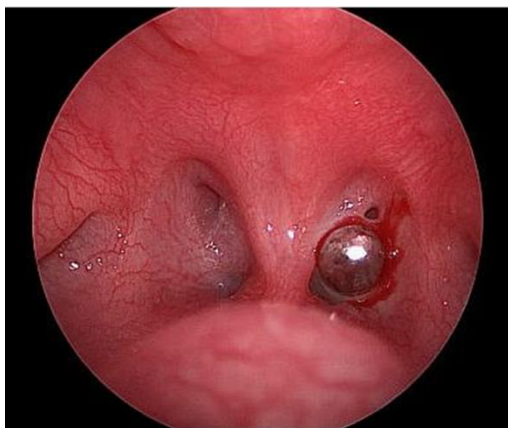


Figure 17: Transnasal puncture of membranous atresia with a urethral sound

- The urethral sound may pass easily in a membranous obstruction or less easily in a purely bony atresia
- The most common position of the membranous section of an atresia is in the inferomedial corner at the junction of the vomer and the palate (*Figure 18*)

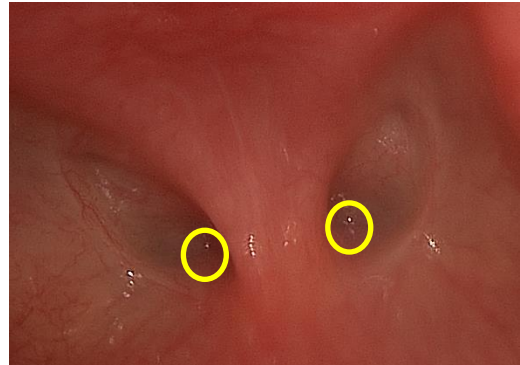


Figure 18: Junction of vomer and palate

- Puncturing the atresia is however not sufficient to relieve the obstruction in the long-term, as rapid restenosis will occur without additional bone removal
- Insert the powered drill transnasally with a 4mm choanal diamond burr.
- Drill the medial pterygoid plate under direct vision taking care preserve as much mucosa as possible (*Figure 19*)



Figure 19: Bilaterally opened choanal atresias after laterally drilling the medial pterygoid plates; vomer intact

- If the vomer impinges on the choana, it can be resected

- If necessary a common posterior choanal opening can be created by removing the posterior few millimetres of the vomer
- Once again as much mucosa as possible is preserved in an attempt to leave as little exposed bone as possible at the end of the procedure
- Obtain haemostasis by using neurosurgical patties soaked in 1:10,000 adrenaline

To stent?

Stents may be fashioned from cut endotracheal tubes, suction catheters, or custom-made silicone stents can be used (*Figure 20*).

Traditionally almost all patients were stented. Currently stenting is however controversial as stents can cause granulations and scarring.

Stents are still preferred in very small babies that are surgically challenging. In such cases stenting preserves patency for the duration of the stent's placement and allows for weight gain to occur prior to revision surgery.

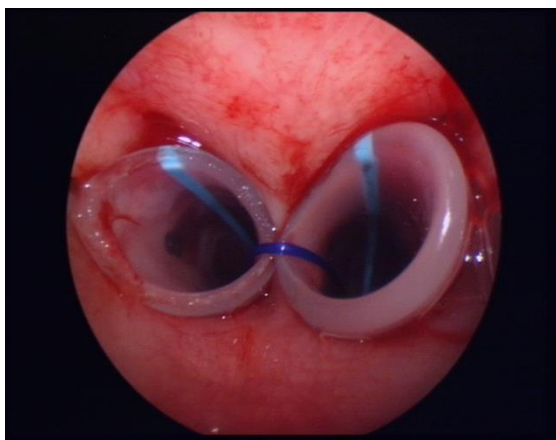


Figure 20: Retropalatal view of bilateral stents in position in the posterior choanae

Mitomycin C

This is another contentious area with some evidence both for and against its use. It is believed to act topically to reduce mucosal restenosis by decreasing fibroblast mediated scarring.

Postoperative considerations

- After successful choanal atresia surgery the child should be extubated and can usually be managed on a general paediatric ward with regular nasal suctioning
- Oral feeding is established as soon as possible
- Postoperative nasal decongestant drops (0.5% xylometazalone) should be used for one week
- Saline and steroid drops should be used for 2 weeks to reduce postoperative inflammation and granulations

Alternative techniques

Transnasal drill-out with operating microscope

If the equipment required to perform a retropalatal endoscopic approach is not available, a standard operating microscope and a small bur can be used transnasally to open the stenosis. A standard ear speculum can be inserted in the nose after adequate decongestion and will allow for a view of the choanae with the use of a microscope.

Non-powered dilation techniques

If the above equipment is not available, transnasal opening and dilatation of the choanal atresia remains a viable and useful surgical option.

The safest way to achieve this is to use curved urethral sounds to perform an initial

puncture of the membranous atresia followed by serial dilatation (*Figures 13, 17*).

- The surgical setup is identical, with the patient positioned supine with a shoulder roll in position, under general anaesthetic with a down-facing Rae endotracheal tube
- Insert a cleft palate/tonsillectomy gag
- Retract the palate using the methods previously described
- A dental mirror and headlight can be used to obtain a view of the posterior choana in order to directly visualise the stenosis
- Prepare the nose with a decongestant
- Insert the smallest urethral sound with the curve pointing towards the floor of the nose and hug the septum to try and puncture the inferomedial corner of the choanal atresia (*Figure 17*). This avoids potential injury of the skull base during this “blind” procedure
- A finger can be placed on the soft palate or in the postnasal space to monitor when the atresia is punctured
- The opening can be further dilated by using serial urethral sounds until an adequate-sized stent (3.5 ET tube) can be passed
- Back-biting forceps introduced down the nose can be used to remove the posterior septum if necessary (*Figure 21*)



Figures 21a, b: Back-biting forceps used to remove bone from posterior septum

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