orphananesthesia

Anaesthesia recommendations for

Choanal atresia

Disease name: Choanal atresia

ICD 10: Q30.0

Synonyms: Atresia of posterior naris; Congenital atresia of choana; Congenital atresia of posterior nares; Atresia of posterior nares

Disease summary:

Choanal atresia is the narrowing and closure of the posterior choanae of the nose. According to Otto [1] it was first described in 1755 by Roederer. The reported incidence is approximately 1 in 7000 live births [2]. Females are more likely to be affected [3] with a ratio of around 2:1 [4].

The obstruction can be bony, membranous, or mixed obstruction. Originally the obstruction was described as bony in 90% of cases and membranous in 10% [5]. More recently a retrospective study with computer tomography (CT) and histologic specimens showed 29% pure bone atresia, 71% mixed membranous/ bony atresia and no pure membranous atresia [6].

Unilateral choanal atresia is more common than bilateral with an estimated high number of undetected cases. Infants with unilateral choanal atresia rarely present with neonatal respiratory distress. They are typically presenting around 5-24 months of age [3, 7] with unilateral obstruction and persistent mucoid rhinorrhoea. Rarely the diagnosis is delayed till adulthood [8].

Neonates are obligate nasal breathers with more than 50% desaturating when nasally obstructed [9]. Bilateral choanal atresia can present at birth with airway obstruction and paradoxical cyanosis which resolves on crying due to oral breathing. The infant may also present with difficulty in feeding and choking on feeds due to the inability to breathe and feed simultaneously. In infants intubated at birth for a secondary airway condition multiple failed extubation attempts may also indicate the diagnosis.

Congenital abnormalities are frequently associated with choanal atresia, particularly in bilateral cases, which are often linked to specific syndromes or multiple congenital anomalies. While unilateral cases are more likely to be isolated, bilateral choanal atresia tends to be associated with a greater number of complications. In one study, 34% of patients with bilateral choanal atresia were found to have additional airway abnormalities, such as tracheomalacia, laryngomalacia, and subglottic stenosis [10]. The condition most commonly associated with bilateral choanal atresia is CHARGE syndrome, characterized by a distinctive set of anomalies that may include coloboma of the eye, heart defects, atresia of the choanae, growth and developmental delays, genital and urinary abnormalities, and ear malformations. Other craniofacial syndromes linked to choanal atresia include Treacher Collins syndrome, Crouzon syndrome, Pfeiffer syndrome, and Apert syndrome, while chromosomal abnormalities such as Trisomy 21 can also be noted.

Beyond craniofacial syndromes, choanal atresia can be associated with dysfunctions in various other organ systems. This emphasises the importance of comprehensive preoperative evaluation, as these less common associations can significantly impact perioperative management.

Metabolic disorders may include disturbances of the pituitary axis in conditions like Pallister-Hall syndrome, or impaired steroid synthesis, seen in Antley-Bixler syndrome, leading to adrenal insufficiency. Additionally, neonatal-onset diabetes mellitus and congenital hypothyroidism may also be present as an associated condition.

Renal malformations are also an important consideration in those with choanal atresia. These patients may present with renal anomalies, such as renal tubular dysgenesis, resulting in significant kidney dysfunction. Additionally, syndromes associated with other kidney malformations, like Beaulieu-Boycott-Innes syndrome and Van Maldergem syndrome, are also seen with choanal atresia.

Medicine is in progress

Perhaps new knowledge

Every patient is unique

Perhaps the diagnosis is wrong

Find more information on the disease, its centres of reference and patient organisations on Orphanet: www.orpha.net

Emergency information

A	**AIRWAY / ANAESTHETIC TECHNIQUE**	High risk for a difficult airway especially in syndromic patients with anatomic abnormalities. Commonly associated with tracheomalacia, laryngomalacia, and subglottic stenosis. Surgery relating to the choanal atresia will require general anaesthesia otherwise all types of anaesthesia can be considered
B	**BLOOD PRODUCTS (COAGULATION)**	Standard management for transfusion and administration of blood products applies. No coagulation disorders associated directly with choanal atresia.
C	**CIRCULATION**	Congenital cardiac abnormalities are often seen as part of the syndromes associated to choanal atresia.
D	**DRUGS**	There are no contraindications to common anaesthetic agents.
E	**EQUIPMENT**	Difficult airway equipment should be available.

Typical surgery

The only treatment for choanal atresia is surgical repair to create patency of the choanae. A variety of surgical approaches have been described: transnasal puncture, transpalatal resection, transnasal endoscopic resection, nasal stenting and tracheostomy [2]. Nowadays, the transnasal endoscopic resection of the choanal atresia is the standard approach as safe and effective technique [11, 12, 13, 14, 15]. The mucosa-sparing surgery and the resection of posterior vomer parts are important factors for surgical success to prevent recurrences [13, 16].

Bilateral choanal atresia is detected in the neonatal period and requires emergent airway management and timely surgical intervention. The definitive treatment of unilateral choanal atresia, where possible, is delayed until a later age (> 7 months) to increase the anaesthetic and surgical safety.

General anaesthesia or sedation may be required to allow imaging studies, particularly CT scans, to be performed.

Type of anaesthesia

General anaesthetic technique is required for surgery.

Necessary additional pre-operative testing (beside standard care)

Detailed history and clinical evaluation are required to exclude associated syndromes and congenital abnormalities. Comprehensive laboratory testing, including genetic analysis, can provide insights into potential underlying syndromes.

Cardiac echocardiography and electrocardiogram may be required if there is concern of congenital cardiac abnormalities.

Imaging studies, such as CT scans of the paranasal sinuses and skull base, and, in rare cases, MRI, can also be beneficial.

The definitive diagnosis of choanal atresia is made with endoscopic visualisation of the atresia plate

Particular preparation for airway management

Neonates with bilateral choanal atresia can present as an airway emergency at birth. Initial management is to provide an adequate airway through the oral route. This can be accomplished using a McGovern nipple, an intraoral nipple with the tip cut off secured around the infant's occiput, or an oropharyngeal airway [17].

Patients with choanal atresia are at high risk for a difficult airway [18], especially syndromic patients with anatomic abnormalities. Adequately sized face masks, oral airways, supraglottic airways and endotracheal tubes should be available. Advanced airway management techniques including fibreoptic bronchoscopy and video laryngoscopy must also be available [18].

There have been case reports of oropharyngeal continuous positive airway pressure (CPAP) being used to prevent the need for and duration of intubation [19]. As this technique requires some time for preparation, it should be considered early in the management plan.

Tracheostomy may be required if definitive surgical management needs to be delayed.

Particular preparation for transfusion or administration of blood products

Standard management.

Particular preparation for anticoagulation

Standard management.

Particular precautions for positioning, transportation and mobilisation

Infants with bilateral choanal atresia are at increased risk of an adverse airway event during transportation. Inter-hospital transfer, such as to a tertiary hospital, should ideally be undertaken after endotracheal intubation if there are concerns with a difficult airway. Patients can be transported with an oropharyngeal airway.

Interactions of chronic disease and anaesthesia medications

Not reported.

Anaesthetic procedure

Infants with bilateral choanal atresia tend to be in the neonatal period so gaseous induction and maintenance tends to be the preferred method. A slow intravenous induction would also be reasonable. There should be a low threshold to use an oral pharyngeal airway to maintain a patent oral airway prior to endotracheal intubation.

Use of a muscle paralysing agent is based on individual clinical judgement and the presence of known or suspected co-morbidity. There is recommendation that in a predicted difficult airway maintaining spontaneous ventilation with a deep intubation may be beneficial [20].

Unilateral choanal atresia, with its older age at operation, can be managed with a gaseous or a total intravenous technique [21].

A difficult airway should be anticipated.

Particular or additional monitoring

Standard monitoring.

Possible complications

Laryngospasm, bronchospasm, desaturation and reintubation are seen as expected complications relating to the difficult intubation [18].

Post-operative care

Post operative extubation should only occur when respiration has completely begun. This may require a period of post operative mechanical ventilator support [21]. Oropharyngeal CPAP can be considered for additional respiratory support post extubation [18].

While post-operative nasal stenting may reduce the rate of restenosis its application should be considered on an individual basis, as it is not standard practice. Stents can lead to complications such as granulation tissue formation and bleeding. In one reported case series, stents were utilised in all patients after bilateral choanal atresia repair but in less than half of unilateral repairs [7]. Therefore, the use, duration, type, and method of securing stents remain subjects of controversy [22].

Topical mucosal care is crucial during the postoperative period. It is recommended to perform daily nasal rinses using sodium chloride 0.9% and to apply corticosteroid nasal sprays for a duration of 6 weeks. Ensuring good parental education is essential to enhance compliance with postoperative care measures.

Disease-related acute problems and effect on anaesthesia and recovery

As mentioned earlier; in bilateral choanal atresia, neonatal asphyxia can present at or shortly after birth. Maintaining a patent oral airway or oral endotracheal intubation is required.

Ambulatory anaesthesia

Due to the significantly higher risk for postanaesthetic adverse airway events ambulatory anaesthesia is not recommended.

Obstetrical anaesthesia

Not reported.

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