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Evaluation and management of upper airway disorders in children

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Upper airway disorders in children may be divided into those that are congenital in origin and those that are acquired. The presentation and management of these disorders is significantly influenced both by the anatomic location of the pathology, which is usually obstructive in nature, and by the severity of the obstruction. This discussion provides an overview of the presentation, diagnosis, management, and potential complications of the most commonly seen upper airway disorders. These disorders are presented within an anatomic framework, progressing from proximal at the nares to distal at the carina.

Upper airway disorders in children may be either congenital or acquired. These disorders occur at various anatomic levels, and their presentation and management are significantly influenced by both the level at which obstruction occurs and the severity of obstruction. It is useful to consider disorders affecting the upper airway within an anatomic framework, progressing from proximal at the nares to distal at the carina. Using this framework, the clinical presentation, diagnosis, management, and potential complications of the most frequently observed upper airway disorders in children will be discussed.

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Congenital nasal pyriform aperture stenosis

Congenital nasal pyriform aperture stenosis (CNPAS) is an uncommon form of nasal airway obstruction that typically presents during the first few months of life when infants are obligate nasal breathers. Bony overgrowth of the medial aspect of the nasal process of the maxilla is the characteristic feature of this disorder. Because the pyriform aperture is the narrowest section of the bony nasal skeleton, even minor anatomic abnormalities in the cross-sectional area of the aperture significantly affect airflow by increasing nasal airway resistance. In most cases, CNPAS is associated with a single central upper incisor, though occasionally it may occur as an isolated anomaly. In rare cases, an association with holoprosencephaly has been described.[1]

Presentation is in the neonatal period and the spectrum of symptoms all result from nasal obstruction. These symptoms are similar to those seen in patients with bilateral choanal atresia and include cyanosis, apnea, feeding difficulties, and labored breathing. Because of this similarity,

evaluation for suspected choanal atresia is sometimes performed, yielding negative results. Clinicians may thus be falsely reassured that there is no significant nasal obstruction.

Diagnosis is made on anterior rhinoscopy, which reveals an anterior bony obstruction of the nasal vestibule. It is confirmed by computed tomography (CT), which may also confirm the presence of a single central upper incisor.

Patients are managed on the basis of the severity of their symptoms. Those with mild symptoms can be managed expectantly. This may be all that is required until growth results in increased nasal airway size. In more severe cases, surgical enlargement is indicated. This is best performed via a sublabial approach, exposing the pyriform aperture and using a diamond burr to remove the excessive bone of the nasal process of the maxillary crest. Nasal stents are generally placed for 2 to 4 weeks.

Choanal atresia

Although several theories of the embryogenesis of choanal atresia have been proposed, this anomaly is thought to be a consequence of the persistence of the nasal buccal membrane. The obstruction may be membranous, bony, or a combination of both, with the latter being the most commonly seen. The atresia may be unilateral or bilateral and though the relative incidence of these two presentations is a subject of controversy, the ratio of unilateral to bilateral cases is likely 1:1. Whether choanal atresia is unilateral or bilateral, it may be associated with a number of congenital anomalies. The best recognized association is with CHARGE syndrome (*c*oloboma, *h*eart defects, *a*tresia, *r*etardation of growth and development, *g*enitourinary disorders and *e*ar abnormalities).[2]

Neonates are obligate nasal breathers during the first 6 weeks of life. As such, whether neonatal nasal obstruction is due to CNPAS, tumor, or choanal atresia, neonates present with apnea during quiet respiration. This is not observed when neonates are upset as they mouth breathe when crying. Once the infant settles down, however, apnea again becomes a risk. Complete obstruction of the posterior nasal passage by choanal atresia does not allow the normal drainage of nasal secretions into the nasopharynx. These secretions must thus passively drain anteriorly and are characteristically copious and tenacious.

Although historically the diagnosis of choanal atresia was suspected when a 6-Fr suction catheter could not be passed through the nose into the nasopharynx, we currently recommend evaluating nasal passages with a thin (1.9 mm) flexible nasal endoscope. The initial management of a child with bilateral choanal atresia is best accomplished by placement of an oral airway or intubation. This will stabilize the child until CT scanning can be performed and, if appropriate, until genetic evaluations can be undertaken. Radiological evaluation with bone/window high-resolution, close-cut CT scanning is recommended. Removing all nasal sections with a soft suction catheter immediately before the scan greatly enhances the quality of the scan. Characteristic findings are of a thin atretic plate at the posterior choana that is commonly both bony and membranous. Frequently there is associated prominence of the bony margins of the choana with either bony overgrowth of the vomer in the midline or medialization of the lateral nasal walls.

The recommended treatment is early surgical repair,[3] unless there is a contraindication such as extreme prematurity or multiple congenital anomalies. In such cases, a temporizing tracheostomy may be the most appropriate treatment. Surgical approaches may be either transpalatal or transnasal. The latter approach is preferable in most children and involves endoscopically guided removal of the atretic plate using urethral dilators, backbiting forceps, drills, powered microdebrider cutters, or a combination of these instruments. In cases in which there is prominence of the bony margins of the choana, the posterior aspect of the vomer may be removed to form a common cavity at the level of the atretic plate. Traditionally, transnasal stents have been placed for several weeks postoperatively until the repair has stabilized. The current trend, however, is for shorter periods of stenting or no stenting at all. Both of these stenting options carry the risk of the development of secondary choanal stenosis. As such, adjuvant therapy, such as the topical application of mitomycin C, is used in some children.

Patients with unilateral disease typically go undiagnosed until later in childhood, presenting with unilateral rhinorrhea and nasal obstruction. CT evaluation is indicated and transnasal repair of the unilateral atresia is usually performed after 2 years of age. Children with choanal atresia associated with CHARGE syndrome are unlike other patients with choanal atresia in two respects. Firstly, the degree of their nasal obstruction at birth is more severe, and as the level of obstruction is not restricted to the choana, repair of the choanal atresia may not prevent the need for tracheostomy placement. Other frequent levels of obstruction include pharyngomalacia, hypopharyngeal collapse, and laryngomalacia. If these levels of obstruction are recognized initially, placement of a tracheostomy tube and late repair of the choanal atresia may be the most appropriate management strategy. Secondly, the results of surgical repair, whether unilateral or bilateral, early or late, are not as successful as in other patients. This may be partially attributed to abnormalities in the skull base seen in patients with CHARGE syndrome. In such patients, the resection of the posterior vomer with a prolonged stenting period is recommended.

Retrognathia/glossoptosis

Retrognathia is associated with a variety of abnormalities, including Pierre Robin sequence (short mandible cleft pal-