Nasal Obstruction in Neonates and Children

Contents

Nasal obstruction in the neonate may lead to serious consequences including respiratory distress or failure to thrive. While bilateral nasal obstruction often presents in the neonatal period, unilateral nasal obstruction may not present until much later in life, with chronic nasal drainage, skin irritation, and congestion (Gnagi and Schraff 2013).

Synonyms and Related Disorders

Choanal atresia; Nasal dermoid; Nasal encephalocele; Nasal Glioma; Nasal obstruction; Nasolacrimal duct cyst; Pyriform aperture stenosis

Genetics/Basic Defects

1. Congenital malformations secondary to aberrant embryogenesis of both the internal and external nose potentially causing nasal obstruction include the following but not limited to (Gnagi and Schraff 2013):
    1. Midfacial hypoplasia
    2. Craniosynostosis
    3. Arhinia (complete absence of the nose)
    4. Nasal hypoplasia (congenitally absent nasal bones)
    5. Complete or partial nasal duplication
    6. Single centrally placed nostril
    7. Supernumerary teeth in the nose
    8. Thornwaldt cyst (a common incidental benign midline nasopharyngeal mucosal cyst)
    9. Nasopharyngeal stenosis (incomplete separation of the soft palate and posterior pharyngeal wall)
    10. Others
2. The most common and clinically significant congenital anomalies: please see the differential diagnosis section.

Clinical Features

1. Signs and symptoms of nasal obstruction (Gnagi and Schraff 2013):
    1. Stuffy nose
    2. Rhinorrhea
    3. Mucus
    4. Stertor
    5. Snoring/snorting
    6. External deformity
    7. Nasal flaring

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8. Chest retractions
9. Cyanosis (+/− cyclical nature)
10. Feeding difficulties
11. Hyponasal cry
12. Failure to thrive
13. Dyspnea/apnea
14. Aerophagia with abdominal distention
15. Difficulty sleeping
16. Epiphora

2. Differential diagnosis (Adil et al. 2012; Gnagi and Schraff 2013):
    1. The most common and clinically significant congenital anomalies:
        1. Choanal atresia:
            1. The most common congenital nasal anomaly
            2. Most commonly associated with CHARGE syndrome (please see the chapter)
            3. Can also be seen with Apert, Crouzon, and Treacher Collins syndromes (please see the chapters)
        2. Congenital nasal pyriform aperture stenosis:
            1. Pyriform aperture: the pear-shaped, bony opening of the nasal cavity
            2. Pyriform aperture stenosis: caused by a bony overgrowth of the medial nasal processes of the maxilla, usually bilateral causing nasal obstruction in neonates
            3. May occur as an isolated anomaly or in association with the absence of the anterior pituitary, diabetes insipidus, submucous cleft palate, and hypoplastic maxillary sinuses or as part of the holoprosencephaly sequence
        3. Midnasal stenosis;
            1. A rare clinical entity secondary to bilateral bony overgrowth midway through the nasal cavity
            2. Usually occurs in children with midfacial hypoplasia
        4. Nasolacrimal duct cysts (dacryocystoceles) (Brugger et al. 2010):
            1. Fluid accumulation and distension of the nasolacrimal duct.
            2. Unilaterally in 50.6% or bilaterally in 49.4%.
            3. Can resolve spontaneously during intrauterine life.
            4. Bilateral involvement with substantial intranasal extension may develop respiratory distress syndrome (Bachelard-Serra et al. 2013).
            5. Usually seen as an isolated abnormality but can be associated with other congenital anomalies (Yazici et al. 2010).
        5. Midline nasal masses:
            1. Nasal/nasopharyngeal dermoid: the most common of the congenital midline nasal mass
            2. Glioma (neuroglial heterotopia)
            3. Encephalocele/meningocele: caused by defective development of the skull and herniation of intracranial contents/meninges
            4. Thornwaldt cyst
    2. Neoplasms:
        1. Teratoma
            1. Histologically containing all three germ cell layers.
            2. Tumors: composed of immature cells carrying a higher risk of malignancy with a worse prognosis.
            3. Alpha-fetoprotein and beta-hCG tumor markers may be elevated.
        2. Hamartoma
        3. Vascular lesions (hemangiomas, arteriovenous malformations, vascular malformations)
        4. Lymphangioma
        5. Lipoma
        6. Neurofibroma
        7. Rhabdomyosarcoma
        8. Lymphoma
    3. Infectious:
        1. Upper respiratory infection
        2. Respiratory syncytial virus
        3. Sexually transmitted diseases:
            1. Chlamydia
            2. Gonorrhea
            3. Syphilis

4. Foreign body:
5. Traumatic/iatrogenic:
    1. Septal dislocation
    2. Septal hematoma
    3. Nasal tip depression
    4. Rhinitis medicamentosa
    5. Instrumentations: suction trauma, nasogastric tube, CPAP, and nasal prongs
2. Inflammatory:
    1. Allergic rhinitis (cow's milk, soy)
    2. Gastroesophageal reflux
    3. Recurrent emesis
    4. Idiopathic
3. Metabolic: hypothyroidism
4. Maternal:
    1. Estrogenic stimuli
    2. Drug ingestion (methimazole, methyldopa, opiates, tricyclic antidepressants, propranolol)
5. Associated syndromes:
    1. Cystic fibrosis
    2. Kartagener syndrome
    3. Charge association
    4. Apert syndrome
    5. Crouzon syndrome
    6. Treacher Collins syndrome
    7. Fetal alcohol syndrome
    8. Down syndrome

Diagnostic Investigations

1. Clinical procedures (Gnagi and Schraff 2013):
    1. Anterior rhinoscopy with an otoscope: help visualize anterior stenosis, masses, or obstructive mucus.
    2. Gently passing a small (5 or 6 French) catheter through the nose into the nasopharynx to confirm an open communication:
        1. Obstruction at the anterior inlet may suggest pyriform aperture stenosis, while obstruction posteriorly (approximately 32 mm) may suggest choanal atresia (Myer and Cotton 1983).
        2. Visualizing or palpating the tube through the mouth confirms that the tube is not coiled in the nose to prevent misdiagnosis. In infants with craniofacial abnormalities or visible nasal masses, care must be taken when attempting to pass a nasal catheter, as these may be associated with skull base defects and risk intracranial passage of the catheter (Ramsden et al. 2009).
    3. Additional studies to assess nasal patency include the use of a tympanometer placed at the nares to confirm or exclude a closed cavity (Effat 2005).
    4. Nasal endoscopy:
        1. A simple and minimally invasive diagnostic procedure.
        2. When a nasal mass is identified on endoscopy, it should be assumed to have intracranial extent until proven otherwise. Therefore, no biopsy of a mass should be performed until appropriate imaging has been undertaken (Jaffe 1981).
2. Radiologic imagings (Adil et al. 2012; Gnagi and Schraff 2013):
    1. Plain radiographs with radiopaque contrast in the nasal cavity: rarely employed because of poor sensitivity and specificity
    2. CT scans: best allow bony definition and typically the test of choice to assess choanal atresia and pyriform aperture stenosis
    3. MRI: better choice to evaluate nasal masses to delineate intracranial involvement and extent

Genetic Counseling

1. Recurrence risk: Recurrence risk depends on the etiology of the nasal obstruction:
    1. Patient's sib:
        1. Autosomal recessive: 25%
        2. Autosomal dominant: not increased unless a parent is affected or having gonadal mosaicism
        3. Sporadic: unknown, probably not increased
    2. Patient's offspring:
        1. Autosomal recessive: not increased unless the spouse is also a carrier

2. Autosomal dominant: 50%
3. Sporadic: unknown, probably not increased

2. Prenatal diagnosis:
   1. Ultrasonography:
      1. Nasal gliomas detected at mid-trimester (De Basio et al. 2006; Grzegorczyk et al. 2010; Tonni et al. 2011; Beegun et al. 2012).
      2. A huge fetal facial mass protruded through the left nostril at 33 weeks of gestation: computed tomography of the neonate suggested a transethmoidal encephalocele. However, MRI showed a huge mass occupying the nasopharynx and the nasal cavity and protruding externally to the face without any intracranial connection. Pathologic examination revealed intranasal glioma (Okumura et al. 2012).
      3. Dacryocystocele: anechogenic cystic masses located anteromedially in the orbits and centered on the medial canthus (Bianchini et al. 2004).
   2. Fetal MRI has been used to confirm the diagnosis of dacryocystoceles with the following triad (Bianchini et al. 2004):
      1. Paraocular cystic mass in the medial canthus region
      2. Nasolacrimal duct enlargement
      3. Intranasal cyst
3. Management (Adil et al. 2012; Gnagi and Schraff 2013): secure an adequate airway (the first goal of therapy):
   1. Choanal atresia:
      1. Delay repair of unilateral atresia until school age to allow the nasal cavity to mature.
      2. Transnasal endoscopic repair: most popular technique today.
      3. Transpalatal approach may also be used.
   2. Congenital nasal pyriform aperture stenosis:
      1. Bilateral balloon dilation
      2. Short-term stenting of the nasal pyriform apertures
      3. May require surgical intervention to drill and widen the pyriform aperture
   3. Nasolacrimal duct cysts:
      1. May resolve spontaneously within the first year of life (Peterson and Robb 1978)
      2. Conservative management: warm compresses and gentle massage for simple dacryocystoceles without intranasal extension
      3. Probing the duct and/or surgical marsupialization of the intranasal cyst (Leonard et al.. 2008)
   4. Congenital frontonasal masses (nasal dermoid, encephalocele, glioma, other neoplasms): surgical intervention with multiple surgical subspecialties
   5. Infectious etiologies: treat the underlying cause
   6. Iatrogenic etiologies: treat the underlying cause
   7. Inflammatory, systemic, and other etiologies: treat the underlying cause
   8. Syndrome associations: treat according to the underlying syndrome

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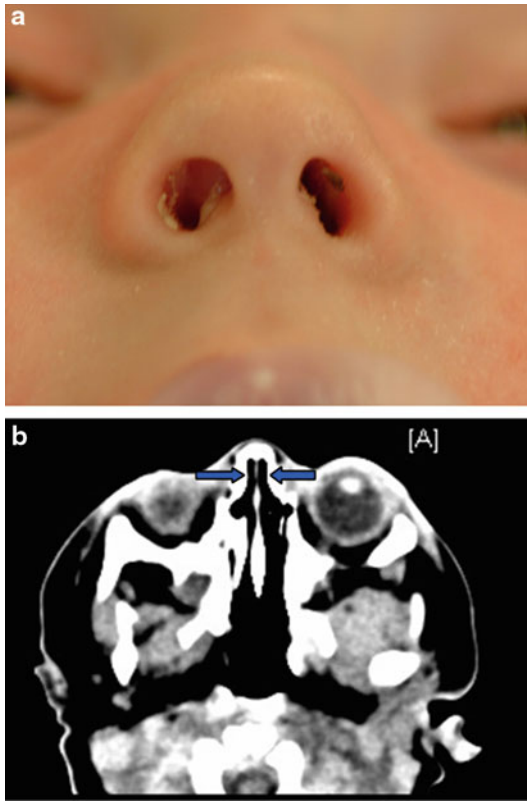

Fig. 1 (**a**, **b**) A 3-week-old Caucasian boy was seen for respiratory distress and inability to feed. Nasal catheters could not be passed on either nasal side and he was referred for evaluation of possible choanal atresia (**a**). He struggled with breathing and desaturated every time he ate. Nasal obstruction was persistent. A flexible pediatric (2.4 mm diameter) endoscope was unable to insert beyond the nasal vestibule on either side. Rigid pediatric endoscope (2 mm Hopkins rod) could not be passed through either nasal passage. A non-contrast axial CT scan (**b**) showed CNPAS with a bilateral combined nasal vestibular aperture width of 5.03 mm (*arrows*) and normal choanae. He underwent bilateral balloon dilation and short-term stenting of the nasal pyriform apertures without the need for additional procedures and remained patent after 12 months at follow-up (Gungor and Reiersen 2014) (Courtesy of Dr. Anil Gungor)

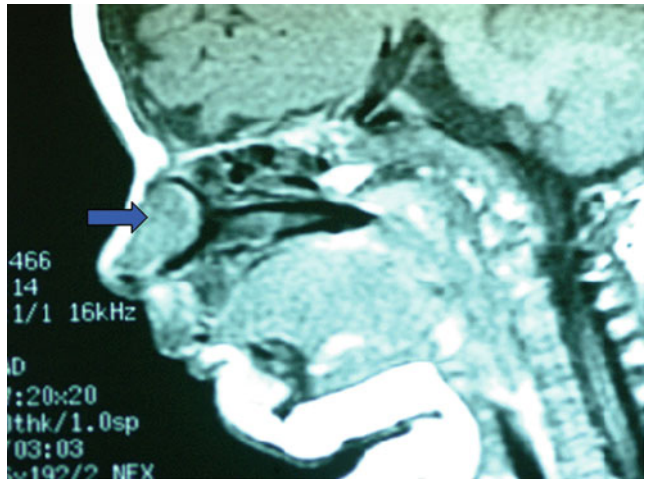

Fig. 2 A 3-month-old boy was seen for a nasal mass present since birth. The mass grew slowly bigger in size, causing nasal obstruction and difficulty in his nasal breathing, accompanied by occasional snoring and intermittent episodic epistaxis. Anterior rhinoscopy showed a large mass originating from the lower lateral cartilage and completely obstructing the right nostril. It appeared to extend to the superior part of the vestibule, but the extent of the entire lesion could not be determined by anterior rhinoscopy alone. Palpation of the mass showed a smooth, rubbery lesion that did not blanche with direct pressure. The lesion was nonpulsatile and did not change in size when the patient cried. A computed tomographic scan and a magnetic resonance imaging scan showed a $0.7 \times 1.8 \times 1.1$ cm soft tissue mass in the anterior right nasal cavity (*arrow*), without extension into the bony nasal framework, nasopharynx, paranasal sinuses, or skull base. No bony erosion was observed. A transnasal excision of the mass was performed with endoscopic guidance. The mass was excised from vestibular skin without difficulty, and a small mucosal attachment from the inferior turbinate was freed with electrocautery. Grossly, the lesion, which was dense and glistening, was a small, tan, white, and focally red mass. Histologically, the mass was a heterotopic neuroglial tissue (nasal glioma) (Roy and Gungor 2014) (Courtesy of Dr. Anil Gungor)