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Congenital nasal pyriform aperture stenosis

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Congenital nasal pyriform aperture stenosis is a rare form of nasal obstruction and must be distinguished from other causes of nasal obstruction. It should be considered in the differential diagnosis of any neonate or infant with signs and symptoms of upper airway compromise. Diagnosis is made on the basis of clinical examination and computerized tomography findings, while treatment often involves surgical enlargement of the bony nasal inlet.

Proper diagnosis and management of respiratory distress in the neonate is extremely important. Because neonates are obligate nasal breathers, neonatal nasal obstruction may have serious consequences. The pyriform aperture is the narrowest, most anterior bony portion of the nasal airway, and a decrease in its cross-sectional area will significantly increase nasal airway resistance. Congenital nasal pyriform aperture stenosis (CNPAS) is a rare, unusual form of nasal obstruction and must be distinguished from other causes of nasal obstruction such as choanal atresia or stenosis. CNPAS presents with symptoms of nasal airway obstruction, which are often characterized by episodic apnea and cyclical cyanosis. It should be considered in the differential diagnosis of any neonate or infant with signs and symptoms of upper airway compromise. The diagnosis is made on the basis of clinical examination and maxillofacial computerized tomography (CT) findings. Treatment of the obstruction often involves surgical enlargement of the bony nasal inlet.

Congenital nasal pyriform aperture stenosis

Neonatal nasal obstruction is a potentially life-threatening condition and an important cause of respiratory distress because neonates are obligatory nasal breathers. Severe bilateral nasal obstruction can lead to apnea, cyanosis, and respiratory distress. CNPAS is a rare form of nasal airway obstruction in neonates that can present as a life-threatening emergency. CNPAS may be isolated or associated with other midline anomalies. Immediate recognition and diagnosis of this condition are critical for restoring the patency of the nasal airway and preventing asphyxia.

The differential diagnosis of neonatal nasal obstruction includes but is not limited to choanal atresia or stenosis, nasal trauma (septal hematoma or subluxation of the septum), cysts (dermoids or epidermoids), skull base defects (meningoencephalocele and encenphalocele), tumors (rhabdomyosarcoma, hemangioma, glioma, lymphangioma, and teratoma), and nasal hypoplasia. CNPAS is most often bilateral and is the result of bony overgrowth of the nasal process of the maxilla into the nasal cavity, which leads to a narrowing of the anterior nasal airway, whereas choanal atresia is the result of uni- or bilateral fibrous or bony obstruction of the posterior nasal cavity. CNPAS has a similar clinical picture to that of choanal atresia or stenosis, and can manifest with respiratory distress, apneic episodes, cyclical cyanosis, inability to nurse, or sudden total airway obstruction, which is relieved with crying.[1] Signs can occur immediately after birth or in a few months of life, depending on the amount of obstruction. Diagnosis is further established by physical examination, showing narrowed anterior nasal fossae, inability to pass a 5F catheter, or 1.9-mm endoscope through the nasal cavity. CT will reveal stenosis at the bony nasal inlet (Figure 1). The presence of a nar-

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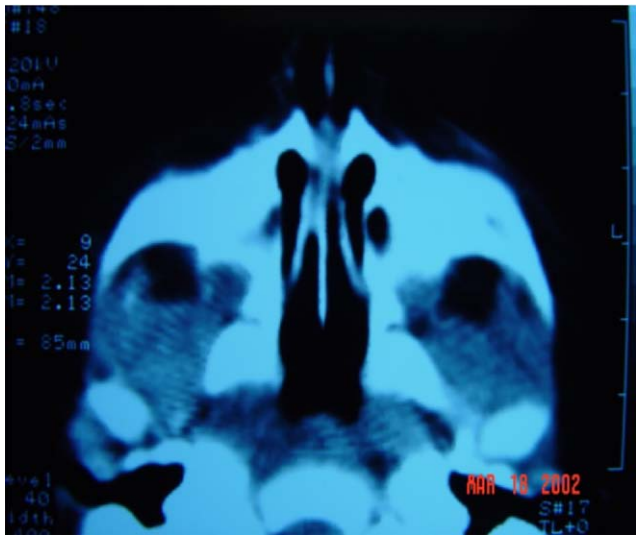

Figure 1 Axial maxillofacial computerized tomography scan showing stenosis of the bony nasal inlet. (Color version of figure is available online.)

rowed anterior nasal inlet and bony overgrowth of maxillary nasal processes confirms the diagnosis of CNPAS. A radiographically measured pyriform aperture width of less than 11 mm in a term infant is considered diagnostic.[2]

CNPAS was first described in 1989 by Brown et al[3] and represents obstruction at the most anterior and narrow aspect of the bony nasal airway, the pyriform. The pyriform aperture is formed laterally by the nasal or frontal processes of the maxilla, inferiorly by the junction of the horizontal processes of the maxilla and anterior nasal spine, and superiorly by the nasal bones.[3] Embryologically, the development of the nasal structures occurs between 5 and 8 weeks of gestation, when the palate develops from 2 primordia called the primary palate and the secondary palate.[4,5] The primary palate is formed from the merging of the medial nasal prominences of the maxilla and later becomes the premaxillary portion of the maxilla.[1,4] At approximately the same time, the lateral nasal prominence fuses with the maxillary prominence to form the lateral nasal wall and the pyriform aperture. The secondary palate develops from 2 horizontal projections of the maxillary eminences, the lateral palatine processes.[4] During the eighth week of development, maxillary ossification begins from the growth center above the canine teeth in the primary palate and extends to the remainder of the maxilla at approximately 4 months of development.[1,4,5] It has been suggested that over-ossification of the nasal process of the maxilla may be responsible for the bony stenosis at the pyriform aperture. Stenosis may also result from hypoplastic growth of the primary palate during fetal life.

The exact pathology of CNPAS is unknown but may represent a genetically heterogenous condition with a variable pattern of inheritance. CNPAS may occur in isolation or as part of several other abnormalities. A relationship between CNPAS and moderate holoprosencephaly and endocrine abnormalities has been suggested.[4,6-8] Craniofacial anomalies occur in up to 40% of cases.[9] The most common associated disorders were solitary median maxillary central incisor and hypopituitary (HP) axis abnormalities, which have been reported in 55% and 40% of cases, respectively. Patients with an abnormal HP axis were noted to be at a higher risk of endocrine dysfunction. Ectopic neurohypophysis was the most frequent HP axis abnormality.[10] Other associated anomalies including shallow sella turcica, craniopharyngeal canal, submucous cleft palate, and hypoplastic maxillary sinuses have also been described. The finding of solitary median maxillary central incisor is thought to be a manifestation of the holoprosencephaly sequence.[1,5,7,11]

The treatment of CNPAS depends on the severity of obstruction and symptoms and the overall prognosis of the infant (Table 1).[9,11] As with any issue involving respiratory compromise, establishing a secure airway is the priority. Placement of a McGovern nipple or oral airway with appropriate monitoring represents the initial intervention.[1] Milder cases of obstruction may be addressed with nasal humidification, suctioning, topical steroid, and decongestant drops. Stenting is not widely advocated because of the risks of mucosal and cartilaginous pressure necrosis.[6,11] Gavage feedings may be needed to optimize nutritional management. Conservative treatment, using the aforementioned measures, has also been recommended in those patients with multiple anomalies and poor prognosis.[11] In patients with better prognoses, surgical treatment is indicated for patients in whom 2 weeks of aggressive medical treatment has failed or those who have severe degrees of nasal obstruction. Timing of surgery should be individualized, and multiple investigators have suggested the application of the rule "10."[1,3] If the patient's respiratory status remains stable, surgical repair may be deferred until the patient reaches 10 lbs, 10 weeks of age, and 10 g of hemoglobin. This application facilitates surgical repair on a more stable and larger patient.[11] However, surgical repair may be performed earlier if clinically indicated.

Table 1 Management of CNPAS in neonates

Indications	Contraindications	Special instruments	Tips and pearls
Obstruction refractory to medical therapy Severe degree of obstruction Failure to thrive	Poor prognosis of patient Unsuitable operative candidate	Otologic instruments Small diamond burr drill Microscope with 300-mm F lens	Avoid dissection along nasal floor Keep dissection plane anterior to inferior turbinate Stents in place for at least 5 days, <15 days

Preoperative evaluation

Preoperative evaluation for subjects with CNPAS involves standard laboratory studies. Maxillofacial CT with 1.5- to 2-mm-thin axial sections in a plane parallel to the hard palate should be used to measure the dimensions of the pyriform aperture and confirm the diagnosis. CT can also evaluate for other anomalies, such as choanal atresia/stenosis, and defines the surrounding bony architecture. If holoprosencephaly is suspected by the presence of a central maxillary incisor or other anomalies, CT or ultrasound of the brain should be performed. The long-term morbidity associated with HP axis abnormalities necessitates that patients be assessed with cerebral MRI prior to surgery. If abnormalities of the HP axis are seen, appropriate biochemical endocrine tests and a referral to an endocrinologist are warranted to prevent life-threatening complications or developmental delays secondary to endocrine dysfunction.

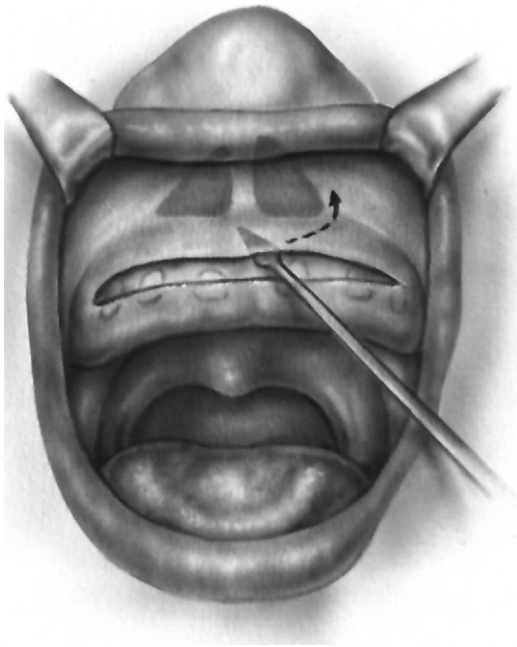

Figure 3 Elevation of the soft tissue to expose pyriform aperture.

Surgical procedure

There are 2 primary approaches for the surgical repair of pyriform aperture stenosis, transnasal, and sublabial. The transnasal approach is described in adults, and is technically difficult in the infant nasal cavity because of poor exposure and an increased risk of injury to mucosa.[1,3,4,12] The more widely accepted and practiced sublabial approach (Figures 2-5) is safe, effective, and well-tolerated in most patients with CNPAS.

The procedure is performed in the operating room with the patient under general anesthesia using magnification with a 300 mm lens, otologic instruments, and a microdrill with diamond burr tips of different sizes. Once the patient is intubated in the supine position, the head is placed in the neutral position on a donut-shaped headrest and draped to protect the eyes, leaving the nose, midface, and mouth uncovered. The endotracheal tube is then secured in a position to allow optimal access to the oral cavity and not to inhibit the operator's movements. The nose is decongested with phenylephrine hydrochloride on neurosurgical pledgets. The upper gingival sulcus and pyriform aperture mucosa are then infiltrated with 1.5 mL of 1% lidocaine with 1:100,00 epinephrine loaded in a dental syringe. A 1.5-cm sublabial incision is made in the sulcus using needle-tipped cautery. The incision is extended down to the bone.

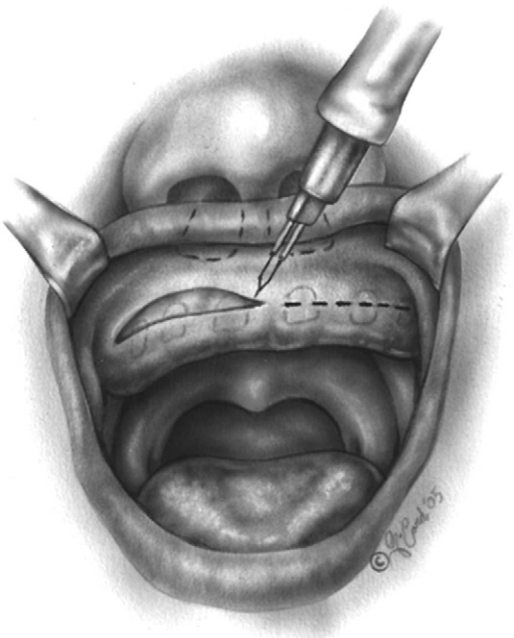

Figure 2 Sublabial incision using electrocautery.

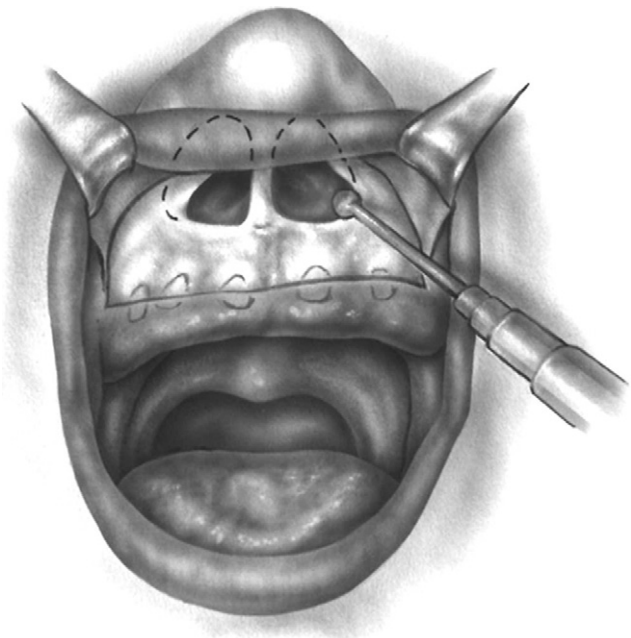

Figure 4 Drilling and expansion of the pyriform aperture.

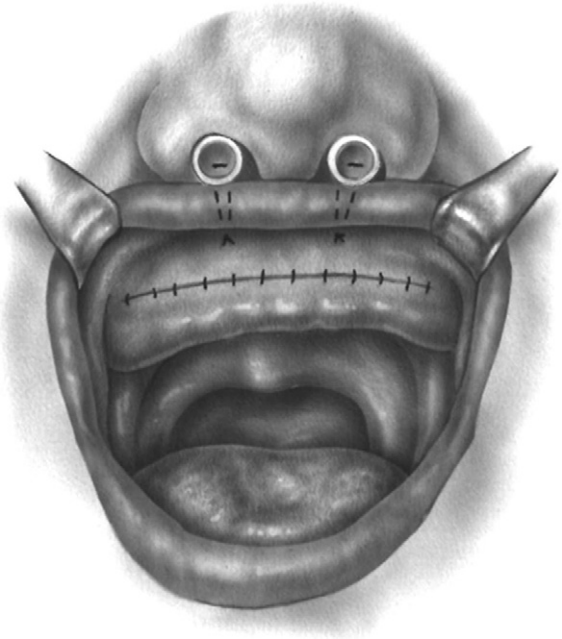

Figure 5 Placement of bilateral nasal stents, knots secured in sublabial sulcus.

Under magnification, gentle subperiosteal dissection is then performed, with the elevation of soft tissue and mucosa, exposing the anterior nasal spine and floor of each nostril. The pyriform aperture should be bilaterally visualized and its bony margin freed up, leaving mucoperiosteum intact along the nasal floor. A 2-mm diamond burr is used to resect the bone from the inferior and lateral margins of the pyriform aperture. An adequate airway is attained when the pyriform aperture provides the passage of a 3.5-mm inner diameter endotracheal tube stent. Dissection is kept anterior to the inferior turbinate to avoid damage to the nasolacrimal duct. Drilling on the nasal floor should be avoided to prevent injury to the tooth buds. At the end of the procedure, the elevated soft tissues are returned to their original location, and a releasing mucosal incision is placed on the floor of the nose. The latter will allow for placement of the stents in the enlarged pyriform aperture. The sublabial incision is closed loosely with interrupted absorbable sutures, such as chromic gut. Modified nasopharyngeal airways or trimmed 3.5-mm endotracheal tubes may be used to stent the enlarged pyriform aperture. We anchor the stents with sutures that originate sublabially in the gingival sulcus, pass through the floor of the nose and stent, and are tied back in the sulcus. Another method involves using a stitch through the columella, with a transverse segment of tubing used to prevent compression and pressure necrosis. The duration of stenting is variable, and can be anywhere between 3 and 14 days, with longer durations used for those patients who receive more extensive dissection. They should be left in place long enough to allow the soft tissues to coapt to the underlying bone, but not produce mucosal damage or necrosis. Van Den Abbeele et al[6] and Lee et al[13] suggest that nasal stents should be maintained for at least 5 days to avoid recurrent stenosis, but should not exceed 15 days to avoid mucosal injury. We use modified nasopharyngeal airways and typically stent for 5 days.

Complications

Potential hazards of this surgery relate to the extent and location of the bony dissection and postoperative stenting. Care must be taken during dissection to avoid damage to the periosteum, nasal mucosa, tooth buds, or nasolacrimal ducts. The tooth buds reach high into the premaxilla, and are present inferior and lateral to the area of dissection. Drilling anteriorly to the tip of the inferior turbinate should ensure avoidance of a nasolacrimal duct. If stents are left in place for insufficient duration, tissue dehiscence or restenosis may occur. If they are left in place for excessive duration, mucosal damage or necrosis may result. Another possible complication is retardation of facial growth; however, it has not been widely reported.

Postoperative care

All patients should be monitored in the intensive care unit for 24-48 hours to ensure airway patency. Nasal irrigations and suctioning may be used to maintain stent hygiene and patency. Once the stents are removed, vasoconstrictive and normal saline drops may be used for several days to minimize edema and crusting. A first-generation cephalosporin and acetaminophen are prescribed for infection prophylaxis and analgesia during the first postoperative week.

Long-term follow-up

Follow-up visits should include interim history detailing airway and feeding difficulties, infections, complications, or readmissions. Physical examination should assess nasal airway patency, facial growth, and eruption of dentition. If the patient has signs and symptoms of stenosis, there may be a need for re-expansion of the pyriform aperture.

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