Congenital Nasal Pyriform Aperture Stenosis

Amin Lim, BA; Richard B. Towbin, MD; Carrie M. Schaefer, MD; Alexander J. Towbin, MD

Abstract

Patients with congenital nasal pyriform aperture stenosis typically present with respiratory distress, including noisy breathing resembling snoring, apneic episodes during wakefulness and sleep, feeding difficulties, and failure to thrive. Sudden airway obstruction may occur but can be temporarily relieved by crying. Imaging plays a key role in diagnosis, with a pyriform aperture width of <11 mm considered diagnostic. While surgical intervention can be curative, the timing of surgery should be individualized based on symptom severity and overall prognosis. Supportive care remains essential to optimize outcomes and ensure the best possible management approach.

Keywords: head and neck, nose, congenital

Case Summary

A 4-week-old child presented with nasal obstruction and feeding difficulties.

Imaging Findings

Axial CT showed narrowing of the pyriform aperture, with the bony aperture measuring <4.5 mm in width with a high degree of obstruction (Figure 1). There were paired central incisors and a normal pituitary fossa and gland (not shown).

Diagnosis

Congenital nasal pyriform aperture stenosis.

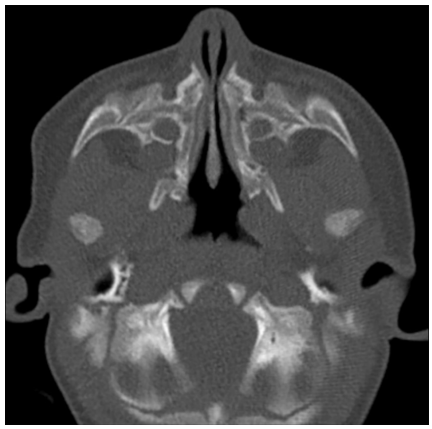
The clinical differential diagnosis of neonatal nasal obstruction includes choanal atresia, nasopharyngeal encephalocele, mucosal edema, dermoid cysts, skull base malformations, tumors, nasal hypoplasia, and nasal trauma.

Discussion

Congenital nasal pyriform aperture stenosis, also referred to as nasal inlet stenosis, is a rare cause of neonatal respiratory distress, with an estimated prevalence of 1 in 25,000 live births.[1] The condition is thought to result from excessive growth of the nasal process of the maxilla, leading to a narrowing of the pear-shaped pyriform aperture and increased airway resistance.[2] This anatomical narrowing restricts airflow, making it difficult for neonates to breathe.[3] Because newborns are obligate nasal breathers, severe nasal obstruction can lead to life-threatening respiratory distress.[4]

Neonates with congenital nasal pyriform aperture stenosis typically present with respiratory distress, noisy breathing resembling snoring, and episodes of apnea both during wakefulness and sleep. Additional symptoms include feeding difficulties, failure to thrive, and sudden airway obstruction, which may be temporarily relieved by crying.[5] The timing of symptom onset depends on the severity of the stenosis, with signs appearing as early as the first hours of life or developing months later.

Figure 1. Axial CT at the level of the nasal cavity showing severe narrowing (arrow) of both nasal pyriform apertures measuring <4.5 mm in width with a high degree of obstruction. There were paired central incisors and a normal pituitary fossa and gland (not shown).



While the exact pathology of congenital nasal pyriform aperture stenosis remains unclear, the condition has been associated with several genetic syndromes. Documented links exist between congenital nasal pyriform aperture stenosis and

Affiliations: University of Texas Medical Branch John Sealy School of Medicin, Galveston, Texas (Lim); Department of Radiology, Phoenix Children's Hospital, Phoenix, Arizona (RB Towbin, Schaefer); Department of Radiology, Cincinnati Children's Hospital, University of Cincinnati College of Medicine, Cincinnati, Ohio (AJ Towbin).

Disclosures: The authors have no conflicts of interest to disclose. None of the authors received outside funding for the production of this original manuscript and no part of this article has been previously published elsewhere.

https://doi.org/10.37549/JPCR-26-0100

semi lobar holoprosencephaly, craniofacial anomalies, solitary median maxillary central incisor, and endocrinologic disorders such as hypopituitary axis abnormalities.[4]

Although awareness of congenital nasal pyriform aperture stenosis has increased in recent years, its rarity and variable presentation continue to pose diagnostic challenges.[4] Distinguishing it from other causes of nasal obstruction requires a combination of clinical examination and maxillofacial CT findings. According to Baker and Pereira, suggestive physical examination findings include a narrowed anterior nasal fossa and the inability to pass a 5-French catheter or a 1.9-mm endoscope through the nasal cavity.[3]

Definitive diagnosis of congenital nasal pyriform aperture stenosis is established using thin-section CT imaging of the nose and face, which directly visualizes the stenotic pyriform aperture.[4] Maxillofacial CT should be performed in a plane parallel to the hard palate using 1.5-2-mm axial sections to accurately measure the dimensions of the pyriform aperture.[4] A pyriform aperture width of <11 mm in a term infant is considered diagnostic.

Beyond confirming the diagnosis, maxillofacial CT is essential for evaluating the severity of obstruction, identifying associated anomalies, and assessing the surrounding bony architecture.[4] In cases where comorbid conditions with a poor prognosis are suspected, additional imaging, such as a CT or US of the brain, should be performed before considering surgical intervention.[4]

The treatment of congenital nasal pyriform aperture stenosis depends on the severity of symptoms, the degree of airway obstruction, and the overall prognosis of the infant.[4] Initial management involves the use of a McGovern nipple or oral airway, along with supportive care such as nasal humidification, suctioning, topical steroids, and decongestant drops.[6] In infants with multiple anomalies and a poor prognosis, conservative management is typically recommended, with prognosis assessed based on comorbidities that contribute to failure to thrive.

Surgical treatment is reserved for infants with severe nasal obstruction who have a good prognosis and no significant comorbidities, or for those in whom 2 weeks of aggressive medical therapy has failed.[7] The "rule of 10" is often applied to determine optimal surgical timing, recommending deferral until the infant reaches 10 pounds, 10 weeks of age, and a hemoglobin level of 10 g/dL, provided that the child's respiratory status remains stable.[7] Until these criteria are met, supportive care should continue to optimize surgical outcomes8,9

Conclusion

Patients with congenital nasal pyriform aperture stenosis typically present with respiratory distress, including noisy breathing resembling snoring, apneic episodes during wakefulness and sleep, feeding difficulties, and failure to thrive. Sudden airway obstruction may occur but can be temporarily relieved by crying. Imaging plays a key role in diagnosis, with a pyriform aperture width of <11 mm considered diagnostic. While surgical intervention can be curative, the timing of surgery should be individualized based on symptom severity and overall prognosis. Supportive care remains essential to optimize outcomes and ensure the best possible management approach[9]

References

1) Tropsek R, Horoi M, Ziereisen F. Congenital nasal pyriform aperture stenosis in association with polymalformative syndrome of the midline. *Cureus*. 2023;15(9):e45153. doi:10.7759/cureus.45153

2) Brown OE, Myer CM 3rd, Manning SC. Congenital nasal pyriform aperture stenosis. *Laryngoscope*. 1989;99(1):86-91. doi:10.1288/00005537-198901000-00016

3) Ey EH, Han BK, Towbin RB, Jaun WK. Bony inlet stenosis as a cause of nasal airway obstruction. *Radiology*. 1988;168(2):477-479. doi:10.1148/radiology.168.2.3393667

4) Baker KA, Pereira KD. Congenital nasal pyriform aperture stenosis. *Oper Tech Otolaryngol Head Neck Surg*. 2009;20(3):178-182. doi:10.1016/j.otot.2009.11.001

5) Serrano TLI, Pfeilsticker L, Silva V, et al. Newborn nasal obstruction due to congenital nasal pyriform aperture stenosis. *Allergy Rhinol*. 2016;7(1):e37-e41. doi:10.2500/ar.2016.7.0146

6) Shikowitz MJ. Congenital nasal pyriform aperture stenosis: diagnosis and treatment. *Int J Pediatr Otorhinolaryngol*. 2003;67(6):635-639. doi:10.1016/s0165-5876(03)00018-1

7) Hui Y, Friedberg J, Crysdale WS. Congenital nasal pyriform aperture stenosis as a presenting feature of holoprosencephaly. *Int J Pediatr Otorhinolaryngol*. 1995;31(2):263-274. doi:10.1016/0165-5876(94)01096-g

8) Van Den Abbeele T, Triglia J-M, François M, Narcy P. Congenital nasal pyriform aperture stenosis: diagnosis and management of 20 cases. *Ann Otol Rhinol Laryngol*. 2001;110(1):70-75. doi:10.1177/000348940111000113

9) Marrugo Pardo GE, Parra Charris JS, Parra Charris AE, Villa Zuluaga DF. Congenital nasal pyriform aperture stenosis: diagnosis, management and technical considerations. *Acta Otorrinolaringologica*. 2020;71(3):154-159. doi:10.1016/j.otoeng.2020.02.001