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Case Report

Congenital Nasal Pyriform Aperture Stenosis

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A B S T R A C T

Introduction: Congenital nasal pyriform aperture is one of the newborns' main aetiologies of respiratory distress. Immediate diagnosis and intervention are mandatory because of the condition's life-threatening characteristics.

Case Presentation: It is a case of a 26-day full-term male infant admitted to the neonatal ICU for severe respiratory distress (stridor, abdominal respiration, rapid and shallow breathing, and chest retractions. The anterior rhinoscopy highlighted a narrowing of the nasal fossa. A nasal CT scan demonstrated a congenital stenosis in the pyriform aperture. The ENT surgeon and pediatric physician indicated urgent surgery to correct the pyriform aperture stenosis. The child improved his nasal breathing and was discharged from the ICU on the 10th day.

Discussion: Congenital stenosis of the pyriform aperture (CSPA) is one of the main aetiologies of newborn nasal obstruction, and it is due to excessive bony growth of the medial nasal process of the maxilla (2), narrowing the bony part of the nasal cavity. The diagnosis of CSPA is based on nasal endoscopy (narrow nasal fossa due to bone protrusion) and a CT scan (confirms the site of obstruction and shows normal choanae anatomy). The correctional surgical intervention of CSPA consists of pyriform aperture enlargement by endoral sublabial approach using small burs (2-3 mm).

Conclusion: CSPA IS considered a minor form of holoprosencephaly other than an isolated deformity as it once was thought to be.

Keywords: Congenital nasal pyriform aperture; Abdominal respiration; Rhinoscopy

1. Introduction

Congenital nasal pyriform aperture is one of the newborns' main aetiologies of respiratory distress. It is due to a bony overgrowth of the nasal lateral process of the maxilla. Immediate diagnosis and intervention are mandatory because of the condition's life-threatening characteristics. It is considered a minor form of holoprosencephaly other than an isolated deformity as it once was thought to be.

2. Case Presentation

It is a case of a 26-day full-term male infant admitted to the neonatal ICU for acute respiratory distress, a weight of 2900 g; at the initial assessment, he showed severe respiratory distress (stridor, abdominal respiration, rapid and shallow breathing, and chest retractions) a small nasal tube could not pass anteriorly as was custom in a normal nasal examination, an anterior rhinoscopy highlighted a narrowing of the nasal fossa. A nasal CT scan demonstrated a congenital stenosis in the pyriform aperture (**Figure 1**). Due to the gravity of the respiratory distress and inability to feed the infant, both the ENT surgeon and pediatric physician indicated urgent surgery to correct the pyriform aperture stenosis. A sublabial approach was conducted, and as the bony margins were exposed, we used small burs 2.0

and 2.5 mm to widen both pyriform apertures laterally and avoid damage to both the nasolacrimal duct and floor of the nasal fossa. A 3.5 mm endotracheal tube passed both pyriform apertures, confirming the procedure's efficacity. We opted for two silastik-made conformers and silastik nasal splints to avoid adherences formation (**Figure 2**). The infant was transferred back to the ICU, where he was intubated for four days. We removed both nasal splints on the 5th day, as the infant was also extubated on the same day. The nasal conformers were drawn on the 7th day postoperatively. The child improved his nasal breathing and was discharged from the ICU on the 10th day.

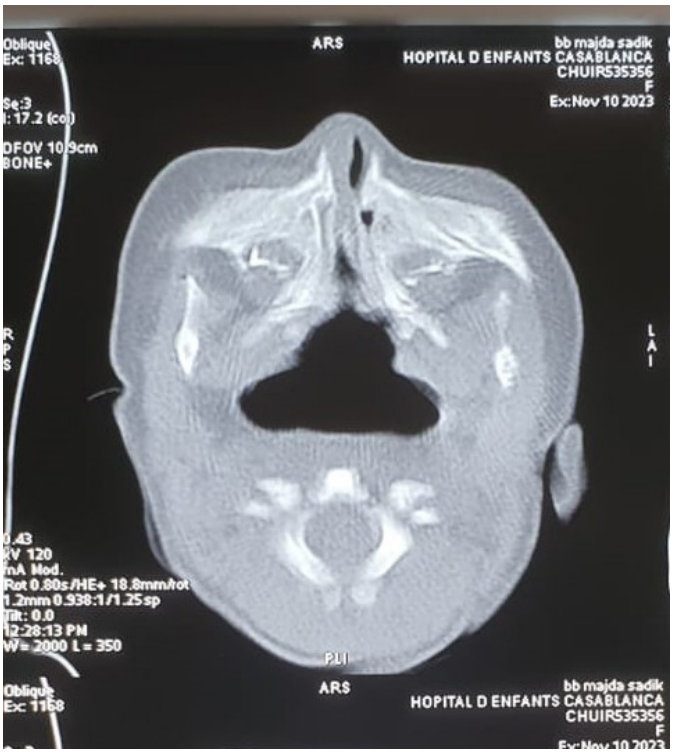

Figure 1: Axial section showing pyriform stenosis and nasal fossa obstruction.

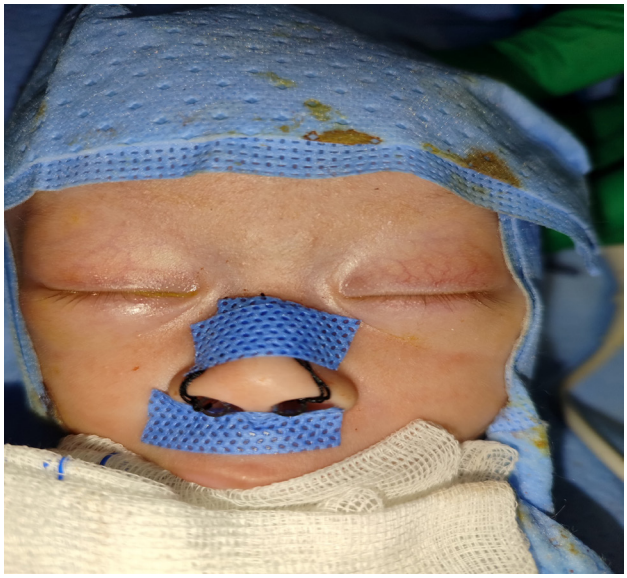

Figure 2: Postoperative dressing and nasal conformers positioning**.**

Discussion

Neonates are nasal breathers, and any cause of nasal obstruction could have severe respiratory consequences[1]. Congenital stenosis of the pyriform aperture (CSPA) is one of the main aetiologies of newborn nasal obstruction, and it is due to excessive bony growth of the medial nasal process of the maxilla[2], narrowing the bony part of the nasal cavity. The main symptomatic presentation is repeated spells of cyanosis relieved with a cry[1,3]. The diagnosis of CSPA is based on nasal endoscopy (narrow nasal fossa due to bone protrusion) and a CT scan (confirms the site of obstruction and shows normal choanae anatomy)[4].

The treatment of CSPA depends on the initial evaluation of the child. In mild cases, we used conservative methods such as nasal decongestants and humidification[3]; otherwise, severe cases of respiratory distress need endotracheal intubation and surveillance in ICU before an eventual surgical repair[6]. The correctional surgical intervention of CSPA consists of pyriform aperture enlargement by endoral sublabial approach using small burs (2-3 mm)[7]. Using nasal conformers and silastik nasal splints is usual to prevent scar-related stenosis and to reduce recurrences and must be retained for at least 6-7 days[8].

Surgical complications consist essentially of damage to the dental germs and nasolacrimal duct, which can be prevented by drilling anteriorly to the head of the inferior turbinate and avoiding injuring the mucosa of the nasal floor[5].

Conclusion

CSPA should be considered a potential cause of nasal obstruction in newborns when there are signs of respiratory distress and difficulty passing a small catheter in the anterior nasal valve. CT is the gold standard examination to confirm the diagnosis, and the therapeutic intervention could vary from conservative to surgical procedure depending on the initial assessment.

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