Congenital nasal pyriform aperture stenosis: a rare cause of upper airway obstruction in newborn

Diana Pinto Silva, Daniela Ribeiro, Sérgio Vilarinho, Luis Dias

Department of ENT, Hospital de Braga, Braga, Portugal

Correspondence to

Dr Diana Pinto Silva,
dianapintosilva@gmail.com

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DESCRIPTION

Congenital nasal pyriform aperture stenosis (CNPAS) is a rare cause of upper airway (UA) obstruction in newborn. Although commonly described as arising from overgrowth of the nasal process of the maxilla, some authors defend that it results from medialisation of normally shaped maxilla.¹ It is associated with other conditions and midline defects like solitary median maxillary central incisor (SMMCI) and holoprosencephaly.^{1,2}

We report a case of a newborn admitted in neonatal care unit on the second day of life with breathing distress, desaturation and cyanosis, reverted by crying. Ear, nose and throat observation revealed intercostal indrawing, oral breathing and absence of upper lip frenulum. On nasal examination, it was impossible to introduce the slim nasendoscope (Xion EF-N slim with 2.8 mm of diameter) in the nostrils, achieved after vasoconstriction and dilation with a 2 mm tube, with confirmation of normal septum and choanal patency. The CT showed pyriform aperture stenosis of 4.7 mm (measured on axial and coronal levels of the inferior meatus) and SMMCI (figure 1). Initially, conservative treatment and airway measures were applied, but once symptoms persisted, a surgical procedure was realised on the 15th day of life. A sublabial approach was performed for removal of the lateral nasal wall using a 2.8 mm diamond burr, widening nasal pyriform aperture. The procedure finished with placement and suturing tubes with 3.5 mm in each nostril (figure 2), removed after

Figure 2 Sublabial surgical approach. Exposure of the sublabial mucosa, mucosal incision to reach pyriform aperture stenosis and nasal process of maxilla, closure of the mucosa and placement of 3.5 mm tubes in the nostrils.

2 weeks, without complications. After 2 months, she experienced weight gain and had no feeding or breathing problems.

CNPAS is a rare condition of breathing distress in newborns. The diagnosis is suggested by physical examination due to failure to pass a nasogastric tube or slim nasendoscope within the first 1 cm of the nostrils and confirmed by CT findings of pyriform aperture width <11 mm (measured on an axial level of the inferior meatus) in a full-term neonate.^{1,2} The main differential diagnose is choanal atresia that is a congenital narrowing of the posterior nasal airway by a bony or membranous septum.^{1,2} First-line treatment is based on airway measures such as nasal saline solutions, decongestants or intranasal steroids, tried at least for 15 days.¹ Surgery is indicated towards conservative management failure or clinical presentation with apnoeic episodes, cyanosis, poor growth and failure to thrive.^{1,3} Current surgical procedure is sublabial approach with drill of the lateral nasal wall, that is effective but may involve complications such as injuries to lacrimal system and to the tooth buds.^{1,3} Postoperative nasal stenting is the norm, although the optimal duration for stenting remains a matter of debate.³ Novel and less-invasive treatments, such as rapid maxillary expansion, allows widening of the nasal base, leading to reduction in nasal airway resistance, having shown good results and may become standard treatment for CNPAS in

Figure 1 CT images (axial and coronal planes) with findings of pyriform aperture stenosis of 4.7 mm (measured at the level of the nasal inferior meatus) and solitary median maxillary central incisor (arrows).

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Learning points

- ▶ Upper airway (UA) obstruction in newborn should be carefully addressed.
- ▶ Congenital nasal pyriform aperture stenosis must be kept in mind as a rare but treatable cause of UA obstruction in neonates.
- ▶ The main goals are establishment of nasal patency, screen and management of other associated conditions, achieved with multidisciplinary teams.

the future.¹ Short and long follow-up should be done carefully by a multidisciplinary team.

CNPAS should be reminded in newborn with UA obstruction. Attempted diagnosis provides adequate management and treatment with re-establishment of nasal patency in first days of life.

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