An infrequent cause of neonatal upper airway obstruction: Congenital nasal pyriform aperture stenosis presenting to a remote facility

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INTRODUCTION

Congenital nasal pyriform aperture stenosis (CNPAS) is a rare condition characterized by nasal obstruction secondary to a hyperostosis of the nasal process of the maxilla or a medialization of otherwise normal maxillary architecture. Its association with solitary median maxillary central incisor (SMMC), panhypopituitarism, and hemivertebrae have led to its description as a microform of holoprosencephaly and a developmental field defect.

Resultant nasal obstruction may be marked by stertor and episodes of apnea or cyanosis. Narrow nasal passages typically impede nasoendoscopic investigation or nasogastric intubation beyond the stenosis. Pyriform aperture width of 11 mm or less measured using computed tomography (CT) is widely accepted as diagnostic for CNPAS, with triangular hard palate and prominent median inferior palatal bony ridge confirmatory.

First-line medical management includes topical nasal saline, intranasal decongestants, continuous positive airway pressure, or high flow nasal cannula therapy. Pyriform aperture width of less than 5.7 mm is associated with a greater risk of invasive intervention, which typically involves sublabial endo-oral incision and drill-out of the inferolateral pyriform borders followed by intranasal stenting, with or without transnasal reduction of the inferior turbinates. A transnasal dilation approach with reduced risks to surrounding structures as compared to open surgery was used in our case with good effect.

CASE REPORT

A full-term male neonate was born at a remote secondary care facility by induced vaginal delivery, indicated by fetal
macrosomia. Birth weight was 4160 g (94th percentile) with Apgar scores of 7, 8, and 10 (respectively at 1, 5, and 10 minutes). At 12 hours of life, increasing subcostal recessions, grunting, hypoxia, and poor oral intake were noted. Empiric intravenous antibiotics and airway support with high flow nasal cannula therapy at 1.5 L/kg in room air were commenced. Airway support was changed to continuous positive airway pressure (CPAP) at 6 cm H2O, 21% FiO2 to facilitate transfer to a larger center, and conservative management with nasal saline and suctioning proved sufficiently effective for the duration of this initial admission. Multiple attempts at passing a size 6 French nasogastric tube failed initially, though was eventually successful on day 2 of life. Successful passage of a flexible nasoendoscope bilaterally revealed enlarged inferior turbinates, crusting, and mucous congestion, though failed to reveal the stenosis at this stage. Improving respiratory distress and 48 hours without apnea allowed for the patient’s discharge from hospital on day 8 of life despite some persisting stertor while awake.

At week 3 of life, the patient presented with respiratory distress and blood-streaked nasal discharge. The patient had achieved a 300 g weight gain since birth despite persisting stertor and difficulty feeding. Conservative management with nasal saline and suction achieved an insufficient response, and the patient was recommenced on high flow 2 L·kg−1·min−1 at FiO2 21% and nasogastric feeding. A nasopharyngeal aspirate was negative for common viruses. In view of ongoing upper airway obstruction with clinical concerns about the possible diagnosis of CNPAS; transfer to a tertiary referral center was organized for a facial CT and ongoing management. Facial CT confirmed the diagnosis, with a 5.4 mm pyriform aperture width, a triangular hard palate, a prominent median inferior palatal bony ridge, and a solitary median maxillary central incisor (Figure 1).

Intensive care unit admission was necessitated at the tertiary center by episodes of desaturation and central cyanosis despite high flow nasal prong support. Due to failing conservative management, definitive surgical management was sought. Under general anesthesia, the nasal passages were visualized using rigid nasoendoscope and dilated to beyond the paranasal sinuses. A 3.5 mm ivory silastic endotracheal tube cut to 5.5 cm in length was used to stent the left nasal passage, and a nasogastric tube was placed in the right nare for enteral feeding. The stent was replaced at day 5 post-dilation under general anesthesia, and again at the bedside multiple times during the patient’s admission. Stent care consisted of hourly topical saline and suctioning to maintain patency, with recruitment of parental involvement supported. At 6 weeks post-dilation, the stent was definitively removed and the patient was discharged.

Follow-up magnetic resonance imaging of the face on day 2 post-dilation confirmed an increase of the pyriform aperture width to 7 mm and ruled out holoprosencephaly and structural pituitary pathology. On follow-up at 3 and 5 months, no further stertor or respiratory distress was noted, and growth was progressing normally.

**DISCUSSION**

The awareness of CNPAS has increased in recent years, though its rarity and variability in presentation can contribute to considerable diagnostic difficulty. The classical presentation of stertor, respiratory distress, cyclical cyanosis, and poor tolerance of oral intake were noted at initial presentation in our case; however, clinical suspicion of CNPAS was allayed due to successful nasogastric intubation and exploration by nasoendoscopy. The diagnosis of CNPAS may not be evident on nasoendoscopy, and all cases of persistent upper airway obstruction should be afforded a high index of suspicion. Thus, clinical assessment in conjunction with nasoendoscopic findings are essential in identifying those patients suited to definitive diagnostic imaging. In clinically difficult cases where diagnostic imaging is not immediately offered, close follow-up and reassessment should be instituted so as to avoid delayed diagnosis.

Fine-slice facial CT is the imaging modality of choice, with a pyriform aperture width of less than 11 mm (measured on an axial level of the inferior meatus) in a full-term neonate accepted as diagnostic. CT imaging was not conducted at the time of initial presentation due to the improving clinical picture and negative nasoendoscopic findings. The clinical benefit of imaging is most apparent in cases refractory to conservative interventions, as the predictive value of imaging regarding surgery is limited. As the effective portion of stenosis may not be restricted to the pyriform rim, development of multifactorial measurement of the nasal architecture is hoped to offer some promise of diagnostic accuracy and treatment...
prediction.10

In this case, first-line conservative interventions including nasal saline, suction, nasal decongestants, CPAP, and high flow nasal cannula therapy failed to prevent further cyanotic episodes and intensive care unit admission. The standard approach after failure of medical management entails invasive sublabial incision and drill-out of the pyriform aperture’s inferolateral margins followed by bilateral stent placement. This approach has a failure rate of 14% and risks damage to the nasolacrimal duct and tooth buds.11

Silva Merea et al12 introduced a combined sublabial drill-out with endonasal inferior turbinate reduction which achieved anecdotally superior results. The authors postulated that turbinate reduction may be the primary mechanism of this approach’s success, though were unable to access the turbinates transnasally without widening the pyriform aperture. Wine et al,8 however, proposed that a similar effect on the inferior turbinates may be achieved through a transnasal dilation using Hegar cervical dilators. Though minimally acting on the maxillary bony architecture, passage of serial dilators ranging from 2 to 5 mm in diameter results in turbinate outfracture and increases patency of the nasal airway.8 8

Following dilation, Wine et al8 reported long-term remission in 2 patients following primary dilation, with the 2 other patients requiring a second dilation. Postprocedural stenting was omitted to avoid stent-related adverse effects, including stent blockage, septal perforation, and alar ulceration. Further, stent care over the standard 1- to 8-week period can be resource intensive and prolong inpatient stay. However, the authors conceded that repeat dilation may have been avoided if stenting were to have been performed.8 8

To increase likelihood of definitive cure in our case, a single stent was used for the left nasal passage. It was important that the requirement for ongoing specialist input was minimized due to the remoteness of his residence, which was more than 1500 km from the nearest pediatric ENT service. A single stent was considered adequate and allowed nasogastric intubation for enteral nutrition via the right nostril, and no stent-related adverse effects were observed. Difficulty was found in parental involvement in stent care at times, however, and postoperative admission was prolonged due to inability to manage this aspect of care on an outpatient basis.

CNPAS is a rare and life-threatening cause of upper airway obstruction in neonates. This case demonstrates that CNPAS should be considered a differential diagnosis for upper airway obstruction in neonates even in the absence of characteristic nasoendoscopic findings or difficult nasogastric intubation. A fine-slice facial CT should be considered where clinical suspicion of CNPAS is high.

CONSENT FOR PUBLICATION
Written consent was provided on behalf of the patient by his parent.

CONFLICT OF INTEREST
None.

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