



# Congenital Nasal Pyriform Aperture Stenosis: Same CT Dimensions, Varied Scenarios

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Indian J Radiol Imaging 2022;32:591–593.

## Abstract

Congenital nasal pyriform aperture stenosis (CNPAS) is a rare cause of neonatal respiratory distress. We report a case series of four infants with similar radiological dimensions but while two needed surgery, two could be managed conservatively. The clinical presentation of the child and the response to conservative treatment, rather than the radiological dimensions were the main predictors for surgical intervention.

## Keywords

- ▶ congenital nasal pyriform aperture stenosis
- ▶ infants
- ▶ radiology
- ▶ CT PNS
- ▶ sub labial approach
- ▶ endoscopic approach

## Introduction

Congenital nasal pyriform aperture stenosis (CNPAS) is a rare cause of nasal obstruction in a neonate with significant breathing difficulty.<sup>1,2</sup> The diagnosis is confirmed by computed tomography (CT) scan of the nose and paranasal sinuses (PNS), which typically shows narrow width of the pyriform aperture.<sup>1,2</sup> This case series highlights the varied clinical presentation of four infants with CNPAS despite having similar radiological dimensions.

### Case 1

A neonate with cerebrocostomandibular syndrome and maternal history of gestational diabetes mellitus (GDM) developed respiratory distress at 2 hours of life. Failure to pass 5 Fr nasogastric tube prompted a diagnosis of choanal stenosis. CT PNS showed narrow pyriform aperture measuring 5 mm (corresponds to the narrowest measurement on axial CT

images at the anterior most portion of the bony nasal cavity between the nasal processes of maxillary bones on either sides), triangular hard palate, single central maxillary mega-incisor tooth, and prominent inferior palatal ridge along the inferior surface of the hard palate suggestive of CNPAS. (–Fig. 1A–C) MRI brain was normal except for showing a dilated nasolacrimal duct. (–Fig. 1D). Conservative treatment was attempted but in view of repeated desaturation and persistent carbon dioxide retention, endoscopic surgical correction was undertaken. Stents modified from silicon Foley's catheter were placed in both nasal cavities.<sup>3</sup> They were kept patent with frequent saline nasal drops and regular nasal suctioning and removed after 4 weeks. The child was asymptomatic on further follow up.

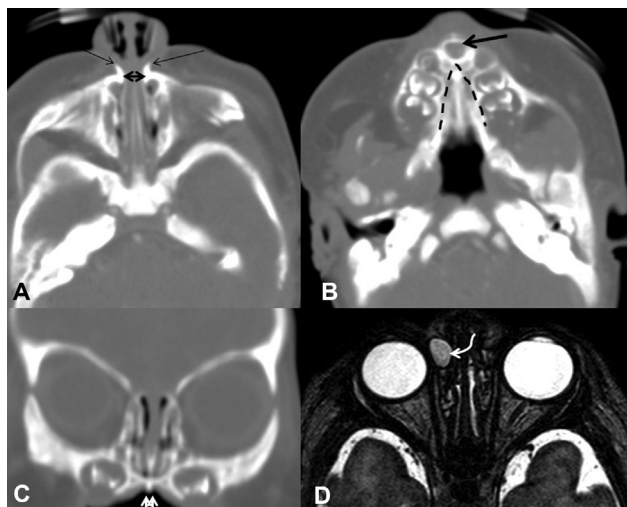
### Case 2

A five-month-old girl presented with breathing and feeding difficulty since birth. There was maternal history of GDM.

published online  
August 30, 2022

DOI <https://doi.org/10.1055/s-0042-1755249>.  
ISSN 0971-3026.

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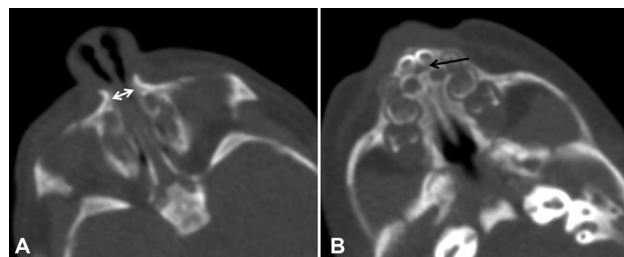


**Fig. 1** Axial CT section at the level of the pyriform aperture (A) and at the level of the hard palate and upper alveolus (B), coronal CT section at mid nasal cavity level (C) and heavily T2-weighted MRI axial section at the level of the orbits (D), showing narrowing of the pyriform aperture measuring 5 mm at level of nasal process of maxilla (double sided arrow in A), thickened medially bowed nasal process of maxilla on both sides (*thin arrows* in A), single mega maxillary incisor (*arrow* in B), triangular hard palate (*interrupted lines* in B), prominent palatal ridge (*double arrows* in C), dilated nasolacrimal duct (*curved arrow* in D).

She required multiple admissions in the peripheral center in the first few months of life. She was fed with orogastric tube throughout early infancy and had significant swallowing difficulty at presentation. CT scan was suggestive of CNPAS (4.5 mm aperture between the nasal processes of maxillary bones on either sides), midline central mega incisor, and prominent midline inferior palatal bony ridge as described in **Fig. 2A–C**. She underwent sublabial approach repair of the stenosis with the placement of bilateral silicon stents as described earlier for 2 weeks. Simultaneous swallowing therapy was initiated. At 2 year follow up, she is doing well with no nasal or feeding problems.



**Fig. 2** Axial CT section at the level of the pyriform aperture (A), coronal CT section anterior to the maxillary sinus (B) 3D reconstruction of the face (C), showing narrowing of the pyriform aperture measuring < 5 mm at level of nasal process of maxilla (*curved arrow* in A), thickened medially bowed nasal process of maxilla on both sides (*straight arrows* in A), single central maxillary megaincisor (*arrow* in B), narrowed pyriform aperture (*curved arrow* in C), and single central incisor (*black arrow* in C).



**Fig. 3** Axial CT section at the level of the pyriform aperture (A) and at the level of the hard palate and upper alveolus (B), showing thickened medially bowed nasal process of maxilla on both sides causing narrowing of the pyriform aperture (*double-sided arrow* in A), absence of the single maxillary incisor (*arrow* in B).

### Case 3

A preterm low birth weight (1.2 kg) newborn was noted to have labored breathing at birth. Maternal history of GDM was present. CT scan showed pyriform aperture narrowing of 5 mm. Other findings were a triangular hard palate and a prominent inferior palatal ridge (**Fig. 3A, B**). There was no central mega-incisor tooth. Conservative treatment was started with saline nasal drops every 2 hours, an oral airway was inserted and the child was shifted out of the ICU to the ward on day 6. On day 26, the child was maintaining saturation in room air even after oral airway was removed. He was discharged and continues to be asymptomatic at 2-year follow up.

### Case 4

A 1-month-old infant presented with feeding and breathing difficulty since birth. CT confirmed CNPAS measuring 4.8 mm between the nasal processes of the maxilla. The acute exacerbation was treated with intravenous antibiotics and noninvasive ventilation for 2 weeks after which the infant was gradually discharged on regular nasal drops. At 2-year follow up, there was good weight gain with no respiratory distress.

## Discussion

CNPAS forms a rare diagnosis of congenital nasal obstruction. It is believed to be due to either deficiency of the primary

palate, or bony overgrowth in the nasal process of the maxilla.<sup>1-3</sup> A strong association with GDM has been described.<sup>2</sup> In our case series, three out of four (75%) infants had GDM. A single maxillary mega-incisor, holoprosencephaly, and cleft palate are some of the abnormalities that can be associated with CNPAS.<sup>1,4</sup> In our series, three out of four patients had mega-incisor and all had prominent palatal ridge. Holoprosencephaly was not seen in any child in our series.

Most children are diagnosed at birth or shortly after birth. The initial suspicion is raised when there is failure to pass a 5-6 Fr nasal catheter.<sup>3,4</sup> The presentation is either acute with apneas, cyanosis, respiratory distress or they can present later with poor feeding, aspiration, and failure to thrive. Nasal endoscopy and the depth at which resistance is encountered can give some indication if the obstruction is in anterior nares, mid nose, or the choana.<sup>1,3</sup> CT is the gold standard for diagnosis. Different authors have mentioned various widths to call it a narrow pyriform aperture on CT ranging from 5 to 11 mm.<sup>1,5,6</sup> In our series, even though the width of the aperture was less than 5 mm in all patients, it was the clinical condition that dictated the need for surgical intervention. The first patient had apnoea and multiple desaturations on weaning oxygen therapy that prompted the need for surgery. In the second patient, swallowing difficulties with nose block necessitated surgery while the last two could be managed conservatively despite having same radiological dimensions. This suggests that while imaging is essential for diagnosis and for ruling out other nasal lesions, the radiologically measured dimensions alone do not decide the need for surgery. This opinion has been shared by other authors.<sup>2</sup> In contrast, some others have opined that a width less than 5.7 mm is more likely to need surgical intervention.<sup>7</sup>

The initial treatment is a trial of medical therapy with decongestants and nasal humidification with saline and topical steroids.<sup>2</sup> While this may be enough in milder cases, severe cases may require airway stabilization using McGovern nipple, continuous positive airway pressure, or even endotracheal intubation.<sup>3</sup> A 2-week waiting time is considered appropriate by most authors.<sup>1,8</sup> The presence of sleep apnea, failure to wean off oxygen or extubate, feeding difficulties, and cyanosis are the main indications for

surgery.<sup>1,8</sup> Both sublabial and endoscopic approaches have shown good surgical results.<sup>1,4</sup>

## Conclusion

This case series highlights that imaging is essential to diagnose CNPAS and rule out other nasal pathologies. However, the radiological dimensions alone do not determine the need for surgery. The surgeon must correlate the radiological findings with the clinical presentation of the child and the response to treatment in the 2-week trial period before deciding on surgical intervention.

## Conflict of Interest

None declared.

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