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# An unusual cause of neonatal respiratory distress

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#### **DESCRIPTION**

Three-week-old female presented with history of choking, crying, respiratory distress with feeds and associated failure to regain birth weight. On examination she had significant respiratory distress with tachypnoea and marked subcostal/intercostal recession. Oxygen saturations were initially 95% in air but dropped to 69% when feeding.

Shortly after admission the patient developed sudden acute upper airway obstruction requiring emergency oral intubation and ventilation. A nasogastric tube was sited, passing easily through the right nostril but not the left. A CT scan undertaken to delineate nasal anatomy showed nasal inlet stenosis due to medialisation of the nasal processes of

the maxilla at the inlet of the nose (figure 1). A diagnosis of congenital nasal pyriform aperture stenosis (CNPAS) was made and subsequent closer inspection of CT images showed the presence of an associated unerupted central upper frontal mega-incisor tooth (figure 2).

CNPAS is rare and should be distinguished from other causes of nasal airway obstruction such as choanal stenosis/atresia. Definitive diagnosis is made by CT, with pyriform aperture width of <11 mm being considered diagnostic in term infants. CNPAS can occur in isolation or be associated with holoprosencephaly, ocular coloboma, pituitary deficiency, a central mega-incisor, clinodactyly, hypoterorism, cleft palate or chromosomal abnormalities. Management



Figure 1 CT scan showing nasal pyriform aperture stenosis measuring 5 mm between the nasal processes of the maxilla (arrows).

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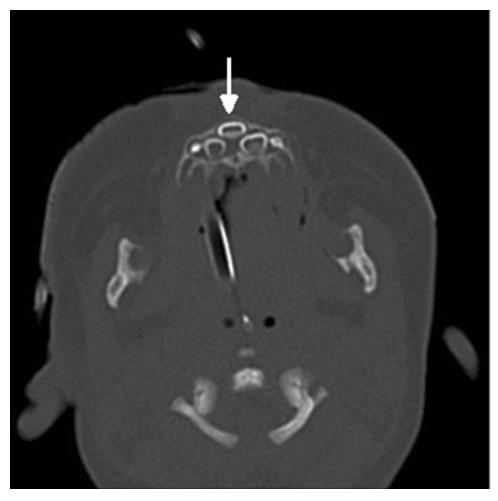


Figure 2 CT scan showing central frontal mega-incisor tooth (arrow).

can be conservative or involve surgical enlargement and temporary stenting. 1

Competing interests None.

Patient consent Obtained.

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