

CASE REPORT

CT findings of congenital neonatal pyriform aperture stenosis

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Abstract

Congenital neonatal pyriform aperture stenosis (CNPAS) is a rare but potentially lethal condition that causes respiratory distress. The characteristic narrowing of the pyriform aperture along with other associated craniofacial dysmorphism is diagnosed using cross-sectional imaging such as computed tomography (CT) and magnetic resonance imaging. CT scan is the imaging of choice for confirming and characterizing CNPAS. Infants are obligate nasal breathers in the first 5 months of life. Hence, a high degree of clinical suspicion, prompt imaging diagnosis and adequate respiratory support is critical to help reduce the morbidity of this condition.

INTRODUCTION

Congenital neonatal pyriform aperture stenosis (CNPAS) is a rare but potentially lethal condition that causes respiratory distress affecting 1 in 50 000 neonates [1]. Patients often present with respiratory distress, poor feeding with bouts of apnea and cyanosis. The diagnosis of CNPAS is made using a computed tomography (CT) scan that reveals a narrowed pyriform aperture with a width of less than 11 mm. While the exact cause of this condition is not clear, CNPAS has been associated with a missense mutation in the Sonic Hedgehog gene, SHH (I111F) [2].

A high degree of clinical suspicion is needed for timely diagnosis as infants are obligate nasal breathers in the first 5 months of life. Severe narrowing of the pyriform aperture significantly increases the resistance of the nasal passage leading to fatigue and respiratory distress [3]. Prompt diagnosis and adequate respiratory support are critical to help reduce the morbidity and long-term sequelae of this condition.

CASE REPORT

A newborn term baby presented with breathing difficulty immediately after birth. Oxygen saturation of 85% under room air was documented. There were associated subcostal recession and rapid breathing of more than 70 breaths per minute. The attending physician was not able to pass a 6 Fr feeding tube into the nasal cavity during routine suction. Oxygen saturation improved after the patient was supported with non-invasive ventilation. On physical examination, the child did not have any midline effects, hypotelorism, microcephaly, ocular abnormalities or simian crease. The rest of the physical examination was unremarkable.

CNPAS was suspected and an unenhanced CT scan of the paranasal sinuses was arranged. Narrowing of the pyriform aperture is the hallmark of CNPAS. CT scan demonstrated bilateral medial displacement of the nasal prominences resulting in extremely narrow nasal passages measuring 6 mm in width

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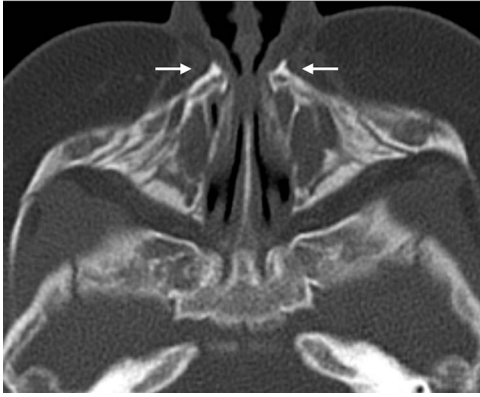


Figure 1: Axial CT image in bone window at the level of the paranasal sinuses showing narrowed pyriform aperture stenosis.

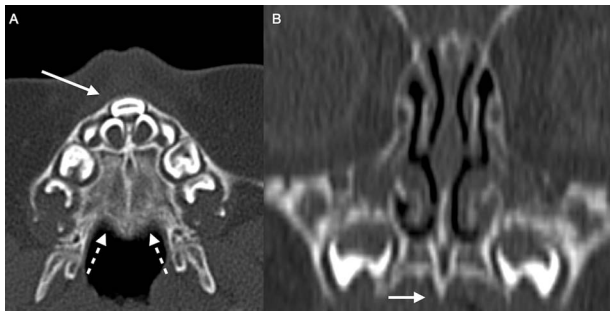


Figure 2: Axial CT image in bone window at the level of the superior alveolar ridge shows solitary median maxillary central incisor (solid arrow, A) and a narrow triangular hard palate (dashed arrows, A). Coronal CT image showing a bony ridge at the inferior surface of the hard palate (B).

(Fig. 1). There was an associated solitary median maxillary central incisor with a narrow triangular hard palate (Fig. 2A). Coronal reformatted CT images revealed a bony ridge along the inferior surface of the hard palate (Fig. 2B).

The child was treated with non-invasive respiratory support, regular suctioning and room air humidification. The child responded well to the conservative management and was discharged after 10 days. MRI was not performed due to restrictions brought about by the ongoing COVID-19 pandemic during the delivery and initial hospital admission. Subsequent follow-up under both paediatric otolaryngology as well as general paediatric clinics was planned.

DISCUSSION

Embryologically, the nasal grooves from the mesodermal layer are transformed into nasal pits in the first 8 weeks after conception [4]. Between the 8th and 24th week, the nasal pits are filled with epithelial tissue which then gets resorbed in normal development. Insufficient resorption of the epithelial plug leads to the formation of prominent nasal processes of the maxilla, resulting in a narrowed pyriform aperture. The reduction in the cross-section area at the pyriform aperture results in a significant increase in the resistance of the nasal passage. Consequently, the neonate needs to exert more effort for respiration.

By consensus, a pyriform aperture width of less than 11 mm is considered diagnostic [5]. CT scan of the paranasal sinuses is the most helpful modality to assess the nasal passage in neonates.

First described in 1989, CNPAS can present as a sporadic anomaly or as part of solitary median maxillary central incisor syndrome (SMMCI) [6]. As such, additional cross-sectional imaging (MRI) also plays an important role in identifying associated nasal anomalies (such as nasoethmoid encephaloceles), congenital brain anomalies (holoprosencephaly) and ocular defects (coloboma).

In our case, a routine examination by the paediatric retrieval team shortly after birth revealed signs of laboured breathing and impending respiratory distress. Routine nasal suctioning plays an important role in clearing the nasal passageway as well as providing a means of determining the presence and location of any possible airway obstruction. In CNPAS, the suction tube will invariably meet resistance after being inserted 2–4 mm into the nostrils, making further advancement impossible.

CNPAS should be distinguished from choanal atresia, which is the narrowing and obstruction of the posterior nasal airways by a bony or membranous septum. The clinical presentation of CNPAS and choanal atresia can be very similar as both cause subcostal recession, tachypnea and cyanosis. However, a CT scan of the paranasal sinuses provides adequate information to distinguish between the two conditions as the management approaches for each condition differ significantly [7]. Neonates with choanal atresia invariably need surgical intervention ranging from transnasal puncture, transpalatal repair, transnasal endoscopic repair and post-operative stenting [7].

Conversely, patients with CNPAS can be managed conservatively or with surgery [8]. Conservative management is preferred when the neonate can maintain good oxygen saturation without signs of respiratory distress. In such cases, meticulous nasal hygiene and suctioning, application of adrenaline saline solution and room air humidification have shown to be effective treatment measures. Non-invasive respiratory support is given during this time and gradually tapered down. Severe CNPAS requires rhinoplasty which entails surgical widening of the nasal passage and the insertion of stents [9]. Surgical intervention is also considered when non-invasive treatment does not help resolve the patient's symptoms. The presence of associated craniofacial anomalies is the main cause of surgical treatment failure in approximately 14% of patients in one relatively sizeable study [10]. CNPAS is a condition that tends to improve as the child grows with an excellent prognosis. Even when initial surgery was inadequate, no recurrence has been reported in the literature [10].

Prompt diagnosis and adequate respiratory support are crucial for neonates with CNPAS. Delay in initiating appropriate treatment puts the child at risk of respiratory distress and apnea which, in severe cases, may lead to ischemic brain injury and death. Diagnosis is made using an unenhanced CT scan of the paranasal sinuses with a pyriform aperture measuring less than 11 mm. Imaging also plays an important role in identifying associated craniofacial dysmorphism, which may affect treatment outcomes.

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CONFLICT OF INTERESTS

All authors have no conflict of interests to declare.

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ETHICAL APPROVAL

Not required.

CONSENT

Written consent has been obtained.

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