Conservative management of congenital nasal pyriform aperture stenosis

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DESCRIPTION

A newborn baby was referred to ENT team with recurrent episodes of desaturations which were worse on feeding and relieved by crying. She was born at 36 weeks with low birth weight (1.9 kgs) to a mother with history of high alcohol intake; Apgar score 6 (1 min) and 9 (5 min). Respiratory distress at birth required a continuous positive airway pressure face mask. A small nasogastric tube was passed via each nostril ruling out choanal atresia. Subsequently episodes of distress and desaturations continued requiring nasal oxygen.

On examination, the baby was noted to have mildly dysmorphic features, specifically small nostrils and a flattened nasal bridge. A CT scan suggested that the pyriform apertures were narrowed (5.3 mm on axial view at the level of the inferior meatus, [figure 1](#)).

Without nasal airway support, the episodes of distress with nasal alar collapse and desaturations as low as 80% SpO₂ continued. It became apparent that oxygen was not required but the presence of the nasal prongs themselves prevented the alar collapse, allowing maintenance of the airway and saturations on air.

The ENT team applied a nasal stent fashioned out of a paediatric curved tip nasal cannula which

Figure 2 Paediatric curved tip nasal cannula with anterior wall cut.

significantly improved her symptoms ([figures 2 and 3](#)). No further desaturations were noted. Mum was trained and observed in application of the nasal stent and the baby discharged from Special Care Baby Unit (SCBU). Over time, the stent use was limited to periods of feeding and overnight sleep. By the fourth month, the stent was no longer required. Subsequent investigations including genetic testing showed bilateral sensorineural hearing loss and Waardenburg syndrome.

Congenital nasal pyriform aperture stenosis (CNPAS) is quite rare estimated at 1 in 25 000 newborns and is frequently missed or even unknown to some neonatal intensivists.^{1,2} Being obligate nasal breathers, this condition among newborns can lead to significant respiratory distress presenting as apnoeic crisis, episodic cyanosis relieved by crying and difficulties to breath on feeding. It is believed that the pathology is due to overgrowth of the nasal processes of the maxilla on the fourth month of foetal development.³

Figure 1 Axial view of head CT scan suggestive of narrowed pyriform aperture.

Figure 3 Modified nasal cannula used as nasal splint.

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Measured on axial CT scan, a distance between maxillary nasal processes at the level of inferior turbinates of less than 11 mm, is considered diagnostic of CNPAS.^{1–3} The management of CNPAS depends on the severity of the symptoms with mild obstruction treated conservatively including nasal splint for a few days while moderate-to-severe obstruction with surgical intervention. Wormald *et al* reported pyriform aperture width of 5.7 mm or less requires surgical management.⁴ This case report, however, had 5.3 mm width with significant desaturations but was successfully managed conservatively with nasal stent although used for longer duration. Lin *et al* found that there was no statistical difference in the initial measured pyriform aperture between patients with CNPAS treated surgically and conservatively which reflects the natural course of this congenital abnormality.⁵ Clinical presentation and symptoms are then more important than

CT measurements in the decision-making process in the management of CNPAS.

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Learning points

- ▶ Congenital nasal pyriform aperture stenosis is a diagnosis that can sometimes be missed and even unknown to some neonatal specialists.
- ▶ Mild-to-moderate nasal pyriform aperture stenosis can be managed conservatively with nasal splints.
- ▶ Clinical presentation is more important than CT scan measurements in the decision-making process of management options.

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