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Novel use of high-flow nasal cannula therapy in the management of pyriform aperture stenosis: case report

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Abstract

Background. Pyriform aperture stenosis is a rare form of congenital nasal obstruction; it poses a management dilemma for otolaryngologists and physicians alike. It can result in poor weight gain and potentially life-threatening airflow obstruction. The challenge lies in the difficulty to predict which patients will require invasive operative management versus conservative therapy alone.

Case report. This case demonstrates the successful use of high-flow nasal cannula therapy in a young child with pyriform aperture stenosis.

Introduction

Pyriform aperture stenosis is a rare form of nasal airway obstruction in the neonate that occurs secondary to narrowing of the anterior bony nasal passageway. As newborns are initially preferential nasal breathers, pyriform aperture stenosis presents early, with potentially life-threatening nasal obstruction.[1,2] Symptoms include laboured breathing, apnoeic events, cyclical cyanosis improved by crying, and failure to thrive.[1,3]

Clinicians are often alerted to the presence of pyriform aperture stenosis in the neonate after difficulty passing a nasoendoscope into the nose during the evaluation of nasal obstruction, and respiratory distress. The diagnosis is confirmed with fine-slice computed tomography (CT). Belden *et al.* suggested a pyriform aperture diameter of 11 mm or less in full-term babies as diagnostic.[4] A recent study by Wormald *et al.* suggested that diameters of less than 5.7 mm will require surgical intervention.[5]

Management of pyriform aperture stenosis is aimed at reducing nasal passage obstruction and improving airflow. Conventional treatment options include conservative approaches, such as topical nasal saline, steroids, decongestants and Silastic™ stents, and surgical procedures to enlarge the pyriform aperture.[6] Below, we present a case of the successful use of long-term high-flow nasal cannula therapy without oxygen supplementation in the conservative management of pyriform aperture stenosis.

Case report

This male patient was born at 37 plus 6 weeks gestation by vaginal delivery, with Apgar scores of 9 and 9, and a birthweight of 2.5 kg. At 11 hours, he developed significant respiratory distress, which required admission to the neonatal intensive care unit and continuous positive airway pressure (CPAP) non-invasive ventilation.

Given no signs of an infectious precipitant and a normal chest X-ray, the ENT department were consulted for an opinion on possible anatomical airway obstruction. On review, a 6 French gauge (outer diameter, 2 mm) suction catheter was able to be passed through both nostrils, thereby excluding choanal atresia. The patient's respiratory effort subsequently improved, except for mild stertor and snoring at night, and he was discharged home at one week of age on intranasal corticosteroids and saline nasal spray.

Two weeks later, the patient re-presented to hospital with bronchiolitis heralded by significant respiratory distress, hypoxia and noisy stertorous breathing. Viral swabs were taken and he was commenced on high-flow nasal cannula therapy. On further ENT review, an inability to pass a 2.7 mm nasoendoscope through either pyriform aperture raised suspicion for anatomical nasal obstruction. A CT of the facial bones confirmed pyriform aperture stenosis, with a distance of 3 mm between the maxillary processes, as well as the presence of a single central maxillary incisor (Figure 1).

The patient underwent further investigation, including magnetic resonance imaging of the brain, renal tract ultrasound, spinal cord ultrasound, chromosome microarray and karyotype, the findings of which were all unremarkable. A clinical genetics review excluded features of a genetic syndromal diagnosis associated with holoprosencephaly.

A period of medical management was instigated, with the judicious use of intranasal dexamethasone drops, intermittent nasal decongestants and nasal saline washes. The patient's clinical picture fluctuated, with periods of clinical stability with no increased

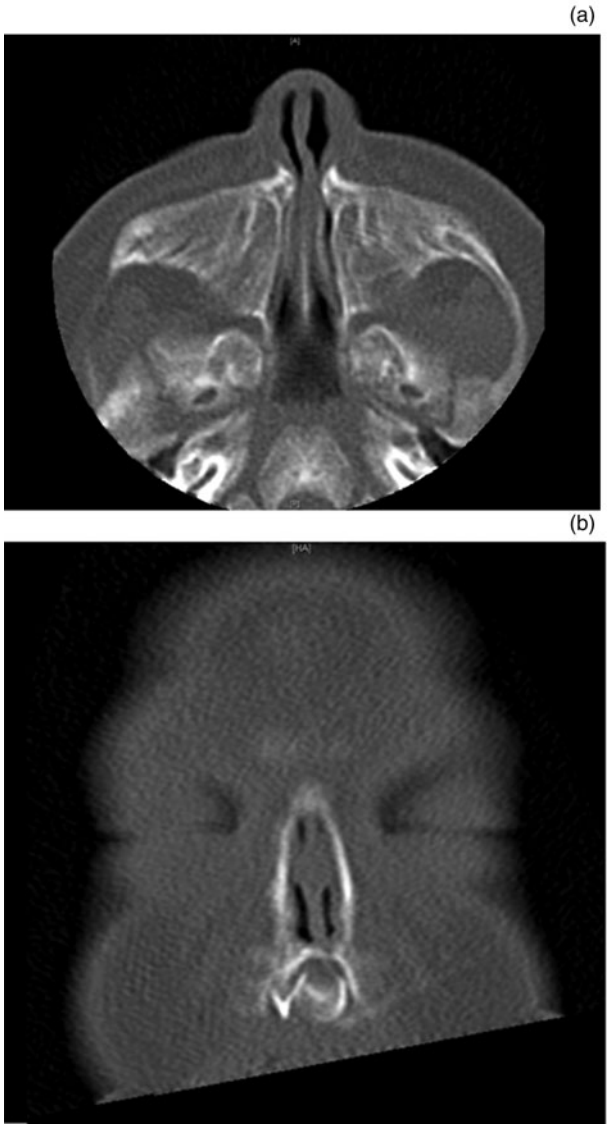

Fig. 1. (a) Axial computed tomography (CT) scan demonstrating a narrowed pyriform aperture. (b) Coronal CT scan demonstrating a single maxillary incisor.

respiratory effort or oxygen requirement. Overnight oximetry, however, demonstrated multiple desaturations, and the patient failed to gain weight between weeks four and five of life.

Surgical intervention was offered, but the patient's parents elected to trial non-invasive ventilation as a first measure. Nasal mask CPAP was commenced, which resulted in fewer overnight desaturations and improved work of breathing during sleep. Unfortunately, this was poorly tolerated; ventilatory support was therefore changed to high-flow nasal cannula treatment in room air, to good effect as seen on repeat overnight oximetry (Figure 2). He was discharged home on overnight high-flow nasal cannula therapy, and with a weaning schedule of decongestants and topical steroids.

The patient was followed up at nine months and was noted to have significant weight gain and growth (Figure 3), and was still tolerating therapy without any facial pressure sores or other complications. Fortunately, he has not required any further hospitalisations at last review (nine months of age).

Discussion

Nasal airflow obstruction, as occurs in pyriform aperture stenosis, can be potentially life-threatening in neonates. Whilst the exact aetiology of pyriform aperture stenosis is unknown, it is hypothesised to occur because of an overgrowth of the maxillary nasal process during fetal development.[4,6,7] Pyriform aperture stenosis can arise alone or in conjunction with a number of craniofacial abnormalities, including a single central maxillary incisor, cleft palate and holoprosencephaly. It often presents in the first hours of life with laboured breathing, respiratory distress, stertor, and cyclical cyanosis that improves with crying.[1,3]

The diagnosis of pyriform aperture stenosis relies upon otolaryngological examination, with fine-slice CT being the imaging modality of choice. An inability to perform nasoendoscopy because of narrow bony nasal inlets and difficulty passing fine-bore nasogastric tubes through either nostril are key features. There is still no clear consensus on diagnostic criteria using CT imaging. Multiple studies have suggested radiologically measured diagnostic pyriform aperture diameters ranging from less than 3 mm to less than 11 mm.[2,4,8]

The overall goal of management of pyriform aperture stenosis is to improve nasal airflow and reduce nasal passage obstruction. Traditionally, conservative management strategies entail the use of topical therapies such as nasal saline, decongestants and intranasal steroids, to reduce mucosal size and secretions. Whilst previously used, intranasal Silastic stents have become less favourable, as their small diameter results in frequent occlusion.

The surgical management of pyriform aperture stenosis involves widening of the stenosed pyriform aperture, which can be performed via a number of approaches. The more favourable method is a sublabial endo-oral procedure, utilising burrs to drill away the nasal process of the maxilla, whilst being mindful of the close proximity of the developing tooth buds, the nasolacrimal duct and the infraorbital nerve. This is followed by nasal stent insertion to reduce the incidence of stenosis recurrence or scar-mediated restenosis. Other options include a transnasal approach, which is limited by the increased risk of soft tissue injury, and balloon dilatation of the pyriform aperture.

The challenge of managing pyriform aperture stenosis lies in the difficulty of patient selection for different management options, with clinicians aiming to implement the least invasive treatment required, as many patients will improve with growth. A lack of data on the natural course of pyriform aperture stenosis makes it difficult for clinicians to predict which patients will not improve with conservative management alone.[3]

There is conflicting evidence on indicators for surgery. In a pooled case series, Wormald *et al.* demonstrated that a pyriform aperture diameter of 5.7 mm or less was associated with the requirement of surgical intervention.[5] However, subsequent studies found no correlation between degree of stenosis and requirement for surgery, instead suggesting evaluation of the patient's symptoms and progress.[3,9] Indications for surgery within these studies were: failure of response to medical treatment, presence of sleep apnoea due to nasal airway obstruction, an inability to insert a 6 French gauge feeding tube, extubation failure, and frequent feeding difficulties with cyanosis. Recently, Moreddu *et al.* elaborated a management algorithm from their experience of managing 10 patients with pyriform aperture stenosis, which suggests undertaking surgery only in those patients who fail to improve after two weeks of medical therapy.[10]

High-flow nasal cannula treatment is a novel method of respiratory support that delivers humidified and warmed gas to the nasal airways (with or without supplementary oxygen),

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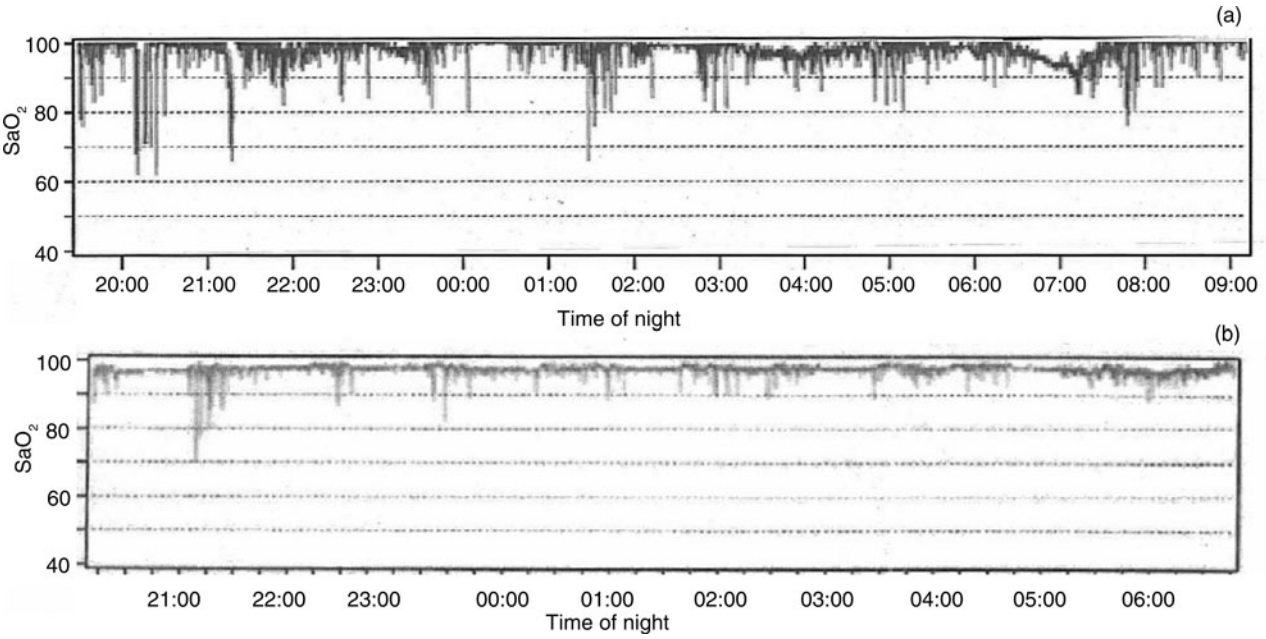

Fig. 2. Overnight oximetry (a) before and (b) after high-flow nasal cannula therapy at eight months' follow up. SaO_2 = oxygen saturation

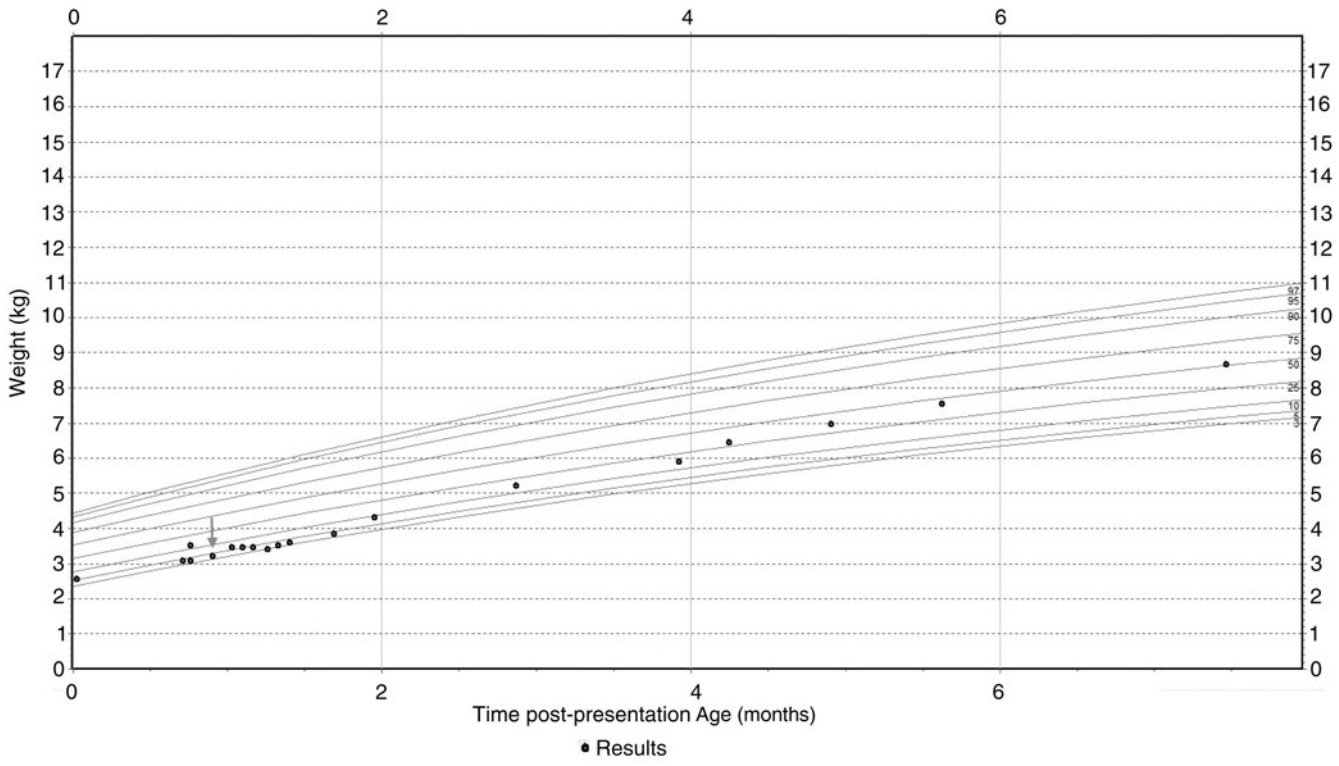

Fig. 3. Growth chart with arrow indicating commencement of high-flow nasal cannula therapy.

whilst providing a small degree of positive airway pressure.[11] High-flow nasal cannula therapy improves oxygenation, increases the end-expiratory lung volume, reduces airway resistance, increases functional residual capacity and alveolar recruitment, and flushes nasopharyngeal dead space, thus helping to decrease the work of breathing.[12] It has been shown to be well tolerated and safe to use in neonates.[13] A recent study demonstrated a reduction in the rates for requirement of ventilatory therapy escalation with high-flow nasal cannula treatment in neonates with bronchiolitis.[11]

One potential benefit of this treatment over classic nasal mask CPAP is a lack of pressure on the maxilla by the delivery mask, preventing the possible development of acquired midface hypoplasia and nasal trauma. Most recently, the use of high-flow nasal cannula therapy has been described for childhood obstructive sleep apnoea. This is especially valuable in children intolerant of nasal mask CPAP.[14–16] However, high-flow nasal cannula therapy is limited by a relative lack of evidence, and the high cost associated with consumables and acquisition of the device. Given its relatively new use, access to such machines is still limited; indeed, the parents in the above case needed to purchase the device privately.

In the case presented above, the infant experienced significant improvement in overnight breathing on long-term,

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home-based, high-flow nasal cannula therapy. This resulted in improvements in feeding, development and weight gain, allowing the patient to avoid invasive surgical management. The benefit seen is likely a result of improved delivery of humidified airflow past the anatomical obstruction. This case demonstrates the benefit of long-term high-flow nasal cannula treatment in avoiding surgery for a patient who would have otherwise been a surgical candidate given the degree of pyriform aperture stenosis and initial failure of traditional conservative therapies. With few adverse effects, long-term high-flow nasal cannula therapy may be an attractive new option to prevent invasive surgical management of pyriform aperture stenosis.

- Pyriform aperture stenosis can be a life-threatening cause of nasal obstruction in neonates
- Management options include conservative topical decongestant strategies and invasive operative procedures
- It poses a management dilemma for clinicians given the difficulty in predicting which patients will experience conservative therapy failure
- High-flow nasal cannula therapy has recently been demonstrated to be beneficial in neonates for obstructive sleep apnoea and severe bronchiolitis
- High-flow nasal cannula therapy may provide an additional conservative treatment option for patients with pyriform aperture stenosis

Conclusion

Pyriform aperture stenosis is a rare but potentially life-threatening condition for neonates. It poses a management challenge, with parents and clinicians alike aiming to undertake the least invasive approach possible. This case demonstrates the first reported successful use of non-invasive, long-term high-flow nasal cannula therapy, and suggests a potential additional treatment avenue for the conservative management of pyriform aperture stenosis.

Competing interests. None declared

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