Congenital nasal pyriform aperture stenosis

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Citation

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Abstract

Congenital nasal pyriform aperature stenosis is a rare congenital abnormality with a frequency less than a third that of congenital choanal stenosis. It typically presents at birth with features of upper airway obstruction and is frequently associated with congenital brain anomalies such as callosal dysgenesis. We present the first such documented case from the Caribbean.

INTRODUCTION

Congenital nasal pyriform aperture stenosis (CNPAS) is a rare congenital abnormality. This condition presents with upper airway obstruction and failure to pass an appropriate-sized feeding tube via the nostrils. It may be confused with choanal atresia which is much more common, however the diagnosis is usually clearly established by Computed Tomography. Surgical management may be necessary in severe cases but conservative management is often sufficient in milder cases. We present the first case reported from the West Indies.

CASE REPORT

A female baby was born to a 16-year-old HIV-negative primigravid woman with a non-reactive VDRL. The antenatal history was complicated only by the development of gestational diabetes. The full term, macrosomic baby was born by spontaneous vaginal delivery. Meconium stained liquor was observed at birth and the early post partum period was complicated by hypoglycaemic spells. She was noted at birth to have respiratory distress with obvious nasal obstruction. A size 4F feeding tube could not be passed through the left nostril. She was assessed as having possible left choanal atresia and meconium aspiration pneumonitis was transferred to a tertiary institution for joint ENT and Paediatric management.

On arrival at the specialist hospital, the neonate was in moderate respiratory distress with a respiratory rate of 76/min. She was acyanotic with mild stertor and tracheal tugging. There was no stridor and her oxygen saturation was 94% on 10L/min of oxygen via head box. Observed dysmorphic features included: brachycephaly, epicanthic folds, retrognathia, short fat legs and rocker bottom feet. There was a 13x5 cm soft swelling in the right parietal region extending slightly across the midline and down to the postauricular area. The spine, hips and external genitalia were normal. Anterior rhinoscopy revealed what appeared to be enlarged inferior turbinates. A size 8F feeding tube could not be passed via the nostrils but a size 6F tube was passed bilaterally. Placement of an oropharyngeal airway resulted in improved oxygen saturation to 96-98%. On cardiovascular examination the heart rate was 128/min with a grade 2/6 ejection systolic murmur heard best lower left sternal edge. The lung fields were clinically clear. Ophthalmologic assessment was normal and excluded the presence of colobomas.

INVESTIGATIONS

An echocardiogram done on the second day of life revealed a small patent ductus arteriosus (PDA) and a patent foramen ovale (PFO). Chest X ray was normal. Audiometric evaluation revealed low-peaked type A tympanograms bilaterally and she met the pass criteria bilaterally on otoacoustic emission testing. Abdominal ultrasound revealed no renal or other abnormalities.

An axial non-contrast CT scan of the paranasal sinuses was done at 3mm x 3mm covering the sinuses and facial bones. The scan was done on a GE Prospeed single slice CT scanner. No multidetector CT scanner was available at our institution. Direct coronal imaging was not attempted as the infant was intubated.

CT revealed narrowing of the anterior nasal cavity bilaterally. There was thickening of the nasal processes of the maxillae with subsequent narrowing of both pyriform apertures. The vomer was not significantly thickened but the

choanae were narrow. There was no evidence of a lacrimal duct cyst. The anterior hard palate was dysplastic with a triangular configuration. There was a midline inferior palatine ridge (Fig.1) and a solitary mega-incisor (Fig.2).

Figure 1

Fig.1 Arrow points to the midline inferior palatine ridge. Note the triangular configuration of the hard palate

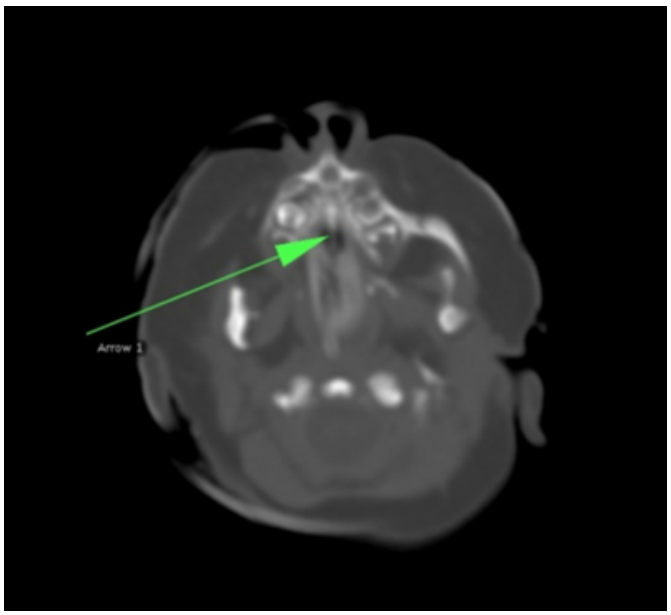
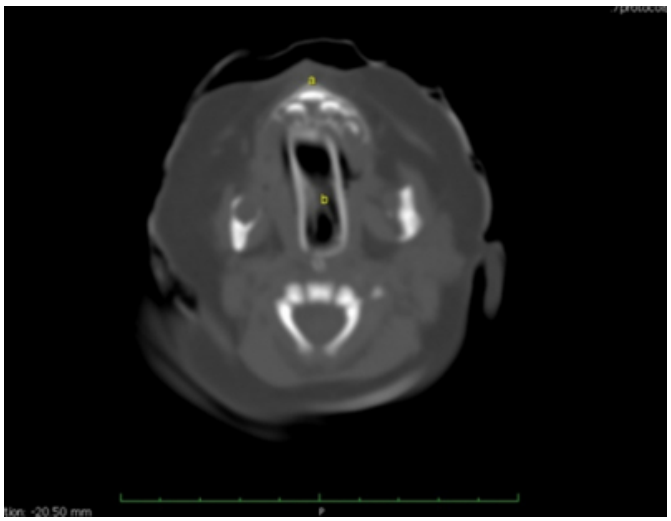


Figure 2

Fig. 2 a: Solitary megaincisor b: Oropharyngeal tube



The following measurements were noted (Fig.3):

Pyriform fossa width: 5.6mm

Interlaminar width (anterior): 5mm

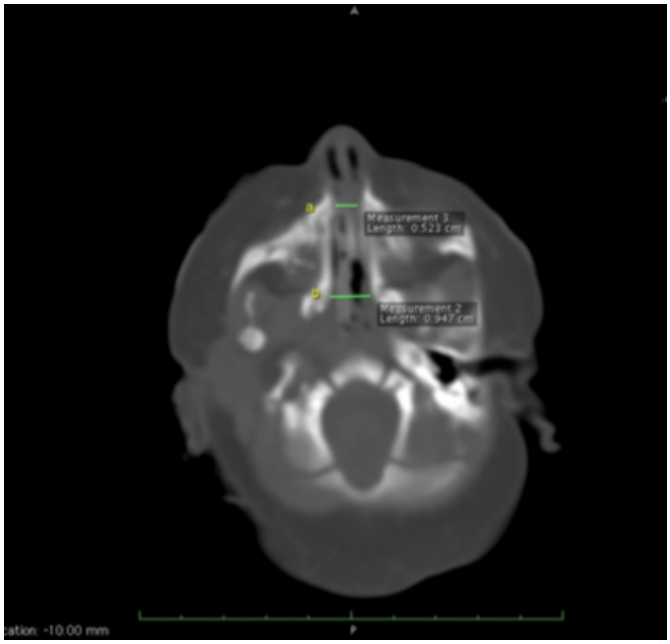
Interlaminar width (posterior): 4.6mm

Choanal width: 9.5mm

The appearances are in keeping with congenital nasal pyriform aperture stenosis (CNPAS).

Figure 3

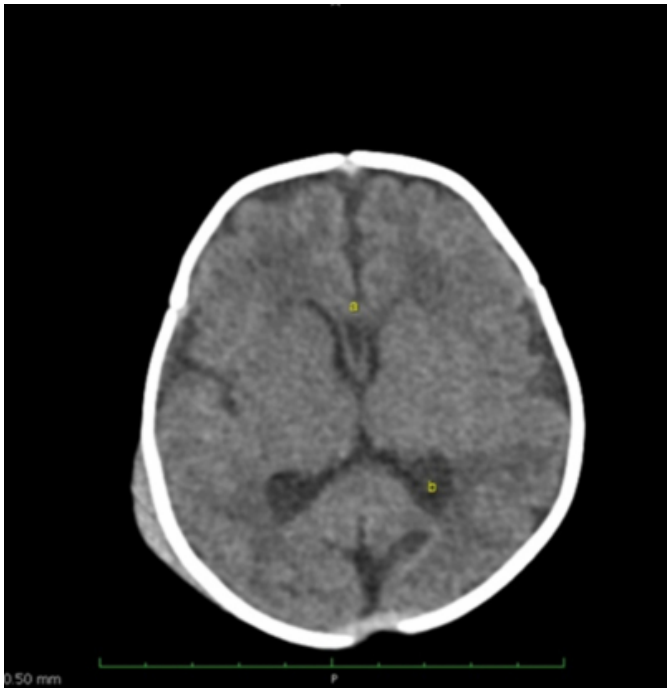
Fig. 3 a: Pyriform aperture measured at the level of the inferior meatus b: Measurement of the posterior choanae A nasogastric tube is in-situ



Given the findings, a CT brain was subsequently done to evaluate the brain for possible associated midline deformities. Images of the brain from CT revealed evidence of callosal dysgenesis (Fig.4.)

Figure 4

Fig.4 a: Widely placed anterior horns of the lateral horns with absence of the septum pellucidum b: Colpocephaly



An MRI was done at 5 months of age which confirmed the CT findings of dysgenesis of the splenium of the corpus callosum. No other abnormality was identified.

TREATMENT

On day 8 of life the nasal cavities were dilated and stented with portions of a size 3.5 endotracheal tube. The post operative recovery was complicated by cardiac failure which was treated with furosemide, aldactone and fluid restriction and she was discharged on the 6th post-operative day. At her second review in the 3rd postoperative week, the tubes were found to be blocked and partially dislodged. They were thus removed and excellent nasal airflow was noted. Two weeks later and again at five months old, the baby was still breathing well and there was no evidence of growth or other developmental delay. Repeat echocardiogram at 5 months of age failed to reveal the PDA and PFO.

DISCUSSION

Congenital nasal pyriform aperture stenosis is rare. While congenital airway obstruction occurs in approximately 1 in 5000 infants[2] the majority are due to choanal atresia. CNPAS has a frequency of less than a third of that for choanal atresia.

Patients typically present at birth with cyclic cyanosis and upper airway obstructive features including apnoeic spells. Presentation later in life may be precipitated by a respiratory tract infection with further compromise of the narrowed upper airway. There may be difficulty passing a nasogastric tube through the nasal inlet and symptoms may be exacerbated during feeding. These symptoms are non-specific and are seen in other causes of congenital upper airway obstruction such as choanal atresia. This condition may be associated with midline facial anomalies. Some authorities believe it is an isolated dysostosis of the pyriform aperture rim without any inter-orbital or midface hypoplasia[5]. Perhaps because this condition is so rare there have not been enough studies evaluating the pyriform aperture in patients with mid-facial anomalies. There is also an association with midline brain anomalies such as callosal dysgenesis/agenesis, holoprosencephaly and Dandy Walker malformations. Our patient demonstrated callosal dysgenesis but had no associated mid-facial anomalies.

This entity was first described clinically in 1989[4]. To our knowledge, this is the first case described in the literature from the West Indies. The aetiology is not known, but there are two main theories. The first suggests that the insult may arise during the formation of the primary palate in the 4th-5th weeks of embryogenesis. The second suggests that it is secondary to bony overgrowth of the nasal processes of the maxilla. Given the high frequency of abnormal incisural dentition, there is stronger support for the first theory.

The best imaging modality for investigating congenital upper airway obstruction is CT scan, which accurately diagnoses CNPAS and excludes other differential diagnoses. The features of CNPAS are well described in the Radiology and Otolaryngology literature[1,3,4,7,8]. Belden et al[2] is widely credited with publishing the first which attempted to fully describe the features of CNPAS. The anomaly is that of a narrowing of the inferior aspect of the entire nasal cavity, with the greatest narrowing being at the anterior and posterior aspects. A width of the pyriform aperture less than 11mm is diagnostic[2]. In our case, the width of the pyriform aperture was 5.2mm. The interlaminar distance (representing the width of the superior aspect of the nasal cavity) was described in that study as similar in patients with CNPAS compared to the controls. The interlaminar distance in our patient was within normal limits. The width of the choana in our patients was narrow which is in keeping with the findings in that study.

The management for mild cases is usually conservative with feeding difficulties overcome by a McGovern nipple. A

diameter of 5mm or more at the level of the inferior meatus on CT scan at birth may indicate a good chance of successful conservative management[6]. Our patient fell into this group and had a successful outcome with simple short-term stenting. Surgical management is required for severe symptomatic patients, which usually include those with a diameter of < 5mm, and in any patient not responding to conservative management. The preferred surgical approach is sublabial with a submucosal drill-out of the maxillary process followed by nasal stenting. Lee et al suggested that stenting for 7 days appear to be adequate. The authors also suggested the use of pre and post operative CT scans to assess surgical results however most authors assess results of surgical management based on follow up with patients being able to maintain an adequate airway[9]. Our patient was followed up for 5 months before defaulting, which is much shorter than we would have wished.

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