Congenital nasal pyriform aperture stenosis in a three-week old girl: a case report and discussion of current treatment strategies

Liselotte Franssen [a], Koen Kaïret [a], Nicolien van der Poel [b]

[a] Department of Pediatrics, AZ Voorkempen, 2390 Malle, Belgium
[b] University Department of ENT, University Hospital Antwerp, B-2650 Edegem, Belgium

Liselotte.franssen@student.uantwerpen.be

Keywords

Congenital nasal pyriform aperture stenosis, CNPAS, nasal obstruction, treatment

Abstract

Congenital nasal pyriform aperture stenosis (CNPAS) is a rare form of nasal obstruction in infants. It can cause respiratory distress and feeding difficulties, as neonates are obligate nasal breathers. The incidence of CPNAS seems to be underestimated. We report a case of CNPAS diagnosed in a three-weeks old neonate. She was initially started on conservative treatment based on topical nasal medication and humification, but without good clinical result. At the hospital heated humidified high-flow nasal cannula (HHHFNC) was started as a temporary treatment. After surgical treatment through sublabial approach, she could be weaned from high flow therapy.

Introduction

Nasal airway obstruction in a newborn can cause respiratory distress and feeding difficulties, as neonates are obligate nasal breathers (1-7). Congenital nasal pyriform aperture stenosis (CNPAS) is a form of nasal obstruction characterised by abnormal narrowing of the anterior inlet of the nasal cavity. It is a condition caused by overgrowth of the medial nasal process of the maxilla (1, 3, 4, 6, 8, 9). It can mimic the clinical findings of choanal atresia, which is the most common cause of neonatal nasal obstruction (1, 3, 6, 9). The incidence of CPNAS seems to be underestimated (7). We report the case of a three-week old girl with CPNAS.

Case report

A three-week old girl, born full term through normal vaginal delivery in a different hospital, presented at our hospital with nose congestion, laboured breathing and feeding difficulty. There were no clear apnoeas or cyanotic incidents. Clinical evaluation showed respiratory distress with inspiratory stridor and subcostal retractions. A notable finding was that we couldn't pass a nasopharyngeal swab. A capillary blood gas demonstrated respiratory acidosis without complete metabolic compensation. Heated humidified high-flow nasal cannula (HHHFNC) without additional oxygen was started. A capillary blood gas showed a clear improvement a day later.

Detailed history showed she had symptoms of obstructive breathing with subcostal and jugular retractions and inspiratory stridor starting shortly after birth. Conservative treatment with rinsing and oxymetazoline drops was initiated, but resulted in no clinical improvement. Upon follow-up consultation it was almost impossible to pass a nasopharyngeal swab. There was suspicion of a choanal atresia and the patient was referred to their ENT department. Initial tests showed there was air passage through both nostrils and no further plan of action was postulated. The patient came to our centre for a second opinion.

As stated above we started with HHHFNC in our hospital on day one. Because weaning from this therapy was not possible without the baby getting very uncomfortable, she was transferred to the ENT department at the University Hospital of Antwerp. Fibreoptic endoscopy showed a bilaterally narrow pyriform aperture and bilateral patent choanae. A computerized tomography (CT) scan confirmed a stenosis of the pyriform aperture with a maximum diameter of 5mm and a generally narrowed nasal cavity, as shown in Figure 1. There was no central mega incisor.

At the age 32 days the pyriform aperture was surgically dilated through a sublabial approach with a placement of stents. Unfortunately, the stents were afunctional because of blockage after one day and, therefore, removed. HHHFNC was restarted and the nose was rinsed and washed with budesonide nasal drops four times a day. She was successfully weaned from HHHFNC after seven days.

As CNPAS can be associated with midline defects, additional technical tests were requested. An echocardiogram showed no abnormalities. MRI of the brain showed no midline defects, but did reveal unrelated sequelae from an earlier birth-related subarachnoid haemorrhage.

Discussion

The exact incidence of CNPAS is unknown. It is said to be a rare form of nasal obstruction, but its frequency is probably underestimated since it has been diagnosed more frequently since it was first described in 1989 by Brown et. al (1). Symptoms include nasal congestion, respiratory distress with retractions, inspiratory stridor, episodes of apnoea and/or cyanosis, difficulty feeding and failure to thrive. These symptoms can occur straight after birth or after a few weeks and are often triggered by an upper respiratory infection which narrows the already compromised airway even further (2, 4-6, 8). Our patient already showed obstructive breathing with subcostal and jugular retractions and inspiratory stridor shortly after birth.

Diagnosis of CNPAS is suspected when the anterior nasal fossae are narrowed upon physical examination and/or when it is difficult to pass a five French catheter or a nasopharyngeal swab. The golden standard tool to establish the diagnosis is a CT scan. The CT scan would show stenosis at the bony nasal inlet. However, there is currently no consensus about the minimal dimensions for CNPAS, as literature describes the dimensions ranging from a width of less than eight millimetres to less than eleven millimetres (2, 4, 6, 8-10). In our case the pyriform aperture width was maximum five millimetres. This would be a definite diagnosis according to both cut off values.

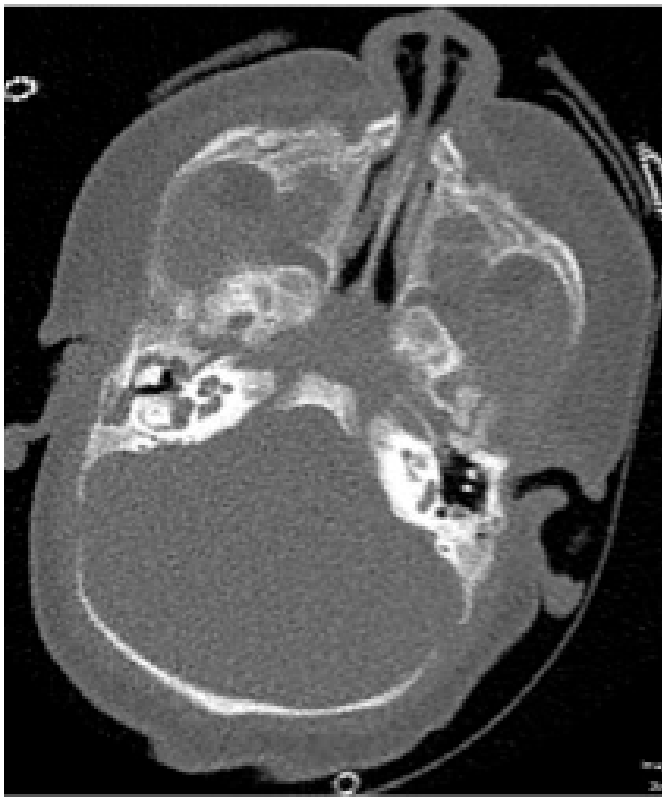
CNPAS can be an isolated phenomenon or it can occur together with other abnormalities. It is associated with midline anomalies, including central nervous system, endocrine and craniofacial abnormalities. Examples of associated anomalies are holoprosencephaly, facial haemangiomas, clinodactyly, pituitary dysfunction and a single central maxillary incisor (2-10).

It is also often associated with a mid-nasal stenosis. Patients with CNPAS should be evaluated to exclude midline anomalies because of this association, especially if there is a single central maxillary incisor, which is seen in up to 60% to 75% of cases (6, 9). Further evaluation includes chromosomal analysis, MRI of the brain and an echocardiogram (2-10).

The choice of treatment is based on the severity of the obstruction and clinical symptoms, as the width of the aperture cannot always predict the need for surgery (3, 9, 10). Initial treatment should include humidification, topical nasal decongestants and steroids. If patients can tolerate conservative management, it is continued until the craniofacial growth improves the nasal airway with time. When patients do not respond to medical treatment alone, they should undergo a surgical procedure. Indications for surgical treatment are respiratory distress or failure to thrive (2-7, 9). The classical surgical technique is widening of the pyriform aperture through a sublabial approach (1, 4, 5, 9, 10). In recent literature, a less invasive dilatation technique using a balloon has also been described. However, this technic carries a risk to the integrity of the nasal septum. (4, 8, 9). Our case was started on conservative management, but without enough improvement. At 32 days old she underwent surgery with the sublabial approach.

In 2020 Fuzi et al. (2) came with a high-flow nasal cannula treatment as a novel method of respiratory support in children with CPNAS. It delivers warmed and humidified gas with or without supplementary oxygen to the nasal airways, combined with some positive airway pressure. Treatment with high-flow nasal cannula decreases the work of breathing by improving oxygenation, increasing the end-expiratory lung volume and dilating the radius of the nasopharyngeal airway which reduces the airway resistance. It also increases the functional residual capacity and alveolar recruitment and flushes the nasopharyngeal dead space. Furthermore, it has been shown to be safe to use and well tolerated in neonates. Its use, however, has been limited by a relative lack of evidence and the high costs associated with the device. As it is still a relatively new method, access to the machines is also limited. In the case mentioned in Fuzi et al. the parents needed to purchase the device privately. In our case, HHHFNC treatment was used temporarily during the hospitalisation period.

Figure 1: CT Scan showing a narrow pyriform apertura of maximum 5mm with a globally narrow cavum nasi.



Conclusion

Congenital nasal pyriform aperture stenosis (CNPAS) is a rare form of nasal obstruction in infants, but it should be part of the differential diagnosis along with (unilateral) choanal atresia. The golden standard to establish the diagnosis is a CT scan revealing stenosis at the bony nasal inlet. As CNPAS has been associated with midline defects, additional evaluation should be performed. Conservative measures, such as nasal decongestants, may be sufficient to improve nasal patency and bridge the time until craniofacial growth improves the nasal airway. In more severe cases, surgical management is necessary. Heated humidified high-flow nasal cannula can be used as a temporary treatment in the hospital setting.

Consent

Written informed consent was obtained from the parents of the patient for publication of this article.

Conflicts of interest disclosure

There are no conflicts of interest.

REFERENCES:

1. Brown OE, Myer CM, 3rd, Manning SC. Congenital nasal pyriform aperture stenosis. The Laryngoscope. 1989;99(1):86-91.
2. Fuzi J, Teng A, Saddi V, Soma M. Novel use of high-flow nasal cannula therapy in the management of pyriform aperture stenosis: case report. The Journal of laryngology and otology. 2020;134(6):558-61.
3. Gonik NJ, Cheng J, Lesser M, Shikowitz MJ, Smith LP. Patient selection in congenital pyriform aperture stenosis repair - 14 year experience and systematic review of literature. International journal of pediatric otorhinolaryngology. 2015;79(2):235-9.
4. Serrano TL, Pfeilsticker L, Silva V, Hazboun I, Paschoal J, Maunsell R, et al. Newborn nasal obstruction due to congenital nasal pyriform aperture stenosis. Allergy & rhinology (Providence, RI). 2016;7(1):37-41.
5. Lee JJ, Bent JP, Ward RF. Congenital nasal pyriform aperture stenosis: non-surgical management and long-term analysis. International journal of pediatric otorhinolaryngology. 2001;60(2):167-71.
6. Sanal B, Demirhan N, Koplay M, Sadikoğlu MY, Gürpinar A. Congenital nasal pyriform aperture stenosis: clinical and radiologic findings and treatment. Japanese journal of radiology. 2009;27(9):389-91.
7. Van Den Abbeele T, Triglia JM, François M, Narcy P. Congenital nasal pyriform aperture stenosis: diagnosis and management of 20 cases. The Annals of otology, rhinology, and laryngology. 2001;110(1):70-5.
8. Gungor AA, Reiersen DA. Balloon dilatation for congenital nasal piriform aperture stenosis (CNPAS): a novel conservative technique. American journal of otolaryngology. 2014;35(3):439-42.
9. Moreddu E, Le Treut-Gay C, Triglia JM, Nicollas R. Congenital nasal pyriform aperture stenosis: Elaboration of a management algorithm from 25 years of experience. International journal of pediatric otorhinolaryngology. 2016;83:7-11.
10. Hui Y, Friedberg J, Crysdale WS. Congenital nasal pyriform aperture stenosis as a presenting feature of holoprosencephaly. International journal of pediatric otorhinolaryngology. 1995;31(2-3):263-74.