Congenital nasal pyriform aperture stenosis in newborn: report on three cases*

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SUMMARY

Congenital nasal pyriform aperture stenosis (CNPAS) is recognized as a cause of nasal airway obstruction in the newborn. The nasal pyriform aperture is narrowed by a bony overgrowth of the nasal process of the maxilla. The CNPAS may occur as an isolated congenital defect or in combination with other abnormalities. Three cases of CNPAS are reported with special attention to the clinical presentation and to the management recommendation prior to surgery. Surgical repair was performed for all these three highly symptomatic patients using a sublabial approach for drilling the nasal process in order to obtain a wider nasal vestibular patency. All of them were equipped with bilateral nasal stenting.

Key words: congenital nasal pyriform aperture stenosis, nasal stent, nasal obstruction, newborn abnormalities, obstructive sleep apnea, single megaincisor, surgical treatment

INTRODUCTION

Congenital nasal pyriform aperture stenosis (CNPAS) was first described in newborns by Brown et al., as a cause of nasal obstruction (Brown et al., 1989). The newborn is an obligate nasal breather and complete nasal obstruction may be a life-threatening event. Non-specific mucosal edema which is the most common cause of neonatal nasal obstruction, is almost always partial and well tolerated. Congenital bony nasal cavity deformities are mostly bilateral and almost complete or complete. CNPAS is secondary to a bony overgrowth of the nasal process of the maxilla, reducing the nasal airway at vestibular level. Diagnosis of CNPAS is often made at birth or in the first weeks of life.

Mildly symptomatic patients may be treated expectantly, while more severe cases may require to be operated in order to obtain a wider nasal airway. Management recommendations based on the patients clinical course, feeding problems and polysomnographic data are therefore very helpful before any operative decision.

CASE REPORT (1)

A male infant born by spontaneous vaginal delivery at 40 weeks of gestation (3180 g) began quickly to have nasal breathing difficulties and received oxygenotherapy. During the first two months of life, the patient had poor nasal respiration, feeding difficulties and apneic episodes. He was transferred to our institution at 10 weeks of life when his weight was 4900 g. Physical examination revealed facial dysmorphy with hypotelorism, down-setted ears and an orbital defect. ENT examination included macroglossia and congenital nasal pyriform aperture stenosis. Fiberoptic examination of the nose was not possibly performed due to a closed bony nasal valve. CT scan of the nose and paranasal sinuses revealed a bony overgrowth of the maxilla reducing the nasal pyriform aperture with a normal nasal cavity and a normal posterior choanae. CT scan of the central nervous system showed corpus callosum agenesis.

A polysomonographic study assessed the diagnosis of severe obstructive sleep apnea syndrome. Chromosome analysis was normal. We decided to set up a cardio-respiratory monitoring and to follow the clinical course of the patient. At 14 weeks of life, an apparent life-threatening event (ALTE) occurred and the feeding problem became more serious. Due to the history of apneic episodes, the ALTE and the feeding problem, operative repair was decided when the child was 16 weeks old and his weight 5200 g. A sublabial approach was performed and the nasal aperture stenosis was drilled away to obtain a wide opening of the nasal vestibule. Bilateral 3,5 mm inner diameter endotracheal tubes were placed in both nasal cavities and

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secured as nasal stents for 53 days. A postoperative polysomnographic study was performed showing the disappearance of the obstructive sleep apnea syndrome. In follow-up at the age of 9 months, the nasal airway remained patent bilaterally and no mega-incisor was observed. Unfortunately, the patient was lost out of sight at 9 months of age because he moved abroad.

CASE REPORT (2)

A male infant was delivered prematurely at 34,5 weeks of gestation (2180 g). At delivery, the newborn had serious difficulties to breathe with subcostal retraction. Oxygenotherapy and placement of a Guedel canula were decided. Soft catheters could not pass through the nasal cavities. Fiberoptic examination revealed a congenital nasal pyriform aperture stenosis. CT scan of the nose and of the paranasal sinuses assessed the diagnosis (Figure 1). Physical examination did not show facial dysmorphy nor other malformations. A polysomnographic study revealed a severe obstructive sleep apnea syndrome. Chromosomal analysis was normal. The newborn was followed up during 16 weeks until feeding problems and failure to thrive urged the surgical repair.

A sublabial approach was performed at 16 weeks of life when his weight was 5260g, drilling away the pyriform aperture stenosis. Endotracheal tube nasal stents 3 mm inner diameter were secured bilaterally during 40 days. A second postoperative polysomnographic study revealed the disappearance of the OSAS. At 6 months follow-up the infant showed a normal nasal airway, no facial dysmorphy and no mega-incisor.

CASE REPORT (3)

A 4-days-old female was evaluated for heavy difficulties in breathing. At birth, respiratory distress needed an oral intubation. Nasal examination revealed a bilateral congenital nasal pyriform aperture stenosis and surgical repair was decided at day 7 using a sublabial approach and a bilateral stenting with an endotracheal tube 3,5 mm inner diameter during 17 days. After airway management, many congenital anomalies were diagnosed which include: central blindness, neural deafness, genital abnormalities, gastro-esophageal reflux, urinary tract abnormalities and central nervous system malformations. Chromosomal analysis demonstrated trisomy 18. At one year follow-up, the nasal airways were patent and oral examination showed a single median incisor.

DISCUSSION

Congenital bony nasal cavity deformities are infrequent but life-threatening causes of nasal obstruction in newborn. Clinical observation and computerized tomography scanning reveals three forms of deformities: posterior choanal atresia, nasal cavity stenosis and nasal pyriform aperture stenosis.

Posterior choanal atresia and nasal cavity stenosis are respectively located at the posterior area and at the middle area of the nasal fossa. Congenital nasal pyriform aperture stenosis (CNPAS) is located at the anterior part of the nasal fossa where small changes in the cross-sectional area of the nasal airway can result in significant increases in nasal airway resistance.

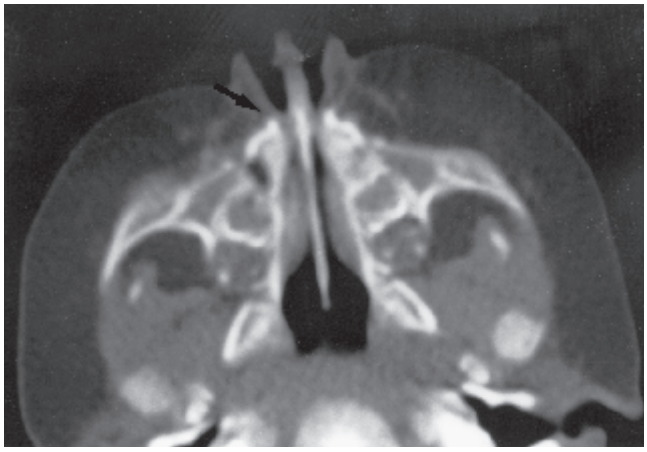

Figure 1. Axial CT Scan revealing pyriform aperture stenosis (Case #2) (Arrow).

Table 1. Choanal atresia (CA) and congenital nasal pyriform aperture stenosis (CNPAS).

CA	**CNPAS**
Posterior area of the nasal fossa	Anterior area of the nasal fossa
Osseous, membranous or both	Osseous
Mostly bilateral	Mostly bilateral
Complete nasal obstruction	Almost complete nasal obstruction
Mostly revealed in the first hour of life when bilateral.	Mostly revealed in the first weeks of life

The nasal airway resistance is increased in CNPAS like in choanal atresia. But it seems that CNPAS is less life-threatening than choanal atresia because the nasal obstruction is almost complete in CNPAS and complete in choanal atresia. Posterior choanal atresia can demonstrate osseous, membranous or both stenosis. Unlike choanal atresia, CNPAS is always osseous. CNPAS is due to an increased bony overgrowth of the nasal process of the maxilla and is always bilateral except of one case described by Tavin (Tavin et al., 1994). Symptoms are very similar to those observed in bilateral choanal atresia and consist in nasal obstruction worsened by feeding and relieved by crying. This nasal obstruction is observed within the first hours or in the first weeks of life. Whereas nasal obstruction due to bilateral choanal atresia is immediately revealed after birth (Table 1).

It has been postulated that CNPAS is due to a overgrowth of maxillary ossification at the area of the nasal process of the maxilla and that this anomaly occurred at 4 months of fetal development (Brown et al., 1989). Rhinoscopic examination reveals narrowed nasal inlet bilaterally. A flexible catheter cannot be passed through the nasal passage and fiberoptic nasopharyngoscopy is stopped by the anterior pyriform aperture stenosis. Computerized tomography confirms the diagnosis making clearly the difference from other congenital bony nasal cavity deformities. Nasal cavity stenosis in CNPAS is located anteriorly with a normal middle segment of the nasal cavity and

a wide posterior choanal opening (Castillo, 1994) (Figure 1). To our knowledge, CNPAS together with choanal atresia has never been demonstrated in the same patient. We advocate the need to perform a polysomnographic study after the CNPAS diagnosis and after the upper airway management. This study allows the clinician an objective measurement of apnea-hypopnea index of nocturnal oxymetry and precise the neurologic status.
Feeding problems and failure to thrive can be discussed with the polysomnographic results between pediatricians and ENT surgeons before any operative decision. Therefore selective criteria and therapeutic guidelines can be proposed. For a mildly affected patient, expectative with transient use of Guedel nipple is indicated. When feeding problems occur, transient feeding with oro-gastric tube may be suitable for a while. For a severely affected patient, surgical repair is indicated at earlier times (Table 2).
Surgery must attempt to obtain a wide pyriform aperture which allows normal nasal breathing. Superior gingivo-labial incision is performed using a sublabial approach. To our opinion, a transnasal approach is not suitable because of an increased risk of secondary local scars leading to fibrous stenosis of the nostril, and also because of less room in the operating field. The pyriform aperture is identified and the nasal mucosa is elevated from the nasal floor. Medial bony overgrowth of the maxilla is demonstrated. The bony obstruction (nasal process of the maxilla) is enlarged by drilling the pyriform aperture backwards till the naso-lacrimal duct and the head of the inferior turbinate. After drilling, the passage of a 3,5 mm inner diameter endotracheal tube in the nasal fossa must be possible. Bilateral stenting is set in place during at least 1 to 4 weeks and then removed under sedation. Care must be taken not to damage deciduous teeth nor the naso-lacrimal duct. Further midfacial an nasal development hypoplasia must be kept in mind though it had never been demonstrated in operated CNPAS patients (Arlis et al., 1992; Brown et al., 1989; Vanden Abbeele et al., 1997). Results of surgical repair are excellent with an improvement of nocturnal oxymetry, of feeding problems and of the patients clinical course. Long term results of surgery seem to be stable and no repetitive procedure was described in the reported cases (Arlis et al., 1992; Brown et al., 1989; Vanden Abbeele et al., 1997). Surgical repair is not indicated for all the CNPAS patients. Brown et al., reported on 6 patients with CNPAS, 5 of whom were operated (Brown et al., 1989). Arlis et al., only operated 2 of the 6 patients with CNPAS (Arlis et al., 1992). Recently Jones et al., reported one case of CNPAS who was not operated (Jones et al., 1998). Our 3 reported patients were severely affected and therefore surgical repair was decided, in regard to: inability to be weaned from the Guedel canula, heavy feeding problems and/or failure to thrive, apparent life threatening event, full nasal obstruction and severe nocturnal oxymetry desaturation. We should recommend to intervene when the infant is 4-6 weeks of age and when the patients clinical course gets no improvement. But earlier surgical repair may be imperious such as in case 3. After successful airway management and surgical wide opening, newborn with CNPAS must be screened for others malformations. In association with CNPAS, single prominent central maxillary incisor has been frequently reported (Arlis et al., 1992, Vanden Abbeele et al., 1997) as the most often encountered developmental anomaly (Figure 2). This anomaly is defined by some authors as a microform of holoprosencephaly which is a developmental field defect (Arlis et al., 1992). When CNPAS is associated with a single central incisor, a microform of holoprosencephaly must be ruled out or confirmed by chromosomal analysis, computerized tomography-scan for central nervous system malformations and assessment of the hypothalamic-pituitary-thyroid-adrenal axis (Arlis et al., 1992). Nevertheless CNPAS may occur as an isolated defect and described as a non specific feature. Similarly single prominent central maxillary incisor may be isolated or associated with other abnormalities (Arlis et al., 1992; Hui Y et al., 1995; Tavin et al., 1994).
CNPAS may be considered as a rare cause of nasal obstruction in the newborn but a more frequent cause considering the congenital bony nasal cavities deformities. Before any operative decision, a polysomnographic study and a critical study of the patient's clinical course are mandatory. Unlike in choanal atresia, surgical repair in CNPAS seems to be associated with good results and with a single step procedure including drilling of the stenosis and stenting of the nasal fossa. The clinical management of the newborn with nasal respiratory distress is based on proper identification of the anatomic abnormality. When CNPAS is considered, early screening for associated abnormalities and central maxillary incisor observation must be kept in mind.

Table 2. Selection criteria for a surgical repair.

– Nasal obstruction
– Failure to thrive
– Feeding problems
– Apparent life threatening event (ALTE)
– Severe obstructive sleep apnea syndrome at polysomnographic study

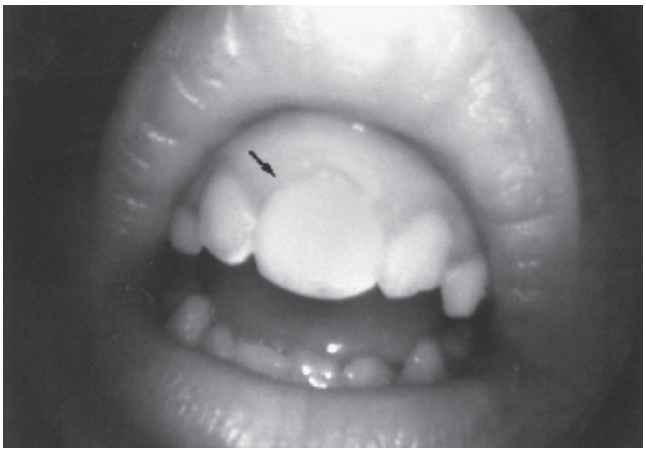

Figure 2. Single prominent central maxillar incisor (Case #3) (Arrow)

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