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Congenital nasal pyriform aperture stenosis: Elaboration of a management algorithm from 25 years of experience

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ABSTRACT

Introduction: Congenital nasal pyriform aperture stenosis (CNPAS) is a rare disease presenting with neonatal respiratory distress, often associated with other anomalies.
Materials and methods: This study reports the clinical and radiological characteristics of the patients managed in The Department of Pediatric Otolaryngology Head and Neck Surgery of La Timone Children's Hospital in Marseille between 1988 and 2014. Pyriform aperture (PA) widths were measured on CT-scans, obtained by using hand calipers at the largest portion of the PA in a plan parallel to the Francfort plan.
Results: 10 patients were included. Average PA width was 6.6 mm, 5/10 patients presented with single central maxillary median incisor, 6/10 patients had associated abnormalities. 8 patients underwent a surgical intervention and 2 patients were medically managed. All the patients had satisfactory nasal airway permeability on late follow-up.

A management algorithm was elaborated. CNPAS should be evoked when breathing difficulties are associated with impossibility of passing fiberscope or nasogastric tube at the nasal inlet. Craniofacial CT-scanning is necessary to make the diagnosis and look for associated abnormalities. Medical treatment associating nasal wash and decongestants should be performed. Surgical intervention is necessary when failure of the medical management.
Discussion and conclusions: Our results were close to those found in the literature in terms of clinical characteristics, associated abnormalities and PA width. However, no objective criterion to decide whether a surgical intervention is necessary or not, has been established so far. The algorithm we propose offers guidelines from diagnosis to treatment, but the management should be adapted based on clinical tolerance.

1. Introduction

Congenital nasal pyriform aperture stenosis (CNPAS) is a rare cause of neonatal airway obstruction leading to respiratory distress. It was identified for the first time in 1952 by Douglas et al. [1], and later radiologically described in 1988 by Ey et al. [2]. However, the first clinical description was published by Brown et al. [3] in 1989.

Few publications in the literature have reported both clinical and radiological characteristics with anatomical measurements on patients with CNPAS. Belden et al. [4] estimated that the lowest pyriform aperture width is about 11 mm on a normal term-born neonatal CT-scan. Although this value is not consensual, all CNPAS patients found in the literature had a PA width measurement under this threshold.

CNPAS can occur as an isolated anomaly or as part of holoprosencephaly spectrum including solitary median maxillary central incisor and other midline anomalies.

Because CNPAS is a rare anomaly, it is mainly diagnosed in specialized pediatric otolaryngology centers. Most of the recent studies focus only on one of the aspects of this craniofacial abnormality, with particular emphasis on trying to find objective criteria for diagnosis and surgical management. However, no clear guidelines have been proposed.

The objective of this study was to present a complete management algorithm for the diagnosis of CNPAS, as well as

Abbreviations: CNPAS, congenital nasal pyriform aperture stenosis; PA, pyriform aperture; SMMCI, single median maxillary central incisor.

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the medical and surgical management. This algorithm was created based on the experience of a tertiary care center pediatric otolaryngology center between 1988 and 2014, reporting clinical and radiological characteristics, management and outcome of patients presenting with CNPAS.

2. Materials and methods

This is a review of all patients presenting with CNPAS and referred to The Department of Pediatric Otolaryngology Head and Neck Surgery of La Timone Children's Hospital in Marseille between 1988 and 2014. Charts were reviewed for details including patient demographics, radiological characteristics, coexisting anomalies, medical management, and, when necessary, surgical treatment and outcome.

CNPAS diagnosis was suspected based on clinical features of airway obstruction: difficult breathing, poor feeding, nasal congestion, episodes of apnea/cyanosis associated with resistance or impossibility felt whilst passing a nasogastric tube or a fibroscope in either nostril. All children underwent craniofacial CT-scan to confirm the diagnosis. Linear measurements were obtained by using hand calipers at the largest portion of the PA in a plan parallel to the Francfort plan (Fig. 1) on CT-scans.

Medical treatment with nasal saline and decongestants (4 drops by nostril of 10% adrenaline saline) was first performed. Persistent airway obstruction symptoms despite this treatment were an indication for surgical intervention.

All operated patients underwent the same surgical procedure. Pyriform aperture was enlarged using a sublabial approach, allowing the elevation of the mucosa and the exposition of the frontal processes of the maxillary bones responsible for the stenosis. Submucosal drilling was then performed from the edge of the pyriform aperture to the head of the inferior turbinate. Stenting of the nasal fossae was done using Portex 3.0 "blue line" endotracheal tubes for a maximum of four weeks. Post-operative stent care consisted if normal saline nasal wash and nasal decongestion using 4 drops of 10% adrenalin saline mixture per nostril 3 times a day.

3. Results

A total of 10 patients were diagnosed with CNPAS. 7 patients were female and 3 were male (Table 1). Three patients were born prematurely, with the lowest gestational age being 35 weeks LMP. Mean birth weight was 2.900 kg and mean term of pregnancy was 38.2 weeks LMP (Table 2). All patients were symptomatic at birth: 8 of the 10 patients presented with respiratory distress necessitating immediate management, and 2 patients presented with noisy breathing. Radiological diagnosis with CT-scanning was performed at a mean age of 22.7 days and a median age of 10 days of life.

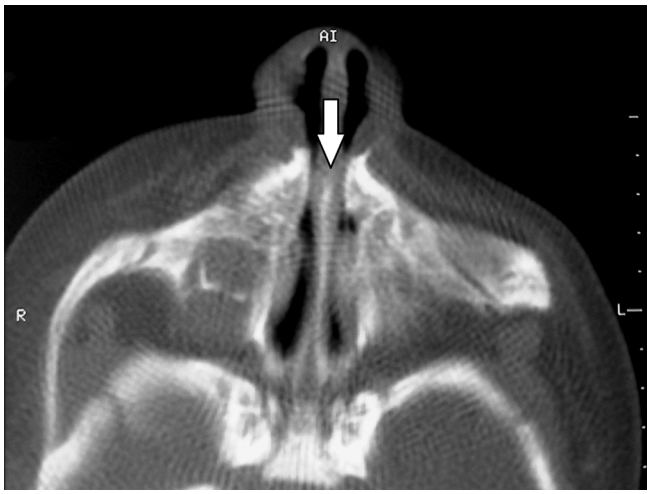

Fig. 1. CT-scan slice highlighting pyriform aperture stenosis (white arrow).

PA widths could be measured on 8 of the 10 patients CT-scans, which was noted to be between 5 and 9 mm (mean: 6.6 mm, median: 6.0 mm), while images were missing in two charts (Table 1). For the 2 patients without images, a copy of the CT-scan report attesting the CNPAS was present in the charts. Six of the 10 patients (Table 1) presented with associated anomalies, which consisted in solitary median maxillary central incisor (5/10) (Fig. 2), unilateral coloboma (1/10), Apert syndrome (1/10) and unilateral cophosis (1/10).

8 patients underwent surgery after failure of medical management. All patients were followed up for a mean period of 55 months (12–193 months), and all patients were found to have satisfactory nasal airway permeability. Two patients underwent adenoidectomy and tonsillectomy for sleep apnea after 3 years of age.

To summarize the data from our experience, we propose a management algorithm of CNPAS (Fig. 3). When CNPAS is suspected due to difficulty in passing a nasogastric tube or a fiberscope at the inlet of the nasal cavity with associated breathing difficulties, craniofacial CT-scanning is the gold-standard radiological imaging, which allows confirmation of the diagnosis (PA width inferior to 11 mm at birth) and evaluation of the other underlying abnormalities. Once confirmed, medical treatment is necessary up to 2 weeks, including nasal washing with saline and decongestant drops. Surgery is only needed in cases of medical

Table 1
Clinical and radiological characteristics of CNPAS patients (NA, non available). 7 patients were female, 3 were male. 5/10 presented with central megaincisor and 4/10 with other associated anomalies. 8/10 underwent surgical intervention.

	Gender	Pyrifom aperture width (mm)	Central megaincisor	Other associated abnormalities	Surgery	Age at surgery (days)
1	Male	5.7	No	No	Yes	9
2	Female	6	No	No	Yes	4
3	Female	7	Yes	No	Yes	19
4	Male	NA	Yes	Right Coloboma	Yes	10
5	Female	5	No	No	Yes	86
6	Female	6	No	No, but history of familial holoprosencephaly	Yes	31
7	Female	NA	Yes	No	Yes	4
8	Male	8	Yes	Apert Syndrome, Polysyndactyly Vestibular Anomalies	Yes	342
9	Female	6	No	Left-sided cophosis	No	NA
10	Female	9	Yes	No	No	NA