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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.

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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.

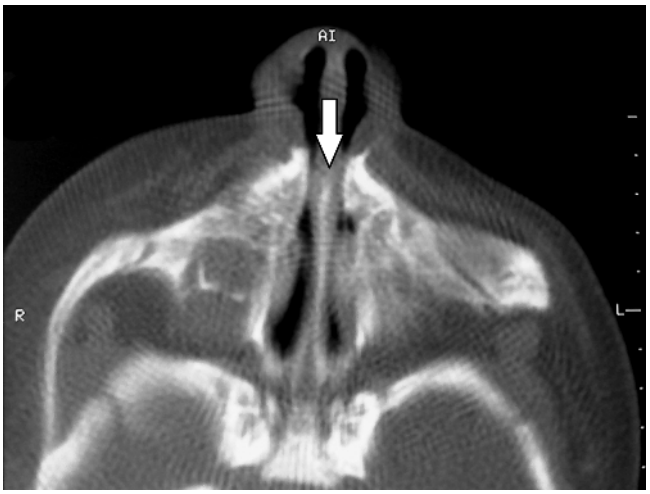

Fig. 1. CT-scan slice highlighting pyriform aperture stenosis (white arrow).

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

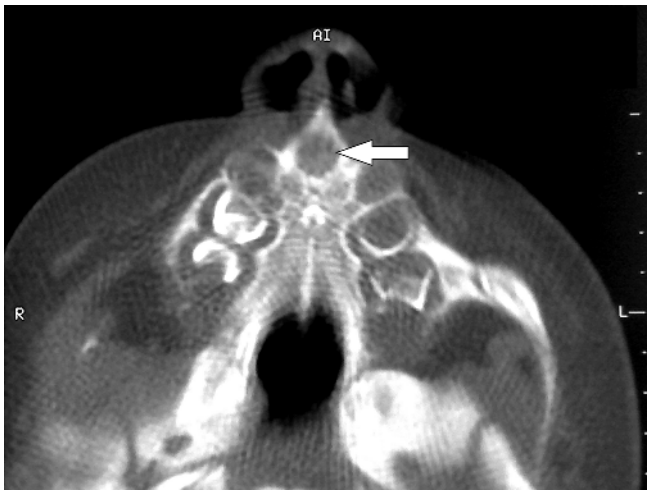

Fig. 2. CT-scan slice highlighting solitary median maxillary central incisor (white arrow).

treatment failure. Depending on the severity of breathing difficulties, evaluation of associated abnormalities by endocrine testing, cranial MRI, ophthalmological examination and cytogenetic testing can be performed prior or after surgery.

Table 2
Demographic and radiologic parameters of CNPAS patients.

	n	Median	Mean	SD	Min	Max
Birth weight (g)	7	2980	2900	722.96	1740	4100
Term (weeks)	8	39	38.2	2.07	35	40
Age at first symptoms (days)	10	0	7.5	19.0	0	60
Age at diagnosis (days)	10	10	22.7	29.7	0	76
Pyriform aperture width (mm)	8	6	6.6	1.3	5	9

4. Discussion

Newborns and infants are obligate nasal breathers; oral ventilation appears between 3 and 6 months of age. Any nasal obstruction during this period can cause respiratory distress, life-threatening dyspnea and eventually a failure to thrive. Congenital nasal airway obstruction occurs in up to 1 in 5000 infants, and most of them are affected with choanal atresia. Nevertheless, other anomalies can occur, such as major septal deviations, nasal cavity masses (teratomas [5] and dacryocystoceles [6]) or CNPAS. CNPAS is a very rare entity and all series in the literature include very few subjects.

Pyriform aperture is bordered superiorly by the nasal bones, inferiorly by the junction of the horizontal process of the maxilla

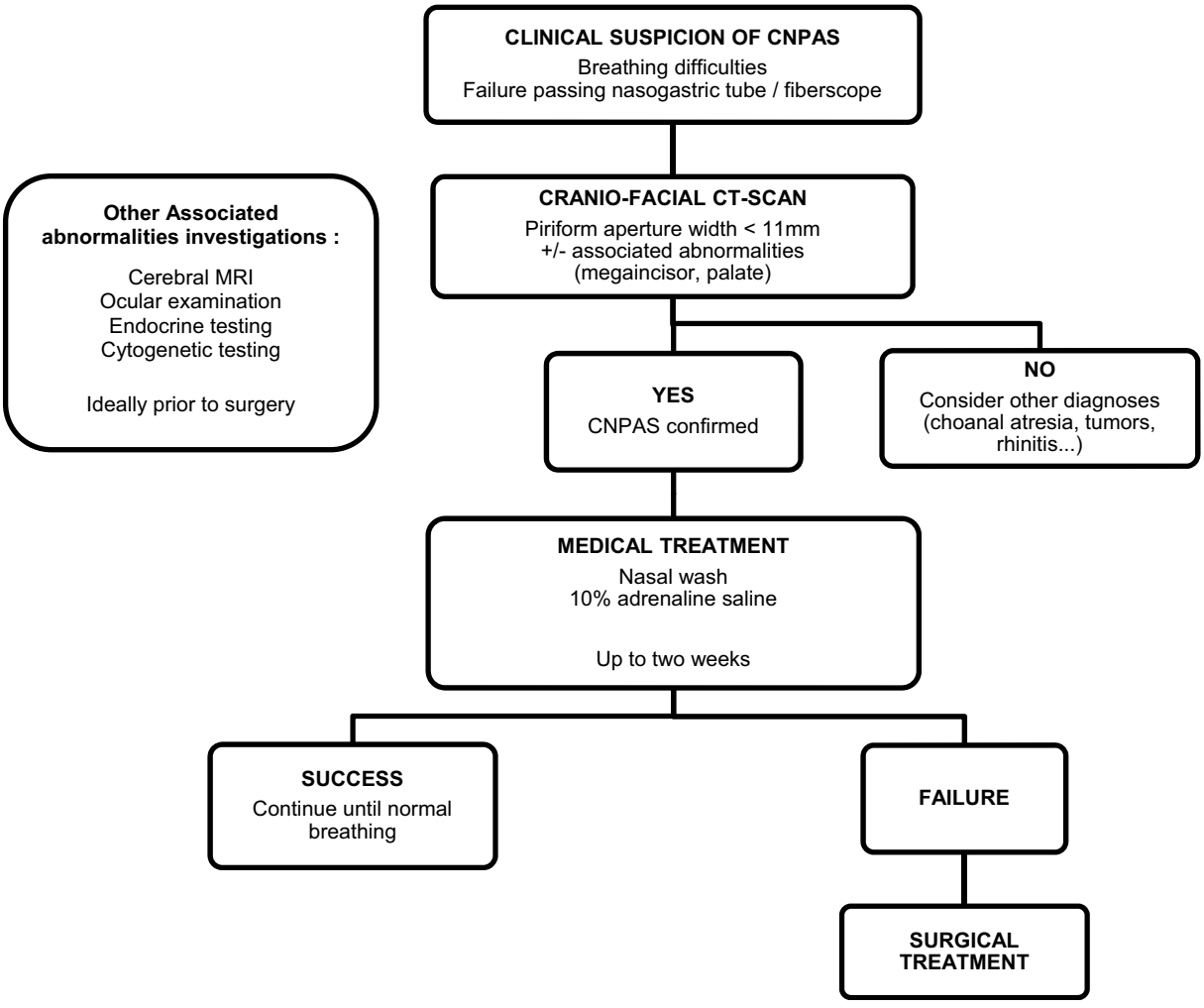

Fig. 3. Management algorithm of congenital nasal pyriform aperture stenosis (CNPAS).

and laterally by the nasal process of the maxilla. CNPAS is characterized by narrowing of the pyriform aperture secondary to bony overgrowth of the nasal process of the maxilla [4,7]. It can be suspected by the finding of very narrow nasal inlet preventing from the introduction of fiberscope or nasal tube. The difference with choanal atresia is in the location of the narrowing: immediately at the entry of the nasal cavity in CNPAS (first centimeter) and more posterior in choanal atresia (around 3 cm). CT-scan criteria for CNPAS include: (1) pyriform aperture width inferior to 11 mm in a term neonate according to Belden et al. [4], (2) triangular shape of the palate, (3) bony overgrowth of nasal process of the maxilla and (4) dental abnormalities such as solitary median megaincisor.

Few series in the literature include anatomical measurements of PA width on CT-scans (Table 3). In our series, mean pyriform aperture width was measured at 6.6 mm, Visvanathan and Wynne [8] reported a mean width of 5.89 mm, Belden et al. [4] of 4.8 mm, Wormald et al. [9] of 5.01 mm, Gonik et al. [10] of 5.49 mm. In all the series, pathological PA widths were found much lower than the 11 mm diagnostic threshold according to Belden et al., which was the lowest PA width obtained from a control group of asymptomatic neonates. However, Belden's series included a small cohort size (3 patients with CNPAS and 10 controls), and does not correlate PA width with clinical symptoms, making its diagnostic utility questionable.

CNPAS may be associated with, or ever considered as a minor form of holoprosencephaly. Holoprosencephaly spectrum includes multiple midline abnormalities. Cerebral abnormalities can include microcephaly, hypoplasy of the corpus callosum or olfactory bulbs and hypopituitarism. Facial anomalies include hypotelorism, ocular abnormalities, nasal defects, cleft lip/palate and dental abnormalities. Ideally, investigations should be performed prior to surgery, but are sometimes delayed when breathing difficulties require immediate surgical intervention. Investigations include facial CT-scanning, cerebral MRI, endocrine testing, ocular examination and cytogenetic testing. Single maxillary central incisor is diagnosed in about 60% of CNPAS cases in the literature (Table 3): Arlis and Ward [11] diagnosed 4 of 6 children (66%) with SMMCI, Lo et al. [12] 15 of 24 children (63%), Devambez et al. [13] 6 of 21 children (29%), Van den Abbeele et al. [14] 12 of 20 children (60%), Visvanathan and Wynne [8] 5 of 10 children (50%), and Belden et al. [4] 4 of 6 patients (66%). CNPAS seems to be predominant in females. In our series, 7/10 patients were female, 8/10 patients in Visvanathan and Wynne [8] series, but Van den Abbeele et al. [14] reported 10 males and 10 females. Only Brown et al. [3] reported CNPAS as an isolated event in 5 of 6 patients.

The occurrence of pituitary dysfunction is frequent and seems to vary between 15% [15] and 22.5% [16]. After initial blood testing, a clinical pituitary dysfunction monitoring for at least one year is recommended. Low or falling height at one year is a good predictor of pituitary dysfunction [15]. Repeated investigations should be required for patients who have abnormal hypothalamo-pituitary axis on MRI with normal blood testing or for patients who develop clinical signs of endocrine dysfunction [16].

Once diagnosed, treatment is either conservative or surgical depending on the severity of symptoms and essentially how the neonate or infant tolerates his nasal obstruction. Medical treatment includes nasal steroids and topical decongestants (10% adrenaline saline). For some authors [14], medical treatment should be performed up to 2 weeks before considering surgery. Failure of medical treatment includes symptoms such as dyspnea, inability to wean from assisted airways, sleep apnea or failure to thrive.

When sufficient, medical treatment should be continued until the craniofacial growth allows normal breathing. There is no unified agreement concerning the modalities for stopping the treatment. In our series, the two medically treated patients had normal breathing after 37 and 94 months of follow-up.

Anticipating the patients who are more likely to undergo surgery is one of the major concerns. Wormald et al. [9] included all patients (38 cases) with PA width measurement in the literature. Their results indicate that 88% of patients with PA width of 5.7 mm or less undergo surgery, versus only 38% of the patients with a PA width superior to 5.7 mm. Yet, the average PA width difference between the two groups (surgery versus conservative treatment) was only 1.3 mm. Presently, given these results, the PA width cannot be used as a decisional criterion.

Surgical treatment is classically performed with a sublabial approach allowing lateral drilling of the nasal process of the maxilla and followed by nasal stenting. Van den Abbeele et al. [14] suggested not exceeding 15 days of stenting, but two of the patients of his series had more than 20 days of stenting without any complications and good final results. Some authors [17] proposed balloon dilatation of the pyriform aperture using the natural plasticity of facial bones and cartilages.

Surgical treatment gives good results. As the dimensions of the nasal cavity follow the craniofacial growth, CNPAS is a neonatal abnormality that tends to improve with growth, and that has an excellent long-term respiratory prognosis. When the surgical intervention is initially sufficient, no secondary recurrence has ever been described in the literature.

Gonik et al. [10] found 9 surgical treatment failures on 63 patients (14%) who underwent surgery. In this study, "failure was determined by the need for additional airway surgery to bypass or further open the nasal airway". Associated craniofacial dysmorphism was the main factor limiting the success of surgery (OR 9.7, $p = 0.013$).

The management algorithm we propose (Fig. 3) is based on our experience and also summarizes the consensual attitude described in the different publications cited above. It is simplified to be easily available to physicians that do not have specialization in pediatric airway and may be faced with neonatal respiratory distress.

Table 3
Summary of the results of the main CNPAS series. PA, pyriform aperture. SMMCI, single median maxillary central incisor.

Authors	Number of patients	Average PA width (mm)	SMMCI
Arlis et al. [11]	6	–	4 (66%)
Belden et al. [4]	6	4.8	4 (66%)
Devambez et al. [13]	21	–	6 (29%)
Gonik et al. [10]	16	5.49	7 (44%)
Lo et al. [12]	24	–	15 (63%)
Van den Abbele et al. [14]	20	–	12 (60%)
Visvanathan et al. [8]	10	5.89	5 (50%)
Wormald et al. [9]	7	5.01	1 (14%)
Present work	10	6.6 (on 8 patients)	5 (50%)

5. Conclusions

Prompt diagnosis and management of CNPAS is necessary and include complimentary examinations due to the high prevalence of associated abnormalities. Diagnosis is made by CT-scanning with pyriform aperture width narrowing usually under 11 mm. Medical treatment should always be performed, and may be associated with surgical intervention when needed. Both medical and surgical management offer good long-term prognosis. The algorithm we propose provides guidelines for all physicians faced with CNPAS. However, in the absence of clear objective criteria, timing and decision-making for a surgical intervention should be based on clinical tolerance.

Conflict of interest

The authors confirm that they have no conflict of interest.

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