Congenital Nasal Pyriform Aperture Stenosis
Is There a Role for Nasal Dilation?

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Infants are obligate nasal breathers, and any degree of nasal airway obstruction can cause significant aerodigestive symptoms. Whereas soft-tissue edema is the most common cause of bilateral nasal obstruction, the differential includes a list of rare diagnoses, including bilateral choanal atresia, midnasal stenosis (congenital bony nasal stenosis), and congenital nasal pyriform aperture stenosis (CNPAS).

The sentinel descriptions of CNPAS occurred in 1988 and 1989 in the radiology and otolaryngology literature, respectively.1,2 Congenital nasal pyriform aperture stenosis is defined as a narrowing of the bilateral nasal cavity at the level of the pyriform aperture due to medial positioning or overgrowth of the maxillary process. In the past 2 decades, numerous case reports have been published regarding the presentation, diagnosis, and treatment of this clinical entity.3-7

The diagnosis of CNPAS must be considered in any infant with bilateral nasal obstruction. The symptoms can include respiratory distress relieved by crying, apnea, nasal congestion, difficulty feeding, and failure to thrive. Physical examination is most notable for being unable to pass a 2.2-mm flexible laryngoscope through either nasal cavity. A maxillofacial computed tomography scan is required to make the diagnosis of CNPAS. The diagnostic criterion for CNPAS requires the pyriform aperture width to be less than 11 mm.3 Commonly, a median central incisor, a triangular-shaped palate, and a median palatal ridge are identified on a computed tomography scan.

Congenital nasal pyriform aperture stenosis associated with a median central incisor may be part of the holoprosencephaly spectrum.8 As such, magnetic resonance imaging of

IMPORTANCE Congenital nasal pyriform aperture stenosis (CNPAS) may require sublabial drill-out of the pyriform aperture when symptoms are severe or refractory to medical therapy. Less invasive nasal dilation decreases potential morbidity to neonates with severe CNPAS.

OBJECTIVE To determine the outcome of patients with CNPAS who underwent nasal dilation alone without other surgical therapy.

DESIGN, SETTING, AND PARTICIPANTS A retrospective case series at a tertiary pediatric hospital involving neonates with CNPAS.

INTERVENTIONS Nasal dilation using Hegar cervical dilators in neonates with severe CNPAS.

MAIN OUTCOMES AND MEASURES Avoidance of sublabial pyriform aperture drill-out and length of stay in the hospital after treatment.

RESULTS Four patients (median age, 15 days) had respiratory distress and feeding difficulties. Nasal stenosis was suspected, and maxillofacial computed tomography scans revealed a mean pyriform aperture width of 4.5 mm. Medical therapy was initiated, but symptoms persisted. Direct laryngoscopy, rigid bronchoscopy, and nasal endoscopy with nasal dilation to at least 4 mm were performed in 4 patients without postoperative stenting. Mean length of stay after treatment was 4 days. Two patients underwent repeat nasal dilation on postoperative days 18 and 23. All 4 patients remained free of nasal disease in a median follow-up of 4.5 months.

CONCLUSIONS AND RELEVANCE Four patients with severe CNPAS were successfully treated with nasal dilation without pyriform aperture bone removal or nasal stenting. This series, while small, suggests that nasal dilation may be a therapeutic option for severe CNPAS that decreases the risks of open surgery and subsequent stent use.

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the brain is recommended in these cases or whenever there is concern for a central neurologic abnormality. Genetics consultation can help determine if other syndromic features are present.

The treatment prescribed for CNPAS consists of initial medical therapy followed by surgical therapy in cases refractory to medical management or with severe initial presentation. Medical therapy includes nasal saline drops, nasal corticosteroid drops, oxymetazoline hydrochloride, temporary stenting of the anterior nares, use of a McGovern nipple to encourage mouth breathing, and/or noninvasive positive pressure ventilation. Surgical therapy consists of a sublabial approach to the pyriform aperture with subsequent enlargement of the pyriform aperture using a powered drill and loupe or microscopic magnification, followed by nasal stenting for 5 to 28 days in most cases.2-7,9

Recognizing that the inferior turbinate arises from the lateral border of the pyriform aperture, we hypothesize that procedures addressing the inferior turbinate may augment medical treatment and reduce the need for sublabial pyriform aperture drill-out. The following series of patients represents our recent experience treating 4 consecutive patients with severe CNPAS and nasal dilation.

Methods

Institutional review board approval from the University of Pittsburgh was obtained for a retrospective review of a case series. All patients were diagnosed with CNPAS and treated with bilateral dilation of the nasal cavities. Presenting symptoms, results of a radiographic workup (width of the pyriform aperture and distance between inferior turbinates), details of the procedure, and subjective outcomes were recorded.

All 4 patients underwent direct laryngoscopy, rigid bronchoscopy, and nasal endoscopy with Hegar cervical dilators (Figure 1). Dilation commenced with a 2-mm dilator and progressed to a maximum of a 4- or 5-mm dilator. All patients were taken to the neonatal intensive care unit and treated with nasal oxymetazoline for 3 days, nasal saline, and nasal corticosteroids. Treatment in the neonatal intensive care unit continued until the patients had obtained all nutritional needs orally and were breathing comfortably. The nasal corticosteroids and saline were continued as outpatient medications.

The procedure was deemed successful if it (and associated medical management) was associated with the resolution of respiratory and feeding problems, allowed outpatient treatment, and avoided the sublabial drill-out procedure. Outcomes were classified as partially successful if the initial symptom improvement did not persist and required revision nasal dilation.

Results

Four consecutive patients with CNPAS sought treatment between October 1, 2011, and August 31, 2012. All 4 patients were undergoing inpatient evaluation for persistent feeding difficulties and failure to thrive. Three of the 4 patients had concomitant respiratory symptoms, including at least one of a variety of symptoms (stertor, noisy breathing, chest retractions, desaturations, and tachyphnea). No patients had stridor or required intubation.

The mean gestational age was 39.8 weeks, while the median age at presentation was 15 days. Bilateral flexible nasal endoscopy (2.2-mm scope) was attempted but unable to be passed in each case. As such, a maxillofacial computed tomographic scan was obtained to identify a site of nasal obstruction. Figure 2 demonstrates a characteristic maxillofacial computed tomographic scan showing the axial and coronal images at the level of the pyriform aperture and other features common to CNPAS. The mean pyriform aperture width was 4.5 mm. The mean distance between the inferior turbinates was 3.5 mm. All 4 patients had a bony ridge of the palate (Figure 2D), and 3 of 4 patients had a triangular-shaped palate and a median central incisor (Figure 2B). Brain magnetic resonance imaging was obtained in 2 of 4 patients to rule out holoprosencephaly. One patient was subsequently diagnosed as having solitary median maxillary central incisor syndrome.

All patients were initially treated with nasal oxymetazoline, saline, and corticosteroids (mometasone or fluticasone). After a median of 2 days of maximal medical therapy with persistent feeding difficulties, all 4 patients underwent direct laryngoscopy, rigid bronchoscopy, and nasal endoscopy with nasal dilation at median age of 18.5 days.

Medical treatment continued after surgery in all patients. All patients had clinical improvement that allowed discharge from the hospital without respiratory or feeding symptoms.
The median length of hospitalization after the nasal dilation was 3.5 days (range, 2-7 days).

Two of the 4 patients with partial recurrence of symptoms underwent repeat dilation a median of 20.5 days after the initial dilation. The median length of hospitalization after the revision procedure was 1.5 days. Subsequent follow-up (range, 2-15 months; median, 4.5 months) on all patients revealed that all 4 patients were thriving without significant respiratory or feeding issues. The Table lists the mean values for weeks gestational age, distances, length of stay, length of follow-up, and need for revision surgery.

### Discussion

Congenital nasal pyriform aperture stenosis was clinically described in 1989. Since then, numerous case series have been published, including radiographic analysis of the condition and reports of surgical therapies. The underlying embryogenesis is not understood, but it appears that the maxillary process is medially positioned compared with a control population. In addition to the pyriform aperture being stenotic, the midnasal and choanal regions of the nasal cavity are also narrow but to a lesser degree.

Currently, no objective criteria exist to determine when CNPAS requires surgical therapy. Lee et al report that nasal obstruction with respiratory distress, apnea, or feeding difficulties indicates surgical management. Recently, Visvanathan and Wynne suggested that a pyriform aperture width less than 5 mm is associated with a need for surgical therapy. In our series, all 4 patients had pyriform aperture widths less than 5 mm, suggesting that their disease severity was similar to the Visvanathan and Wynne cohort in whom medical therapy was unsuccessful and ultimately required pyriform aperture drill-out.

Nasal dilation with Hegar dilators was associated with successful treatment in a small but consecutive series of children with severe CNPAS (pyriform aperture width...
<5 mm). Use of Hegar cervical dilators is advantageous due to their rigidity and straight shape (Figure 1). Urethral sounds were not used because the curvature was suboptimal for anterior nasal cavity dilation. Possibly, balloon dilation may be useful as well, but their use incurs additional cost.

It is notable that 2 patients returned to the clinic with increased nasal symptoms approximately 3 weeks after their initial dilation. Repeat dilation was easily performed, and each patient was discharged home again after a short hospital stay. Revision dilation may not have been required if stenting had been used at the first dilation. Due to the difficulties maintaining the stents (frequent suctioning) and possible complications (septal ulceration or perforation and nasal ala ulceration), repeat nasal dilation appears to be a simpler solution. Since the purpose of dilation was to allow normal respirations, oral nutrition, appropriate weight gain, and the avoidance of sublabial drill-out, we considered these patients to have a successful outcome despite requiring 2 dilations.

Avoiding the sublabial drill-out is advantageous for a couple of reasons. The procedure typically removes 1 to 2 mm of bone from the lateral wall of the pyriform aperture, followed by stenting with a 3.5-mm endotracheal tube from 5 to 28 days.7−9 As such, the risks include injury to the nasal mucosa, nasolacrimal duct, tooth buds, nasal ala, and septum. Devambez et al6 reported that septal ulceration was common (24%) after drill-out and stenting for CNPAS. Furthermore, the maintenance of a stent can be challenging for medical providers and families.

We hypothesize that in many patients with CNPAS, the most medial structure in the nasal cavity is the inferior turbinate (Figure 2B). Our series offers support for this concept, since the median distance between the inferior turbinates was less than the width of the pyriform aperture, suggesting that the inferior turbinate is the flow-limiting structure. Capitalizing on the inferior turbinate being the most medial structure in the nasal cavity, nasal dilation causes isolated turbinate outfracture without addressing the more lateral nasal process of the maxilla. Our experience suggests that dilation improves the airway sufficiently to allow subjective improvement in symptoms and outpatient medical treatment. The traditional drill-out procedure for CNPAS increases the nasal airway because it widens the bony aperture of the nasal cavity, and stents displace the inferior turbinate laterally.6 However, stenting and lateral displacement of the turbinates alone may be a significant factor in improving the nasal airway.

Important in the analysis of any treatment option is the consideration for health care use. The length of hospitalization after nasal dilation ranged from 2 to 7 days (median, 3.5 days). This compares favorably with the largest surgical CNPAS series reporting a mean length of hospitalization of 18.7 days (range, 10-31 days).6 The decreased length of hospitalization and lack of stents may improve the parental quality of life associated with CNPAS. In addition, the overall cost of caring for patients with CNPAS is minimized by reducing inpatient health care costs and decreasing the number of days lost from work for the parents. Thus, nasal dilation is a simple, less invasive technique, possibly capable of offering symptomatic improvement with better health care use. As such, it could augment medical therapy and decrease the need for traditional sublabial pyriform aperture drill-out.

It is unclear what happens to patients with CNPAS as they age, but some authors have suggested that children become less symptomatic due to subsequent growth.10 In the future, prospectively observing patients with CNPAS who are treated medically and surgically with nasal dilation and pyriform aperture drill-out would help to truly determine if dilation has a clinical effect, when dilation should be used, and if treatment modality has an effect on future width of the pyriform aperture in older children and postpubertal adolescents and adults.

The limitations of this study are the lack of a control and another experimental group. A control group would have been helpful for comparing the outcomes of groups that underwent medical therapy alone. However, in our series, all infants were treated with dilation because medical therapy was unsuccessful. In addition, comparison with an experimental group that received pyriform aperture drill-out would have been useful. The infrequency of this disease, however, makes an ideal study with multiple cohorts impractical. The infrequency also limited the sample size of this initial report of a novel therapeutic approach.

Conclusions
Nasal dilation may be an effective management option for severe CNPAS based on symptomatic improvement of 4 consecutive patients with feeding and respiratory difficulties secondary to CNPAS. We propose that this simple surgical procedure be considered a treatment option in addition to those currently in use. Further experience with this nasal dilation will help to elucidate its optimal use and role in a treatment algorithm for CNPAS.

CORRECTION

Error in Online Author Name: An error appeared in the first author’s name in the byline of the online version of the Original Article titled “The Health Impact of Chronic Recurrent Rhinosinusitis in Children” (doi:10.1001/archotol.126.11.1363), originally published in the November 2000 print issue of Otolaryngology–Head and Neck Surgery (2000;126[11]:1363-1368). The first author’s name appeared correctly in the print and downloadable pdf versions of the article. The first author’s name is Michael J. Cunningham, MD.