Congenital Nasal Pyriform Aperture Stenosis: Diagnosis and Treatment

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Congenital nasal pyriform aperture stenosis is an unusual form of nasal airway obstruction in the neonate. Pediatric plastic surgeons are often involved in the management of these children and should recognize this condition and know the treatment options. Fifteen cases of children with congenital nasal pyriform aperture stenosis were reviewed for presentation of the disorder, management, and effectiveness of treatment, making it the largest series to date. There were nine male patients and six female patients in the series. They all experienced varying degrees of nasal obstruction at birth and were managed on the basis of the severity of their symptoms. Twelve patients were treated surgically in the first year of life, with a mean age at operation of 97 days (range, 3 to 362 days). Two patients required surgical intervention during their teenage years (age, 14 and 18 years) because of persistent symptoms, and one patient (age, 2 years) with mild symptoms was managed medically. Associated craniofacial anomalies were present in six cases (40 percent). Surgical enlargement of the pyriform aperture was successfully performed through an upper buccal sulcus incision in 14 patients. Preoperative symptoms of upper airway obstruction were improved in all patients at an average follow-up of 2.4 years (range, 1 month to 5 years). Congenital nasal pyriform aperture stenosis varies in presentation and severity, occurring either as an isolated congenital anomaly or in association with developmental craniofacial anomalies. It can be effectively managed by surgical enlargement of the pyriform aperture without significant recurrence or long-term morbidity. (*Plast. Reconstr. Surg.* 109: 1506, 2002.)

Nasal airway obstruction is a potentially life-threatening condition in the newborn because of obligate nasal breathing. The pyriform aperture is the narrowest, most anterior bony portion of the nasal airway, and a decrease in its cross-sectional area will significantly increase nasal airway resistance. Congenital nasal pyriform aperture stenosis is an unusual form of nasal obstruction and is not unique to the craniofacial population. It should be considered in the differential diagnosis of any neonate or infant presenting with signs and symptoms of upper airway compromise.[1,2] It is important to differentiate this level of obstruction from the more common posterior choanal stenosis or atresia. Craniofacial surgeons are often involved in the management of children with upper airway obstruction and should be able to recognize this condition and know the treatment options. Congenital nasal pyriform aperture stenosis presents with symptoms of nasal airway obstruction often characterized by episodic apnea and cyclical cyanosis. The diagnosis is suggested by history and physical examination; however, it should be confirmed radiographically by a computed tomographic scan of the nasal cavity (Fig. 1). Numerous questions remain regarding the exact cause and degree of association with other craniofacial developmental anomalies.

In 1995, Burstein and Cohen[1] described the initial experience of four patients with nearly complete pyriform aperture stenosis. The purpose of our report is to expand this series, specifically in the areas of presentation, associated anomalies, and management strategies.

Patients and Methods

Patients

A retrospective review was performed of 15 cases of children diagnosed with congenital nasal pyriform aperture stenosis who were seen between January 1992 and December 2000 by the craniofacial team at Children's Healthcare of Atlanta at Scottish Rite Hospital. The series consisted of nine boys and six girls. Five patients were born prematurely. Discharge data-

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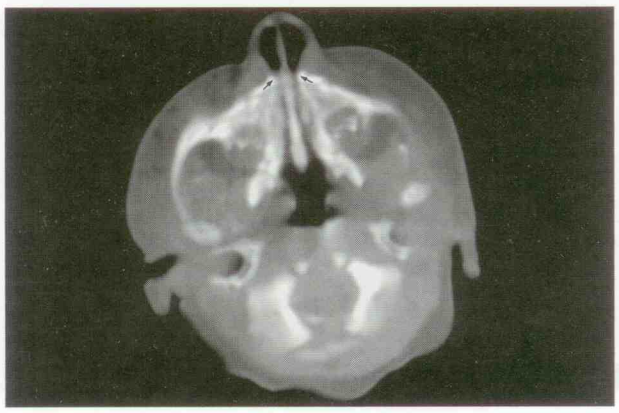

FIG. 1. Computed axial tomographic image taken at the level of the lower nasal cavity, demonstrating focal stenosis of the nasal pyriform aperture (*black arrows*) by the nasal process of the maxilla.

bases, medical charts, and office notes were used to analyze details of their diagnosis, preoperative assessment, initial management, surgical operations, and outcome. The degree of presentation was reviewed and the various treatment options were evaluated. Special attention was directed toward any associated physical or developmental anomalies. Postoperative courses were reviewed with particular focus on symptomatic recurrence and developmental abnormalities.

Clinical presentation represented a spectrum of severity, with varying degrees of stridor, congestion, snorting, and intermittent episodes of apnea. All patients in the series were symptomatic at birth. Symptoms were often more pronounced with feeding or with infections of the upper respiratory tract and were relieved by crying. Actual cyanotic episodes with desaturations were present in 47 percent of the patients ($n = 7$), as documented by sleep studies or continuous pulse oximetry. The inability to pass a feeding catheter through the anterior naris was occasionally experienced on physical examination, further suggesting the diagnosis of congenital nasal pyriform aperture stenosis. A controlled airway was established with endotracheal intubation or oropharyngeal tubes when necessary. The diagnosis was suspected clinically by the previously mentioned presentations and confirmed radiographically with a computed tomographic scan of the nasal cavity and maxilla (Fig. 1). Occasionally, the diagnosis was established by using nasal endoscopy.

Treatment was either medical ($n = 1$) or surgical ($n = 14$), depending on severity of the symptoms. Eighty percent of the children ($n = 12$) were treated surgically in the first year of life, with six of these treated before 1 week of age (Table I). Their mean age at operation was 97 days (range, 3 to 362 days). Two patients were managed conservatively, at first, and required surgical intervention during their teenage years (age, 14 and 18 years) because of persistent symptoms. Along with enlargement of their pyriform aperture, these adolescents also underwent an inferior turbinectomy and septoplasty. One patient (age, 2 years) was diagnosed with congenital nasal pyriform aperture stenosis at birth and has been followed medically.

TABLE I

Profile of Patients with Congenital Nasal Pyriform Aperture Stenosis Showing Management and Associated Anomalies

Case No.	Additional Diagnosis	Age at Surgery	Management	Associated Craniofacial Anomalies	Outcome
1	CA	3 days	PAE, CA repair	Apert syndrome	No recurrence
2	CA	28 days	PAE, CA repair	Crouzon's	No recurrence
3	CA	300 days	PAE, CA repair	Achondroplasia, MF hypoplasia	Early recurrence
4		89 days	PAE	Hydrocephaly	No recurrence
5		14 years	PAE, septoplasty, turbinectomy	Narrow vertical MF, SCMI	No recurrence
6		18 years	PAE, septoplasty, turbinectomy	MF hypoplasia	No recurrence
7		9 days	PAE	None	No recurrence
8		362 days	PAE	None	No recurrence
9		360 days	PAE	None	No recurrence
10		6 days	PAE	None	No recurrence
11		3 days	PAE	None	No recurrence
12		4 days	PAE	None	No recurrence
13		4 days	PAE	None	No recurrence
14		5 days	PAE	None	No recurrence
15		N/A	Medical management	None	Occasional snorting

CA, choanal atresia; PAE, pyriform aperture expansion; MF, midface; SCMI, single central maxillary incisor.