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Patient selection in congenital pyriform aperture stenosis repair – 14 year experience and systematic review of literature

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ABSTRACT

Purpose: Congenital nasal pyriform aperture stenosis (CNPAS) is a rare cause of respiratory distress in neonates that may necessitate early surgical intervention. Restenosis and granulation are postoperative concerns that may prompt a return to the operating room. Reoperation places children at increased risk of perioperative complications and prolonged hospital stays. We are presenting a review of our institutional experience of 16 patients treated for CNPAS over a 14 year period and a systematic review with pooled data analysis to determine the effect of craniofacial and neurologic anomalies on surgical success.

Methods: Retrospective chart review of all cases of CNPAS treated at our tertiary children's hospital between 1999 and 2013. Systematic review of English language literature was conducted adhering to the PRISMA statement to determine the effect of neurologic anomalies and craniofacial dysmorphism (CFD) on surgical failure for CNPAS treatment. Univariate and exact multiple logistic regression were used for analysis of an individual patient data analysis.

Results: 10 patients had surgery and 6 were treated medically. Average pyriform apertures were 5.71 ± 1.72 mm for the surgical group and 4.83 ± 1.26 mm for the medical group ($p = 0.38$). 31% had neurological impairments. 31% had craniofacial dysmorphisms (CFD). 2 patients developed restenosis and 1 required tracheotomy. Both of these patients had other CFDs. Literature review captured 63 surgical patients and 9 failures in 6 series of CNPAS. 4.6% of patients without CFD and 36.8% of patients with CFD required surgical revision ($p = 0.023$, OR13.8).

Conclusion: When repairing CNPAS, co-morbidities must be considered. Impaired respiration, central neurologic deficits and extensive craniofacial anomalies may require additional surgeries or an alternative approach.

1. Introduction

Congenital nasal pyriform aperture stenosis (PAS, CNPAS) is an uncommon cause of nasal obstruction in children and neonates. It is caused by bony overgrowth at the pyriform aperture or the maxillary nasal inlet. PAS was first described in adults in the 1950s but it was not until 1989 that Brown formally identified a congenital etiology in children [1,2]. Since that time its management has evolved from simple transnasal dilation to sublabial and submucosal approaches. Recently there has been debate regarding which children can be managed medically and which ultimately will require surgery. We present our experience with 16 consecutive CNPAS patients treated at our tertiary care children's hospital between 1999 and 2013. Our series identified two patients, with multiple comorbidities, that failed initial surgical management requiring other operative interventions. There are few large series describing the management of CNPAS and little is known about the mechanisms of surgical failure. It is understood in other airway surgeries that neurologic and craniofacial anomalies can limit success [3,4]. We performed a systematic review to assess whether craniofacial dysmorphism

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(CFD) and neurologic anomalies (NA) can predict postoperative difficulty after surgical correction of CNPAS.

2. Methods

Retrospective review of consecutive patients treated for CNPAS at the Steven and Alexandra Cohen Children's Medical Center of New York was performed. Children were managed by three surgeons (JC, MS, LS) between 1999 and 2013. Medical and surgical management choices were determined by each child's presentation. All patients were trialed on medical management after the diagnosis of PAS. Medical management included nasal saline irrigations, topical nasal steroids and topical nasal decongestion. If the child was not able to be weaned from high pressure or high flow supplemental oxygen and take adequate oral nutrition, surgery was considered. Surgical approach was via a sublabial incision in all cases [5]. Sub-periosteal dissection was carried to the inferior turbinates and then a cutting drill bit was used to remove adequate bone so that a 3.0 or 3.5 endotracheal tube stent could fit adequately in each nasal airway. Care was taken to avoid injury to the nasolacrimal duct. If needed, the inferior turbinates were also out-fractured. The stents were secured with an endolumenal suture through the septum. Stents were left in place for seven days while children were kept in a monitored setting.

Systematic review was performed on all published series of pyriform aperture stenosis adhering to the PRISMA guidelines (Fig. 1) [6]. Papers were identified via Pubmed and Google Scholar searches using the following search terms: "pyriform/piriform aperture stenosis/narrowing", "pediatric/congenital nasal obstruction", "PAS", "CNPAS", and "CNPS". Independent review was performed with the following inclusion criteria: unique patients with the diagnosis of CNPAS, surgical repair of stenosis, at least 3 patients in the series, individual patient data available for comparison and analysis that includes comorbidities and outcome. Exact multiple logistic regression was carried out (PROC LOGISTIC, SAS Institute, Cary, NC), stratifying by author. Details are further described in the following section.

3. Results

Sixteen children were managed for PAS during the 14 years reviewed. Excluding the child that was identified at 3 years of age, the average age at diagnosis was 4.2 days old (Table 1). The workup for most children began shortly after birth. Most pregnancies were uncomplicated and full-term. There were two twin pregnancies. Family history was rarely contributory; however, one twin with PAS had a genetically identical twin sibling with holoprosencephaly (HPE) and single mega-maxillary incisor (SMMI), but without PAS. Genetic work up for this family did not identify a known mutation. One was identified to have Aperts syndrome but was included as the obstruction was only at the pyriform aperture; he did not require surgical intervention.

Table 1 describes our medical and surgical management of PAS patients. The average size of the pyriform aperture was 5.71 mm ($\pm$1.72 mm, $n = 9$) in patients undergoing surgery and 4.83 mm ($\pm$1.26 mm, $n = 3$) in those undergoing medical management ($p = 0.38$). Surgery was offered to those children that could not be weaned off of high-flow supplemental oxygen or non-oral enteral feeding modalities. Ten patients proceeded to surgery while six were weaned successfully with medical management. After surgery all patients were weaned off of supplemental oxygen and feeding except for one that required two subsequent dilations and revisions, and another child that eventually received a tracheotomy. Two patients were gastrostomy dependent postoperatively related to congenital intestinal malrotations.

Comorbidities in our patients are listed in Table 1. Seven out of 16 patients had a SMMI (43%). One patient had a concomitant subglottic stenosis identified during screening bronchoscopy. Most children had normal neurological development, but 6/16 (37%) had documented developmental delays. While SMMI was relatively

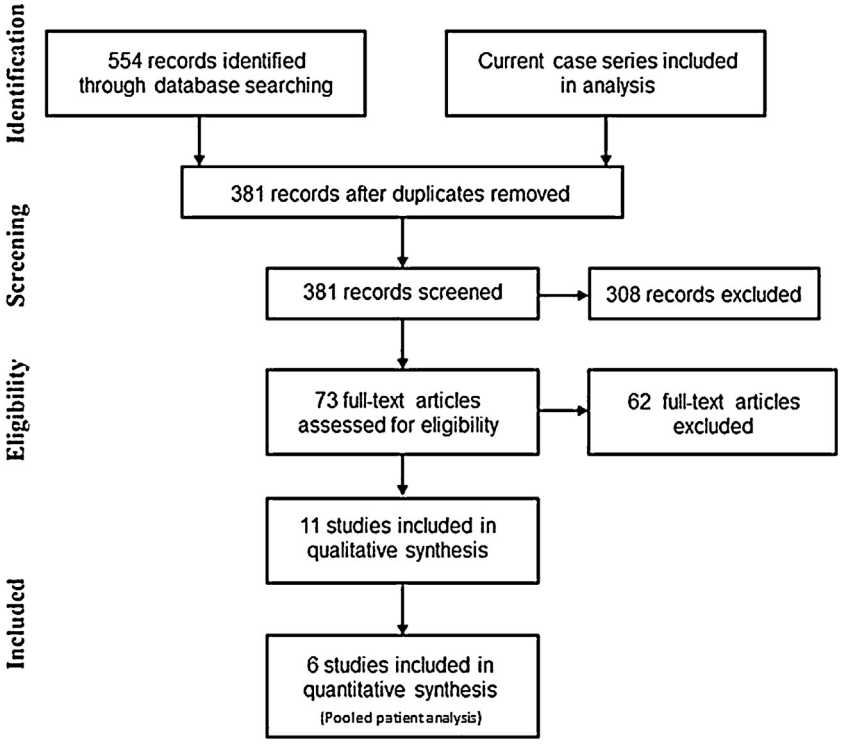

Fig. 1. PRISMA flowchart of systematic review inclusion [4].