Using the Maxillary-Nasal Angle to Evaluate Congenital Nasal Pyriform Aperture Stenosis

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**Importance**
Congenital nasal pyriform aperture stenosis (CNPAS) is a rare cause of nasal airway obstruction in newborns. The decision to operate is made clinically. Although pyriform aperture width is used for diagnosing CNPAS, it does not fully characterize stenosis of the nasal cavity.

**Objective**
To determine the utility of additional metrics for evaluating CNPAS.

**Design, Setting, and Participants**
The medical records of 13 patients with CNPAS treated from 2007 through 2012 at a single tertiary pediatric facility were retrospectively examined. Data on patient demographic characteristics, known genetic abnormalities, and hospital courses were extracted. Computed tomographic images were evaluated for pyriform aperture width; maxillary-nasal angle (MNA), defined as the angle between the anterior maxilla and anterior-posterior nasal axis; and choanal width.

**Interventions**
Medical management and surgical management.

**Main Outcomes and Measures**
Pyriform aperture width, MNA, and choanal width.

**Results**
Six of 13 patients underwent medical management, and 7 patients underwent surgical treatment. For patients who were managed medically as compared with those managed surgically, the evaluation revealed a larger pyriform aperture width (median [interquartile range {IQR}], 5.6 [5.4-6.1] vs 4.6 [4.5-4.7] mm; \( P = .03 \)) and MNA (median [IQR], 70° [63°-73°] vs 59° [59°-64°]; \( P = .048 \)) but no significant difference in choanal width (median [IQR], 11.0 [9.6-12.2] vs 11.9 [10.3-11.9] mm; \( P = .76 \)).

**Conclusions and Relevance**
The MNA, when used in conjunction with pyriform aperture width, provides additional pertinent information to supplement clinical decision making in the evaluation of patients with CNPAS. These measurements may be helpful in identifying patients who should undergo surgical intervention, although additional studies would be required to allow predictive use of the MNA.
dromes that cause significant nasal and midfacial abnormalities. The clinical course of CNPAS can range from patients managed medically to those who require multiple surgical procedures. There are currently no strategies described in the literature using measurement of pyriform aperture width to help predict patient tolerance of medical management alone. However, use of imaging metrics to characterize the anterior and posterior nasal cavity might allow for refined prediction of those patients who will require surgical intervention. Our objective was to determine the ability of additional CT characteristics besides pyriform aperture width to predict the need for surgical intervention in patients with a diagnosis of CNPAS.

Methods

After approval was obtained from the institutional review board for the Research Institute at Nationwide Children’s Hospital in Columbus, Ohio, and need for informed consent was waived due to the retrospective nature of this study, a retrospective review was performed to search for patients with CNPAS. Medical records of patients treated from 2007 through 2012 with International Classification of Diseases, Ninth Revision, Clinical Modification diagnostic code 748.0 (choanal atresia) or 748.1 (other congenital anomaly of nose) or Current Procedural Terminology code 30540 (intrasal choanal atresia repair) or 30465 (repair of nasal vestibular stenosis) were screened for a diagnosis of CNPAS, defined as pyriform aperture width of less than or equal to 11 mm on CT imaging. Patients with syndromes that cause significant nasal and midfacial abnormalities such as Noonan syndrome, Apert syndrome, and hemifacial microsomia with heminasal hypoplasia were excluded.

Patients were separated into 2 cohorts: those who underwent surgical intervention and those who received medical management alone. Medical records were reviewed to document clinical characteristics including presentation, medical or surgical management, number of procedures, radiologic findings, placement of a gastrostomy tube prior to discharge, and diagnosis of a genetic abnormality prior to discharge. On CT imaging, the pyriform aperture width was defined as the width of the pyriform aperture at the inferior meatus, the choanal width was defined as the distance across the nasal cavity at the posterior choana, and a novel measurement, the maxillary-nasal angle (MNA), was defined as the angle between the anterior maxilla and the anterior-posterior nasal axis (Figure). Data and imaging were independently reviewed by 2 pediatric otolaryngologists (T.P.M., J.M.G.) with no discrepancies in interpretation identified.

Comparisons were made between the population who underwent medical management and the population who underwent surgical management. Categorical variables were compared using Fisher exact tests. Pyriform aperture width, MNA, and choanal width were compared using Wilcoxon exact tests with medians and interquartile ranges (IQRs) reported. A Pearson correlation coefficient was calculated for the correlation between pyriform aperture size and MNA. All analyses were performed using SAS, version 9.3, and P < .05 was considered statistically significant.

Results

Of 13 patients who met inclusion criteria, 7 patients (54%) were treated surgically using an open sublabial approach with drill-out of the pyriform aperture bone in a submucosal fashion. Six patients received medical management alone (Table). There were no differences in presenting symptoms, physical examination findings, or length of stay. Eight patients had genetic abnormalities, including sonic hedgehog mutation, double aortic arch, 22q deletion syndrome, double-outlet right ventricle, 18p deletion, holoprosencephaly, hypopituitarism, and hypothalamic hamartoma. All patients who were managed surgically had nasal stents placed at the completion of surgery, with the duration of stenting ranging from 1 to 8 weeks after the primary procedure.

Several patients had complications from surgery. Two patients required tracheotomies, with 1 due to acquired subglottic stenosis. Two patients developed nasal synechiae after the initial surgery, necessitating additional surgical procedures. Two of the 13 patients were identified as deceased at the time of medical record review; 1 death was associated with likely stent plugging, whereas the other death was due to cardiac comorbidities.

Computed tomographic imaging was available for 11 patients. The median (IQR) pyriform aperture width was 5.6 (5.4-6.1) mm in the medical group and 4.6 (4.5-4.7) mm in the surgical group (P = .03). The median (IQR) MNA was 70° (63°-73°) for those managed medically and 59° (59°-64°) for pa-
patients who required surgery (P = .048). The median (IQR) choanal width was 11.0 (9.6-12.2) mm for patients managed medically and 11.9 (10.3-11.9) mm for patients who underwent surgery (P = .76). The angular measurement and pyriform aperture size were not statistically significantly correlated (Pearson correlation coefficient, 0.10; P = .76).

### Discussion

For those patients with CNPAS who require surgical management, one important consideration is whether nasal cavity narrowing is limited to the pyriform aperture or if it extends into the middle and posterior nasal cavity. Although CNPAS is defined on the basis of the diameter of the pyriform aperture, the anatomy of the nasal cavity is more complex than this definition would suggest. The present study demonstrates that patients who underwent surgery had smaller MNAs and pyriform aperture widths. The MNA accounts for the width of the nasal cavity, measuring whether the narrowing is isolated to the area of the pyriform aperture or if the entire nasal cavity is long and narrow. The absence of correlation of the MNA and pyriform aperture width suggests that these measurements provide 2 separate pieces of information instead of expressing similar information about the nasal cavity.

The decision to operate on patients with CNPAS is based on clinical evaluation. The management of CNPAS is highly variable and depends on a variety of factors including degree of respiratory distress, severity of symptoms, and response to initial medical treatment. Although a consensus for surgical criteria for CNPAS does not exist, Van den Abbeele et al recommend treating patients medically for up to 2 weeks before considering surgery, and proceeding with surgery if the patient continues to demonstrate increased work of breathing, cyanosis, or oxygen desaturation. In our study population, patients were managed conservatively for a median (IQR) of 10 (9-25) days before undergoing surgical correction based on clinical evaluation.

Disadvantages of surgical correction include formation of nasal synechiae and unnecessary exposure to surgery and general anesthesia because many patients will outgrow their disease as the pyriform aperture expands and they are no longer obligate nasal breathers. Alternatives to surgical correction include tracheostomy placement, medical management, and serial dilation. Medical management can expose patients to unnecessary risks as well; patients with inadequately treated CNPAS are at risk of poor nutrition and failure to thrive. Wine et al recently presented a case series of patients with CNPAS whose medical management failed who were treated successfully with serial dilation, but these patients still required general anesthesia, intubation, direct laryngoscopy, and bronchoscopy. Conservative measures such as medical management and serial dilation are effective in some patients, but because up-front surgery may provide better control of the neonatal airway and definitive cure without the need for multiple procedures, correctly determining which patients would benefit from a conservative or surgical management strategy is important.

Determining the anatomy of the entire nasal cavity is important for surgical planning. Koga et al reported a series of neonates with bony nasal stenosis that extended posteriorly into the nasal cavity. If the obstruction extends more posteriorly, initial surgical correction of the pyriform aperture stenosis may not provide an adequate nasal airway. The patient...
may continue to have respiratory distress, feeding difficulties, and other problems. This may require the patient to return to the operating room for further surgical widening of the nasal airway. Evaluating the posterior nasal cavity on CT scan prior to surgery may help to determine whether the neonate initially requires more extensive surgery and may help the surgeon decide to manage the patient medically or to avoid nasal surgery altogether and bypass the obstruction via tracheostomy.

The MNA and pyriform aperture size, when considered together, provide information about the anterior and posterior nasal cavity to help the surgeon decide whether it is appropriate to proceed with surgery. Our data revealed a larger MNA for patients who were managed medically as compared with those managed surgically. In addition, the degree of pyriform aperture stenosis was less severe in the medically managed group. To our knowledge, these associations have not been previously reported in the literature, and whereas there is no clear division between the 2 groups, decreased MNA and pyriform aperture width may be predictors for reduced disease severity. It is possible that use of additional metrics would further characterize the nasal cavity and add even more information for clinical decision making, but the additional benefit of using the MNA over other methods such as 3-dimensional modeling is that the MNA is easy to measure with minimal technology. We did consider another measurement, choanal width, but this measurement was not associated with a need for surgery in our patient population.

Although our analysis indicates that MNA measurement could be a helpful tool in determining the type of treatment that patients with CNPAS receive, there were limitations to the present study. The sample size was 13 patients, 11 of whom had CT imaging. Any correlation between CT imaging findings and the need for multiple surgical procedures could not be determined with sufficient power. The present study also demonstrates associations between CT measurements and medical vs surgical management based on our group practice. It is our standard practice to treat patients with medical management first and proceed to surgical management in cases of medical failure.

In the evaluation of patients for CNPAS, the anatomical cause of obstruction of airflow may be more complicated than stenosis of the pyriform aperture alone. For this reason, the decision to operate on patients with CNPAS remains a clinical decision. Without clear surgical criteria for CNPAS and given the many postoperative complications that are possible, MNA measurement may help better select those patients who can tolerate conservative management. Additional studies that evaluate the utility of this diagnostic measurement may allow surgeons to differentiate which patients are more likely to require surgery and which may be managed medically.

## Conclusions

The decision to operate on patients with CNPAS is a clinical decision. Measuring both the MNA and pyriform aperture width may be helpful in identifying those patients who will likely need surgical management, as well as for planning the extent of surgery. Improved preoperative planning as afforded by use of the MNA may result in a reduction in exposure to multiple surgical interventions or pyriform aperture dilations, but our small sample size limits the predictive ability of this novel measurement.