Abstract

Objective: To introduce and outline the advantages of using 1% ophthalmic dexamethasone drops intranasally (ODDI) to treat nasal obstruction in patients with congenital pyriform aperture stenosis (PAS). Study Design: Retrospective review from 2006-2009. Setting: Tertiary referral center. Subjects and Methods: All children diagnosed with PAS were included for review. Computer-tomography measurements were taken for pyriform aperture and midnasal diameters. Symptoms at initial presentation and treatments were reviewed for each patient. Outcome measures included the cessation of obstructive symptoms per care giver reports, ability to successfully taper steroid drops, and avoidance of surgical intervention. The dose of 1% ophthalmic dexamethasone began with 2 drops each nostril BID and then was increased or tapered depending on symptoms of obstruction. Results: Five children were diagnosed with PAS at our institution in the past three years. Four of these patients were incidentally found to have characteristics consistent with solitary median maxillary central incisor syndrome (SMMCIS), and all patients had midnasal cavities less than or equal to 11 mm between lateral nasal walls. One patient was initially treated at an outside facility with surgical repair of PAS after “failed conservative therapy.” A second patient was treated surgically at our facility because of respiratory insufficiency requiring intubation. Both patients presented to us with continual nasal obstruction after surgery and responded well to ODDI without requiring any further surgical intervention. Three other patients presented with apnea and desaturations. All three responded well to ODDI without requiring surgery. All five patients in follow up (avg. 1 year) have been weaned off their steroid drops and are breathing well with no dysphagia. Conclusion: SMMCIS and midnasal stenosis (MNS) are often found in children with PAS. MNS is typically not addressed in patients with PAS and is difficult to treat surgically. We have found that these patients may not respond well to surgical intervention. In the past, conservative measures for PAS have mostly included nasal saline sprays and vasoconstrictors; however, our results demonstrate the benefit of ODDI in treating PAS. We recommend using ODDI as conservative management prior to surgical repair of PAS, especially in the setting of MNS.

Introduction

The diagnosis of congenital nasal pyriform aperture stenosis (CNPAS) requires a pyriform aperture width less than 11 mm on computed tomography (CT).1-2 It is often accompanied by a solitary median maxillary central incisor.3-4 Midnasal stenosis (MNS), although not well defined in the literature, may occur simultaneously with CNPAS and contribute to symptoms of nasal obstruction in some patients. We also suspect that midnasal narrowing may contribute to persistent symptoms in CNPAS after surgical treatment in some patients and possibly presents a stronger justification for conservative treatment.

CNPAS patients have been taught from the outset that the pyriform aperture diameter is greater than 5 mm.5 Conservative treatments reported in the literature include humidification, decongestants, nasal saline sprays, and vasoconstrictors.6 Side effects include mucosal ulcerations, inflammation, and seep perforation.7 To our knowledge, conservative nasal steroid treatment and the associated side effects have not been formally described in the literature for CNPAS. Some treat CNPAS conservatively for 7-10 days before progressing to sublabial surgical intervention.8 Surgical risks include tooth bud and nasolacrimal duct damage, restenosis, and granulations.9 Post surgical medical and nasal developmental hypoplasia and sepal ulcerations have also been observed.10 A therapy that avoids the drawbacks of current conservative methods, gives prompt relief of nasal obstruction, and avoids surgical risks could be of significant benefit to CNPAS. This study will evaluate use of 1% ophthalmic dexamethasone drops inanally (ODDI) to treat CNPAS nasal obstruction. We examine the pyriform aperture and midnasal diameters, and treatment outcomes.

Results

Three female patients were initially given conservative ODDI. Two other patients underwent surgery as initial CNPAS intervention. One patient presented after birth with cyanosis during feeding. CT showed a 4.7 mm pyriform aperture diameter and the interuncinate distance was 11 mm. ODDI was begun after diagnosis and was tapered from two drops to one and then to twice daily over one month before she was released. Improvement occurred and surgery was unnecessary. At follow up, the PA diameter and interuncinate distance had increased to 8.8 and 15.5 mm.

Patient 2 presented with nasal obstruction at birth with CNPAS. 5 days of Afrin raised the oxygen saturation percentage from the 70’s to the high 90’s, but obstruction and dysphagia persisted intermittently. The patient started ODDI and after 9 days was tapered to one drop per nostril per day. Nasal obstruction and feeding improved. She was eventually weaned off ODDI.

Patient 3 presented at birth with sleep apnea, desaturations in the 80’s, and nasal obstruction. ODDI was commenced after diagnosis. At 2 weeks she was breathing freely through her nose, retractions subsided and dysphagia improved moderately. By two months, however, she was not receiving ODDI consistently due to caregiver noncompliance and her symptoms returned. After reinstating ODDI at one drop per nostril per day, symptoms again improved and stabilized. She was weaned completely off ODDI after 6 months.

Patient 4 was intubated when 3 days old due to severe respiratory failure from CNPAS. Surgical correction was performed at six weeks. Nasal obstruction persisted post-surgically, however, and ODDI was instituted. After little improvement, his dose was increased to four times daily. The increase improved the obstruction and allowed adequate nasal breathing. Drops were tapered until 1.5 years old. After weaning ODDI, he was sleeping restlessly. Although he had no significant sleep apneas, he continued breathing persistently, and at 3 years old this was treated with Flonase pm, which helped alleviate his symptoms.

Patient 5 is a 4 year old female who presented with obstructive sleep apnea, nasal obstruction, and a history of CNPAS. Although his outside CT was not available for review, he had undergone a sublabial repair at an outside institution, which initially improved his symptoms, but nasal obstruction and sleep disordered breathing eventually developed. He was treated with pm Afrin postoperatively for several months, which was not successful at treating his symptoms. The child was a candidate for either surgical PA repair or ODDI with adenotonsillectomy. Her parents elected for the latter. Nasal obstruction continued, however, and she required valvular stenosis repair. Breathing had improved after this surgery and she required no further treatment for nasal obstruction. The patient was subsequently lost to follow up.

Discussion

For most neonatal patients with CNPAS, surgical is generally not recommended until they reach 10 pounds, 16 weeks, or 20 grams of hemoglobin.6 Conservative therapies or ventilator support may help manage symptoms until surgery can be performed in most cases. ODDI offers an effective alternative to surgery. This case series is the first to show ODDI to be an effective option of therapy for some patients with CNPAS. ODDI was effective in patients 1, 2, and 3 from the onset and averted surgery entirely rather than deferring it. In patients 2 and 3, ODDI provided rapid breathing improvements and was weaned within 2 weeks.

Patients 4 and 5 had severe cases of CNPAS that required surgery initially. ODDI was effective in alleviating post-surgical nasal obstruction in these patients. While ODDI helped Patient 4 avoid further surgery, patient 5 only had mild improvements from ODDI and required ventilator stenosis repair as well as an adenotonsillectomy.

Ingestion or absorption of dexamethasone can cause adrenal insufficiency, immunosuppression, or metabolic side effects, and patients should be closely monitored by a pediatric endocrinologist until further data on safety and side effects are available.11 While no side effects were observed in patients 1, 2, 3. or 5, patient 4 required a longer course of ODDI, which resulted in iatrogenic Cushing symptoms and adrenal suppression. These effects are likely minimized by close monitoring by endocrinology, low dosing, and careful, slow tapering. Further studies on the safety and side effects are required.

Conclusion

This is the first case series to illustrate the use of ODDI in the management of CNPAS. We found that ODDI may be an effective conservative therapy in some patients with CNPAS. Further studies are needed to document the results and safety of ODDI therapy in these patients. Whether the presence of MNS should be considered in management decision making is uncertain. A consensus definition of MNS may be helpful to stratify patients when comparing conservative and surgical treatment options in the future.

Methods and Materials

Five children evaluated for CNPAS at our tertiary referral center from 2006-2009 were reviewed retrospectively. CT measurements of the pyriform aperture diameter and the midnasal diameter (interuncinate distance) were documented. All patients, initially received two drops in each nostril twice daily. Dosage was modified and tapered based on symptoms. Pediatric endocrinology was consulted for each patient to help diagnose and manage side effects. Outcome measures included the cessation of airway obstruction, ability to taper steroid drops, necessitation of surgical intervention, and evidence of adrenal suppression.

Table 1: Patient Characteristics

<table>
<thead>
<tr>
<th>Patient</th>
<th>PA diameter (mm)</th>
<th>Interuncinate Distance (mm)</th>
<th>Age of ODDI (mo)</th>
<th>Duration of ODDI (months)</th>
<th>Surgical intervention required</th>
<th>Follow up (Months)</th>
<th>Adrenal suppression</th>
<th>Weight % (age) on last FU</th>
<th>Height % (age) on last FU</th>
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</tr>
</tbody>
</table>

ODDI: ophthalmic dexamethasone drops intranasally; NA: not available; N: no; Y: yes; Mo: months; FU: follow up

1Based on CDC growth charts

References

2. Hall MC. Congenital nasal pyriform aperture stenosis (CNPAS): a case series. Graysville, Ohio, University of Cincinnati College of Medicine, 2010.