American Journal of Otolaryngology and Head and Neck Surgery
Research Article
Published: 04 Sep, 2023

Congenital Nasal Piriform Aperture Stenosis – A New Paradigm for Surgical Management

*Vijayasekaran S**

Perth Children's Hospital, University of Western Australia, Australia

Abstract

Congenital Nasal Piriform Aperture Stenosis (CNPAS) is a rare cause of nasal obstruction in infants, characterized by narrowing of the nasal maxillary processes to <11 mm. For patients who fail conservative management, surgery has traditionally been *via* sublabial approach to drill-out the nasal aperture, with nasal stenting for up to 1 month. Radiological evidence shows that the entire length of the nasal cavity is narrowed in these patients which is not addressed by drill-out procedures. The first report of balloon dilation to treat CNPAS was in 2014 and the author has employed this technique since 2017. Including the present series of 4 patients there have been 10 patients with CNPAS managed by dilation alone, of whom 6 required a single dilation procedure, 3 required a single repeat dilation, and 1 who required an open drill-out procedure after repeated dilation. Three out of 9 patients successfully treated with dilation alone had a stent placed post-dilation. Balloon dilation is technically easier to perform than open drill-out, is associated with fewer complications, rapid recovery, and reduced length of hospitalization. Correcting the entire length of the nasal cavity could have long term benefits on development of nasal structures and the need for additional surgery later in life. The requirement for stents after balloon dilation remains undefined. Balloon dilation represents a new treatment paradigm for patients with CNPAS and could be used as the first-line treatment in patients requiring surgery. There is still a role for drill-out procedures in patients who fail repeated dilation.

Keywords: Congenital nasal piriform aperture stenosis; Pyriform stenosis; Balloon dilation

Introduction

Congenital Nasal Piriform Aperture Stenosis (CNPAS) is a rare cause of nasal obstruction that usually manifests soon after birth as respiratory distress, cyanosis when feeding, apnoea, chronic nasal congestion, and failure to thrive. Presenting symptoms can range from mild to severe, necessitating urgent life-saving intervention in some cases. CNPAS was first described in 1952 [1]. The first Computer Tomography (CT) radiological description of CNPAS distinguishing it from choanal atresia was reported by Ey et al. in 1988, and the first case series was reported the following year [2,3].

The piriform aperture is the anterior limit of the nasal cavity formed superiorly by the nasal bones, inferiorly by the maxillary processes, and laterally by the frontal processes of the maxillary bone. CNPAS is suspected when there is failure to pass a nasal tube or endoscope and is confirmed by CT or Magnetic Resonance Imaging (MRI) which shows narrowing of the aperture due to overgrowth of the nasal processes of the maxilla [4]. Other common radiological findings include a triangular palate and dental abnormalities such as solitary median mega-incisor [4].

There is much about CNPAS that remains elusive. The true incidence is not known but CNPAS is thought to occur in approximately 1:25,000 births, and may be more common in females [4,5]. The narrowed pyriform aperture is a result of failed reabsorption of epithelial cells that plug the nostrils between 8- and 24-weeks' gestation [6]. The aetiology is unclear but approximately 60% of cases also present with a solitary median maxillary central incisor which is found in some children with a missense mutation in the Sonic Hedgehog gene (I111F) at 7q36 [5,7]. Other malformations reported in children with CNPAS include other neurological deficits and/or craniofacial dysmorphisms in approximately one-third of patients in one series, as well as holoprosencephaly, hypopituitarism, cardiac, and urogenital malformations [5,8-10]. Gestational diabetes is present in up to 75% of cases [11,12]. In view of the high rate of associated abnormalities, cross-sectional imaging plays an important role in determining the overall management strategy.

OPEN ACCESS

Correspondence:
Shyan Vijayasekaran, Perth Children's Hospital, University of Western Australia, Australia, Tel: +61893280580

Received Date: *11 Aug 2023*
Accepted Date: *29 Aug 2023*
Published Date: *04 Sep 2023*

Citation:
Vijayasekaran S. Congenital Nasal Piriform Aperture Stenosis – A New Paradigm for Surgical Management. Am J Otolaryngol Head Neck Surg. 2023; 6(5): 1244.

In 2022 our group published a review of CT scans of 14 infants with CNPAS and 100 controls of comparable age to assess the anteroposterior length of the bony nasal septum and the width of the entire nasal cavity. We measured the widest part of the piriform aperture, the choana, and at positions 25%, 50% and 75% of the distance between the aperture and the choana [13]. We found that the nasal cavity was significantly narrower in patients with CNPAS compared to controls not only at the piriform aperture, but at points 25%, 50% and 75% along the entire length of the bony nasal septum. While the mean width of the choana was lower in cases than controls (11.7 mm *vs.* 13.5 mm), the difference was not statistically significant (p=0.4) [13]. Our results confirmed the findings initially published in a smaller study by Reeves et al. [14], and questions whether the traditional surgical approach that corrects narrowing of the piriform aperture but does not address more distal narrowing of the nasal cavity could be further optimized.

Management Options

The natural history of the development of piriform aperture is gradual widening with age until around the age of 10 years. The distance between the nasal maxillary processes is approximately 1.5 mm wider at 4 months of age than at 1 month of age in patients with CNPAS, and a proportion of cases can be managed conservatively on the expectation that symptoms will resolve with time [15]. Conservative treatment includes a constellation of nasal hygiene, nasal suctioning, adrenaline solution or other decongestants, humidification, nasal stents, steroid drops, oropharyngeal airway, and the use of a McGovern nipple for feeding [16,17]. Some authors advocate a 2-week trial of medical management before embarking on surgical intervention [5,18]. However, up to 70% to 80% of patients may eventually require surgery [9,19].

When to operate?

Belden et al. 1999 was the first to use CT findings to diagnose CNPAS and determined that narrowing of the piriform aperture to <11 mm was diagnostic in infants [4]. This definition is still in use today, although there is poor correlation between the degree of narrowing and the clinical picture. Moreddu et al. [5], proposed a management algorithm for CNPAS but did not specify an aperture width at which surgery should be considered. This is in keeping with the general principle that clinical symptoms are the most important factor guiding the decision to operate [11-13,16]. There have been several attempts to identify the degree of narrowing of the nasal aperture that predicts a need for surgery. A case study of 34 patients found a statistically significant association between piriform aperture size <6 mm and the need for surgical intervention (p=0.031) [9]. There was no association between surgery and other congenital anomalies, gestational age, or low birth weight. Conversely, a smaller case series of 10 patients found no difference in mean piriform aperture width in patients treated surgically (5.71 mm) or conservatively (4.83 mm), but noted that surgical revision was required in 37% of patients who had craniofacial dysmorphism [10]. Our group conducted an analysis of 38 patients (including 7 patients treated by the author and 31 patients from other published series) and found a significant difference in aperture width between surgically and medically treated patients (5.1 mm *vs.* 6.4 mm, p<0.05). An aperture width of ≤ 5.7 mm had 88% sensitivity and 73% specificity for identifying patients in whom surgery was required [19]. The rate of surgical intervention was almost 12-fold higher in patient with piriform aperture width ≤ 5.7 mm. Quantification of the risk of surgery provides useful information for paediatric teams and for parents and carers when considering the potential disease course. However, this information needs to be considered in the context of the severity of symptoms, the patient's response to conservative efforts, and the presence of comorbidities. Infants with very small apertures can be managed successfully with conservative measures [11,12,16].

Surgical options

Open surgery *via* a sublabial approach: Surgical intervention for CNPAS has traditionally been *via* sublabial approach to drill out the nasal aperture followed by nasal stenting for up to 1 month. Maintenance of patent stents over this period can be challenging for families and healthcare providers. Risks include those associated with anesthesia in the very young neonate and potential disturbances to maxillofacial and dental development. Columellar necrosis has been reported in one out of 20 surgical cases, development of synechia, and septal ulceration with septal perforation are other potential complications [18,20]. While the traditional surgical approach relieves acute obstruction at the piriform aperture, it does not address the narrowing of the nasal cavity beyond the aperture, potentially contributing to rates of surgical revision.

Nasal dilation: The use of dilatory techniques is technically easier and less invasive that drill-out procedures and has been associated with a substantially reduced length of hospital stay (3.5 days *vs.* 18.7 days, respectively) [21]. As yet, published experience using dilation procedures is limited to 10 patients (Table 1).

Gungor et al. [22], and Wine et al. [21], both reported different dilation methods in 2014. Wine performed nasal endoscopy with Hegar cervical dilators (2 mm to max 4 to 5 mm) on 4 patients. Two patients required re-dilation which was conducted as a minor procedure. Gungor performed balloon dilation of 5 min duration on each side followed by nasal stents for 12 days [22]. Sitzia et al. [23], used Hegar dilators to 4 mm followed balloon dilation to 8 mm in an infant. One side of the nasal cavity required re-dilation after 1 month.

The author has employed balloon dilation for surgically managed CNPAS since 2017. To date, 4 patients have been treated (Table 1). Approval for the evaluation of these outcomes was obtained from the institution. One patient (piriform aperture width 4.2 mm) failed repeated balloon dilation and required open surgery after 3 balloon dilations. This patient had multiple comorbidities. Three patients (mean width 5.4 mm) required a single balloon dilation only with successful resolution of symptoms until the last review at age 3, 5 and 6 respectively. Two patients had short term nasal stenting using a modified 3 mm ivory Portex endotracheal tube (Smith Medical, USA). These were the first 2 patients in our series and the stents were placed as part of our protocol to treat CNPAS based on the previous 7 cases who were treated by open drill-out [13].

Including this series there have been 10 patients in the published literature managed by dilation alone. Six patients (mean aperture width 4.9 mm) were managed by a single dilation procedure. Three patients (mean aperture width 4.4 mm) required a single repeat dilation, and only 1 patient (aperture width 4.2 mm) required an open drill out procedure after repeated dilation. Only 3 out of 9 patients who were successfully treated with dilation alone had a stent placed post dilation. Due to the small number of patients, statistical analysis has not been performed.

Balloon Dilation – A New standard of care for CNPAS? The nasal drill-out procedure has been used for decades to manage

Table 1: Summary of published experience using nasal dilation for surgical management of CNPAS.

Author	Case No.	Mean aperture width (mm)	Other anomalies	Procedure	Nasal stents	Outcome
Wine et al. [21]	1	4.2	4 patients had a median bony ridge of the palate, 3 had a triangular-shaped palate and a median central incisor, 1 had solitary median maxillary central incisor syndrome.	Direct laryngoscopy, rigid bronchoscopy, and nasal endoscopy with Hegar cervical dilators (2 mm progressing to 4-5 mm)	No	1 x repeat dilation
	2	4			No	
	3	4.9			No	1 x repeat dilation
	4	4.9			No	
Gungor et al. [22]	5	4.08	None	7 mm airway balloon dilated to 10 m wate for 5 minutes each side. Placement of nasal trumpets for 12 days	Nasal trumpets for 12 days	Patent nares 1 year later
Sitzia et al. [23]	6	4.01	Ogival palate, depression of the median palatine raphe	2 mm diameter Hegar cervical dilator, followed by 3- and 4-mm dilators and an 8 mm airway balloon	No	1 x repeat balloon dilation (1 side of the nasal cavity only)
Current series	7	5	None	6-7 mm balloon held at pressure for 60 seconds, repeated twice	Yes	1 x balloon dilation
	8	5.5	4q35 deletion, atrial and ventricular septal defects, common truncus arteriosus, ventriculoperitoneal shunt		Yes	1 x balloon dilation
	9	5.7	None		No	1 x balloon dilation
	10	4.2	Semi lobar holoprosencephaly, global developmental delay, cerebral palsy, visual impairment central diabetes insipidus		Yes for drill-out	3 x balloon dilations and triamcinolone injection. Sublabial approach and drill out.

CNPAS. Recent radiological analysis shows that the bony abnormality associated with CNPAS is not limited to the nasal aperture but affects the entire nasal cavity [13]. The long-term clinical implications of this narrowing on patient symptoms, medication requirements, and the frequency of additional surgical procedures later in life are not known. However, it seems reasonable to assume that correcting the narrowing of the entire nasal cavity is likely to have long term benefits over and above that of correcting the area of most acute stenosis.

Advantages and disadvantages of balloon dilation compared to open drill-out: Because of rarity of CNPAS, it is not feasible to conducted randomized clinical trials to test the efficacy and safety of different surgical strategies. Balloon dilation techniques offer an alternative treatment approach that has proven effective and well tolerated in the patients treated using this method to date. Balloon dilation is technically easier to perform than open drill-out, is not associated with the complications associated with open surgery and potentially prolonged use of stents. It addresses the narrowing of the whole nasal cavity which could have long term advantages in terms of development of nasal structures and the need for revision of procedures that concentrate on the piriform aperture. The procedure is associated with a shorter recovery period and duration of hospitalization, with the attendant benefits to direct healthcare costs and costs to families. The need for stents remains undefined, but the available (albeit limited) evidence suggests a reduced requirement for their use after balloon dilation.

The most clinically important disadvantage is the potential need for revisions. Revisions are easily and rapidly performed. However, some patients could require multiple re-dilations. There is therefore a need to balance multiple anesthetics versus the risks and benefits of the more invasive drill-out procedure that might provide a more definitive outcome for some patients.

Conclusion

As yet clinical experience with balloon dilation is limited. However, randomized controlled trials are not feasible to perform because of the rarity of CNPAS and all available data are based on case reports or case series. Including this series, there are 10 reported cases of the use of dilation to treat CNPAS. In six patients a single dilation procedure alone was required. Balloon dilation could be used as the first-line treatment for CNPAS in patients who require surgery. There is still a role for drill-out procedures in patients who fail repeated dilation.

References

1. Douglas B. The relief of vestibular nasal obstruction by partial resection of the nasal process of the superior maxilla. Plast Reconstr Surg. 1952;9(1):42-51.
2. Ey EH, Han BK, Towbin RB, Jaun WK. Bony inlet stenosis as a cause of nasal airway obstruction. Radiology. 1988;168(2):477-9.
3. Brown OE, Myer CM, 3rd, Manning SC. Congenital nasal pyriform aperture stenosis. Laryngoscope. 1989;99(1):86-91.
4. Belden CJ, Mancuso AA, Schmalfuss IM. CT features of congenital nasal piriform aperture stenosis: Initial experience. Radiology. 1999;213(2):495-501.
5. Moreddu E, Le Treut-Gay C, Triglia JM, Nicollas R. Congenital nasal pyriform aperture stenosis: Elaboration of a management algorithm from 25 years of experience. Int J Pediatr Otorhinolaryngol. 2016;83:7-11.
6. Papesch E, Papesch M. The nasal pyriform aperture and its importance. Otorhinolaryngol Head Neck Surg. 2016;1(4):89-91.
7. Nanni L, Ming JE, Du Y, Hall RK, Aldred M, Bankier A, et al. SHH mutation is associated with solitary median maxillary central incisor: A study of 13 patients and review of the literature. Am J Med Genet. 2001;102(1):1-10.
8. Visvanathan V, Wynne DM. Congenital nasal pyriform aperture stenosis: A report of 10 cases and literature review. Int J Pediatr Otorhinolaryngol. 2012;76(1):28-30.
9. Chakravarty PD, Sim F, Slim MAM, Patel N, Wynne DM, Clement WA. Congenital nasal pyriform aperture stenosis; our experience of 34 cases. Int J Pediatr Otorhinolaryngol. 2023;166:111491.
10. Gonik NJ, Cheng J, Lesser M, Shikowitz MJ, Smith LP. Patient selection in congenital pyriform aperture stenosis repair – 14-year experience and systematic review of literature. Int J Pediatr Otorhinolaryngol. 2015;79(2):235-9.

11. Shah GB, Ordemann A, Daram S, Roman E, Booth T, Johnson R, et al. Congenital nasal pyriform aperture stenosis: Analysis of twenty cases at a single institution. Int J Pediatr Otorhinolaryngol. 2019;126:109608.

12. Naina P, Dahiya V, Irodi A, Varghese AM. Congenital nasal pyriform aperture stenosis: Same CT dimensions, varied scenarios. Indian J Radiol Imaging. 2022;32(4):591-3.

13. Pollaers K, Herbert E, Giblett N, Thompson A, Vijayasekaran S, Herbert H. Re-thinking congenital piriform aperture stenosis: Modern imaging demonstrates narrowing of the full nasal cavity length. Int J Pediatr Otorhinolaryngol. 2022;158:111184.

14. Reeves TD, Discolo CM, White DR. Nasal cavity dimensions in congenital pyriform aperture stenosis. Int J Pediatr Otorhinolaryngol. 2013;77(11):1830-2.

15. Lin KL, Lee KS, Yang CC, Hsieh LC, Su CH, Sun FJ. The natural course of congenital nasal pyriform aperture stenosis. Laryngoscope. 2016;126(10):2399-402.

16. Abelardo E, Manuelian C, Devarajan D, Jones G. Conservative management of congenital nasal pyriform aperture stenosis. BMJ Case Rep. 2021;14(3):e241187.

17. Collins B, Powitzky R, Enix J, Digoy GP. Congenital nasal pyriform aperture stenosis: Conservative management. Ann Otol Rhinol Laryngol. 2013;122(10):601-4.

18. Van Den Abbeele T, Triglia JM, Francois M, Narcy P. Congenital nasal pyriform aperture stenosis: Diagnosis and management of 20 cases. Ann Otol Rhinol Laryngol. 2001;110(1):70-5.

19. Wormald R, Hinton-Bayre A, Bumbak P, Vijayasekaran S. Congenital nasal pyriform aperture stenosis 5.7 mm or less is associated with surgical intervention: A pooled case series. Int J Pediatr Otorhinolaryngol. 2015;79(11):1802-5.

20. Devambez M, Delattre A, Fayoux P. Congenital nasal pyriform aperture stenosis: Diagnosis and management. Cleft Palate Craniofac J. 2009;46(3):262-7.

21. Wine TM, Dedhia K, Chi DH. Congenital nasal pyriform aperture stenosis: is there a role for nasal dilation? JAMA Otolaryngol Head Neck Surg. 2014;140(4):352-6.

22. Gungor AA, Reiersen DA. Balloon dilatation for Congenital Nasal Piriform Aperture Stenosis (CNPAS): A novel conservative technique. Am J Otolaryngol. 2014;35(3):439-42.

23. Sitzia E, Santarsiero S, Tucci FM, De Vincentiis G, Galeotti A, Festa P. Balloon dilation and rapid maxillary expansion: A novel combination treatment for congenital nasal pyriform aperture stenosis in an infant. Ital J Pediatr. 2021;47(1):189.