

Balloon Dilatation and Rapid Maxillary Expansion: A Novel, Combined Treatment for Congenital Nasal Pyriform Aperture Stenosis in an Infant

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Case report

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Abstract

Background

Congenital nasal pyriform aperture stenosis (CNPAS) is a rare condition isolated or inserted in a multi-formative syndrome, which still encounters management difficulties. There are no specific treatment protocols and the use of traditional surgery represents an anachronism since cognitive assumptions and non-invasive or minimally invasive therapeutic innovations would currently be available. However, the rarity of the disease and the infantile context makes RCTs difficult to process.

Case presentation

We expose the case of a one-month-old caucasian male infant suffering from CNPAS. The patient presented to the Emergency Department of the Bambino Gesù Pediatric Hospital with a clinical picture characterized by nasal obstruction, noisy breathing, feeding difficulties and suspected sleep apnea. During hospitalization he underwent overnight pulse oximetry, airway endoscopy and maxillofacial CT with a final diagnosis of CNPAS with moderate obstructive sleep apnea. We treated him successfully through an innovative therapeutic strategy that required the collaboration of a team of ENT surgeons and orthodontists.

Conclusions

The combination of minimally invasive balloon surgery and the application of a palatal device may represent a successful strategy for patients with CNPAS, probably also applicable to other types of nasal bone stenosis. Future studies may allow the definition of treatment strategies of these pathologies, we hope in the context of novel practical guidelines.

Background

Nasal obstruction is a frequent cause of morbidity in infant. Although the most common cause is mucosal edema, other less common pathologies must be considered, including congenital nasal masses and congenital bony stenosis. Basing on the site of straitening, congenital bony stenosis may be classified in anterior (congenital nasal pyriform aperture stenosis, CNPAS), middle (midnasal stenosis) and posterior (choanal atresia) [1].

First described by radiologists in 1988 and one year later even by ENT specialists [2,3], congenital nasal pyriform aperture stenosis (CNPAS) is a rare condition characterized by a narrowing of the nasal cavity at the level of the pyriform aperture due to a medial positioning or an overgrowth of the maxillary process. Due to the frequent association with other craniofacial or neurologic anomalies, the incidence of isolated CNPAS is not clearly known [4-7]. The main pathogenetic theory explains CNPAS as an abnormal overgrowth of the nasal process of the maxilla associated with an overlapping of the lateral palatal shelves during the fusion of the secondary palate in the fourth month of gestation [8].

As in other causes of congenital nasal stenosis, morbidity of CNPAS may vary from a severe respiratory distress requiring immediate oral or orotracheal respiratory assistance, to a mild condition that may cause difficulty in proper feeding and consequent impairment in growth. Even in the mildest conditions, early diagnosis is advisable because neonates are obligate nasal breathers and they may not tolerate nasal obstruction for a long time. Severe nasal obstruction is usually identified at birth by the inability to pass the nasal tube. However, in some cases the condition remains undiagnosed until the onset of symptoms such as noisy breathing, tachypnea, cyanosis, desaturations, aspiration, difficult feeding and reduced growth. Acute respiratory distress and cyanosis are typically relieved with crying and tend to return with rest (paradoxical cyanosis).

Basic physical examination aims to assess a noisy nasal breathing, a reduced nasal airflow and any signs of chronic respiratory fatigue. If a congenital nasal obstruction is suspected, nasal endoscopy should be performed to orient the clinician. In CNPAS the passage of the 2.2 mm flexible laryngoscope is usually obstructed at the entrance of both the nasal cavities. Diagnosis of CNPAS must be confirmed by maxillofacial CT study of the axial and coronal sections. The transverse width of the piriform aperture normally ranges from 8.8 to 17.2 mm, but a measurement <11 mm taken at the axial level of the inferior meatus in a term neonate is considered diagnostic for CNPAS by most of the authors [9]. CT scans usually show a bony crest along the median axis of the hard palate (median palatal ridge) or a triangular shaped palate. Frequently CNPAS coexist with congenital midnasal stenosis, which is a rare condition due to unequal growth of the lateral wall of the nose or excessive in folding of the nasal septum. Other possible CNPAS-associated features found in CT may involve abnormal dentition; a central mega-incisor coexists in 50% of cases (solitary median maxillary central incisor syndrome, SMMCI). CNPAS with SMMCI may be part of the holoprosencephaly spectrum. Hypopituitarism may coexist as well. Magnetic resonance of the brain and genetic counseling are recommended to assess any neurological or syndromic abnormalities [10-12].

Several treatments have been proposed for CNPAS, basing on the severity of the nasal respiratory impairment. Mild conditions require medical treatment followed by a close watchful waiting. Nasal decongestants or corticosteroids, saline irrigations, anti-reflux medicaments, oxymetazoline hydrochloride, humidifiers, mouth breathing device (e.g., McGovern nipple) or non-invasive positive pressure ventilation are described as first level strategies in different studies [13, 14].

In severe conditions, or when no improvement is observed, current recommendations suggest a surgical correction of the stenosis. The traditional surgical technique consists of performing an osteotomy of the nasal lateral wall by drilling through a sublabial incision, with the aid of a loupe of microscopic magnification, followed by the placement of nasal stents to be kept in place for 5-28 days [15]. Some authors combined osteotomy with the reduction or resection of the inferior turbinates, without positioning nasal stent [16]. Although generally effective, surgery may cause considerable complications such nasal scarring, restenosis, nasolacrimal duct injury, anomalies of the facial mass development and abnormal dentition [17]. Furthermore, age-related surgical risk should be considered, especially when CNPAS is associated with other craniofacial anomalies. Therefore, most authors have been moving towards less

invasive approaches, such as balloon dilatation of pyriform aperture with or without subsequent nasal stenting [18, 19]. Rapid maxillary expansion (RME) and surgically assisted rapid palatal advancement (SARPE) were proposed to obtain an enlargement of the nasal space by means of a palatal distraction in infants with CNPAS or midnasal stenosis [20, 21]. In order to propose a rapid, long effective and safe treatment, we present a case of CNPAS infant treated through the combined use of the balloon dilatation and an innovative removable oral device for rapid palatal expansion that we called Neonatal Palatal Expander Plate (NPEP).

Case Presentation

A one-month-old caucasian male infant was referred to the Emergency Care Unit of Bambino Gesù Children Hospital (Palidoro, Rome, Italy) for noisy nasal breathing, nasal obstruction and suspected respiratory distress. The neonate was born at full term (38 weeks) after a normal pregnancy, from caesarean section, birth weight 3028 grams and APGAR score 6 due to respiratory distress and mild hypotonus, for which he was subjected to positive mask ventilation (PEEP) and then to CPAP for 20 hours, finally kept at high flow for 3 days. Neonatologists did not highlight any particular problem and the patient was discharged in apparent good condition. In the following days, the parents noticed an increased difficulty in keeping nasal breathing during the meal. Despite the fragmented supply of breast milk, his weight gain rate was still maintaining within normal limits, but he looked very irritable. Awakenings with crying were very frequent during sleep. Cyanosis and apnea had not been observed, but his nose seemed chronically congested despite the daily nasal irrigations and aspirations. The parents did not report any family history of congenital malformations.

At the time of our first evaluation his appearance was rosy and the oxygen saturation was 97% in ambient air. His crying was valid and he did not show signs of chronic respiratory distress. Specillation of the nasal cavities showed a significant impediment in the passage, particularly in the right nares. The palate had an ogival aspect, with a noticeable depression in correspondence of the median palatine raphe (Figure 1). Nasal endoscopy performed in wakefulness with a 2.2 mm pediatric flexible endoscope did not allow the nasal cavity to be exposed for the inability to advance with the instrument.

The infant was hospitalized to carry out diagnostic investigations. During the first night of hospitalization, he underwent an overnight pulse oximetry to assess the severity of respiratory distress in sleep. Overnight pulse oximetry showed an average oxygen saturation 97.5% and minimum oxygen saturation 80%, with an oxygen desaturation index (ODI) 8.4. These parameters were compatible with a moderate obstructive sleep apnea (Figure 2).

Head CT scan (Figure 3) showed a bilateral stenosis of the pyriform aperture, with an overall transverse diameter of 4.01 mm. Choanal diameters entered the normal range.

In consideration of the infant's symptoms, we planned a complete airways endoscopy in general anesthesia with simultaneous balloon-dilation of the pyriform stenosis. In the same operative setting, impression of the upper jaw was performed with addition silicon by a pediatric dentist (Figure 4, A and B).

Endoscopy confirmed the radiological appearance of stenosis of the anterior and middle third of the nasal cavities, with choanal patency and normal appearance of the larynx, trachea and bronchi.

The dilation of the stenosis was performed by passing a Hegar number 3 and subsequent dilation with an 8-millimeter balloon, resulting in an increased nasal respiratory space. During the procedure no complications occurred. Parents were instructed to carry out daily nasal irrigation with saline solution to remove secretions. Steroid nasal spray was administered once daily for one month in order to reduce nasal inflammation and discourage a restenosis.

The special removable palatal device which we called Neonatal Palatal Expander Plate (NPEP) was constructed by a dental technician on the upper arch cast. The device was a mucous anchoring plate made of transparent acrylic resin extended towards the oral vestibule with flanges to ensure good retention. In the center of the device, a 12 mm screw was placed in correspondence of palate median suture. To avoid any risk of suffocation, a safety wire was inserted through two holes in the canine region. The safety wire was a surgical silk suture thread (without needle) and about 70 centimeters long. The device was placed in the same setting of surgery and the parents were instructed to wear the device almost 14 hours/day and to turn the screw twice a day for 20 days, then once a day for 12 days. The total amount of the screw expansion was 11.5 mm (Figure 5).

Overnight pulse oximetry and nasal endoscopy were repeated after one and then after two months post-procedure. No clusters of desaturation occurred in both the records. The first control endoscopy was performed under general anesthesia. Patency of the piriform aperture appeared optimal in the right nasal cavity, while on the left there was a non-obstructing small bone spur of the lateral wall. We considered appropriate to perform a second balloon dilation only at left. The two-month endoscopic check-up was performed while awake in outpatient clinic and confirmed complete restoration of patency in both nasal cavities. The removable palatal expander was prescribed as retention period for 12 hours a day for about 30 days.

Discussion And Conclusions

Treatment of CNPAS as well as midnasal stenosis should be proportionate to the infant's clinical severity, in terms of respiratory distress and ability to feed. In mild conditions medical therapy may support the patient until somatic growth leads to spontaneous recovery. In unresponsive or worsening patients or in moderate/severe stages, anatomical correction of the defect is required. Despite its effectiveness, traditional surgical approach had many post-operative complications, especially in infants with multiple malformations.

Nasal balloon is a minimally invasive approach that allows to obtain an immediate opening of the bony stenosis due to the plasticity of maxillofacial bone and cartilage of the infants. Gungor et. al [18] successfully treated a 2-week-old neonate with balloon dilatation of pyriform aperture followed by nasal stenting for 12 weeks and topical steroids for 2 months. Because of the traumatic effect on the nasal mucosa and the poor tolerance in infants, some authors [19] performed the technique without a post-

operative nasal stenting, but they reported relapses due to a subsequent restenosis. Nasal stenting may be useful to stabilize the dilation, but it is difficult to manage in infants, in addition to the risk of damaging nasal mucosa and causing epistaxis.

In the last decades management of nasal obstruction with OSAS in children has benefited from the collaboration between dentists and ENT specialists. Research has been refined towards increasingly less invasive, rapid and effective long-term therapeutic strategies, with a significant reduction in morbidity. Rapid maxillary expansion (RME) is routinely used by orthodontists to increase the transverse palatal diameter when constricted, with benefit on nasal breathing by widening the nasal base [20]. Collares et al [21] treated a CNPAS infant applying a fixed REM device under general anesthesia. Although the remodeling of the maxilla in the infant takes less time than in the adult, a CNPAS infant may need an immediate restoration of the nasal patency and the number of days necessary for the active phase may lead to morbidity in moderate or severe obstruction. In fact, the initial risk of worsening respiratory distress due to oral space reduction need to be considered.

Surgically assisted rapid palatal advancement (SARPE) is a technique of palatal distraction that has been used to provide space for crowded maxillary dentition and in maxillary hypoplasia. A recent and successful application to a midnasal stenosis case was described by Graham et al [22]; they performed surgical dissection until exposing nasal floor before drilling the lateral nasal wall and finally they applied a fixed device.

In our experience, the ideal treatment for CNPAS - as well as for midnasal stenosis - would be to combine the mini-invasive approach with immediate nasal patency guaranteed by balloon dilation, with a stable result over time such as that resulting from maxillary expansion. The advantage of using a mobile device equipped with a safety cord found outside the mouth is to avoid a sedation or a general anesthesia to remove it. Furthermore, the application of screws in the maxilla is an invasive and not risk-free procedure.

In conclusion, the application of minimally invasive technologies and the collaboration between complementary branches determined the therapeutic success in this clinical case. We believe that a similar approach may be applied also to other types of nasal stenosis (e.g., mindasal stenosis). Further studies are needed to outline therapeutic strategies within shared protocols.

Abbreviations

CNPAS: congenital nasal pyriform aperture stenosis

SMMCI: solitary median maxillary central incisor syndrome

RME: rapid maxillary expansion

SARPE: surgically assisted rapid palatal advancement

NPEP: Neonatal Palatal Expander Plate

PEEP: positive-end expiratory pressure

CPAP: continuous positive airway pressure

Declarations

Ethics approval and consent to participate: The ethics committee of our organization approved the publication of clinical data for research purposes.

Consent for publication: Written informed consent for publication of clinical details and/or clinical images was obtained from the parent of the patient. A copy of the consent form is available for review by the Editor of this journal.

Availability of data and materials: All data generated or analysed during this study are included in this published article. We cited any publicly available data on which the conclusions of the paper rely in the manuscript including a persistent identifier.

Competing interests: The authors declare that they have no competing interests.

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Authors' contributions: "E.S. and S.S. analyzed and interpreted the patient data. E.S., T.F.M. and G.D.V. performed airways endoscopy and balloon dilatation. A.G. and P.F. designed and built the palatal device. E.S., S.S., G.A. and P.E. were the major contributors in writing the manuscript. All authors read and approved the final manuscript."

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Figures



Figure 1

Clinical evaluation before treatment showed an ogival aspect of the palate with a noticeable depression in correspondence of the median palatine raphe.

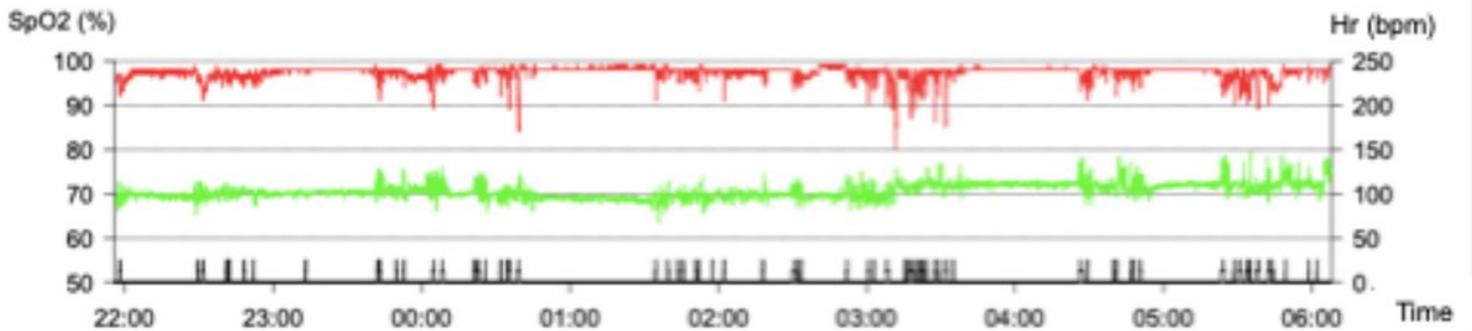


Figure 2

Overnight pulse oximetry of the patient performed before treatment showed clusters of desaturation and concomitant increase in heart rate. Parameters' analysis was conclusive for a moderate obstructive sleep apnea.



Figure 3

Pre-operative head CT scan showed pyramidal aperture stenosis. The measurement was taken at the axial level of the inferior meatus, as recommended by radiological guidelines.



Figure 4

On the left (A), impression of the upper jaw performed during the operative session. On the right (B), impression of upper jaw performed with in addition silicon.

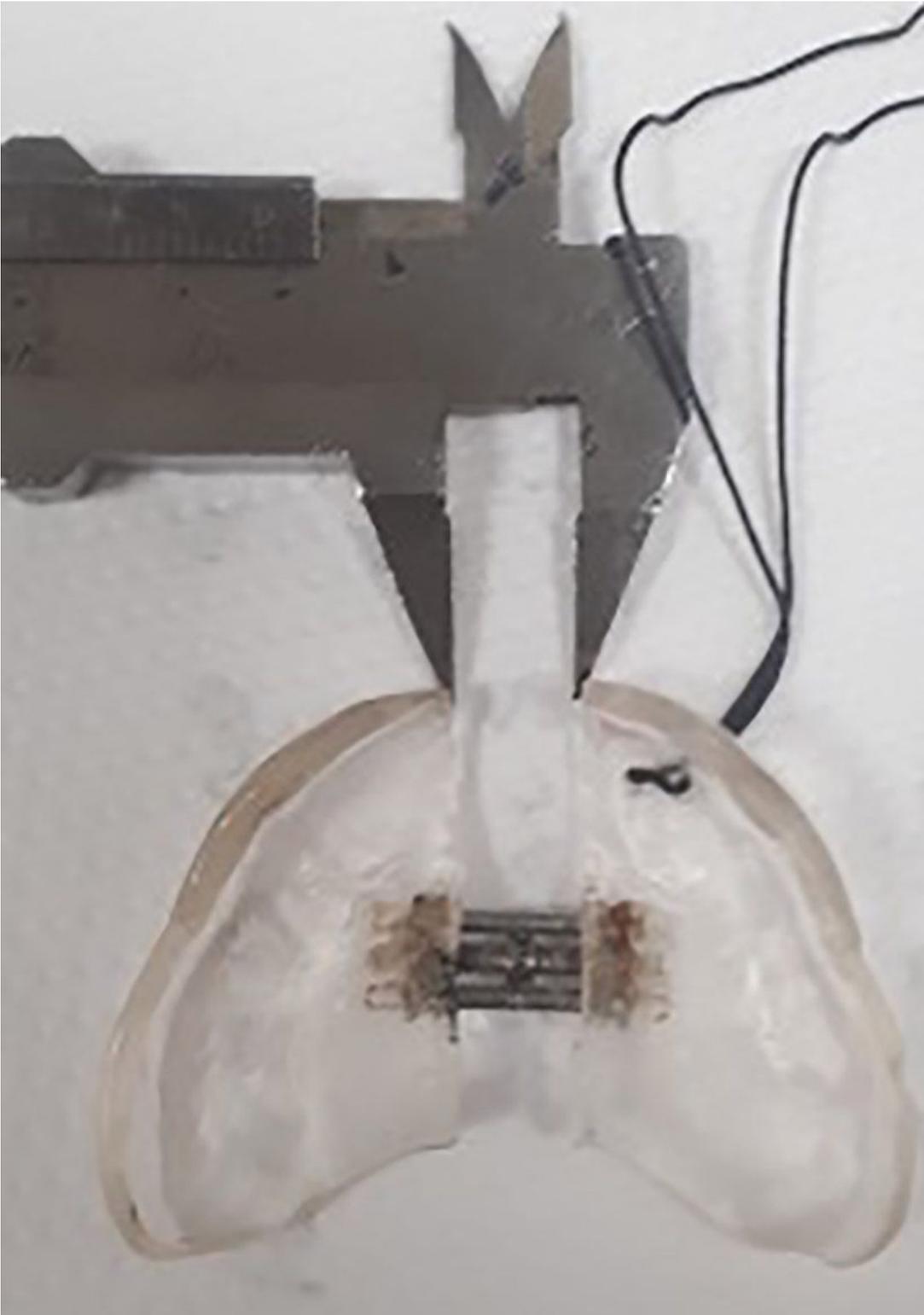


Figure 5

Neonatal Palatal Expander Plate (NPEP) made of transparent acrylic resin and a 12 mm screw in correspondence of palate median suture. The total amount of the screw expansion was 11.5 mm.