Case Report

Bilateral choanal stenosis in a craniosynostosis child: a case report

H. Y. Tan*, Noor Azrin M. Anuar, Halimuddin Sawali

Department of Otorhinolaryngology, Queen Elizabeth Hospital, Kota Kinabalu, Sabah, Malaysia

Received: 20 November 2018
Accepted: 10 December 2018

*Correspondence:
Dr. H. Y. Tan,
E-mail: klyhop@hotmail.com

ABSTRACT

Congenital bilateral choanal stenosis is a rare developmental condition and is highly associated with craniosynostosis syndromes. It can present with life threatening asphyxia. Diagnosis is made clinically using simple bedside tests and nasoendoscopy. Computed tomography of paranasal sinus confirms the diagnosis and facilitates pre-operative planning. Although the definitive treatment is surgery, the surgical options are based on involvement of unilateral or bilateral as well as bony or membranous. We report a rare case of bilateral choanal stenosis in a child with Pfeiffer syndrome who presented with severe respiratory distress at day 32 of life. Clinically, there was failure to insert suction catheter size 6 French through both nasal cavities. A high-resolution computed tomography (HRCT) of paranasal sinus confirmed the diagnosis of bilateral choanal stenosis. Endoscopic repair of the choanal stenosis was performed and stent was inserted for 14 days. Post-operatively, there was no evidence of restenosis after 3 months of clinical follow up.

Keywords: Choanal atresia, Choanal stenosis, Craniosynostosis syndrome, Pfeiffer syndrome

INTRODUCTION

Congenital choanal atresia is a rare condition which was first described by Roederer in 1755. The incidence is reported to be 1 in 5000 to 8000 live births and is twice more common in females. Bilateral congenital atresia are commonly associated with other congenital abnormalities namely CHARGE syndrome (coloboma, heart defects, choanal atresia, growth retardation, genital abnormalities, and ear abnormalities) and other craniosynostosis syndromes such as Crouzon, Pfeiffer and Apert syndrome. The deformity is made of 90% bony (30% pure bony and 70% mixed bony and membranous) and 10% membranous. Choanal stenosis is postulated to be minor forms of the choanal atresia or thin atresia segment perforated by early suctioning. Various definitive surgical techniques has been described and surgery is done via transpalatal or transnasal approach.

We described a rare case of bilateral choanal stenosis in a craniosynostosis child and its management modalities.

CASE REPORT

A term boy, with an uneventful antenatal history, was born via spontaneous vaginal delivery with a birth weight of 3.47 kg. Both parents are nonsanguineous and phenotypically normal. The APGAR score was 4, 8 and 10 at 1st, 5th and 10th minute respectively with 1 cycle of cardiopulmonary resuscitation and 1 cycle of positive pressure ventilation given in the immediate postpartum period. He was subsequently stable under supplementary 3 litres nasal prong oxygen and was admitted to neonatal ward. Upon examination, he was noted to have craniosynostosis with bilateral proptosis and hypoplastic maxilla (Figure 1). He was clinically diagnosed with Pfeiffer syndrome. He was then discharged after weaning off oxygen supplementation and establishing breastfeeding.

He presented again at day 32 of life with coryza and stertor with an episode of desaturation. Upon assessment by otorhinolaryngologist, noted congested nasal cavity
with inability to pass a 2.7mm 0° rigid scope, choanal atresia was suspected when Ryle’s tube size 6 French failed to pass through both nasal cavity. This was then confirmed by high-resolution computed tomography of cranium and paranasal sinuses, which revealed mixed bony-membranous bilateral choanal stenosis which was worse on the left side and features of craniosynostosis which was fusion of coronal, sagittal and lambdoid sutures resulting in turcephaly, hypoplastic maxilla and failure of formation of supraorbital ridges (Figure 2).

Child was then intubated for respiratory distress and underwent dilatation of bilateral choanal stenosis via transnasal approach under general anaesthesia. Coblator was used to address the membranous part of the stenotic segments and subsequently followed by dilatation using Bougie and Haegar dilators. A modified endotracheal tube size 3.5 mm, folded and a perforation fashioned at the bend, was used as a stent and placed in the nasal cavity with the perforation placed at the nasopharynx. The stent was secured with silk sutures. Regular saline drops were instilled into the stent with regular suctioning to prevent formation of crusting and blockage. Stent was then removed after 14 days postoperatively. As of follow-up at 3 months post-operatively, there was no recurrence of symptoms or any complications.

The child was scheduled for a series of further operations to correct his craniosynostosis.

DISCUSSION

Bilateral choanal stenosis is a medical emergency as neonates are obligate nasal breathers. Initial management is to maintain an adequate oral airway via McGovern nipple or oropharyngeal airway. Definitive management of choanal atresia and stenosis is by surgical correction. Various surgical methods have been described over the years; however, the optimal technique is still shrouded with controversies.

Endoscopic resection was first reported by Stankiewicz.5 Our patient underwent transnasal endoscopic resection with stenting and to date (3 months post-operatively) has favourable results. Nasal cavity is decongested and a 2.7mm 0° rigid endoscope is used to visualize the stenotic segment. The stenotic segment is dilated using power instruments and dilators. Heengerer reported 18% of patients who underwent transnasal resection developed restenosis requiring revision surgery.6 Transpalatal resection offers good exposure to the choanal abnormality, posterior vomer and lateral nasal wall. A U-shaped flap is raised on the palate, conserving the greater palatine arteries. The choanal abnormality can be removed under direct visualization. Ferguson reported 90% of patients treated with transpalatal resection did not require any further surgery.7 However, it is associated with a high rate of crossbite abnormalities.8 Other possible complications are palate flap necrosis or fistula.

Postoperative stenting of nasal cavity has routinely been part of the management to maintain patency of nasal cavity and restore nasal breathing. Various methods of stenting have been described by multiple authors. Most common material used is modified endotracheal tube in a similar fashion as our patient.9-11 Modified endotracheal tube has the required rigidity to maintain patency. However, care has to be taken during refashioning to ensure no sharp edges are present that might cause mucosal injury during placement and removal. Other
materials used are silastic stents and Teflon stents, both which are softer.\textsuperscript{10,12} Softer materials are believed to reduce the risk of granulation tissue and hence restenosis.\textsuperscript{10} Stents are regularly instilled with saline and suctioned to prevent crusting and blockage.

Traditionally stents are left in-situ for 6-8 weeks for reepithelization of the neochoana.\textsuperscript{3} However, Pasquini suggested a shortened stenting period and usage of soft stents reduces the risk of granulation tissue formation and postoperative infection, hence, reducing the risk of restenosis.\textsuperscript{10} In our case, stent was kept in-situ for 2 weeks with favourable outcome.

However, stents are not without its risk, such as patient discomfort, septal or columellar necrosis, nasal and paranasal infection and intranasal synechiae. In view of the morbidities associated with stenting, several studies argued that postoperative stenting is not a must.\textsuperscript{11,13} Their argument being that with the advancement in endoscopic surgery, surgeons have a better view of the posterior nasal cavity. Removal of atresia segment can be done with more precision, hence, reducing the need to protect the denuded tissue from exuberant granulation tissue and synechiae formation. Nevertheless, the literature of both for and against stenting is limited and consists of case series. Further multicentre and well-controlled study is needed to produce a conclusion.

CONCLUSION

Bilateral choanal atresia and stenosis are medical emergencies. Transnasal endoscopic resection is the technique of choice owing to its favourable outcome and lower morbidity as compared to transpalatal approach. However, the evidence of postoperative stenting is inconclusive and further study is needed.

Funding: No funding sources
Conflict of interest: None declared
Ethical approval: Not required

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