

Bilateral Congenital Choanal Atresia in an 11-year-old Girl: An Unusual Presentation

SH Chandrashekharayya, Prakash S Handi, Mohammed Riyas

ABSTRACT

Congenital choanal atresia (CCA) is characterized by narrowing of posterior nasal apertures in newborns present with nasal obstruction and respiratory difficulty. Although unilateral obstruction goes unnoticed at birth, bilateral case is an emergency situation because newborn is an obligate nasal breather. Here, we present an unusual case of bilateral choanal atresia in an 11-year-old girl, which is a rare finding. Surgical correction was done by transnasal approach using endoscopes and otological drill.

Keywords: Choanal atresia, Bilateral, Transnasal endoscopic approach.

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INTRODUCTION

Congenital choanal atresia (CCA) is a rare anomaly of posterior nasal cavity causing nasal obstruction in newborns with incidence of 1 in 7,000/8,000 live births. Female babies are affected more compared to males with predilection for right side.¹ First reported by Roederer in 1755, Otto gave it a status of anatomical disorder. Emmert corrected it surgically for the first time in 1851.²⁻⁴ Nearly 90% of the defects are bony type, 10% are membranous. However, with advancement in imaging studies, recent reports say that 30% are purely bony, 70% are mixed bony-membranous type of anomalies.⁵ CCA can occur as an isolated entity or may have association with other defects as in CHARGE syndrome (coloboma, heart disease, choanal atresia, retarded development, genital hypoplasia and ear anomalies).¹

Surgical treatment is the accepted modality of management. Various approaches have been used to repair choanal atresia. Among them transpalatal, transnasal, sublabial transnasal, transantral and transseptal approaches have been described along with historical perspective by Pirsig.⁶ With development of endoscopes and powered instruments, transnasal endoscopic repair has become the most successful and accepted modality of treatment.⁷⁻⁹ Postoperative stenting is a controversial issue, some authors favoring it and some against it. Some of the studies have shown benefits of stent placement.¹⁰

CASE REPORT

An 11-year-old girl presented to us with a history of bilateral nasal obstruction and excessive nasal discharge. On clinical examination patient had all features of adenoid facies *viz* open mouth, vacant look, high arched palate and pinched nose. On examination of nose, bilateral mucoid nasal secretions with bilateral hypertrophy of inferior turbinate were noted. The clinical diagnosis of adenoid hypertrophy was done and patient was posted for diagnostic nasal endoscopy to confirm the diagnosis, as the patient did not have any tonsillar symptoms. On diagnostic nasal endoscopy to our surprise patient had complete obstruction of both posterior nasal openings with no communication to the nasopharynx (Fig. 1). On further careful history taking, grandmother revealed that baby was born in home and had difficulty in breathing as well as feeding, they took a help of local midwife who advised them to feed the baby with cup and spoon. Patient had mouth breathing since then.

Patient was subjected to computerized tomography (CT) of nose and paranasal sinuses. CT revealed bilateral choanal atresia, bony type, with thickening of vomer in midline. Patient was taken up for transnasal endoscopic repair under general anesthesia. Using zero degree endoscope, secretions were cleared from the nose and decongestion was done, then a mucosal flap elevated to expose the bony atretic plate. It was punctured with a trocar at the weakest point taking care not to injure the posterior nasopharyngeal wall. Then opening was widened using an otologic drill and curettes



Fig. 1: Endoscopic picture showing atretic plate

taking care not to injure vestibular skin. Widening was continued medially and inferiorly. Thick posterior vomer removed using kerrison punch (Fig. 2). Widening was continued until an opening of 5 mm was obtained, raw bony surface was covered with mucosal flaps. Same procedure was repeated on other side and a satisfactory opening was achieved (Fig. 3). Soft silicon stents were placed and secured to the vestibule by stitch (Fig. 4).

Stents were removed after 48 hours. Patient was advised saline nasal douche, decongestant and steroid nasal drops. Diagnostic endoscopy was done after 1 week, 1 month and third month; patency was satisfactory.

DISCUSSION

Articles in excess of 300 have been published on CCA, this reflects the difficulty and controversy related to the management of CCA. An ideal technique for correction of the condition is one which offers easy access, good visualization, reduced operative time with low recurrence

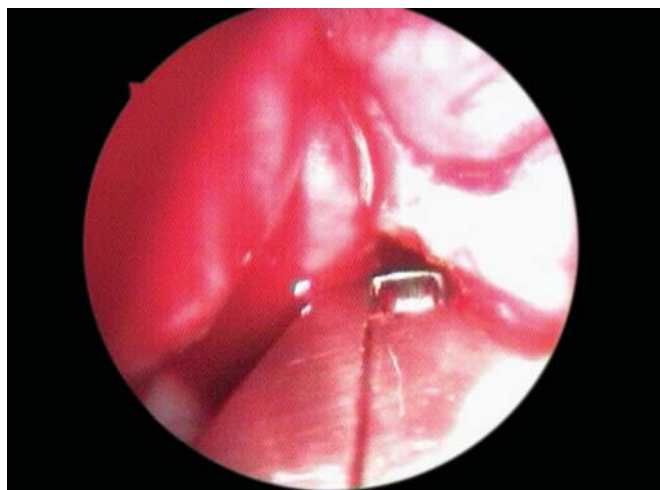


Fig. 2: Punch being used to remove thick vomer



Fig. 3: Endoscopic picture showing newly created choanal opening. Adenoids can be seen



Fig. 4: Postoperative stenting

rate. With advent of the endoscopes, transnasal endoscopic approach has become most accepted technique.¹¹

Bilateral CCA presents as a medical emergency as newborn is an obligate nasal breather. Ability to open mouth in response to nasal obstruction develops only after few weeks. Newborn can breathe through mouth only during crying or if any artificial airway is inserted into the mouth. In our case patient had difficulty in breathing at birth and difficulty improved while crying, but how the baby survived, until the reflex opening of mouth to nasal obstruction developed remains unclear. Probably the knowledge of local midwife who managed the baby during this period needs to be studied. Bilateral CCA presenting late in life is a very rare phenomenon with only few cases being reported in literature.³

Among the various approaches for treatment of choanal atresia, transpalatal route was widely used before the advent of endoscopes. Management by transpalatal route is associated with longer operating time, more blood loss and long recovery time. There is a risk of palatal perforation also.¹² We managed our case with transnasal endoscopic technique and stenting for a short period. This technique gave us satisfactory results.

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ABOUT THE AUTHORS

SH Chandrashekharayya (Corresponding Author)

Associate Professor, Department of ENT, S Nijalingappa Medical College and HSK Hospital and Research Center, Bagalkot-587102 Karnataka, India, Phone: 9448580785, e-mail: drcschent@rediffmail.com

Prakash S Handi

Assistant Professor, Department of ENT, S Nijalingappa Medical College and HSK Hospital and Research Center, Bagalkot, Karnataka India

Mohammed Riyas

Postgraduate Student, Department of ENT, S Nijalingappa Medical College and HSK Hospital and Research Center, Bagalkot, Karnataka India