Case Report

UNUSUAL CHOANAL ATRESIA IN CHILDREN

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ABSTRACT

Choanal atresia is a developmental failure of the nasal cavity to connect with the nasopharynx. The cause is either unilateral or bilateral nasal obstruction. Bilateral choanal atresia is a very rare condition and diagnosed soon after birth because neonates are obligate nasal breathers. Delayed diagnosis can cause death due to respiratory distress. The objective of this study was to report bilateral choanal atresia in 5-year-old boy who survived by breathing through his mouth using a simple tool made by his parent. It was a very rare condition because the posterior nasal cavity was covered by medial wall malformations of both maxillary sinuses which fused with the posterior nasal septum. The surgery method for this patient was posterior septectomy with transnasal endoscopic to create a neochoanae. The post-operative patient could breathe normally through his nose. Six weeks later, the nasal endoscopic evaluation indicated the patient's patent neochoana.

Keywords: *Bilateral choanal atresia; children; posterior septectomy; human and health*

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Highlights:

1. Unusual CA in a 5-year-old boy with paranasal sinuses malformation in the ORL-HNS.
2. Transnasal endoscopy posterior septectomy was successfully done.

INTRODUCTION

Choanal atresia (CA) is a rare case, with only 10 cases reported in literatures (Anajar et al. 2007; Verma et al. 2016; Kars et al. 2020; Tatar et al.2012). Choanal atresia is a failure in connecting the nasal cavity to the nasopharynx. It occurs in 1:8000 of live births, more prevalent in women than men, 2:1 respectively (Sutikno & Thaufiqurrakhman 2021).

Choanal atresia is a relatively uncommon but well-recognized condition characterized by the anatomical closure of the posterior choanae in the nasal cavity. The posterior choanae anatomic boundaries include the undersurface of the body of the sphenoid bones superiorly, the medial pterygoid lamina laterally, the vomer medially, and the horizontal portion of the palatal bone inferiorly. The actual narrowing could be caused by one of the abovementioned bony components. Correctly identifying and addressing the point of obstruction is the key to surgical success.

teristics of CA follow a "2–1" rule, namely the ratio of unilateral to bilateral CA, female to male, and the right-sided to left-sided CA (Hengerer et al. 2008). It is a very rare condition that causes a blocked nose, due to the failure of posterior choanal formation. Roederer first described the discovery of this disease in 1755 (Flake & Ferguson 1964). Emmert et al. were the first to demonstrate the use of curved trochar in transnasal repair of bilateral CA in a 7-year-old boy (Kwong 2015). This malformation occurs in 1 out of 5,000-7,000 births with a female and male ratio of 2:1. Neonates are obligate nose breathers so bilateral CA results in fetal severe respiratory distress (Leung et al. 2014).

The diagnosis is confirmed by nasal endoscopy and CT scan. This malformation requires immediate treatment. Repair of CA can be done with transnasal puncture technique, transpalatal, or transnasal endoscopy (Kwong 2015).

This was a case report of a male neonate with CA that survived his 5 years of life and underwent repair with posterior septectomy technique using a transnasal endoscopy approach. Surgery for CA is performed under general anesthesia. A self-retaining nasal speculum is used to expose the nasal cavity and the atretic plate (Balasubramanian 2015). The case in this study was very interesting because of the unusual anatomical malformations of the patient's nasal cavity.

CASE REPORT

A 5-year-old boy came to the outpatient ORL-HNS Department of Dr. Soetomo General Academic Hospital, Surabaya, Indonesia, on Ma 3, 2017, with nasal obstruction since birth, came out with clear and dilute liquid from both nostrils, distorted odor, and widening nose (Figure 1).

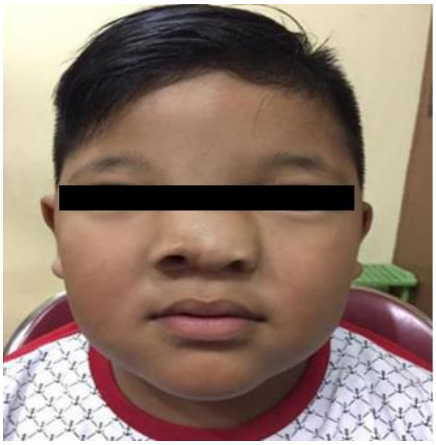

Figure 1. Clinical appearance of the patient.

From history-taking, there was no problem during pregnancy and delivery. The patient was born at a primary medical service in Maluku and got helped by a midwife. After the birth, the patient was not crying and had cyanosis with no spontaneous breath from the nose. The patient's father put his finger within the baby's mouth to support breathing. To replace his father's fingers, he made rolled gauzes (Figure 2). Then the patient was consulted to higher medical service and obtained oxygen support. When he was 3 months old, a pediatrician tried to insert a nasogastric tube but failed. The pediatrician suggested performing another diagnostic at a higher medical service in Java. In the ORL-HNS Department of Dr. Soetomo General Academic Hospital, the patient was diagnosed with CA and Congenital Talipes Equinovarus (CTEV). The previous doctor decided to correct CTEV first and then the choanal atresia when the patient was older.

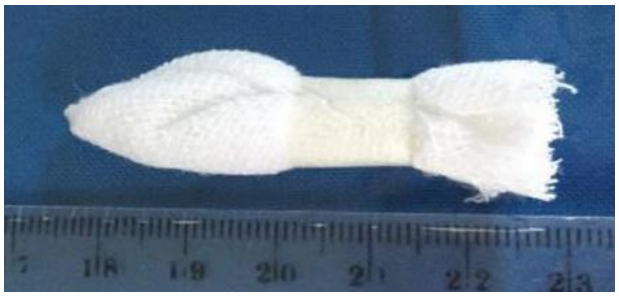

Figure 2. McGovern nipple was made personally by the patient's parents.

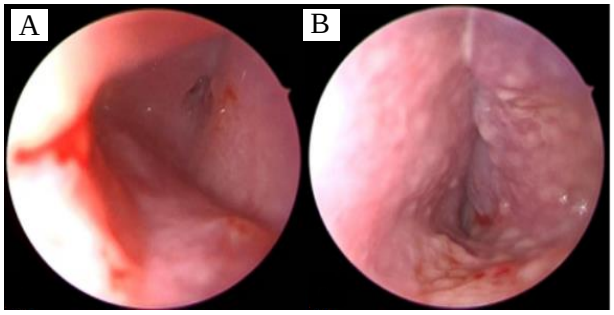

Figure 3. Pre-operative nasoendoscopy of right (A) and left (B) nasal cavity.

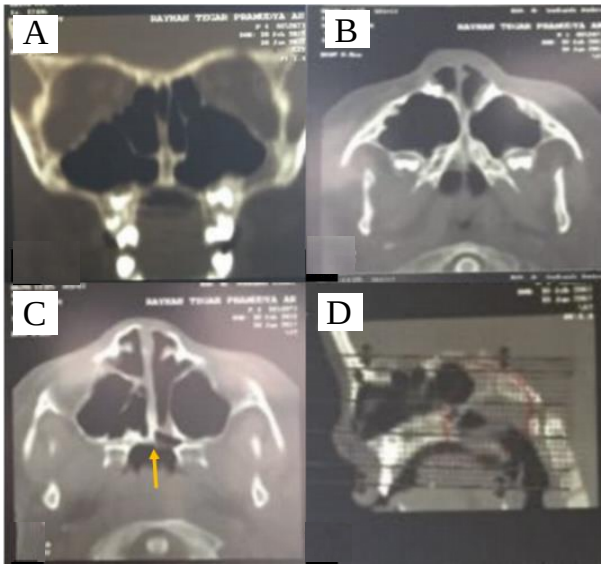

Figure 4. Paranasal sinuses CT scan. Coronal (A), axial (B, C), and sagittal views show bone-type choanal atresia with true choanal behind the malformation paranasal sinuses (yellow arrow).

From the physical examination, anterior rhinoscopy of the nasal cavity appeared narrow. Right, left middle, and inferior turbinates were not found. Medial walls malformation of both maxillary sinuses that merge with a posterior nasal septum in medial line indicated no defect of the palate. Pre-operative nasoendoscopic exploration confirmed the anterior rhinoscopy findings (Figure 3).

The CT scan result on January 10, 2017 from Soebandi Hospital when the patient was 5 years old showed that both maxillary antrum medial walls merged to nasal septum with the entire sinus walls intact. The structure covered the true choana on the posterior aspect. This finding supported the claim of bony type bilateral CA with paranasal sinuses malformation (Figure 4).

The surgery was done in two steps. First, surgery was performed on July 28, 2017 to break down both left and right medial wall maxillary sinuses. We made a wider passage by drilling the thick nasal floor and reducing the

malformed mucosa on both nasal cavities in the next surgery (Figure 5).

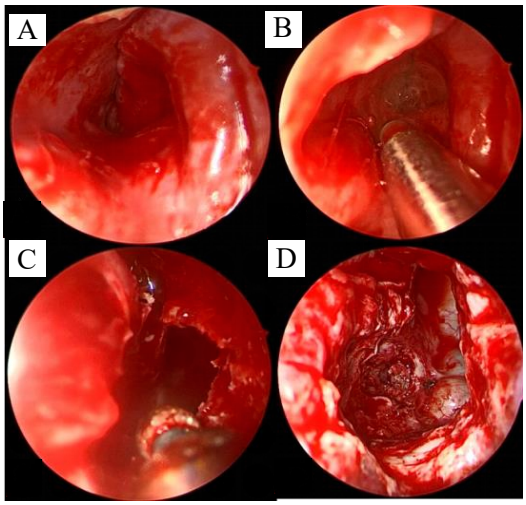

Figure 5. First surgery for malformed nasal floor(A). In the next step, we made a wider passage by drilling malformed nasal floor (B) and breaking down both maxillary sinuses (C). The nasal cavity (D) became wider.

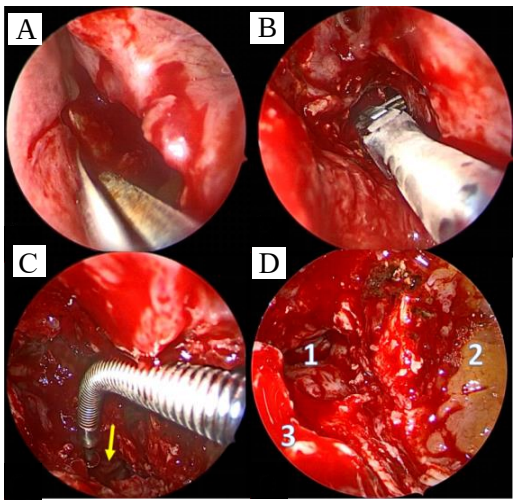

Figure 6. The second surgery used the posterior sepctectomy technique (A, B), and neochoana creation (C). The surgery was done on both sides of the posterior nasal cavity and the maxillary antrum, resulting in a single sinus cavity connected to the neochoana (D): 1. Velopharyngeal valve, 2. Left nasal cavity and maxillary anthrum, 3. Right nasal cavity and right maxillary anthrum.

The second surgery was performed on December 8, 2017. We did posterior septectomy with the incision of the posterior septal mucosa, then the septal mucosa was separated from the vomer. After the bone was exposed, the posterior septal bone was encrusted and extracted, making the left and right maxillary antrum, sphenoid, and posterior nasal cavity became single sinus cavity. This procedure created a neochoana. Then the neochoana was optimally expanded with "mushroom" to prevent restenosis. The surgery result was the creation of a neochoana that connected left-right nasal cavity (Figure 6). Restenosis prevention was performed by giving systemic corticosteroids for up to two weeks post-therapy.

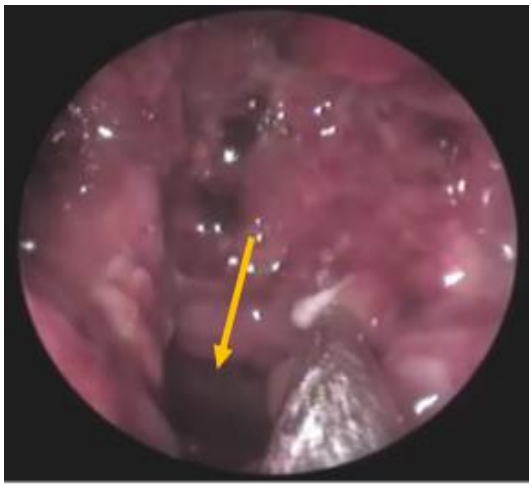

Figure 7. Nasoendoscopy showing a patent neochoana (yellow arrow).

A week later, the patient said in the first surgical follow-up that there was no nasal obstruction. From nasoendoscopy evaluation, hard and loose crust were found. In the second surgical follow-up another week later, the patient still had no nasal obstruction. The nasoendoscopy evaluation found that the neochoana looked patent and no sign of restenosis (Figure 7). The patient could not come to the next surgical follow-up at the hospital six months later, but sent a video that showed he could breathe normally through his nose.

DISCUSSION

Choanal atresia is a very rare malformation with an incidence of 1 in 5,000-7,000 births, with 50% being bilateral CA cases (Elumalai & Jeyapaul 2016).In the case of bilateral CA,patients can have acute respiratory distress with intermittent cyanosis.Feeding difficulty and choking can be the initial alerting progressive airway obstructions because these indicate an inability to feed and breathe simultaneously. Neonates with bilateral CA can also be present with a history of multiple failed extubating attempts, especially in secondary airway issues (Elumalai & Jeyapaul 2016). The incidence of bilateral CA in Indonesia has not been widely reported. In Makassar, Indonesia, a 16-day-old girl has been reported with bilateral CA (Perkasa 2013). At Dr. Soetomo General Academic Hospital, Surabaya, Indonesia, there was only one case of bilateral CA throughout 2017 which is reported in this paper, a boy who was able to survive

until 5 years of life without prior specific medical treatment.

Choanal atresia is a malformation, which 41% to 72% appears with no syndromic facial disorders, such as curved ceiling, cleft lip, and auricular deformity. Choana atrophy is associated with chromosome-based syndrome components, such as coloboma, heart deformity, choana atresia, stunted growth and development, genital abnormalities, and ear disorders (CHARGE), while Treacher Collins and Tessier syndromes are present in 4% of the cases (Bovo et al. 2017, Maruvala et al. 2017, Saitabau et al. 2018, Wormald et al. 2016). What the patient in this study had was bilateral CA with a facial deformity and congenital anomaly of lower extremities of CTEV.

Neonates with bilateral CA are born with acute respiratory distress with rapidly improving intermittent cyanosis when crying because neonates are obligate nose breathers for up to 4 to 6 weeks of life (Jašić et al. 2016, Gupta & Kour 2017). The typical clinical finding is cyclic cyanosis, blue spells, and respiratory difficulty that worsen during feeding or when a child falls asleep with their mouths closed (Gupta & Kour 2017). The timing of surgery for bilateral CA must be guided by clinical tolerance of the nasal obstruction and the presence of associated abnormalities. Normally, children with bilateral CA undergo surgery within the first week (Moreddu et al. 2018).

McGovern nipple is a non-surgery medical tool used for treating this condition. If it fails, the use of the endotracheal tube and even tracheotomy can be a definitive airway for patients until choanoplasty is successfully performed. The airway is kept patent by an oral airway. Soft rubber nipple can be used with the tip of the nipple being cut and put into the neonate's mouth (Jašić et al. 2016). Unlike the other cases that get medical attention, our patient succeeded in going through the ONB phase without medical treatment. This could happen because he used a simple rolled gauzes as his personal McGovern nipple. It supported him to survive without breath distress until we performed the surgery.

There are two types of CA, membranous and bony type. It can be classified as unilateral or bilateral CA. The diagnosis of bilateral CA can be suspected if a neonate is born with respiratory distress accompanied by a nasogastric catheter that cannot enter the nasopharynx through the nose on both sides (Leung et al. 2014, Wormald et al. 2016). Clinical examination with nasoendoscopy and CT scan are the gold standards for the diagnosis (Pokharel et al. 2013, Dharmaputri et al. 2017). Endoscopic examination of the nasal cavity should be performed with a flexible nasopharyngoscope or with a rigid rhinoscope. This examination makes it possible to confirm the diagnosis of atresia whether it is unilateral or bilateral and to assess the condition of the nasal cavity. In this case, the patient was diagnosed with bilateral bony type CA with a very rare malformation of paranasal sinuses.

Various operation techniques can be done, such as transnasal puncture, transpalatal, or transnasal endoscopic repair. The mean success rate with transnasal endoscopic repair was 85.3% out of 238 cases in a meta-analysis of 20 studies (Durmaz et al. 2008). Risk factors for restenosis include nasopharyngeal reflux, gastroesophageal reflux, age <10 days (associated with limited visualization in noses of neonates and limited resection of the vomer), bilateral CA with purely bony atretic plate, and the presence of associated malformations (Kim et al. 2012).

Post-operative restenosis remains a common complication of the endoscopic CA repair (Asma et al. 2013). Frequent post-operative use of nasal saline irrigation and periodic endoscopic surveillance or second look procedures can improve the primary repair success and reduce the rate of restenosis (Rodriguez et al. 2014). Operation choice is based on the CA type. These techniques allow the restoration of nasal patency using the natural airway. Without detachment of the palatal fibromucosa, optical magnification of the operated area allows precise excision of the atretic plate.

Superior visualization of the atretic plate and the surrounding intranasal anatomy provides accurate removal of obstructing soft tissue and bone (Moreddu et al. 2018, Patel & Carr 2018). Endoscopic approaches lead to short operative times, minimal bleeding, early feeding after surgery, and short hospital stay (Moreddu et al. 2018).

Exposed posterior vomer and lateral pterygoid lamina is elevated by making a mucosal incision and a mucosal flap. A diamond burr on an angled handpiece is used to drill the atretic bony plate. It is perforated at the junction of the hard palate and the vomer. Incidentally, this is the thinnest part of the atretic plate. To improve visualization, the inferior turbinate can be out fractured or even be trimmed. After drilling, care is taken to preserve the mucosal flaps. A silastic stent is placed into each nostril passing through the drilled neochoana. This helps in reducing the incidence of restenosis. The stent is kept in place for at least 6 weeks (Kastrati et al. 1997). The first procedure was demonstrated using endoscopic techniques for transnasal CA repair by Stankiewicz (1990). Since then, there have been numerous reports on different modifications of the endoscopic techniques.

After adequate nasal decongestion, a 2.9- or 4.0- mm Hopkins rigid endoscope is introduced in the nostril to visualize the atretic plate. A laterally based mucosal flap is then raised to expose the bony part of the atretic plate.

The thinnest section of the atresia, usually found at the junction of the hard palate and vomer below the tail of the middle turbinate, is the ideal point of entry into the nasopharynx. The posterior bony septum (i.e., vomer) was removed to create a "neounichoana" (Ibrahim et al. 2010) using a powered instrument. No more than one-third of the bony septum is removed to prevent the potential adverse effect on the nasal growth centers. Avoiding bone ridges and covering the exposed bone surface with mucosa is essential to prevent post-operative restenosis (Rodriguez et al. 2014). For the patient in this case, who had a very rare malformation type, the use of posterior septectomy with transnasal endoscopy surgery brought a satisfying result. During surgical follow-ups that were done one week until six months after surgery, the patient had no complaints and could breathe through his nose normally.

Strength and limittion

The case provides clear and specific information about choanal atresia. It is a rare condition that can be life-threatening if not diagnosed early, which is to report a case of bilateral choanal atresia in a 5-year-old boy who survived with the help of a simple tool made by his parent and the surgery was successful in creating a patent neochoana.

CONCLUSION

This case reported unusual CA in a 5-year-old boy with paranasal sinuses malformation in the ORL-HNS Department of Dr. Soetomo General Academic Hospital, Surabaya, Indonesia. After being diagnosed, the patient underwent repair with posterior septectomy technique using a transnasal endoscopy approach. Transnasal endoscopy posterior septectomy was successfully done. In the evaluations until six months after surgery, the patient had no complaints, distorted odor, widening nose, clear and dilute liquid coming out of both nostrils. He also could breathe normally through the nose.

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Conflict of interest

None.

Funding disclosure

None.

Author contribution

YSI and IK contributed the design of this study, manuscript preparation and drafting, write the manuscript and revise it, and also supervision

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