

Case Report: Revision Choanoplasty in a 15-year-old Teenager with Bony and Membranous Type of Bilateral Choanal Atresia

Case Report

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ABSTRACT

Introduction: Choanal atresia is a rare congenital disorder with an incidence of 1:5000 to 1:9000 births and is rarely found in teenager. The incidence is slightly higher in female than male. Bilateral choanal atresia in newborns caused respiratory distress, as they were an obligate nasal breather. They were rarely survived without emergency treatment. A transnasal choanoplasty was the mainstay treatment for choanal atresia.

Case report: a 15-years-old male reported with bony and membranous type of bilateral choanal atresia. He initially complained of nasal obstruction and rhinorrhea. There was a history of respiratory distress that was alleviated with crying when he was born. The nasal endoscopic and CT-scan examinations showed septal deviation and bilateral choanal atresia of bony and membranous type in the right and left nasal cavity, respectively. The patient underwent septoplasty before choanoplasty. A stent was used to prevent stenosis. However, one month after choanoplasty, a total restenosis of choana was observed in the right nasal cavity and followed by mild stenosis in left nasal cavity. Further revision of choanoplasty with steroid diluted tamponade and steroid injection in the choanal circumference were done to repair the restenosis. The post-operative evaluation showed a mild restenosis with a patent choana.

Conclusion: Treatment of bilateral choanal atresia with a transnasal endoscopy approach facilitates choanoplasty with minimal complications. However, complications in the form of recurrent stenosis after surgery remains to be a challenge.

Key Words: Bilateral Choanal Atresia, Choanal Atresia.

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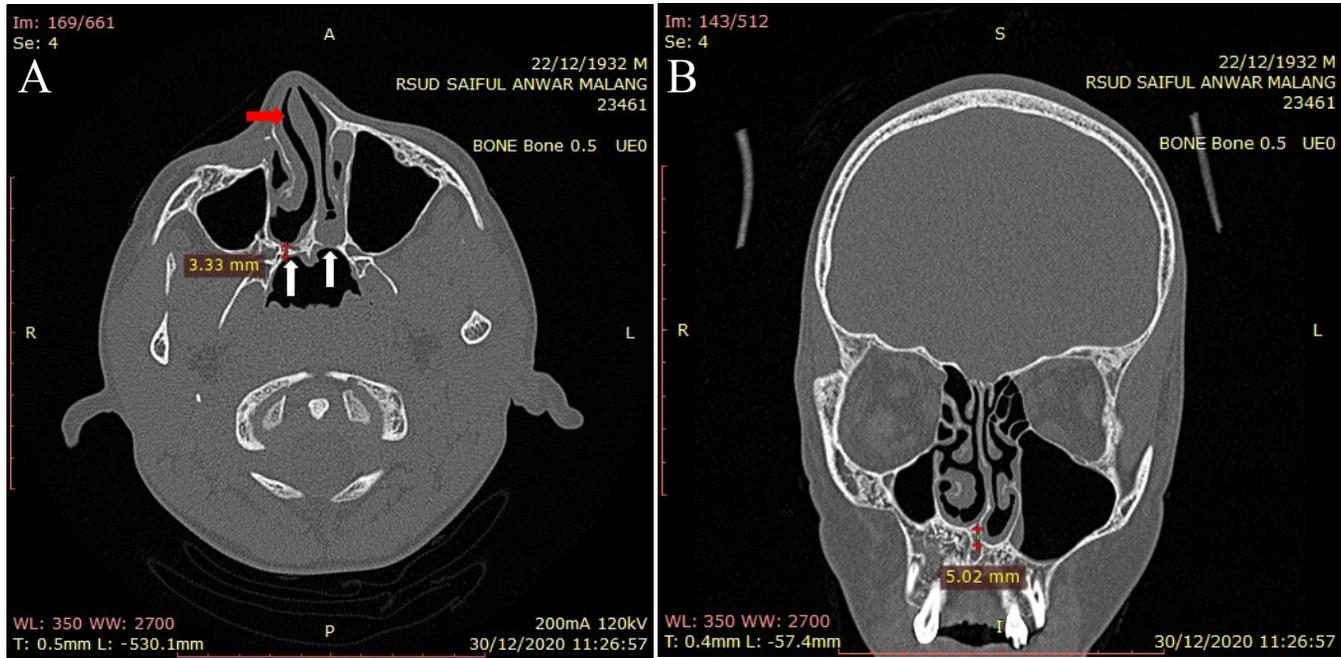
CASE REPORT

A 15-year-old boy presented with nasal obstruction with profuse watery rhinorrhea and rhinolalia. He cannot breathe through nose. He had history of respiratory distress which was alleviated with crying when he was born. Other apparent congenital malformation of palpebral coloboma was observed. He presented neither growth or developmental disorder. There was no heart defect, genitourinary and ear abnormalities. A catheter insertion test was failed to reach nasopharynx suggesting an obstruction. Nasal endoscopy examination showed total choanal obstruction with septal deviation. The CT-scan result showing bilateral choanal atresia with bony type atresia in the right side and membranous type atresia in the left side. There was a marked difference in the level of the nasal floor. The right nasal floor was higher than the contralateral side. (Picture 1.B)

A septoplasty procedure was performed to provide access to choanae before choanoplasty. The transnasal endoscopic choanoplasty procedure was done in both side of the choanae. The membranous type atresia in the left side was incised using sickle knife to make an opening to nasopharynx. Then, it was widened and trimmed with microdebrider to achieve choanal diameter of approximately 8 mm. While, the bony type atresia in the right side undergone similar procedure with additional drilling with diamond bur to make an opening to nasopharynx and widened the choanae. A higher nasal floor in the right nasal cavity compared to the left, limited the diameter of choanal opening. The right choanal diameter of approximately 6 mm was able to be made. A stent made from nasogastric tube was maintained for 2 weeks. (Picture 2.B)

A weekly follow up nasal endoscopy was done to evaluate choanal patency. At approximately one month after choanoplasty, a mild stenosis with choanal diameter of approximately 5 mm was observed in left choanae and total stenosis in the right choanae. In order to repair the restenosis, a revision choanoplasty procedure was performed. The right choana was drilled to achieve a diameter of 8 mm and steroid diluted tamponade was put in the choanae after the procedure. In addition, steroid injection was administered in both choanae. After the procedure, the patient experienced an improvement in

nasal obstruction. Post-operative nasal endoscopy showed a left choanal patency and right choanal mild stenosis with a diameter of 3 mm. Further nasal endoscopic evaluation was planned for the patient but he was not returned. The choanal nasal endoscopic image after the first choanoplasty and revision surgery can be seen in Picture 2.



Picture 1: Axial (A) and Coronal (B) Section CT-Scan. A. Bilateral choanal atresia with bony (right) and membranous (left) type (white arrow) with bone thickness of 3.33 mm and septal deviation (red arrow). B. Coronal section showing a higher level of nasal floor was seen in the right side with 5.02 mm difference.

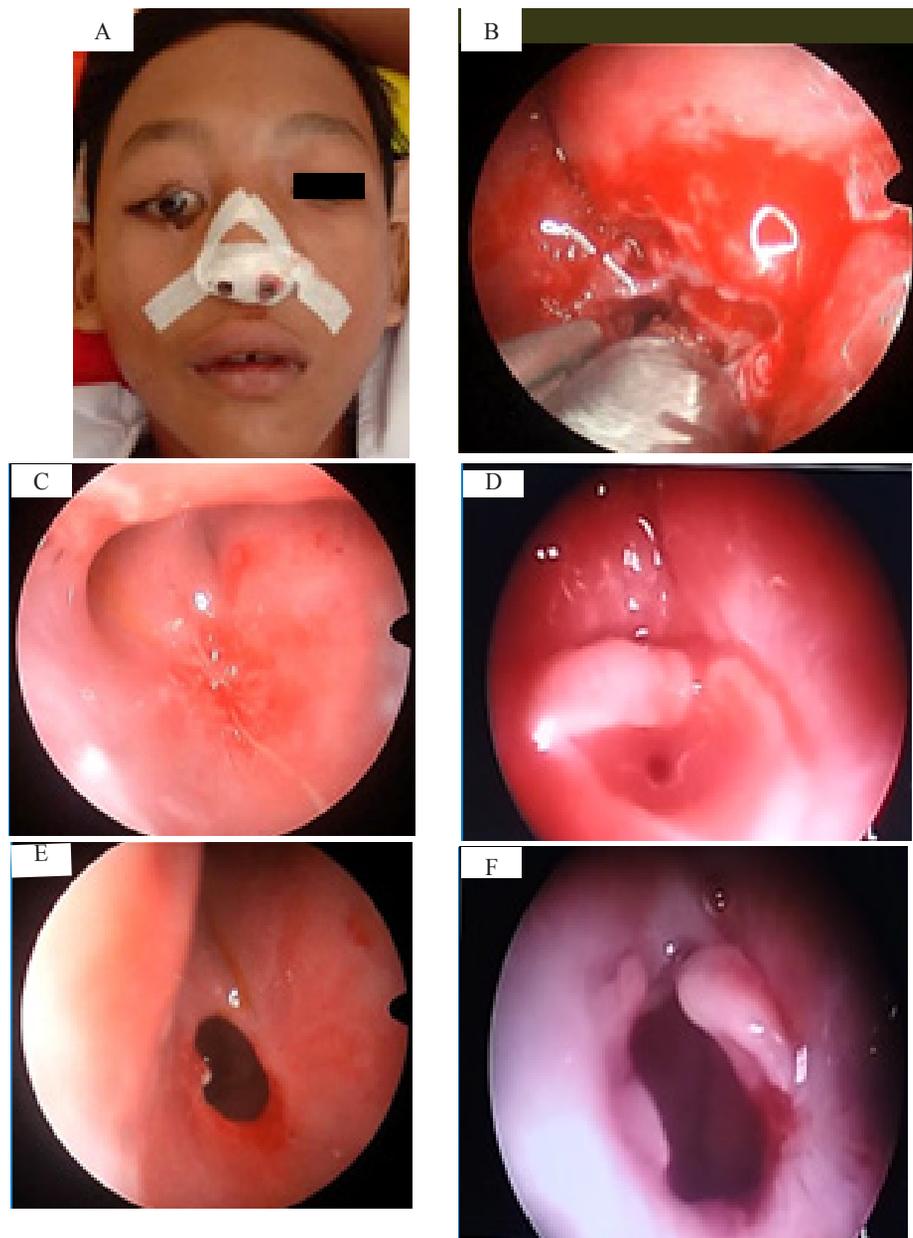


Figure 2: The clinical picture and choanal image from nasal endoscopy. A. the patient with nasal stent after the first choanoplasty procedure. B. Bur drilling to widen the bony choanal atresia. C&D. The comparison of the right choana with total restenosis after first choanoplasty (C) and patent choana after revision choanoplasty (D). E&F. The comparison of the left choana with mild restenosis after first choanoplasty (E) and wide and patent choana after revision choanoplasty (F)

DISCUSSION

Choanal atresia is a rare congenital condition marked with the obliteration of posterior choanae. The incidence is estimated around 1:5000 to 1:9000 for every newborn.^[1] There is slightly higher predominance in girls than boys. A third of choanal atresia cases involved bilateral choanae.^[2] In the condition of bilateral obstruction, an emergency treatment to secure airway was needed as newborn until the age of six months were an obligate nasal breather.^[3, 4] In our case, bilateral choanal atresia was diagnosed when he was a teenager. He had a history of respiratory distress when born that was alleviated when crying. The bilateral choanal atresia was rarely found in boys. Moreover, it is rarely survived through childhood. Instead of respiratory distress, the patient complained with nasal obstruction with profuse rhinorrhea. These symptoms were frequently found in bilateral choanal atresia in teenagers or adults.^[1] Furthermore, there was no other apparent congenital or growth abnormalities found in the patient, except for the palpebral coloboma. Although choanal atresia were associated with CHARGE syndrome (Coloboma, Heart defect, Atresia choanae, Retarded growth, Genitourinary hypoplasia and Ear anomalies), this case showed no typical sign of CHARGE syndrome.^[5]

The etiology of choanal atresia was not well understood. It was hypothesized that the persistent buccopharyngeal membrane in the foregut during the embryonic growth causing choanal atresia. The other hypothesis was the persistent mesodermal adhesions in the posterior nasal aperture. To this date, the accepted hypothesis of choanal atresia was the bucconasal membrane of Hochstetter that persist between oral and nasal cavity. The other widely accepted theory was neural crest migration failure due to the local growth factor.^[3, 6]

Initially, the clinical diagnosis of choanal atresia can be made with the failure of soft catheter insertion from the nasal cavity into the nasopharynx. Further evaluation with nasal endoscopic and CT-Scan provide reliable imaging to support the diagnosis and plan for treatment.^[4] In this case, the nasal endoscopy showed a narrow right nasal cavity compared to the left side with septal deviation. The CT-scan provide more clear view showing the bilateral choanal atresia with different types in each side. The bony type and membranous type were seen in the right and left choanae, respectively. The difference in choanal atresia type in each side add on the uniqueness and rarity of this case. This difference required different surgical approach for choanoplasty.

The technological advancement improved the surgical approach greatly from the transpalatal approach to transnasal endoscopic approach for choanoplasty. The transpalatal approach provide a good result for thick atresia. However, the transpalatal surgery have possible risk of palatal fistula and possible midface deformity.^[7] Following the technological advancement, the transnasal endoscopic approach gaining the popularity for its advantages. The biggest advantage of this approach was it provide access for minimal invasive surgery. Currently, this approach has become the standard therapy for choanal atresia. The endoscopic approach provided an optimal visualization of the choanae. In our case, an initial septoplasty was performed before choanoplasty to provide better access to choanae. A transnasal endoscopic approach choanoplasty was performed in both choanae. The newly formed choanae was made as wide as possible using microdebrider for the membranous type and bur for widening the bony part. This approach enabled precise choanal construction without risking of damaging surrounding structures.

The stent made from nasogastric tube was maintained for two weeks to ensure choanal patency. One month after surgery, a mild stenosis was observed in the membranous side and total restenosis in the bony side. It is known that restenosis was the common complication of choanoplasty. There were 9-36% cases of restenosis occurred within 3-21 weeks of surgery.^[8] There was a controversy around the use of stent which might promote stenosis. It is argued that the stent induces granulation tissue formation and inflammation.^[9] However, it is advised to be used in the bilateral choanal atresia. Moreover, the stent use within the duration between 2-6 weeks can minimize the risk of restenosis.^[4] Therefore, the post-operative use of stent in bilateral choanal atresia was considered to be more beneficial despite the risk of restenosis.

There were numerous factors that can lead to restenosis. It is known that the bony type of choanal atresia was more likely to have revision surgery compared to the membranous type. The choanal atresia that was associated with other congenital malformation have a higher risk of restenosis after surgery.^[10, 11] In our case, the restenosis might be affected by the higher level of the right nasal floor which limited the size of the newly formed choanae. In comparison with the membranous type in the left side, the diameter of the newly formed choanae was smaller. An inadequate bone removal may also contribute to restenosis. It was suggested that the rate of restenosis was higher in boys than girls. However, further research was needed to evaluate the role of gender to restenosis because of possible confounding factors affecting the association.^[12] Thus, these factors remains to be challenging in treatment of bilateral choanal atresia.

The revision surgery often necessary in restenosis. The diameter of choana of less than 4 mm required revision surgery.^[9] A total stenosis was occurred in the bony side of the choanal atresia in our case. A revision choanoplasty was performed. The choanae was tamponade with triamcinolone acetonide gauge after the procedure. Additional steroid injection surrounding the choanae was administered in both choanae. The follow up result show a recurrent stenosis but with intact patency of the choanae and improved symptoms. The steroid was use for anti-inflammatory effect and preventing granulation tissue formation. A recent case series reported other application of steroid in choanal atresia repair.^[13] The steroid eluting stent provide potential in reducing restenosis after choanoplasty. However, further research was needed to explore the long-term effects. Other operative technique recommended the use of flap to prevent restenosis. The combination of flap and fibrin glue provide a good hemostasis and prevent granulation formation.^[7] We acknowledge that the recurrent stenosis occurred even after revision surgery, but the use of steroid shows potential effect in ensuring choanal patency.

CONCLUSION

Treatment of bilateral choanal atresia with a transnasal endoscopy approach facilitates choanoplasty with minimal complications. However, complications in the form of recurrent stenosis after surgery remains to be a challenge.

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The authors provide equal contribution in patient's examination, treatment, follow up and writing the case report.

CONFLICT OF INTEREST

There are no conflicts of interest.

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