

CASE REPORT

Management of congenital choanal atresia (CCA) after multiple failures: A Case Report

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SUMMARY

Nasal obstruction in neonates is a potentially fatal condition because neonates are obligatory nasal breathers. Bilateral choanal atresia is therefore a neonatal emergency. Several approaches for corrections of choanal atresia are available including the helium laser: YAG. A 5-year-old Chinese girl born with bilateral choanal atresia, had birth asphyxia that required intubation. She underwent multiple surgeries for correction of choanal atresia at other hospitals but failed to improve. She was referred to Universiti Kebangsaan Malaysia Medical Center (UKMMC) after presenting with intermittent respiratory distress and cyanosis following an upper respiratory tract infection. A repeat computed tomography (CT) scan done preoperatively showed complete bony stenosis over the left choana and finding was confirmed by examination under general anesthesia. She underwent endoscopic transnasal removal of left bony atretic plate. There was no intra or postoperative complications. During follow up 10 years later, the airway on both sides remains patent.

KEY WORDS:

Choanal atresia; stenosis; laser; computed tomography

INTRODUCTION

Neonates are obligatory nasal breathers and compromise of the nasal passages is potentially life threatening. Congenital choanal atresia (CCA) occurs in approximately one in 7000 live births, 45% are bilateral and of the unilateral cases, 71% involve the right choanae¹. Male to female ratio is about 2:1. In 90% of cases, there are bony atretic plate obstructing the posterior choanae and another 10% are membranous type. The etiology of this anomaly is due to persistence of the embryological bucconasal membrane. This membrane divides the nasal cavity from the stomatodeum until it breakdowns at the seventh week, allowing communication through the primitive nares. Other possible etiology of choanal atresia could be due to persistence of epithelial cells in the nasal cavities. These epithelial cells proliferate between the sixth and eight week.

CASE REPORT

A 5-year-old Chinese girl was referred from one of the state hospital with a history of intermittent respiratory distress preceded by upper respiratory tract infection. She had a history of birth asphyxia requiring intubation for a week. The

attending pediatrician had difficulty in introducing a Ryle's tube through both nostrils. Preliminary diagnosis of bilateral choanal atresia was made. A CT scan of paranasal sinuses demonstrated bilateral choanal atresia (Fig 1).

She underwent transnasal perforation of the atresia at the age of one week and the stent was placed in-situ for six weeks. However, removal of the stent precipitated another episode of cyanosis, requiring intubation. A week later she underwent a second operation and the stent was left in-situ for two months. After removal of the stent she had difficulty in breathing during feeding. She occasionally turned cyanosed following an upper respiratory tract infection. Two months later, a transnasal endoscopic surgery using a drill to create an opening on both posterior choana was performed. Silicon stents were left for 8 weeks. Upon removal of the stent, the right posterior choana was noted to have a small aperture, covered with granulation tissue. The left posterior choana had a good opening. Six months later she was re-admitted for a similar problem. Examination under general anaesthesia revealed a small aperture covered with granulation tissue. The left choana was occluded by a bony segment. She underwent dilatation using Hegars dilator for both choanae. Three years later, she was referred to our center with a history of intermittent respiratory distress preceded by upper respiratory tract infection. Preoperative CT scan showed a bony atresia on the left choanae and the right choana was normal. She underwent another examination under general anesthesia; using endoscopic approach. A diamond burr was used to create an opening on the left posterior choanae. This time, the posterior part of the septum was removed. When reviewed in the clinic last December, she had no respiratory problem and no cyanosis. Clinical examination revealed patent airways on both nostrils.

DISCUSSION

The spectrum of presentation in choanal atresia patients ranges from respiratory difficulty only during upper respiratory tract infection to acute total obstruction. In cases of unilateral stenosis/atresia or in those who has better-compensation, they may be managed by using the McGovern nipple to establish an oral airway which allows definitive correction to be deferred until approximately one year. If the atresia is unilateral, the newborn may not have severe airways problem at birth. The baby may remain asymptomatic until the first respiratory infection develops.

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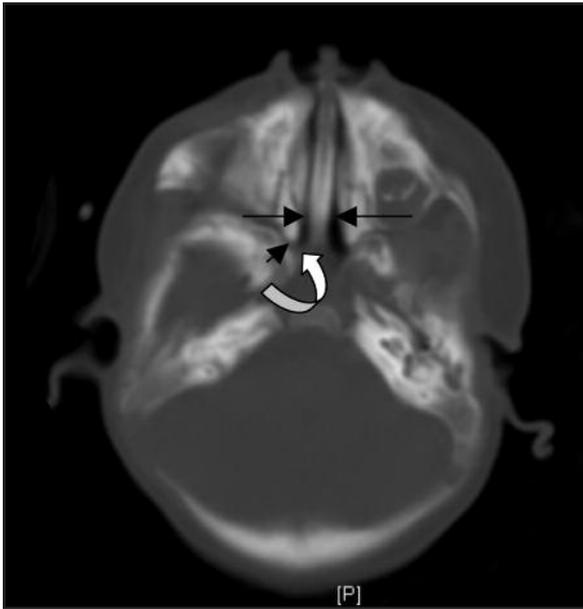


Fig. 1 : Axial section of HRCT demonstrating widening of the vomer (long arrows), bowing of the posteromedial maxilla (short arrow), and narrowing of the choana anterior to the pterygoid (curved white arrow).

Computed tomography is used to confirm the diagnosis, identify the type of CCA and delineate the extent of atresia. The main disadvantages of using CT in pediatric populations are the high radiation exposure dose and the need for sedation to avoid movement artifacts, the latter could render the examination of fine details less reliable. Using contrast material in the nose will help to overcome that. The application of a low-dose technique using 50–60 mA can avoid a high radiation dose to the child without any effect on the diagnostic accuracy. Multislice CT can clearly delineate the posterior choanae and their communication with the nasopharynx².

Many methods of surgical correction have been used, for example transpalatal, transseptal and transnasal. Transantral approach has been described but has become obsolete. Transnasal puncture is performed under general anesthesia with oral intubation. The nasal cavity is packed with ribbon gauze that has been soaked with cocaine mixed with adrenalin. The puncture is performed bimanually with the surgeon's index finger directly palpating the posterior choana to prevent inadvertent injury to the spinal cord and any other structures in the base of skull. A Hegar dilator or mastoid curette may be used to create the initial puncture. The created opening may be enlarged with a Hegar dilator.

Friedman *et al* had studied 65 children presenting with choanal atresia. In all cases; correction was performed by a transnasal approach under endoscopic control. A 120-degree Hopkins rod telescope was used to visualize an atretic plate from nasopharynx. Straight urethral sounds were used to perforate the plate followed by use of a drill to remove the bony component³. His group found that 74% were asymptomatic following correction. Those neonates with

bilateral choanal atresia who were stented with Portex endotracheal tube for at least 12 weeks had the best outcome³.

Sadek advocated a new stenting technique that may reduce the incidence of restenosis in choanal atresia after decanulation⁴. In his study, eight neonates with bilateral bony CCA were treated by endonasal perforation. He used a modified stenting technique utilizing endotracheal tube placed beyond the atretic plate. He claimed that this stent aided epithelization of new tract and thus preventing restenosis. He followed up the patients ranging from eighteen months to eleven years. Seven out of eight patients did not require further treatment and one required unilateral dilatation⁴.

The transpalatal approach was popular previously in infants with bilateral bony atresia. Palatal exposure may be obtained with Brown-Davis or Dingmans self-retaining retractors. The authors do not advocate this approach as it may cause disturbance of the palatal growth later and it has a higher complications rate. Nowadays many surgeons use the endoscopic approach in the treatment of CC.

Fong *et al* reported 8 patients who underwent procedures for correction of choanal atresia using helium: YAG laser. All patients were placed under general anesthesia with laser endotracheal tubes. A nonreflecting black ear speculum was then placed into the nasal cavity. A black malleable retractor was placed in the nasopharynx to protect the posterior pharyngeal wall. The patient's head and body were protected from the laser beam. The helium: YAG laser was set at 5 watts super pulse with an interval of 0.2 seconds. The area of the atresia was vaporized with the laser starting medially moving laterally until the malleable retractor could be visualized. Two of 8 patients had restenosis on the second look procedure that required formal revision. The average follow up for these patients is 2.5 years. The revision rate is 23%. None of these patients suffered any complication⁵.

Antibiotics are administered postoperatively to prevent infection especially when the stent is in-situ. Regular suction and local care of stent are important to ensure the patency of the tube for breathing. Intra-operative complications risks are dependant on the type of approaches. For transpalatal approach, intraoperative complications are bleeding from the greater palatine arteries, injury to the basocciput, and injury to the content of incisive foramen. Early postoperative complications may include airway obstruction secondary to edema of the soft palate or tongue, nasal infection, necrosis of palatal mucosa, velopharyngeal incompetence and pressure necrosis of the columella of the alar rim.

Operative complications for transseptal approach includes cerebrospinal leak from excessive removal of the bony septum. Other complications such as septal hematoma and septal perforation are the complications of transseptal approach. Late postoperative complications include restenosis of the choana. Patient may require dilatation or revision surgery. This complication can be prevented by adequately removing bone, preserving mucosal flaps and stenting for 2 months. Our patient underwent five operations

for correction of choanal atresia. The last follow up which was 2 years ago and she was asymptomatic.

CONCLUSION

Congenital choanal atresia is the developmental failure of the nasal cavity to communicate with nasopharynx. Patients may require multiple surgeries to achieve an adequate airway.

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