



Bilateral Congenital Choanal Atresia: A Case Report

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Abstract: Choanal atresia is an uncommon congenital anomaly of nose with an incidence of approximately 1 in 5000-7000 live births. Choanal atresia is caused by failure of resorption of the bucco-pharyngeal membrane during embryonic development. Choanal atresia has a significant incidence of associated defects. The most common associated congenital anomaly is CHARGE association. Bilateral choanal (BCA) atresia presents with severe respiratory distress and cyanosis at birth and is alleviated by crying whereas unilateral atresia often remains undetected till late in life. Bilateral choanal atresia is managed with an oropharyngeal airway. Surgery is the definitive treatment with two main approaches, namely transnasal or transpalatal. We discuss successful management of a neonate with bilateral choanal atresia who presented with respiratory distress and feeding difficulty since birth.

Keywords: Choanal atresia, neonate, congenital anomalies, endoscopic surgery.

Choanal atresia is an uncommon congenital anomaly of nose with an incidence of approximately 1 in 5000-7000 live births.¹ It is more common in females with female to male ratio of 2:1. Generally, 65% to 75% of patients with choanal atresia are unilateral, whereas the rest are bilateral.² About 30% cases are pure bony, whereas 70% cases are mixed bony-membranous.³ Bilateral choanal atresia (BCA) presents with acute respiratory distress in neonates whereas unilateral atresia often remains undetected till late in life.

Case Report: A three day old female child weighing 2.5 kg presented with respiratory distress and feeding difficulty since birth. Child was born vaginally at term in our hospital. She was examined by neonatologist and based on history and physical examination diagnosis of choanal atresia was made which was confirmed by inability to pass suction catheter through nostrils. An oropharyngeal airway was put and fixed with tape in her mouth to relieve respiratory distress and intravenous fluid was started. She was referred to otolaryngology department for correction of choanal atresia. On physical examination, child was having respiratory difficulty with chest retraction. Auscultation revealed bilateral crepitations and spasm in chest and normal heart sounds. Routine investigations

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including Hb, BT, CT and urine examination were normal. Patient was nebulised with salbutamol and oxygen inhalation via face-mask was started. On operation table, routine monitoring was instituted. Patient was preoxygenated with 100% oxygen and induced with inj atropine, fentanyl, thiopentone and scoline and intubated with endotracheal tube of size 3.5mm. Anaesthesia was maintained with oxygen, nitrous oxide, halothane and atracurium. Intravenous fluid was given with 0.45 % saline with 5% dextrose.

The patient was operated by transnasal approach with the help of endoscope. Bone plate was perforated with suction tube and appropriately dilated. Stenting was done with the help of 3.0mm portex endotracheal tube to maintain the patency of newly formed lumen. The whole procedure lasted for about one and half hours. At the end of the surgery, inhalational anaesthetic and nitrous oxide were switched off and neuromuscular blockade was reversed with inj. atropine and neostigmine. The ETT was kept in situ till the child was fully awake and had regained adequate muscle power. The trachea was then extubated and the child observed in the operating room for another 30 min and then shifted to recovery room. Patient was discharged on 5th post-operative day with the advice of weekly follow-up. Stent was removed after 4 weeks and during this period patient was advised antibiotics to prevent any infection.

Discussion: Choanal atresia is the developmental failure of the nasal cavity to communicate with the nasopharynx. It is generally thought to be secondary to persistence of either the nasobuccal membrane of Hochstetter or the buccopharyngeal membrane from the foregut. This membrane normally ruptures between the fifth and sixth weeks of gestation to produce choanae. Failure of this membrane to rupture causes atresia of choanae.⁴ Choanal atresia is often associated with other congenital anomalies. The most common associated congenital anomaly is CHARGE association (C = coloboma, H = heart disease, A = atresia of choanae, R = retarded growth and development, G = genital hypoplasia, E = ear deformities or deafness).⁵ It can also be associated with Treacher-Collins syndrome and congenital malformations such as syndactyly or polydactyly, microcephaly, oesophageal atresia and craniostosis⁶.

Choanal atresia can be unilateral or bilateral. Bilateral choanal atresia presents very early in the life. Most patients with bilateral choanal atresia are detected within the first month of life. Respiratory distress occurs in patients with bilateral choanal atresia at or shortly after birth as neonates are obligate nasal breathers. They present with cyclic cyanosis relieved by crying. On the other hand, patients with unilateral choanal atresia rarely present with immediate or severe airway obstruction. They normally present within first 18 months of life with feeding difficulties and nasal discharge, but may present with unilateral nasal obstruction and discharge in later life.

There are numerous ways to diagnose choanal atresia. The simplest method is to pass a soft, red-rubber catheter (no. 8 French catheter) or 2.6 mm feeding tube through the nose into the nasopharynx. If the catheter is visualized in the oral cavity or oropharynx, a nasal airway is present. Inability to pass that catheter to the pharynx raises the possibility of choanal atresia. Management of these patients varies and depends on age, type of atresia, and general condition of patients. Because infants are obligate nasal breathers, bilateral choanal atresia is a life-threatening situation since, if



not promptly recognized, it can lead to severe asphyxia and death immediately after birth. Immediate management of bilateral choanal atresia involves training the infant to breathe through the mouth with the aid of an indwelling oral appliance such as a McGovern nipple or an oropharyngeal airway. This temporary oral airway maintenance is needed until adequate or clinically patent choanae are established for breathing. Patient should be evaluated fully before planning surgical correction. There are various surgical approaches to correct this problem. The transnasal approach is currently the preferred one and can be performed in a minimally invasive fashion with endoscopic instrumentation. It is a safe and rapid procedure even in very young children, with no complications and a high rate of success.

Neonates with BCA have unique anaesthetic and surgical requirements regarding equipments, fluid and drug therapy, anaesthetic dose and environmental control. During first few weeks of life, neonates are vulnerable to so called flip-flop circulation. Hypoxia, hypercapnia, acidosis, infection and hypothermia can cause sudden increase in pulmonary artery pressure resulting in shunting of blood through the patent foramen ovale or re-opening of ductus arteriosus. Ventricles are also less compliant, sensitive to volume overloading, have a rate dependant cardiac output and are more susceptible to cardiac depression by potent anaesthetic agents⁷.

Neonates have immature kidney. At one month the kidneys are only about 60% mature. Hepatic metabolic capacity is also immature at birth. Both these factors have important clinical implication regarding neonate's ability to excrete some medications and make them vulnerable to drug over-dose⁷. Temperature regulation is an important issue in neonate. Because of large body surface-to-weight ratio, they are especially vulnerable to intra-operative hypothermia. Thus, an understanding of basic differences in physiology, pharmacology and pharmaco-dynamic responses and the underlying pathology of surgical problem is essential for development of safe anaesthetic/surgical plan.

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