



Modern approach in the treatment of choanal atresia



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Abstract:

Introduction: Choanal atresia is a well-recognized condition characterized by an anatomical closure of the posterior choanae in the nasal cavity.

Case presentation: We report the case of a term female infant from first normal pregnancy. During routine delivery room aspiration an obstruction of the upper airways was suspected due to failure of passing a suction catheter through the nasal passages. The infant was transferred to neonatal intensive care unit where after initial sedation with phenobarbital and midazolam was intubated and assisted ventilation was initiated. A nasogastric tube was inserted for feeding. The diagnosis of bilateral bony choanal atresia was confirmed by a CT scan. On the 7th day of life, the infant was transported from the NICU of ACC University Hospital Tokuda to the Department of ENT Surgery in University Hospital “Queen Giovanna” – ISUL for surgical treatment. Under general anesthesia a transnasal endoscopic choanoplasty was performed and bilateral stents were inserted. After the surgery the patient was transported back to the NICU of ACC University Hospital Tokuda in a stable condition for additional intensive care and treatment.

Discussion: Complex pathologies must be managed in a coordinated way by the ‘multidisciplinary team’ approach. Bilateral choanal atresia in the infant may present as a life-threatening respiratory distress immediately after birth or later at the first feeding. When diagnosed the main priority is to ensure free airway. This can be achieved by the use of different oropharyngeal devices or by endotracheal intubation. The perioperative anesthetic management of neonates is challenging even for the experienced anesthesiologist. Neonatal anesthesia demands profound understanding of the rapidly changing physiology of the neonate and both the pharmacokinetics and pharmacodynamics of the medications used to provide anesthesia. Conclusion: Successful management of complicated medical cases with complex pathologies require modern approach and close coordination between different specialties. Our therapeutic success is a result of the close collaboration between the anesthesia, surgery and neonatology teams.

Key words: Choanal atresia, multidisciplinary team, modern approach

INTRODUCTION

Choanal atresia (CA) is a well-recognized condition characterized by the anatomical closure of the posterior choanae in the nasal cavity¹. It occurs in one in 5000–8000 neonates, twice as common in females, twice as often on the right and twice as often unilaterally (i.e., the rule of two)^{4, 12}. Anatomic boundaries of the posterior choanae 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. Different types of atresia are recognized: pure bony atresia, pure membranous atresia, mixed bony/membranous atresia, unilateral or bilateral. The rarest type is pure bony bilateral atresia^{2, 3}. There are several theories regarding the embryogenesis of CA, but it is generally considered to be secondary to the persistence of either the nasobuccal membrane of Hochstetter or the buccopharyngeal membrane from the foregut. This membrane normally ruptures between the 5th and 6th weeks of

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gestation to produce the choanae. Failure of this membrane to rupture causes atresia of choanae⁶. Bilateral CA is a life-threatening disorder. Neonates are obligate nasal breathers with a superior position of the epiglottis above the soft palate. With patent choanae, an infant can breathe and feed simultaneously with a low aspiration risk. Bilateral CA due to nasal obstruction in the infant may occur as a life-threatening respiratory distress immediately after birth, or later as respiratory distress when the first feeding is initiated⁵. In case of bilateral CA, the immediate ensuring of free airway is crucial. Free airway could be ensured by different oropharyngeal devices or by endotracheal intubation. After that a nasogastric or feeding tube should be inserted. The definitive treatment of the condition is to ensure the patency of the nasal passages as soon as possible. Before the surgical treatment, it is first necessary to carry out urgent diagnostic procedures to determine associated diseases and risk factors, especially cardiological factors⁷. Then, if the procedure under general anesthesia is safe, the ear, nose and throat (ENT) surgeon will decide on the timing of the surgical treatment. The methods for surgical treatment are: transnasal perforation, transpalatal resection, transnasal endoscopically assisted perforation and transnasal endoscopic choanoplasty. A stent can also be inserted regardless of the used method to maintain choanal patency. Transnasal endoscopic choanoplasty is considered the surgical technique of choice for the treatment of CA by most ENT surgeons. It is based on the endoscopic removal of the atretic area and the posterior part of the nasal septum, thereby creating a single choana⁸⁻¹¹.

Choanal atresia can be associated with other congenital anomalies. One of the most common associations is the CHARGE syndrome (C=coloboma, H=heart disease, A=atresia of choanae, R=retarded growth and development, G=genital hypoplasia, E=ear deformities or deafness). Other anomalies associated with CA include polydactyly, nasal-auricular and palatal deformities, Crouzon's syndrome, Down syndrome, Treacher-Collins syndrome, DiGeorge syndrome, craniosynostosis, microencephaly, meningocele, meningoencephalocele, facial asymmetry, hypoplasia of the orbit and midface, cleft palate, and

hypertelorism¹³. This requires a multidisciplinary approach in the management of patients with CA.

CASE REPORT

We present a case of a term newborn female infant, from first normal pregnancy and spontaneous vaginal delivery at 40 gestational weeks, with birth weight 3080 g and height 50 cm. The Apgar score was 8/8 at 1st and 5th min respectively. During routine delivery room aspiration an obstruction of the upper airway was suspected due to failure of passing an 8-Fr suction catheter through both nasal passages. The skin was pale with persistent acrocyanosis at 5th min. The infant was admitted to Neonatal Intensive Care Unit (NICU) in deteriorated general condition with oxygen desaturation, marked cyanosis and inspiratory dyspnea with improvement when crying. The infant was placed in an incubator for thermal comfort and standard monitoring was applied including pulse oximetry, non-invasive blood pressure and ECG. The general physical examination on admission found pale skin with persistent acrocyanosis after the 5th min after birth, on auscultation breathing was weakened vesicular with crepitations in the bases of the lungs, with respiratory rate of 58-62/min. Hemodynamic was stable with rhythmic heart activity and heart rate of 140-156/min. Neurological status showed decreased muscle tone. After sedation with phenobarbital and midazolam, the infant was intubated and assisted ventilation was initiated on sIPPV regiment with the following initial settings – FiO₂ – 40%, fr – 25/min, peep-4, pin-22mmbar. A nasogastric tube was inserted for enteral feeding. Laboratory and microbiological examinations were assigned. An empiric antibiotic therapy was started. Within the 1st 24 hours the ventilatory parameters were successfully adapted to sIMV regiment with the following settings – FiO₂ – 30%, fr – 18/min, peep-4, pin-24mmbar. CT scan of the head was performed and confirmed the diagnosis of pure bony bilateral CA. After consultation with ENT surgeon and anesthesiologist the infant was prepared for surgical treatment.

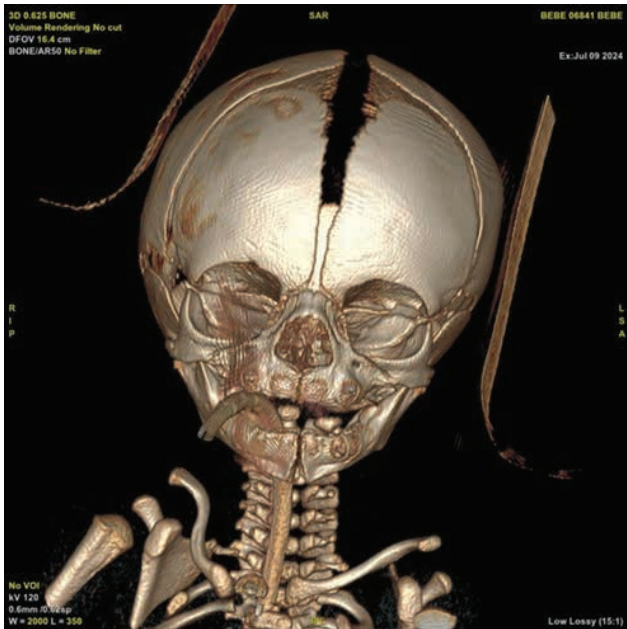


Fig.1. CT scan of the head confirms the diagnosis of pure bony bilateral choanal atresia

On 7th day of life (DOL) the infant was transported from the NICU of ACC University Hospital Tokuda to the Department of ENT Surgery at University Hospital „Queen Giovanna” – ISUL for surgical treatment. Under general anesthesia transnasal endoscopic choanoplasty was performed with insertion of bilateral stents.



Fig.2. Transnasal endoscopic choanoplasty was performed with insertion of bilateral stents

After the operation the patient was transported back to the NICU of ACC University Hospital Tokuda in a stable condition. Spontaneous nasal breathing was immediately recovered. The patient was successfully extubated on the same day. Within the next couple of days bottle feeding was successfully established. Twelve days after the operation the nasal stents were removed, and a day after the infant was discharged from the hospital in a stable general condition.



Fig.3. After the operation the patient is in stable condition

At the age of 1 month and 16 days the infant was readmitted to the NICU in deteriorated general condition with history of recent, feeding and breathing difficulty. Standard monitoring was applied – pulse oximetry, non-invasive blood pressure and ECG. The patient had marked subcostal retractions, on auscultation breathing was weakened vesicular. The respiratory rate was 52-61/min. Hemodynamic was stable with rhythmic heart activity and heart rate of 130-145/min. Neurological status showed normal muscle tone. A 6-Fr suction catheter hardly passed through the right nasal passage and failed to pass through the left one. On the next day the infant was transported by the neonatal team to the Department of ENT Surgery in University Hospital „Queen

Giovanna” – ISUL for reevaluation. Under general anesthesia a transnasal endoscopic revision of the choanoplasty was performed with dilation and insertion of bilateral stents. At the end of the operation the infant was extubated and transported back to the NICU of ACC University Hospital Tokuda in a stable condition. Sufficient bottle feeding was restored and the patient was discharged from the hospital in good general condition with follow-up recommendations.

DISCUSSION

Complex pathologies must be managed in a coordinated way by the ‘multidisciplinary team’ approach. The first members of our multidisciplinary team were the neonatologists. Neonatology is a subspecialty of pediatrics that consists of the medical care of newborn infants. Choanal atresia is a pathology of the nasal cavity and the treatment is surgical, especially when it is bilateral. Since the transnasal endoscopic choanoplasty is the gold standard for surgical treatment of CA, we needed an experienced in endoscopic surgery ENT specialist in our team, and of course an available special endoscopic equipment for such precise surgery of a newborn infant. And last, but not least, such operation should be performed under general anesthesia so we needed an anesthesiologist with experience in both ENT surgery anesthesia and infant anesthesia. Which were the main challenges of managing this „multidisciplinary situation”? First, the situation required close coordination between specialists from three different departments in two different hospitals. Second, there was a very short time to manage the legal issues of performing a surgery on a newborn infant in another hospital with no Neonatal or Pediatric departments. And third – to organize and coordinate the transport of a newborn infant between the hospitals in conditions of prolonged mechanical ventilation by the neonatology team.

The breathing in the newborn infant is obligatory through the nose which makes simultaneous breathing and eating possible. The infant begins to learn to breathe through the mouth between

the fourth and sixth weeks of age. At six months of age, mouth breathing is usually well established. A bilateral CA in the newborn infant may present as a life-threatening respiratory distress immediately after birth or later at the initiation of the first feeding. The clinical picture of nasal obstruction consists of cyanosis during feeding and improvement with crying, episodes of apnea, snoring, nasal flutter, nasal congestion, intercostal and jugular retractions and sleep disturbances^{14, 15}. When bilateral CA is diagnosed the main priority is to ensure free airway. This can be achieved by the use of different oropharyngeal devices or by endotracheal intubation. In the presented case we choose endotracheal intubation because we needed to perform a transport of the patient to another hospital and the endotracheal tube is the most secure device for airway management.

One of the most critical parts of this multidisciplinary treatment was the anesthesiological management. The perioperative anesthetic management of neonates is challenging even for the experienced anesthesiologist. Neonatal anesthesia demands understanding of the rapidly changing physiology of the neonate and both the pharmacokinetics and pharmacodynamics of the medications used to provide anesthesia. The small size of neonates require experience in vascular access and airway management. They are vulnerable to respiratory and cardiac events which require rapid detection and correction. Common anesthesia practices in neonates include the use of monitoring (ECG, non-invasive blood pressure, temperature, pulse oximetry and end-tidal carbon dioxide). Care must be taken to preserve normal temperature during transport to and from the operative suites and well has heating the operative environment. Continuous glucose delivery throughout the duration of the procedure is required. General anesthesia with placement of endotracheal tubes is generally required for the safe care of the neonate. Anesthesia typically consists of a balanced combination of volatile agent supplemented with narcotic for hemodynamic stability¹⁶. In the presented case the opioid of choice was fentanyl. Fentanyl based analgesia in neonates can lead to delayed awakening so it must be very precisely carried



out. We did not use muscle relaxation because the neurological status showed decreased muscle tone. After the anesthesia spontaneous breathing was quickly recovered, mechanical ventilation was ceased and the patient was extubated on the same day. Bottle feeding was quickly successfully established and the patient was discharged from the hospital in a stable general condition.



Fig.4. Simultaneous bottle feeding and breathing after the operation

CONCLUSION

Successful management of complicated medical cases with complex pathologies require modern approach and close coordination between different specialties. Our therapeutic success is a result of the close collaboration between the anesthesia, surgery and neonatology teams.

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