Case Report

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Modified transnasal endoscopic repair of bilateral choanal atresia with pulmonary atresia

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ABSTRACT

Congenital choanal atresia is due to failure in the development of communication between the nasal cavity and nasopharynx in newborns. This condition is life threatening when it is bilateral and causes severe respiratory distress immediately after birth as children are obligatory nose breathers. We present a one day old child who presented to us with severe respiratory distress, which we diagnosed as choanal atresia. The child also had tertology of fallot with pulmonary atresia and had to be intubated as oxygen saturation was not adequate. Surgery was undertaken on emergency basis on day one of life which helped in extubation of the child. This case highlights the importance of prompt diagnosis of a case bilateral choanal atresia, and also the importance of early surgery which can lead to significant improvement.

Keywords: Bilateral choanal atresia, CHARGE syndrome

INTRODUCTION

Congenital choanal atresia is defined as a congenital failure in the development of communication between the nasal cavity and nasopharynx in newborns. It is the most common congenital anomaly of the nose. Most of the patients present with unilateral choanal atresia. Neonates with bilateral choanal atresia have severe respiratory distress at birth. We present a case of congenital bilateral choanal atresia in which we describe novel techniques in the surgical management like usage of micro ear instruments and stenting.

CASE REPORT

A full term live male baby weighing 2.58 kg delivered by emergency caesarean section in view of oligohydramnios/fetal distress presented to the casualty with severe respiratory distress and cyanosis. The child did not cry spontaneously, cried on stimulation and bag and mask

ventilation. On examination pulse rate was 112/min, respiratory rate was 80 per minute, child was cyanosed SpO2 was maintained at 82% on hood. There was no nasal flaring, cold spatula put at the nostrils showed no fogging, an attempt to insert nasogastric tube failed bilaterally. In view of severe respiratory distress and exaggerated work of breathing the child was intubated using 3.5 size endotracheal tube and connected to ventilator.

Echocardiogram was done for the child and showed Tetrology of fallot with pulmonary atresia. Prostaglandin infusion was started. A non contrast CT scan of the nose and PNS (Figure 1) showed bony atresia of bilateral choana and hence the child was diagnosed as a case of bilateral congenital choanal atresia. The baby was immediately posted for surgery on day 1 of life and a transnasal endoscopic approach was used to remove the atretic plates (Figure 2). The unique use of ear instruments (Figure 3) like Rosen's circular knife for

mucosal incision, Hartmann's alligator forceps and connecting both the choana is to be noted. Red rubber tubes passed through bilateral nostrils through choana were used as stents (Figure 4).

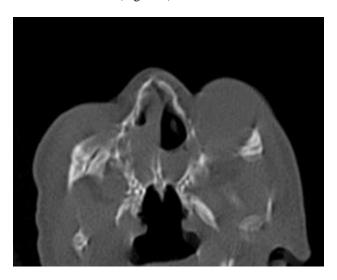


Figure 1: CT Picture showing bilateral choanal atresia.



Figure 2: Endoscopic view of atretic plates.



Figure 3: Instruments used for surgery.



Figure 4: Postoperative image showing bilateral red rubber tubes used as stents.

The child was extubated on postoperative day 1 after removal of the nasal red rubber tube stents and maintaining a saturation of 87% with oxygen hood. Frequent saline irrigation was done post operatively along with instillation of steroid nasal drops and frequent dilation with steroid coated nasogastric tubes. There was an immediate improvement of cyanosis and oxygen saturation of the child after surgery.

DISCUSSION

Congenital choanal atresia is defined as a congenital failure in the development of communication between the nasal cavity and nasopharynx in newborns. It is the most common congenital anomaly of the nose. Most of the patients present with unilateral choanal atresia. Neonates with bilateral choanal atresia have severe respiratory distress at birth. Congenital choanal atresia is seen approximately in every 7000 to 8000 live births and it is presented with upper airway symptoms and obstruction. Neonates are obligate nasal breathers for 3 to 4 weeks after birth. Therefore neonates with bilateral choanal atresia have severe respiratory distress immediately after birth.

The incidence of associated congenital anomalies in choanal atresia is 10% to 50%. ⁴⁻⁷ The most common being the CHARGE syndrome. Although Choanal atresia is a rare entity, it is the most common congenital anomaly of the nose. Approximately 60 to 70% of the cases are unilateral and the rest present as bilateral choanal atresia. ^{4,7} Our case is a case of bilateral choanal atresia with congenital heart disease in the form of tetrology of fallot with pulmonary atresia. Failure of passing a soft

catheter from nares to nasopharynx is the simplest diagnostic method of CCA.

Flexible nasopharyngolaryngoscopy and paranasal sinus CT are helpful to confirm the diagnosis, assess the type of atretic plate, and evaluate the extent of atresia. In recent years its reported that every congenital choanal atresia which was thought of membranous also has a bony component. Structure is usually bony in 30% and osteomembranous in 70% patients. In our case it was a complete bony atresia.

Many surgical modalities have been implemented for treatment. These are transantral, transpalatal, transseptal, and transnasal approaches.^{3,7} The trans-palatal approach offers a very wide field for operation, making corrective manoeuvres easier, but is more invasive and susceptible to complications like bleeding, fistulas, infections and growth defects of the jaw and the palate bone. The transnasal endoscopic approach is the most common technique used, the most common techniques for incision are: double mucosal anterior and posterior low-hinged flap, side-hinged double flap, upper hinged flap, four flaps with cruciate incisions, double nasal and septal flap, and multiple flaps secured with fibrin glue, so as to obtain mucosal flaps for the re-covering of the raw areas at the level of the medial lamina of the pterygoid process and the posterior part of the septum.⁸⁻¹⁰

CONCLUSION

We would like to highlight the early diagnosis, timing of surgery on the day one of life, use of ear instruments for surgery due to the very narrow nasal cavity of the newborn. We also highlight the decision to operate on day one of life as soon as diagnosis is made which resulted in postoperative extubation of the child. The need to address bilateral choanal atresia in a new born as an emergency has been highlighted by our case.

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