Choanal Atresia: Birth Without Breath

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Abstract Bilateral Choanal atresia is a medical emergency. Corrective surgery is the mainstay of the treatment. Hegar's dilator was used in all cases to break the bony/membranous atretic plate. The 22 cases of choanal atresia all operated by the first author were included in this study. Eight cases were 1–5 year old with bilateral choanal atresia and all required immediate surgery as they had repeated attacks of respiratory distress and cycle of cyanosis. Eight cases were of CHARGE Syndrome. All the cases were operated under general anaesthesia. Hegar's dilators were used and nasal stents were placed in all cases. Although complete nasal patency was achieved by surgery, in 8 cases, neonates could not survive due to the CHARGE Syndrome. The mortality was unrelated to the surgery. The 14 cases which were not related to CHARGE Syndrome had a good postoperative recovery. 3 cases were above 15 years old with unilateral complete bony/membranous choanal atresia and they presented with continuous nasal discharge. Hegar's dilators are a safe and simple method of surgery for choanal atresia. Nasal stenting is mandatory to prevent restenosis.

Keywords Choanal atresia · Hegar's dilator · Atretic plate · CHARGE syndrome

Introduction

Choanal atresia is one of the most common congenital anomaly of the nose [1]. Roeder first described the choanal atresia in 1755 then this entity was further described by Otto [2, 3]. The incidence of this abnormality is between 1 in 5000 and 1 in 8000 [4, 5]. The word “choana” is a greek from the word Xovan meaning funnel. It is life threatening condition in the neonates as neonates are obligate nasal breathers for the first 6 weeks of life. After the childbirth as soon as the crying ceases, the child enters the state of respiratory distress.

Embryology

Choanal atresia occurs due to the persistence of buccopharyngeal or naso-buccal membrane [4]. During embryologic development the nasal cavity is formed from the nasal placodes on the lateral surface of head. These placodes sink into the mesoderm which form the nostril and anterior nasal cavity. The lateral palatal shelves fuse in an anterior-to-posterior direction which form the hard and soft palate. The nasal cavity formed as the lateral palatine processes fuse in posterior direction. There is absorption of thin membrane which separates the oral and nasal cavity forming the posterior choana [6]. The factors which may cause atretic choana are medial outgrowth of the vertical or horizontal process of the palatal bone and persistent mesenchyme with misdirection of developmental flow [7].

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Pathophysiology

The epiglottis is more superiorly placed in the neonate as compared to adult. When infant swallows, the larynx rises above epiglottis and touch the nasopharynx and locks between soft palate and side of nasopharynx. During inspiration neonate sucks the tongue and the vacuum is created in the oropharynx which helps to move soft tissue of the floor of mouth up and back towards soft palate. During expiration the pressure in the airway causes soft palate to push forward against the soft tissue and tongue in the mouth, also obstructing the oral airway [8]. The oral airway is invariably blocked in quite respiration as the infant is obligate nasal breather. The infant's breath through the mouth only during crying. The 1/3rd reduction in the diameter of the nasal airway increase nasal airway resistance by 81 times resulting in severe distress in neonates. The medical emergency would exist in complete absence of patent both nasal cavities. The infant will become cyanotic which is relieved by crying or gasping when the child opens the mouth widely. Crying instantly releases the air obstruction and the cyanosis disappears. If the crying stops, the mouth closes and the cycle repeats itself. In resting state child has severe retractions, struggles to breath with rapid development of cyanosis. The infant with bilateral choanal atresia develops rarely an ability to breathe through mouth [9].

47% of patients with choanal atresia without chromosomal abnormalities have CHARGE Syndrome. The CHARGE Syndrome is comprised of *Coloboma* of the Iris with or without microphthalmia, *Heart* defects (i.e., atrial septal defects), choanal atresia (*Atresia* of the choana), *Retarded* growth and abnormalities of the *Genitourinary* system (i.e., cryptorchidism, microphallus, hydronephrosis) and *Ear* abnormalities with associated deafness.

It is very important to diagnose choanal atresia immediately after birth. If there is any doubt regarding the patency of neonate's airway by the paediatrician or any supporting staff a catheter should be passed through each nostril into the child's oral cavity. To confirm the diagnosis of suspected Choanal atresia lateral X-ray is taken after radioopaque dye is put into the nasal cavity. If an atresia is demonstrated a computerized tomography scan would further delineate characteristics of choanal atresia (Fig. 1).

The benefits of obtaining a CT Scan are as follows:

1. Confirm the diagnosis and if it is on one or both sides.
2. Determine the anatomy of the atretic area, including the width of the Vomer bone and the medial displacement of the sides of the lateral wall of the nose.
3. To measure the thickness of the atretic plate and the presence and thickness of a bony plate.

Fig. 1 Computerised tomography showing bony atretic plate

4. To determine if any other sites of obstruction or abnormalities exist in the nasal, nasopharyngeal or sinus cavities.

Materials and Methods

It is a retrospective study of 22 cases of choanal atresia over the period of 20 years. It is one of the largest series of choanal atresia in the world. All cases were operated by the first author. All the patients details including sex, age at presentation, type and site of atresia, associated congenital abnormalities, method of surgical repair and postoperative complications were maintained for analysis and interpretation. A 3-tier system was used for diagnosis. In the first tier diagnosis was made by clinical presentation of neonates. In the second tier suspected cases it was attempted to pass a small suction catheter through the nares. And in the 3rd tier confirmation was done by CT Scans and Nasal Endoscopy. Though there are various methods of surgery like transnasal, transpalatal and transseptal, only transnasal route was used in all the patients. Curved Hegar's dilators from obstetrics and gynecology were used for perforating the atretic plate and to dilate the opening following it. A 0° 2.5 mm nasal endoscope was used for examination prior to and after the dilatation. The stenting after the perforation of atretic plate was done by Portex endotracheal tubes by using the procedure of "Vikas Sinha Stenting" in all the patients. Period of stent placement depended on the patient and generally varied between 7 and 15 days. Prophylactic postoperative antibiotic cover was given to all the patients. All the patients needed stay in Intensive Care Unit for 48–72 h, while some patients had a prolonged stay. Few needed subsequent serial dilatation at 6 weeks based on Nasal Endoscopy and CT scan findings.

Surgical Technique

The surgery was performed under general anaesthesia with cuffed endotracheal tube to prevent the aspiration of blood during surgery. Hegar's dilators from the Department of Obstetrics and Gynaecology was used to perforate the atretic plate. Hegar's dilators were used in all the cases to perforate the atretic plate. The Hegar's dilator has the ideal curvature to follow the sloping contour of nasal floor. The nasal endoscopy was performed in all cases to reconfirm the diagnosis and to identify clinically bony/membranous atretic plate. The typical hard bony feeling by the suction canula gives the fair amount of idea of bony/membranous atretic plate. The smallest size of Hegar's dilator was used to perforate the atretic plate (Fig. 2a). The atretic plate is almost always thinnest and weakest at the junction of floor and posterior end of septum and can be easily perforated. The dilator is passed along the floor of the nose staying against the septum to avoid penetrating the basal-sphenoid. The floor of the nose of infant is not straight but curved, paralleling the curve of the hard palate. The index finger of the left hand was placed at the nasopharynx (Fig. 2b) against the nasopharyngeal side of the atretic plate mainly to safeguard accidental sudden perforation and prevent the accidental hitting the base of skull. When the atretic plate was touched by the dilator, the dilator is carefully forced through it with gentle uniform pressure and screwing movement till the atretic plate is perforated. The atretic plate is serially punctured and dilated using increasing sizes of Hegar's dilators. Both sides were dilated serially to prevent shifting of the septum. The maximum size of dilator (Fig. 2c) used for the dilatation approximates a size

that is slightly smaller than the child's nostril. A similar procedure is repeated on the other side. Surprisingly there is little bleeding during entire procedure.

Stenting Procedure

A non-cuffed Portex endotracheal tube is used for the stenting. The size of endotracheal tube should be approximately the size of last Hegar's dilator used. The tube is folded exactly equal from the mid point thus making two exactly equal lengthly. The posterior fenestration (outer curved side) of the tube at the mid-point was created with a No. 11 blade (Fig. 3a) for the air to pass from nose to the nasopharynx while anterior part of the tube (inner curved end) remains in continuity (Fig. 3b). Smaller vents or holes were made in a different site and the axis for the drainage of nasal secretions.

Two red rubber catheters are passed from both nasal cavity to be taken out from the oral cavity (Fig. 4a). The two ends of newly created stents were tied with the red rubber catheter and pulled retrograde from the nasal cavity (Fig. 4b). The both rubber catheters are pulled together and simultaneously so that posterior fenestration lies exactly against the midline of nasopharynx. Care should be taken not to twist the stent. An equal length of two tubes outside the nose will ensure that posterior fenestration is in the midline and nasopharynx. A small red rubber catheter is passed through the stent to prevent the blockage from the blood clot (Fig. 4c). The tube is secured at the nostril with silk sutures (Fig. 4d). The two ends of the tubes are also sutured together to prevent posterior slipping of stent at the nasopharynx. The continual friction of silk suture against the columella causes pressure necrosis of the columella

Fig. 2 **a** Insertion of the smallest size Hager's dilator. **b** The surgeon's finger placed in the nasophaynx as a safe guard. **c** Dilatation with Hager's dilators

Fig. 3 **a** Creation of fenestra in endotracheal tube. **b** Endotracheal stent showing vent at posterior wall of endotracheal tube for air to pass through. The anterior wall in continuity. The small vents at different sides of tube for nasal secretion drainage

Fig. 4 **a** Red rubber catheters passed from nose to oropharynx. **b** The red rubber catheters are attached to the stent, so the stent can be position in the choana when they are pulled through nose. **c** The two ends of stent are delivered through nose, centering the mid portion at choana for respiration. **d** The stent is sutured with silk sutures

which can be prevented by the placing a small piece of red rubber catheter over the columella. The excess length of tube is cut. All the post op neonates were kept in the Neonatal Intensive care unit for regular suction of nasal passage for 1 week.

Results

All the cases had established air passage completely on the operation table. This was the criteria used for successful operation. Out of total 22 cases, 14 (64%, $n = 22$) cases

were male and 8 (36%, $n = 22$) females. The age variations varied from youngest of 3 days to oldest 31 year old female. 10 (45%, $n = 22$) patients had bilateral choanal atresia while 6 (27.5%, $n = 22$) cases had right side unilateral and 6 (27.5%, $n = 22$) cases left sided unilateral; choanal atresia. Most commonly diagnostic maneuver used to establish the diagnosis was the inability to pass a rubber catheter through the nose into the nasopharynx. Nasal endoscopy was performed in all cases with 0° Endoscope. CT scan were performed in all cases which depicted eight cases as bony and fourteen cases were of mixed variety i.e., both bony and membranous and the findings were further confirmed postoperatively. All the cases were operated endonasally along with perforation of atretic plate using Hegar's dilators. In all the cases adequate airway was established on the table. No life threatening intraoperative complication like skull base injuries or spinal injuries were observed in any of the cases operated. In all the patients, subsequent stenting was done using Portex non-cuffed endotracheal tube by 'Vikas Sinha stenting technique', the size of the tube was approximately the size of the last Hegar's dilator used while dilatation. Eight cases (36%, $n = 22$) of neonates were associated with CHARGE Syndrome and all failed to survive for more than 5–10 days due to complications of CHARGE Syndrome and unrelated to the outcome of the surgery. Stents were kept for a period of 6–8 weeks and regular follow up nasal endoscopy was done weekly for suctioning and visualizing the size of the airway. Eight patients (36%, $n = 22$) required repeated dilatation after 6 weeks of dilatation and stenting. Columellar necrosis developed in three cases (14%, $n = 22$) which compelled us to remove the stent around 3 weeks after dilatation in both the cases. All the patients were regularly followed up for a period of 12–24 months. In three patients, granulations developed around the stent. The stent was removed subsequently and Foleys catheter was used as a stent where the balloon part of the catheter was insinuated at choana like a dumbbell, where the balloon part of the catheter was on either side of choana.

Discussion

It was first described by Flake and Ferguson [10], later the first transnasal operative repair was performed by Emmerth. It is of utmost importance to be aware of this condition or else it may lead to disastrous consequences like death of a child. An oropharyngeal tube or airway must be used as an emergency procedure. The diagnosis of this condition can be done clinically by placing a laryngeal mirror under to nostril to check for fogging or inability to pass a catheter through nose. Choanography can be used followed by CT Scan to confirm. One of the major

complications of choanal atresia is restenosis of the atretic plate. Significant debate exists in the literature on whether placing stents in the operative site after the procedure provides improved outcomes. Stents are usually made out some type of plastic tubes. Portex and manipulated endotracheal tubes are often used. As a proponent of stenting, Friedman et al. [11] reviewed 46 cases of choanal atresia and cited a favorable prognosis (defined as less operations needed) with children who had stents in for greater than 12 weeks. Later, Gujarathi et al. [12] used stents in place for a median of 12 weeks. In addition to stents, the anti-neoplastic agent Mitomycin C has been used as a treatment adjunct to try to decrease stenosis. Mitomycin works by inhibiting fibroblast growth and proliferation, which may help keep granulation tissue at bay.

The five different surgical approaches used are Trans-nasal, Trans-palatal, Trans-septal, Trans-antral, Sublabial-Transnasal [11, 13]. The most popular are the trans-nasal approach. Hegar's dilator was used in all our patients as Hegar's dilators curve attunes with the natural curve of the palate and the floor of the nose. Its blunt curvature has less tendency of damaging surrounding structure. In our study the male female ratio 1.5:1 which is compatible with previous studies done by Sinha [14], Goseph et al. [15] (2:1). In our study bilateral atresia was seen in 45% ($n = 22$) and unilateral in 55% ($n = 22$) which is comparable to Newman et al. [16] (44% bilateral cases, $n = 43$). Portex endotracheal uncuffed tube was used as stent and its size was dependent on size of last Hegar's dilator which could be introduced without resistance as done by Sinha [14], Gujarati et al. [12] and Sharma et al. [17]. Nasal stents were used for a period of 6 weeks which is comparable to Al-Ammar [18] and earlier study of Sinha et al. [14]. Use of stent is controversial [19] as stated in literature because it may cause discomfort, localized infection and ulceration as in our study 3 (14%, $n = 22$) developed columellar necrosis. Eight cases (36%, $n = 22$) requires revision dilatation after 6 weeks of stenting, variable results have been reported in study of Uzimefuna et al. [20] (57% in cases with stent and 33% in cases without stent) and Velegrakis et al. [21] (57% in bilateral and 25% in unilateral). A stable child can be discharged within 1 week however the patients parent should come back immediately if baby has difficulty in breathing, baby has yellowish ooze from nose, baby has bleeding from nose, baby has high temperature of 37.5 °C or more, stent becomes loose or falls out. Some of the major complications with use of stents include foreign body tissue reaction, Nasal alar erosion, columellar pressure necrosis. If skull base is violated by sharp instruments or too much force on bone it may result in CNS Trauma.

Conclusion

“Birth without breath” is the precise description of choanal atresia. The diagnosis and management of this condition needs the combined efforts of both the paediatrician and E.N.T. surgeon. Amicable and prompt referral to the ENT surgeon can be of immense value so as to buy adequate time for the surgeon to decide and act as per the patient’s clinical and radiological findings. Whenever faced by child with nasal/upper airway obstruction or respiratory distress choanal atresia must be considered in the differential diagnosis. If bilateral atresia is found, this is considered an upper airway emergency and the proper measures should be taken to stabilize the patient related with other features of CHARGE Syndrome first before considering surgical intervention. The surgery for choanal atresia should be undertaken as soon as the diagnosis is made. Intranasal Hegar’s dilator method is a very safe method to perforate the atretic plate whether bony or membranous. Controlled pressure of the Hegar’s dilator using a twisting movement is the key for the successful controlled perforation of the atretic plate. Repeated serial dilatation may be performed in cases where early signs of restenosis can be visualized on CT scan and nasal endoscopy.

When undergoing surgical treatment, one must consider the most effective approach and possible treatment adjuncts like Mitomycin C and stenting to give the patient the best outcome possible. Stenting with either an endotracheal tube or balloon is mandatory to prevent restenosis. Sometimes excessive granulation tissue forms and this can be prevented by usage of soft Polyvinyl Chloride (Portex) tube for stenting, avoiding excessive damage of mucosa during surgery and lastly by use of broad spectrum antibiotics during the whole period of stenting.

Compliance with Ethical Standards

Conflict of interest The authors declare that they have no conflict of interest.

Ethical approval All procedures performed in studies involving human participants were in accordance with the ethical standards of the institutional and/or national research committee and with the 1964 Helsinki declaration and its later amendments or comparable ethical standards.

Informed consent Informed consent was obtained from all individual participants included in the study.

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