Choanal Atresia - Corrective Surgery Using Hegar's Dilators


*Dean, Professor E.N.T., **Assistant Professor, ***Professor and Head, ****Junior Resident, *****Senior Resident

Institution: Department of Otolaryngology and Head and Neck Surgery, M.P.Shah Medical College, Jamnagar (Gujarat), India

Corresponding Author:
Dr. Vikas Sinha
Dean, Prof E.N.T.
M.P.Shah Medical College
Jamnagar (Gujarat), India
E mail dr_sinhavikas@yahoo.co.in

Abstract

Background and objective: Bilateral choanal atresia is a medical emergency and surgery is the only means of treatment for this condition. Hegars dilator was used in all cases to break the bony and membranous atretic plate.

Material and Method: Seven cases of choanal atresia were studied. Six cases were 1-5 days old with bilateral complete choanal atresia. They all required immediate surgery as they had repeated attacks and cycles of cyanosis. Four neonates were associated with the CHARGE syndrome. All of the cases were done under general anesthesia. Hegars dilators were used in all cases and nasal stents were placed in all cases.

Results: Although complete nasal patency was achieved by surgery, in four cases, neonates could not survive due to the CHARGE syndrome. The mortality was unrelated to the surgery. The two cases which were not associated with CHARGE syndrome had a good postoperative recovery. One case was 14 years old with a unilateral, complete bony and membranous choanal atresia. This patient required surgery due to continuous nasal discharge.

Conclusion: Hagers dilators are a safe and simple method of surgery for choanal atresia. Nasal stenting is mandatory to prevent restenosis.
Introduction

In 1775, Roeder first described Choanal atresia and in 1829 the entity was further defined by Otto. The derivation of the word "Choana" is Greek from the word Xovan, meaning funnel. The term "Posterior Choana" would literally mean a posterior funnel. However, its usage in modern day medicine is somewhat of a misnomer, since its meaning is the posterior aperture of the nasal cavity. Choanal atresia is one of the more commonly observed congenital abnormalities of the nose. In the neonate it is life threatening, since neonates are obligate nasal breathers for the first six weeks of life, and will enter a state of severe respiratory distress after the crying from childbirth has ceased. The incidence of this abnormality is between one in 5000 to one in 8000 births.

Embryology: The embryonic origin of choanal atresia is thought to be a persistent bucco-pharyngeal or naso-buccal membrane. During embryologic development, the nasal cavities form from nasal placodes on the lateral surface of the head. These placodes sink into the mesoderm and form the nostrils and the anterior nasal cavity. This process is influenced by the lateral palatal shelves which fuse in an anterior-to-posterior direction and form much of the hard and soft palate. The nasal cavity is influenced and formed as the lateral palatal processes fuse in a posterior direction.

The membrane which separates the oral and nasal cavities thins and is finally reabsorbed. The reabsorption of this membrane forms the posterior choana. If the membrane fails to reabsorb then a choanal atresia will be present. Factors which may cause an atretic choana are a medial outgrowth of the vertical or horizontal process of the palatal bone and persistent mesenchyme with misdirection of developmental flow.

Pathophysiology: In the neonate, the epiglottis is more superiorly positioned then the adult. When an infant swallows, the larynx rises and the epiglottis enters the nasopharynx and locks between the soft palate and the sides of the nasopharynx.

During inspiration, a neonate sucks the tongue and a vacuum is created in the oropharynx. This helps to move the soft tissues of the floor of the mouth up and back towards the soft palate. During expiration, the pressure in the airway causes the soft palate to push forward against the soft tissues and tongue in the mouth, also obstructing the oral airway.

The oral airway is blocked with quiet respiration. The infant is an obligate nasal breather, and only breaths through his mouth during crying. Reduction in the diameter of the nasal airway by one third can increase nasal airway resistance by 81 times and result in respiratory distress in neonates. Nasal stenosis has the same effect and if both nasal cavities have a complete absence of a patent airway, a medical emergency exists. The infant will become cyanotic, which is broken by crying or gasping when the mouth is open widely. In a resting state, the child has severe retractions, struggles to breath with the rapid development of cyanosis. Crying instantly relieves the airway obstruction and the cyanosis disappears. If the crying stops, the mouth closes and the cycle repeats itself. Rarely, an infant with bilateral choanal atresia develops the ability to mouth breath. If this happens the medical emergency subsides but the child still requires treatment.

Forty-seven percent of patients with choanal atresia without chromosome anomalies have the 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 imperative to diagnosis choanal atresia immediately after birth. If any doubt regarding the patency of a neonate’s airway exists in the recovery room, a catheter should be passed through each nostril and into the child’s oral cavity. The passage of the catheter down each nostril should be performed on every newborn in the the recovery room.

To confirm the diagnosis of a suspected choanal atresia, a lateral x-ray is taken after radiopaque dye is placed into the child’s nasal cavity. If an atresia is demonstrated, a Computerized Tomographic scan is obtained to further delineate the characteristics of the choanal atresia. For the best results, the nasal cavity should be suctioned, immediately before the test to remove any nasal secretions.

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

Methods

This is the prospective study of 7 cases of choanal atresia. The 6 cases had bilateral bony choanal atresia and all were of 1-5 days old. One case was an adult, 14 year old girl, having unilateral bony and membranous choanal atresia. All the surgery was done by the first author. Each patient was investigated in detail with respect to sex, age of presentation, type and site of atresia, and associated other congenital anomalies. The diagnosis was made clinically by the inability to pass a small suction catheter through the nares and confirmed by CT scanning and nasal endoscopy. The immediate management of a child with respiratory distress was the insertion of an oral airway.

All patients underwent surgical repair of choanal atresia through a transnasal technique. A curved Hegars dilator from obstetrics and gynecology was used to perforate the atretic plate, transnasally, in bony and membranous atresia and subsequently to dilate the opening. A 4 mm, 0 degree nasal endoscope was used for visualization before and after the dilatation. All patients were stented using portex endotracheal tubes. They were inserted by using the procedure of “Vikas Sinha Stenting” for a period of 7 to 15 days depending on the tolerability of the patient. All patients were given postoperative prophylactic antibiotics.

Surgical Technique: Surgery was performed under general anesthesia with a cuffed endotracheal tube to prevent the aspiration of blood during surgery. Hegars dilators were used to perforate the obstructing plate (Fig 1-a and Fig 1-b). Hegars dilators have the ideal curvature to follow the sloping contour of the nasal floor. The smallest size of Hegars dilator was used to perforate the atretic plate (Fig 2-a). The atretic plate is almost always thinnest and weakest at the junction of the floor and the posterior end of the 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. It must be remembered that the floor of the nose of the 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 2-b) against the nasopharyngeal side of the atretic plate mainly to safeguard accidental sudden perforation and prevent the accidental hitting or damage of the base of the skull. When the atretic plate was touched, the dilator was carefully forced through it with gentle uniform pressure and screwing movement till the atretic plate was perforated. The atretic plate is serially punctured and dilated using increasing sizes of Hegars dilator (Fig 2-c). Both sides were dilated, serially, to prevent shifting of the septum. The maximum size of the dilator used for the dilation approximates a size that is slightly smaller than the child's nostril. A similar procedure was repeated on the other side. Surprisingly, there is little bleeding during the entire surgery.

12 Year Old Indian Female With Left Choanal Atresia

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Figure 1-A: Pre operative bony atretic plate
Figure 1-B: Pre operative bony atretic plate

Figure 2-A: Insertion of the smallest size Hager's dilator
Figure 2-B: The surgeon's finger placed in the nasopharynx as a safeguard
Figure 2-C: Dilatation with larger sized Hager's dilators

Stenting procedure: Vikas Sinha stenting - A noncuffed portex endotracheal tube was used for stenting. The size of the tube should be approximately the size of the last Hegars dilator used. The tube is folded exactly equal from the midpoint thus making two exactly equal lengths. The posterior fenestration (outer curved side) of the tube at the midpoint was created with a No. 11 blade (Fig 2-d) for the air to pass from the nose to the nasopharynx while the anterior part of the tube (inner curved end) remains in continuity² (Fig 2-e). Smaller vents or holes were made in a different site and axis for the drainage of nasal secretions. Two red rubber catheters are passed from both nasal cavities to be taken out from the oral cavity (Fig 2-f). The two ends of the newly created stent were tied with a red rubber catheter and pulled retrograde from the nasal cavity. Both rubber catheters are pulled together and simultaneously (Fig 2-g) so
that posterior fenestration lies exactly in the midline at the nasopharynx. Care should be taken so as not to twist the stent. An equal length of the two tubes outside of the nose will ensure that the posterior fenestration is in midline and in the nasopharynx. A small red rubber catheter is passed through the stent to prevent blockage from a blood clot (Fig 2-h). The tube is secured at the nostril with silk sutures (Fig 2-i) taking care not to damage the columella by placing a small gauze piece at the columella. The continual friction of silk suture against the columella causes pressure necrosis on the columella. The excess length of tube is cut. All the postoperative neonates were kept in the neonatal intensive care unit for regular suction of the nasal passage for one week.
Surgical Technique: View YouTube Video: https://youtu.be/0NOLGTskRuA


Results

All the cases had established air passage completely on the operation table. This was the criteria used for the successful operation. Four cases of neonates were associated with CHARGE syndrome. They all died within 2-4 days due to CHARGE syndrome unrelated to surgery. Two children, who were otherwise normal, survived this postoperative period and stenting was continued for 4 weeks and afterword it was removed. One adult patient had the stenting continued for 3 weeks but postoperative columella necrosis developed (Fig 3-a) which forced us to remove the stent. The columellar skin was sutured with primary silk sutures (Fig 3-b) and healing was excellent (Fig 3-c) due to very good blood supply of this region. The nasal balloon catheter was used (Fig 4-a) for a further 2 weeks to prevent restenosis (Fig 4-b). Nasal endoscopy was done every 2nd or 3rd day to ensure proper positioning of the nasal balloon (Fig 4-c). The balloon was removed in-between to visualize any possible restenosis. The Karrison bone punch was used under direct vision of the 0 degree 4 mm nasal endoscope to nibble any bony growth. Saline nasal drops were used as needed to prevent nasal crusting. Nasal endoscopy showed a wide nasal cavity at the choanal (Fig 5-a). CT scan confirmed a successful surgery with an absence of choanal atresia (Fig 5-b).
Figure 3-A: Columellar necrosis

Figure 3-B: Fine silk sutures at columella

Figure 3-C: Excellent healing and final appearance of the columella

Figure 4-A: Anterior nasal balloon catheter

Figure 4-B: Anterior nasal balloon catheter placed into the naries

Figure 4-C: The balloon portion of anterior nasal balloon catheter snuggly fitting the choana

Figure 5-A: Postoperative view of the patient’s wide choana

Figure 5-B: Six week post operative CT Scan showing a wide nasal airway

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Discussion

Roederer first described choanal atresia in 1755 and Emmerth first performed a transnasal operative repair. He used a curved trocar to perforate the membranous atretic membrane.11,12

There are a number of methods to diagnosis choanal atresia on physical examination: A laryngeal mirror can be placed under the nostril to test for fogging, failed attempts to pass a catheter through the nose, or the absence of movement of a wisp of cotton placed in front of the nostril. The time-tested method for diagnosis is choanography where contrast material is placed in the infant's nostril and a lateral x-ray is obtained. If this test is positive, a CT scan should be performed, which will give an accurate measurement of the atresia’s thickness along with its composition (membranous or bony).

There are five different surgical approaches that have been described for surgical treatment of choanal atresia: (1) trans-nasal; (2) trans-palatal; (3) trans-septal; (4) trans-antral and (5) sublabial-transnasal.13,14 The two most popular techniques that are used in newborn infants with choanal atresia are trans-palatal and trans-nasal approaches. With the advent of the endoscope and powered instruments, the transnasal approach becomes more popular. It provides direct and excellent visualization of the nasal cavity and choana. Hegars dilator was used in all our patients. The dilator's curve conforms to the natural curve of the palate. It is also blunt so there is less of a chance of damaging surrounding structures. We determined the size of the stent by the size of the last number of Hegar's dilator used for dilatation. The Hegars' dilators were also used by Shama, et al., in a large series of 14 patients with choanal atresia.15 There is some controversy regarding the use of a stent. Because a stent may cause discomfort, localized infection and ulceration, along with circumferential scar tissue and injury to surrounding normal tissue. A properly sized stent left in the nasal cavity maintains a circumferential haemostatic pressure.2 Once removed may cause restenosis. Indwelling nasal stents are difficult to manage and may migrate or break.11 However most surgeons are using stent in the post-operative period.16 The overall failure rate of surgery using stents is about 30%.17 Tracheostomy has little place in the management of airway problems caused by choanal atresia. Endotracheal intubation is contraindicated unless the infant's condition is so poor or there is an unrelated airway obstruction.18

Conclusion

The surgery for choanal atresia should be undertaken by the E.N.T. surgeon as soon as the diagnosis is made by neonatologists or pediatricians. The Hegars dilator is a very safe method to perforate the atretic plate whether bony or membranous. The finger on the surgeon's other hand is inserted into the nasopharynx to prevent any damage to the base of the skull in case of accidental sudden perforation of the atretic plate. Controlled pressure of the Hegars dilator using a twisting movement is the key for the successful controlled perforation of the atretic plate. Stenting with either an endotracheal tube (“Vikas Sinha Stenting”) or balloon is mandatory to prevent restenosis.

References


Figure 1A
Figure 1B

Figure 2A