Chapter 4 – Surgery for Choanal Atresia

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Since the first description of choanal atresia by Roederer in 1755, more than 350 publications have described multiple surgical techniques for correcting this congenital defect.[1,2] Bilateral choanal atresia is a medical emergency. The diagnosis of bilateral choanal atresia requires a high index of suspicion and it should be suspected by the pediatrician in the delivery room if the child has the characteristic cyclic apnea, cyanosis, and respiratory distress, which are temporarily relieved by crying.

The embryologic basis for choanal atresia is not entirely clear, but Hengerer and Strome suggest four "theories" to explain it: (1) persistence of the buccopharyngeal membrane from the foregut, (2) persistence of the nasobuccal membrane of Hochstetter, (3) abnormal persistence of mesoderm-forming adhesions in the choanal region, and (4) misdirection of mesodermal flow secondary to local factors.[3] An excess of this displaced mesenchymal tissue in the midfacial region creates a tangential atretic plate that blocks the nasal airway from the posterior vomer across the lateral nasal wall. Ninety percent of these plates are bone and can be as thick as 12 mm.

In addition to the atretic plate, other anatomic changes may include a diminished nasal airway secondary to septal deflection toward the obstructed side, a widened vomer, medial bowing of the lateral nasal wall, and narrowing of the nasopharynx. This combination of anatomic variations supports the concept of a generalized defect in mesenchymal cell flow involving development of the skull base. It is quite important to understand not only the anatomy and embryology but also these associated abnormal anatomic findings to safely correct choanal atresia.

Congenital atresia is uncommon and occurs once in every 5000 to 10,000 live births.[4] Approximately 50% of patients with choanal atresia have additional congenital anomalies such as Treacher Collins syndrome, Apert's syndrome, trisomy D, and the cluster of defects known as the *CHARGE association* (*c*olobomas, congenital *h*eart defects, choanal *a*tresia, *r*etarded development, *g*enital hypoplasia, and *e*ar anomaly).[5] Another important single associated anomaly is cardiovascular malformation, which is 20 times more frequent in infants with choanal atresia than in the general population. Mental retardation, micrognathia, deafness, cleft palate, and facial palsy may also be encountered as single associated anomalies.[6] Unilateral choanal atresia may not be detected until long after the newborn period despite long-term unilateral rhinorrhea and nasal obstruction.[7]

As part of the standard evaluation of newborn infants, a catheter should be passed through each nostril into the nasopharynx. Infants with choanal atresia or severe stenosis may be identified with this maneuver. Infants are obligate nasal breathers, and nasal obstruction produces respiratory distress and marked feeding difficulty. An oral airway, once placed and secured, stabilizes the patient and allows complete evaluation and surgical planning.[8] A firm, curved dilator can be used to gently palpate the atretic plate, or the area can be inspected with a telescope once the mucus has been aspirated from the nasal airway.[9] Djupesland and colleagues described the use of acoustic rhinometry.[10] They compared acoustic measurements in five infants with congenital respiratory distress caused by bilateral choanal atresia with their computed tomography (CT) scans and concluded that acoustic rhinometry represents a new and valuable tool in the diagnosis of congenital choanal atresia. This technique may also be helpful in the postoperative evaluation.

Cotton and Stith described the use of a standard endotracheal tube placed transorally into the esophagus as their choice of airway in the case of bilateral choanal atresia.[6] The tube acts as an airway by stenting the patient's oral airway open and may also be used for gavage feeding.

Unilateral choanal atresia may go unrecognized until later in life because the associated respiratory distress may not be present at birth. When this condition goes unrecognized, patients are initially seen later in childhood or as teenagers with unilateral nasal obstruction and chronic rhinorrhea. Unilateral atresia is more common than bilateral atresia. This disorder affects females more often than males and is found more commonly on the right side when it occurs unilaterally. A bony atretic plate is found in 90% of patients, and a membranous plate is found in the remaining 10%.[11]

The most widely accepted method of correcting the problem of unilateral or bilateral choanal atresia has been the transpalatal technique. However, with more recent technologic advances, such as endoscopes for functional endoscopic sinus surgery (FESS) and the microdébrider, transnasal endoscopy has emerged as an equally acceptable technique and is particularly appealing because of widespread familiarity with endoscopic sinus surgery by present-day otolaryngologists. A survey of pediatric otolaryngologists was completed in 1999 to determine their preferred techniques for correction of choanal atresia. Endoscopic techniques were favored (85%) over transpalatal repair (Fig. 4-1).[7]

Pirsig pointed out that the ideal procedure for choanal atresia should create a normal unobstructed nasal passage, prevent damage to any growing structures important in facial development, be technically safe, and require a brief operative time and a short hospital stay and convalescence.[12]

A constant gradual evolution plus reevaluation of the management of patients with choanal atresia is apparent. Beinfield in 1956 performed transnasal curettage with progressively larger curettes to perforate the atretic plate. The choanae were stented open for several months.[13] Other transnasal approaches that have been described for the treatment of choanal atresia include the external rhinoplasty technique, the transseptal technique, and the sublabial approach.[9]

Richardson and Osguthorpe[8] performed a transnasal drill-out procedure with an operating microscope and otologic instrumentation. Stankiewicz in 1990[1] described a similar procedure that incorporated modern endoscopic telescopes for transnasal removal of the atretic plate. Such instruments provide excellent visualization of the entire atretic plate and make it possible to perform precise surgery on patients of all ages. These techniques are generally applied to infants withbilateral choanal atresia to obviate the necessity for prolonged use of an oropharyngeal airway.

More recently, Fong and coauthors reported using the holmium: YAG laser to correct choanal atresia in eight patients.[14] The authors emphasized the safety of the procedure and the advantage of a flexible fiberoptic system, good hemostasis, and better bone-cutting characteristics than with the CO_2 laser, which is in widespread clinical use.

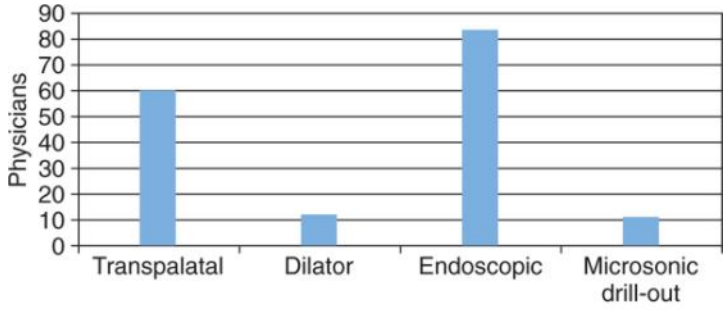

Figure 4-1 Survey of the number of physicians who perform the specific type of procedure for repair of choanal atresia.
(Reprinted with permission from Park AH, Brockenbrough J, Stankiewicz J: Endoscopic versus traditional approaches to choanal atresia. Otolaryngol Clin North Am 33:77-90, 2000.)

PATIENT SELECTION

An early surgical approach to bilateral choanal atresia is currently advocated for several reasons. With medical management, the risk for pneumonia as a result of inadequate pulmonary ventilation and inhalation of food is high in these patients. The medical approach may also downgrade the chance for survival, particularly if the patient has other congenital anomalies. Improvement in techniques of neonatal anesthesia is another factor that has enhanced the safety of infants undergoing early surgery for this condition. Arguments against early surgical repair include claims of higher rates of choanal stenosis. The procedure should be delayed until associated medical problems have been stabilized.

It is also important that the patient be thoroughly evaluated for the more severe congenital anomalies, such as cardiac abnormalities, that can be seen in association with choanal atresia. Other craniofacial anomalies should be considered when planning the surgical approach but should not delay definitive treatment. A child with unilateral atresia who has significant symptoms should also be operated on soon after diagnosis. Surgery remains the definitive treatment once other medical factors have been resolved or at least stabilized.

April and Ward in 1996 introduced a technique involving the use of powered instrumentation with protected blades and drills that allows more precise removal of the atretic plate, thereby contributing to a more satisfactory surgical procedure.[15] Rombaux and associates reported a series of seven children with both bony and mixed soft tissue/bony unilateral choanal atresia who were treated between 1999 and 2001 with powered instrumentation.[16] A dissector tip was used to open the posterior choana in the inferior medial aspect of the atretic plate. A microdébrider was then used to enlarge the opening. The opening was coated with mitomycin C and left unstented. There was no excessive operative bleeding. One patient required a repeat endoscopic procedure 9 months later because of stenosis, and minor synechiae developed in another patient but did not require treatment. No other complications were reported. The mean number of surgical repairs per patient in the powered instrumentation group was 1.14, and the success rate after 12 months was 85.7%; in contrast, in the standard treatment of unilateral choanal atresia without powered instrumentation (n = 19), the mean number of surgical procedures per patient was 1.89, and the success rate at 12 months was 47.3%.[16] Because of the small number of patients, however, the results did not reach statistical significance. In addition, biases favoring a more recent technique such as the microdébrider may be due to other improvement in techniques, such as computer-assisted navigation. Nonetheless, there is a trend toward superiority of results with use of the microdébrider, as well as the appeal of using this instrument because of improved visualization and efficiency of tissue removal. The authors suggest that the reason for improved success in the treatment of unilateral choanal atresia with powered instrumentation may be that it allows faster healing with less scarring, thus reducing the risk for stenosis.[16]

PREOPERATIVE PLANNING

Before the availability of CT, propyliodone in peanut oil (Dionosil Oily) was instilled in the nasal cavity after the mucus was removed, and a cross-table lateral view of the patient was taken. Failure

of propyliodone to enter the nasopharynx was taken as evidence of posterior choanal atresia; however, this rather simple technique did not always provide the information necessary for planning the management of these patients.

Radiographic evaluation of choanal atresia is now carried out with CT to help differentiate bony from membranous atresia. This is important for surgical planning because management of membranous atresia is less complex. CT also identifies alterations frequently present in the posterior aspect of the nasal septum and reveals the configuration of the lateral nasal wall and nasopharynx (Fig. 4-2).[17] Brown and coworkers reviewed CT scans of 47 patients with choanal atresia from the literature and 16 from their own clinical experience and classified the atresia as pure bony, mixed bony-membranous, and pure membranous.[18]

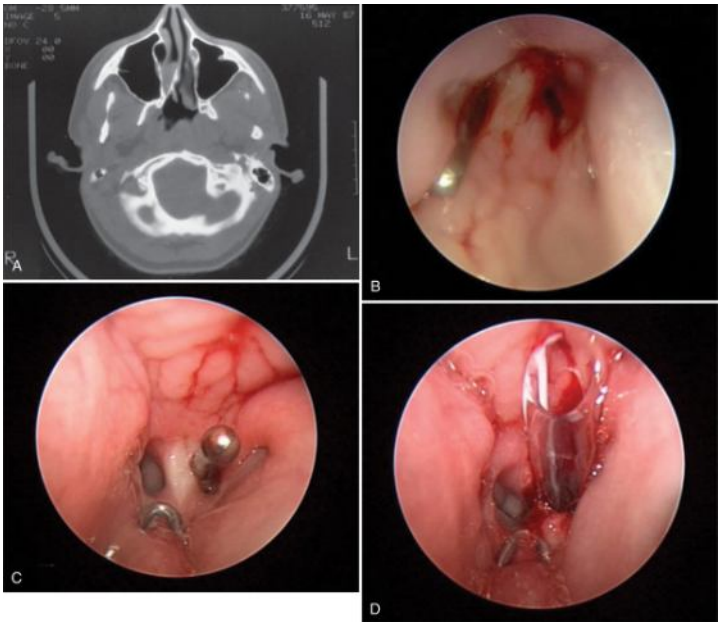

Figure 4-2 **A**, Computed tomography scan demonstrating bony choanal atresia of the right side. **B**, Ninety-degree telescopic view of unilateral choanal atresia. **C**, Puncturing choanal atresia. **D**, Stenting of the posterior choana.

SURGICAL TECHNIQUE

Transnasal Puncture Technique

Under general anesthesia, a small Lempert curette or urethral sound is placed through the nose to puncture the atretic plate. The surgeon's index finger is inserted into the nasopharynx to protect the skull base (Fig. 4-3). After puncturing the atresia, a stent is inserted to prevent restenosis. Transnasal puncture usually requires repeated revisions or dilatation. Complications include leakage of cerebrospinal fluid and meningitis. This is ablind technique, and in the era of nasal endoscopy, it is contraindicated.

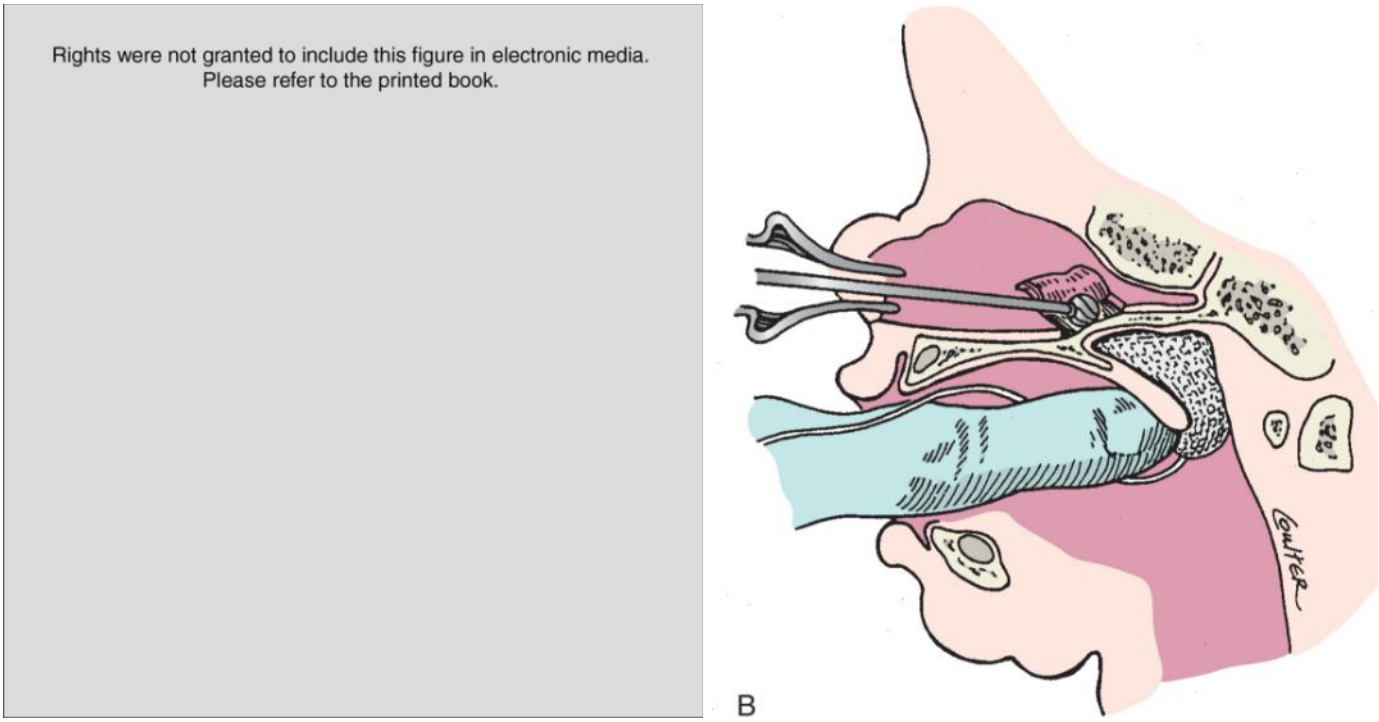

Figure 4-3 **A** and **B**, Removal of atretic bone inferiorly and medially.
(Reprinted with permission from Kenna MA, Rahbar R: Congenital nasal malformations. In Bluestone CD, Rosenfeld RM [eds]: Surgical Atlas of Pediatric Otolaryngology. BC Decker, Ontario, Car 2002, p 303.)

Transpalatal Technique

The patient is placed in a modified Trendelenburg position with the shoulders extended over a folded towel. A Dingman mouth retractor is used because the retraction of the cheeks that it affords allows valuable additional exposure. The area along the line of incision is infiltrated with lidocaine (Xylocaine 1%) with epinephrine 1:100,000 to improve hemostasis. An incision is made in the midline through the mucoperiosteum down to palatal bone. The mucoperiosteum is then undermined with a sharp elevator as far posterior as the junction of the hard and soft palates. The greater palatine vessels on one side may be clamped and cut if necessary. Cutting both vessels deprives the soft palate of its blood supply, which may cause necrosis or prevent postoperative wound healing and is a severe technical error. The muscular aponeurosis and nasopharyngeal mucosa of the soft palate are incised transversely, and the palate is retracted posteriorly and superiorly toward the nasopharynx.

To remove the bony atresia and establish a patent nasal airway, the bone of the posterior third of the hard palate is removed with a rongeur until the nasal mucosa is identified (Fig. 4-4). The bony

atresia plate and the posterior third of the vomer are removed with a rongeur or a curette (Fig. 4-5). The nasal mucosal flap should be preserved whenever possible to prevent postoperative stenosis. Failure to remove a portion of the vomer and hard palate predisposes to eventual stenosis of the choana.

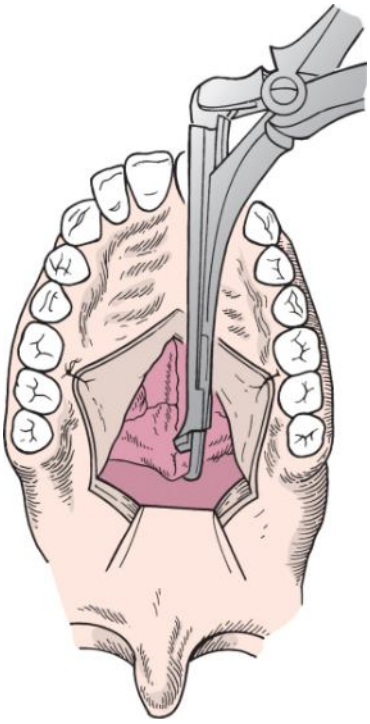

Figure 4-4 A rongeur is used to remove the posterior aspect of the hard palate.

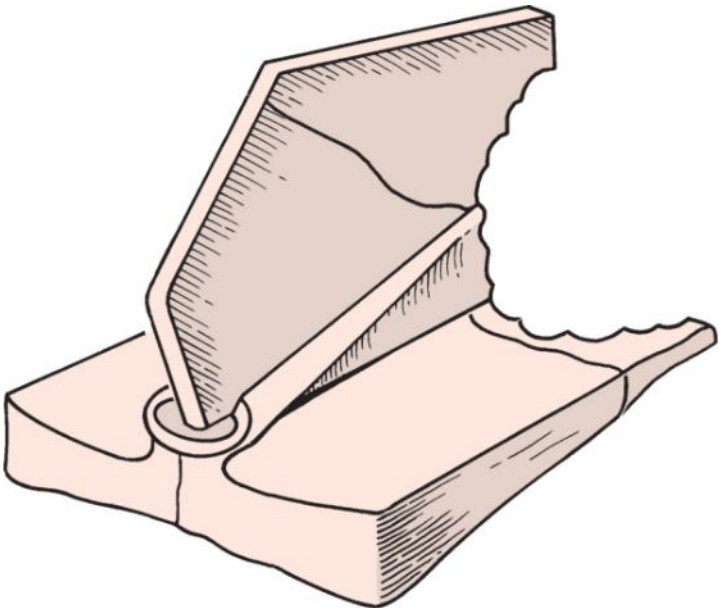

Figure 4-5 Removal of a portion of the hard palate and vomer.

This procedure must be done under direct vision, although the use of a laryngeal mirror to adequately visualize the atretic plate is helpful. After the atresia has been corrected, soft Silastic stents should be inserted (see Fig. 4-2D). We have used a nasopharyngeal airway with the flange placed in the nasopharynx to good advantage (Fig. 4-6). The airway is sutured to the septum. This helps reconstitute nasal breathing and provides a stent that is left in place for 2 months to ensure healing and prevent stenosis.

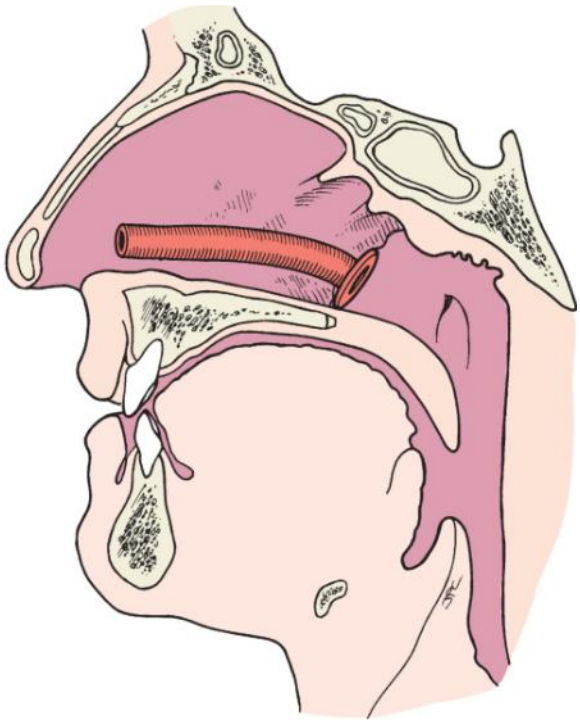

Figure 4-6 Use of a nasopharyngeal airway as a stent.

At the end of the procedure, the palatal flap is returned to the anatomic position and the wound is closed with 3-0 silk suture on a cutting needle. The silk seems to hold the knot in the oral cavity better than absorbable or nylon suture does. The patient is allowed to awaken from the anesthesia and is extubated.

Transnasal Endoscopic Technique

Under general anesthesia, a cottonoid or cotton pledget soaked in oxymetazoline (Afrin) is placed in the nasal cavity to decongest the mucosa. A 2.5- to 4.4-mm scope is placed in the nasal cavity after the pledgets are removed, which produces excellent visualization. Powered instrumentation is then used to remove the atretic plates and vomer. A backbiting rongeur can be introduced through the nasal cavity to remove a portion of the vomer. An endoscope placed in the contralateral nasal cavity can provide excellent visualization during this important step.

Sublabial Transseptal Repair

The technique is said to be helpful in neonates with unfavorable nasal anatomy or craniofacial anomalies. After a sublabial incision into the mucosa of the floor of the nose, the septum is elevated and the atretic plate is exposed and removed.

POSTOPERATIVE MANAGEMENT

Antibiotics are administered to prevent infection. Suctioning plus cleaning of the stents is important for personal hygiene and to ensure patency of the tube for nasal breathing. Local care includes cleansing around the stents with hydrogen peroxide to control any crusting that may develop in the area of the nasal ala. After the crusts are removed, antibiotic ointment is applied to the vestibular skin. The use of nasal steroid spray for 6 weeks after stent removal to reduce granulation tissue formation has been suggested.[9] The stents are removed after 2 months under general anesthesia in pediatric patients, at which time the granulation tissue may be removed with nasal endoscopes.

PEARLS

- The systematic use of CT scans to evaluate the patient provides valuable information for planning.
- The use of nasal endoscopes and powered instruments provides a precise technique for removal of an atresia plate with a lower risk of complications.
- Stenosis may be prevented by adequately removing bone, preserving mucosal flaps, and stenting.
- Complications may be avoided by meticulous attention to surgical technique, the use of a properly sized stent, and placement of an airway.
- The use of nasal steroid spray after removal of stents may be helpful.

PITFALLS

- In the transpalatal approach there may be intraoperative problems such as bleeding from the greater palatine arteries, injury to the basiocciput during removal of the atresia plate, and injury to the contents of the incisive foramen.
- Early postoperative complications may include airway obstruction secondary to edema of the soft palate or tongue.
- Late complications include nasal infection, necrosis of the palatal mucosa, velopharyngeal incompetence from either muscular dysfunction or overly long stents, oral antral fistula, and stenosis of the choana.
- Pressure necrosis of the columella or the alar rim may be caused by improperly sized stents.
- Leakage of cerebrospinal fluid as a result of injury to the cribriform plate with or without meningitis has been reported.[19]