Operative Management of Choanal Atresia

A 15-Year Experience

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Objective: To analyze factors affecting 15-year surgical outcomes of choanal atresia repair.

Design: Case series.

Setting: Tertiary care pediatric hospital.

Patients: Between April 17, 1996, and March 23, 2010, a total of 42 patients aged 3 days to 15 years underwent endoscopic or transpalatal choanal atresia repair by our pediatric otolaryngology faculty.

Main Outcome Measures: Reoperation and restenosis rates, with consideration of effects of mitomycin C therapy, stenting, and postoperative dilation.

Results: Three of 42 patients were excluded because of inadequate follow-up data; the follow-up time for the remaining 39 patients averaged 6.3 years (range, 1-14.9 years). Excluding 6 patients whose initial repair was performed by other physicians, 31 of 33 patients in whom we performed initial repair had a total of 43 endoscopic surgical procedures (19 patients had unilateral procedures, and 12 patients had bilateral procedures), and the other 2 underwent bilateral transpalatal repair. Of the total 43 sides we operated on endoscopically, 9 sides (21%) required revision surgery, including excision of scar tissue or additional drilling of persistent bony stenosis. No significant difference was observed in the rate of restenosis among cases treated endoscopically with mitomycin C (22 of 43 operative sides, \(P = .13\)), with stenting (36 of 43 operative sides, \(P = .99\)), or with subsequent dilation (\(P = .45\)). When we used stents, they were usually (in 28 of 36 patients) left in place for 15 days or longer.

Conclusion: Our revision rate after initial endoscopic repair of choanal atresia was low and was unaffected by adjuvant mitomycin C therapy or stenting.


Choanal atresia (CA) is an uncommon malformation of the upper airway but is one that may lead to significant neonatal airway obstruction. Each year, 1 in 7000 to 1 in 8000 children are born with CA. In most children, the atresia is unilateral (occurring at a 2:1 ratio), and half of all children with CA have associated anomalies, with the most common being CHARGE syndrome.

The developmental abnormality in CA seems to be persistence of the nasobuccal membrane (of Hochstetter), which typically is resorbed about the sixth week of gestation. Other implicated mechanisms include persistence of the buccopharyngeal membrane of the foregut, incomplete resorption of the nasopharyngeal mesoderm, or abnormal migration of neural crest cells. Anatomically, CA manifests as medialization of the lateral pterygoid plate (creating the lateral aspect of the obstruction), an enlarged vomer (the medial aspect of the obstruction), and obstruction of the nasalpharyngeal airway on the affected side, with complete obstruction in bilateral cases. The obstruction is most commonly purely bony, but it may be membranous or a combination of the two. Initial manifestations range from life-threatening airway obstruction to bothersome nasal airway obstruction or rhinorrhea, which is more typical of the unilateral condition.

Bilateral CA with complete obstruction is typically diagnosed at birth; it usually manifests as cyclical cyanosis that improves with crying. Inability to pass a suction catheter through the nasal passages should prompt a more thorough evaluation of nasopharyngeal anatomy, generally by fiber-optic endoscopy, possibly followed by computed tomography. Until the CA can be repaired, a McGovern nipple may be used to maintain the infant’s airway and to allow feeding. When CA is unilateral, surgical repair can often be delayed until the nasal passages are

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larger, potentially improving the outlook for successful repair.

After the initial description of CA by Roederer in 1755, numerous authors have reported on various repair techniques and adjuvant therapies. However, few studies in the English-language literature present outcomes in series of more than 30 patients. In addition, controversies exist about the benefits of stenting, the use of adjuvant mitomycin C therapy, and the timing of repair in unilateral CA. To address these questions, we examined outcomes of CA repair (the need for revision surgery) at our pediatric teaching hospital during the past 15 years, including comparison of results in patients who received stents and in patients who were operated on with adjuvant mitomycin C.

METHODS

PATIENTS AND INTERVENTIONS

Approval for this study was obtained from the University of Alabama at Birmingham Institutional Review Board for Human Use. For this study, we conducted a retrospective medical record review to identify all the patients who were operated on for CA by the faculty of Pediatric ENT Associates and Children’s Hospital of Alabama between April 17, 1996, and March 23, 2010. Of 42 patients identified, all had undergone a full history intake and physical examination to determine whether associated congenital anomalies were present, and formal genetic testing was performed in many patients. All 42 were operated on by fellowship-trained pediatric otolaryngologists (W.P.S., J.S.H., A.L.W., and B.J.W.) using transnasal endoscopic or transpalatal techniques.

Medical records were analyzed for age at diagnosis of CA, age at surgery, type of atresia (bony or mixed, including membranous), the presence of syndrome-associated anomalies, technique for initial repair (endoscopic or transpalatal), and the use of intraoperative mitomycin C therapy, as well as follow-up time and whether CA was unilateral or bilateral, whether a stent was placed at the conclusion of the initial operation and the time until stent removal, whether subsequent dilations were performed, and whether revision surgery was required. Revision surgery was considered any surgical intervention except serial dilation or stent or granulation tissue removal.

Based on the medical record review, 3 of 42 patients were excluded because of inadequate data for analysis or insufficient postoperative follow-up time. Six of the remaining 39 patients had initially been operated on for CA by other physicians and were excluded from calculations of restenosis and reoperation rates.

Statistical analyses were performed using available software (GraphPad; GraphPad Software, Inc.). Fisher exact test was used, with $P \leq .05$ accepted as statistically significant.

ENDOSCOPIC OPERATIVE TECHNIQUE

After induction of general anesthesia and endotracheal tube placement, the patient is suspended with a Dingman mouth gag. A telescope (120° rod lens; Karl Storz) with palate retractor is used to obtain direct endoscopic visualization of the posterior choana. The atretic choana is then punctured with a van Buren urethral sound (typically 10F catheter) under direct visualization. Even in cases of purely bony atresia, choanal puncture can typically be performed successfully using the sound. In cases in which the bony atresia cannot be punctured in this way, a microdebrider drill may be used. Once an aperture has been created in the atresia plate, progressively larger urethral sounds are used to dilate the opening until it is of sufficient size to allow passage of the endoscope into the nasopharynx.

A key step is performed next, namely, removal of the posterior hypertrophied vomer. This is usually achievable using endoscopic backbiting forceps, although occasionally a microdebrider is required to complete this portion of the operation. The microdebrider drill is then used to remove the posterolateral sphenopalatine wall. Once an adequate opening has been made in the atretic choana, the surgeon may decide to place a nasal stent, particularly if stenosis is severe. For stenting, a 3.5-mm uncuffed endotracheal tube is typically used. The tube is modified by bending it in half lengthwise and cutting halfway through its diameter, allowing the tube to maintain this bend. The tube is then placed circumferentially around the posterior choanal repair site, with the cut portion facing posteriorly. The ends of the tube will protrude slightly from the right and left nares. A 0 polypropylene suture is passed through the stent on both sides, going around the posterior choana, and is then tied anteriorly. The stent is designed so that the anterior aspect rests just behind the nasal ala intranasally to prevent columellar necrosis. The parents or caregivers are instructed to move the stent back and forth at least daily to prevent synechiae formation and to irrigate and suction the tubing at least 3 times per day.

When a stent has been placed, we usually remove it in the operating room. The timing is largely based on the impression of the attending surgeon at the initial repair, although most are removed within 14 to 21 days. When mitomycin C therapy is used, it is most commonly applied at the time of stent removal.

TRANSPALATAL REPAIR TECHNIQUE

Indications for transpalatal repair included exceptionally thick bony atresia or nasal alar stenosis that prevented the use of transnasal endoscopic instrumentation. For transpalatal repair, the palate is injected with topical anesthetic. An incision is made starting behind the maxillary tuberosity and is carried along the palatoalveolar ridge to the nasopalatine foramina. Flaps are raised in the submucoperiosteal plane to the border of the hard palate, taking care to avoid the neurovascular bundle. A releasing incision is then made at the junction of the hard and soft palates. The soft palate is retracted posteriorly. Then, a cutting burr or rongeurs are used to remove a portion of the posterior hard palate. Bone is also removed laterally if needed to obtain an aperture through which a 14F catheter (or larger) urethral sound dilator can pass easily.

After achieving an adequate opening in the choana, the palatal mucosa is closed with absorbable suture. Finally, if the surgeon chooses, a U-shaped stent fashioned from an endotracheal tube (as already described) is secured in each operated choana.

RESULTS

During the past 15 years, our specialty hospital group of faculty pediatric otolaryngologists evaluated and surgically repaired congenital CA in 42 patients aged 3 days to 15 years at the time of surgery. Of 39 patients, 23 (59%) had unilateral atresia, with the right side affected in 14 (61%) (Table). In 12 of 39 patients (31%), CA was part of a syndrome, most commonly CHARGE syndrome. Six of 39 patients (15%) with complete (bony) bilateral CA were aged 3 days to 11 months (mean, 102 days) at the time of initial surgery; 1 patient with bilateral partial membranous atresia was 15 years old at the time of initial surgery.

Of 33 patients receiving primary surgery at our institution, the initial procedure was performed endoscopi-
cally in 31 patients and transpalatally in 2 patients. Choanal atresia was bony (in 6 patients) or membranous (in 3 patients) in a few cases and was mixed in 30 of our 39 patients followed up for 1 year or longer. Many authors categorize CA only as bony or mixed. Using these categories, among 39 patients followed up for 1 year or longer, 6 (15%) had bony atresia, and 33 (85%) had mixed atresia. The mean follow-up time after surgery was 6.3 years (range, 1-14.9 years) (Table).

### OVERALL REVISION RATES

Overall, 21% (9 of 43) of repaired choanae that underwent endoscopic repair required revision surgery. The mean time from initial repair to revision was 1.3 years (range, 35 days to >9 years). Thirty-three percent (1 of 3) of those with purely bony atresia required revision surgery vs 20% (8 of 40) of those with mixed or membranous atresia (*P* = .51). Four of 10 choanae (40%) associated with syndromic disease required revision surgery compared with 5 of 33 choanae (15%) related to isolated CA (*P* = .17). Eight of 19 patients (42%) with unilateral disease underwent revision surgery compared with 2 of 24 patients (8%) with bilateral atresia. At the last follow-up visit, complete restenosis was absent in all of these patients. Two patients had partial narrowing discovered on in-office nasal endoscopy, but both were asymptomatic and did not undergo further treatment.

### MITOMYCIN C THERAPY

Mitomycin C therapy was used as an adjunct to surgery in approximately half of atretic choanae treated endoscopically (22 of 43 operative sides [51%] in 31 patients). No significant difference was observed in the rate of revision surgery among those treated (7 of 22 [32%]) vs not treated (2 of 21 [10%]) with mitomycin C (*P* = .13) (Figure).

### STENTING

Intranasal stents were used in 36 of 43 choanae (84%) operated on endoscopically. When we used stents, they were usually (in 28 of 36 patients) left in place for 15 days or longer. The use of stents did not significantly alter revision rates (*P* > .99) (Figure). Similarly, revision rates among 36 cases of congenital CA (in 31 children) did not differ significantly whether the time between placement and removal of the stent was 1 to 14 days (3 of 8

#### Table. Characteristics of Choanal Atresia in 39 Patients Followed Up for 1 Year or Longer After Initial Surgical Repair

<table>
<thead>
<tr>
<th>Characteristic</th>
<th>All Patients, No./Total No. (%)</th>
<th>Patients Undergoing Initial Surgery by Us (n = 33)</th>
</tr>
</thead>
<tbody>
<tr>
<td>Unilateral choanal atresia</td>
<td>23/39 (59)</td>
<td>19 Endoscopic</td>
</tr>
<tr>
<td>Right side only</td>
<td>14/23 (61)</td>
<td>...</td>
</tr>
<tr>
<td>Left side only</td>
<td>9/23 (39)</td>
<td>...</td>
</tr>
<tr>
<td>Age at diagnosis of choanal atresia, mean (range)</td>
<td>...</td>
<td>33.0 mo (Birth to 10 y)</td>
</tr>
<tr>
<td>Age at surgery, mean (range)</td>
<td>...</td>
<td>35.0 mo (3 wk to 10 y)</td>
</tr>
<tr>
<td>Bilateral choanal atresia</td>
<td>16/39 (41)</td>
<td>2 Transpalatal, 12 Endoscopic</td>
</tr>
<tr>
<td>Age at diagnosis of choanal atresia, mean (range)</td>
<td>...</td>
<td>0.2 mo (Birth to 2 mo)</td>
</tr>
<tr>
<td>Age at surgery, mean (range)</td>
<td>...</td>
<td>2.8 mo (3 d to 1 y)</td>
</tr>
<tr>
<td>Type of atresia</td>
<td></td>
<td></td>
</tr>
<tr>
<td>Bony</td>
<td>6/39 (15)</td>
<td>...</td>
</tr>
<tr>
<td>Mixed, including membranous</td>
<td>33/39 (85)</td>
<td>...</td>
</tr>
<tr>
<td>Presence of syndrome-associated anomalies</td>
<td>12/39 (31)</td>
<td>...</td>
</tr>
<tr>
<td>Technique for initial repair</td>
<td></td>
<td></td>
</tr>
<tr>
<td>Endoscopic</td>
<td>36/39 (92)</td>
<td>31/33</td>
</tr>
<tr>
<td>Transpalatal</td>
<td>3/39 (8)</td>
<td>2/33</td>
</tr>
<tr>
<td>Intraoperative mitomycin C used at the end of initial endoscopic repair by us</td>
<td>22/43 (51) Sides of 31 patients</td>
<td>...</td>
</tr>
<tr>
<td>Stent placed at the end of initial endoscopic repair by us</td>
<td>36/43 (84) Sides of 31 patients</td>
<td>...</td>
</tr>
</tbody>
</table>

*The mean follow-up time was 6.3 years (range, 1-14.9 years).*

*Excludes a 15-year-old patient with bilateral partial atresia.*

*Thirty-three patients had primary repair by us; 31 (19 unilateral and 12 bilateral) of those were repaired endoscopically, and the other 2 had transpalatal repairs.*

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**Figure.** Percentage of 43 cases of congenital choanal atresia (in 31 patients) that required revision surgery after initial endoscopic repair. Revision rates were similar whether mitomycin C was used (22 of 43 operative sides, *P* = .13) and whether intranasal stents were used (36 of 43 operative sides, *P* > .99). Seven of 22 patients who were treated with mitomycin C required revision surgery. Eight of 26 patients who underwent stenting required revision surgery.
ENCE was not significant herein (P = .19).

### POSTOPERATIVE DILATION

Postoperative dilation was performed in 25 of 43 operated sides (58%). Four of 25 sides (16%) treated by dilation eventually required revision surgery compared with 3 of 18 sides (28%) of patients not treated by dilation (P = .45).

### AGE AT SURGERY

Patients with unilateral CA were seen for care at ages ranging from birth to 10 years, with a mean age at diagnosis of 33 months (Table). The age range for initial surgical repair was 3 weeks to 10 years. Among 19 children undergoing revision surgery after endoscopic repair of unilateral CA, the need was not significantly related to age at the time of initial surgery for unilateral CA for those younger than 6 months (2 of 5 [40%]), age 7 to 24 months (2 of 7 [29%]), or older than 24 months (3 of 7 [43%]) (P = .33).

### COMMENT

Analysis of our long-term (mean, 6.3 years) results of initial endoscopic repair of CA in 31 patients (43 operated sides) found that revision surgery was eventually needed in 21% (9 of 43) of cases. No significant differences were observed in revision rates whether mitomycin C therapy was used or a stent was placed or whether the patient was younger than 6 months, between 6 and 24 months, or older than 24 months at the time of initial repair.

Some large differences between groups were not statistically significant. However, we found that the trends among our data generally matched those in the literature.

### TYPE OF ATRESIA

One trend we noted was that patients with bony atresia (1 of 3 [33%]) were more likely to need revision surgery than patients with mixed or membranous disease (8 of 40 [20%]). Other authors have reported that the management of bony atresia is more challenging than the management of atresia with a membranous component, possibly due to neo-osteogenesis as described by Ayari et al.9

We also found that patients with syndrome-associated atresia were more likely to need revision surgery than patients with isolated disease (4 of 10 choanae [40%] vs 5 of 33 choanae [15%], P = .17). Studies3,10-12 have also found higher rates of restenosis in patients with syndromic CA, although 2 recent studies3,7 found no significant difference in the 2 groups.

### MITOMYCIN C THERAPY

The use of mitomycin C therapy is thought by many surgeons to prevent tissue regrowth and to prolong patency of CA repair. Although some authors still promote its use in preventing restenosis, others have found that it does not improve outcomes.3,14 Although the difference was not significant herein (P = .13), we noted a higher proportion of revisions in cases treated with this agent (7 of 22 treated sides vs 2 of 21 untreated sides) (Figure). Mitomycin C therapy was not used routinely and often was used in the more difficult cases, which could account for the increased incidence of restenosis in patients treated with this agent.

### STENTING

Stents were placed at the end of the procedure in 36 of 43 choanae (84%) we operated on endoscopically. The rationale for placing a stent is that it will prevent narrowing of the newly formed lumen as it heals. However, opponents of stents argue that they damage the nasal mucosa, resulting in granulation tissue, scar formation, bacterial overgrowth, and impaired drainage of mucus, all of which work against the end goal of patency.4,10,13 Several publications4,6,10,16,17 and a Cochrane review18 report no difference in patency rates for patients who receive or do not receive stenting after surgery; our findings are in agreement with these authors (Figure). Publications of retrospectively collected data showing no difference in revision rates with stenting continue to emerge; however, more prospective trials are needed to definitively decide the issue.

Although we found no difference in revision rates when stents were or were not used, we suspect this may be due to selection bias because the decision to stent was at times based on greater severity of disease. Because of this, we do not conclude from our results that stents should be universally used or abandoned but believe that the decision to stent is best left to the judgment of the attending surgeon at the time of repair.

Regarding duration of stenting, some authors report that a short period of stenting (as short as 5-7 days) minimizes the negative effects of the stent on the healing process while still improving patency rates.7,10 However, other investigators have proposed that prolonged stenting (as long as 12 weeks) is necessary.3 We found a trend in our study for a lower revision rate when stents were left in place for 15 to 28 days (7% [1 of 14] revision rate) vs 1 to 14 days (38% [3 of 8] revision rate), although the difference was not significant (P = .19).

### AGE AT SURGERY

The optimal age to perform repair of unilateral CA has been debated.6,20,21 In our series, the revision rate was somewhat lower among patients operated on between age 6 and 24 months (2 of 7) compared with those younger than 6 months (2 of 5) or older than 24 months (3 of 7), although the difference was not significant. Other authors routinely wait to operate on patients with unilateral CA who are not having serious airway or feeding problems until the child is about 12 months of age.6,22 The rationale for delaying repair as long as possible is that the surgically created aperture does not enlarge as the surrounding tissues grow so that the aperture becomes narrower as the child matures.

Weaknesses of our study were largely due to its retrospective nature. With surgical procedures being performed by 4 different attending surgeons (W.P.S., J.S.H.,...
A.L.W., and B.J.W.), we were unable to completely control for the differences in techniques. All 4 surgeons use the same instruments and generally follow the same steps in repair. Because we did not detect a skewed number of revisions based on attending surgeon, it is unlikely that any small differences in repair created significant differences in outcomes. All 4 surgeons planned second-look procedures in the operating room for stent removal when stents were used. As mentioned previously, the timing of stent removal was largely based on the impression of the attending surgeon at the initial repair, although most stents are removed within 14 to 21 days. The retrospective nature of our study also meant that other markers for success were not routinely recorded (such as choanal diameter or degree of stenosis).

Our overall revision rate after initial endoscopic surgery in this study was 21% (9 of 43) compared with rates reported in the literature of between 12% and 54.7%. Our rate is somewhat higher compared with that of some other authors. We believe that this difference is accounted for by 3 factors. First, we defined revision surgery more broadly than others as including any trip to the operating room for intervention other than removal of a stent (with associated mild granulation tissue) or dilation alone. Second, we reported data from long follow-up periods (up to 14.9 years), with several patients requiring revision surgery years after the initial repair. Third, we may have been more likely than others to recommend revision surgery in pursuit of alleviating symptoms and achieving normal apertures, as demonstrated by the fact that almost all of our patients were disease free at the last documented clinic visit. Certainly, varying definitions of successful surgery exist within the literature, with some authors using the need for any further operative intervention and others using eventual patency rather than need for revision surgery. It is impossible to know how many of the patients on which we (and other authors) performed further debridement at second-look procedures for stent removal would have eventually required any additional intervention for disease symptoms.

In conclusion, transnasal endoscopic repair is effective for bilateral and unilateral CA. Delaying repair of unilateral CA until a child is at least 6 months old may lower the risk that revision surgery will be needed as or after the child matures. Our revision rates were similar with or without the use of mitomycin C therapy or stenting, although these adjunct treatments were more often used in patients having atresia of greater severity.

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Author Contributions: Drs Newman, Harmon, Woolley, and Wiatrak had full access to all the data in the study and take responsibility for the integrity of the data and the accuracy of the data analysis. Study concept and design: Newman, Harmon, and Wiatrak. Acquisition of data: Newman, Harmon, Shirley, Hill, Woolley, and Wiatrak. Analysis and interpretation of data: Newman, Harmon, and Wiatrak. Drafting of the manuscript: Newman and Wiatrak. Critical revision of the manuscript for important intellectual content: Newman, Harmon, Shirley, Hill, Woolley, and Wiatrak. Statistical analysis: Newman and Harmon. Administrative, technical, and material support: Wiatrak. Study supervision: Harmon, Shirley, Hill, Woolley, and Wiatrak.

Conflict of Interest Disclosures: None reported.

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REFERENCES