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Original Investigation

Surgical and Audiometric Outcomes for Repair of Congenital Aural Atresia and Hypoplasia

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IMPORTANCE Surgical repair of congenital aural atresia and hypoplasia (CAAH) is technically challenging. Long-term surgical and audiologic outcomes of atresiaplasty are incompletely understood.

OBJECTIVES To review the surgical outcomes for CAAH and analyze the hearing results.

DESIGN, SETTING, AND PARTICIPANTS A retrospective medical record review of CAAH outcomes was performed during an 11-year period from January 1, 2004, through December 31, 2014. The data analysis was undertaken from December 1, 2014, through January 31, 2015. The mean clinic follow-up time was 3.9 years, and the mean audiologic follow-time was 2.8 years. The study included 98 patients aged 5 to 66 years (mean age, 16.6 years) with CAAH who underwent a total of 104 operations.

INTERVENTIONS Surgical repair of CAAH.

MAIN OUTCOMES AND MEASURES Preoperative and postoperative pure-tone averages (PTAs), speech reception thresholds (SRTs), air-bone gaps (ABGs), and interaural PTA and SRT differences were compared. Factors that affect hearing outcomes were analyzed. The complication rates were reviewed and compared with results from similar studies.

RESULTS In the 98 patients with CAAH, the mean improvement in ABGs and SRTs was 26.7 and 25.9 dB, respectively, resulting in a postoperative ABG of 30 dB or less in 4 of 5 cases. The mean postoperative PTAs and SRTs were 36.9 and 34.3 dB, respectively. Patients with a functional native ossicular chain (36 of 104 [34.6%]) had significantly superior audiometric outcomes when compared with patients in whom a reconstruction prosthesis was required during primary or revision operations. Audiometric results from hypoplasia surgery were not significantly different from those of atresia surgery; results in patients with craniofacial syndromes were similarly not significantly different from those in patients with sporadic CAAH. We report a low incidence of meatal stenosis.

CONCLUSIONS AND RELEVANCE The mean hearing outcomes for this group compared favorably with other series. The need for ossicular chain reconstruction was associated with poorer audiometric outcomes. The safety profile and the demonstrated hearing improvement of CAAH surgery suggest that it remains a favorable option for patients.

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Congenital aural atresia and hypoplasia (CAAH) refer to a spectrum of otologic deformities that result from abnormal development of the first and second branchial arches.[1] Congenital aural atresia and hypoplasia result in varied degrees of malformation that affects the ossicles, middle ear space, tympanic membrane (TM), bony and cartilaginous external canal, and pinna. Kiesselbach[2] is credited with undertaking the first surgical correction of aural atresia in 1883. This attempt resulted in facial paralysis, and the notoriously difficult procedure did not gain popularity for decades secondary to frequent complications and poor surgical outcomes. In the late 1970s, Jahrsdoerfer et al[3,4] emerged as key proponents of atresia repair and provided remarkable improvements in management principles and surgical technique.

Despite continued advancements, atresiaplasty remains an exceedingly challenging procedure. In light of the technical simplicity and effectiveness of bone-anchored hearing devices, there are some who question the appropriateness of atresia repair because of its higher incidence of complications and less established long-term results. Several informative retrospective case series[5-9] evaluating these concerns have been reported, including a prior review of 45 patients (54 ears).[10] Nonetheless, atresiaplasty results remain a topic of interest. We present the outcomes of 98 patients (104 ears) who underwent surgery for CAAH during an 11-year period at a tertiary care institution.

Methods

Surgical Technique

With the patient under general anesthesia, a 0.008-in split-thickness skin graft is harvested with a dermatome from the inner aspect of the upper ipsilateral arm. The graft is placed, dermis side up, on occlusive petrolatum gauze, and 1 × 5-cm strips are created (eFigure 1 in the Supplement), set aside, and kept moist. The donor site is cauterized with Monsel solution, cleansed with saline, and dressed with antibiotic ointment, nonadherent pads, and dry gauze. After local injection, a postauricular incision is made and taken down to the areolar temporoparietal fascia plane. The ear is reflected anteriorly. Superior and posterior elevation of the skin and soft tissue is performed sharply. A 3 × 3-cm temporoparietal fascia graft is then harvested, pressed, and set aside to dry, and a T-shaped musculoperiosteal incision is made over the mastoid cortex with cautery. The tissues are elevated with a periosteal elevator, and the temporomandibular joint is identified anteriorly.

Drilling begins with a 4-mm cutting burr along the linea temporalis posterosuperior to the glenoid fossa, preserving the posterior wall of the fossa. The tegmen mastoideum and glenoid fossa serve as the key landmarks superiorly and anterioinferiorly, respectively, and drilling continues medially with progressively smaller cutting and diamond burrs through the atresia plate while avoiding contact with the ossicles or entrance into the mastoid air cells. A stapes curette is used to carefully enter the epitympanum or middle ear space, and all surrounding bone is removed from around the ossicular chain so that the ossicles are centered in the neotympanic ring (eFigure 2 in the Supplement). The neotympanic ring is made as large as possible given anatomical constraints. In this manner, the large TM will take advantage of its ratio with the oval window for maximal amplification. The ossicles are then inspected. An ossicular chain reconstruction (OCR) is performed if there is any evidence of ossicular chain fixation, discontinuity, or a hypermobile-fibrous incudostapedial connection. Once the neocanal is of suitable size, 2 drill holes are made at the posterior edge of the lateral ear canal using a 1-mm diamond burr.

Attention is then turned to the creation of the soft-tissue meatus. After injection of additional local anesthetic, a posteriorly based rectangular skin flap is incised and elevated. The underlying soft tissue and cartilage are then excised to create a lumen through which the neocanal is visualized (eFigure 3 in the Supplement). Next, the anteriorly based musculoperiosteal flap is reflected anteriorly, trimmed in length, and then sutured to the medial aspect of the tragal skin or cartilage (eFigure 4 in the Supplement). This procedure creates a smooth anterior canal contour from the tragus down to the TM. The areolar temporalis fascia graft is trimmed to size and shape. The leading edge is tucked medial to the bony ledge of the neotympanic ring anteriorly and then draped out over the neocanal superiorly and posteriorly to cover any air cells exposed during canal creation. If an adequate malleus is present, a small slit incision is made on the graft to accommodate the handle of the malleus.

In contrast to the large skin graft described by Jahrsdoerfer et al,[3,4] the strips of split-thickness skin grafts are then placed, beginning anteriorly, circumferentially over the neocanal and fascia graft in a wallpaper-like fashion. Once in position, the occlusive petrolatum gauze is carefully removed, and 2 beveled pope ear wicks (eFigure 5 in the Supplement) are placed in the ear canal and expanded with saline solution. The pointed end of the bevel maintains a sharp anterior sulcus. The lateral ends of the skin grafts are folded over the end of the wicks, and postauricular closure is initiated. The postauricular musculoperiosteal tissues are approximated with interrupted sutures of 3-0 polyglactin. The postauricular skin incision is then closed with inverted, interrupted subcuticular 3-0 polyglactin sutures. Superior repositioning of the auricle is often necessary to align the bony and soft-tissue openings of the canal. A tacking suture from the deep auricular cartilage to the temporalis fascia helps hold this new position. Two horizontal mattress sutures of 5-0 polyglactin are passed through the distal portion of the posteriorly based meatal skin flap and the previously drilled bony suture holes at the posterior edge of the ear canal (eFigure 6 in the Supplement). These flap sutures are placed before the postauricular closure is completed. Tying the sutures is performed via the canal meatus; this creates a meatus that requires no stent-type packing. The lateral ends of the skin grafts are unfurled and extended out through the ear canal meatus, and excess skin is trimmed. The cut edges of the skin graft are sutured to the edges of the meatal skin with interrupted sutures of 5-0 fast-absorbing chromic gut. At the conclusion of the procedure, 3 to 4 untrimmed pope ear wicks are inserted into the lateral canal and hydrated with sterile saline solution.

In the postoperative period, patients are instructed to avoid getting any water into the ear canal and to instill antibiotic drops twice daily. The first postoperative visit is at 3 weeks, and all wicks are removed. The healing of the TM and skin grafts are assessed, and new pope ear wicks are placed. The wicks are removed at the next visit 4 weeks later. After another 4 weeks, the ear canal is thoroughly cleaned, and the first postoperative audiogram is obtained. Long-term care of the ear is critical for lasting results. Because reconstructed ear canals do not clear squamous debris normally, shed skin will accumulate and adhere to the canal lining. The neocanal should accordingly be carefully cleaned twice per year. For 3 consecutive nights before a scheduled cleaning, patients are asked to instill baby or mineral oil into the ear to loosen the squamous debris and facilitate atraumatic cleaning

Data Collection

A retrospective medical record review was undertaken for all patients who underwent CAAH repair from January 1, 2004, through December 31, 2014, with one of us (R.A.C.) at a tertiary referral center. The data analysis was undertaken from December 1, 2014, through January 31, 2015. A preoperative Jahrsdoerfer scale score was not determined, but when the preoperative computed tomographic scans are reviewed, it is insured that the middle ear and mastoid are well aerated, the oval window and round window are patent and well formed, the facial nerve is in a favorable position, and the tegmen is not excessively low riding. Hearing also needs to be appropriate. In highly motivated adults, surgery may be contemplated even if their preoperative speech discrimination score is suboptimal (60%-70%).

Basic demographic data and preoperative and postoperative audiologic data were collected. These data were composed of the pure-tone average (PTA), speech reception threshold (SRT), and air-bone gap (ABG) and were recorded in decibels of hearing loss. The PTA and ABG were calculated as a mean of the pure-tone responses at 0.5, 1.0, 2.0, and 3.0 kHz. The interaural difference of the PTA and SRT was calculated in patients with unilateral CAAH. The most recent audiogram was used for postoperative data in each case. Exclusions were made if preoperative and postoperative data were not available or if surgery was performed without an attempt at hearing rehabilitation. Surgical complications and rates of revision surgery were assessed. Complications included chronic myringitis, TM perforation, exposed bone, prosthesis displacement, external auditory canal stenosis, TM lateralization, sensorineural hearing loss, and temporomandibular joint prolapse. Audiologic outcomes in patients undergoing OCR were compared with those in patients with an intact ossicular chain. Results in patients with hypoplasia were compared with those in patients with canal atresia. Results in patients with craniofacial syndromes were compared with those in patients without associated craniofacial deformities.

Table 1. Patient Characteristics

Characteristic	Finding (98 Patients and 104 Cases)[a]
Bilateral atresia or hypoplasia	20/98 (20.4)
Male sex	59/98 (60)
Mean age, y	16.6
Syndromic patients	13/98 (13.3)
Revision cases[b]	7/104 (6.7)
Prior microtia repair	59/104 (56.7)
Mean clinic follow-up,[c] y	3.9
Mean audiologic follow-up,[d] y	2.8

[a] Data are presented as number (percentage) unless otherwise indicated.

[b] Prior surgery performed by another physician.

[c] Mean time from surgery to most recent clinic visit.

[d] Mean time from surgery to most recent audiogram.

Ethical Considerations

This retrospective review was approved by the Southern California Permanente Medical Group Institutional Review Board. Informed consent was not required.

Statistical Analysis

Data entry and statistical analysis were completed with SPSS statistical software, version 8 (SPSS Inc). Continuous variables of equal variance were compared with a 2-tailed *t* test, and those of unequal variance were compared with a Mann-Whitney log-rank test. Categorical data were compared with a 2-tailed Fisher exact test. $P \leq .05$ was considered statistically significant.

Results

A total of 104 patients who underwent a total of 110 CAAH repair operations were identified. Six exclusions were made: 4 owing to insufficient audiogram data and 2 for cases in which hearing rehabilitation was not attempted (one owing to an absent oval window and another in which reconstruction was prohibited by a facial nerve overlying the oval window). As such, 98 patients, totaling 104 CAAH repairs, were ultimately included in the analysis (**Table 1**).

Audiologic data for all 104 cases are given in **Table 2**. Statistically significant improvements in the PTA, SRT, and ABG were all achieved. These metrics underwent mean improvements of 24.5, 25.9, and 26.7 dB, respectively. Fifty percent of the cases resulted in a postoperative PTA of 30 dB or better. An ABG of less than or equal to 30 dB was achieved 81% of the time. In cases of unilateral CAAH, the mean postoperative interaural differences for the PTA and SRT were 27.8 dB and 25.4 dB, respectively.

Complications of the 104 CAAH repairs are tabulated in **Table 3**. At least 1 complication occurred in 43 of the 104 cases, yielding an overall complication rate of 41.3%. Chronic myringitis, prosthesis displacement, and sensorineural hearing loss were the 3 most common complications and occurred in 10 (9.6%), 9 (8.7%), and 9 (8.7%), respectively. Sensorineural hearing loss, defined as a 15-dB hearing loss in one or more frequencies, typically affected higher frequencies. This complication is attributed to acoustic trauma and vibration injury during the drilling process and was thought to result in largely subclinical deficits. Soft-tissue stenosis and TM lateraliza-

Table 2. Summary Audiologic Data for 104 Cases

Parameter	Preoperative, dB	Postoperative dB	*P* Value[a]	Improvement, dB	Postoperative Value ≤30 dB, No. (%)	Interaural Difference, dB
Mean PTA	61.4	36.9	<.001	24.5	53 (50.9)	27.8
Mean SRT	60.2	34.3	<.001	25.9	42 (40.4)	25.4
Mean ABG	49.5	22.8	<.001	26.7	85 (81.0)	NA

Abbreviations: ABG, air-bone gap; NA, not applicable; PTA, pure-tone average; SRT, speech reception threshold.

[a] Type 1, 2-tailed *t* test comparing preoperative and postoperative means.

tion were less common complications, occurring in 6 cases (5.8%) each. There were no cases of facial nerve paresis or paralysis. Overall, 26 of 104 cases (25.0%) required revision surgery, with canalplasty (9 of 26 [34.6%]), tympanoplasty (6 of 26 [23.1%]), and OCR (7 of 26 [26.9%]) being the 3 most common revisions performed.

No statistically significant differences were found between hypoplasia and atresia cases (**Table 4**). The 3 most common craniofacial syndromes represented were hemifacial microsomia (4 of 13 [30.8%]), Goldenhar syndrome (2 of 13 [15.4%]), and PARC (poikilodermia, alopecia, retrognathism, and cleft palate) syndrome (2 of 13 [15.4%]). No statistically significant differences were found between syndromic and nonsyndromic cases. Multiple statistically significant differences were found that revealed improved results when an intact and appropriately mobile native ossicular chain was present that did not require reconstruction. The rate of revision surgery was compared between patients who underwent OCR in their initial surgery vs those who did not undergo OCR in their initial surgery. Revision surgery was performed in 15 of the 60 OCR patients and 11 of the 41 non-OCR patients ($P > .99$), whereas OCR was performed in 11 of the 60 OCR patients and 5 of the 41 non-OCR patients ($P = .58$). The overall revision rate was assessed as that at which an OCR was performed. No statistically significant difference in either parameter was found between the OCR and non-OCR groups.

Table 3. Complications of the Congenital Aural Atresia and Hypoplasia Repairs in 104 Cases

Complication	No. (%)
General complications	29 (27.9)
Chronic myringitis	10 (9.6)
SNHL[a]	9 (8.7)
Soft-tissue stenosis	6 (5.8)
TMJ prolapse	2 (2.0)
Middle ear adhesions	1 (1.0)
Bony exposure requiring STSG	1 (1.0)
Complications affecting conductive apparatus	24 (23.0)
Loose or malfunctioning prosthesis	9 (8.7)
TM perforation	6 (5.7)
TM lateralization	6 (5.8)
Ossicle fixation	2 (1.9)
Prosthesis extrusion	1 (1.0)
Overall complication rate	43 (41.3)

Abbreviations: SNHL, sensorineural hearing loss; STSG, split-thickness skin graft; TM, tympanic membrane; TMJ, temporomandibular joint.

[a] Defined as a 15-dB loss in one or more frequencies.

Discussion

Consistent with several other series[5-9] with long-term follow-up, persistent and statistically significant hearing improvements have been found. Of note, these results reflect only the net change between the preoperative audiogram and the most recent postoperative audiogram. As such, we cannot comment on the stability of the postoperative hearing results over time. However, given the overall favorable hearing results, ongoing audiologic stability would be expected.

Similar to other series,[6-8,11,12] we have reported a relatively high rate of complications (41.3%). It is noteworthy that most of these complications were conservatively resolved and many had a minimal clinical effect. Soft-tissue stenosis and TM lateralization occurred relatively infrequently at a rate of 5.8% each. This is an improvement in the rate of TM lateralization, which was 18% in a prior publication.[10] Our current rate of TM lateralization of 5.8% is consistent with most other studies.[6,7,12-14] Our rate of soft-tissue stenosis (5.8%) is consistent with a prior series[10] and other publications.[7,15] However, many other studies[11,13,14,16] have quoted this complication at significantly higher rates of 20% to 30%. In a prior series, the ear canal was filled with antibiotic ointment at the time of surgery in a significant portion of the cases. This ointment was then suctioned out about 2 weeks postoperatively and wicks placed. After personal communication with Dr Jahrsdoerfer, one of us (R.A.C.) began placing wicks at the time of surgery. This method has resulted in much better drum position and stability. Regarding meatal stenosis, we attribute our low rate of meatal stenosis to the use of the posteriorly based meatal skin flap (sutured to the posterior bony ear canal) and the anteriorly based periosteal flap (sutured to the medial aspect of the tragal area), which serve to hold the meatus open. In addition, this design avoids a circular cicatrix at the meatus.

Atresia can be considered the most extreme form of hypoplasia in which an external canal passage is completely absent. In light of prior evidence of disparate postoperative audiologic results between these 2 groups,[8] we compared hearing outcomes between patients with canal atresia and those with hypoplasia. As indicated in Table 4, audiologic outcomes between these 2 subgroups were found to be comparable. Although the acoustics of an atretic canal would be expected to be less favorable than those of a hypoplastic canal, it appears as though these discrepancies can be overcome with adequate surgical technique. By meticulously clearing the middle ear of bony overgrowths impinging the ossicles, creating a

Table 4. Subgroup Audiologic Outcomes

	Hypoplasia Cases	Atresia Cases	*P* Value[a]	Syndromic Cases	Nonsyndromic Cases	*P* Value[a]	OCR Cases[b]	Non-OCR Cases	*P* Value[a]
No. of patients	38	66		14	90		69	36	
Postoperative PTA, dB	35.2	37.9	.39	36.3	37	.94	39.2	32	.02
Postoperative SRT, dB	32.2	35.5	.25	32.9	34.6	.84	37.3	28.1	.001
PTA improvement, dB	24.3	28.8	.13	19.5	23.1	.34	25.8	31.1	.14
SRT improvement, dB	21.8	28.2	.07	21.1	26.6	.26	23.2	31.7	.02
Interaural PTA difference, dB	26.3	28.4	.54	32.8	27.5	.37	28.7	25.5	.34
Interaural SRT difference, dB	23.8	26.1	.48	28.8	25.3	.64	27.7	20.5	.02

Abbreviations: OCR, ossicular chain reconstruction; PTA, pure-tone average; SRT, speech reception threshold.

[a] Type 1, 2-tailed *t* test comparing results between the 2 subgroups; a Mann-Whitney log-rank test was used in instances of unequal variance.

[b] Includes data from initial surgery and any revision cases in which OCR was performed.

widely patent and appropriately oriented external canal, and creating as large a TM as possible, the hearing outcomes are optimized. Our similar results between syndromic and nonsyndromic cases correlate with a prior study from Sakamoto and colleagues.[17]

In a prior series, no significant differences in audiologic outcomes were found between cases in which an OCR was performed compared with cases in which an intact and mobile ossicular chain was encountered.[10] In the current larger series, statistically significant differences between these 2 groups were found in 4 of the 6 audiometric parameters assessed. Similar findings have been described previously by Dobratz and colleagues.[18] In addition, this is conceptually consistent with prior evidence revealing the ability of the Jahrsdoerfer scale score to predict hearing outcomes, given that 4 of the 10 points on this scale reflect the status of the ossicles.[4,19] There is potential for bias here, however, because associated anatomical abnormalities, such as a contracted middle ear space, may be a confounding variable.[20] Nonetheless, overall rates of revision and revision OCR were not statistically different. This finding is in contrast to the findings of the aforementioned study by Dobratz et al[18] in which statistically significant higher rates of revision were found in cases with OCR.

Despite the relatively high rates of complications and revision surgery, CAAH repair remains a favorable option for affected patients, including those who require OCR. It is generally agreed on that, on average, osseointegrated bone conduction devices impart superior audiologic outcomes compared with atresiaplasty.[21] In addition to being audiologically superior overall, the hearing results with these devices are predictable and consistent, whereas audiologic results with atresiaplasty are more variable.[22] However, one would be remiss to conclude that bone-anchored devices are superior based on this fact alone. For patients with more severe degrees of atresia, with Jahrsdoerfer scale scores of 6 or below, for example, most otologists would recommend a bone conduction device. For less severe atresia, however, CAAH repair remains the optimal choice for many patients. The advantages of CAAH repair are many and include the possibility of normal or near-normal hearing without the use of sound amplification of any kind. For others, a simple hearing aid can be used in a reconstructed canal to achieve normal hearing. Atresiaplasty provides a cosmetically superior outcome and allows for direct surveillance for infection and cholesteatoma. In contrast to the chronic and often frequent wound care problems of a percutaneous abutment,[23] the reconstructed canal typically requires only semiannual cleaning. A CAAH repair allows for superior sound localization[24] and the use of prevalent in-the-ear headphones and accessories. For these reasons, the benefits of CAAH repair often outweigh the potentially improved audiologic results of bone conduction devices in appropriately selected patients.

Conclusions

The CAAH repair imparts advantageous audiologic outcomes and a favorable safety profile. Hearing outcomes for patients with canal atresia are not different from those with hypoplasia. Patients with craniofacial syndromes have equivalent outcomes to those with isolated CAAH. Audiologic results of patients with intact ossicular chains are superior to those requiring OCR, although the need for revision surgery is not different between these 2 groups. The CAAH repair remains a favorable option for appropriately selected patients with atresia.

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Study concept and design: All authors.
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Drafting of the manuscript: Moss, Lin.
Critical revision of the manuscript for important intellectual content: Lin, Cueva.
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