ear structure and function. Regaining normal structure also aids hearing restoration. In this series, using the ossicle cup and columella, 80% of the patients eventually had a hearing gain within 20 dB of the preoperative bone-hearing level.

Since the surgical alternative for these patients was a radical mastoidectomy, the results of the closed tympanomastoidectomy with secondary staged surgical procedures to validate the excision of cholesteatoma are justified by normal structure and function being maintained. As the facts become known, the prevailing surgical opinion regarding the excision of cholesteatoma could be the closed tympanomastoidectomy.

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High-resolution *CT* scanning and auditory brain stem response in congenital aural atresia: Patient selection and surgical correlation

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Thirty patients with congenital aural atresia underwent CT scanning and/or auditory brain stem response (ABR) testing in a 20-month period. Eighteen patients had unilateral atresia and 12 had bilateral atresia. Twelve patients subsequently had surgery for repair of their atresia. CT scanning was not electively done until the patient was at least 2 years of age, while ABR testing was often performed in the first few months of life. Nineteen patients had CT scanning and 27 had ABR testing. The CT technique was found to offer specific advantages not previously observed in other methods of radiographic evaluation: (1) the course of the facial nerve was more easily traced and (2) the presence (or absence) of a stapes was more easily noted. The ABR was measured for monaural air-conduction as well as mastoid-placement bone conduction click stimuli; simultaneous multielectrode two- or four-channel recordings were employed. With this measuring technique it was not only possible to enhance wave I detection but, more important, the laterality of ABR wave I could be noted. (OTOLARYNGOL HEAD NECK SURG **93:**292, 1985.)

There are two absolute indications for surgery in congenital aural atresia: (1) radiographic evidence of an inner ear and (2) audiometric evidence of cochlear function. Until a few years ago polytomography and behavioral audiometric testing were the benchmarks for investigating patients with congenital aural atresia. The results were frequently equivocal and at times misleading. On rare occasions a nonhearing ear was inadvertently operated on. A "surgical exercise" of this nature did little to bolster the confidence of the surgeon.

Much has changed. High-resolution CT scanning has replaced polytomography as the radiographic procedure of choice, and auditory brain stem response (ABR) testing has either replaced or augmented behavioral methods of testing hearing in the young child. The task of patient selection has been made easier and the surgical result is more predictable. It is only proper that the patient has been the ultimate benefactor.

ABR and congenital aural atresia. Early assessment of auditory function in children is important for otologic and audiologic reasons. Accurate description

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of the functional status of the middle ear and sensorineural apparatus contributes to decisions regarding surgical correction of defects and the selection of appropriate amplification. The assessment of auditory function in neonates and young children is confounded by two main problems. First, it is not possible to obtain valid pure-tone speech audiometry results for each ear of a neonate or young child. Second, the serious hearing impairment associated with congenital aural atresia often creates a masking dilemma. That is, the intensity level of noise that is necessary to effectively mask the ear not being tested exceeds the interaural attenuation of the head, resulting in crossover of the noise to the test ear. Therefore, even with a cooperative young child, it is often not possible to define sensorineural function for each ear when a bilateral conductive deficit is suspected.

ABR audiometry has clinically documented value in the assessment of auditory function in neonates and young children.[1-3] The ABR is not influenced by a patient's state of arousal or by anesthetic agents; therefore, with the use of sedation, valid results can be obtained under earphones and with bone-conducted stimulation. Although ABR audiometry solved the problem of auditory assessment of the young child, traditional ABR test techniques were still confounded by the masking dilemma.

The advent of multichannel ABR recording has provided a means for describing sensorineural function in each ear, even in patients with a maximum conductive hearing impairment. The ABR is traditionally recorded with one electrode on the vertex and a second electrode on the mastoid of the side that is stimulated. In recent years, however, multiple-channel ABR measurement has been reported. For the most part these studies were conducted with experimental equipment on normal subjects.[4-6] Simultaneous multichannel recordings of the ABR are now clinically feasible with commercially available equipment. We hypothesize that, by comparing ABR recordings for the electrode on the mastoid ipsilateral to the stimulus vs. the contralateral electrode, peripheral (eighth cranial nerve) auditory function can be routinely specified for each ear without the use of masking noise.[7] We have studied multichannel ABR recordings in a series of patients with congenital aural atresia to evaluate the clinical value of this technique.

CT in congenital aural atresia. The importance of preoperative evaluation of the temporal bone with polytomography prior to surgical repair of congenital atresia of the ear has long been recognized.[8] Recently, high-resolution CT techniques rather than polytomography have been used for the preoperative evaluation of this malformation with high clinical and operative correlation. The CT technique offers the advantage of visualization of soft tissue as well as bony structures and delivers a reduced radiation dose to the critical organ (lens of the eye) in comparison with polytomography. The use of axial and coronal CT in each case compensates for the lack of direct sagittal images. The present high-resolution CT technique preserves detail yet produces life-sized images.

CT is useful in providing the following information preoperatively: (1) course of the facial nerve in relation to the planned surgical approach, (2) thickness and form of the bony atretic plate, (3) degree of development of the ossicles and whether they are fused to each other and/or to the bony atretic plate, (4) degree of development of the inner ear structures, including the internal auditory canal, labyrinthine windows, and cochlea, (5) degree of mastoid pneumatization, (6) extent of the bony external ear canal, if present, and relative contribution of soft tissue to the atresia, (7) evaluation of possible congenital cholesteatoma, and (8) the development, size, and aeration of the middle ear cavity.

METHOD

Thirty patients with congenital aural atresia were evaluated in a 20-month period. Twenty-seven patients underwent ABR testing, 19 had high-resolution CT, and 12 were operated on. The operations reviewed in this study form part of a larger series of 155 operations for congenital ear malformations. Our protocol was more heavily weighted toward early ABR testing in patients with bilateral atresia. Often, ABR testing was performed within the first few months of life. CT scanning was not electively done until the patient was at least 2 years of age. When possible, both ABR testing and CT scanning were carried out under general anesthesia in one sitting—usually at 3 to 4 years of age. Surgery was most often performed between 4 and 6 years of age.

We intended to provide, through surgery, serviceable hearing (25 dB or better) in at least one ear prior to the child entering school. When the external ear deformity was severe, that is, grade III microtia, surgery for hearing rehabilitation was delayed until the auricle was reconstructed. Patients with grade I microtia routinely had surgery for hearing first, irrespective of the need for any minor cosmetic alteration of the auricle. Patients with grade II microtia were treated on an individual basis according to their needs.

ABR data were collected with commercially available instrumentation (Nicolet CA-1000/DC-2000, Pathfinder II). Stimuli were clicks of 0.1 msec duration presented at a rate of 21.1/sec with TDH-39 earphones and MX-41 AR cushions enclosed within aural domes coupled to circumaural pads. Stimuli were always pre-

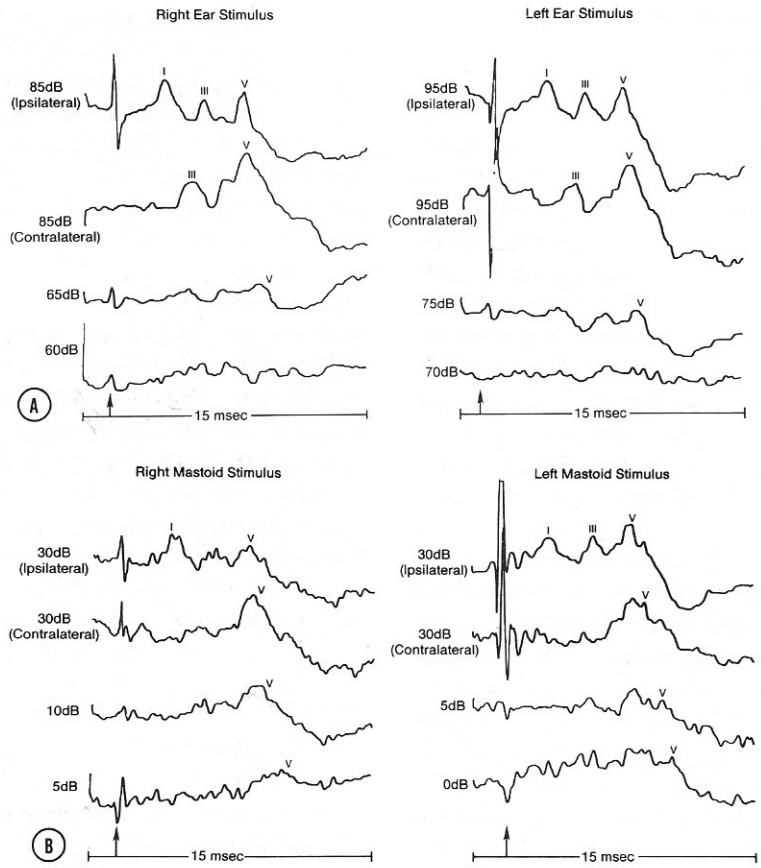

Fig. 1. ABR recordings for monaural, air-conduction stimulation **(A)** and bone-conduction stimulation **(B)** from 6-year-old girl with bilateral congenital aural atresia (case 1). Note wave I component for ipsilateral, but not contralateral, electrode array at highest stimulus intensity level.

sented monaurally to the right and left ears. Bone-conduction stimuli were presented to the right or left mastoid with a standard bone oscillator (B71A). The evoked neural signal was detected simultaneously with three standard EEG disk-type electrodes and filtered at settings of 3000 Hz. Interelectrode resistance was always less than 5000 Ω. Two electrode arrays were employed simultaneously in all ABR recordings. One of the arrays consisted of an electrode pair with a voltage-positive electrode high on the forehead and a second electrode on the earlobe ipsilateral to the stimulus. The other array consisted of the forehead voltage-positive electrode with the second electrode on the earlobe contralateral to the stimulus. A low forehead ground electrode was used for all recordings.

High-resolution CT of the temporal bones utilized the technique proposed by Chakeres and Spiegel.[9] Contiguous 1.5 mm slices were performed in one axial and one coronal plane using the GE 8800 scanner. We have found the 30-degree axial and 105-degree coronal planes to be most helpful, utilizing the 0-degree axial and 70-degree coronal planes only when necessary. Each plane, however, offers certain advantages and visualizes some structures better than others. Therefore the program should be tailored to the specific structures deemed critical to the patient. When the temporal bone is very small, as in infants or small children, 1.5 mm slices at 1.0 mm intervals with a 0.5 mm overlap may be performed for better detail of minute structures such as the ossicles.

The raw data are saved on a separate tape for processing at a later time (after the patient scanning for that day is complete). At the time of scanning, X and Y coordinates are selected for purposes of centering the enlarged images. Images are reconstructed from the raw data with a standard GE bone algorithm program and are displayed and photographed with an extended window of 4000 and a target factor of 3.5.

The right and left sides are reproduced separately using the X and Y coordinates chosen previously as the center of the enlarged images. On the newer GE 9800 scanner with PA software, life-sized images are pro-

duced by utilizing the 512 matrix, a display field of view of 12.8, the specified X and Y coordinates, the multiple display photographic format, and the large image size on the filming device.

More rapid computer processing of data on the 9800 machine also allows immediate production of the enlarged images rather than delaying this procedure as on the 8800 machine. It is important to emphasize that simple magnification techniques may not be utilized, as detail will be lost.

Although other authors believe that a single axial series of images with reformatted coronals may be used as an alternative method,[10] we believe that accuracy is improved with both axial and coronal direct images.

The method of using surgical repair to improve hearing has been outlined in detail elsewhere.[11] In brief, a standard technique was used for the canaloplasty and the meatoplasty, whereas the ossicular reconstruction was individualized, depending on the condition of the middle ear at the time of surgery.

CORRELATING ABR, CT, AND SURGERY

CASE REPORTS

Case 1. K.R.C., a 6-year-old girl with Treacher Collins syndrome and bilateral ear atresia, had ABR assessment carried out previously on two separate occasions at another audiology facility. The report of the first assessment, made when the child was 1 year 10 months of age, stated that "the overall pattern of test results suggests a moderate conductive hearing loss in the right ear." It continued, "We were unable to adequately evaluate hearing sensitivity in the left ear, as we could not effectively mask the right ear." A second assessment at 5 years of age (1983) yielded similar findings; however, the report further stated that "air-conduction sensitivity in the right ear is approximately 60 dB HL" and "the ABR response suggests a profound, primarily sensorineural loss in the left ear." If these opinions were valid, the patient would not be a candidate for surgery, as the ear under consideration would have been the only hearing ear.

At 6 years of age the patient underwent conventional audiologic testing at our facility; the results indicated a severe, bilateral (65 to 75 dB) pure-tone hearing impairment by air conduction. However, unmasked bone-conduction responses were within normal limits. Speech threshold values were in close agreement with pure-tone findings (within 7 dB). ABR testing was carried out in the operating room under general anesthesia (Fig. 1). With right ear air-conduction stimulation there was a well-formed and repeatable ABR at stimulus intensity levels of 85 dB down to 65 dB. A clear wave I component was observed when recordings were made with an electrode on the earlobe ipsilateral to the stimulus. However, with a contralaterally placed electrode no wave I was noted. There was a similar pattern to the tracings when the left ear was stimulated, although the threshold appeared to be somewhat elevated (75 dB) in comparison with the right ear. For bone conduction an ABR was observed on each side

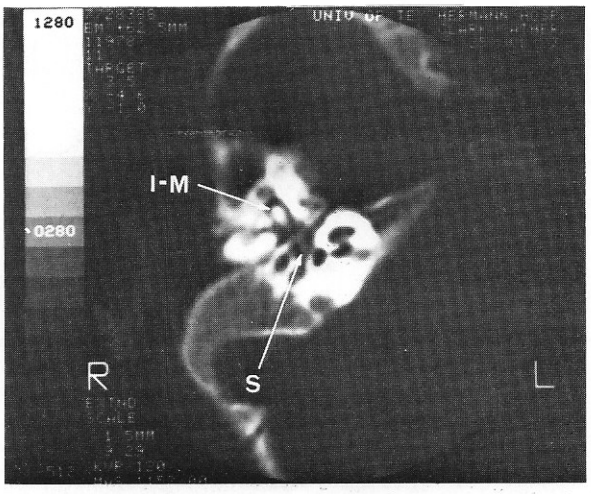

Fig. 2. Thirty-degree axial view of right ear in 6-year-old girl with Treacher Collins syndrome and bilateral atresia. Note fused incus-malleus complex *(I-M)* and stapes *(S)*.

at intensity levels of 30 dB down to 0 and 5 dB. Again, however, a clear wave I component was apparent only for the ipsilateral electrode array. With air and bone-conduction stimulation, brain stem transmission times were well within normal. These ABR results indicated a conductive hearing impairment of about 60 to 65 dB for the right ear and 70 to 75 dB for the left ear. Analysis of multichannel ABR recordings suggested normal sensorineural function bilaterally.

High-resolution CT scanning of the temporal bones showed that the inner ear structures were normal, there was bilateral atresia, and the middle ear structures were markedly underdeveloped. In the right ear (the ear under consideration for surgery) there was a fused incus-malleus complex that was situated more anteriorly than the cochlea. A stapes was identified in a small middle ear space and was separate from the fused incus-malleus. The mastoid had not pneumatized; instead, there was only bone marrow (Fig. 2).

The right ear was operated on for atresia repair on September 16, 1983. The anatomy of the temporal bone was greatly distorted. A small tympanic bone remnant was identified on the lateral surface of the temporal bone. On drilling, the first landmark to be encountered was the horizontal semicircular canal, which was inadvertently blue-lined. A nerve measuring 1 mm in diameter was noted emerging from the middle ear where one would normally find the chorda tympani nerve. Stimulation of this nerve failed to evoke movement of the face. The middle ear space contained some gluelike secretions and the lining mucosa was thickened. The stapes and oval window were not initially identified and were presumed to be absent.

While additional bone was being drilled away in an attempt to visualize a round window, a second nerve was identified and, on stimulation, the face moved well. Since the oval window was believed to be absent, a decision was made to drill a new oval window through the promontory. As thickened mucosa was being dissected from the medial wall of the

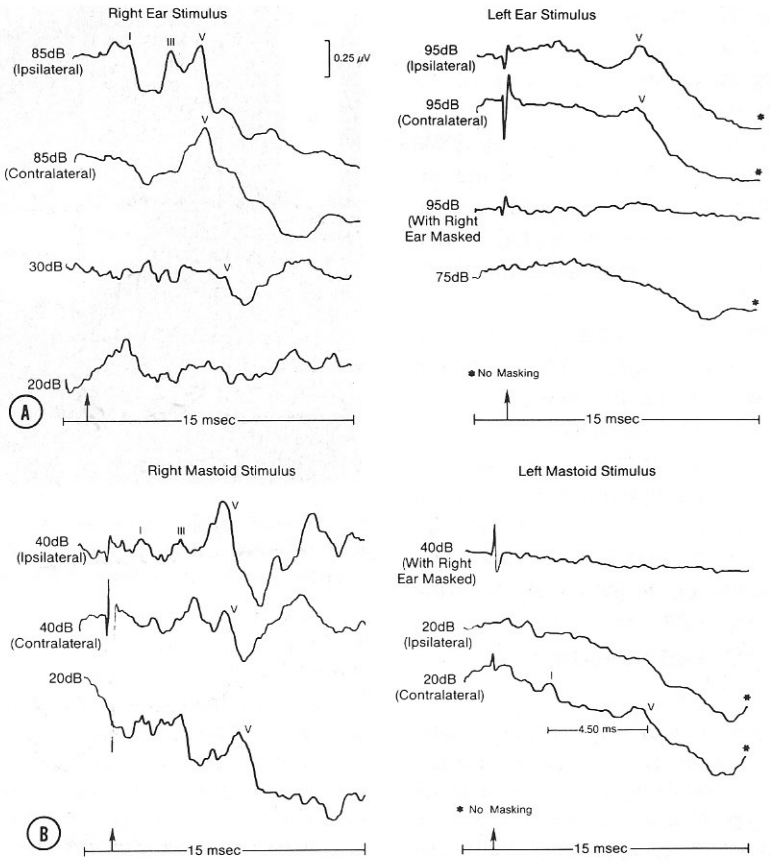

Fig. 3. ABR recordings for monaural air-conduction stimulation **(A)** and bone-conduction stimulation **(B)** from 21-year-old man with unilateral (left) congenital aural atresia (case 2). Note wave I component for ipsilateral, but not contralateral, electrode array, with stimulus right.

middle ear, a stapes remnant was palpated. Further dissection revealed a monopedal stapes partially hidden beneath the facial nerve. A partial ossicular replacement prosthesis was modified to fit the malformed stapes. The eardrum, the external auditory canal, and the meatus were constructed in the usual manner.

A postoperative pure-tone hearing threshold of 20 dB was achieved.

COMMENT. This patient would not have been subjected to a potentially high-risk operation if indeed there was a profound or total sensorineural hearing loss in one ear. However, once it was determined that there was good cochlea function bilaterally, the risk of opening one ear was justified.

Case 2. L.R.G., a 21-year-old man with unilateral ear atresia, was referred because of persistent purulent drainage from a left meatal fistula and recurring abscess formation behind the left ear. The patient had earlier undergone 10 operations to improve the appearance of the left auricle. During one operation the plastic surgeon tried to create an ear canal. The fetid odor from the ear greatly bothered the patient.

Prior to being referred to our facility the patient underwent audiologic evaluation elsewhere. Hearing was normal in the right ear, and no apparent responses for air-conduction stimuli presented to the left ear. Bone-conduction sensitivity for the left ear seemed to be within normal limits through 1000 Hz, with a 40 to 60 dB sensorineural loss for frequencies above 1000 Hz. At our facility no response could be elicited for the left ear by air-conduction or bone-conduction at equipment limits.

ABR audiometry was carried out under general anesthesia prior to definitive surgery on the left ear (Fig. 3). Right ear air-conduction stimulation yielded a well-formed ABR at intensity levels of 85 dB down to 30 dB. Latency values for all wave components were well within normal limits. A repeatable wave I component was observed with the ipsilateral electrode array but not with the contralateral electrode. ABR recordings with a left ear air-conduction stimulus at maximum intensity levels (95 dB) and no masking noise in the right ear yielded a broadly shaped wave V component at prolonged latency values. However, with adequate masking in the right ear there was no repeatable ABR. Bone-conduction findings showed that right mastoid stimulation produced a well-formed response at intensity levels down to 20 dB and below. All wave components were clearly visible, and latency values were well within normal limits. The contralateral electrode array failed to reveal a well-formed wave I component. With

Table 1. Hearing improvement (N = 12)

Before	After
55	25
68	18
55	15
40	10
55	25
65	20
65*	45-65
75	35
50	15
50	15

There was no attempt at hearing rehabilitation in 2 patients.
*Fenestrated.

left mastoid stimulation and adequate masking in the right ear there was no ABR at maximum bone-conduction equipment limits (40 dB). When masking was removed from the right ear, there appeared to be a reliable ABR for left mastoid stimulation when recorded with the contralateral electrode array. In summary, ABR findings were consistent with normal auditory sensitivity in the right ear and a severe (or worse) hearing deficit in the left ear.

Both high-resolution CT in the axial plane and lateral polytomography showed a large, soft-tissue lesion filling the middle ear space and thinning the bone laterally.

Surgery revealed a massive cholesteatoma that had expanded the tympanic bone remnant from within and greatly enlarged the area of the middle ear. There was no bony ear canal, although there was a small bony dehiscence on the surface of the domelike tympanic bone. The etiology of the cholesteatoma—congenital or acquired—is conjectural.

COMMENT. Important ABR information garnered preoperatively indicated that the left ear had a profound or total sensorineural hearing loss, and ossicular reconstruction was clearly not indicated.

SURGICAL RESULTS

Twelve patients with congenital atresia of the ear had surgery. In two patients no attempt was made at hearing rehabilitation. Both patients had recurrent abscesses of the lateral face and neck and both had a huge cholesteatoma medial to the atresia. In both patients the cholesteatoma or the accompanying infection had destroyed the middle ear structures and in one patient had caused a profound or total loss of hearing.

The hearing results in the remaining 10 patients are illustrated in Table 1. Eight patients attained a postoperative hearing threshold of 25 dB or better and one reached a threshold of 35 dB. The speech reception threshold was the parameter routinely measured; however, in two patients the pure-tone average was used. One child (3 years of age) refused to verbalize, while another child had delayed speech development. In one

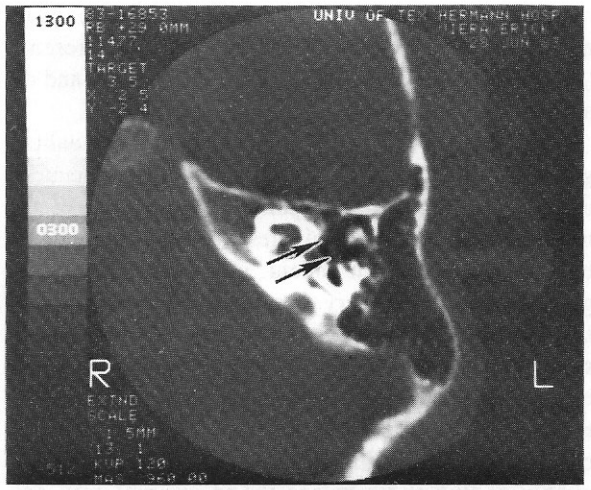

Fig. 4. Thirty-degree axial view of left ear. Note bare horizontal portion (arrows) of facial nerve crossing middle ear. There is complete atresia, and malleus is in bony union with atretic plate.

patient there was no change in hearing acuity. In this patient the oval window was noted to be absent and a fenestration of the horizontal semicircular canal was performed. Initially there was a slight hearing improvement of 20 dB that later regressed to preoperative levels.

In the remaining eight patients the average gain was 37 dB. The range of hearing improvement was 30 to 50 dB. No patient's hearing was made worse.

One patient sustained a temporary facial weakness after the facial nerve was decompressed and manipulated to better visualize the stapes and oval window. There was full return of facial function in 1 month.

DISCUSSION

Multichannel ABR recordings appear to offer a clinically feasible method of assessing auditory function, including sensorineural status, in neonates and young children with congenital aural atresia. Information for both ears can be quickly obtained with the patient either sedated or under general anesthesia. On the average, testing required less than 45 minutes. In addition, analysis of ABR recordings for electrodes placed on the earlobe or mastoid ipsilateral vs. contralateral to the stimulus permits evaluation of sensorineural function for each ear. With this measuring technique it not only was possible to enhance wave I detection but, more important, the laterality of ABR wave I could be noted. It was therefore possible to describe ear-specific sensorineural auditory function, even in patients with severe bilateral hearing deficits.

The CT technique was found to offer specific advantages not previously observed in other methods of

radiographic evaluation: (1) the course of the facial nerve was more easily traced (Fig. 4), (2) the presence (or absence) of a stapes was more easily noted, and (3) a covert cholesteatoma was better seen.

The benefits of these refined testing modalities greatly strengthen the hand of the surgeon. Accurate information from the preoperative assessment of patients with congenital aural atresia enables a surgeon to be more flexible, even lenient, in his selection of surgical candidates. Surgery on selected patients with unilateral atresia is appropriate and yields predictable results. In certain high-risk (poor-result) patients with bilateral atresia (i.e., those with scant mastoid pneumatization, a small middle ear space, absent or markedly deformed ossicles, and only a small, amorphous nubbin of skin and cartilage for an external ear), corrective surgery on one ear by an experienced operator is now justified.

We believe that multichannel ABR and high-resolution CT offer distinct advantages in the clinically challenging young patient with congenital aural atresia.

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Persistence of the stapedial artery: A histopathologic study

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Persistence of the stapedial artery is a rare event. Fewer than 30 cases have been reported since the discovery of this artery in 1836. We carried out a histopathologic study on three temporal bones from two patients who had this anomaly, and were able to trace the full course of the artery. In two specimens a large stapedial artery persisted and substituted for the middle meningeal artery. In the third, a small, persistent stapedial artery ended in the arterial plexus surrounding the facial nerve. Persistence is discussed in terms of embryogenesis, developmental theories, histologic findings, and clinical significance. The material suggests that the stapedial artery can persist to varying degrees (OTOLARYNGOL HEAD NECK SURG **93:**298, 1985.)

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