Pediatric Congenital Conductive Hearing Loss: Radiographic and Operative Findings

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Introduction

The otherwise healthy pediatric patient who presents with congenital conductive hearing loss (CHL) and a normal otoscopic exam can be a diagnostic challenge. Computed tomography (CT) of the temporal bones is the imaging modality of choice. However, the sensitivity of CT for detecting middle ear anomalies in the setting of congenital CHL has not been well-established and varies widely in the existing literature. Furthermore, the decision to operate on patients with congenital CHL in the form of an exploratory tympanotomy with possible repair of middle ear anomaly – especially in a patient with a negative CT scan – is controversial.

Given the rarity of this entity, data is limited. Consequently, it can be difficult to counsel patients and their families about the expected outcomes of evaluation and treatment. Here we present a retrospective review of patients presenting to the Children's Hospital of Pittsburgh of UPMC with congenital CHL and a normal otoscopic exam to (1) determine how frequently CT of the temporal bones identifies the cause of CHL and (2) describe the intra-operative findings and hearing results in patients who undergo exploration and repair.

Methods

- ❖ IRB-approved retrospective chart review of patients evaluated in otolaryngology clinic at a tertiary care academic children's hospital for congenital CHL from January 2003 to October 2015
- ❖ Inclusion criteria:
  - Congenital CHL by history
  - Normal otoscopic exam
  - CT temporal bone evaluation
- ❖ Exclusion criteria:
  - Patients with syndromes or major malformations involving the outer ear
  - Patients with history of prior tubes or diagnosis of middle ear disease (such as recurrent acute otitis media, chronic otits media with effusion, and cholesteatoma)
- ❖ Main outcomes measured:
  - Age of presentation
  - Gender
  - Laterality of hearing loss
  - Severity of hearing loss
  - Frequency of abnormal CT findings (as reported on original CT read)
  - Frequency of operative intervention
  - Type of intra-operative findings
  - Type of operative repairs
  - Pre- and post-operative hearing loss severity and air conduction hearing levels

Results

Table 1. Characteristics of patients (n = 39)

Age of presentation by patient (n = 39)	
	Average = 7.5 years Range = 1 to 17 years
Gender by patient (n = 39)	
	Females = 17 (43.6%) Males = 22 (56.4%)
Laterality of hearing loss by patient (n = 39)	
	Bilateral = 11 (28.2%) Left = 20 (51.3%) Right = 8 (20.5%)
Severity of hearing loss by ear (n = 50)	
	Mild = 22 (44%) Mild to moderate = 15 (30%) Moderate = 11 (22%) Moderately severe = 1 (2%) Severe = 1 (2%)

Table 2. Types of radiographic findings in patients with abnormal CT (n = 7)

Ossicular malformations	1	Dysplastic stapes
	2	Oval window stenosis
	3	Oval window stenosis
	4	Incudomalleal widening
	5	Dysmorphic ossicles fused to epitympanum
Other	6	Superior semicircular canal dehiscence
	7	Middle ear mass
	Patients that underwent surgery: n = 0	

Table 3. Intra-operative findings and hearing outcomes in patients undergoing surgery (n = 7)

	Ossicular abnormalities	Teunissen-Cremers	Repair	Severity of hearing loss		Air conduction (dB HL)		
				Pre-op	Post-op	Pre-op	Post-op	Change
1	Thickened, immobile stapes footplate with abnormal posterior crus and fibrous anterior crus	Class 1	Stapedotomy, PORP (bucket handle prosthesis)	Moderate	Normal	45	6.25	-38.75
2	Fixed stapes with minimal anterior crus; malformed incus	Class 2	Stapes mobilization	Mild to moderate	Normal	42.5	12.5	-30
3	Stapes with no obturator foramen and small footplate, minimal mobility; mucosal attachments at incudostapedial joint	Class 2	IS joint separated, stapedectomy, PORP (bucket handle prosthesis)	Moderate	Normal	48.8	17.5	-31.25
4	Fixed stapes; incus fixed to stapes	Class 2	IS joint separated, stapes mobilized, PORP	Mild to moderate	Mild	41.3	30	-11.25
5	Fibrous attachments at IS joint; mobile stapes footplate	Class 3	TORP	Moderate	Mild	40	25	-15
6	Malformed malleus connected to tympanic wall; mobile stapes footplate	Class 3	Bony attachment drilled, ossicular chain mobile after	Mild	Normal	25	11.3	-13.75
7	IS discontinuity, stapes not attached to visible footplate, ossified OW with overhanging FN	Class 4	Aborted	Moderately severe	Moderately severe	57.5	61.3	+3.75
	Patients that had abnormal pre-operative CT findings: n= 0					Average change (dB HL)		= -15.5

Discussion

In our case series of 39 patients, 7 patients (17.9%) had abnormal CT findings. Five patients had evidence of an ossicular malformation, while 2 patients had other abnormal middle ear and inner ear abnormalities (middle ear mass and superior semicircular canal dehiscence, respectively). However, none of these patients opted to undergo surgery, thus prohibiting validation of their radiographic findings. A separate group of 7 patients (17.9%) underwent exploratory tympanotomy. Interestingly, all patients who underwent surgery had pre-operative CT's that were read as normal.

Given the low incidence of positive CT's and the 100% false negative rate of CT's within the operative group, our series indicates that the sensitivity of CT for detecting congenital middle and inner ear anomalies is low.

Regarding patients' intra-operative findings, Teunissen-Cremers class 2 and class 3 ossicular anomalies were most commonly identified, which is similar to previously reported incidences. Six of 7 patients underwent repair of ossicular anomaly. One patient was aborted secondary to unfavorable anatomy. All patients who underwent repair demonstrated improvement in post-operative hearing, with an average improvement of 15.5 dB (95% CI = 4.2 – 26.7, p = 0.02).

Conclusions

- ❖ The detection rate of abnormalities on CT in patients with congenital CHL is low.
- ❖ Operative exploration is performed infrequently, but when performed, is successful and results in significant hearing gain.

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