

On-line Table 1: Results of CT studies of CH^a

Hearing Category	No. of Ears	Min.	Max.	Mean	SD	P Value
SNHL (all)	162	2.5	6.1	5.23	0.46	.002 ^b
Males	90	4.0	6.1	5.29	0.37	.004 ^b
Females	72	2.5	6.1	5.16	0.55	.33
CHL (all)	101	4.2	6.1	5.32	0.37	.09
Males	61	4.2	6.1	5.38	0.39	.17
Females	40	4.7	6.1	5.24	0.33	.60
Mixed HL (all)	25	3.4	5.5	4.67	0.69	<.00001 ^b
Males	11	3.4	5.5	4.53	0.69	<.00001 ^b
Females	14	3.5	5.4	4.79	0.70	.008 ^b
Normal (all)	74	4.5	6.3	5.42	0.38	1
Males	52	4.5	6.3	5.48	0.38	1
Females	22	4.6	6.0	5.28	0.35	1
Unknown (all)	60	4.6	5.9	5.29	0.30	.04
Males	32	4.6	5.9	5.32	0.35	.06
Females	28	4.6	5.6	5.26	0.26	.82
Total (all)	422	2.5	6.3	5.26	0.45	
Males	246	3.4	6.3	5.32	0.43	.009 ^c
Females	176	2.5	6.1	5.18	0.47	

Note:—Min. indicates minimum; Max, maximum.

^a Two-sample *t* test *P* values are for comparisons among SNHL, CHL, mixed HL, or unknown-hearing ears and normal-hearing ears divided by all, male, or female.

^b Statistically significant differences (*P* < .01).

^c For the comparison of male and female CHs, multivariate linear regression controlling for age and ICW was used.

On-line Table 2: Patients with cochlear hypoplasia^a

Patient	Age (yr) Sex	L CH (mm)	R CH (mm)	TYPE HL	Diagnosis	CT Findings
1	6.3 Female	2.8	2.5	SNHL, bilaterally	Bilateral vestibulocochlear dysplasia, L Mondini malformation	Bilateral vestibulocochlear dysplasia R common chamber malformation, absent vestibular aqueduct, aplastic modiolus L Mondini malformation (hypoplastic cochlea with partition defect)
2	1.9 Male	3.4	3.6	Mixed HL	BOR syndrome	Bilateral dilated vestibular aqueduct with small modiolus, trumpet-shaped IAC, small mass in R middle ear (possible congenital cholesteatoma)
3	0.8 Female	3.7	3.5	Mixed HL	BOR syndrome	Bilateral tympanostomy tubes, hypoplasia of modiolus, vestibular ectasia, dilated vestibular aqueducts, Mondini deformities (cochlear ectasia with partition defects), question of ossicular fusion R middle ear and mastoid air cell opacification; absent vs hypoplastic stapes L hypoplastic mastoid with soft tissue thickening at L mesotympanum
4	6.7 Female	3.6	4.3	Mixed HL, bilaterally	CHARGE syndrome	Bilateral hypoplastic SCCs, prominent EAC, vestibular dysplasia, cochlear ectasia with partition defects, hypoplastic stapes R sclerosis of ossicles, poorly visualized oval window, L poorly defined modiolus
5	0.1 Male	4.2	3.8	Mixed HL, bilaterally	CHARGE syndrome	Bilateral absent SCCs, middle ear/mastoid air cell congestion or inflammation, normal middle ear morphology
6	6.6 Male	4.0	4.5	SNHL, bilaterally	EVA syndrome	Bilateral enlarged vestibular aqueducts, R cochlear implant, L hypoplastic modiolus
7	7.1 Male	4.2	4.3	CHL, bilaterally	Bilateral class II microtia and EAC atresia	R malformed IAC, partially formed middle ear cavity, poorly defined ossicles, narrow EAC, normal SCCs/ vestibule L absent lateral SCC, vestibular dysplasia, ossicular dysplasia, absent membranous EAC, normal cochlea

Note:—R indicates right; L, left; IAC, internal auditory canal; SCC, semicircular canals; BOR, branchio-oto-renal; EVA, enlarged vestibular aqueduct; EAC, external auditory canal.

^aCHs >2 SDs below the mean CH.