Temporal bone computed tomography findings in bilateral sensorineural hearing loss

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Abstract
Aim—To examine the yield of computed tomography (CT) of the temporal bones when investigating sensorineural hearing loss (SNHL) and to identify factors associated with CT findings.
Methods—Retrospective analysis of 116 consecutively investigated children with bilateral SNHL at the audiology department of Great Ormond Street Hospital, London. Main outcome measures were CT results, hearing loss parameters, history, and clinical examination.
Results—A total of 33 (28.4%) CT scans were identified as abnormal. Children with profound and/or progressive hearing loss and/or craniofacial abnormalities were more likely to have an abnormal CT scan and together accounted for 25 abnormal CT scans. Sex, consanguineous parents, or family history of SNHL were not associated with CT findings. Dilated vestibular aqueduct was significantly correlated with the presence of progressive SNHL.
Conclusions—All children with SNHL should undergo radiological investigation of the petrous bones/inner ear; abnormalities are more likely to be found in cases with craniofacial abnormalities, or profound or progressive hearing loss. The decision whether to perform a CT or magnetic resonance imaging will depend on scanner availability, expertise, and management considerations, but cochlear implant candidates will require both.
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Computed tomography (CT) of the petrous bones is a first line recommended investigation of sensorineural hearing loss (SNHL).[1] In cases of bilateral SNHL, the CT scan may be abnormal in 6.8%[2] to 12.8%,[3] and in up to 30% of cochlear implant candidates.[4] However, some authors[3] argue that CT may be unjustified for children with stable hearing impairment and no other abnormalities.

Our aim was to examine the CT yield in the investigation of SNHL in children, to identify factors correlated with CT findings, and to assess the effect of the CT findings on management.

Materials and methods
The study included patients with SNHL who had been investigated regarding the aetiology of their hearing loss at the audiology department, Great Ormond Street Hospital between January 1996 and June 1998.

Criteria for inclusion in the study were:
- Bilateral SNHL, defined as:
  - air conduction thresholds in the standard pure tone audiogram (PTA), averaged across 500, 1000, 2000, and 4000 Hz, worse than 20 dB hearing loss in the better hearing ear or
  - high frequency hearing loss, with low and mid frequency thresholds better than 20 dB hearing loss and average threshold at 4000 and 8000 Hz worse than 20 dB hearing loss in the better hearing ear
  - air–bone conduction gap of 10 dB hearing loss or less, as identified by masked bone conduction audiometry, in both ears.
- Bilateral SNHL, as above, and fluctuating conductive type hearing loss consistent with the presence of otitis media with effusion.

Hearing loss was classified by averaging air conduction thresholds across 500, 1000, 2000, and 4000 Hz[1] of the better hearing ear, as: mild, > 20 and $\leqslant 40$ dB hearing loss; moderate, > 40 and $\leqslant 70$ dB hearing loss; severe, > 70 and < 95 dB hearing loss; profound, $\geqslant 95$ dB hearing loss.

High frequency SNHL was defined as average threshold across 500–4000 Hz better than 20 dB hearing loss, and average threshold of 4000 and 8000 Hz over 20 dB hearing loss.

CT scans of the petrous bones and internal auditory meatuses were performed on a Siemens Somaton +4 scanner in 1 mm serial axial and coronal cuts. Axial scans were oriented parallel to the infraorbital–meatal line, while coronal scans were tilted at a 105° angle to this line.

STATISTICAL ANALYSIS
The Statistical Package for the Social Sciences (SPSS) was used to analyse the study results. The results were summarised as means, medians, and percentages. Subgroups within the study population were compared by χ^2 and Mann–Whitney tests. A p value of 0.05 or less was accepted as indicating statistically significant results.

Results
Review of the records identified 116 children with bilateral SNHL who underwent CT investigation during the period of the study. Nine children had progressive hearing loss (7.8%). Males predominated in the sample (70 males (59.8%) and 46 females (40.2%)), but there was no significant difference in the degree of the SNHL between boys and girls (Mann–

Table 1   Epidemiological data summary

Total number	116
Age (years), mean (SD)	9.1 (4.47)
Age at diagnosis (years), mean (SD)	3.2 (2.5)
Male, n (%)	70 (59.8)
Progressive, n (%)	9 (7.8)
Parents related, n (%)	19 (16.4)
Family history of SNHL, n (%)	34 (29.3)

Whitney test, p >> 0.05). The child's parents were related in 19 cases (16.4%). There was a first or second degree relative with documented SNHL of possible genetic origin in 34 cases (29.3%). Table 1 summarises these results.

There were 38 cases (32.8%) with profound SNHL (table 2). The severity of SNHL (as an average or as percentage of profound cases, for example) was not significantly different between the two sexes (p >> 0.05).

All 116 children had CT scans of the petrous pyramids and internal acoustic meatuses (IAM). A total of 33 CT scans (28.4%) were reported as showing abnormalities of the inner ear, internal auditory meatuses, or other associated structures. Table 3 summarises these results.

The diagnosis of the aetiology of hearing loss was based on CT findings and other investigations as well as on the family history of hearing loss, the child's medical history, and audiometric patterns. Identification of factors that might have caused the hearing loss was possible in 93 cases (80.2%) and unknown in 23 (19.8%). Of the 93 cases with an available (presumptive) diagnosis:

- 51 (44%) were classified as genetic: 20 indicating autosomal recessive inheritance, nine indicating autosomal dominant, four indicating mitochondrial, one indicating X linked, five chromosomal, and 12 genetic syndromic (three

Table 2   SNHL characteristics

	n (%)
Mild	14 (12.1)
Moderate	24 (20.7)
Severe	32 (27.6)
Profound	38 (32.8)
High frequency SNHL	3 (2.6)
Missing data*	5 (4.3)

*Missing data applies to cases in which the degree of SNHL was not fully established (information only from auditory brainstem response (ABR)—all five cases with ABR thresholds exceeding 70 dB nHL).

Table 3   CT scan results

CT report	n	%
Normal	83	71.6
Common cavity deformity	1	0.9
Mondini	3	2.6
Mondini and dilated vestibular aqueduct	4	3.4
Dilated vestibular aqueduct	6	5.1
Narrow IAM	3	2.6
Abnormal vestibule/semicircular canals	7	6.0
Abnormal cochlea, vestibule, and IAM	5	4.3
Cerebral abnormalities	1	0.9
Miscellaneous*	3	2.6

The common cavity deformity consists of absent or dilated cochlear basal coil widely communicating with the vestibule. "Mondini" describes a cochlea with a basal turn and a distal sac. Dilated vestibular aqueduct refers to middle third diameter of more than 1.5 mm.
*Two cases with skull base deformity and one with high jugular bulb on one side.

branchio-oto-renal syndrome (BOR)—all with Mondini type cochlea; eight Pendred syndrome—four with dilated vestibular aqueduct and four with dilated vestibular aqueduct and Mondini type cochlea; and one Usher I syndrome*)
- 10 (8.6%) congenital syndromic (four CHARGE, one VATER RAPADILINO*, one case first arch syndrome, four unclassified)
- five (4.3%) congenital—unclassified (sporadic or autosomal recessive, all with abnormal CT scans)
- six (5.2 %) following prenatal complications
- seven (6%) following perinatal complications
- two (1.7%) ototoxic (gentamycin)
- six (5.2%) post-meningitic
- four (3.4%) post-viral infections
- two (1.7%) post-traumatic.

*Note: Usher I syndrome is an autosomal recessive disorder characterised by early onset retinitis pigmentosa, severe to profound hearing loss, and absent vestibular responses.[7] The acronym CHARGE describes the association of colobomata, heart defect, atresia of the choanae, retarded growth/development, genital hypoplasia, and ear anomalies or deafness. The acronym VATER describes the association of vertebral defects, anal atresia/stenosis, tracheo-oesophageal fistula, radial defects, and renal anomalies.[5] The acronym RAPADILINO describes radial defects, absent/hypoplastic patellae and high/cleft palate, diarrhoea (and dislocated joints), little size, a long/slender nose, and normal intelligence, with hearing loss also a feature.[6]

Table 4 summarises these results (diagnostic labels against number of positive CT findings in each subgroup).

The presence of progressive hearing loss was a significant predictor of positive (abnormal) CT outcome (p = 0.01564, Fisher's exact test). Six of nine cases (66.6%) with progressive SNHL had a positive CT scan, while only 26 of 103 (25.24%) children with non-progressive hearing loss had a positive CT scan. (Note: there were four missing data regarding progressiveness of SNHL.) Cases with dilated vestibular aqueduct (DVA), with or without other abnormalities, were more likely to have progressive SNHL than the rest of the sample population (p = 0.02397, Fisher's exact test). Three of nine cases (33.3%) with DVA had a progressive hearing loss but

Table 4   Aetiological diagnosis of bilateral SNHL and abnormal CT scans

Diagnostic labels	n (%)	Abnormal CT scan (n)
Genetic	51 (44)	21
Congenital syndromic	10 (8.3)	6
Congenital non-syndromic	5 (4.3)	5
Prenatal	6 (5.2)	0
Perinatal	7 (6)	0
Ototoxic	2 (1.7)	0
Post-meningitic	6 (5.2)	1
Post-childhood infections	4 (3.4)	0
Post-traumatic	2 (1.7)	0
Unknown aetiology	23 (19.8)	0
Total	120	33 (27.5%)

only six of 107 cases (5.6%) without DVA had progressive hearing loss.

Children with profound SNHL were more likely to have an abnormal CT scan (p = 0.00583), as 17 of 38 (44.7%) profoundly hearing impaired children had an abnormal CT scan, while of the remaining 75 children with mild, moderate, severe, and high frequency SNHL, only 15 (20%) had abnormal scans. Craniofacial abnormality was also significantly correlated with an abnormal CT scan (p = 0.00794): nine of 16 (56.25%) children with craniofacial abnormalities had a positive CT scan, but only 24 of 100 (24%) children with no craniofacial abnormality had an abnormal CT scan. Neither consanguineous parents, nor a family history of hearing loss, nor sex of the patient were significantly correlated with an abnormal CT scan (p > 0.05).

Discussion

We found a 28.4% yield of CT in identifying abnormalities relevant to the cause of SNHL in children in our tertiary tier audiology department. This is similar to the findings of Woolford *et al*,[4] who reported 29.5% abnormal CT scans in adults and children awaiting cochlear implantation. Shusterman *et al*,[3] however, reported a yield of 12.85%, while an earlier study by Zalzal *et al*[2] reported an incidence of 6.8% abnormal CT findings in children with SNHL. This wide range of CT yields may be explained partly by the improvement of imaging techniques and better understanding of SNHL related inner ear abnormalities over the years, as well as by the different populations targeted by the different studies.

The male preponderance in children with SNHL (59.8% in our study) has been commented on by several authors.[8] [9] We found no statistically significant sex difference, in terms of either severity of hearing loss or CT findings. A convincing explanation for the male preponderance is still lacking, although possible explanations include preferential referrals of boys to tertiary audiology centres, male children being more susceptible to hearing damage, and genetic differences.

DVA was the most common CT abnormality in our series (10 of 116 cases), as in other studies.[10] DVA was an isolated finding in six (60%) and an associated finding in four (40%) of our cases, in contrast with the findings of Valvassori and Clemis,[11] that DVA in association with other cochlear abnormalities is more prevalent than as an isolated finding. Three of our nine cases with DVA had progressive hearing loss (information was missing regarding progressiveness of hearing loss in one case), similar to the report of Antonelli *et al*.[12] Cases with DVA were more likely to have progressive hearing loss than the rest of the sample population, and this association was statistically significant. This information was particularly relevant in terms of management, as hearing thresholds may deteriorate after trauma when DVA is present,[12] [13] so we advised all patients with DVA to avoid contact sports.

Mondini-type dysplasia, defined as presence of a cochlea with a normal basal turn and a distal sac,[14] was the third commonest abnormality, as an isolated finding in three cases, and in association with dilated vestibular aqueduct in four cases. Jackler *et al*[15] found this abnormality, described as incomplete partition, in 41 of 98 cases with abnormal CT scans (41%), and proposed that this deformity corresponds to an arrest in development during the seventh week in utero. These authors also reported the presence of dilated vestibular aqueduct in 10 of 41 ears (24%), but estimated that DVA was under reported in their study, as lateral polytomograms were not available for all their patients.

Four of our children with DVA and four children with DVA and Mondini were thought to have Pendred's syndrome, an autosomal recessive condition that consists of congenital hearing loss and thyroid goitre or positive perchlorate test.[16] Phelps *et al*[16] reported dilated vestibular aqueduct present in all 120 cases with Pendred's syndrome they reviewed, while Mondini-type dysplasia was also a common finding. Branchio-oto-renal syndrome (BOR) is an autosomal dominant disorder, with hearing loss, preauricular pits, renal anomalies, branchial fistulae, pinna, and external auditory canal deformities as major features.[17] Our three cases with BOR syndrome had a Mondini-type abnormality of the cochlea, in accordance with previously published reports.[17]

Abnormalities of the semicircular canals were the second commonest abnormality in our series, occurring in nine of 116 cases (7.75%). The semicircular canals were absent or dysplastic as an isolated finding in three cases (two with the CHARGE association and one VATER RAPADILINO syndrome), in association with a vestibular abnormality in four cases (two were CHARGE cases), and in association with a vestibular and cochlear abnormality in two cases. Semicircular canal and vestibular abnormalities are frequent findings in cases with SNHL,[15] while deformed or absent semicircular canals is the hallmark of the CHARGE association.[18] The acronym CHARGE was coined by Pagon *et al*[19] to describe the association of colobomata, heart defect, atresia of the choanae, retarded growth/development, genital hypoplasia, and ear anomalies or deafness.

The CT was abnormal in one of six (16.6%) patients with previous documented meningitis. Brookhouser *et al*[20] identified 10 abnormal CT scans in 57 conducted (18%) on similar cases. CT scan did not identify any abnormalities in either of our two patients with post-traumatic hearing loss.

Profound or progressive hearing loss and craniofacial abnormalities were significant predictors of abnormal CT findings (p < 0.05) and together accounted for 25 of 33 abnormal CT scans (75%). However, adherence to this joint criterion (presence of profound and/or progressive SNHL and/or craniofacial abnormalities) for CT scanning investigations would have missed one in four cases with abnormal CT scan, or 6.8% of cases of our total sample.

The CT was instrumental in the aetiological diagnosis in all our cases and helped to clarify

management issues in most. The CT findings helped substantiate the diagnosis of genetic hearing loss in 21 cases and of a syndromic diagnosis in six, while we are keeping under review the five "congenital" classified cases with abnormal CT scans, as evidence supporting the diagnosis of an autosomal recessive pattern which may emerge in the future.[21] The above mentioned families have received or are about to receive genetic counselling (apart from a few who declined this offer). Furthermore, additional investigations were initiated in 13 cases, on the basis of the CT results. All three patients with narrow IAM had magnetic resonance imaging (MRI) performed, to identify whether nerve VIII was present and to decide on hearing amplification issues; all 10 cases with DVA were referred for a perchlorate test to exclude Pendred's syndrome. In addition, all patients with DVA were advised to abstain from sports with a high risk of accidents, as their hearing thresholds might deteriorate after trauma.[12][13]

We communicated normal CT findings by letter to patients and discussed them at a regular follow up visit, while abnormal CT findings were personally communicated to parents at an early appointment. We believe that parental stress is reduced and the child's hearing impairment better accepted when we give thorough information on the cause and prognosis of hearing loss for the child and family members. Counselling parents and child is a significant part of the child's management, and CT can provide excellent information for this purpose. CT is the investigation of choice to assess the middle ear.[23] However, CT may not show the presence of labyrinthitis obliterans reliably in all post-meningitic cases,[22] or show the presence or absence of nerve VIII,[23] so, for these purposes, thin section high resolution MRI is indicated. This is particularly the case for cochlear implant candidates, in order to avoid inappropriate cochlear implantation.[23]

CONCLUSION

All children with SNHL must have radiological investigations of the petrous bones/IAMs. The CT scan is more likely to be abnormal in the presence of craniofacial abnormalities, and/or profound or progressive hearing loss. However, not performing CT scan in those with neither profound nor progressive hearing loss, or craniofacial abnormalities may leave up to one of four CT documentable abnormalities un-

identified. The decision whether to do a CT or MRI will depend on scanner availability, expertise, and management considerations, but cochlear implant candidates will require both.

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