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Diagnostic Yield of Computed Tomography Scan for Pediatric Hearing Loss: A Systematic Review

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Abstract

Background—Computed tomography (CT) has been used in the assessment of pediatric hearing loss, but concern regarding radiation risk and increased utilization of magnetic resonance imaging (MRI) have prompted us toward a more quantitative and sophisticated understanding of CT's potential diagnostic yield.

Objective—To perform a systematic review to analyze the diagnostic yield of CT for pediatric hearing loss, including subgroup evaluation according to impairment severity and laterality, as well as the specific findings of enlarged vestibular aqueduct and narrow cochlear nerve canal.

Data Sources—PubMed, EMBASE, and the Cochrane Library were assessed from the date of their inception to December 2013. In addition, manual searches of bibliographies were performed and topic experts were contacted.

Review Methods—Data from studies describing the use of CT in the diagnostic evaluation of pediatric patients with hearing loss of unknown etiology were evaluated, according to a priori inclusion/exclusion criteria. Two independent evaluators corroborated the extracted data. Heterogeneity was evaluated according to the I^2 statistic.

Results—In 50 criteria-meeting studies, the overall diagnostic yield of CT ranged from 7% to 74%, with the strongest and aggregate data demonstrating a point estimate of 30%. This estimate corresponded to a number needed to image of 4 (range, 2–15). The most commonly identified findings were enlarged vestibular aqueduct and cochlear anomalies. The largest studies showed a 4% to 7% yield for narrow cochlear nerve canal.

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Supplemental Material

Additional supporting information may be found at <http://otojournal.org/supplemental>.

Author Contributions

Jenny X. Chen, analysis, interpretation, drafts, final approval; **Bart Kachniarz**, analysis, interpretation, drafts, final approval; **Jennifer J. Shin**, concept, design, analysis, interpretation, drafts, final approval.

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Conclusion—These data, along with similar analyses of radiation risk and risks/benefits of sedated MRI, may be used to help guide the choice of diagnostic imaging.

Keywords

hearing loss; imaging; computed tomography; diagnosis; pediatric; infant; child; adolescent; systematic review

Introduction

Hearing loss is a regularly encountered pediatric problem with significant implications for childhood development. Approximately 9% to 16% of school-age children are affected by some form of hearing impairment,^{1–4} and studies of affected students have shown that they are prone to significantly worse academic performance, behavior, and self-esteem than their normal hearing peers.^{1,5–8} The diagnostic assessment of pediatric hearing loss may involve a range of studies, such as genetic testing, electrocardiogram, and imaging evaluation. Imaging has classically been performed with computed tomography (CT),⁹ which has the capacity to identify anomalies of the cochlea, vestibular aqueduct, and other key aspects of the temporal bone. Concerns regarding the attendant radiation exposure have been raised, refuted, and debated in public forums such as the *New York Times* and *Newsweek*,^{10–12} bringing into question what role CT should have in the evaluation of our affected youth (Paul H. Ellenbogen, MD, FACR, e-mail communication, April 7, 2014). More recently, magnetic resonance imaging (MRI) has also been used in the evaluation of infants and children with hearing loss,¹³ either in concert with or in lieu of CT. The decision to use either or both modalities is multifaceted¹⁴ and ideally involves a thorough understanding of the unique benefits and risks associated with each option.

Diagnostic test selection involves a variety of factors, including the clinical pretest probabilities, diagnostic yield, potential harms, and additional available test options. Accordingly, the decision to pursue a CT scan for pediatric hearing loss involves an understanding of not only the specific patient characteristics but also (1) the expected diagnostic yield, (2) the potential risks of the attendant radiation, and (3) the additionally available imaging options. Specific patient characteristics, such as whether hearing loss occurs in isolation or with other clinical findings, as well as the type, severity, and laterality of the impairment, may also influence the decision.^{13,14} In addition, the clinical implications of potentially expected findings play a role. Our overarching goal was thus to investigate the 3 aspects listed above, so as to provide caregivers with concrete, evidence-based information upon which to base the decision to obtain a CT scan in the setting of pediatric hearing loss. Systematic reviews provide a rigorous method to evaluate the current best evidence regarding a specific clinical question and are among the highest levels of evidence available.^{15–17} The objective of the current systematic review was to evaluate the first of the 3 enumerated concepts above, in order to support decisions regarding CT in pediatric patients with hearing loss (sensorineural, mixed, or conductive). More specific, the goal of this systematic review was to determine (1) the prevalence of imaging-identified diagnoses in those undergoing CT for hearing loss, (2) subgroup-specific diagnostic yield according to

hearing severity and laterality, and (3) the prevalence of specific diagnoses among those with abnormal findings on CT.

Methods

A computerized search was performed to focus on the diagnostic yield of CT scan for infants, children, and adolescents with hearing loss. Computerized and manual searches were performed to identify all relevant data. A PubMed search of MEDLINE from 1966 to December 2013 was performed. Articles that mapped to the medical subject heading “tomography, X-ray computed” (exploded) and those that mapped to keywords “computed tomography” were collected into a first group. Next, articles mapping to the exploded medical subject headings “hearing loss,” “ear, inner/diagnosis,” “ear, inner/pathology,” and “ear, inner/radiography” as well as the keyword “hearing” were collected into a second group. Articles that mapped to the exploded medical subject headings “child” and “infant” and those that mapped to the keywords “pediatric” and “newborn” were then collected into a third group. The 3 groups were then cross-referenced (Appendix S1, available at <http://otojournal.org>) and limited to those with human subjects and English language. Case reports as defined by the database’s publication type variable “case reports” were excluded.¹⁸ Two independent searches were performed by individuals blinded to the others’ results. In addition, searches with corresponding terms were repeated in EMBASE and the Cochrane Library to December 2013. In accordance with standard systematic review techniques, all journals indexed to these databases were included by default, thus spanning the range of all available impact factors.

This initial computerized search yielded a total of 794 studies. The abstracts were evaluated according to the inclusion/exclusion criteria described below. Reference lists from criteria-meeting publications and narrative reviews were manually searched for additional studies, yielding 53 additional potential articles. Experts in the field were contacted for any additional reports of published or unpublished data. Titles and abstracts for all identified studies were reviewed, and ultimately, 379 full articles were evaluated (Figure 1 and Figure 2).

Inclusion/Exclusion Criteria

Articles identified by the search strategy described above were evaluated to identify those that met the following inclusion criteria: (1) patient population younger than 21 years with unilateral, bilateral, conductive, mixed, or sensorineural hearing loss (SNHL); (2) CT temporal bone or head performed for the purpose of diagnosing or guiding management of hearing loss; and (3) outcome measured in terms of the proportion of those undergoing CT in which the imaging establishes a diagnosis of a temporal bone anomaly or further delineates the specific types of anomalies identified. Prospective, retrospective, and comparative studies as well as case series were included. Articles were excluded if (1) patients were older than 21 years; (2) hearing results were not delineated; (3) hearing loss was temporary; (4) no CT of the temporal bone or head was performed; (5) CTs were obtained for reasons not associated with hearing loss; (6) the cause of hearing loss in the study population had already been previously fully identified; (7) syndromic patient

population; (8) no quantitative data were presented; and (9) isolated case reports. Case reports were defined according to a standard definition of a “single clinical observation whose principal purpose is to generate hypotheses regarding human disease or provide insight into clinical practice.”^{19,20} This process yielded 50 studies that met our inclusion criteria.

Manual Search

In general, a computerized search has limitations, particularly if the topic assessed is diagnosis. The sensitivity and specificity of the best single term and combinations for high sensitivity MEDLINE searches are just 0.80 and 0.77, respectively.²¹ Accordingly, a systematic review standardly includes a manual search to supplement the computerized inquiry.²²

The manual search for this query resulted in 53 titles and 5 additional criteria-meeting papers, as depicted in the more detailed flow charts in Figure 2. Considering that 50 criteria-meeting studies were included in the end, the number of papers identified by manual search falls within expected parameters, given the sensitivity and specificity described above.

Data Extraction

Data extraction additionally focused on potential sources of heterogeneity or bias among those results and study identification (author, year of publication, full reference citation). Extracted data included (1) the number/percentage of patients with CT scans that revealed a new diagnosis of temporal bone anomaly, (2) the number/percentage of subsets of specific types of anomalies identified by CT, (3) consecutive or nonconsecutive status of reported patients, and (4) the mean follow-up time. Also collated were (1) age at CT, (2) the extent of hearing loss in patients studied, (3) types of hearing loss studied (mild, moderate, severe, profound, unspecified; bilateral or unilateral; sensorineural, mixed, or conductive), and (4) study design with potential confounders. Two reviewers corroborated extracted data independently using standardized tables. In accordance with data demonstrating that overall “study quality” ranking scales may be misleading or give heterogeneous results,^{23–26} we focused on evaluation of data quality by consistent factual description of individual elements of study design with attention to prospective/retrospective analysis and assessment of consecutive patients.

Quantitative Data Analysis

The extracted data were analyzed for heterogeneity to determine if pooling of data would be appropriate. Data were examined in subsets according to clinical hearing loss characteristics: severe to profound, bilateral, unilateral, and no conductive/mixed component. Studies of children with severe, bilateral hearing loss were included in both related subsets. Diagnostic yield was defined as the proportion of patients affected according to the defined imaging modality: $\text{yield} = (\text{number of patients with imaging-established diagnoses})/(\text{number of patients imaged})$. Nearly all studies reported their findings per patient, but in the minority instance when it was reported per ear, the data were nonetheless included in the systematic review and numerical analyses in the translated per-patient increment, since the decision to image is made at the level of the patient, rather than 1 ear at a time. In the single instance

where data were reported solely on a per-ear basis,²⁷ these data were withheld from the aggregate analyses so as to not confound the per-patient measurement.

For counts of all diagnoses, any reported CT finding made by the imaging modality indicated was enumerated, also at the patient level. Thus, every effort was made to (1) ideally use a composite total number of affected patients from the primary report, and (2) account for the potential for overlapping diagnoses in a single patient when 1 was not provided. For this latter reason, if the affected number of patients was reported such that it was unclear whether the findings did or did not overlap within the same patients, the individual numbers were not simply summed to establish a total. In the case where more than 1 system was used to evaluate a single diagnosis in the same subset of patients, the system that the authors espoused in conclusion was used in the analysis.²⁸

Heterogeneity among studies was evaluated using the I^2 statistic, which is a measure of the variation between studies that exceeds that from chance alone. Perfectly homogeneous studies have a theoretical I^2 value of 0%. The range from 0% to 40% is thought to represent unimportant heterogeneity, whereas the overlapping values of 30% to 60% and 50% to 90% have been postulated to represent moderate and substantial heterogeneity, respectively.^{29,30} Since the number of studies in subgroup analyses was often small or results were notably variable, 95% confidence or “uncertainty” intervals were calculated.³¹ An a priori plan was made to pool data for a formally presented meta-analysis in the instance where the group/subgroup’s point estimate for I^2 was $< 60\%$ and the 95% confidence interval (CI) overlapped by 0% to 40%.

Meta-analyses were performed using a random effects analysis, according to the standard technique of DerSimonian and Laird^{32,33} to obtain a weighted pooled risk difference or pooled proportion. Statistical analyses and calculations were performed in Stata 12.0 (College Station, Texas, USA), Medcalc (Ostend, Belgium), and Microsoft Excel (Redmond, Washington, USA). Since no group or subgroup analyses met the a priori heterogeneity threshold described above, the data for meta-analysis are not formally presented in full (ie, with forest plots and tables for each subset), as their pooled accuracy is less certain.^{34,35} The aggregate estimates are, however, presented in tabular format for reader interest, with the associated due caution in the setting of notable heterogeneity.

Results

Study Characteristics

The 50 criteria-meeting studies relevant to the diagnostic yield of CT scans for temporal bone anomalies included a total of 5757 subjects.^{27,28,36–82} Forty-one studies were retrospective case series. The remaining studies included prospective case series,^{36,37,56,57} 1 prospective cohort study,⁵⁵ 1 case-control study,⁴⁸ 1 cross-sectional study,²⁷ 1 study with both prospectively and retrospectively recruited patients,⁸³ and 1 historical inception study.⁵⁸ Fourteen restricted their analyses to patients with severe to profound SNHL (Table 1). Eleven studies included only patients with bilateral hearing loss (Appendix S2, available at <http://otojournal.org>), and 7 studies included only patients with unilateral hearing loss

(Table 2). Twenty-seven studies did not specify or categorize the types of hearing loss of patients studied (Table 3).

Heterogeneity among studies was large ($I^2 = 90\%$; 95% CI, 89%–92%), such that interpretation of pooled data for the entire group of publications should be done with caution. For reader interest, however, the overall data are demonstrated in a forest plot (Figure 3), and the pooled diagnostic yield (random effects) is noted to be 30% (95% CI, 26%–34%). The heterogeneity among studies as calculated by I^2 remained substantial, even when stratified by study characteristics and severity, laterality, or type (conductive/mixed/sensorineural) of hearing loss (Table 4). In the setting of substantial heterogeneity, pooled data should be viewed with caution³⁴ but are presented for the overarching data set to help provide a visual summary of the body of relevant studies.

Severe to Profound Hearing Loss

Fourteen studies specifically evaluated the yield of diagnostic CT in pediatric patients with severe to profound SNHL (Table 1). The percentage of CT scans of patients with profound hearing loss that revealed new diagnoses of temporal bone anomalies ranged from 16%¹⁸ to 74%.⁴³ The 2 prospective studies found that 43% (19/44)³⁷ or 49% (33/67)³⁶ of patients had diagnostically valuable CT scans. Nine of 14 studies had consecutive patients, with the same range of diagnostic yield. Ten of 14 studies limited their patients to cochlear implant candidates or patients who had already received cochlear implants and reported the same range. Among the 4 remaining studies of patients with severe to profound SNHL of unknown etiology, the study with the largest sample size found that 18% (43/245) had diagnostically valuable CT scans.⁴² The 2 studies that reported the highest diagnostic yields were among those with the smallest sample sizes: 74% (25/34)⁴³ and 70% (7/10).⁴⁸ Overall, the most common diagnostic findings on CTs of patients with profound hearing loss were enlarged vestibular aqueduct (EVA) and cochlear dysplasia.

Bilateral Hearing Loss

Eleven studies evaluated CT findings in infants and children with bilateral HL (Appendix S2). Nine of these had patients with cochlear implants and are thus represented in the subsets of both Table 1 and Appendix S2. The percentage of CT scans of patients with bilateral hearing loss that revealed new diagnoses ranged from 10%⁸¹ to 74%.⁴³ Nine of 11 studies had consecutive patients with the same diagnostic range. There was a single prospective study, which reported a diagnostic yield of 49% (33/67).¹⁵ The most common findings associated with profound hearing loss were EVAs and cochlear dysplasias or malformations. The 2 reports not restricted to cochlear implant candidates^{81,82} included patients with the range of mild to profound hearing loss and demonstrated a 10% and 28% yield, respectively, of new diagnoses identified via CT scan.

Unilateral Hearing Loss

In 7 case series of patients with unilateral hearing loss, the primary outcome measure was the proportion of patients who received CTs that diagnosed new temporal bone anomalies (Table 2). Six retrospective studies evaluated consecutive patients; the seventh study⁸³ included both prospective and retrospective patients. The percentage yield ranged from

18%⁵³ to 67%.⁵⁰ Song et al⁴⁹ had the largest study population (n = 322) and reported a 29% diagnostic rate. Each of the remaining retrospective case series had n = 69 patients or fewer. Across all 7 studies, 45%⁸³ to 76%⁴⁹ of patients had profound hearing loss or worse. In these studies of unilateral hearing loss, the most common CT-established diagnoses included EVA, cochlear malformation, and atypical internal auditory canal (IAC).

Unspecified/Uncategorized/Range of Types of Hearing Loss

Twenty-seven studies either did not specify the range of hearing loss studied or studied a wide range of types of hearing loss (Table 3). Twenty-one studies reported results for consecutive patients. Three studies had nonconsecutive patients,^{27,63,74} and 3 studies did not report the consecutive status of patients.^{67,75,80} Among all studies, the percentage of newly diagnosed temporal bone anomalies ranged from 7%⁷⁵ to 64%.⁷⁹ One study followed a prospective cohort and found a 30% diagnostic yield with no significant difference in CT findings according to severity of SNHL.⁵⁵ Two smaller prospective case series with consecutive patients reported yields of 27%⁵⁶ and 22%,⁵⁷ and 1 historical inception cohort reported a yield of 26%.⁵⁸ One cross-sectional study of CTs in patients with SNHL compared to those with normal hearing found statistically significant differences in percentages of ears diagnosed with (1) narrow IAC, (2) EVA, and (3) cochleovestibular abnormalities.²⁷ Across all studies, the most common diagnoses were EVA and cochlear anomalies.

Prospective Data

Of the 50 studies examined, only 5 used a prospective study design. The largest was reported by Preciado et al⁵⁵ and described consecutive patients. In this prospective cohort study of children with a range of hearing loss, 30% of CT scans (45/150) provided new diagnostic information. Wu et al³⁶ reported that 49% of consecutive patients with cochlear implants (33/67) had newly identified anomalies. Declau et al⁵⁶ and Denoyelle et al⁵⁷ reported diagnostic yields of 27% (9/33) and 22% (7/32), respectively, in prospective consecutive series. Ma et al³⁷ described a prospective case series of children with profound hearing loss who had a 43% (19/44) diagnostic yield; the consecutive status of patients was not reported. Four of 5 prospective studies^{36,37,55,57} determined that EVA was the most common diagnostic entity, whereas 1 study⁵⁶ did not report the specific types of anomalies identified. Preciado et al⁵⁵ reported that there were no statistically significant differences in diagnostic yields of CT for patients of different levels and types of hearing loss.

Enlarged Vestibular Aqueduct

Enlarged vestibular aqueduct was the most common diagnosis in 25 out of 50 studies. Four studies reported solely EVA findings.^{28,60,62,78} The largest prospective study reported that 16% of pediatric patients with hearing loss of unknown etiology were diagnosed with EVA via CT scan.⁵⁵ Other studies showed that EVA accounted for as many as 75% of abnormal CT findings.⁵¹

The aggregate data were analyzed for heterogeneity, which was substantial even when this diagnosis alone was considered, $I^2 = 91\%$ (95% CI, 89%–93%) (Table 4). This heterogeneity remained high even when the EVA diagnosis alone was analyzed in the

subgroup defined by severe to profound hearing loss ($I^2 = 96\%$; 95% CI, 94%–97%). When EVA was evaluated in the subset of unilateral hearing loss, heterogeneity was less ($I^2 = 67\%$; 95% CI, 27%–85%), which was the lowest among all of the study subgroups but still notable. Among this subset, the pooled data reflected a diagnostic yield of 11% (95% CI, 7%–17%).

A sensitivity analysis for the EVA subset was performed, as 1 study evaluated 2 different criteria for EVA diagnosis/measurement in the same patient population.²⁸ Regardless of whether the Cincinnati or Valvassori criteria were used for that study in the aggregate analysis, the composite data showed similar results.

Narrow Cochlear Nerve Canals/Internal Auditory Canals

Diagnoses of narrow, stenotic, small, or absent cochlear nerve canals (CNC) or IACs were described in 31 out of 50 studies. Overall, the reported diagnostic yield of CT scan for narrow CNC or IAC in pediatric patients ranged from 0% to 54%.⁵⁰ The largest studies showed a 4% to 7% prevalence of this finding.^{38,49} Heterogeneity among these studies was high, regardless of whether subsets of severe to profound or unilateral hearing loss were considered (Table 4). For reader interest, it is noted that the pooled data for the narrow CNC subset was 4% (3%–7%), although again these aggregate data should be viewed with some caution.

Discussion

The data from this systematic review demonstrate a wide range of diagnostic yields in temporal bone CTs (7%⁷⁵ to 74%⁴³) obtained for pediatric hearing loss. The largest prospective study and aggregate data show a 30% yield for all diagnoses combined.⁵⁵ This yield suggests that in order to obtain 1 new diagnostic result, 4 patients need to undergo CT⁵⁵ (range, 2–15).^{43,75}

Certain diagnoses may alter the management strategy for the presenting patient and were therefore considered in more detail in our analysis. More specific, although controversial, some practitioners may instruct families of children with EVA to avoid contact sports or other activities with an inherent risk of head trauma.⁸⁴ In addition, other providers use the finding of EVA to prompt testing for mutations in the PDS or EYA genes.^{85,86} The strongest data suggested that the diagnostic yield for EVA was 16%. The subset with the least heterogeneity occurred in the circumstance when the diagnosis of EVA was considered exclusively in studies of unilateral hearing loss.

The finding of a narrowed or absent cochlear nerve canal may also affect counseling if cochlear implantation is being considered, as an absent nerve in particular may prompt consideration of alternate interventions such as auditory brainstem implant. A narrowed cochlear nerve canal may additionally suggest that future additional hearing deterioration will be limited,⁸⁷ providing valuable prognostic information for the family. The largest studies showed a 4% to 7% diagnostic yield for a narrow CNC, and again, these data had substantial heterogeneity.

When considered in aggregate, these CT data did not suggest that discrete patterns of hearing loss had statistically significant differences in diagnostic yield, although it was suggested in some individual study results. Regardless of whether hearing loss was severe, bilateral, unilateral, sufficient to warrant cochlear implant candidacy, or unspecified, the range of diagnostic yield was wide and heterogeneity was high even within subgroups, making it difficult to draw conclusions about differential evaluation of specific types of hearing loss. Even when studies were limited to subgroups with objectively defined diagnostic criteria, heterogeneity within the group remained high (Appendix S3, available at <http://otojournal.org>). Prospective studies had the tightest range of diagnostic yield (22%–49%, in comparison with 7%–74% in other studies), which suggests that further prospective analysis with a priori, clearly defined diagnostic criteria may yield more specific data that could guide evaluations specific to the severity, laterality, and type of hearing loss.

The wide range of observed estimates of diagnostic yield for CT scans may be partially attributable to differences in diagnostic criteria used in each study to determine what constitutes specific anomalies. For example, Dewan et al²⁸ demonstrated that application of the Valvassori criteria (> 1.5 mm at the midpoint) versus the Cincinnati criteria (midpoint or opercular width greater than the 95th percentile) resulted in an approximately 30% difference in rates of EVA diagnosis. In addition, other authors reported using still different criteria for the same diagnosis (eg, > 2 mm),²⁷ whereas others did not report on their defined criteria at all. Differences in CT equipment and protocols may have also contributed to variability in the reported yields⁸⁸; included studies used scans of different slice thicknesses (0.625-mm slices⁷⁶ to 1-mm slices⁶⁹) and a variety of levels of radiation (eg, 1 study estimated a mean dose of 29 mGy⁵⁴ whereas another estimated doses ranging from 35.55 to 44.44 mGy⁷⁶).

Regardless, understanding the associated numbers needed to image (ie, number of patients who need to undergo CT in order to yield 1 diagnosis) provides information that may be weighed against the associated numbers needed to harm (ie, the number of patients who need to undergo CT in order for 1 malignancy or other adverse effect to develop), and this latter concept is addressed in a separate systematic review.⁸⁹ The most rigorous data from that sister study show that if every excess brain cancer after brain CT were attributable only to the imaging itself, then approximately 1 in 4000 pediatric brain CTs would be followed by a malignancy (mean estimated radiation dose 40 mSv per scan) or 1 brain tumor per 10,000 patients (10 mGy per scan, < 10 years of age at exposure).^{90,91} As temporal bone CT confers a smaller radiation dose than brain CT, the associated risks of radiation are anticipated to be even lower, although direct patient data from temporal bone imaging are very limited. Overall, these data suggest that CT for pediatric hearing loss is more than 1000 times more likely to yield a diagnosis than have a subsequent malignancy. The CT-specific data in the present systematic review also provide a context in which to consider the diagnostic yield of other imaging evaluation options for pediatric hearing loss (ie, MRI). All of these data together form the basis for decisions analysis on a population level^{92,93} as well as for shared, informed decision making^{94–96} with families in circumstances where the risks and benefits of CT are weighed together.

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Figure 1.
Flow diagram showing the stages of identification of studies.

Figure 2.
Flow diagram showing the stages of identification of studies by citation source.

Figure 3.

Forest plot of the diagnostic yield for all studies, all diagnostic findings. The wide range of data is demonstrated. The pooled proportion yield should be interpreted with caution due to the substantial heterogeneity among studies. This pooled estimate is thus presented for interest but not as a meaningful single estimate of the effects of the studies, given the heterogeneity observed (Table 4). The studies are presented in an order that parallels the order in the tables presenting the remainder of the results (prospective precedes retrospective; severe to profound and bilateral precedes unilateral and unspecified).

Diagnostic Yield of CT Scan in Children with Severe to Profound Hearing Loss of Unknown Etiology.^a

Table 1

First Author, Year	Study Design	Percentage (Proportion) with New Diagnoses	Types of Anomalies Identified, Percentage of All Anomalies (Proportion)	Age Group	Severity of Hearing Loss	Additional Comments
Prospective studies						
Wu, 2008 ³⁶	Prospective case series with chart review of patients with cochlear implants	49% of patients (33/67)	EVA, 58% (19/33) SCC Dysplasia, 30% (10/33) Aplasia, 3% (1/33) Vestibule Enlargement, 18% (6/33) Hypoplasia, 12% (4/33) Aplasia, cochlea, 0% (0/33) Cochlea Incomplete partition, 33% (11/33) Common cavity, 9% (3/33) Hypoplasia, 9% (3/33) Aplasia, 3% (1/33)	Ages 1–14 years (mean 4.7 years) at implantation	Cochlear implant patients	Consecutive
Ma, 2008 ³⁷	Prospective case series with chart review of patients with SNHL	43% of patients (19/44)	36 malformations in 36 ears Michel malformation, 3% (1/36) Common cavity, 8% (3/36) IP-I, 8% (3/36) IP-II, 14% (5/36) Vestibular/SCC malformation, 36% (14/36) EVA, 44% (16/36) IAC malformation, 22% (8/36)	3–19 years (mean 11 years)	Profound SNHL (mean response threshold 88 dB HL)	Consecutive status of patients NR
Retrospective studies						
Papsin, 2005 ³⁸	Retrospective case series with chart review of cochlear implant recipients	35% of patients had cochleovestibular anomalies (103/298)	Incomplete partition, 14% (42/298) EVA, 12% (37/298) Posterior labyrinth anomaly, 9% (26/298) IAC/cochlear canal anomaly, 4% (11/298) Hypoplastic cochlea, 5% (16/298) Common cavity deformity, 3% (8/298)	Mean age 5.3 years	Cochlear implant patients	Consecutive
Drvis, 2008 ³⁹	Retrospective case series with chart review of	16% (44/270)	Inner ear malformation, 100% (44/44) EVA, 41% (18/44)	5 months–14 years (mean 3.9 years)	Cochlear implant patients	Consecutive

First Author, Year	Study Design	Percentage (Proportion) with New Diagnoses	Types of Anomalies Identified, Percentage of All Anomalies (Proportion)	Age Group	Severity of Hearing Loss	Additional Comments
	cochlear implant candidates cochlear implant candidates		Vestibulocochlear dysplasia, 27% (12/44) Mondini malformation, 23% (10/44) Ossified cochlea, 9% (4/44)			
Lin, 2011 ⁴²	Retrospective case series with chart review of patients with severe to profound SNHL	18% of patients (43/245)	Total: Cochlear dysplasia, 58% (25/43) Vestibule/SCC dysplasia, 58% (25/43) IAC/cochlear aperture anomaly, 42% (18/43) EVA, 30% (13/43) Isolated: IAC/cochlear aperture anomaly, 28% (12/43) EVA, 16% (7/43) Cochlear dysplasia, 12% (5/43) Vestibule/SCC dysplasia, 9% (4/43)	Children-specific ages NR	Severe to profound HL	Consecutive
Trimble, 2007 ⁴⁰	Retrospective case series with chart review of cochlear implant candidates	59% of patients (54/92)	EVA, 48% (26/54) Cochlear dysplasia, 24% (13/54) Narrow CNC, 15% (8/54) Small bony island of lateral SCC, 7% (4/54) Modiolar deficiency, 6% (3/54) Labyrinthine ossification, 4% (2/54)	7 months–17 years (mean 4.7 years)	Cochlear implant candidates	Consecutive; more than 1 anomaly per patient was noted in some cases
Kong, 2009 ⁴¹	Retrospective case series with chart review of cochlear implant candidates	16% of patients (inner ear malformation) (11/68) 3% of patients (narrow IAC) (2/68)	Not specified whether the 11 inner ear malformations and 2 narrow IACs occurred in overlapping patients	1–15 years old (mean 5.4 years)	Cochlear implant candidates	Consecutive; follow-up time > 6 months
Seicshnaydre, 1992 ⁴³	Retrospective case series with chart review of cochlear implant recipients	74% of patients (25/34)	Narrowed basal turn, 32% (8/25) ^b Bony lip at round window, 32% (8/25) Ossified cochlea, 16% (4/25) Widened cochlear aqueduct, 12% (3/25) Bulbous IAC, 4% (1/25) Right Mondini, left aplasia, 4% (1/25)	2.5–15 years	Cochlear implant patients	Consecutive

First Author, Year	Study Design	Percentage (Proportion) with New Diagnoses	Types of Anomalies Identified, Percentage of All Anomalies (Proportion)	Age Group	Severity of Hearing Loss	Additional Comments
Bath, 1993 ⁴⁴	Retrospective case series with chart review of cochlear implant recipients	42% of patients (11/26)	Partially ossified cochlea, 42% (11/26) Patent cochlea, 58% (15/26) Only the cochlea was examined.	2.4–11 years (mean 5.3 years) at operation	Cochlear implant patients	Consecutive
Dewan, 2009 ²⁸	Retrospective case series with chart review of cochlear implant recipients	Cincinnati criteria: 57% (64/112) Valvassori criteria: 25% (28/112)	The focus of this study was to evaluate 2 separate criteria to diagnose EVA. Other CT-identified anomalies were NR. EVA 57% (64/112), Cincinnati criteria 25% (28/112), Valvassori criteria	Mean age of 5.2 years (SD = 4.4 years)	Cochlear implant patients	Consecutive
Nikolopoulos, 1997 ⁴⁵	Retrospective case series with chart review of cochlear implant recipients	19% of patients (21/108)	At least partial obliteration of cochlea, 86% (18/21) Congenital malformation of cochlea, 10% (2/21) Stenotic IAC, 5% (1/21)	21 months–16 years (mean 5.4 years)	Cochlear implant patients	Consecutive status of patients NR
Van Wermeskerken, 2007 ⁴⁶	Retrospective case series with chart review of congenitally deaf patients with cochlear implants	18% of patients (9/51)	EVA, 55% (5/9) IP-I, 11% (1/9) IP-II, 55% (5/9) SCC dysplasia, 33% (3/9) Wide IAC, 22% (2/9)	Those with abnormal findings; 2–6 years old at implantation (mean 3.9, SD 1.5)	Cochlear implant patients	Consecutive status NR; follow-up of 12–48 months
Komatsubara, 2007 ⁴⁷	Retrospective case series of patients with congenital hearing loss	60% of patients (9/15) had cochlear nerve deficiency	Only cochlear nerve deficiencies reported.	6 months–13 years (mean 5.4 years)	Severe SNHL	Consecutive status of patients NR
Kochhar, 2009 ⁴⁸	Case-control study comparing patients with HL of DFNB1 and non-DFNB1 etiology	70% of patients (7/10) with non-DFNB1 SNHL	Site of anomaly: Cochlear basal turn lumen, 43% (3/7) Vestibule width, 57% (4/7) Lateral SCC island width, 57% (4/7) Vestibular aqueduct width, 14% (1/7) Coronal cochlear height, 14% (1/7)	Mean age at scan: 41.2 months (range 9–156 months)	Severe to profound HL	Consecutive status of patients NR

Abbreviations: CNC, cochlear nerve canal; CT, computed tomography; EVA, enlarged vestibular aqueduct; HL, hearing loss; IAC, internal auditory canal; IP-I/IP-II, incomplete partition type 1 or 2; NR, not reported; SCC, semicircular canal; SD, standard deviation; SNHL, sensorineural hearing loss.

^aIndividual anomalies may overlap within patients or may not have been completely reported, so percentage numbers do not always sum to 100%.

^b Four CT scans had 2 separate findings each.

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Diagnostic Yield of CT Scan in Children with Unilateral Hearing Loss of Unknown Etiology.

First Author, Year	Study Design	Percentage (Proportion) with New Diagnoses	Types of Anomalies Identified, Percentage of All Anomalies (Proportion)	Age Group	Extent of Hearing Loss	Additional Comments
Song, 2009 ⁵⁰	Retrospective case series with chart review	29% of patients (93/322)	Cochleovestibular malformations, 53% (49/93) IP-II, 30% (28/93, 20 combined with EVAs) IP-I, 11% (10/93) Common cavity, 6% (6/93) Cochlear aplasia, 2% (2/93) Cochlear hypoplasia, 2% (2/93) Complete labyrinthine aplasia, 1% (1/93) Vestibular malformations, 29% (27/93) Malformed IAC, 25% (23/93) Malformed SCC, 4% (4/93) Malformations of vestibular or cochlear aqueducts, 18% (17/93) EVA, 18% (17/93)	6 months–15 years (mean 7.9 years)	Mild to severe, 24% (78/322); profound, 76% (244/322)	Consecutive patients; follow-up time 6 months–7 years (mean 30 months)
Masuda, 2013 ⁵⁰	Retrospective case series with chart review of patients with unilateral SNHL	67% of patients (46/69)	Cochlear nerve canal stenosis, 70% (32/46) Associated malformations, 59% (19/32) IAC malformation, 48% (22/46) Narrow, 43% (20/46) Enlarged, 2% (1/46) Absent, 2% (1/46) Cochlear malformation, 30% (14/46) Cochlear aplasia, 0% (0/46) Common cavity deformity, 4% (2/46) Cochlear hypoplasia, 2% (1/46) Incomplete partition, 24% (11/46) Vestibular/SCC malformation, 11% (5/46) Bilateral EVA, 4% (2/46)	0–15 years (mean 4.3 years)	Mild HL, 9% (6/69); moderate HL, 19% (13/69); severe HL, 10% (7/69); profound HL, 62% (43/69)	Consecutive
Haffey, 2013 ⁵¹	Retrospective case series with chart review of patients with unilateral SNHL	32% of patients (20/61)	EVA, 75% (15/20) Mondini, 40% (8/20) Mastoiditis/COM, 25% (5/20) SCC dehiscence, 15% (3/20) High jugular bulb, 5% (1/20) Cholesteatoma, 5% (1/20) Bony deformation of incus, 5% (1/20)	0–17 years (mean 5.6 years)	Type of HL: low frequency, 1% (1/79); mid-frequency, 22% (17/79); high frequency, 37% (29/79); flat, 41% (32/79)	Consecutive; follow-up time of 5 years
Brookhouser, 1991 ⁵²	Retrospective case series with chart review of patients with unilateral SNHL	18% of patients (10/57)	EVA, 30% (3/10) Cochlea and SCC malformation, 20% (2/10) Widening and shortening of IAC, 30% (3/10) Fractures of temporal bone, 20% (2/10)	19 years	Of the 10 abnormal CTs: borderline, 20% (2/10); moderate, 10% (1/10); severe, 20% (2/10); anacusis, 50% (5/10)	Consecutive; follow-up data available for periods of 1–15 years for 105 patients
Bamiou, 1999 ⁵³	Retrospective case series with chart review of patients with unilateral SNHL	31% (11/35)	Unilateral EVA, 18% (2/11) Bilateral EVA, 18% (2/11) Cochlear hypoplasia, 18% (2/11) Narrow IAC, 9% (1/11) Labyrinthitis ossificans, 27% (3/11)	Children: mean age of 11.1 (SD 3.6)	Mild, 6% (2/35); moderate, 17% (6/35); severe, 14% (5/35); profound, 63% (22/35)	Consecutive

First Author, Year	Study Design	Percentage (Proportion) with New Diagnoses	Types of Anomalies Identified, Percentage of All Anomalies (Proportion)	Age Group	Extent of Hearing Loss	Additional Comments
Cama, 2012 ⁵⁴	Retrospective case series with chart review	64% of patients (14/22)	Enlarged lateral SCC, 9% (1/11) EVA, 29% (4/14) Common cavity, 7% (1/14) Cochleovestibular hypoplasia, 7% (1/14) Hypoplasia of handle of malleus, 7% (1/14) Labyrinthine ossification, 14% (2/14) High jugular bulb dehiscence with the vestibular aqueduct, 36% (5/14) Narrow IAC, 7% (1/14)	Birth-8.5 years (mean 4.6 years)	Profound HL, 73% (16/22)	Consecutive patients; follow-up time 1–5 years
Neary, 2003 ⁸³	Case series with chart review: 37 patients recruited retrospectively, 19 recruited prospectively, 1 excluded; 39 had CT scans	28% of patients (11/39)	EVA, 5% (2/39) Various abnormalities of external auditory canals, middle ear structures, and SCC, 10% (4/39) Aplasia of the cochlea + dysplasia of SCC + small IAC, 3% (1/39) Narrow IAC, 3% (1/39) Occlusion of central neural foramen + small IAC, 3% (1/39) EVA + severe dysplasia of SCC, 3% (1/39) Small middle ear + abnormal ossicles + dysplastic SCC, 3% (1/39)	Diagnosis 3–13 years (mean 6 years)	For the entire study: mild, 9% (5/55); moderate, 32% (18/55); severe, 13% (7/55); profound, 38% (21/55); dead ear, 7% (4/55)	Consecutive status of retrospective case series NR; prospective children recruited consecutively

Abbreviations: COM, chronic otitis media; CT, computed tomography; EVA, enlarged vestibular aqueduct; HL, hearing loss; IAC, internal auditory canal; IP-I/IP-II, incomplete partition type 1 or 2; NR, not reported; SCC, semicircular canal; SD, standard deviation; SNHL, sensorineural hearing loss.

Table 3
Diagnostic Yield of CT Scan in Children with Unspecified, Uncategorized, or a Range of Hearing Loss of Unknown Etiology.

First Author, Year	Study Design	Percentage (Proportion) with New Diagnoses	Types of Anomalies Identified, Percentage of All Anomalies (Proportion)	Age Group	Extent of Hearing Loss	Additional Comments
Prospective and cross-sectional studies						
Preciado, 2005 ⁵⁵	Prospective cohort study of patients with SNHL	30% of patients (45/150)	EVA, 53% (24/45) Cochlear dysplasia, 13% (6/45) Cochlear hypoplasia, 4% (2/45) Multiple abnormalities, 29% (13/45) Diagnostic yield for ^a : Patients with bilateral severe-to-profound loss, 27.8% (10/36) Patients with bilateral moderately severe loss, 37.9% (11/29) Patients with bilateral mild-to-moderate loss, 23.4% (15/64) Patients with unilateral SNHL, 42.8% (9/21)	1 week–18 years (mean 4.8, SD 4.8)	Bilateral severe to profound, 24% (36/150); bilateral moderately severe, 19% (29/150); bilateral mild to moderate, 43% (64/150) Unilateral SNHL, 14% (21/150)	Consecutive
Declau, 2008 ⁵⁶	Prospective case series of patients with SNHL	27% of patients (9/33)	NR	36–86 days (median 50 days)	Bilateral HL, 59% (68/116) Unilateral HL, 41% (48/116) Median hearing threshold was severe HL	Consecutive
Denoyelle, 1999 ⁵⁷	Prospective case series of patients with SNHL	22% of non-DFNB patients (7/32)	Bilateral EVA, 57% (4/7) Cochleovestibular dilation or I AC dilation, 29% (2/7)	4–20 years (median 7 years)	Mild, 11% (6/57); moderate, 19% (11/57); severe, 26% (15/57);	Consecutive

First Author, Year	Study Design	Percentage (Proportion) with New Diagnoses			Types of Anomalies Identified, Percentage of All Anomalies (Proportion)	Age Group	Extent of Hearing Loss	Additional Comments
McClay, 2002 ²⁷	Cross-sectional study of random sample of temporal bone CTs obtained in patients with versus without HL	Anomaly	Ears with SNHL	Ears without SNHL	Pericochlear osteodystrophy, 14% (1/7)	2 months–5 years	profound, 44% (25/57)	Nonconsecutive comparison group: children with no SNHL
		Narrow IAC ^b	4% (8/185)	1% (4/309)	Pericochlear osteodystrophy, 14% (1/7)			
		EVA (> 2 mm) ^b	5% (9/185)	0% (0/309)	Pericochlear osteodystrophy, 14% (1/7)			
		Cochleovestibular ^b	17% (32/185)	0% (0/309)	Pericochlear osteodystrophy, 14% (1/7)			
		Wide IAC	0.5% (1/185)	4% (11/309)				
Retrospective studies	Historical inception cohort	Bulbous IAC	9% (16/185)	8% (24/309)				Among 113 children with hearing loss: bilateral SNHL, 64% (72/113); unilateral SNHL, 36% (41/113)
Ghogomu, 2014 ⁵⁸								Consecutive
		31% of patients (30/98)						
Preciado, 2004 ⁵⁹	Retrospective case series with chart review of patients with SNHL							Consecutive
		29% of patients (149/511, 50 had CT and MRI; CT only: 31% [143/461])						

First Author, Year	Study Design	Percentage (Proportion) with New Diagnoses	Types of Anomalies Identified, Percentage of All Anomalies (Proportion)	Age Group	Extent of Hearing Loss	Additional Comments
Lee, 2009 ⁶⁰	Retrospective case series with chart review of patients with SNHL who underwent GJB2 testing	NR for all anomalies 26% of patients had EVA (108/412)	Small IAC, 3% (5/149) Small IAC, 3% (5/149) Cochlear hypoplasia, 3% (4/149) Multiple abnormalities, 9% (13/149)	"Children" ages NR	mild to moderate, 39% (253/650); high frequency SNHL, 6% (39/650)	Consecutive
Chan, 2011 ⁶¹	Retrospective case series with chart review of patients with congenital SNHL	14% of patients (32/225)	NR	Mean 5.8 years	Unilateral and bilateral mild to severe HL	Consecutive
Arijmand, 2004 ⁶²	Retrospective case series with chart review of patients with SNHL	9% of patients had EVA (19/221) (only EVA documented)	Isolated EVA, 79% (26/33) EVA with cochlear anomalies, 3% (1/33) EVA with SCC anomalies, 6% (2/33) EVA with cochleovestibular anomalies, 3% (1/33) EVA with SCC and vestibular anomalies, 9% (3/33)	1 month–17.2 years (mean 5.5 years)	NR	Consecutive
Wiley, 2011 ⁶³	Retrospective case series with chart review of patients with permanent HL	46% of patients (67/161)	EVA, 30% (20/67) Cochlear dysplasia, 18% (12/67) Hypoplastic cochlea, 9% (6/67) Deficient modiolis, 7% (5/67) EVA and cochlear dysplasia, 6% (4/67) Cochlear partitioning defect, 4% (3/67) Mondini, 3% (2/67)	1 month–19.7 years (median 69.7 months)	SNHL, 86% (171/198) Mixed HL, 8% (17/198) Conductive HL, 4% (7/198) Auditory neuropathy, 2% (3/198)	Nonconsecutive

First Author, Year	Study Design	Percentage (Proportion) with New Diagnoses	Types of Anomalies Identified, Percentage of All Anomalies (Proportion)	Age Group	Extent of Hearing Loss	Additional Comments
Wu, 2005 ⁶⁴	Retrospective case series with chart review of patients with SNHL	37% of patients (59/160)	EVA and hypoplastic cochlea, 3% (2/67)	1–18 years (mean 5.3 years)	NR	Consecutive; minimum follow-up period of 6 months (mean 3.4 years)
			EVA and hypoplastic cochlea, 3% (2/67)			
Antonelli, 1999 ⁶⁵	Retrospective case series of patients with SNHL or mixed HL	31% of patients (49/157)	EVA and hypoplastic cochlea, 3% (2/67)	0–18 years (mean 7.2 years)	NR	Consecutive
			EVA and hypoplastic cochlea, 3% (2/67)			
			EVA and hypoplastic cochlea, 3% (2/67)			
			Other, 13% (9/67)			
			Unknown, 3% (2/67)			
			Brain finding, 3% (2/67)			
			Bilateral anomaly, 93% (55/59)			
			Unilateral anomaly, 7% (4/59)			
			Individual diagnoses described relative to the total number of ears (n = 114): EVA, 58% (66/114)			
			SCC dysplasia, 16% (30/114)			
			Vestibule Enlargement, 42% (48/114)			
			Hypoplasia, 4% (5/114)			
			Aplasia, 2% (2/114)			
			Cochlea Incomplete partition, 49% (56/114)			
			Common cavity, 7% (8/114)			
			Hypoplasia, 4% (4/114)			
			Aplasia, 2% (2/114)			
			EVA, 53% (26/49, 21 bilateral, 5 unilateral)			
			Anomalies in: Labyrinth, 29% (14/49)			
			Cochlea, 43% (21/49)			

First Author, Year	Study Design	Percentage (Proportion) with New Diagnoses	Types of Anomalies Identified, Percentage of All Anomalies (Proportion)	Age Group	Extent of Hearing Loss	Additional Comments
Billings, 1999 ⁶⁶	Retrospective case series with chart review of patients with SNHL	15% of patients (23/156)	Modiolus, 41% (20/49) Modiolus, 41% (20/49) Oval window, 6% (3/49) Round window, 14% (7/49) IAC, 4% (2/49)	1 month–13 years at diagnosis (mean 3.52 years)	Total population (n = 301); bilateral mild-moderate, 10% (30/301); bilateral severe-profound, 70% (211/100); profound, 67% (67/100); unilateral severe-profound, 20% (60/301)	Consecutive
			Abnormalities included EVA, 35% (8/23) Vestibulocochlear dysplasia, 30% (7/23) Mondini malformation, 17% (4/23)			

First Author, Year	Study Design	Percentage (Proportion) with New Diagnoses	Types of Anomalies Identified, Percentage of All Anomalies (Proportion)	Age Group	Extent of Hearing Loss	Additional Comments
Arcand, 1991 ⁶⁷	Retrospective case series with chart review of patients with SNHL and mixed HL	25% of patients (33/130)	EVA, 55% (18/33) (+ other ear abnormalities) Bilateral vestibular dilation, 3% (1/33) Right Mondini dysplasia and left hypoplastic SCC, 3% (1/33) Severe cochlear hypoplasia and vestibular dilation, 3% (1/33) Severe cochlear and SCC hypoplasia with vestibular dilation, 3% (1/33) Bilateral cochlear hypoplasia, 3% (1/33)	Mean age of 6 years (3.1 years was age of EVA patients)	Among EVAs: sequential audiograms available for 13 cases—6 progressive HL (5 bilateral, 1 unilateral) with mean HL of 50 dB, average progression of 30 dB of loss 7 stable HL, with mean HL of 60dB	Consecutive status of patients NR; mean follow-up period of EVA cases: 4 years
Simons, 2006 ⁶⁹	Retrospective case series with chart review of patients with unilateral or bilateral asymmetric HL	41 % of patients (50/123)	Unilateral SNHL EVA, 30% (15/50) EVA+ IEA, 4%(2/50) IEA, 12% (6/50) Small IAC, 4%(2/50) Asymmetric SNHL EVA, 26% (13/50) EVA+ IEA, 10% (5/50) IEA, 4%(2/50) Small IAC, 2% (1/50)	0–17 years (mean 5.2 years)	NR	Consecutive
Adachi, 2010 ⁶⁸	Retrospective case series with chart review	28% (34/121)	Inner ear/IAC anomaly, 17% (20/121) Middle/external ear anomaly, 12% (14/121)	Infants (age at first visit: 5 days–8 months, mean 19 days)	ABR > 50 dB bilaterally (CTs done in “habilitation” group)	Consecutive; HL identified by newborn screening
Mafong, 2002 ⁷⁰	Retrospective case series with chart review of patients with SNHL	37% of patients (33/90)	Isolated, 55% (18/33) EVA, 21% (7/33)	1–18 years (mean 9 years)	Bilateral SNHL, 83% of total	Consecutive

First Author, Year	Study Design	Percentage (Proportion) with New Diagnoses	Types of Anomalies Identified, Percentage of All Anomalies (Proportion)	Age Group	Extent of Hearing Loss	Additional Comments
Cross, 1999 ⁷¹	Retrospective case series with chart review of congenital SNHL	11% of patients (8/71)	Lateral SCC dysplasia, 15% (5/33)	13-20 years	Hearing impairment of greater than 50 dB	patients in study (95/114) (including those without CT scan data) Unilateral SNHL, 11% (13/114) Moderate to profound HL, 81% (92/114) Mild HL, 19% (22/114)
			Lateral SCC dysplasia, 15% (5/33)			
			Lateral SCC dysplasia, 15% (5/33)			
			Cochlear dysplasia, 9% (3/33)			
			Otic capsular lucency, 3% (1/33)			
			Small IAC, 3% (1/33)			
			Hypoplastic cochlea, 3% (1/33)			
			Multiple inner ear malformation, 27% (9/33)			
			Cochlear dysplasia, 21 % (7/33)			
			EVA, 18% (6/33)			
Shusterman, 1992 ⁷²	Retrospective case series of SNHL	13% of patients (9/70)	Lateral SCC dysplasia, 18% (6/33)	1.0-20.9 years (mean 6.8 years)	Bilateral, 66% (6/9) Unilateral, 33% (3/9) Profound or severe, 22% (2/9) Moderate, 22% (2/9)	Consecutive
			Abnormalities not related to HL, 18% (6/33)			
			Bilateral EVA, 50% (4/8)			
			Unilateral EVA, 13% (1/8)			
			Bilateral Mondini abnormality, 38% (3/8)			
			Bilateral dilation of SCC, 13% (1/8)			
			Unilateral dysplasia of middle and external ear, 13% (1/8)			
			Bilateral congenital anomaly of SCC, 11% (1/9)			
			Narrow external auditory canals, 11% (1/9)			
			Congenital anomaly of cochlea and ossicles, 11% (1/9)			

First Author, Year	Study Design	Percentage (Proportion) with New Diagnoses	Types of Anomalies Identified, Percentage of All Anomalies (Proportion)	Age Group	Extent of Hearing Loss	Additional Comments
			Asymmetry in posterior wall of IAC, 11% (1/9) Asymmetry in posterior wall of IAC, 11% (1/9) Asymmetry in posterior wall of IAC, 11% (1/9) Enlarged/dilated endolymphatic duct, 33% (3/9) Fewer turns in cochlea, 11% (1/9) Dilated cochlear aqueduct, 11% (1/9)		Mixed, 11% (1/9) Low frequency, 11% (1/9)	
Coticchia, 2006 ⁷³	Retrospective case series with chart review of patients with SNHL	25% of patients (17/69)	EVA, 47% (8/17) Membranous defects, 24% (4/17) Miscellaneous, 29% (5/17)	NR "children"	NR for study population	Consecutive
Huo, 2012 ⁷⁴	Retrospective case series of patients with SNHL	31% of patients (20/65)	33 ears (of 130 ears) in 20 patients EVA, 45% (15/33) Cochlear malformations, 30% (10/33) IAC malformation, 24% (8/33) Vestibular malformations, 21% (7/33)	1–14 years (mean 3.78 years)	NR	Nonconsecutive; multislice spiral CT used
Zalzal, 1986 ⁷⁵	Retrospective case series with chart review of SNHL	7% of patients (3/44)	SCC malformation, 15% (5/33) 1 brainstem mass 1 asymmetric IAC 1 middle ear fluid (exploratory tympanotomy revealed perilymph fistula)	9 months–12 years	NR	Consecutive status of patients NR
Miyasaka, 2010 ⁷⁶	Retrospective case series with chart review of unilateral or bilateral SNHL	29% of patients (6/21) 23.8% of ears (10/42)	Michel deformity, 17% (1/6) Cochlear aplasia, 17% (1/6) Incomplete partition type 1, 17% (1/6) EVA, 33% (2/6) Duplication of IAC, 17% (1/6)	1–13 years (mean 7 years)	NR	Consecutive

First Author, Year	Study Design	Percentage (Proportion) with New Diagnoses	Types of Anomalies Identified, Percentage of All Anomalies (Proportion)	Age Group	Extent of Hearing Loss	Additional Comments
Sudden, mixed, or conductive hearing loss Tarshish, 2013 ⁷⁷	Retrospective case series with chart review of patients with sudden SNHL	13% of patients (2/14)	Absent CNC, 50% (3/6) Absent CNC, 50% (3/6) Closed CNC, no cochlear dysplasia, 17% (1/6)	0.25–18 years at onset of sudden SNHL	Mild/moderate, 21% (3/14) Severe, 64% (9/14) Profound, 14% (2/14)	Consecutive
			EVA, 50% (1/2) Soft tissue density, 50% (1/2)			
Yaeger, 2006 ⁷⁸	Retrospective case series with chart review	13% of patients with nonsyndromic HL had EVA (25/191)	Only EVA reported	“Children” Ages NR	Bilateral HL, 76% (381/500) Unilateral HL, 24% (119/500) SNHL, 99% (495/500) Conductive, 1% (5/500) A range of severity of HL (mild to profound)	Consecutive
Whittemore, 2012 ⁷⁹	Retrospective case series with chart review of patients with persistent conductive HL after tube placement	64% of patients (16/25)	Middle ear abnormalities, 25% (4/16) Inner ear abnormalities, 25% (4/16) Lesions resulting in third window effect, 31% (5/16) Cholesteatoma, 6% (1/16) Abnormalities leading to diagnosis of CHARGE syndrome, 13% (2/16)	Age at tympanostomy tube placement: 9 months–16 years (mean 5.92 years)	Bilateral HL, 56% (22/39) Unilateral HL, 44% (17/39)	Consecutive

First Author, Year	Study Design	Percentage (Proportion) with New Diagnoses	Types of Anomalies Identified, Percentage of All Anomalies (Proportion)	Age Group	Extent of Hearing Loss	Additional Comments
Ohlms, 1999 ⁸⁰	Retrospective case series with chart review of patients with SNHL or mixed HL	25% of patients (14/56)	Bilateral cochlear agenesis, 14% (2/14) Mondini malformation, 7% (1/14) EVA, 7% (1/14) Additional details NR	0–10 years at diagnosis (mean 2 years)	84% of patients with severe or profound HL (96/114) including those without CT data)	Consecutive status of patients NR

Abbreviations: ABR, auditory brainstem response; CNC, cochlear nerve canal; CT, computed tomography; EVA, enlarged vestibular aqueduct; HL, hearing loss; IAC, internal auditory canal; IEA, inner ear abnormality; MRI, magnetic resonance imaging; NR, not reported; SCC, semicircular canal; SD, standard deviation; SNHL, sensorineural hearing loss.

^aThere are no statistically significant differences.
^bThere is a statistically significant difference between SNHL and non-SNHL groups.

Table 4
Heterogeneity and Aggregate Results among Studies of Children with Hearing Loss, Proportion with Diagnostic Yield.

Studies Included	Imaging Findings Included	Hearing Loss Characteristic	No. Studies in Group/Subgroup	I ² (95% CI)	Diagnostic Yield (95% CI) ^a
All studies	All findings	All	n = 49 ^b	90% (89–92%)	30% (26%–34%)
All studies	All findings	Severe to profound hearing loss	n = 14	93% (91–95%)	39% (29%–40%)
All studies	All findings	Bilateral hearing loss	n = 11	94% (91–96%)	36% (25%–47%)
All studies	All findings	Unilateral hearing loss	n = 7	88% (78–94%)	38% (25%–51%)
All studies	All findings	Unspecified/range of hearing loss	n = 26 ^b	88% (83–91%)	24% (20%–29%)
All studies	All findings	No mixed or conductive hearing loss	n = 46	89% (86–91%)	29% (25%–33%)
Prospective studies	All findings	All	n = 5	66% (12–87%)	35% (26%–44%)
Studies with consecutive patient status specified	All findings	All	n = 35	91% (89–93%)	30% (25%–34%)
All studies	Enlarged vestibular aqueduct	All	n = 40	91% (89–93%)	11% (8%–15%)
All studies	Enlarged vestibular aqueduct	Severe to profound hearing loss	n = 12	96% (94–97%)	15% (7%–25%)
All studies	Enlarged vestibular aqueduct	Unilateral hearing loss	n = 7	67% (27–85%)	11% (7%–17%)
All studies	Enlarged vestibular aqueduct	No mixed or conductive hearing loss	n = 38	91% (89–93%)	12% (8%–15%)
Studies with consecutive patient status specified	Enlarged vestibular aqueduct	All	n = 29	88% (83–91%)	11% (8%–15%)
All studies	Narrow cochlear nerve canal/internal auditory canal	All	n = 31	88% (84–91%)	4% (2%–7%)
All studies	Narrow cochlear nerve canal/internal auditory canal	Severe to profound hearing loss	n = 11	86% (77–92%)	4% (1%–8%)
All studies	Narrow cochlear nerve canal/internal auditory canal	Unilateral hearing loss	n = 7	94% (89–96%)	9% (1%–21%)
All studies	Narrow cochlear nerve canal/internal auditory canal	No mixed or conductive hearing loss	n = 30	87% (83–90%)	5% (3%–7%)
Studies with consecutive patient status specified	Narrow cochlear nerve canal/internal auditory canal	All	n = 24	89% (85–92%)	5% (2%–7%)

^a Pooled diagnostic yields in the setting of substantial heterogeneity (ie, I² > 60%) should be interpreted with caution.

^b In the quantitative aggregate analysis, “all studies” includes all studies reporting data on a finding per patient basis. One publication²⁷ reported findings on a per ear basis alone; that publication would have appeared in the groups/subgroups of all studies, all findings, all hearing loss; all studies, all findings, unspecified/range of hearing loss.