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Abnormal Cochleovestibular Anatomy and Imaging: Lack of Consistency Across Quality of Images, Sequences Obtained, and Official Reports

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Abstract

Objectives: There are significant variations across centers on how to acquire and interpret imaging of children with congenital sensorineural hearing loss and cochleovestibular abnormalities. This study assesses the quality of imaging, sequences included, and accuracy of official radiology reports, to determine if these children are being assessed appropriately.

Methods: This study is retrospective review of CTs and MRIs from 40 pediatric patients diagnosed with profound sensorineural hearing loss and cochleovestibular structure/nerve abnormalities presenting to a tertiary referral academic center. Images were reviewed by two experienced neuroradiologists and a neurotologist. Findings were compared to official reports, when available.

Results: Twelve (30%) patients had an MRI only, while 28 (70%) had both an MRI and a CT. There were 3 (10.7%) CTs and 7 (17.5%) MRIs noted to be of poor quality. Children received an average of 6.8 (±2.7) CT acquisitions and 10.9 (±5.7) MRI acquisitions. There was non-concordance between the official report and expert review for 27 (71.1%) ears on CT and 27 (56.3%) ears on MRI.

Conclusions: These data demonstrate high variability in protocols and quality of medical imaging of children with sensorineural hearing loss. Interpretation of images is highly discordant

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between official reports and tertiary review. Given these results, we recommend that these children be imaged and evaluated at centers with neuroradiologists who are experienced in interpreting congenital abnormalities of the cochleovestibular system.

Keywords

Sensorineural hearing loss; cochlear implant; radiology

1. Introduction

Congenital hearing loss affects 1.4 per 1,000 infants screened annually in the United States[1]. Hearing loss has a profound impact on quality of life; children with hearing loss have lower IQ, difficulties in language acquisition and socialization and poor academic performance[2-4]. However, the advent of cochlear implantation in this population has resulted in vast improvements in neurocognitive development and socialization. Children who meet the criteria for cochlear implantation are able to achieve educational outcomes similar to that of normal hearing children[5].

Currently, imaging evaluation of a cochlear implant candidate includes structural Magnetic Resonance Imaging (MRI) to assess the cochleovestibular anatomy and the nerves in the internal auditory canal (IAC) and cerebellopontine angle (CPA) cistern. High definition Computed Tomography (CT) can be added to better view the bony cochleovestibular anatomy. These procedures guide the surgeon and determine which electrode will suit the inner ear anatomy[6].

While the majority of children with congenital sensorineural hearing loss have no gross bony or nerve abnormalities, roughly 20% have anomalies in the cochleovestibular structure or cochleovestibular nerve (CVN)[7]. For these children, high quality imaging of the temporal bone and IAC is essential because it assesses the functional potential of the CVN and determines whether or not a child would benefit from a cochlear implant. This information can result in the denial of the procedure by a surgeon or insurance. Correct depiction of anatomy affects the decision of the surgeon to pursue cochlear implantation, surgical approach, post-implant expectations, and post-surgical device programing by audiologists[8, 9] It is also important for guiding clinician presurgical discussions with families about risks and prognosis[8].

Previous studies[10-12] have shown a low association between imaging and baseline auditory function as well as post-implant outcomes. One of the reasons may be due to difficulties with imaging. Many of these malformations are rare and concordance of CT interpretations with surgical findings is related to the experience of the surgeon and neuroradiologist with these anomalies[13]. In order to correctly interpret these images, radiologists need to understand the embryogenesis of the inner ear and the classification systems for anatomical malformations. Unfortunately, there are a variety of classification systems and they do not always accurately describe the full range of potential abnormalities.

Another difficulty is with the acquisition protocol itself. There is a general consensus that high resolution CT with fine cuts of the temporal bone and high resolution T2-weighted

MRI are appropriate ways to image the inner ear[9, 14-16]. There is debate over whether both CT and MRI are needed. Some believe that a dual modality approach is preferred[17], while others believe the cost and risks do not always favor this approach[18]. There is no research about the current state of imaging protocols, quality of imaging or radiologists' interpretations across different centers. Additionally, there are no generally accepted imaging protocols for imaging the inner ear and many protocols are facility and physician dependent.

The purpose of this study is to determine the quality of imaging, protocols used, and accuracy of official radiology reports in children with congenital sensorineural hearing loss. We analyzed the imaging results from a group of children with abnormal cochleovestibular anatomy and/or and abnormal CVN to determine if these children are being imaged and assessed appropriately.

2. Materials and Methods

We retrospectively reviewed imaging (MRI and CT) for pediatric patients diagnosed with profound sensorineural hearing loss and abnormal cochleovestibular anatomy and/or an abnormal CVN. These patients presented to a tertiary referral academic center for second opinion. Imaging was obtained at 18 different centers. Forty children and 80 ears with imaging were included in the final analysis. Images were reviewed by two experienced neuroradiologists and a neurotologist, and a consensus interpretation was obtained.

The modiolus, cochlear aperture, cochlea, vestibule, semicircular canals and IAC were described. The cochlear aperture diameter and IAC midpoint diameter were measured from the widest point. Cochlear aperture was measured on CT if available and if no CT was available MRI was used. IAC diameter was measured on MRI. The fluid status in the IAC was reported, and the number of nerves visible in the CPA cistern and the IAC were counted. A cochlea was reported as abnormal if there was abnormal cochlear partitioning, hypoplasia of the cochlea or cochlear bud, or if the cochlea was absent. The vestibular system was reported as abnormal if the semicircular canals were dysplastic or absent, if the vestibule was enlarged or dysplastic or if the vestibular aqueduct was enlarged. The IAC should contain four nerves which include the facial nerve and 3 branches of the cochleovestibular nerve: the cochlear division and two vestibular divisions. The CPA cistern should contain two nerves: the facial nerve and the cochleovestibular nerve.

Images were rated as poor in quality if there was difficulty assessing any of the structures of the inner ear due to image resolution and/or artifact. The sequencing acquisition protocols of the images were assessed, and the number of acquisitions were counted to determine the variation in acquisition protocols across centers. Sequences that needed to be repeated due to motion artifact were not counted. When available, official reports from external radiologists were compared to our experienced specialist readings for concordance. CTs were concordant if there were matching descriptions for the modiolus, cochlear aperture, cochlear anatomy, vestibular anatomy and IAC. MRIs were concordant if there were matching descriptions of the nerves in the IAC for those who had both MRI and CT. For those who underwent MRI

only, images were concordant if both of the criteria previously mentioned for CT and MRI were met.

Descriptive statistics are presented separately for the cochleovestibular anatomy depicted by imaging, characteristics of imaging studies, and concordance of interpretations. Associations between imaging characteristics and number of nerves in the IAC were conducted using linear regression at a significance level of $p<0.05$. Written informed consent was obtained from at least one parent for all subjects. This project was approved by the Institutional Review Boards at the Keck School of Medicine of the University of Southern California and the University of California San Diego, School of Medicine.

3. Results

3.1 Participant characteristics and Cochleovestibular Anatomy

We collected bilateral imaging data on 40 patients with known bilateral or unilateral CVN abnormalities, or "absent auditory nerves." The patients age ranged from 0.25 to 14 years of age at the time of imaging collection and the average age was 1.61 years (SD=2.19). Eighteen patients (45.0%) were female and 22 (55.0%) were male. Sixty-five ears (92.9%) had an abnormal modiolus, while 9 (11.3%) were normal. The anatomy of the modiolus could not be determined for 6 ears (7.5%). For images where cochlear aperture size could be reliably determined, the mean cochlear aperture size was 0.53mm (SD=0.72) and 71 (97.3%) ears had a cochlear aperture diameter of less than 2.0mm. Forty-three cochlea (53.8%) were abnormal. The vestibular system was abnormal in 55 (68.8%) ears. The average IAC midpoint diameter was 3.27mm (SD=1.37) and 17 ears (21.3%) measured less than 2.0mm. Fifty-eight ears (72.5%) had sufficient fluid in the IAC to determine the nerve contents. Sixty ears (75.1%) had two discernable nerves in the CPA cistern while 14 ears (17.5%) had four nerves in the IAC. In depth information on the cochleovestibular anatomy of the patients is described in Table 1.

3.2 Imaging Characteristics

Twelve patients (30%) had an MRI only, while 28 (70%) had both an MRI and a CT. Nineteen (67.9%) CTs had an official CT report and 16 (40%) had an official MRI report included in their imaging. In terms of quality of imaging three (10.7%) CTs and seven (17.5%) MRIs were of poor quality, such that there was difficulty appreciating anatomical structures. Patients received an average of 6.8 (±2.7) CT acquisitions and 10.9 (±5.7) MRI acquisitions. Twelve patients (30%) received gadolinium-based intravenous contrast for their MRI imaging (Table 2).

None of the measurements of bony architecture were predictive of the number of nerves present in the IAC including: a small cochlear aperture (<2.0mm) (p=0.24), an abnormal cochlea (p=0.73), an abnormal vestibular system (p=0.47), or a small IAC (<2.0mm) (p=0.20). There is no clear consensus on what constitutes a small cochlear aperture or IAC. However, a cochlear aperture less than 2.0mm[8] and an IAC diameter of less than 2.0mm[19] are generally considered small.

3.3 Concordance with Official Reports

Specialist review was not concordant for 27 (71.1%) of 38 ears where official CT reports were obtained and 27 (56.3%) of 48 ears with official MRI reports. For participants with MRI only, 6 out of 8 ears (75%) were non-concordant. Four of those (66.7%) were non-concordant on the basis of comments about cochlear anatomy and/or the vestibular system. Overall, 9 out of 52 ears (17.3%) had an imaging report that was concordant in all modalities for which an official report was obtained. Four children (15.4%) had concordant imaging reports bilaterally. The most common non-concordant items on the official reports in descending order of frequency were incorrectly described vestibular system, missed modiolar abnormality, missed absent cochlear aperture, other incorrectly described cochlear architecture, incorrectly counted nerves in the IAC, and missed bifid IAC. Additionally, 11 (68.8%) of the official MRI reports did not comment on the nerves in the CPA cistern (Table 3). Official reads were not counted as discordant if they did not comment on the nerves in the CPA cistern if the number of nerves in cistern were normal. However, there was one case where the individual had distinct nerves present in the IAC but no discernable nerves in the CPA cistern (Appendix B).

Abnormalities were often described incorrectly. For example, a patient with a common cavity deformity on the left and an incomplete partition-type I on the right was initially described as having bilateral Michel Deformities (Figure 1). Another patient with absent cochlea bilaterally was first described as having common cavity malformations bilaterally. There were also cases where abnormalities were reported as normal. For example, one official report noted the vestibular system to be normal bilaterally but on expert review was found to have an enlarged vestibule and no semicircular canals bilaterally. Further examples of discordant interpretations are presented in Appendix A.

4. Discussion

This retrospective review of 40 MRI and CT scans obtained during evaluation for CI revealed that there is a large variation in imaging protocols for the management of children with congenital sensorineural hearing loss. The quality of image acquisition for both MRI and CT scans are frequently suboptimal, making it difficult to discern key anatomical features. Children were often administered gadolinium when no tumor was suspected based on available medical history. There was a wide variation in the number of acquisitions used to assess the labyrinthine anatomy and the neural contents of the IAC, with many receiving a large number of acquisitions. Additionally, official reads were often non-concordant with expert review.

Although some have argued that images can be reconstructed or reformatted[9], directly acquired high resolution images may be needed to evaluate the fine labyrinthine structure and structure of the nerves in the IAC[8] (Figure 2). In 10% of CTs and 17% of MRIs, acquisition techniques, not due to artifact, led to difficulties characterizing key structures including the status of the nerves in the IAC. These findings are of crucial importance for the decision to pursue a CI, and poor-quality imaging could lead to a misdiagnosis of anatomy, which could affect surgical risks and outcomes.

The results of this review also support a dual imaging approach for the evaluation of these children. The combination of CT and MRI provides more complete information about the bony anatomy and the status of the cochleovestibular nerves. Previous research has found that CT is superior to MRI in determining certain components of the inner ear[18, 20], while MRI is crucial for visualizing the CVN[21, 22] We found that a small cochlear aperture, an abnormal cochlea, an abnormal vestibular system and a small IAC midpoint did not predict how many nerve bundles were seen in the lateral IAC, which emphasizes that assumptions about nerves cannot be made from CT measurements alone. This supports prior research that highlights the limitations of using CT measurements of as a biomarker of CVN status[14], and is the primary reason why MRI has become the preferred modality for imaging in children with congenital sensorineural hearing loss.

Prior research has shown that a significant number of abnormalities can be detected in one modality and missed in another for both CT and MRI[23]. We found that non-concordance in reports was higher in patients who had MRI alone compared to patients who had both CT and MRI. This was most often due to incorrectly reported cochlear and vestibular structures, highlighting the fact that bony anatomy was more difficult to assess correctly without complementary CT imaging. Dual scans often require increased cost, the need for sedation, and potentially multiple appointments which can inconvenience families. However, the likelihood of having a more complete understanding of the anatomical structure in these children is higher when utilizing both modalities. These factors, as well as the unique risks of MRI and CT, sedation and exposure to ionizing radiation respectively, should be considered when determining the imaging approach of choice in these patients. Another important factor for clinicians to consider is whether there is a need to visualize the entire brain or other neuroanatomical structure beyond the IAC and temporal bones. As a result of the interplay of these factors, some have suggested algorithms for how to decide on imaging modality for pediatric patients[18].

Our results also suggest that imaging protocols being used in children with congenital sensorineural hearing loss are highly variable across centers and may expose these patients to unnecessary risks. A large number of children were given gadolinium-based contrast when not necessary for the imaging acquisition based on patient history. Additional CT imaging acquisitions increase radiation dose and additional MRI sequences increase the time of the study and may increase the amount of sedation used in these children. These results support the need for a standardized protocol for assessing the inner ear in pediatric patients. The high variation across centers in terms of number of acquisitions obtained highlights the fact that a succinct acquisition protocol would be beneficial in this population to reduce radiation and sedation exposure while still maintaining adequate visualizations of key structures.

Finally, concordance between official reports and specialist review was exceedingly low. Less than half of the ears had matching interpretations either on CT or MRI. Only 9 ears had fully concordant reports across all modalities where reports were available, which means that only four children overall had concordant reports for both ears. More than half of the reports conflicted with expert review of the cochlea and number of nerves in the IAC- two of the most important imaging findings which determine surgical approach and device selection

and assist in predicting surgical risks and CI outcomes. However, it is important to note that general radiologists have important roles in assessing temporal bone imaging studies including in the diagnosis of conductive hearing loss due to otitis media with effusion ossicular chain abnormalities and skull base tumors. Given the complexity of abnormalities that can present with congenital sensorineural hearing loss as well as the differing level of surgeon comfort with these abnormalities, this population would likely benefit from experienced temporal bone radiologists.

5. Conclusion

Accurate interpretation of MRI and/or CT of children with congenital sensorineural hearing loss is critical because it determines the surgical candidacy for cochlear implant and prevents inappropriate denial of CI by the patient's insurance based on misdiagnosis of certain anatomical variants. Therefore, it is crucial for the patient and the surgeon that images are acquired consistently and reported accurately. Given the high incidence of inadequate images, variations in imaging protocols, and discordance in radiologists' interpretations, we recommend that these children be imaged and evaluated at tertiary referral centers which have experienced neuroradiologists who are familiar with these rare inner ear malformations.

Supplementary Material

Refer to Web version on PubMed Central for supplementary material.

Acknowledgments

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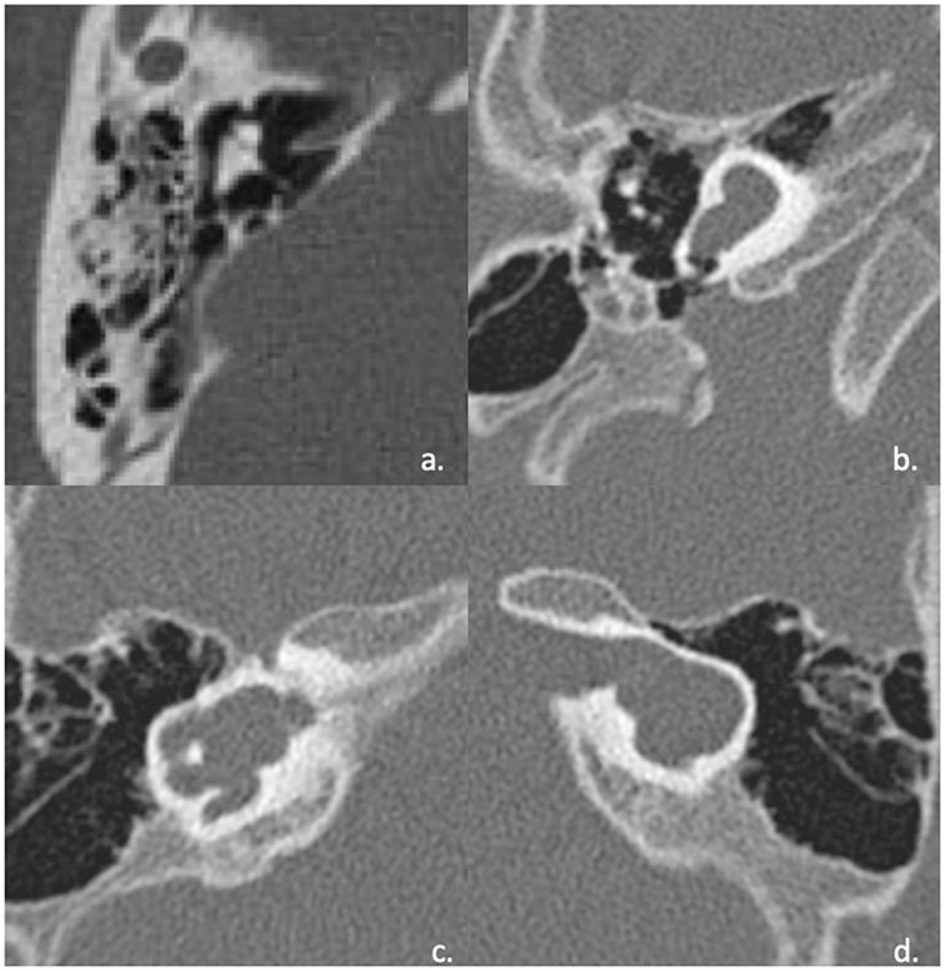

Figure 1.

Example of Discordant Interpretation of the Cochlea. Axial CT demonstrating (a) an example of Michel Aplasia- complete lack of any labyrinthine differentiation and (b) the right temporal bone in the patient who was described as having a bilateral Michel aplasia who was described by our team as having an IP-I (c) the same patient's right vestibular anomalies and (d) left common cavity are shown

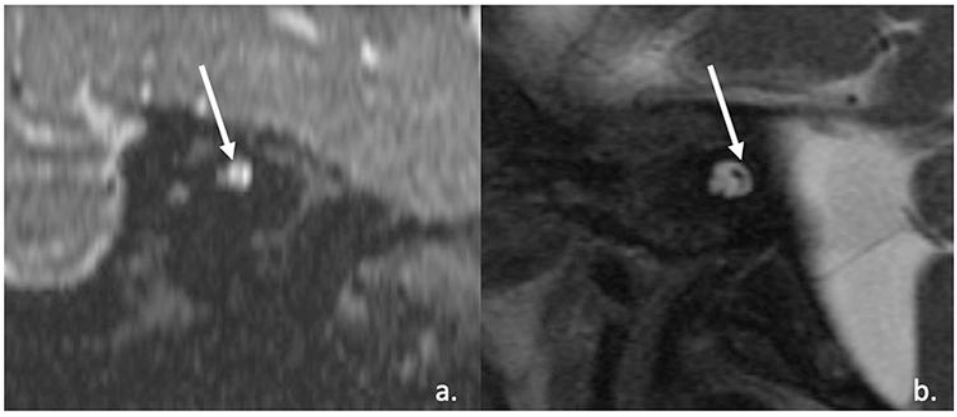

Figure 2.

Reformatted versus Directly Acquired Images. White arrow indicating internal auditory canal (IAC). Compare (a) reformatted oblique cuts through the IAC to (b) directly acquired cuts through the IAC.

Table 1.

Characteristics of Cochleovestibular Anatomy

Patient Characteristic	Description	Number
Age	Years	1.61 (±2.19)
Gender	Male	22 (55.0%)
	Female	18 (45.0%
Modiolus	Normal	9 (11.3%)
	Absent	17 (21.3%)
	Absent Cochlea	12 (15.0%)
	Bony	29 (36.3%)
	Deficient	5 (6.3%)
	Dysplastic	1 (1.3%)
	Slightly dense	1 (1.3%)
	Cannot be determined	6 (7.5%)
Cochlear Aperture*	Size (in mm)	0.53 (±0.72)
	Size <2.0mm	71 (88.8%)
	Size ≥2.0mm	2 (2.5%)
	Cannot be determined	7 (8.8%)
Cochlea	Normal	37 (46.3%)
	Absent	12 (15%)
	Common Cavity	3 (3.8%)
	Complete partition	2 (2.5%)
	Dysplasia	4 (5.0%)
	Hypoplasia	4 (5.0%)
	IP-I	15 (18.8%)
	IP-II	3 (3.8%)
Vestibular System	Normal	25 (31.3%)
	Abnormal vestibule	41 (51.3%)
	Abnormal vestibular aqueduct	3 (3.7%)
	Abnormal SCCs	49 (61.3)
IAC	Size (in mm)	3.27 (±1.37)
	Size <2.0mm	17 (21.3%)
	Size ≥2.0mm	63 (78.8%)
Nerves in Cerebellar Cistern	0	2 (2.5%)
	1	15 (18.8%)
	2	60 (75.1%)
	Cannot be determined	3 (3.8%)
Nerves in IAC	No Fluid	20 (25.0%)
	Cannot be determined	1 (2.5%)
	0	3 (3.8%)
	1	9 (11.3%)
	2	20 (25.0%)

Patient Characteristic	Description	Number
	3	13 (16.3%)
	4	14 (17.5%)
	Cochlear division hypoplastic	9 (11.3%)

IAC= Internal Auditory Canal, CVN= cochleovestibular nerve, IP-1=incomplete partition type I, IP=II= incomplete partition type II, SCCs=semicircular canals, Continuous variables are reported

Table 2.

Imaging Quality and Procedures

Imaging Characteristic	Description	Number
Imaging Modality	MRI	12 (30%)
	CT and MRI	28 (70%)
Official Reports Available	CT	19 (67.9%)
	MRI	16 (40.0%)
Poor image Quality	CT	3 (10.7%)
	MRI	7 (17.5%)
Number of Sequences	CT	6.8 (±2.7)
	MRI	10.9 (±5.7)
Gadolinium for MRI	No	28 (70.0%)
	Yes	12 (30.0%)

CT= Computerized tomography scan, MRI= Magnetic Resonance Imaging. Variables are reported as N(%) and N(± SD). Imaging results are reported by patient.

Table 3.

Concordance Between Official Reports and Specialist Review

Characteristic	Description	Number
Official Report Available	CT	19 (47.5%)
	MRI	24 (60.0%)
	At least one (CT, MRI or both)	23 (57.5%)
CT Concordance (by ear)	Concordant	11 (28.9%)
	Non-concordant	27 (71.1%)
MRI Concordance (by ear)	Concordant	21 (43.8%)
	Non-concordant	27 (56.3%)
Overall concordance	By ear	9 (17.3%)
	By child	4 (15.4%)
Reason for Non-Concordance	Incorrectly Described Vestibular Anatomy	14 (66.7%)
	Missed Modiolar Abnormality (n=33)	21 (63.6%)
	Missed Absent Cochlear Aperture (n=13)	8 (61.5%)
	Incorrectly Described Cochlear Anatomy (n=19)	11 (57.9%)
	Incorrectly Reported Nerves in IAC (n=46)	26 (56.5%)
	Missed Bifid IAC (n=3)	1 (33.3%)
Status of Nerves in Cerebellar	Included in Official Report	5 (31.2%)
Cistern	Not Included in Official Report	11 (58.8%)

CT= Computed Tomography, MRI= Magnetic Resonance Imaging. Variables are reported as N(%). Official reports are counted by patient. Concordance is counted by ear. Non-concordance data was based on ears that had official reports available.