

IMAGING FEATURES OF CHILDREN WITH HEARING LOSS

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ABSTRACT

Objectives: To determine the related factors of sensorineural hearing loss and the imaging features of this issue among children in Children's Hospital 1.

Methods: From July 2022 to July 2023, a prospective study was conducted on 72 children with a diagnosis of sensorineural hearing loss at the Children's Hospital 1.

Results: The degree of hearing loss most severe-to-profound hearing loss (≥ 90 dB) was 51.4% as many as 46 children (63.9%) with asymmetrical hearing loss in most of the subjects. Most of the children with bilateral congenital sensorineural hearing loss (SNHL) are in the age > 5 years old (mean age: 5.8 years). There were 19.4% of cases having abnormal inner ear images on temporal bone computed tomography (CT scan) Through brain, the percentage of abnormal cranial nerve 8th was 6.6%, including 2 cases of aplasia cranial nerve 8th and 2 cases of hypoplasia cranial nerve 8th.

Conclusion: There was a delay in the diagnosis of prelingual sensorineural hearing loss, with a high proportion of the severe - to - profound among children presenting with a diagnosis. Besides diagnosing the causes of hearing loss, CT scan and MRI also assist in determining the feasibility of surgical intervention based on the images they provide. Audiometry tests can indicate the extent of hearing loss and profound deafness caused by cochlear damage. However, when a CT scan is conducted, an MRI can offer us precise information regarding the causes, potential recovery, and the techniques and approaches for intervention.

Keywords: Sensorineural hearing loss, hearing loss, children.

I. BACKGROUND

Hearing is extremely important and indispensable for the development of children, especially in the first 5 years of their lives [1, 2]. According to the World Health Organization, there are currently 466 million people with hearing loss in the world, of which 34 million are children.

Hearing loss has numerous causes and the exact cause cannot always be determined. Most children with hearing impairment show delayed detection and diagnosis of pre-lingual hearing loss, with a high proportion of severe cases in most children who come for hearing tests. Families often lack

understanding and awareness of their child's auditory responses, and typically only seek medical attention and conduct hearing function tests when the child displays clear signs such as delayed speech and psychomotor development [3].

Some studies have shown that in the absence of other defects, if children with congenital deafness intervened appropriately before 6 months of age, they may have the same hearing level as their peers in terms of language development. The diagnosis and treatment of sensorineural hearing loss require a combination of audiometric parameters and diagnostic imaging, including temporal bone CT scan and MRI of the ears - brain [1, 2].

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Functional hearing tests will help accurately diagnose the degree of hearing loss and the location of the damage. CT scan allows for a detailed evaluation of the external, middle, and inner ear structures, aiding in the diagnosis of malformations and assessment of anatomical variations to determine surgical indications and evaluate surgical difficulties. MRI is a method that helps identify abnormalities in the brain and determine the presence of the auditory nerve and is also one of the factors determining surgical indications and reinforcing the incomplete images provided by the CT scan. 6-9. We conducted a study to determine the prevalence of factors associated with sensorineural hearing loss and the imaging features related to this issue among children at Children's Hospital 1.

II. MATERIALS AND METHODS

2.1. Study settings

From July 2022 to July 2023, we conducted a case series study on 72 pediatric patients with the diagnosis of sensorineural hearing loss who admitted to Children's Hospital 1.

2.2. Study design and participants

Inclusion criteria: aged under 16; confirmed diagnosis of sensorineural hearing loss at the Audiology Unit of Children's Hospital 1 by pure tone audiometry, conditioned play audiometry (CPA), auditory brainstem response (ABR) or auditory steady-state response (ASSR); having the results of other audiometric tests including tympanography and acoustic reflex testing; and having the results of temporal bone CT scan and brain MRI.

Exclusion criteria: Unknown family history of the patients, unknown obstetric history of the patient's mothers, medical history of the child and mother (before and during pregnancy), and relatives of patients who disagreed to participate in the study.

2.3. Conducting method

Firstly, all subjects who met the admission criteria were explored the clinical characteristics including the reasons for detecting hearing loss, the age of that detection, their medical history and obstetric history of their mothers related to

sensorineural hearing loss; and hearing loss history of their parents and siblings. Next, the audiometric parameters comprising hearing threshold, severity of the hearing loss, the results of tympanography and acoustic reflex were recorded. Finally, the patients were conducted temporal bone CT scan and brain MRI according to the standards and evaluated the results of captured images.

2.4. Statistical method

The data were entered using Epidata 3.1 software and were analysed using Stata 14.0 software.

2.5. Ethical considerations

This study was approved by the Medical Ethics Committee of University of Medicine and Pharmacy at Ho Chi Minh City and the Ethics Committee of Children's Hospital 1.

III. RESULTS

3.1. Characteristics of the study sample

The average age of children when hearing loss detection was 5.8 years (range: 0.8 - 14.2 years). More than 80% of children with hearing loss was female, and the majority of them lived in rural areas.

3.2. Clinical characteristics

Approximately 22% of hearing loss cases were detected before the age of 2 years and more than 40% of cases were detected after the age of 6 years, but no cases were identified at an earlier stage and referred to the Department of Otorhinolaryngology in Children's Hospital 1. The most common reasons for hearing loss detection were decreased hearing level (62%), speech delay (28.3%), and only 2.2% children were hearing loss screening.

Among the risk factors for hearing loss, infants who had past medical history of being treated in neonatal resuscitation unit for more than 5 days accounted for the majority with 23 cases, representing 59% of the total 4 cases, accounting for 10.3% had a history of treatment requiring respiratory support during the neonatal period, 1 case had undergone extracorporeal membrane oxygenation (ECMO), and 1 case had jaundice and required blood transfusion. There were also 3 cases (7.7%) with craniofacial abnormalities (Table 1).

Table 1: Hearing loss risk factors JCIH 2007

Hearing loss risk factors		Number (cases)	Rate (%)
During and after birth	Neonatal resuscitation more than 5 days	23	59.0
	Extracorporeal membrane oxygenation	1	2.6
	Assisted ventilation	4	10.3
	Using auditory poisoning drugs	0	0
	Hyperbilirubinemia	1	2.6
	Blood transfusion need	3	7.7
	Craniofacial abnormalities	0	0
	Clinical syndrome associated with sensorineural hearing loss	0	0
	Postpartum infection with positive blood cultures (bacterial meningitis, Herpes and Varicella viruses)	0	0
	Head injury	1	2.6
	Children must be treated with chemotherapy and radiotherapy	0	0
Family medical history	family history of sensorineural hearing loss	5	12.8
Total		39	100

The results demonstrate that 13.9% of cases have hearing loss in only one ear and 86.1% in both ears. In the latter group, the percentage of children who have asymmetrical hearing loss in two ears was 63.9%. Out of 72 cases, The highest proportion of severe hearing loss with 51.4%, ranking second was severe hearing loss accounting for 23.6%, and third place was moderate hearing loss about 19.4% of which there were 3 cases of sudden deafness (Table 2).

Table 2: Hearing loss degree

Hearing loss degree	Right ear		Left ear	
	Number (cases)	Rate (%)	Number (cases)	Rate (%)
Normal (< 25 dB)	2	2.8%	8	11.1%
Light (26 - 40 dB)	3	4.2%	1	1.4%
Medium (41 - 70 dB)	13	18.1%	14	19.4%
Severe (71 - 90 dB)	17	23.6%	12	16.7%
Profound deafness (> 90dB)	37	51.4%	37	51.4%
	N = 72	100%	N = 72	100%

Temporal bone CT scan and ears-brain MRI results

Regarding external ear canal and middle ear abnormalities, the results of temporal bone CT denote that there were 81.5% of cases having normal middle ear imaging; while mastoid hypoventilation, tympanic fluid accumulation, and mastoid hypoplasia accounted for 4.9%, 4.9% and 2.5%, respectively (Table 3).

Table 3: CT scan image of the external ear canal and middle ear

		Number (cases)	Rate (%)
The external ear canal and middle ear	Normal	66	81.5%
	High riding jugular bulb	3	3.7%
	Mastoid hypoventilation	4	4.9%
	Tympanic fluid accumulation	4	4.9%
	Destruction of ossicles	2	2.5%
	Mastoid hypoplasia	2	2.5%

When investigating inner ear abnormalities, CT scan results indicate that there were 2 cases of Cystic hypoplastic cochlea (CH-Type 2), accounting for 2.7%, 3 cases of incomplete partition Type I (IP – I), and 2 cases of hypoplastic cochlea less than 2 turns (CH-Type 3). The prevalence of enlarged vestibular aqueduct was 2.5%, and 3 cases of narrow internal auditory canal with 4.1% (Table 4). Similar to the image of the cochlea on CT scan of the temporal bone, the MRI image had the same results (Table 5).

Table 4: CT scan image of cochlear.

CT scan Results		Number (cases)	Rate (%)
Cochlear	Normal	65	90.5%
	CH - Type 2 (Cystic hypoplastic cochlea)	2	2.7%
	CH - Type 3 (less than 2 turns)	2	2.7%
	Incomplete partition Type I (IP - I)	3	4.1%
	Total	N = 72	100%

Table 5: CT scan image of the vestibular and semicircular canals

CT scan Results	Number (cases)	Rate (%)
Normal	61	84.9
Cochlear vestibular dysplasia	4	5.5
Cochlear vestibular hypoplasia	2	2.7
Widening vestibule drain	2	2.7
Narrowing of the inner ear canal	3	4.2
Total	72	100

In the study results, through MRI, we recorded that the rate of normal cranial nerve 8th was 94.4%, whereas abnormal rate was 6.6%, including 2 cases of aplasia cranial nerve 8th and 2 cases of hypoplasia cranial nerve 8th (Table 6) This result contributes to confirm the patient's diagnosis of the location of the lesion causing hearing loss, and plays a role in choosing the surgical ear.

In addition, MRI results reveal that there were 87.5% of cases having normal brain imaging, 8 cases had sequelae of brain parenchymal injury suspected by fetal infection, accounting for 11.1%, and 1 case had ventricular dilatation (Table 7).

Imaging features of children with hearing loss

Table 6: Cranial nerve 8th MRI image

Cranial nerve 8 th MRI image	Number (cases)	Rate (%)
Normal	68	94.4
Nerve aplasia	2	2.8
Nerve hypoplasia	2	2.8
Total	72	100

Table 7: MRI imaging of cerebellopontine angle and brain parenchyma

	Number (cases)	Rate (%)
Normal	63	87.5
Sequelae of brain parenchymal injury	8	11.1
Ventricular dilatation	1	1.4
Total	72	100

When analyzing the relationship between structural abnormalities on temporal bone CT scan and MRI of the ear-brain and the severity of hearing loss from severe to profound, the results of our study are shown in Table 8.

Table 8: Correlation of hearing loss with number of structural abnormalities

Number of Structural Abnormalities Hearing loss degree	0	1	2	≥ 3	Total
Normal	0	0	0	0	0
Light level (26 - 40 dB)	0 (0%)	0 (0%)	0 (0%)	1 (100%)	1 (100%)
Medium level (41-70 dB)	14 (57.1 %)	4 (19.0%)	3 (14.3%)	2 (9.5 %)	21 (100%)
Severity (71 - 90 dB)	11 (57.9 %)	4 (21.1%)	2 (91.5%)	2 (10.5%)	19 (100%)
Profound deafness (> 90dB)	22 (71.6 %)	3 (9.7%)	3 (9.7%)	3 (9.7%)	31 (100%)
Total	47 (65.3%)	11 (15.3%)	8 (11.1%)	6 (8.3%)	72 (100%)

Table 9: Frequency of abnormalities and severe to profound hearing loss in the Right ear

Number of Structural Abnormalities	Number (cases)	Rate (%)	OR	p
0	34/47	72.3 %	-	< 0.005
1	9/11	81.8 %	0.52 (0.11 - 3.05)	0.522 (> 0.005)
2	5/8	62.5 %	1.56 (0.32 - 7.52)	0.573 (> 0.005)
≥ 3	5/6	83.3 %	0.53 (0.056 - 4.91)	0.571 (> 0.005)

Table 10: Frequency of abnormalities and severe to profound hearing loss in the left ear

Number of Structural Abnormalities	Number (cases)	Rate (%)	OR	p
0	34/47	72.3 %	-	< 0.005
1	8/11	72.7 %	0.98 (0.22 - 4.27)	0.979 (> 0.005)
2	4/8	50.0 %	2.61 (0.56 - 12.03)	0.217 (> 0.005)
≥ 3	3/6	50.0 %	2.61 (0.46 - 14.65)	0.274 (> 0.005)

In our study, we observed that 47 out of 72 cases, accounting for 65.3%, exhibited different degrees of hearing impairment without any abnormalities in the structure of the temporal bone CT scan or MRI of the ear-brain. Additionally, 11 out of 72 cases showed 1 abnormality, while 8 out of 72 cases had 2 or more abnormalities, representing proportions of 11.1% and 8.3% respectively. When analyzed using a simple linear regression model, the results were not statistically significant (Table 8).

IV. DISCUSSION

In terms of clinical characteristics, a study of 72 children diagnosed with hearing loss revealed an average diagnostic age of 5.8 years (71 months), with the oldest being 14.2 years (171 months) and the youngest being 0.8 years (10 months). These findings are consistent with the study conducted by Pham Dinh Nguyen in 2018 at Children's Hospital 1, in which the youngest age being 1 month old and the oldest being 12 years old, [4].

The results of this study are consistent with study of Wiranadha (2020), [5], in which the rate of severe and profound hearing loss account for 71.01%. Similarly, in Vietnam, Tai Tan Pham Doan (2017), [6] recorded that rate of hearing loss was 67% (> 90dB).

Thus, this rate is lower than ours, which can be explained by differences in population characteristics and sampling criteria. The reason for these results is

not due to higher prevalence of severe and profound hearing loss in the community, but rather because severe and profound hearing loss is easier to detect than children with mild hearing loss.

Regarding imaging features, when referring to hearing loss related to abnormalities of structures in the inner ear or vestibulocochlear nerve. The authors McClay J.E. et al. (2008), [7] and Coticchia J.M. et al. (2006), [8] reported a higher prevalence of inner ear abnormalities in children with severe-to-profound SNHL compared to those with mild or moderate SNHL. They have shown that the majority of SNHL patients have normal morphology, with reported abnormality rates of approximately 25% on CT scan and around 40% on MRI. In our study, we also observed similar trends in the overall abnormality rate, with approximately 35% of cases exhibiting abnormalities of varying degrees and there were differences in the specific rates within

each group. Based on the findings from the research by McClay J.E. and Coticchia J.M., the incidence of abnormalities in inner ear structures in children with severe-to-profound SNHL appears to be higher compared to those with mild or moderate SNHL, which have differences from our previous discussion.

According to research conducted by McClay J.E. et al., it was discovered that out of 271 cases of SNHL, 87 cases (32%) showed abnormal cochlear imaging. Abnormalities included Cystic hypoplastic cochlea (CH - Type 2), Incomplete partition Type I (IP - I), hypoplastic cochlea less than 2 turns (CH-Type 3), cochlear nerve in 29% of the cases, and an absent (aplastic) cochlear nerve in 14% of the cases. We also noted similar abnormalities, albeit in a smaller proportion. Furthermore, among the 72 cases examined, 2 cases (2.8%) displayed hypoplasia of the cochlear nerve, while an additional 2 cases (2.8%) aplastic cochlear nerve. We would like to emphasize that despite having a much smaller sample size and limited data collection time compared to foreign authors, the observed abnormalities were still diverse and similar to other studies.

Caselman and Doris emphasized the need to always take MRI for all patients with congenital hearing loss [9,10]. O Chin and colleagues conducted a study to analyze the results of CT scan and MRI on 240 congenital hearing loss children, in order to determine whether MRI alone is enough to evaluate structural abnormalities of the inner ear before surgery. The results show that the combination of CT scan and MRI may be effective but insignificant [11]. The question of whether a combination of CT scan and MRI is necessary for the diagnosis of hearing loss is still controversial. However, according to the results of this study, we found that CT scan only recorded 3 cases of narrow internal auditory canal, while MRI detected 4 cases of cranial nerve 8th abnormalities. Thereby, this result may indicate that doctors could have missed abnormal images if merely using CT scan.

In addition to diagnose the causes of hearing loss, CT scan and MRI also assist in determining the feasibility of surgical intervention based on the images they provide. Audiometry tests can indicate the extent of hearing loss and profound deafness caused by cochlear damage. However, when a CT

scan is conducted, an MRI can offer us precise information regarding the causes, potential recovery, and the techniques and approaches for intervention.

When comparing the occurrence frequency of structural abnormalities in the study, we discovered that among the severe cases, there were common abnormalities in the cochlea. These findings align with a study conducted by McClay J.E and colleagues, who also observed common abnormalities such as cochlear malformation. Additionally, when comparing our results to a study conducted in Vietnam by author Nguyen Xuan Nam [12], where only 1 out of 146 ears had Mondini malformation with an incomplete cochlear structure of 2.5 turns, we also observed 2 out of 72 cases with ears having only 2 turns. According to Aschendoff in 2009, Mondini malformation is the most prevalent abnormality among cochlear anomalies, accounting for 45% of cases.

In previous studies, many authors have found a correlation between the number of abnormalities and the severity of hearing loss. For example, McClay J.E, [7] reported that ears with severe or profound hearing loss had more abnormalities than ears with mild or moderate hearing loss (66/138 [48%] vs. 23/80 [29%]; $P = 0.006$). Children with moderate or severe-profound hearing loss had more inner ear abnormalities than children with bilateral moderate or severe hearing loss (28/45 [62%] vs. 54/144 [38%]; $P = 0.004$). In our study, we found that 47/72 (65.3%) cases of hearing loss of all degrees did not have any structural abnormalities on CT of the temporal bone or MRI of the ear-brain. Next, 11/72 cases had 1 abnormality, and the frequency in the groups with 2 abnormalities and ≥ 3 abnormalities was 8/72 and 6/72 cases, accounting for 11.1% and 8.3%, respectively. When analyzed with a simple regression model, the results were not statistically significant. However, as mentioned above, all cases with abnormalities had severe profound hearing loss degree.

We noticed a significant difference in the proportion between the two groups with and without structural abnormalities. This led to results that differed from those of other authors who conducted similar studies. Possible explanations for the difference in results may be that the different sampling criteria and populations, the rate of

abnormalities may fluctuate from those studied in previous studies.

V. CONCLUSION

Therefore, in addition to diagnosing the cause of hearing impairment, CT scan and MRI also help determine whether surgical intervention may be necessary based on the images they provide. Auditory function tests may indicate the extent of hearing loss and profound deafness due to damage to the cochlea, but they alone may not be sufficient. Still, it is through CT scan and MRI that we can identify the cause, the potential for recovery, and the methods and means of intervention.

Abnormal images may be missed if only images from the bone CT scan, and the information provided by the MRI of the brain and ear help to reinforce the incomplete images provided by the CT scan.

Disclosure

The authors report no other conflicts of interest in this work.

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REFERENCES

1. World Health Organization Library Cataloguing-in-Publication Data, Childhood hearing loss: Strategies for prevention and care. 2016.
2. Yoshinaga - Itano C, Sedey AL, Coulter DK, Mehl AL. Language of early - and later - identified children with hearing loss. *Pediatrics*. 1998;102(5):1161-71.
3. Culbertson SR, Dillon MT, Richter ME, Brown KD, Anderson MR, Hancock SL, et al. Younger Age at Cochlear Implant Activation Results in Improved Auditory Skill Development for Children With Congenital Deafness. *J Speech Lang Hear Res*. 2022;65(9):3539-3547.
4. Phạm Đình Nguyên. Nghiên cứu khiếm thính không mắc phải ở trẻ em. 2018.
5. Wiranadha IM, Hartayanti A. Characteristics of Congenital Sensorineural Hearing Loss in Children at ENT Outpatient Clinic Sanglah General Hospital Denpasar in 2017. *Biomedical and Pharmacology Journal*. 2020;13.
6. Phạm Doan Tân Tài, Quang PD, Nhu NT, Hung DX. Đánh giá tình trạng nghe kém tiếp nhận - thần kinh và các yếu tố liên quan ở trẻ tại Bệnh Viện Nhi Đồng 1. *Tạp Chí Y Học TP Hồ Chí Minh*. 2017;23(4):173-178.
7. McClay JE, Booth TN, Parry DA, Johnson R, Roland P. Evaluation of pediatric sensorineural hearing loss with magnetic resonance imaging. *Arch Otolaryngol Head Neck Surg*. 2008;134(9):945-52.
8. Cotichchia JM, Gokhale A, Waltonen J, Sumer B. Characteristics of sensorineural hearing loss in children with inner ear anomalies. *Am J Otolaryngol*. 2006;27(1):33-8.
9. Casselman JW, Offeciers FE, Govaerts PJ, Kuhweide R, Geldof H, Somers T, et al. Aplasia and hypoplasia of the vestibulocochlear nerve: diagnosis with MR imaging. *Radiology*. 1997;202(3):773-81.
10. Bamiou DE, Worth S, Phelps P, Sirimanna T, Rajput K. Eighth nerve aplasia and hypoplasia in cochlear implant candidates: the clinical perspective. *Otol Neurotol*. 2001;22(4):492-6.
11. Chin O, Dharsono F, Kuthubutheen J, Thompson A. Is CT necessary for imaging paediatric congenital sensorineural hearing loss? *Cochlear Implants Int*. 2020;21(2):75-82.
12. Nguyen Xuan Nam. Nghiên cứu thăm dò chức năng nghe, chẩn đoán hình ảnh và đánh giá kết quả thính lực của trẻ cấy điện cực ốc tai. Luận Án Tiến Sĩ, Đại Học Y Hà Nội. 2017.