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RESEARCH

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Incidental MRI brain findings in children with sensorineural hearing loss

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Abstract

Background Sensorineural hearing loss is one of the leading causes for cognitive dysfunction. Incidental brain abnormalities are frequently seen in patient`s MRI. Our aim was to highlight the incidence of brain abnormalities in children with sensorineural hearing loss and to consider brain screening as a part of the standard cochlear implant MRI protocol.

Methods This retrospective study included 385 prelingually deaf mute children who were referred for pre-cochlear implant imaging evaluation in the period from January 2020 to June 2022. We evaluated brain images for any structural or white matter abnormality.

Results We detected brain abnormalities in 62 patients (16.11%), 27 (7.01%) with white matter lesions and 35 patients (9.1%) with other structural brain abnormalities. The commonest white matter lesions were bilateral focal lesions (5.71%). The commonest structural brain abnormality was arachnoid cyst (2.86%). Four patients had two coincidental brain abnormalities. No significant correlation was found between ear abnormalities and white matter lesions or structural brain abnormalities ($P > 0.05$).

Conclusions The incidence of brain abnormalities in children with sensorineural hearing loss is not uncommon. Pre-implant MRI screening of the brain helps to obtain best outcomes.

Keywords MRI, Brain, Sensorineural hearing loss, Cochlear implantation

Background

Sensorineural hearing loss (SNHL) is one of the leading causes for cognitive dysfunction [1]. Children with SNHL have to fit eligible audiological, mental and imaging criteria before proceeding for cochlear implantation (CI) [2]. MRI is an important tool for preoperative evaluation of inner ear and cochlear nerve prior to CI [3, 4].

Incidental brain abnormalities are frequently seen in children with SNHL. Most of them do not contraindicate CI [2]. Many lesions may be missed clinically as they may have no manifest neurological dysfunction [3]. Some structural abnormalities may have no impact on perceptual processing or cognition [3, 4]. However, other lesions like white matter (WM) abnormalities may have an impact on cognitive function or neurological development [2–5]. White matter lesions may slow down auditory impulses transmission or may cause nerve fiber injury in different brain structures with consequent lag in language development [6–8]. Despite that the effect of WM lesions on post-implant hearing improvement is still unclear [4, 8], they may have an impact on post-implantation auditory and speech performance [3]. Early CI in prelingually deaf mute children increases the likelihood of obtaining better auditory development, receptive and expressive language outcomes [9, 10]. Better outcomes found in patients aged below 6 years [11, 12].

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MRI has the advantage to assess the brain for any incidental pathological or structural abnormality [2]. The ability to perform MRI after CI is still a matter of debate and is not widely accepted to be safe. Also, images will be suboptimal due to the distortion effect of the implanted magnet [8]. Ideally, we need to remove the magnet from the implanted package and replace it after performing MRI, a condition that is not practical and needs general anesthesia [7, 8].

Before surgery, we aim to accurately predict the expected benefit from CI [13]. Brain screening provides a baseline information before MRI scanning becomes relatively contraindicated due to the applied implant [7].

Our aim was to highlight the incidence of brain abnormalities in children with SNHL and to emphasize the importance of considering brain screening as a part of the standard CI MRI protocol to gain valid preoperative information and to guarantee best outcomes.

Methods

This retrospective study included 385 prelingually deaf mute children who were referred from ENT Department or outpatient clinic to the Diagnostic Radiology Department for pre-CI imaging evaluation in the period from January 2020 to June 2022. The number of patients underwent CI surgery by the time of the study was 224.

Approval of our institute Research Ethics Committee was obtained.

Inclusion criteria

- Children with SNHL 5 years old or less.
- Both genders were included.
- Patient fulfilled audiological and phoniatric criteria eligible for CI.

Exclusion criteria

- Patient older than 5 years, as we were concerned with the younger prelingually deaf mute children in our study trying to enroll them for early CI to attain the best post-operative outcomes.
- Patients with no or missing imaging records.

MRI protocol

MRI was done using 1.5-T MRI unit (Philips, Acheiva, Netherlands) using a dedicated head coil. Patients lie in neutral supine position after a light sedation (chloral hydrate, syrup, 500 mg/5 mL, 30–50 mg/kg as a single dose 15–30 min before examination) was given. The ear protocol included axial and coronal 3D balanced turbo gradient echo (B-TFE sense) sequences on cerebellopontine angle and inner ear with sagittal oblique T2-weighted 3D Drive clear sequence perpendicular on each internal auditory canal. Brain protocol included axial fluid-attenuated inversion recovery (FLAIR). In case of incidental brain abnormality was found, the protocol was extended for a full brain imaging protocol including axial and coronal T2 WI, sagittal T1 WI and axial DWI.

Image analysis

Images were reviewed and analyzed for any inner ear abnormality or cochlear nerve abnormality by a single radiologist with 9 years experience in cochlear implant imaging, all findings were recorded. The brain images were evaluated for any structural abnormality or WM abnormality. We simply classified WM lesions into focal or diffuse, unilateral or bilateral regardless their severity and their impact on the surgery.

Statistical analysis

Data were analyzed using STATA version 14.2 (Stata Statistical Software: Release 14.2 College Station, TX: StataCorp LP.). Quantitative data were represented as mean, standard deviation, median and range. As the data were not normally distributed, Kruskal–Wallis test was used for comparison of three groups and Mann–Whitney test was used to compare two groups. Qualitative data were presented as number and percentage and compared using either Chi-square test or Fisher exact test. P value was considered significant if it was less than 0.05.

Results

This retrospective study carried out on 385 patients, 199 (51.69%) were females and 186 (48.31%) were males. Their age ranged from 1 to 5 years, the mean age ± SD was 2.23 ± 1.07, the median (range) was 3 (1:5).

Brain abnormalities were detected in 62 patients (16.11%), 27 patients (7.01%) had WM lesions, and 35 patients (9.1%) had other structural brain abnormalities. In the 27 patients with WM lesions, they were unilateral in 4 patients (1.04%), bilateral focal in 22 patients (5.71%), bilateral diffuse in 1 patient (0.26%) (Fig. 1) (Table 1). In the 35 patients with other structural brain abnormalities, arachnoid cyst was found in 11 patients (2.86%) (Fig. 2A), diverticular outpouching of foramen of Luschka in 4 patients (1.04%) (Fig. 2B), diffuse brain atrophy in 3 patients (0.78%), Chiari I malformation in 3 patients (0.78%) (Fig. 2C), periventricular leukomalacia in 2 patients (0.52%) (Fig. 3A), Chiari II malformation in

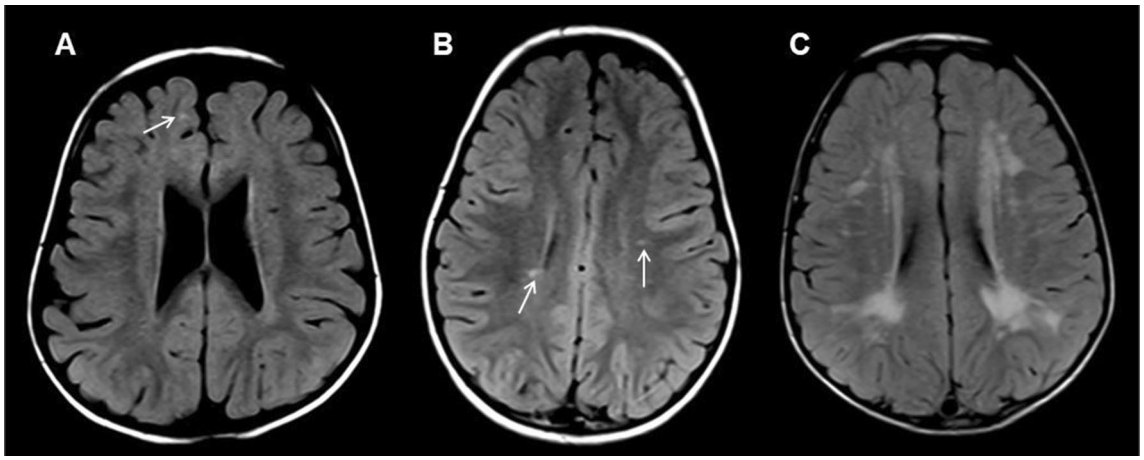

Fig. 1 Axial FLAIR images show WM lesion in in three different patients. **A** Unilateral focal lesion (arrow) at right frontal lobe in a 5 years old child, **B** bilateral focal lesions (arrows) at parietal lobes in a 3 years old child, **C** Bilateral diffuse lesions in a 3 years old child

Table 1 Distribution of white matter lesions in the studied population

White matter lesion	Summary statistics
Bilateral focal WM lesion	
Negative	363 (94.29%)
Positive	22 (5.71%)
Bilateral diffuse WM lesion	
Negative	384 (99.74%)
Positive	1 (0.26%)
Unilateral focal WM lesion	
Negative	381 (98.96%)
Positive	4 (1.04%)
Unilateral diffuse WM lesion	
Negative	385 (100%)
Positive	0
WM lesion	
Negative	358 (92.99%)
Unilateral	4 (1.04%)
Bilateral	23 (5.97%)

1 patient (0.26%), venous angioma in 1 patient (0.26%), tentorial incisura hernia in 1 patient (0.26%) (Fig. 3B), focal cerebral encephalomalacia in 1 patient (0.26%), focal dysplastic cerebral cortex in 1 patient (0.26%), olivopontocerebellar atrophy in 1 patient (0.26%), Joubert syndrome in 1 patient (0.26%) (Fig. 3C), isolated lateral ventricular dilatation in 1 patient (0.26%). Four patients had two coincidental brain abnormalities including cerebellar hemiatrophy and subependymal heterotopia in 1 patient (0.26%) (Fig. 4), cerebellar hemiatrophy and veli interpositi cyst in 1 patient (0.26%), corpus callosum agenesis and heterotopia in 1 patient (0.26%), Blake pouch cyst and hypoplastic brainstem in 1 patient (0.26%) (Table 2). No significant correlation was found between patients age and WM lesions ($P=0.056$), gender and WM lesions ($P=0.31$), patients age and structural brain abnormality ($P>0.05$) and between gender and structural brain abnormality ($P>0.05$).

Ear abnormalities were found in 101 patients (26.24%), unilateral in 15 patients (3.90%) and bilateral in 86 patients (22.34%).

In the 27 patients with WM lesions, bilateral ear abnormality found in 5 patients (18.52%). No significant correlation between WM lesions and ear abnormalities ($P=0.80$) (Table 3). We had 224 out of 385 patients underwent CI surgery by the time of the study. There were 18 patients with bilateral cochlear nerve aplasia, 2 patients with bilateral Michel deformity, one patient with Michel deformity on the right and rudimentary otocyst on the left, surgery was contraindicated in all of them. Some patients did not proceed into the surgery due to socioeconomic reasons, others were in the waiting list by the time study was ended. None of the brain abnormalities found in our study was considered as a contraindication for surgery.

Of the 101 patients with ear abnormalities, WM lesions were found in 5 patients (4.95%), unilateral in 1 patient (0.99%) and bilateral in 4 patients (3.96%). Other structural brain abnormalities were found in 15 patients (14.85%). No significant correlation was found between ear abnormalities and WM lesions or structural brain abnormalities ($P>0.05$) (Table 4).

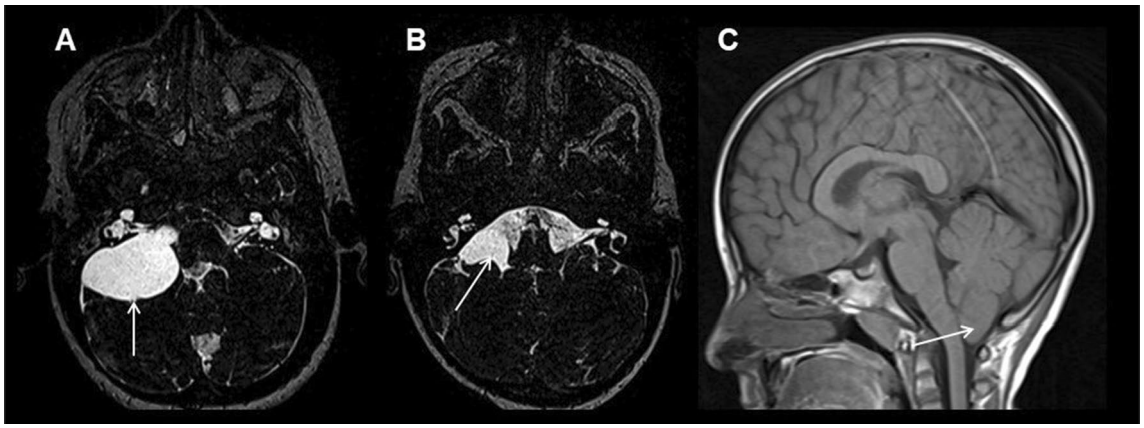

Fig. 2 **A** Right CPA cistern arachnoid cyst (arrow) in a 4 years old child with bilateral incomplete partition type 1, **B** Diverticular outpouching (arrow) of right foramen of Luschka in a 3 years old child, **C** Chiari I malformation (arrow) in a 5 years old child

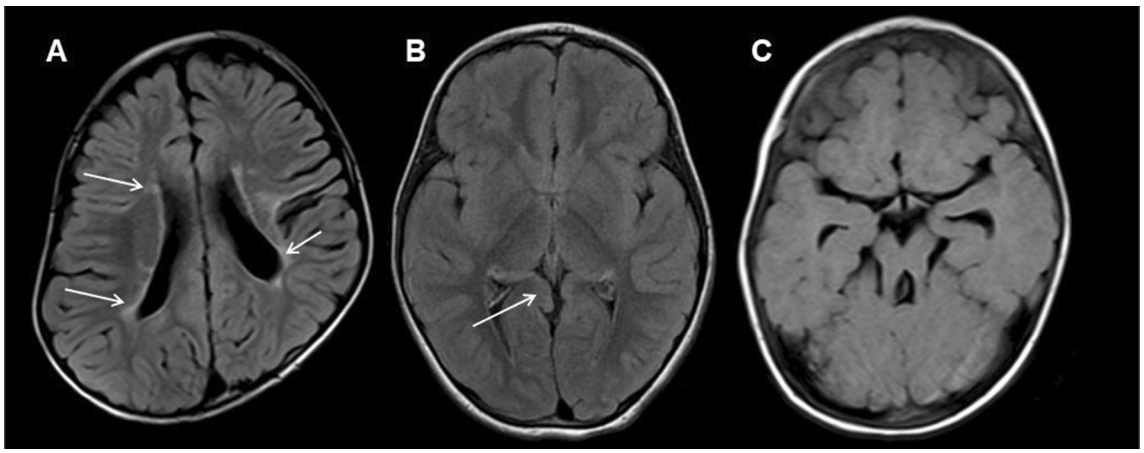

Fig. 3 **A** Periventricular leukomalacia (arrows) in a 4 years old child, **B** Right tentorial incisura hernia (arrow) in a 3 years old child, **C** Joubert syndrome in a 2 years old child

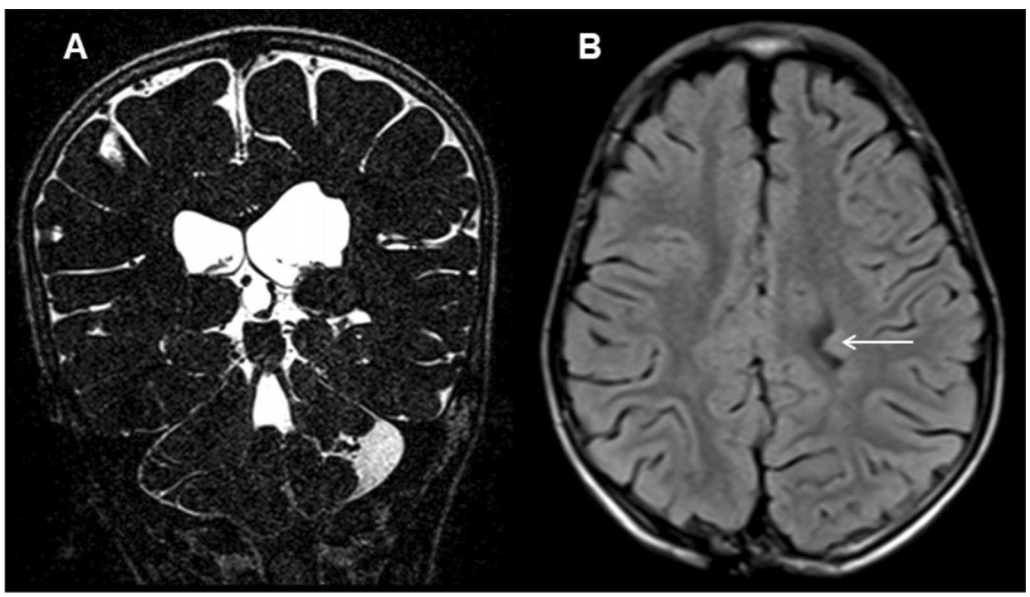

Fig. 4 **A** Left cerebellar hemiatrophy and **B** subependymal heterotopia (arrow) in a 4 years old child

Table 2 Distribution of the structural brain abnormalities in the studied population

Structural brain abnormality	Summary statistics
Single structural brain abnormality	
Arachnoid cyst	11 (2.86%)
Diverticular outpouching of foramen of Luschka	4 (1.04%)
Diffuse brain atrophy	3 (0.78%)
Chiari I malformation	3 (0.78%)
Periventricular leukomalacia	2 (0.52%)
Chiari II malformation	1 (0.26%)
Venous angioma	1 (0.26%)
Tentorial incisura hernia	1 (0.26%)
Focal cerebral encephalomalacia	1 (0.26%)
Focal dysplastic cerebral cortex	1 (0.26%)
Olivopontocerebellar atrophy	1 (0.26%)
Joubert syndrome	1 (0.26%)
Isolated lateral ventricular dilatation	1 (0.26%)
Two coincidental structural brain abnormalities	
Cerebellar hemiatrophy and subependymal heterotopia	1 (0.26%)
Cerebellar hemiatrophy and veli interpositi cyst	1 (0.26%)
Corpus callosum agenesis and heterotopia	1 (0.26%)
Blake pouch cyst and hypoplastic brainstem	1 (0.26%)
Total	35 (9.1%)

Discussion

Obtaining optimal post-CI outcome is a sophisticated status that necessitates a combination of meticulous preoperative assessment, optimal surgical performance and tailored post-implant rehabilitation programs. Successful surgery is one of the main steps that aims to lead such patients to a life closer to normal life. Actually, achieving such a goal requires the success of a number of other important elements, bearing in mind that all of these elements have the same degree of importance.

One of the main elements that requires careful evaluation before proceeding into surgery is the ability to detect any brain abnormality like WM lesions that may act as obstacle or expected to cause slow response to post-implant rehabilitation programs or requires a special type of rehabilitation program [1].

Cerebral structural or WM abnormalities are a frequent findings in SNHL patients [8]. In our study, brain abnormalities were detected in 62 patients (16.11%). This is in accordance with the incidence reported by Xu et al. (14.6%) [4], Hong et al. (18%) [5] and Lapointe et al. (20%) [6]. Higher incidence reported by Trimble et al. (40%) [14] and by Jonas et al. [7] and Walton et al. (20–56%) [15].

The WM lesions in our study were detected in 27 patients (7.01%), unilateral in 4 (1.04%), bilateral focal in 22 (5.71%) and bilateral diffuse in 1 (0.26%). Our results are in accordance with Wang et al. [8] and Archbold et al. [16] who reported WM lesions in 9.1% and 10%, respectively. Jonas et al. [7] reported WM lesions in 22%.

White matter lesions were found in 27 (43.55%) out of the 62 patients with brain abnormality in our study. The WM lesions accounted for 50% out of brain abnormalities in the studies conducted by Davis et al. [17], Fortnum et al. [18] and van Beeck et al. [19]. Higher incidence (69.6–70%) reported by Busi et al. [20], Jonas et al. [7], Xu et al. [4] and Proctor et al. [21].

In our study, 35 patients (9.1%) had other structural brain abnormalities. Arachnoid cyst was the commonest and accounted for 2.86%. Four patients had two coincidental brain abnormalities.

Lapointe et al. [6] reported that none of the patients with a brain abnormality had an inner ear abnormality

Table 3 Relation between white matter lesion and ear abnormality

Variable	No WM lesion N = 358	Unilateral WM lesion N = 4	Bilateral WM lesions N = 23	*P* value	P1	P2	P3
Rt inner ear abnormality							
Negative	267 (74.58%)	3 (75.00%)	19 (82.61%)	0.69	1	0.39	1
Positive	91 (25.42%)	1 (25.00%)	4 (17.39%)				
Lt inner ear abnormality							
Negative	272 (75.98%)	3 (75.00%)	19 (82.61%)	0.77	1	0.47	1
Positive	86 (24.02%)	1 (25.00%)	4 (17.39%)				
Inner ear abnormality							
Negative	262 (73.18%)	3 (75.00%)	19 (82.61%)	0.8	0.91	0.48	0.72
Unilateral	15 (4.19%)	0	0				
Bilateral	81 (22.63%)	1 (25.00%)	4 (17.39%)				

P value compared the three group, P1 compared none with unilateral WM abnormalities, P2 compared none with bilateral WM abnormalities, P3 compared unilateral with bilateral WM abnormalities

Table 4 Relation between ear abnormalities and brain abnormality

Variable	No ear abnormality	Unilateral ear abnormalities	Bilateral ear abnormalities	*P* value	P1	P2	P3
	N = 284	N = 15	N = 86				
WM lesion							
Negative	262 (92.25%)	15 (100%)	81 (94.19%)	0.8	0.53	0.79	0.63
Unilateral	3 (1.06%)	0	1 (1.16%)				
Bilateral	19 (6.69%)	0	4 (4.65%)				
Arachnoid cyst							
Negative	279 (98.24%)	14 (93.33%)	81 (94.19%)	0.09	0.19	0.057	1
Positive	5 (1.75%)	1 (6.67%)	5 (5.81%)				
Diverticular outpouching of Luschka foramen							
Negative	281 (98.94%)	15 (100%)	85 (98.84%)	0.92	1	1	1
Positive	3 (1.06%)	0	1 (1.16%)				
Other brain abnormalities							
No abnormality	272 (95.77%)	15 (100%)	78 (90.70%)	0.33	0.72	0.17	0.47
One abnormality	10 (3.52%)	0	6 (6.98%)				
Two abnormalities	2 (0.70%)	0	2 (2.33%)				

P value compared the three group, P1 compared none with unilateral ear abnormalities, P2 compared none with bilateral ear abnormalities, P3 compared unilateral with bilateral ear abnormalities

and none of the patients with inner ear abnormality had a brain abnormality. We have 101 patients (26.24%) in our study with ear anomalies, 5 of them (4.95%) had WM lesions and 15 (14.85%) had other structural brain abnormalities. Yet, no significant correlation was found between ear abnormalities and WM lesions or structural brain abnormalities ($P > 0.05$).

In the 27 patients with WM lesions in our study, bilateral ear abnormality found in 5 patients (18.52%). No significant correlation was found between WM lesions and ear abnormalities ($P = 0.80$). This is in accordance with Xu et al. [4] who reported inner ear anomalies in 3 (13.04%) out of the 23 patients with brain abnormalities. Teagle et al. [22] reported that brain abnormalities were more common in patients with bilateral rather than unilateral SNHL.

The high signal generated by WM lesions on MRI indicates myelin injury. This is expected to impede and slow down impulses transmission through the brain [2] and consequently may impair cognitive and neurodevelopmental progress [3, 4].

Whether WM lesions have an impact on post-CI outcome or not is still a matter of debate [4, 7, 8]. Authors reported delay in cognitive performance in children with WM lesions when compared to normal children [2]. In the contrary, others reported no significant difference between children with brain abnormalities and the control group [3]. Better improvement reported after CI in children with focal WM lesions than those with diffuse lesions. Also, SNHL patients with structural brain abnormalities got benefit from CI [4].

Given the above, we aim to highlight that the occurrence of brain abnormalities in children is not uncommon. As brain abnormalities may delay the neurodevelopmental progress in children without SNHL, it is expected to be more worse when associates SNHL, thus adding more difficulty and is expected to impede the post-CI learning outcome.

It is their rights that children with SNHL to have MRI screening of the brain as a part of pre-CI planning in order to obtain best outcomes and before the implanted electrode becomes an obstacle for post-CI imaging.

Limitations

Our study was a retrospectively descriptive study. We reported the incidence of brain abnormalities in prelingually deaf mute children. The impact and the severity of the lesions on the post-implant auditory and speech performance were not correlated. Further prospective studies to correlate the impact of the abnormal brain findings on the post-operative outcomes are recommended.

Conclusions

The incidence of brain abnormalities in children with sensorineural hearing loss is not uncommon. The neurodevelopmental progress can be more worse when sensorineural hearing loss is associated with brain abnormality. Pre-implant MRI screening of the brain helps to obtain best outcomes.

Abbreviations

SNHL	Sensorineural hearing loss
CI	Cochlear implantation
WM	White matter

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Author contributions
MH conceived the study and designed it. MH and AA contributed equally to data collection and data analysis. Manuscript was written by MH and HA. Statistical analysis done by AA. All authors have read and approved the manuscript.

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Availability of data and materials
Data will be available upon request via contacting the corresponding author.

Declarations

Ethics approval and consent to participate
The study approved by the Medical Research Ethics Committee of the Faculty of Medicine, Sohag University in Egypt in compliance with the Helsinki Declaration (DoH-oct20081).

Informed consents
Informed consents were not obtained as this was a retrospective study.

Consent for publication
Consents were not obtained as this was a retrospective study.

Competing interests
The authors declare that they have no competing interests.

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