CASE REPORT

Intisari Sains Medis 2024, Volume 15, Number 1: 387-390
P-ISSN: 2503-3638, E-ISSN: 2089-9084

Intisari Sains Medis

Published by Intisari Sains Medis

Serial case report: Radiological images of congenital sensorineural hearing loss patients with and without hypoplasia of the vestibulocochlear nerve

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Received: 2024-01-16
Accepted: 2024-03-10
Published: 2024-04-08

ABSTRACT

Background: Congenital Sensorineural Hearing Loss (SNHL) is a hearing loss caused by damage to the inner ear (cochlea), vestibulocochlear cranial nerve (N.VIII), or the innervation pathway from the inner ear to the brain that is acquired from birth. If hearing loss occurs from birth or before the period of speech development is not detected. Thus, this case report compares two patients with congenital sensorineural hearing loss with and without vestibulocochlear nerve hypoplasia.
Case Description: The first case was a 4-year-old girl with the main complaint of not hearing and responding to calls since the age of 1.5 years. Temporal CT scan and MRI results did not show existing organ and structural abnormalities, so suspected retro-cochlear auditory neuropathy. Patients undergo the installation of a cochlear implant in the left ear with good results. Case second, a 3-year-old girl with Down Syndrome who has not been able to speak since birth. CT scan results showed left and right basal cochlea 1.5 turns and bilateral jugular notch dehiscence, while MRI results showed vestibulocochlear nerve hypoplasia, narrowing of the IAC opening width 1.5 turns cochlea. If this happens, a cochlear implant cannot be done because abnormality finding is a contraindication operation.
Conclusion: CT-scan and MRI examination are necessary to evaluate the structures and organs in the outer, middle and inner ear because the presence or absence of structural abnormalities found in the patient will significantly impact further management.

Keywords: CT-scan, MRI, congenital sensorineural hearing loss, vestibulocochlear nerve hypoplasia.
Cite This Article: Priyastana, A.A.G.A., Ayusta, I.M.D., Martadiani, E.D. 2024. Serial case report:Radiological images of congenital sensorineural hearing loss patients with and without hypoplasia of the vestibulocochlear nerve. *Intisari Sains Medis* 15(1): 387-390. DOI: 10.15562/ism.v15i1.1989

INTRODUCTION

Sensorineural Hearing Loss (SNHL) is a disturbance not only hearing loss caused by damage to the inner ear (cochlea), vestibulocochlear nerve, or track innervation from the ear into the brain. This disease is the most common reason for not hearing enough permanently. Sensorineural Hearing Loss is usually influenced by age or caused by congenital abnormalities.[1]

Congenital sensorineural hearing loss is generally bilateral and is divided into severe and very severe. If hearing loss occurs from birth or before the period of speech development is not detected, it will disrupt speech, language, and cognitive development. The leading causes of SNHL in babies include congenital abnormalities, genetics, disease, or abnormalities when the child is in the womb and the birth process. The risk factors include bacterial and viral infections in pregnant women such as toxoplasma, rubella, cytomegalovirus, herpes (TORCH), and syphilis can cause hearing loss in babies, use of drugs that damage the cochlea, inflammation of the lining of the brain, and high bilirubin levels.[2]

Diagnosis of SNHL was made based on anamnesis, physical examination, and radiology imaging. Radiology imaging that became the primary choice are computed tomography (CT) scan and Magnetic Resonance Imaging (MRI). This advanced examination is perfect for evaluating congenital SNHL patients suspected of having organ or structural abnormalities in the outer, middle, and inner ear.[3]

Malformation of the outer and middle ear (congenital atresia) is divided into three grades. Malformation grade 1 is a mild abnormality of the external acoustic meatus, normal tympanic cavity or little hypoplastic, deformed ossicles, and well-aerated mastoid. Malformation grade 2 includes malformation intermediate, MAE is not formed, the tympanic cavity narrows, deformation and fixation of the ossicles, and decreased pneumatization of mastoid air cells. Malformation grade 3 covers abnormality from severe middle ear hypoplastic, severe bone hearing impairment and no pneumatization of mastoid air cells. Other abnormalities that can occur include ear fistulas in the middle ear, congenital cholesteatoma (congenital

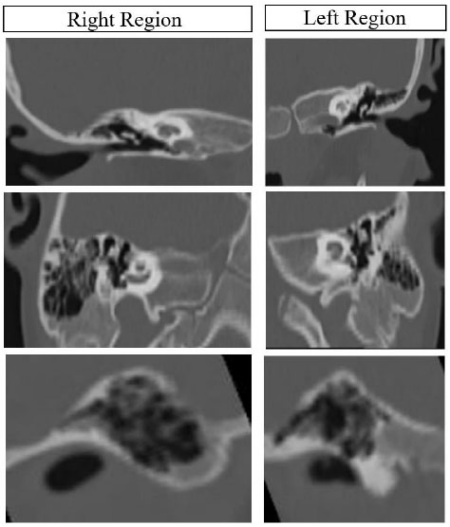

Figure 1. Axial, coronal, and sagittal sections of temporal CT-scan. There are no structural abnormalities in the outer, middle, and inner ear.

epidermoids), congenital dermoid, and malformations of muscle in the middle ear. Whereas abnormalities ear, including complete labyrinthine aplasia (Michel deformity), rudimentary otocyst, cochlear aplasia, common cavity deformity, cochlear hypoplasia, incomplete partitions, enlarged vestibular aqueduct, malformations of the semicircular canals and vestibule, and abnormalities of the vestibulocochlear nerve.[4,5] Based on CT-scan and MRI images, this case report will discuss two patients with congenital sensorineural hearing loss with and without vestibulocochlear nerve hypoplasia.

CASE DESCRIPTION

Case 1

The patient was a girl, 4 years old, with complaints of being unable to hear and respond since the patient was 1.5 years old. The patient was born by cesarean operation due to premature rupture of membranes. The birth weight was 2800 grams, and I cried immediately. Complete immunization history following age. The patient can still carry out activities according to children his age. A family history of the disease was denied. The results of the external ear examination did not reveal any abnormalities, so the next CT-scan and MRI examination

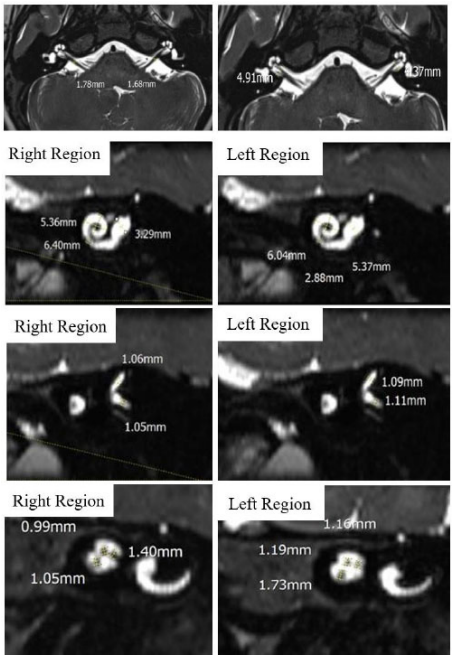

Figure 2. MRI showed no abnormalities in the basal cochlea, vestibulocochlear, or facial nerves.

were to examine the head and ear. The patient was planned for surgery to install cochlear implants in the right and left ear if no contraindications were found on radiological examination.

The results of the CT-scan examination of the head in axial, coronal, and sagittal sections (Figure 1) show no visible abnormalities in the structures of the outer, middle, and inner ear, as well as in the bones of the temporal and mastoid regions.

MRI Temporal in Axial T1WI, Sagittal T2WI, Coronal T1WI 2mm, Coronal T2WI 2mm, Axial T1 2mm, Axial T2 2mm, MIP IAC dextra et sinistra (Figure 2) showed there were no abnormal lesions or abnormalities in the bones of the temporal region, mastoid, external, middle and inner ear structures, cochlea, cochlear nerve, vestibular nerve, and right and left facial nerves. The patient underwent a cochlear implant installation in the left ear with good results. Until now, the patient was still under control for periodic evaluation and speech delay therapy.

Case 2

The patient was a girl, 3 years old, with a chief complaint of being unable to speak,

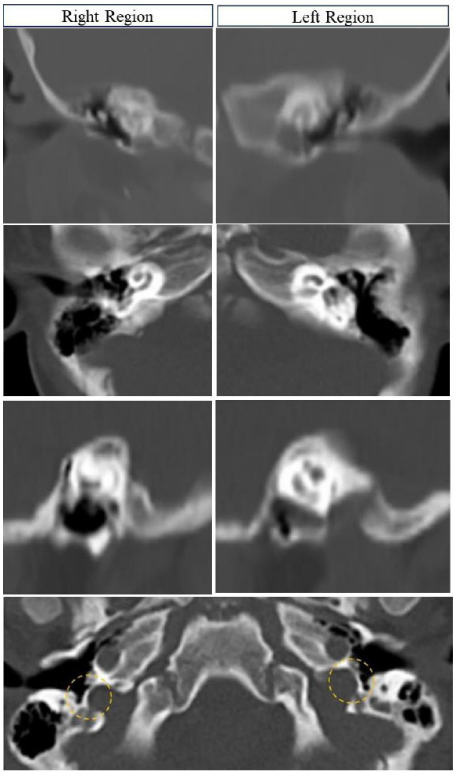

Figure 3. Axial, coronal, and sagittal CT scans showed left and right basal cochlea 1.5 turns and bilateral jugular notch dehiscence (circle yellow).

hear, and respond since birth. Patient with a history of Down Syndrome, birth by cesarean operation because of premature rupture of membrane (PROM), birth weight about 2700 grams, and did not cry immediately. History of illness or abnormality in the family was denied. Immunization history was complete by age. Outer ear examination showed no abnormality.

Results of CT-scan examination of the head in axial, coronal, and sagittal sections (Figure 3) showed abnormality in the right and left basal cochlea 1.5 turns accompanied by bilateral jugular notch dehiscence. MRI Temporal in Axial T1WI, Sagittal T2WI, Coronal T1WI 2mm, Coronal T2WI 2mm, Axial T1 2mm, Axial T2 2mm, MIP IAC dextra et sinistra showed vestibulocochlear nerve hypoplasia accompanied by a narrowing of the right and left IAC opening width and 1.5 turns on the right and left cochlea (Figure 4).

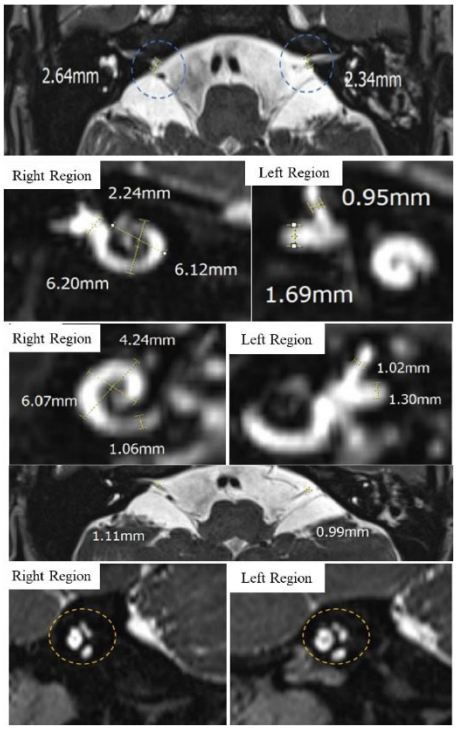

Figure 4. MRI showed the IAC opening width narrowing on the right and left (blue circle). No visible vestibulocochlear nerve structures in the right and left IAC (yellow circles).

DISCUSSION

Congenital SNHL can be caused by various factors, such as the presence of a tumor or mass in the cerebellopontine angle (CPA), infection or inflammation in the vestibular area, or congenital abnormalities such as aplasia and hypoplasia of the middle and inner ear structures.[6,7]

CT-scan and MRI examination of the patient in the first case showed no structural abnormalities in the outer, middle, or inner ear. The patient also did not have a history of trauma, auto-immune disease, metabolic disease systemic infection, or the use of ototoxic drugs, so it was suspected that the cause of SNHL was retro-cochlear auditory neuropathy. Management of congenital SNHL due to retro-cochlear auditory neuropathy can include the placement of a cochlear implant. The patient had a cochlear implant installed in the left ear with satisfactory results. The patient in the second case is a child, a 3-year-old girl with a chief complaint, who cannot speak since birth and has a history of Down Syndrome. No abnormalities on examination of the outer ear. Temporal CT-scan results show right basal cochlea left at 1.5 turns and bilateral jugular notch dehiscence. In contrast, temporal MRI results showed bilateral hypoplasia of the vestibulocochlear nerve accompanied by a narrowing of the IAC opening width and 1.5 turns of the cochlea.

Due to the vestibulocochlear nerve's formation during the embryonic period, patients with congenital diseases, including Down syndrome, may have developmental issues in this nerve. From the third to the 24th week of pregnancy, there are several phases in which the embryological development of the inner ear occurs, with specifics revealed at each. The process starts with localized thickening of the ectoderm (otic placode). The statoacoustic ganglion cells follow the otic placode, which emerges from the ectoderm's surface on each side of the rhombencephalon in the third week of gestation. Otic placodes invade during the fourth week of pregnancy, resulting in otic pits. By the end of the fourth week, these otic cysts, also known as otocysts, auditory vesicles, and the statoacoustic ganglia, are formed. The vestibular portion and the labyrinthine cochlea result from the otocyst's division into the dorsal utricular and ventral saccular sections in the fifth week. The semicircular canal (SSC) superior, posterior, and lateral canals begin to develop with statoacoustic ganglion divides.[8–10]

The anterior side of the cochlea lengthens, and the membranous cochlea starts to develop near the conclusion of the sixth week of pregnancy. With the saccule and utricle present, the SCC is fully formed. After the seventh week of gestation, a macula is present. The membrane cochlea runs about 2.5 turns, and the cochlear duct displays sensory ridges. Beginning at the base of the cochlea and progressing towards the apex is the process of cochlea lumen development. The vestibular labyrinth's fluid secretion regulates these alterations in part. The cochlea had 1.5 twists at the conclusion of the eighth week of pregnancy. The ductus reuniens connects the saccule to the utricle during the nine-week gestation. Cartilage is formed by the mesenchyme that surrounds the cochlear duct. A portion of cartilage remains between the scala tympani and scala vestibuli, forming the modiolus; epithelial cells start to differentiate into sensory cells of the organ of Corti. At the tenth week of gestation, the cochlea almost reaches its adult form and reaches 2.5 turns. The cartilage surrounding the cochlea experiences vacuolization to form the scala tympani and scala vestibuli; the original cochlear duct forms the scala media, The cochlear duct's epithelium begins to thicken between weeks 11 and 24 of pregnancy, while the labyrinth is still developing. Week 16 indicates the beginning of the otic capsule's ossification; Week twenty coincides with the membranous labyrinth's maximal size and the cochlear duct's most significant length; Ossification of the perilymphatic space and otic capsule is finished (weeks 23–24).[8–10]

By the theory, the patient in the second case probably experienced impaired embryological development, especially from the 5th to 10th week of pregnancy, because the cochlea was only 1.5 turns in rotation accompanied by vestibulocochlear nerve hypoplasia and bilateral narrowing of the IAC opening width. In this patient, installation of a cochlear implant cannot be done because these findings are contraindicated for operation.

The limitations of this case report can be seen in the first case, where the leading cause of congenital SNHL was not known through CT scan or MRI examination, so retro-cochlear auditory neuropathy was suspected. Then further tests such as ABR (Auditory Brainstem Response) or BERA (Brainstem Evoked) were needed. The strength of this study is that CT scans and MRI examinations are essential to evaluate the structure of the patient's outer, middle, and inner ear because the presence or absence of structural abnormalities plays a critical role in selecting further treatment.

CONCLUSION

The hypoplasia of the vestibulocochlear nerve narrowing of the IAC opening width bilaterally and 1.5 turns of the basal cochlea are rare diseases and available cause sensorineural hearing loss and congenital deafness. These diagnoses have

been considered necessary before surgery for candidates for cochlear implants. Head CT-scan and MRI examination Temporal Focus is essential in diagnosing SNHL and congenital deafness cases.

INFORMED CONSENT

Informed consent was obtained before the procedure, including permission to publish all photographs and images.

CONFLICT OF INTEREST

When creating this case series, the writers declared no conflicts of interest.

FUNDING

The author provides funds to write this case series. No other funding source is involved.

AUTHOR CONTRIBUTION

From the stage of case discovery to the interpretation of the radiography data and the clinical outcomes acquired, all authors have contributed equally to this case report, published in a scholarly journal.

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