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Congenital sensorineural hearing loss due to Inner ear malformations: Spectrum on CT and MRI

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Abstract
Introduction
Inner ear malformations is an important cause of congenital sensorineural hearing loss .Cross sectional imaging plays a crucial role not only in the diagnosis but also for preoperative evaluation in candidates deemed fit for cochlear implant.
Aim
To study the various spectrum of SNHL on HRCT temporal bone and MRI cochlea.
Material And Methods
A prospective study was conducted to study the spectrum of cases in 30 patients evaluated over a period of 2 years.
Results
Out of 30 patients, 18 were females and 12 were males with a mean age of 4 years. All patients presenting with congenital SNHL were evaluated with HRCT/MRI or both . Audiometry and BERAtestswere also performed in all cases. Around 2/3rd of the cases were positive for varying types of inner ear malformations as per the Sennaroglu classification.
***Conclusion** :*
As imaging plays a key role in the evaluation and management of congenital SNHL ,knowledge about the anatomy and classification is a must for accurate diagnosis .
***Key words** – SNHL,IP type 1,2,cochlea,VA*

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I. Introduction

Inner ear malformations spectrum is an important cause of congenital SNHL which itself is a major contributor of childhood disability. Cross-sectional imaging plays a vital role in the evaluation not only for diagnosing the cause but also for choosing candidates for cochlear implant .CT provides intricate details about the osseous anatomy and inner ear bony malformation .When performed preoperatively, CT also provides other details like width of the cochlear aperture and/or associated external ear and middle ear abnormalities,integrity of ossicularchain and presence of anatomic variants like high riding /dehiscent jugular bulb.MRI complements CT by providing details about membranous labyrinth and cochlear nerve evaluation which is crucial before cochlear implant procedure.The cochlear nerve can sometimes be absent even in the presence of normal IAC and bony labyrinth.(1)

II. Material And Methods

This was a prospective study conducted in the radiology department of a tertiary health center. Inclusion criteria included patients with congenital SNHL sent for imaging while exclusion criteria included those who had causes other than congenital /inner ear malformations. All patients with congenital SNHL were evaluated with HRCT/MRI or both also including T2WI axial screening of the brain to evaluate brainstem or any intracranial abnormality. Audiometry and BERA tests were also performed in all cases

III. Results with tables and images

Out of 30 patients, 18 were females and 12 were males with a mean age of 4 years. Around 2/3 rd of the cases were positive for varying types of inner ear malformations as per the Sennaroglu classification
Spectrum of congenital inner ear malformations classified as per Sennaroglu and Saatci(2)

Time of Insult (week)	Malformation	Features
Third	Labyrinthine aplasia (Michel deformity)	Complete absence of cochlea and vestibule
Third-to-fourth	Cochlear aplasia	Complete absence of cochlea; vestibule present
Fourth	Common cavity	Single cystic cavity representing cochlea and vestibule, without any differentiation
Fifth	Cystic cochleovestibular anomaly (IP-I)	Cystic-appearing cochlea lacking entire modiolus and cribriform area; large cystic vestibule
Sixth	Cochlear hypoplasia	Cochlea and vestibule are separate but are smaller than normal; hypoplastic cochlea resembles small bud off the IAC
Seventh	Incomplete partition type II (IP-II)	Cochlea consists of 1.5 turns, in which middle and apical turns coalesce to form a cystic apex; vestibule and VA may be enlarged

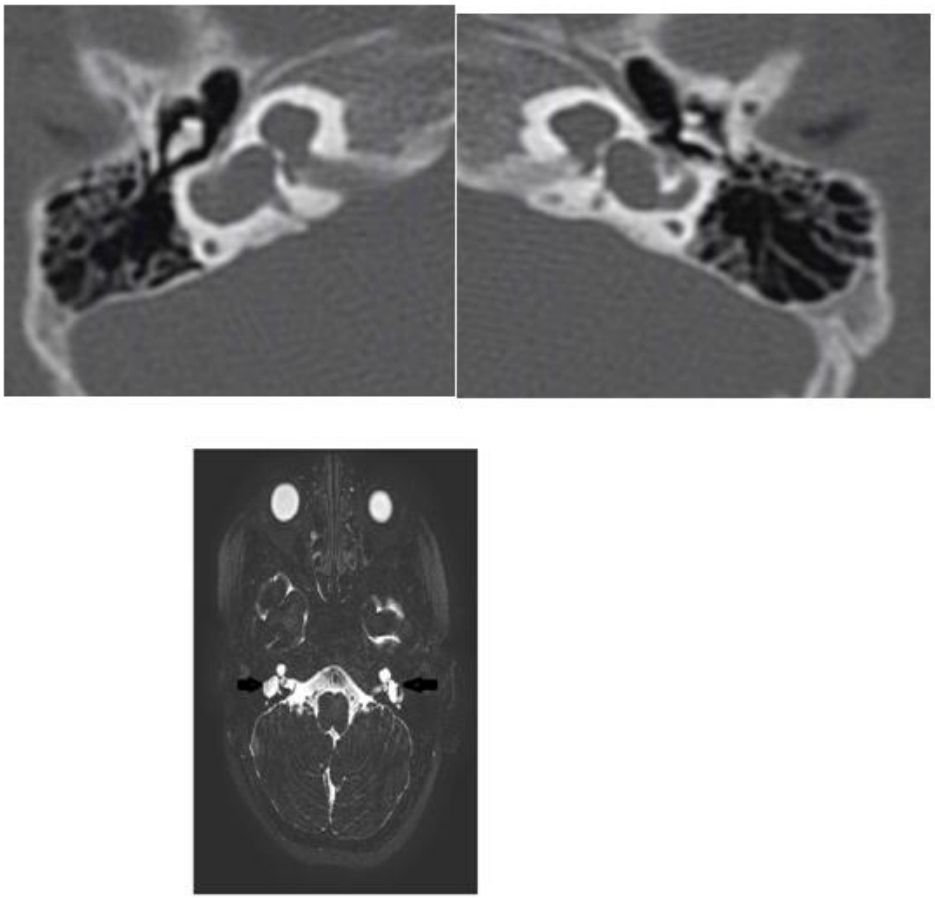

Figure1,2 and 3 :3D CISS axial MR sequence and HRCT temporal bone reveals bilateral cystic dysplastic cochlea and vestibular aqueduct consistent with Incomplete partition type 1 malformation.

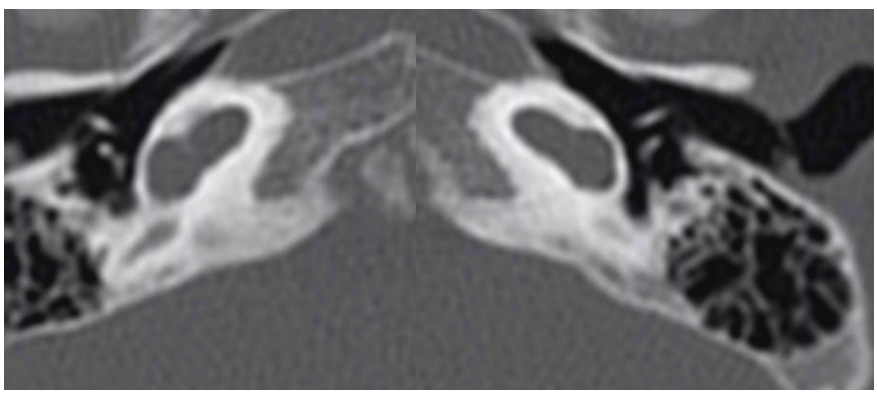

Figure 4,5 reveals HRCT images of bilateral common cavity malformation.

Figure 6:3D CISS axial MR image reveals fusion with cystic dilatation of middle and apical turns of left cochlea with markedly attenuated modiolus consistent with incomplete partition (IP II) congenital malformation.

IV. Discussion

The various anomalies in inner ear malformation spectrum include:

a) Complete labyrinthine aplasia:This is characterized by the complete absence of inner ear structures. HRCT reveals narrow, atretic IAC while MR confirms absence of cochlear nerve [(3)]

b) Cochlear aplasia: Dense otic bone is noted at the expected site of cochlea while the vestibule and semicircular canals may be dysplastic or normal. The absence of the bulge of the cochlear promontory(producedby the basal turn of the cochlea)helps to differentiate it from labyrinthitis ossificans .

c)Common Cavity.— There is confluence of the cochlea and vestibule forming a cystic cavity with no internal architecture The width of the cavity (10mm)is typically greater than its height(7mm)

d)Type I Incomplete Partition (cystic cochleovestibular malformation): This is characterized by absent modiolus; the cochlea has a cystic appearance. The vestibule is often dilated, forming a figure of eight [(4)]The presence of a separate vestibule differentiates this condition from a common cavity malformation. The vestibular aqueduct is normal.

e)Type II Incomplete Partition- known as Mondini deformity ,this is the most common type of cochlear malformation in our study .In this malformation the cochlea consists of 1½ turns, the basal cochlear turn is normal but the middle and apical turns coalesce to form a cystic apex. The modiolusis seen only at the level of the basal turn. It presents with a triad consisting a normal basal turn and cystic apex of cochlea, enlarged vestibular aqueduct and vestibule with normal semicircular canals [(4)]

f) Varying degrees of malformation of semicircular canals and vestibular aqueduct may be noted.

g)Enlarged vestibular aqueduct- CT reveals enlargement of the bony vestibular aqueduct,(more than 1.5 mm) while in MRI there is enlargement of the endolymphatic duct and sac [(5)] This is a crucial finding in preoperative imaging as it carries a risk of CSF gusher at the time of surgery.

h) IAC and Cochlear Nerve Anomalies.—The normal diameter of the IAC ranges from 2 to 8 mm(average 4 mm), diameter of less than 2 mm is considered stenotic [(1)]

i) T2WI screening of brain is mandatory to evaluate for brainstem or any neuroparenchymal abnormality.

V. Conclusion

Imaging plays a very crucial role in the evaluation of congenital SNHL .A thorough knowledge of the anatomy and classification of inner ear malformations is indispensable for the accurate diagnosis, which has a vital role in the management and prognosis.

Abbreviations

SNHL	Sensorineural hearing loss
VA	Vestibular aqueduct
IAC	Internal auditory canal
IP	Incomplete partition
BERA	Brainstem evoked response audiometry.

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