



Congenital hearing loss. Is CT enough?



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Abstract The aim of this study is to evaluate the value of high resolution CT scan (HRCT) in diagnosis and management of congenital hearing loss.

Patients and methods: This is a prospective study including 60 patients, 24 males and 36 females aged from 1 to 7 years, who were presented by unilateral or bilateral congenital conductive (CHL) or/and sensorineural hearing loss (SNHL). All patients were evaluated by HRCT scan with post-processing multiplanar reconstruction (MPR). Only three Results: External auditory canal atresia (EACA) was diagnosed in 10 patients, while both EACA and middle ear anomalies were diagnosed in another 6 patients. Variable inner ear anomalies were diagnosed in the patients with SNHL. One patient with SNHL had cochlear nerve aplasia that was missed by HRCT and was diagnosed by MRI, in addition to the CT diagnosed vestibulocochlear anomalies.

Conclusion: Although HRCT scan can be used as a solitary diagnostic imaging tool for diagnosis of pure CHL, combined CT and MRI examination is crucial for preoperative evaluation of SNHL patients.

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1. Introduction

Congenital hearing loss is one of the developmental disorders that may be not clearly identifiable at birth. Delayed identification and lack of appropriate early management of hearing loss in children have potential impact on child educational, cognitive and social development. The goal of early diagnosis and correction of congenital hearing loss is to maximize perception of speech and the resulting attainment of linguistic-based skills.

2. Physiology of hearing

Hearing is a complex physiological process, which is mediated through interaction of many fine structures. External auditory

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canal (EAC) is the first station; it serves as collecting apparatus that collects sound energy focusing it on tympanic membrane. The tympanic membrane transmits this sound energy as vibrations, through the ossicles of middle ear. Middle ear is a conductive way that transmits the sound vibrations to the inner ear through foot plate of stapes in the oval window. The vibrating foot plate causes fluid motion in the membranous labyrinth of the inner ear. Finally, the inner ear cochlea is the converter operator, which converts these vibrations into nerve impulses. These impulses are transmitted by the cochlear nerve, through the brain stem into the cerebral auditory area, which is located in the temporal lobe.^{1,2}

3. Embryology of the ear

EAC develops from the ectoderm of the first branchial groove, where epithelial cells proliferate forming a solid core of cells, known as meatal plug. This solid core will soon recanalize to form the epithelial lined EAC. Middle ear develops from the endoderm of the first branchial pouch and maintains its pharyngeal communication through the auditory (Eustachian) tube. First and second pharyngeal arches mesoderm are responsible for ossicles' generation. The first arch is precursor of epitympanic ossicles that are head of malleus, body and short process of incus. The second arch forms the mesotympanic ossicles that are long process malleus, long process incus, stapes head, neck and crura, while stapes footplate is formed from the otic capsule.³

Inner ear begins to develop at the 4th week of gestation, as localized ectodermal thickening in the developing embryo known as otic placode. The otic placode subsequently invaginates to form otocyst. At the fifth week, a diverticulum buds from the otocyst, forming the endolymphatic sac, followed by the cochlea and vestibule. The membranous cochlea achieves 1–1.5 turns at the end of 6 weeks, while 2.5 turns are formed at the end of the 7th week. The semicircular canal develops from the otocyst at 7–8 gestational weeks. Full embryological development of inner ear structures occurs at the end of 8 weeks.^{3,4}

4. Incidence of congenital hearing loss

Records of prevalence of congenital hearing loss vary, depending on the criteria used to define the different degrees of hearing loss and the characteristics of the studied population. Congenital hearing loss may be conductive and/or sensorineural, unilateral or bilateral, symmetrical or asymmetrical, progressive or stable. In some published studies,⁵ profound congenital hearing loss is estimated to occur in approximately 1 in 1000 births. In other researchers' records, the incidence of congenital bilateral hearing loss (greater than 40 dB) was 2–4 per 1000 live births, in developed countries. In developing countries, the incidence was estimated to be not less than 6 per 1000 live births.⁶

5. Etiology

Congenital hearing loss may occur as a result of genetic or non-genetic factors. Non-genetic factors (35%) may be due to maternal infections, such as rubella or cytomegalic or herpes simplex viruses, prematurity, low birth weight, birth injuries, toxins including drugs and alcohol consumed by the mother during pregnancy, complications associated with the Rh factor incompatibility, maternal diabetes, toxemia during pregnancy and perinatal anoxia.⁷

Hearing loss due to genetic factors (more than 50%) can present at neonatal period or develop later on childhood period. This type may be due to autosomal recessive or autosomal dominant inheritance. Some other uncommon types are inherited via X-linked roots. There are many genetic syndromes that include hearing loss as one of their symptoms e.g. Down syndrome, Treacher Collins syndrome, Crouzon syndrome, Alport syndrome, Klippel-Feil, Norrie disease and Waardenburg syndrome.⁷

6. Role of imaging

Early diagnosis through a clear accurate imaging tool is a cornerstone for management planning. It guides the selection of candidates, who will gain benefits from hearing aid implants or corrective surgeries. The advent of high resolution multislice computed tomography (HRCT) scan of temporal bone was expected to play a very valuable role in preoperative assessment and postoperative evaluation. Post processing 2D and 3D multiplanar reconstruction is the discerning gain of multislice CT application in temporal bone imaging.⁸

7. Radiological anatomy of the ear

As known, the ear consists of external, middle and inner parts. External ear is formed of the auricle and EAC, which is divided into membranous and bony channels, while middle ear is formed of tympanic cavity and the antrum. The antrum is the air space located superior and posterior to the tympanic cavity having a communicating channel with the epitympanic recess called aditus ad antrum. The middle ear space contains three articulating ossicles (malleus, incus and stapes); that-together with the tympanic membrane-play the major role in sound wave conduction. These conducting structures transfer sound waves to the stapes footplate, which lies in contact with the base of the cochlea in the oval window.^{9,10}

The inner ear consists of the osseous labyrinth which encloses the membranous labyrinth. The osseous labyrinth

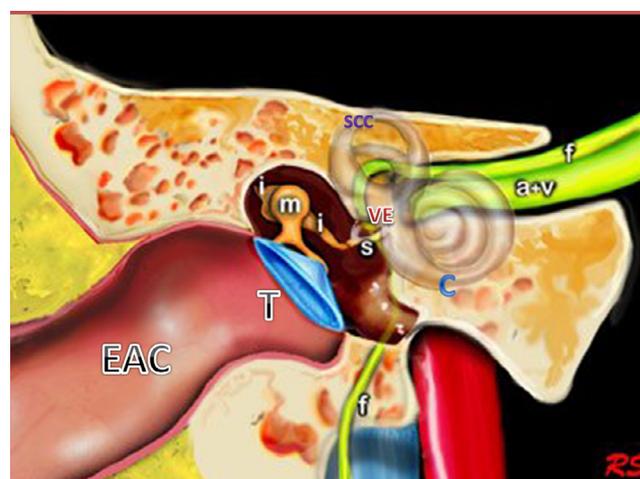


Figure 1 Coronal model of petrous bone showing different parts of the ear: EAC: external auditory canal, T: tympanic membrane, i: incus, m: malleus, S: stapes, SSC: semicircular canal, C: cochlea, f: facial nerve, a + v: auditory and vestibular nerves, VE: vestibule. Bee.¹⁰

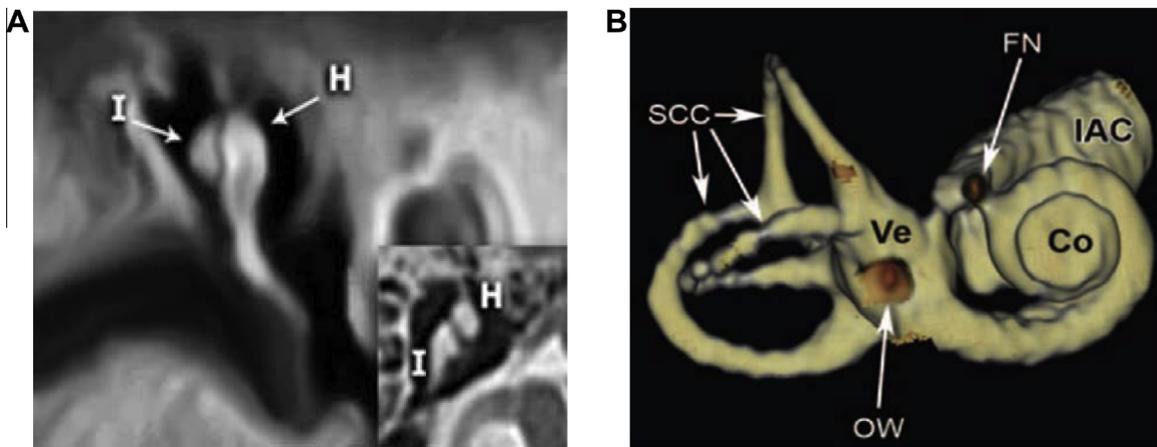


Figure 2 A-Coronal reconstruction shows incus (I) is positioned posterolateral to the malleolar head (H). B- Normal anatomy of the bony labyrinth. Three-dimensional VR CT image (anterolateral view) shows the normal bony labyrinth, which consists of the cochlea (Co), vestibule (Ve), and semicircular canals (SCC). FN = facial nerve canal, IAC = internal auditory canal, OW = oval window. Bee.¹⁰

?> consists of the vestibule, cochlea, semicircular canals, vestibular aqueduct, and cochlear aqueduct. There is a space between the osseous labyrinth and membranous labyrinth, which is filled with perilymph, in contrast to the membranous labyrinth which is filled with endolymph. The vestibule is the central rounded portion of the osseous labyrinth, which has communication with the cochlea anteriorly, while it communicates the semicircular canals and the vestibular aqueduct posteriorly. It envelopes utricle and saccule, that are parts of the membranous labyrinth. The vestibular aqueduct is a thin linear tube, which contains the endolymphatic duct and sac and communicates the utricle and saccule, it measures 1.5 mm in

diameter. The semicircular canals are three canals; superior, lateral and posterior. The cochlea consists of a spiral canal that has 2½ turns around a central column of bone (modiolus). The cochlear aqueduct is a thin osseous canal just inferior and parallel to the internal auditory canal (IAC) (Figs. 1 and 2).^{10,11}

8. Patients and methods

This is a prospective temporal bone HRCT scan study, for 60 patients presented with congenital hearing loss, in AL-Mana General Hospital – Saudi Arabia (AGH), from March 2010

Table 1 Patients’ age and sex.

Age-group	Male	Female	Total (patients)	Gender
1–3 years	6 (10%)	13 (21.7%)	19 (31.7%)	
3–5 years	7 (11.7%)	8 (13.3%)	15 (25%)	
5–7 years	11 (18.3%)	15 (25%)	26 (43.3%)	
Total (patients)	24 (40%)	36 (60%)	60	

Table 2 Type of the deficits.

Type	CHL		SNHL(IE)
	EAM	EAM and ME	
No. of patients (60)	10 (16.6%)	6 (10%)	44 (73.3%)
Mean age (years)	1.1		4.3
P-value	0.033		

CHL: conductive hearing loss, SNHL: sensorineural hearing loss, EAM: external auditory meatus, ME: middle ear, IE: inner ear.

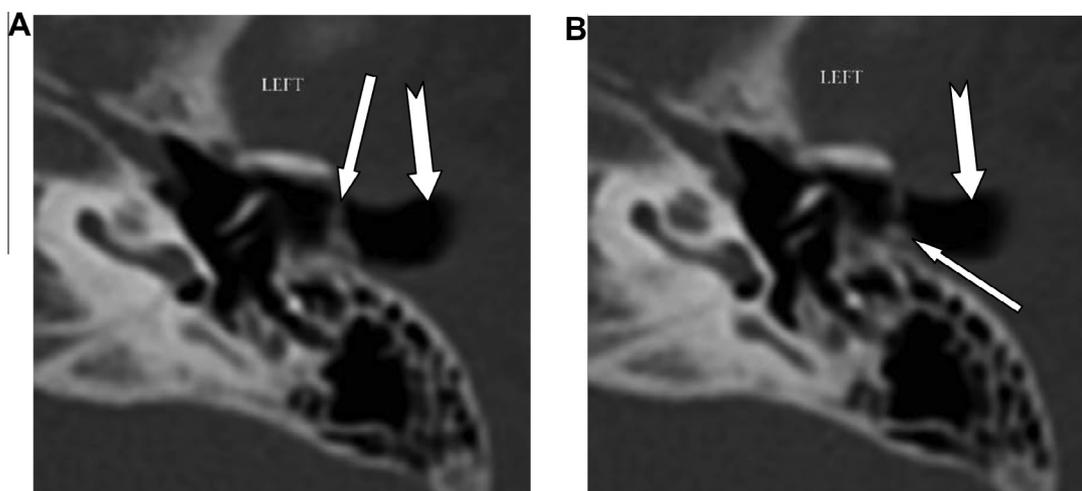


Figure 3 A and B: Axial HRCT scan showing external auditory atretic bony plate (arrow), with normal looking membranous EAC (notched arrow), tympanic membranes and middle ear cavity and ossicles, inner ear and mastoid aeration (High Jahrsdoerfer score = 9).

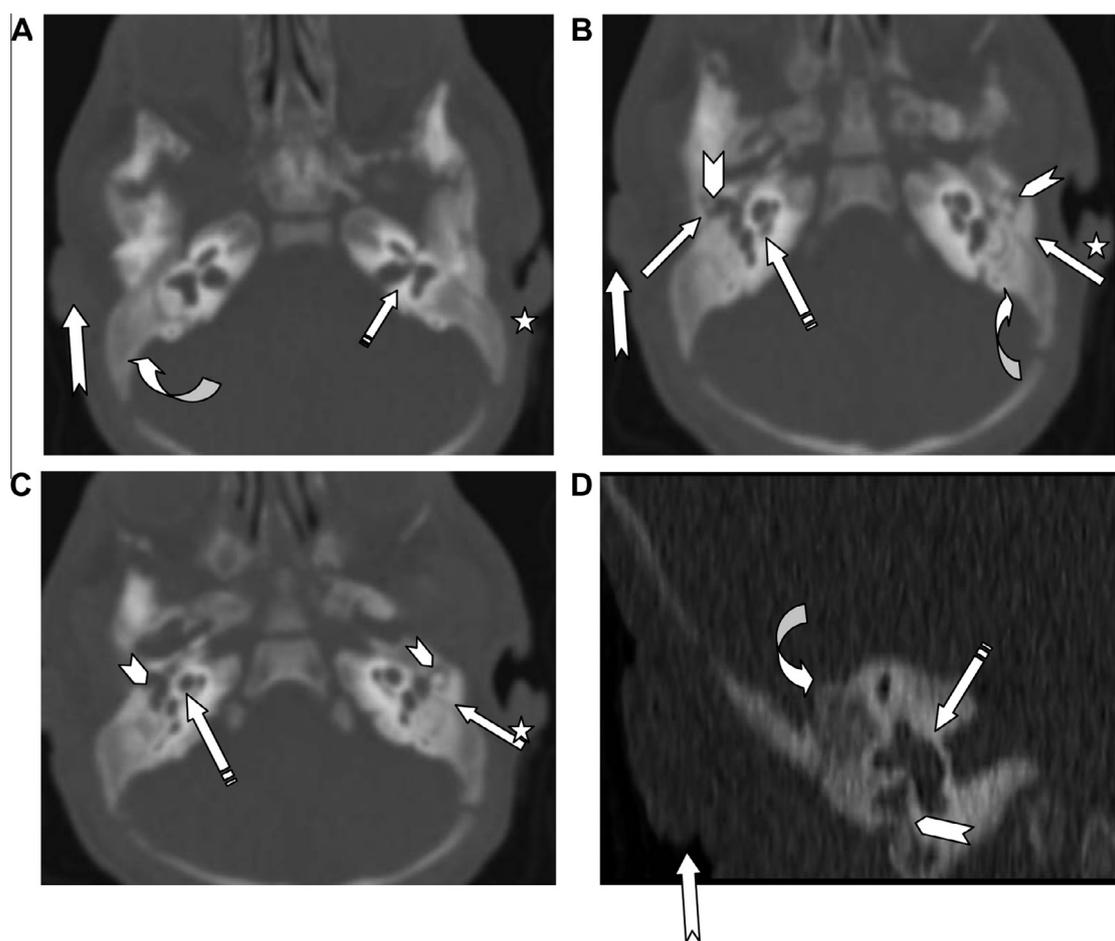


Figure 4 HRCT scan: A–C: axial cuts; D: coronal reformat of right ear; showing bilateral mixed external auditory canal atresia (arrows) with deformed right pinna (notched arrow) and small middle ear cavities with hypoplastic deformed ossicles, more severe on the right side (chevrons), non-pneumatized mastoids (curved arrows), normal left ear pinna (star) and normal inner ear structures (striped arrows).

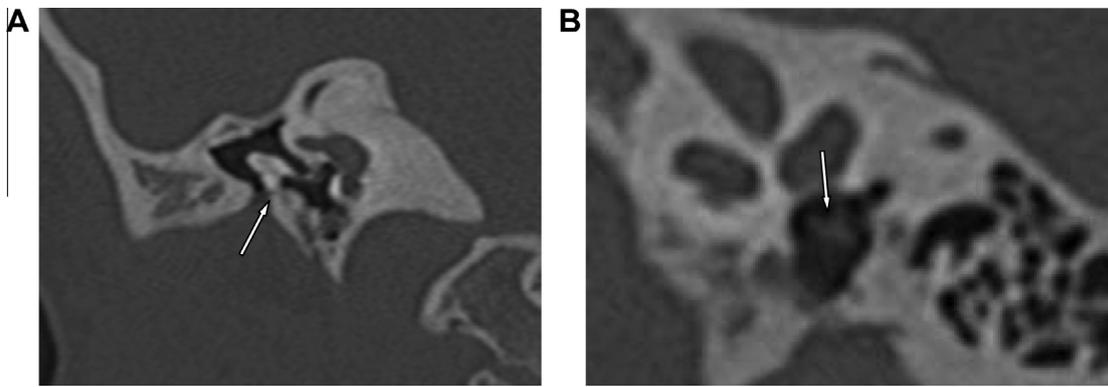


Figure 5 HRCT scan petrous bone coronal reformat showing: (A) external auditory meatus bony atresia with short handle of malleus fused to the atretic plate (arrow). (B) Magnified view showing: Dysplastic stapes with short anterior crus disconnected from the foot plate (arrow).

to November 2012. The study protocol was approved by the scientific and ethics committee of the hospital, with signed consents from all study candidates' parents or responsible relatives. First, audiometry studies were performed in audiology department and interpreted by experienced audiologists under the supervision of the Otolaryngology author. All CT images were interpreted, reported and adopted with consensus of the three radiology authors.

Reports were edited carefully fulfilling standardized check list formula, covering all possible affected parts of different ear segments (external, middle and inner). This reporting formula represents expansion of the standardized international scoring system developed by Jahrsdoerfer et al.,¹² that was widely applied for external and middle ear anomalies, in attempt for obtaining guidelines for management of these anomalies. These guidelines help selection of candidates with high scores, who were in need for surgical repair with expected good prognostic values.

Scans were made on a 64-section spiral CT scanner (Somatom Sensation; Siemens Medical Solutions, Malvern) and Toshiba Aquillion CX 128 (Toshiba Medical Systems Europe B.V., Zilverstraat 12718 RP Zoetermeer – The Netherlands).

The CT scan studies were initially done with axial cuts, using parameters of: 120 kV, 180 mAs, 1-s rotation time, 0.5-mm section thickness, 0.5-mm collimation, 0.2 reconstruction increment, 1-mm table feed and rotation, high resolution algorithm with 512 × 512 matrix, and 9-cm field of view. Bismuth eye shield was routinely used for all patients (AttenuRad; F&L Medical Products Co, USA) for lens protection.

Only three patients who got profound sensorineural hearing loss did additional MRI petrous examination under anesthesia, before cochlear implant procedure. MRI examinations were done through MRI Philips Intera 1.5 T, Closed Magnet, Philips. Medical Systems, 5656 AE Eindhoven – Netherlands.

9. Imaging post-processing

Axial images were transferred and stored through the RIS (Radiology information system) and PACS (Picture archiving and communication system). In the radiology department reporting room, post-processing was done through work station Osirix software (Osirix Foundation California, USA). Multiplanar reconstruction was made to evaluate the different

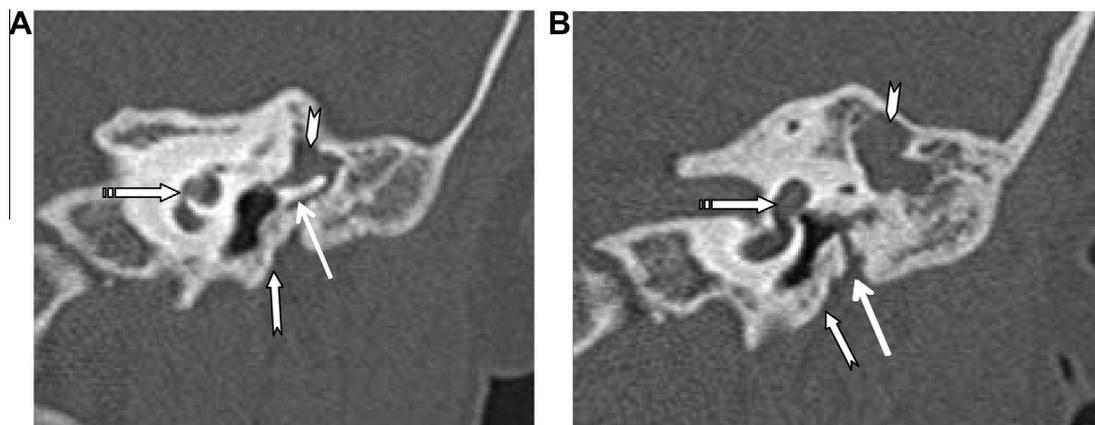


Figure 6 Coronal reformat HRCT temporal bone of patient with combined left external and middle ear anomalies with low Jahrsdoerfer score: (A) Atretic bony plate (notched arrow), hypoplastic tympanic cavity with opacified epitympanum (chevron), malleuincudal fusion (arrow). (B) External auditory atretic plate (notched arrow), aberrant mastoid segment of facial nerve canal (arrow), non-pneumatized mastoid air cells (chevron). Normal inner ear structures are noted (stripped arrow in A and B).

segments and fine structures of the ossicular chain, stapedial footplate, oval and round windows, cochlea, vestibular aqueduct, cochlear aqueduct, semicircular canals and facial nerve and skull base vessels. Evaluation of these structures in the three orthogonal planes (axial, coronal, and sagittal) limits missing of subtle anomalies. For obtaining perfect reconstructed images, the axial cuts were initially done with 1-mm section thickness and an overlap of 0.5 mm and images were displayed at average window values of 800 HU and a window width of 4000 HU.

10. Results

The study included 36 female and 24 male children with age 1–7 years; 19 (31.6%) patients (1–3 y), 15 (25%) patients

Table 3 Detailed CT findings. CT scan findings of different affected segments of external, middle and inner ears in patients with conductive or/and hearing loss (60 patients).

Structure	CT description	No. of Patients
EAC	Normal	44 (73.3%)
	Membranous	4 (6.7%)
	Bony or mixed atresia	12 (20%)
Size of the tympanic cavity	Good	50 (83.3%)
	Moderate	4 (6.7%)
	Poor	6 (10%)
Aeration of mastoid	Good	50 (83.3%)
	Moderate	4 (6.7%)
	Poor	6 (10%)
Malleus and Incus	Normal	54 (90%)
	Dysplastic	5 (8.3%)
	Absent	1 (1.7%)
Stapes	Normal	59 (98.3%)
	Dysplastic	1 (1.7%)
	Absent	0 (0%)
Oval window	Open	59 (98.3%)
	Closed	1 (1.7%)
Round window	Open	60 (100%)
	Closed	0 (0%)
Facial nerve	Normal	56 (93.3%)
	Slightly aberrant	3 (5%)
	Markedly aberrant	1 (1.7%)
Arteries and veins	Normal	58 (96.6%)
	Slightly aberrant	1 (1.7%)
	Markedly aberrant	1 (1.7%)
Cochlea	Normal	32 (53.3%)
	Hypoplasia	28 (46.7%)
	Aplasia	0 (0%)
Vestibule	Normal	41 (68.3%)
	Deformed	19 (31.7%)
	Aplasia	0 (0%)
Semi-circular canals	Normal	42 (68.3%)
	Deformed	18 (31.7%)
	Aplasia	0 (0%)
Vestibular aqueduct	Normal	23 (38.3%)
	Ectasia	37 (61.7%)
Total		60

(3–5 y) and 26 (43.3%) patients (5–7 y) (Table 1). 10 patients (16.6%) were presented with conductive hearing loss (CHL), due to EAC atresia, while, another 6 patients (10%) were suffering from CHL due to mixed EAC and middle ear anomalies. 44 patients (73.3%) were complaining of sensorineural hearing loss (SNHL) due to inner ear different part anomalies. Children with CHL were presented at mean age of 1.1 years, in comparison with children with SNHL who were presented at mean age of 4.3 years with significant mean age susceptibility difference ($P < 0.005$). (Table 2)

External auditory canal atresia (EACA) was purely found in 10 patients (16.7%) (Fig. 3), while combined EACA and middle ear anomalies were diagnosed in six patients (10%) (Figs. 4–6) (Table 2). The middle ear anomalies were mainly expressed in the form of ossicular dysplasia/aplasia, however additional important items should be fulfilled in the report. These items include size and aeration of the tympanic cavity, aeration of mastoid cells, any aberrant courses of the facial nerve (found in one patient (Fig. 6)) or the skull base vessels and the degree of development of the round and oval windows (Table 3). In SNHL patients – 44 (73.3%) – the findings detected by CT scan were semicircular canal dysplasia or hypoplasia, vestibular aqueduct dilatation, vestibular and cochlear abnormalities, seen in 30%, 63.3%, 31.6%, and 46.6% of patients, respectively. These different inner ear anomalous parts were presented separately or in different combinations. (Table 3, Figs. 7–9).

Using Jahrsdoerfer's scoring system we classified the severity of conductive hearing loss anomalies in our study, into severe with score < 5 in 3 patients (5%), intermediate with score 5–8 in 4 patients (6.7%) and mild with score > 8 in 9 patients (15%) (Table 4).

Comparative data analysis of the different groups was done through the online website (<http://www.graphpad.com/quick-calcs/contingency1.cfm>), with application of Fisher's exact test to estimate P -values that reflect the susceptibility difference of the study groups. P values less than 0.05 are consistent with statistically significant difference.

11. Discussion

Congenital hearing loss is not uncommon congenital anomalies, if not early diagnosed and perfectly managed; it will impede the child speech, language, and cognitive development. This study was planned to check the diagnostic utility of HRCT scan as an imaging tool, in diagnosis and management of children with congenital hearing loss. As expected, children with SNHL were presented at significantly later age groups in comparison with CHL patients. This may be explained by the alarming auricle deformity or the membranous atresia, easily noticed in patients with aural atresia.¹³

In patients with congenital aural atresia, regardless of the fact that clinical examination is diagnostic for absent external auditory canal and associated microtia, the important role of CT scan examination before surgical repair is the evaluation of the middle ear structures to exclude associated anomalies, that were detected in six patients (10%). So, HRCT scan can perfectly differentiate if it is only external auditory atresia (Fig. 3), or associated with anomalous middle ear structures (Figs. 4–6). This could be simply explained by the common embryological branchial origin of these structures. External

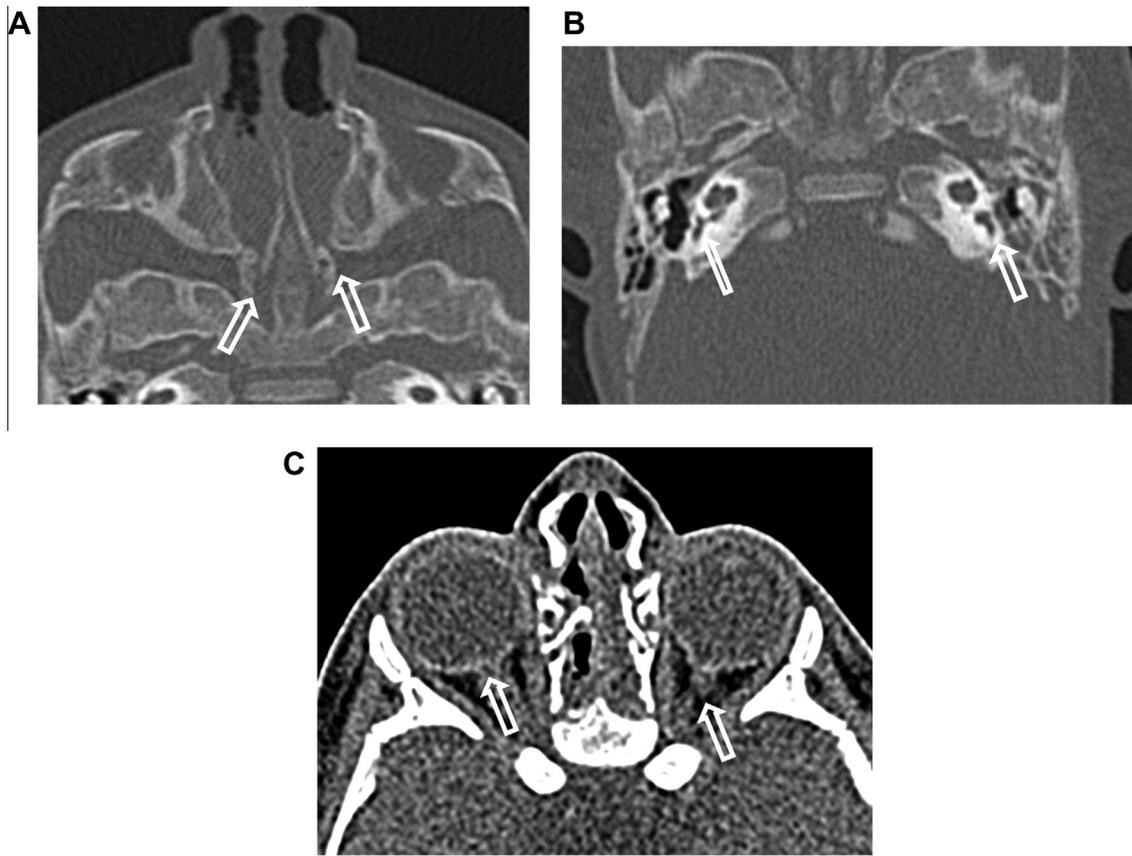


Figure 7 Axial HRCT scan showing multiple anomalies in patient with CHARGE syndrome: (A) Choanal atretic bars (arrows), (B) semicircular canal hypoplasia (arrows), (C) bilateral ocular coloboma (arrows).

auditory canal forms from first branchial cleft, malleus and incus from first branchial arch while, mastoid air cells, tympanic cavity and Eustachian tube forms from first branchial pouch.¹⁴

In middle ear anomalies, whatever in conjunction with external ear anomaly or isolated, there is a broad variety of ossicular aplasia/dysplasia like bony bridges with malleus and incus fixation, fusion, disjunction or defective parts of the ossicular chain. HRCT scan with post-processing multiplanar reconstruction is highly sensitive for demonstration of these anomalous fine defects of the ossicular structures (Figs. 4-6). Patients with severe CHL – 3 patients (5%) – were advised for insertion of titanium prosthesis, which is consid-

ered better compared to gold or Teflon, as stated by Singh.¹⁵ Reconstructive surgery was not encouraged in such patients due to expected improper prognosis.¹⁶

In addition to this ossicular anomaly diagnostic sensitivity, one of the major benefits of preoperative HRCT scan is clear demonstration of the course of the tympanic and mastoid segments of facial nerve. Normally, the tympanic portion of the facial nerve lies below the horizontal semicircular canal and above the anterior margin of the oval window. An aberrant course of facial nerve or skull base vascular structures can be infrequently associated with external and middle ear anomalies, as seen in one of our candidates (Fig. 6). Aberrant



Figure 8 Axial HRCT scan in two different patients with congenital SNHL showing: (A) globular apical turn of the cochlea (incomplete partition type II) (arrow). (B) Bilateral large vestibular aqueducts (arrows).

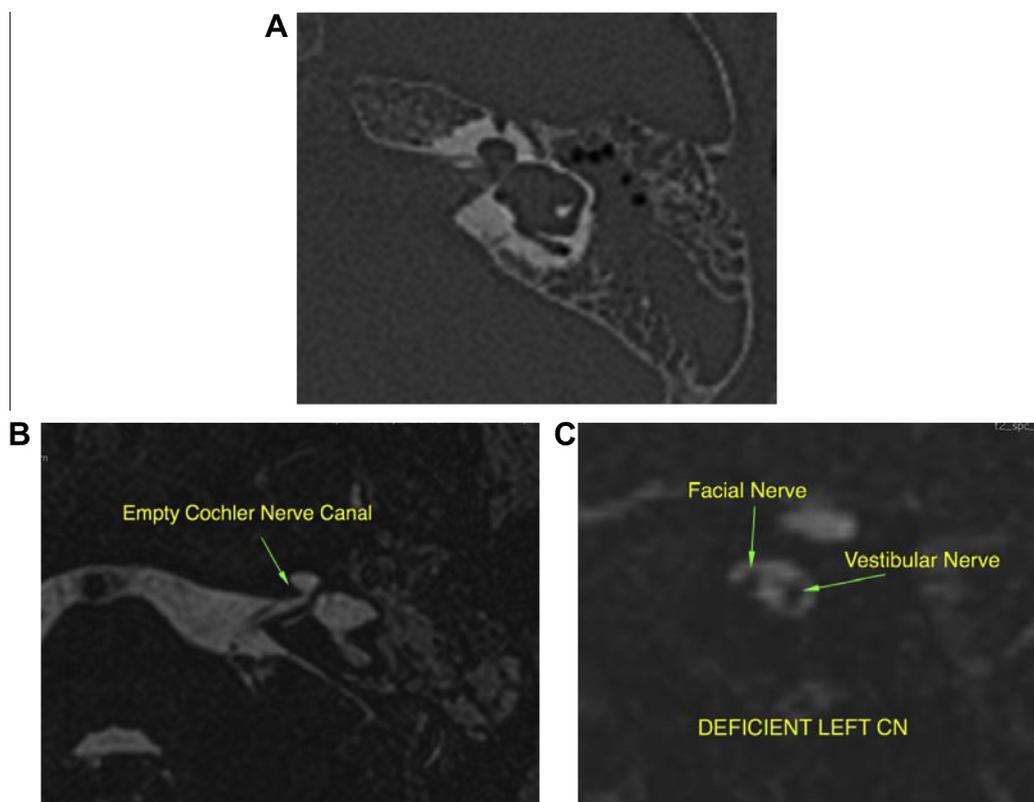


Figure 9 (A) Cystic vestibulocochlear malformation (incomplete partition type I) (arrow) with normal caliber IAC. MRI High resolution T2 of petrous bone axial (B) and sagittal (C) showing absent cochlear nerve with empty CSF filled canal with preserved facial and vestibular nerves. (Labeled arrows). . .patient is unfit for cochlear implant.

Table 4 Jahrsdoerfer scoring of patients with aural atresia and/or Middle ear anomalies.¹²

Anatomic structure	Score	Degree	No. of patients
Stapes bone	2	Score < 5 = severe	3
Oval window open	1		
Middle ear space	1	Score 5–8 = intermediate	4
Facial nerve	1		
Malleus-incus complex	1		
Mastoid pneumatization	1	Score > 8 = mild	9
Incus-stapes connection	1		
Round window	1		
External ear	1		
Total Possible Score	10	Total	16

intratympanic course of the facial nerve, limits the insertion of prosthesis needed for treatment of middle ear anomaly conductive hearing loss, especially in cases with hypoplastic tympanic cavity. Careful detailed description of the aberrant course of the skull base vessels, is mandatory to protect against life threatening iatrogenic injury of these major vessels.^{17–19}

In patients with inner ear anomalies (Figs. 7 and 8), cochlear implantation is indicated for severe or profound sensorineural hearing loss in patients who are refractory to conventional hearing augmentation tools. A cochlear implant processes the sound wave received by the device and converts it into an electrical impulse. This process bypasses the severely degenerated hair cells in the organ of Corti and then travels normally along the remainder of the auditory pathway, hence the crucial need to check intact cochlear nerve before

implantation. It was previously considered that hypoplastic IAC, which could be easily diagnosed by HRCT scan, is the indicator of cochlear nerve aplasia, as reported by Shelton et al.²⁰

However, as we faced in one patient, it is not uncommon to get inner ear canal of normal caliber, despite cochlear nerve aplasia (Fig. 9). This could be explained by the presence of other components of the vestibulocochlear nerve (vestibular nerve) that still can promote the development of normal caliber of internal auditory canal. High resolution axial and sagittal MRI T2w sequence is the reliable imaging sequence for demonstration of four nervous components of the IAC (facial, cochlear, superior and inferior vestibular nerve). Also, MRI can help in the diagnosis of other associated cranial, ocular, or cervical anomalies that may be found in syndromic

congenital SNHL e.g. Apert disease, Cruozon disease, Kleipel Feil syndrome, Hurler syndrome and Waardenburg syndrome.

So, combined HRCT scan and high resolution MRI of the temporal bone are considered a mandatory preoperative investigation tool, for facilitating accurate diagnosis, which guides the ideal management tool. This is consistent with similar data published by Varsha M. Joshi et al.²¹

12. Conclusion

- Regaining the ability to hear has an immeasurable positive psychological and emotional impact on the self-esteem of a young child with congenital hearing loss.
- Temporal bone CT is a good reliable single imaging modality for identifying bony abnormalities of the external auditory canal and middle ear as well inner ear bony labyrinth anomalies.
- The deficiency of cochlear nerve could not be constantly diagnosed by CT scan detected hypoplasia of the bony internal auditory canal, as it may be normal if vestibular nerve components are spared, however it can be clearly diagnosed by MRI.
- Combined HRCT scan and MRI petrous bone are an essential preoperative investigation tool, before any surgical repair procedure.

13. Limitation of the study

Lack of MRI examination for all candidates—only three did MRI examination—represents limitation of this study as regards radiological evaluation of neural defects in all candidates.

Conflict of interest

None declared.

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