CT Scan Value Of Temporal Bone In Assessment Of Congenital Deafness

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Summary:

Background:. Computed tomography (CT) of the temporal bone is the first-line recommended imaging modality for SNHL. Because it can identify inner ear malformations that may be responsible for hearing impairment.

Objectives: To demonstrate CT abnormalities encountered in children with congenital deafness and to assess the value of CT in the prediction for cochlear implantation. Also to evaluate the incidence and types of inner ear abnormalities in children with congenital deafness identified with CT scan for implantation difficulties.

Patients & Methods: This is a cross sectional study carried out during the period from October 2009 to October 2010 at Baghdad medical city complex on children patients who are suffering from congenital deafness. The study included 60 patients (120 ears), 27 males and 33 females, were evaluated by CT scan of temporal bone before cochlear implants.

Results: Most of the cases undergo CT scan examination were normal (80%). Enlargement of the vestibular aqueduct is most common (10%) causes of congenital inner ear malformations, followed by cochlear malformation (8.3%) while dysplasia of lateral semicircular canal found in (3.3%) of cases.

Conclusion: High resolution CT scan is recommended in all patient for pre implant analysis of temporal bone morphology due to its reliability and easy availability. CT scan is the modality provided critical information on abnormalities of the otic capsule, pneumatisation of the mastoid, middle ear abnormalities, cochlear ducts patency and vascular abnormalities- thus helping to assess the suitability of the ear for implantation, determine the side to be implanted and to find any associated abnormality which could adversely influence the surgery or post operative period.

Key word: temporal bone, inner ear malformations, CT scan, cochlear implant

Introduction;

The cochlear implant (CI) is a highly technological device that is surgically inserted in the cochlea of patients with severe to profound bilateral sensorial deafness.(1) To achieve this goal, the implant must be placed well within the cochlear lumen.(2). Given such importance, the ideal evaluation would include high resolution computed tomography (CT) of the temporal bone and the central nervous system (CNS), Thus, CT scan is always done.(1) Therefore, a detailed preoperative radiological assessment of the temporal bone has become vital for implantation of these multichannel cochlear implant devices. Imaging is required to determine the suitability of the ear to receive an implant, the choice of the ear to be operated upon and also detects additional findings that may have a bearing on the surgery or subsequent patient management.(2).

Most inner ear malformations arise when formation of the membranous labyrinth is interrupted during the first trimester of pregnancy. This interruption may be either a result of inborn genetic error or a consequence of a teratogenic exposure.(3)

Approximately 20% of cases of congenital SNHL will demonstrate inner ear malformation with modern imaging technology. (3)

Table (1): Classification of inner ear malformation (3)

1.	Complete labyrinthine aplasia (Michel)
2.	Cochlear anomalies
•	Cochlear aplasia
•	Cochlear hypoplasia
•	Incomplete partition (Mondini)
•	Common cavity
3.	Labyrinthine anomalies
•	Semicircular canal dysplasia
•	Semicircular canal aplasia
4.	Aqueductal anomalies
•	Enlargement of the vestibular aqueduct
•	Enlargement of the cochlear aqueduct
5.	Internal auditory canal anomalies
•	Narrow internal auditory canal
•	Wide internal auditory canal
6.	Eighth nerve anomalies
•	Hypoplasia
•	Aplasia

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Patients and Methods:

This is a cross sectional study conducted at Baghdad medical city complex during the period from October/ 2009 to October /2010.

It involved (60) patients (120 ears) (27males & 33 females), age from 2 to 7 years all of them presented with sever to profound SNHL.

Full history was taken regarding the following: Age. Gender. Degree of deafness. & Family history.With An attempt was made to ascertain the etiology of deafness by means of a meticulous history.

Etiology factors; congenital (idiopathic), prematurity, difficult labour (hypoxic ischemic), infectious, kerenictereus (neonatal jaundice),ototoxicity & trauma.

CT scan technique and protocol:- All 60 patients underwent multi slice (Siemens or Toshiba) spiral CT scan native study (without contrast) of temporal bone. A contiguous (0.5 to 1mm) thickness axial section parallel to the orbitomeatal line. With technique of 120 to 140 Kv & 94 or 110mA. All images were targeted using high-bone & soft tissues detail algorithms.Infants and very young children required heavy sedation or general anesthesia.

Table 2 Pre implantation HRCT Checklist

External ear & EAM	Presence , patency
Middle Ear	Integrity of ossicles; otitis media; cholesteatoma
Cochlea	presence, patency, malformations type
Vestibule & SCC	presence, malformation
Internal auditory canal	Width
Cochlear aqueduct	Width
Vestibular aqueduct	Width
Mastoids	degree of pneumatisation; Mastoiditis
Associated intracranial finding	

Result:

Age Distribution**:** The age range from 2yr to 7yr , with mean age (4,5y) with peak age at presentation was 5yr (33.3 %), lowest presentation at (2 & 7years), (table 2).

Table (3) age at presentation

Age	No.	Percentage
2	2	3.3 %
3	8	13.3 %
4	16	26.7 %
5	20	33.3 %
6	12	20 %
7	2	3.3 %

Gender distribution: The incidence of congenital hearing loss was slightly higher in females (33 females & 27 males). (Figure 1)

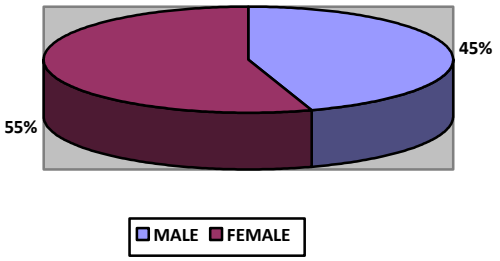

Fig. 1: incidence in male & females

CT Scan Findings: Data from all preoperative CT examinations were reviewed and colleted as described in Tables (4) & figure (2 & 3) and were as follow:-

Two patients were having external ear malformation (microtia & atresia of EAC) (3.3%), one of them has associated unilateral inner ear malformation on the epsilateral side (common cochlear cavity).Three patients had otitis media, one case showed deformed ossicles & one case showed acquired type cholesteatoma.

Inner ear malformations were found in 12 patients (20%). Three patients (5%) showed multiple (more than one) congenital anomalies that account (25 %) of all inner ear malformation. Congenital cochlear malformations were found in **five** cases (8.3%) of all patients who have SNHL & represent (41.7%) of all congenital inner ear anomalies, three cases were incomplete partition (**Mondini's dysplasia**), the other two cases showed **common cavity** cochlear malformation ,(one of them showd bilateral common cavity). two cases of Mondini showed associated inner ear anomalies (one cases showed wide VA & other has wide CA). Two patients (3.3%) had Vestibular & SCC anomalies which is dysplasia of lateral SCC. Which represent (16.7 %) of all inner ear malformations.One patient (1.7%) have widening of cochlear aqueduct & was associated with Mondini's dysplasia .while in two patients we could not recognize cochlear aqueduct.Six patients (10 %) have widening of vestibular aqueduct which represent (50%) of total inner ear anomalies, with two patients had associated congenital inner ear anomalies, one case has bilateral widening of vestibular aqueduct. Narrow IAM found in one patient (1.6%) how have SNHL, (less than 2.5mm). Three patients have Mastoiditis. While three patients were found to have high jugular bulb.One patient was found to have small bony defect in the occipital bone.

Table 4. HRCT Findings.

Part of ear	Appearance	No.	Frequency
External ear			
	Normal	58	96.7%
	Microsia & atresia of EAC	2	3.3%
Middle ear	Normal	55	91.6%
	Ossicles deformities	1	1.7%
	OM	3	5 %
	Cholesteatoma	1	1.7%
Inner ear			
	Normal	48	80%
	Cochlear malformation	5	8.3%
	Vestibule & SCC malformation	2	3.3%
	Wide Cochlear aqueduct	1	1.7%
	Wide Vestibular aqueduct	6	10%
	Narrowing IAM	1	1.7%
Mastoid			
	Normal pneumatisation	55	91.7%
	Extensive pneumatisation	2	1.7%
	Sclerosis	0	0%
	Mastoiditis	3	5%
Others	High jugular bulb	3	5%
	Carotid canal anomalies	0	0
	Associated finding (encephalocele)	1	1.7%

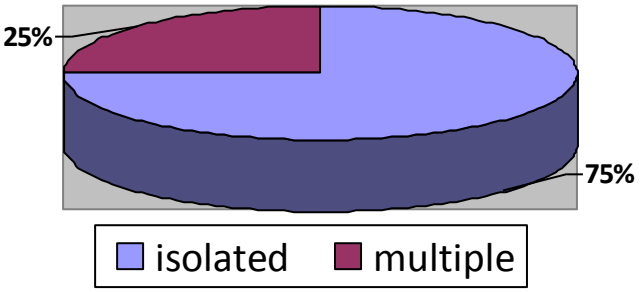

Fig.3: incidence of multiple & isolated anomalies

Table (5): Incidence of congenital inner ear malformations.

Type of malformations	No. of patients	Percentage
Normal	48	80 %
Vestibular aqueduct	6	10 %
Mondini's dysplasia	3	5 %
Common cochlear cavity	2	3.3 %
Vestibule & SCC	2	3.3 %
Cochlear aqueduct	1	1.6 %
IAM	1	1.6 %

(Note: the percentage in this table above is more than 100% because three patients have multiple anomalies).

Discussion:

Imaging plays an important role in pre-operative assessment of cochlear implants candidates, it is an established fact that radiologists are increasingly called upon to play a key role in preoperative selection of candidates. (4- 5)

Radiographic imaging of the temporal bone can identify inner ear malformations that may be responsible for hearing impairment. Computed tomography (CT) of the temporal bone is the first-line recommended imaging modality for SNHL. (6)

Incidence of congenital ear malformations:

Congenital bony inner ear malformation:- The congenital inner ear malformations were found in 12 patients (20%). Which is approximately similar to large Korean study found that 22% incidence of anomalies on radiologic imaging in 590 ears with profound SNHL.(7) While Derek D Mafong, et al found Approximately 25% of patients with congenital hearing loss will have bony inner ear malformations on computed tomography (CT) of the temporal bone.(6)D E Bamiou, et al found (28.4%) CT scans were identified as abnormal.(8) Antonelli and colleagues found anomalies in 31% of 157 children with SNHL of variable degree. (7) This range of CT yields may be explained partly by the improvement of imaging techniques. (8)Congenital cochlear anomalies:- The incidence of cochlear anomalies in our study found in 5 patients (8.3%) from all patients who have SNHL & 38.3% from all congenital inner ear anomalies. Mondini's dysplasia found in 3 patients (60 %), while common cavity. Found in two patients (40%) (These anomalies are not an absolute contraindication for an implant but since surgical experience was limited early in our series, these cases were provisionally excluded from surgery). Two patients with Mondini's deformity have associated widening VA & second patient show widening CA. Reilly found cochlear anomalies in only 4% of SNHL evaluated by high-Resolution computed tomography (CT).(7) H. Ric Harnsberger, et al. found congenital cochlear malformation in (4.8%) of SNHL.(4)D E Bamiou et al found that the cochlear malformations is the third commonest abnormality (6 %) after dilated VA & abnormal vestibule & SCC, its isolated finding in three cases (2.6%), and in association with dilated vestibular aqueduct in four cases(3.4%),while common cavity found in one patient (0.9%).(8) Urman and Talboto found common cavity in three (33%) out of ten patients who have cochlear anomalies presented with SNHL. (9).

Vestibular & lateral semicircular canal anomalies:- Regarding vestibule & lateral semicircular canal anomalies, its third most common anomaly in our study, occur in two of 60 cases (3.3%). & (13.3%) of all inner ear malformation which is enlargement of lateral SCC, one patient from those has associated widening VA.

The incidence of deformities of the semicircular canals (SCCs) and inner ear aqueducts has been less well studied than that of cochlear deformities. In a series of patients with radiographically detectable

malformations of the inner ear, the cochlea was involved in 76%, the semicircular canals were involved in 39%, and the vestibular aqueduct (VA) was affected in 32% of ears.(7) D E Bamiou et al found deformities ofSCC was second most common anomaly, occur in 9 patients from 116 cases (7.55%) of those 2 patient have associated anomalies. (8) While Derek D. Mafong found it's second most common 11%, & 5% as isolated anomalys in 6% as multiple anomalies.(6)

This wide range in percentage is possibly due to variation in reading of CT scan & introduction of advanced technology. Dilated vestibular aqueduct:- Regarding dilated VA, it was the most common CT finding in congenital inner ear anomalies, occur in six patients (10%) of all patients with SNHL & 50% of patients who have congenital inner ear anomalies, its an isolated finding in four cases (66.7%) and an associated finding in two cases (33.3%) (One with Mondini's & other one with V/SSC. Urman and Talboto found VA enlargement was 18 (50 %) in 32 patients who have inner ear anomalies with SNHL.(9) while D E Bamiou et al found isolated enlarged VA in (60%) and associated finding with other anomalies in (40%).(8) At least 40% of children with large VA will develop profound SNHL.(10) The presence of large VA may also indicate additional malformations and has been associated with lateral semicircular canal dysplasia, and Mondini's deformity. (11)

Dilated Cochlear Aqueduct**:-** We found that CA was dilated in only one patient (1.7%) which represent 12% of all patients who have congenital inner ear malformations also we found this anomaly associated with Mondini's dysplasia **.** Narrow Internal Auditory Canal:- Narrow IAC found in 1 patient (1.6%) of all patients who have SNHL & represent 7.7% of patients who have inner ear malformation. Which is approximately similar to Derek D. Mafong, et al, who found it in 1% in patient with SNHL.(6)While D E Bamiou *et al*, found narrow IAM in 2.6% of patients with SNHL.(8) The importance of small internal auditory canal is that it is considered as contraindication to implantation on that side.(11)

Conclusion:

In congenital SNHL. Approximately 80% of inner ears demonstrate no radiographically detectable abnormality. Enlargement of the vestibular aqueduct is most common (10%), followed in frequency by cochlear deformities (8.3%) & Lateral semicircular canal dysplasia (3.3%). CT is considered the modality of choice for accurate imaging of the bony labyrinth anomalies due to its reliability & easy availability. CT scan is the modality provided critical information on abnormalities of the otic capsule, pneumatisation of the mastoid, middle ear abnormalities, cochlear ducts patency and vascular abnormalities- thus helping to assess the suitability of the ear for implantation, determine the side to be implanted and to find any associated abnormality which could adversely influence the surgery or post operative period.

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