TEMPORAL BONE IMAGING IN UNILATERAL SEVERE TO PROFOUND SNHL.

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ABSTRACT:

Background: *The effect of unilateral sensor ineural hearing loss (UHL) on the quality of life is well documented. Studies found a direct link between UHL and educational and social delays. Radiological studies aimed to detect the associated anomalies of the earen countered a wide variation in the rates of incidence.*

Aim of the work: *Our study aims to detect and give an estimate about the rate of incidence of different anomalies.*

Patients and methods: *A review of the medical records of Audiology department in Eldemerdash hospital between 2014 to 2017 was done. Only cases with severe to profound USNHL diagnosed before age of 12 years were included. Clinical examination, full audiological assessment, MRI and CT temporal bone were done for all cases and results were reviewed for each patient.*

Results: *50 cases were included (8 cases with severe USNHL and 42 cases with profound USNHL). The mean age of diagnosis was 7.6 years.Only16 cases (32%) had risk factors known to contribute to SNHL. The most common abnormality found was cochlear nerve (CN) deficiency in 22 cases (44%).Bilateral findings were present in 4 cases (8%).*

Conclusion: *Radiological evaluation is not only for detection of the cause but also allows proper counselling and exploring the possible options of rehabilitation.*

Key words: *unilateral hearing loss, pediatric hearing loss, USNHL, imaging in hearing loss, congenital SNHL, CN deficiency.*

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INTRODUCTION*:*

Severe to profound USNHL means that one ear doesn't give a serviceable hearing. In Egypt, both the absence of a functioning screening system and the patients' preference to depend on the better hearing ear instead of seeking medical advice, makes the determination of the incidence of pediatric USNHL difficult. Using temporal bone imaging, the detected incidence of inner ear anomalies in pediatric USNHL ranges between 25% and 58%.[1-8]

History taking and full examination should be done as SNHL can be caused by abnormalities on the cellular level which couldn't be detected by imaging techniques.[9] Recently, cochlear implant is proposed as a treatment to restore binaural hearing in these patients which entails proper anatomical to decide the possibility of surgery and the possible outcome in those cases.[10]

PATIENTS AND METHODS:

We included 50 cases of severe to profound USNHL. Inclusion criteria was unilateral severe to profound SNHL with first audiological diagnosis of HL before the

age of 12 years. Excluded subjects were cases with mild and moderate degrees of hearing loss in the affected ear, bilateral HL, conductive component in HL and asymmetrical HL also cases whose age of diagnosis was after 12 years and individuals with history of ear surgery were excluded. HL was categorized according the WHO classification as severe SNHL if hearing threshold average between 61–80 dB in frequencies between 500 Hz and 4 KHz, and more than 80 dB as profound.[11] The normal ear should have a hearing threshold of 20dB or better in all tested frequencies. Audiologic assessment was repeated twice at two different times to ensure consistent data.

Both CT and MRI were done for all cases and were reviewed by radiologist blinded to the laterality of HL. In order to overcome the gender, age and environmental exposure the normal hearing ear was used as the control.

MRI was performed as internal auditory canal (IAC) protocol 1.5-T scanner. The caliber of the cochlear nerve(CN) was compared to the facial nerve, the superior and inferior vestibular nerves, and the CN in the normal hearing side. The CN was described to be hypoplastic when it appeared smaller in size compared with the other nerves of the IAC. The CN was considered aplastic when it could not be visualized in all planes.

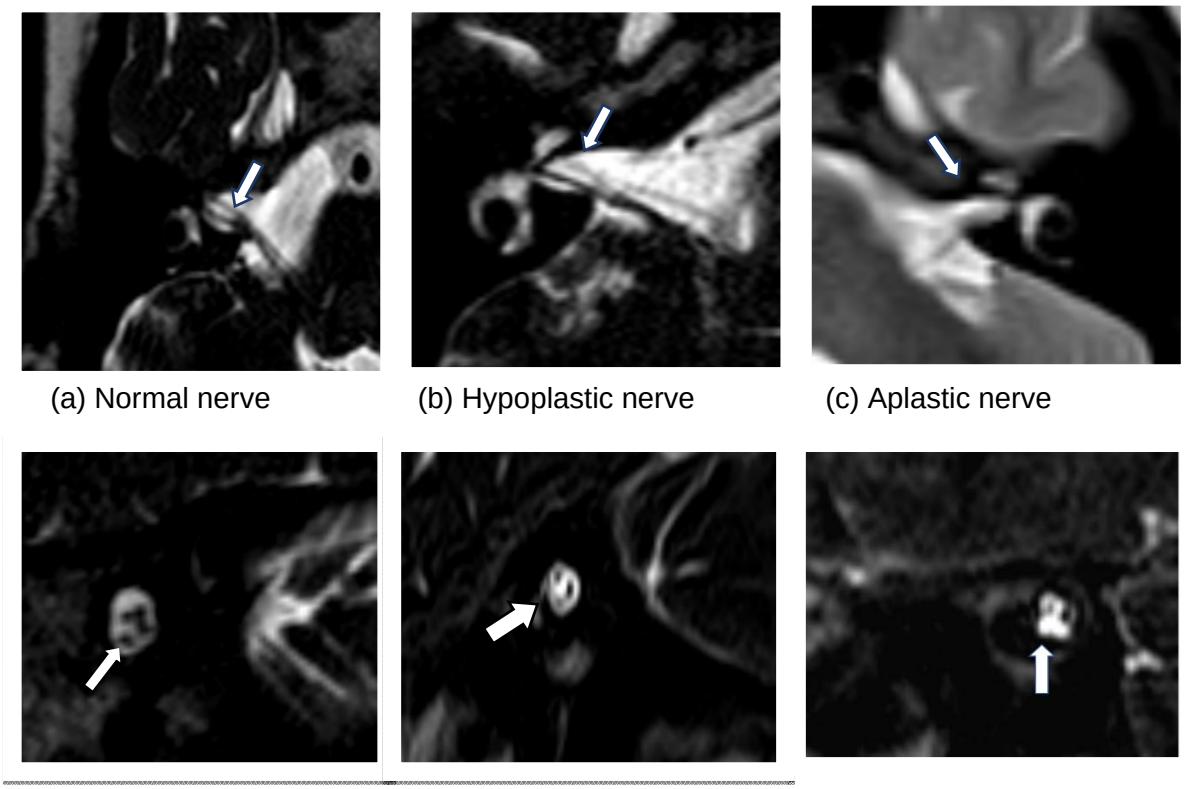
Non-contrast CT temporal was done with the cuts 1 mm wide. The protocol included axial, coronal planes with oblique sagittal reconstruction plane. Under Sennaroglu's classification, the inner ear malformation is divided into 8 categories, while the CN anomalies were described as a separate anomaly. [12]

RESULTS:

This study involved 50 subjects. Their ages at the time of evaluation ranged from 4 to 12 years, with mean age of 7.6 years ± 2.4, while the median age was 7 years. Sixteen patients (32%) had risk factors known to contribute to SNHL. (Table 1).Physical examination of all cases was unremarkable. None of them had history of vertigo, tinnitus or other associated neuropathies.

Table (1): Risk factors assessment for hearing loss.

Cases	Number	Percentage
No risk factors	34	68%
with risk factors	16	32%
• Perinatal insults	6	12%
• Consanguinous marriage	4	8%
• Familial history for HL	4	8%
• Viral infection	2	4%



(a) Normal nerve (b) Hypoplastic nerve (c) Aplastic nerve

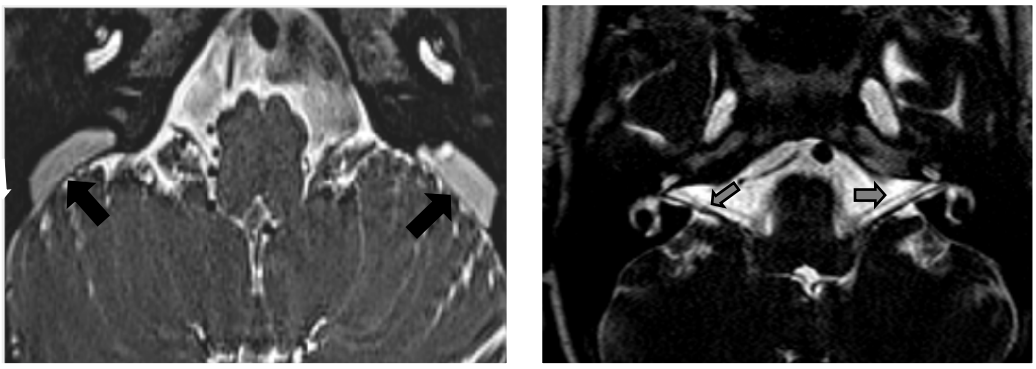
Fig (1): MRI axial and oblique sagittal cuts

Twenty-four cases were normal in both CT and MRI while 36 of them had anomalies detected in either or both imaging. The anomalies were either isolated or associated with other anomalies.

The most common was CN deficiency in 22 cases. Dysplastic cochlea and enlarged vestibular aqueduct (EVA) were each detected in 4 cases. All 4 cases with EVA were found associated with CN deficiency; furthermore, IP2 was found in 2 of them. All cases had normal IAC, SCC, vestibule. (Table 2).Bilateral CN deficiency present in 8% (n=4) with the affected side smaller than the other but both are smaller compared to the facial nerve.

Table (2) MRI findings in the affected ear

Affected ear	NO.	%
cochlear nerve (CN)		
Normal	28	56
Hypoplastic	16	32
Aplastic	6	12
Cochlea		
Normal	46	92
Dysplastic	4	8
vestibular aqueduct		
Normal	46	92
Enlarged (EVA)	4	8
Cochlea and nerve abnormality		
Normal nerve and dysplastic cochlea	2	4
Abnormal nerve and dysplastic cochlea	2	4
Other findings	4	8



(a) bilat EVA and IP2 (b) CN hypoplasia

Fig (2) MRI axial cuts

CT identified anomalies in 8 cases (16%) regarding the deaf ear. Bony cochlear nerve canal (BCNC) anomalies represent (8%) of the affected cases. It was found completely occluded in 1 case with absent CN and hypoplastic modiolus in MRI. A stenotic canal was found in 3 cases with hypoplastic nerve. Dysplastic cochlea as IP2 and EVA were each found in 2 cases (4%)(Table 3).

Table (3) abnormal CT findings in the affected ear

	NO.	%
BCNC		
Normal	46	92
Abnormal	4	8
Cochlea		
Normal	48	96
Dysplastic	2	4
Vestibular aqueduct		
Normal	48	96
Enlarged	2	4

MRI was superior in diagnosis inner ear anomalies, it identified 100% of anomalies while CT identified only 50%. (Table 4). CN deficiency present in 22 cases in MRI but 63.6% of them (14 cases) had normal CT findings.

Table (4) The sensitivity of CT to MRI in diagnosis of abnormality

	Cochlear anomalies	Vestibular aqueduct anomalies
MRI	8 cases (100%)	4 cases (100%)
CT	4 cases (50%)	2 cases (50%)

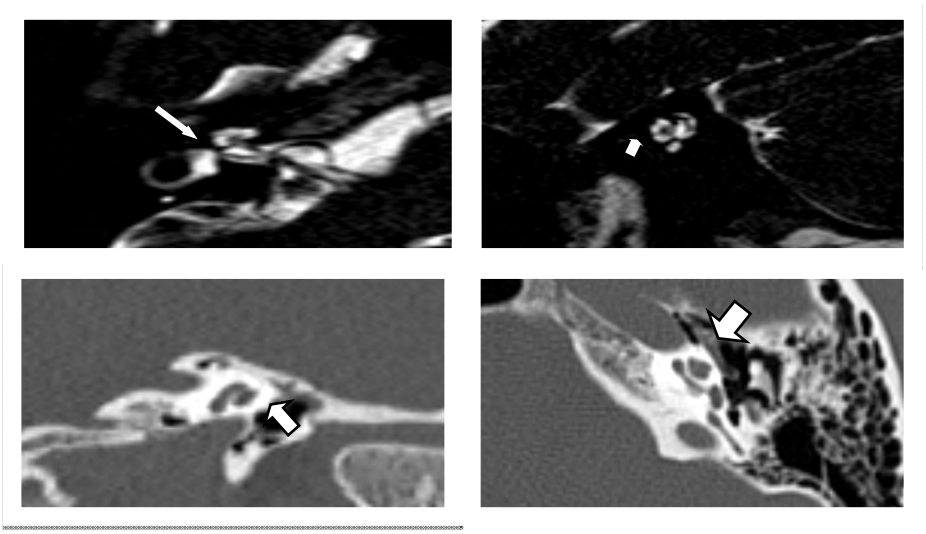


Fig (3): IP 2 in both CT and MRI

DISCUSSION:

The age of detection of USNHL is crucial for early diagnosis and intervention. Also, it plays a key role in identification of the possible cause of HL. Many studies evaluated the screening systems in their countries, and all concluded that the mean age of diagnosis decreased significantly after newborn screening system was introduced.[13, 14]. It was reported to be between 1 and 3.7 years of age. [3, 15, 16]In our study, the mean age of diagnosis was 7.6 years ± 2.4.This is probably due to lack of primary neonatal screening in Egypt. The screening allows an accurate diagnosis of the cause of hearing loss. Prior to screening, the most common etiology was idiopathic 41% followed by congenital factor 27% and other risk factors 22%. After screening, there was a shift in percentage of different etiologies. Congenital factors became the most common etiology 45% followed by idiopathic 31%.[17]In 68% of our cases, no evident etiology could be found, while the perinatal period events, familial history of HL, consanguineous marriage and viral infections were 12%, 8%, 8% and 4% respectively. The high incidence of idiopathic cases may be attributed to lack of proper documentation during the perinatal period as we rely only on history taking which might be deficient in some points.

Many inner ear anomalies could be detected by MRI as a single modality. In comparison with CT; MRI is more accurate in delineating the inner ear structures.[9] The detected incidence of inner ear anomalies in pediatric USNHL ranges between 25% and 58%, where the most common positive finding was CN deficiency with a rate of occurrence between 14 % and 58%.[1–8]In our sample, MRI identified abnormalities in 52% of cases. The most common anomaly was CN deficiency44%(32% had hypoplastic nerve, while 12% had an aplastic nerve). In previous studies, CN hypoplasia ranged between 7.1% and 42.5% while CN aplasia was between 11.9% and 68%.[17,19,20,21]

In cases with USNHL; the normal hearing ear is considered to have a normal structure by default. Recently, studies including USNHL reported bilateral lesions. While Song and colleagues[22]included cases of post traumatic and post meningitis SNHL reported a rate of incidence of bilateral anomalies in 19.4% of cases using CT scan, Bamiou and colleagues[18]included cases with syndromic hearing loss reported an

incidence rate 20%.' In this study, bilateral findings were present in 8% (n=4) of the cases. All had bilateral hypoplastic CN detected by MRI with the affected side smaller than the other but both nerves are smaller when compared to the facial and vestibular nerves. Two of them were associated with bilateral EVA and cochlear IP2. None of them had signs of an existing syndrome. This can be due to non-syndromic genetic abnormalities.

The term of isolated CN deficiency means a dysplastic CN without any other deformity. The incidence of isolated CN deficiency in this study was 28%. This goes with the finding of previous studies where the incidence was 27%.[20]

In our study, the cochlea was dysplastic in 8% (n=4) and EVA occurred in 8%.All cases had normal IAC, SCC and vestibule

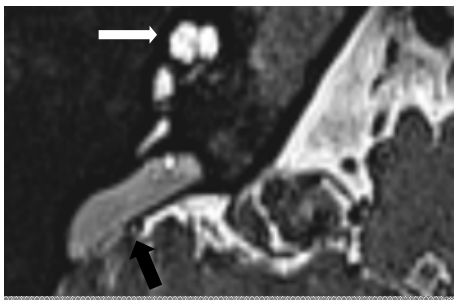


Fig (4): MRI axial cuts showing right EVA with IPII.

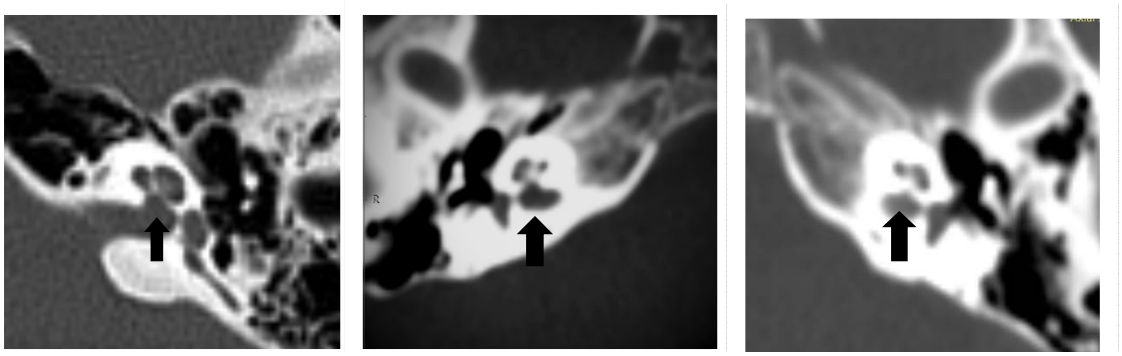
This is different from previous reports where the least mentioned rate of cochlear anomalies was 20.3%, the incidence of EVA ranged from 4% to 75% and the reported incidence of IAC and vestibularmal formations was 23% and 7.2% respectively[1,4,5,20, 23.]

In many studies, EVA can occur as an isolated lesion but usually it is a part of a wider developmental abnormality. EVA has a wide spectrum of audiological presentation ranging from mild to profound HL. Also, it can be of sudden onset, fluctuating or progressive course. EVA can be unilateral or bilateral; when bilateral, it can cause asymmetric HL.[4, 23–25]In our study, there were 2 cases of bilateral EVA with bilateral hypoplastic CN and bilateral IP2, who were presented only with USNHL. This presentation is not common for bilateral EVA but in routine audiological follow up for one of them, the child had a deterioration in hearing in the presumed normal ear.

Developmental venous anomalies (DVA) are the most common vascular lesion in CNS imaging. Most of them are asymptomatic or uncomplicated. It is extremely rare in the CPA and IAC. They can cause SNHL if caused compression of the CN. There were 2 case reports of DVA causing USNHL in children. Although rare, it should be considered in the differential diagnosis of USNHL.[26 -28]Vascular loops are a normal variant of vascularity present in 8% of cases in this study. They were bilateral with no mass effect. So, they are considered harmless.

The CT sensitivity (as a single modality) in SNHL reported by previous studies ranged between 7% and 44%.[1, 23,22]Our study was within this range, where the CT was able to detect the possible reason of USNHL in 16 % of cases.

In literature BCNC caliber of less than 1.4 mm was considered stenotic. Canal stenosis incidence was between 46.4% and 85% of congenital cases. Also, many studies found a direct connection between the width of the canal and CN deficiency with sensitivity up to 84% of cases.[16, 5,23,29]



(a) Normal BCNC (b) stenotic BCNC (c) occluded BCNC

Fig (5): CT axial cuts

The numbers derived from our study don't line up with this data. The incidence of BCNC anomalies was only 8% of total cases. In this study CN deficiency incidence was 44%. But CT didn't give a hint in most cases of CN deficiency. All cases with BCNC anomalies had CN deficiency but 36% of cases with CN deficiency had a normal BCNC. The stenosis of BCNC can be a good indicator for CN deficiency. But normal BCNC doesn't indicate a normal nerve.

In 2018, Sunwoo and colleagues[16] reported a 100% incidence of CN deficiency and 85% incidence of BCNC in cases of congenital SSD. Also, Masuda in 2013[23] gave an incidence of BCNC anomalies of 46.4% in USNHL. This discrepancy can be owed to the type of included samples. Sunwoo depended upon cases diagnosed before the age of 1 with profound USNHL after failing neonatal screening tests and excluded cases with apparent risk factors for HL, Masuda depended on a similar sample but didn't exclude cases with risk factors which gave an incidence of BCNC anomalies of 46.4%. A study in 2010 used a sample of children with auditory neuropathy (AN). They reported an incidence rate of 81.6% for BCNC anomalies and CN deficiency.[30]This is because they included cases diagnosed as AN, whom, by default have an audiometric evidence of normal cochlear function and thus, will probably have a normal cochlea in imaging. So, these findings cannot be generalized.

Degeneration of the cochlear nerve theory can explain the absence of the CN without any other anomaly. The nerve gradually degenerates, and the CN becomes hypoplastic or aplastic. Therefore, in some cases a well developed BCNC seen on CT with absent CN on MRI.[20,31,32]Regarding our study, there is no data from neonatal screening to determine the exact timing of onset of HL which makes the study includes cases of acquired HL where the cochlear nerve may be injured and partially or completely degenerated leaving a normal BCNC in CT.

In similar studies, when both CT and MRI were used together, the results were positive in 69% of cases.[4]We used both CT and MRI which led to a positive finding in 52% of cases with anomalies identified in one or both imaging.

Conclusion:

Pediatric USNHL can be due to inner ear anomalies. Radiological investigations are mandatory for all cases as it can identify the possible cause of HL. These valuable results over weigh the risk of radiation exposure or sedation and stresses over the fact that USNHL can be a sign of a life-threatening condition such as the risk of

meningitis or the possibility of progressive HL in the contra lateral ear.

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تصوير العظم الصدغي في حالات فقدان السمع الحسي الشديد الى الكلى بجهة واحدة

حسن وهبة* و لبنى الفقى* و وفاء الخوليو سامر إبراهيم و توجان طه و رضوى حلمى**

المقدمة: أوضحت دراسات عديدة تأثير ضعف السمع أحادي الجهة علي جودة حياة المرضي ولذلك ظهرت عدة أبحاث أشعة تحاول توضيح الأسباب التشريحية للمرض ولكن العديد منها واجة اختلافات كبيرة في نسب ظهور التشوهات المختلفة.

الهدف من البحث: تحديد نسب حدوث التشوهات المختلفة المسببة لضعف السمع.

المرضي والوسائل: تممراجعةسجلات المرضي في قسم السمعيات في مستشفى الدمرداش في الفترة من ٢٠١٤ وحتى٢٠١٧.اشتمل البحث على حالات الضعف السمعي الحسي الشديد الي الكلي أحادي الجهة والذين تم تشخيصهم في سن أقل من ١٢ عاما. تم عمل فحص طبي، أبحاث سمع، اشعة مقطعية ورنين مغناطيسي لجميع الحالات.

النتائج:أدرجت ٥٠ حالة (٨ حالات تعاني من ضعف سمعي شديد أحادي الجهةو٤٢ حالة تعاني من ضعف سمعي كلي أحادي الجهة). وجد ان متوسط عمر التشخيص هو ٦.٧ و٣٢% فقط من الحلات لديها عوامل خطورة قد تساعد على ظهور الضعف السمعي. تشوهات العصب السمعي مثلت نسبة ٤٤% من أجمالي التشوهات في حين ٨% من الحالات تعاني من تشوهات بالجهتين.

الاستنتاجات: فحوصات الأشعة التشخيصية لا تساعد فقط على معرفة الأسباب لكن تساعد على تقييم مدي تناسب وسائل التأهيل المختلفة وشرح المعلومات للمرضي وذويهم.