Temporal bone imaging in unilateral severe to profound SNHL

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professional levels.

May his soul rest in peace

List of abbreviation

- **ABR**---------- auditory brain stem response
- **AN**---------- auditory neuropathy
- **BAHA**---------- bone anchored hearing aid
- **BCNC**---------- bony cochlear nerve canal
- **CI**---------- cochlear implant
- **CIC**---------- complete in the canal
- **CMV**---------- congenital cytomegalovirus
- **CN**---------- cochlear nerve
- **CPA**----------cerebellopontine angle
- **CROS**--------contralateral routing of signal hearing aid
- **DVA**---------- developmental venous anomaly
- **EAC**---------- external auditory canal
- **EVA**---------- Enlarged vestibular aqueduct
- **HL**---------- hearing loss
- **IAC**---------- internal auditory canal
- **IP**---------- incomplete partition of the cochlea
- **JCIH**---------- Joined committee of infant hearing
- **OAE**---------- otoacoustic emission

- **SCC**---------- semicircular canal
- **SD**---------- speech discrimination
- **SDT**---------- speech discrimination threshold
- **SNHL**---------- sensorineural hearing loss
- **TORCH**---------- Toxoplasmosis, Other (syphilis, varicella-zoster, parvovirus B19), Rubella, CMV and Herpes infections,
- **USNHL**---------- unilateral sensorineural hearing loss

Abstract:

Objectives: Our study aims to detect and give an estimate about the rate of incidence of different anomalies. ***Methods:*** A review of the medical records of audiology department in Eldemerdash hospital between 2014 to 2017. Clinical examination, full audiological assessment, MRI and CT temporal bone were done for all cases and results were reviewed for each patient. Only cases with severe to profound USNHL diagnosed before age of 12 years were included. ***Results:*** 50 cases were included (8 cases with severe USNHL and 42 cases with profound USNHL) with slight male predominance (60%) and left side affection (52%). The mean age of diagnosis was 7.6 years. Only (32%) of cases had risk factors known to contribute to SNHL. The most common abnormality found was cochlear nerve (CN) deficiency in 44% of cases. Bilateral findings were present in 8% of the cases. ***Conclusion:*** Radiological evaluation is not only for detection of the cause, but also allows proper counselling and exploring the possible options of rehabilitation. Currently, cochlear implant (CI) is the only method to regain bilateral hearing. That's where radiological evaluation importance is most clear.

Key words: unilateral hearing loss, pediatric hearing loss, USNHL, imaging in hearing loss.

Introduction

Introduction

Worldwide, determination of the exact incidence of pediatric unilateral sensorineural hearing loss (USNHL) is difficult because children with mild USNHL pass unnoticed during early life but it manifests later as speech or language problems. Besides, the current screening techniques may fail to diagnose unilateral hearing loss. (**Ghogomu et al., 2014).**

In Egypt, many patients with unilateral hearing loss don't seek medical advice or consider treatment options and prefer to depend on the better hearing ear. So, they don't get included in studies which makes the determination of the actual incidence in the pediatric population difficult. Also, a functioning screening system is still under development; so, it is difficult to point out and follow up of children with risk factors of hearing loss.

The effect of Unilateral HL on speech and language development as well as the quality of life is well documented over the last few decades. These studies found a direct correlation between USNHL and educational and social delays due to absence of binaural hearing specially in a challenging noisy environment as schools. (**Levi et al., 2012).**

Pediatric USNHL can be caused by different insults such as infections, prematurity with NICU admission or Jaundice, exposure to ototoxic medications and hereditary hearing loss. **(Robson, 2006).**

With the awareness of the functional effect of USNHL on the affected child, a lot of studies aiming to find out the cause was done, especially those documenting inner ear anomalies.

Putting into consideration that congenital SNHL can be caused by abnormalities of the membranous labyrinth cells and functions that could not be detected by imaging techniques. A proper history taking, and full examination and investigation should be done for all cases. There is no single investigation that can cover all the aspects of HL. **(Bamiou et al., 1999).**

Since 1791- Mondini's research- the abnormalities of the temporal bone associated with congenital sensorineural hearing loss (SNHL) are investigated.

Both CT and MRI were used in the process.

CT is less time consuming, more affordable and less likely to require sedation than MRI. But w hile CT could identify bony malformations of the inner ear; MRI gives an excellent visualization of the inner ear soft tissue structures in addition to the central auditory pathway.

Audiological assessment determines the degree and type of HL and the possible reason, yet imaging investigations give an idea of the cause of HL and the anatomy of both the affected and normal ears and can expect the development of HL in the presumed normal ear. Other tests may include lab and genetic testing. Putting those investigations in a specific order is difficult and should be tailored based on the history and general examination of each case. (**Lieu et al., 2012).**

The available methods of rehabilitation of severe and profound USNHL is versatile. In some cases of USNL, no intervention was done to help the child other than preferential seating in class.

Hearing aids can be useful to compensate for hearing loss that can be effective and accepted by the patients such as:

- The conventional CROS (contralateral routing of signal) where a microphone placed near the deaf ear and an amplifier near the normal ear. The two units communicate by wireless transmission. In many countries it is considered the standard care in several countries. The main effect is to alleviate the head shadow effect. **(Hol et al., 2010).**
- CIC/ CROS where a high-power hearing aid tightly fits into the patient's deaf ear and vibrates inside the EAC in response to sound. those vibration are transmitted through bone conduction to the cochlea in the normal ear. (**Marx M et al., 2019).**

- BAHA: a surgically implanted device that conveys sound from the deaf ear to the cochlea of the normal ear. It utilizes the high speed at which sound can travel through bone. It is approved for patients 5 years old and older. Candidacy for BAHA includes all causes of profound SNHL, whether congenital or acquired. It improves understanding of speech in noise and patient satisfaction. There are risks for BAHA such as losing the fixtures, and failed osseointegration. (**Christensen et al., 2010)**

- Intraoral bone conduction device is a non-surgical prosthetic device formed of 2 parts; a microphone inserted in EAC of the deaf ear and a small device look like a teeth prothesis fixed to the molars of the upper jaw - instead of the internal part of BAHA - on the side of good hearing and eliminates the need for surgery but this requires EAC and molars in good condition; Having previous dental work might affect the results.

Neither of these modalities allows binaural hearing to develop or sound localization ability because they deliver only unilateral auditory input to the brain through contralateral routing of signal depending on one functioning cochlea. **(Mertens et al.,2017).**

cochlear implantation in USNHL is an emerging intervention that forms another option to patients which can help in the development of true binaural hearing at the brain level. Bilateral auditory stimulation

can therefore be achieved in those patients which represents the most significant difference from other types of hearing aid devices. (**Marx M et al., 2019).**

- By using the knowledge derived from all investigations, it is possible to determine which options can suite each case and family counseling on the behalf of the child could be done based on a complete knowledge.

Aim of study