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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.

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 perinatral 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

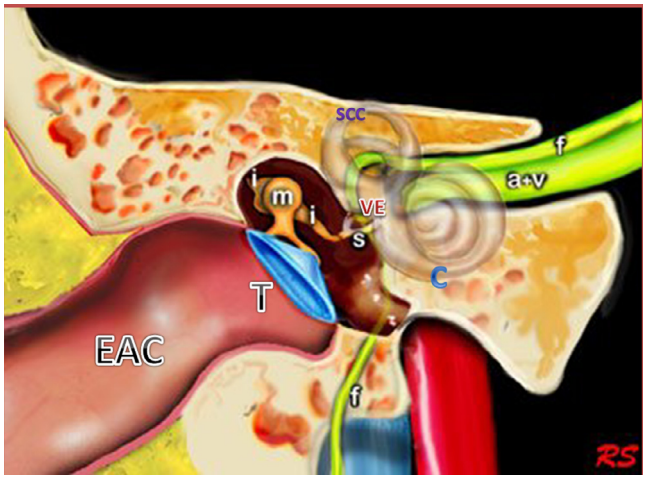

Figure 1 Coronal model of petrous bone showing different parts of the ear: EAC: external auditory canal, T: tympanic membrane, i: incus, m: malleus, S: stapes, SSC: semicircular canal, C: cochlea, f: facial nerve, a + v: auditory and vestibular nerves, VE: vestibule. Bee.[10]