Introduction

The advent of the cochlear implant (CI) has brought the world of sound to a growing population of patients. The number of patients who are candidates for cochlear implantation has increased steadily as we have worked to find new ways to incorporate cochlear implantation in the care of patients. Fishman emphasizes the role of cochlear implantation especially in patients who have difficult anatomy or underlying pathology that make cochlear implantation more difficult. The question is who fits these criteria for cochlear implantation? Second, who doesn't fit these criteria? And lastly, what makes the patient difficult to implant? ***(Fishman, 2012).***

Since their development 35 years ago, cochlear implants have revolutionized the management of children and adults with deafness or profound sensorineural hearing loss ***(Clark et al., 1977; House, 1978; Hochmair et al., 1979; Wilson et al., 1991).*** These first true bionic devices of the human body nowadays enable congenitally deaf children to attend mainstream schooling, education and pursuit of professional career like their normal hearing peers ***(Dorman and Gifford, 2010; Niparko et al, 2010).*** Over the last decade, the selection criteria for CI have expanded to include children with special auditory, otology, and medical problems. Definitive indications for implantation in these unique pediatric populations currently are still in evolution ***(Hang et al., 2012).***

Sensorineural hearing loss (SNHL) is a major cause of childhood disability worldwide, with an estimated prevalence of 1 in 2000 neonates and 6 in 1000 children by 18 years of age ***(Billings and Kenna, 1999).*** Early diagnosis and treatment of SNHL in children is critical because it is well recognized that a delay in identification of hearing impairment can adversely affect speech and language development, academic achievement, and social and emotional development. The institution of universal neonate hearing screening in the United States has altered the management of childhood SNHL by substantially lowering the average age at diagnosis from 24-30 months before the introduction of screening programs to 2-3 months ***(Harrison et al., 2003).***

It has been estimated that approximately 30-40% of children with sensorineural hearing loss may have additional disabilities, most commonly syndrome abnormalities, autism, cerebral palsy, and neuro-degenerative conditions ***(Wiley et al., 2005).*** One multi-centre study found that five percent of the pediatric cochlear implant population had additional needs, mostly visual impairment and learning disabilities ***(Uziel et al., 1995).*** In these highly heterogeneous groups, the use of standard outcome measures and subsequent analysis is even more problematic than usual ***(Filipo et al., 2004).*** Cochlear implantation in syndromic children has been relatively uncommon in the past because of their complex medical needs and difficulties with surgery and auditory

rehabilitation in these patients. In the literature, small series of cochlear implants in individual syndromes have been reported ***(Bajaj et al., 2012).***

SNHL results from congenital or acquired conditions that affect hair cell function of the cochlea ***(Fatterpekar et al., 1999).*** For infants and children with severe to profound SNHL, CI surgery remains the best habilitative/rehabilitative option, improving sound and speech perception abilities in the vast majority of implant recipients ***(Papsin and Gordon, 2007).*** Although variables such as age at implantation, duration of hearing loss, and the presence of residual hearing are among the most important determinants of CI performance, outcomes vary widely, even among similar cohorts ***(Lazaridis et al., 2010; and Vlastarakos et al., 2010).***

What is not fully understood is whether the degree of inner ear dysplasia conferred on imaging studies can help predict clinical outcomes. Unlike the vast majority of adult CI candidates with post lingual SNHL who have normal anatomy, 20% of pediatric candidates have cochlea-vestibular anomalies, as described by ***(Jackler et al., 1987).*** Therefore, imaging is an important component of the evaluation of a child with congenital SNHL ***(McClay et al., 2008).*** Computed tomography (CT) of the temporal bones remains the modality of choice at most CI centers ***(Adunka et al., 2007)*** and an absent cochlea or auditory nerve (AN) would preclude surgical intervention ***(McClay et al., 2008).***

In equivocal cases (small or absent cochlear nerve canal, hypoplastic cochlea, or profound thresholds), high-resolution magnetic resonance imaging (MRI) can help complement the CT findings, and reformatted constructive interference steady-state or fast imaging employing steady-state acquisition sequences can help determine the presence of a small but present AN. As a result, a higher rate of inner ear dysplasia is seen on temporal bone MRI compared with CT in pediatric patients with SNH ***(McClay et al., 2008).***

The rate of hearing loss in children has been estimated as 2.5 per thousand. The latter figure including those cases of genetic hearing loss that present later in childhood or hearing loss of an acquired etiology. The advent of neonatal hearing screening has significantly increased early detection of significant childhood hearing loss and resulted in earlier intervention with respect to both hearing amplification and cochlear implantation. Normal hearing is a pre-requisite for adequate speech and language development and therefore identification and management of hearing loss is critical to ensure an optimal outcome in this regard ***(Papsin and Gordon, 2007).***

Prior to the 1990s, inner ear dysplasia was felt to be a contraindication to implantation ***(Balkany et al., 2005)*** due to concerns about (1) electrode placement and stability, (2) abnormal or absent neurons, (3) risk of cerebrospinal fluid (CSF) leak, and (4) facial nerve injury ***(Luntz et al., 1987).*** The first CI was performed in pediatric and adult patient with anomalous cochlear anatomy, the patient was found to

have a Mondini deformity on imaging performed after successful surgery ***(Mangabeira-Albernaz, 1983).*** Since then, a number of articles have described CI surgery in patients with cochleovestibular malformations ***(Incesulu et al., 2008; Shim et al., 2006).***

Over the last decade, the selection criteria for CI have expanded to include children with special auditory, otology, and medical problems. Included within this expanded group of candidates are those children with auditory neuropathy spectrum disorder (ANSD), cochleo-vestibular malformations, cochlear nerve deficiency (CND), associated syndromes, as well as multiple medical and developmental disorders.

Historically, inner ear anomalies have been categorized based on an arrest in development during different stages of embryogenesis. In 1987, Jackler et al classified these malformations into 5 types: (1) complete labyrinthine aplasia, (2) cochlear aplasia, (3) cochlear hypoplasia, (4) incomplete partition (IP) and (5) common cavity.

In 2002, Sennaroglu et al. proposed a new classification system that distinguished Mondini malformations from more severe abnormalities by sub classifying incomplete partition into IP-I (less than 1.5 turns) and IP-II (between 1.5 and 2.75 turns, or a classic Mondini deformity accompanied by a dilated vestibule and enlarged vestibular aqueduct ***(Sennaroglu et al., 2002).*** Although CI surgery for children with inner ear dysplasia is

routinely performed today, the impact of anatomic anomalies on long-term CI outcomes is not completely understood ***(Munro et al., 1996).***

A large study by Papsin ***(Papsin, 2005)*** described more challenging surgery in these patients due to abnormal facial nerve anatomy and CSF leakage, but there were no statistically significant data on speech perception outcomes. Multiple small studies have examined outcomes in CI users with inner ear anomalies ***(Bloom et al., 2009; Aschendorff et al., 2009; Loundon et al., 2008).*** In order to better understand how one is able to implant cochlear devices in those difficult ears, one must understand the principal goals of cochlear implantation. The principal cause of hearing loss in most people is damage to the sensory hair cells. This severs the connection to the central auditory system.

The function of CI is to bypass the missing or damaged hair cells and stimulate directly the surviving neurons. Histological analysis has shown that peripheral neurons of a spinal ganglion cells generally undergo retrograde degeneration. However, the cell bodies of the spinal ganglion cells are much more robust and remain intact even years after hearing loss ***(Wilson, 2009).***

The cochlear implantation is used to excite the nodes around the spiral ganglion cells. There is question to how many surviving cells are left in the cochlea depending upon the pathology. Most of these have shown that the surviving cell counts vary from location to location and from cochlea to cochlea. The minimum number of auditory neurons

necessary for speech recognition is unclear. Multiple histological analyses have been performed studying the ganglion cell count and various disorders. Patients with Labyrinthitis ossificans can have between 6000-28,000 ganglion cells. Patients with Mondini deformity can have 7000-16,000 ***(Almond and Brown, 2009).***

Patients diagnosed with a congenital SNHL and cochleovestibular malformation are regarded as CI candidates. However, what contraindications can occur when implanting this subset of patients? What complications can occur with implanting patients with cochleovestibular malformations? Are still unclear? ***(Eisenman et al., 2001).***

The type of malformation generally dictates the surgical approach for implantation. With the exceptions being complete labyrinthine and cochlear aplasia ***(Buchman et al., 2011),*** all cochleovestibular malformations can be implanted, and most can be approached with the mastoidectomy with posterior tympanotomy approach (MPTA). The exception is common cavity (CC) malformation, which may be accessed directly via a transmastoid labyrinthotomy ***(Buchman et al., 2011).*** The electrode choice can differ between the types of cochlear malformation. Many malformed cochleae do not have the neuroepithelium in normal location. This is the target for the cochlear implantation. With few exceptions, normal electrode arrays are used. After implantation, device programming can be more

challenging. Frequent adjustment in the program is necessary in these patients as stimulation of the facial nerve (FN) is more common ***(Eisenman et al., 2001).***

Aim of the Work

To review the literature regarding the current concepts, future strategies and prognosis of cochlear implantation in congenital inner ear malformations.

Chapter (I): Embryology and Surgical Anatomy

In order to better understand both the role of cochlear implantation and the process by which it works, we must first understand the anatomy and embryology the ear better. To quickly summarize the physiological cause of hearing, sound waves travel through the ear canal vibrating the tympanic membrane. This then causes the ossicular chain to vibrate. The stapes attached the oval window (OW) causes a hydraulic like motion pushing fluid waves to the cochlea. This causes motion of the basilar membrane which is then sensed by the hair cells. Hair cells motion then activate or inhibit chemical transmitter release. The nearby neurons are then activated based on these changes and transmit the signal to the central auditory complex. This is what creates sound in the normal working ear ***(Almond and Brown, 2009).***

Embryology

Development of the Temporal Bone (TB)

Although inner and middle ear (ME) structures have completed development long before birth, the mastoid and tympanic bones, in particular, manifest postnatal growth and development. Knowledge of these developmental changes is imperative for the otologic surgeon contemplating operative intervention in the very young pediatric patient or cochlear implantation in the profoundly deaf infant or child. In the neonate, the squama is disproportionately large in comparison with that of the adult (Fig.1).

The mastoid process is essentially nonexistent, and the tympanic bone is a relatively flat ring, rather than a cylinder. The relative position of the entire TB in the

neonate (Fig.1) is inferolateral in comparison with the TB in the adult and its more lateral orientation. The FN, in the absence of a mastoid process, exits the stylomastoid foramen to emerge on the lateral aspect of the skull and thus is especially vulnerable to injury if a standard postauricular incision is performed.

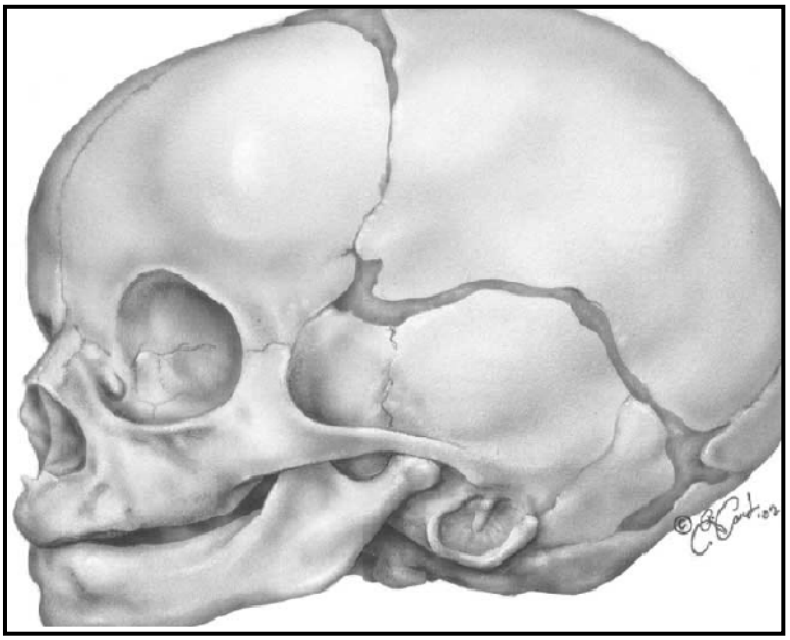

Fig. (1); Skull of the infant, shows the TB with absent mastoid process and laterally extended external auditory canal (EAC) ***(Anson, and Donaldson, 1981)***

After the first year of life, the mastoid process begins development both laterally and inferiorly, with the mastoid tip deriving from the petrous portion of the mastoid ***(Anson and Bast, 1980).*** Similarly, the tympanic ring extends laterally, completing the formation of the bony external auditory canal (EAC), the sheath of the styloid process, and the nonarticular part of the glenoid fossa (Fig.2).

In the 1-year-old infant, opposing spurs of growing bone at the ventral aspect of the bony EAC fuse, dividing the original EAC into the adult EAC and an inferior channel, known as the foramen of Huschke. The adult EAC is cranial to, and larger than, the foramen of Huschke (Fig.2). This secondary foramen closes in late childhood ***(Anson et al., 1955).*** With these changes in the mastoid and tympanic bones, the lateral aspect of the TB is vertically oriented, and the FN is buried beneath the protective barrier of the mastoid process. The lateral growth of the tympanic ring, as mentioned previously, carries the tympanic membrane from the nearly horizontal orientation of the neonate to the adult angulations by age 4 or 5 years.

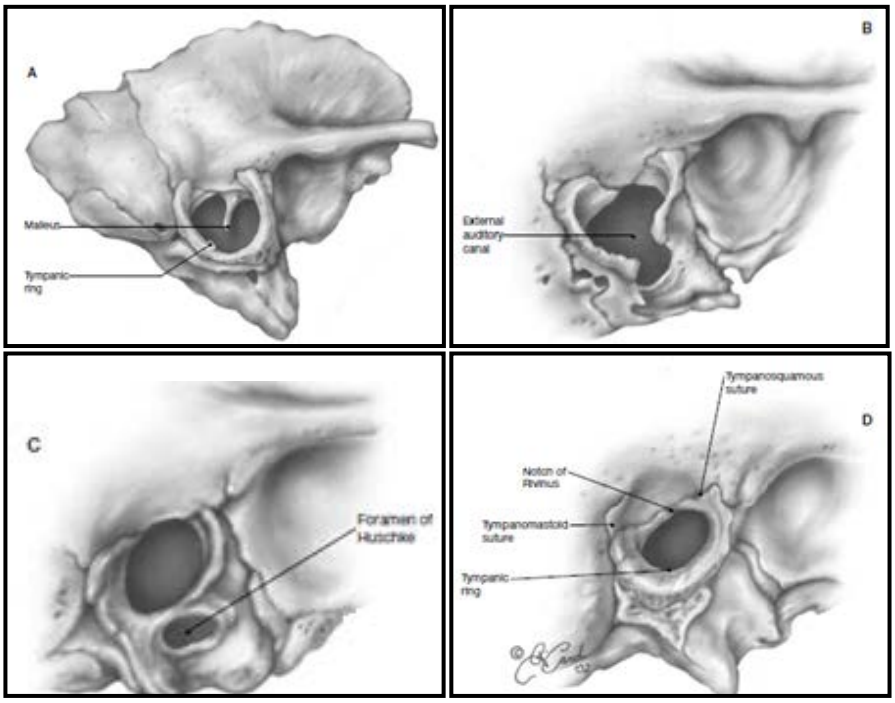

Fig. (2): Postnatal development of the tympanic portion of the temporal bone (TB). *A*, Neonate. Note the flat tympanic ring and the exposed stylomastoid foramen. *B*, Infant, The notch of Rivinus and the foramen of Huschke are becoming evident. *C*, Infant. *D*, Adolescent ***(Anson, and Donaldson 1981).***

With a view toward cochlear implantation in the infant or young child, one study suggested that the dimensions that show significant growth, continuing into the teenage years, include the depth of the tympanic cavity (as measured by the distance between the tympanic membrane (TM) and the stapes footplate) and the length, width, and depth of the mastoid ***(Eby and Nadol, 1986).***

The inner ear commences to develop at approximately 3 to 4 weeks of intrauterine life. The otic placode, a plaque like thickening of surface ectoderm dorsal to the first branchial groove, appears at the end of

the third week on each side of the developing hindbrain (fig.3). Approximately 2.5 cm of slack to accommodate anticipated growth. The facial recess, on the other hand, should be adult size at birth ***(Eby, 1996).*** If genetic control is defective or any noxious agent is present during this time, the deformity will be severe. Occasionally there is complete absence of the labyrinth (Michel's syndrome) ***(Jackler et al., 1987).*** Failure of cochlea development late in the third week of gestation results in a cochlear aplasia ***(Ozgen et al., 2009)*** (Fig.7).

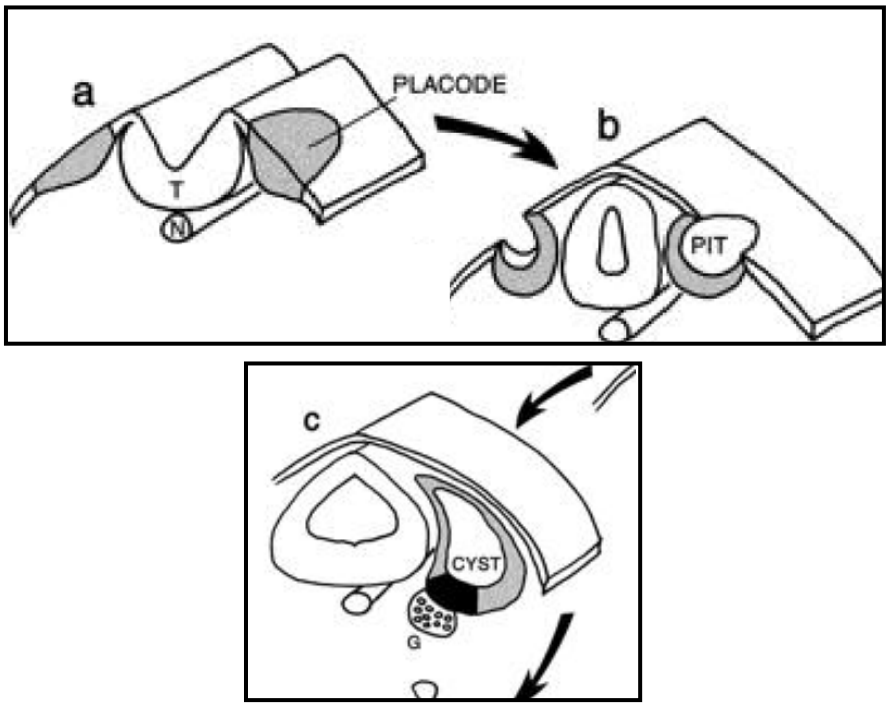

Fig. (3): (a) Otic placode is an ectodermal thickening adjacent to the rhombenscephalon. (b) Invaginates to form the otic pit. (c) Pinching off of otic pit to form otic cyst ***(Streit, 2007).***

The endolymphatic appendage appears at this stage, considerably in advance of the semicircular and cochlear ducts ***(Streeter, 1906).*** Expansion of the auditory pit and fusion of overlying tissue create the otocyst (otic vesicle), separated from the surface (Fig. 3). This occurs roughly in the fourth gestational week. The mesenchyme tissue that surrounds and differentiates in conjunction with the otocyst is the future otic capsule (bony labyrinth). It becomes bilobed as the pars superior forms the vestibular system and the pars inferior forms the cochlea and saccule (Fig.4). If development is arrested at this stage, the cochlea may only develop one turn or a CC (Fig.7). Development then involves elongation of the otocyst and the appearance of three deepening folds (I, II, and III) (Fig.5).

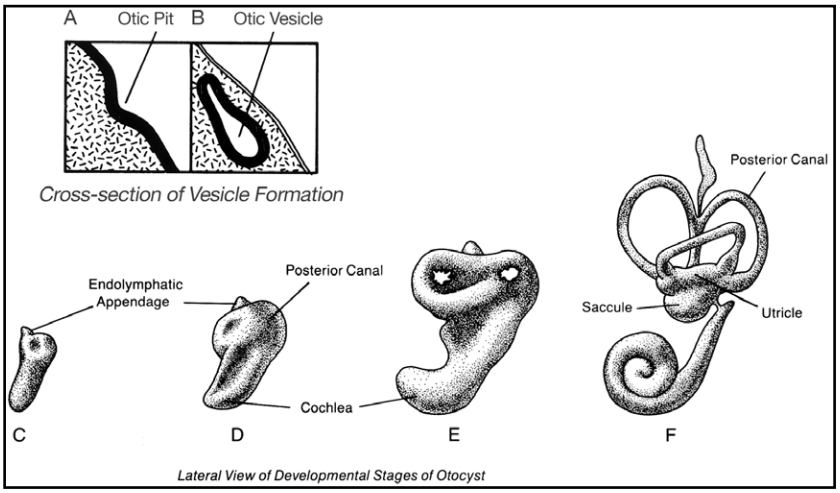

Fig. (4): The evolution of the endolymphatic (otic) labyrinth. *A* = 22 days, *B* = 4 weeks, *C* = 4½ weeks, *D* = 5½ weeks, *E* = 6 weeks, and *F* = 8+ weeks. After Streeter.23 Reproduced with permission from Gulya AJ, Schuknecht HF. Anatomy of the temporal bone with surgical implications ***(2nd ed. Pearl River (NY): Parthenon Publishing Group; 1995).***