Original Article

Assessment and management of patients with severe-to-profound sensorineural hearing loss

Rizwan Ahmad

Assistant Professor, Department of ENT and HNS DMCH Laherisarai, Bihar, INDIA.
Email: kajal6160@gmail.com

Abstract

Background: Severe-to-profound sensorineural hearing loss (SNHL) is a significant impediment to an individual's ability to communicate successfully. If hearing loss occurs prior to the development of speech, additional reading and language difficulties confront the individual. Although postlingual hearing loss does not necessarily affect speech and language development, it can also have a profoundly negative impact on a person's ability to function. The successful application of cochlear implantation in children and adults with severe-to-profound SNHL has been pivotal in helping overcome these functional deficits. This chapter reviews various etiologies of severe-to-profound SNHL, highlights aspects involved in patient evaluation, and discusses potential management options for rehabilitation. Particular focus is given to the role of cochlear implants in the management of patients with severe-to-profound SNHL. **Materials:** Total 46 Children were enrolled in my study, and hearing level was examined by audiometry on first day then 1 week, and 3 months interval, According to the degree of the initial hearing loss. All children's were attending on OPD& IPD in the department of ENT in Darbhanga Medical College & Hospital. **Results:** The criteria used to determine cochlear implant candidacy have evolved fairly rapidly over the past several years. At present, pure-tone averages should exceed 70 dB sound pressure level, whereas word recognition scores should be less than 50% in the best-aided condition using the Hearing in Noise Test or comparable testing methods. As discussed, a trial of appropriate amplification should be undertaken before using a cochlear implant. Additional criteria include inner ear anatomy amenable to receiving a cochlear implant, a lack of medical contraindications, appropriate patient/family expectations and support, ear free of active infection, and age older than 12 months. As implant technology improves, indications for cochlear implantation will expand to include those with better residual hearing. **Conclusion:** It is important to recognize that the use of cochlear implants has been transformational. Within the pediatric population, early use of cochlear implants has allowed language development equivalent to that of hearing children and has provided hearingimpaired children an opportunity to participate and thrive in mainstream educational settings. Certainly, there is great individual variability among cochlear implant users, and it is difficult to prognosticate expected outcomes for anyone potential candidate. Nevertheless, there is growing evidence that the average cochlear implant user will benefit greatly from the use of this device.
Key Words: Hearing Loss, Cochlea, Ear.

Dr. Rizwan Ahmad, Assistant Professor, Department of ENT and HNS DMCH Laherisarai, Bihar, INDIA.
Email: kajal6160@gmail.com
Received Date: 15/02/2017 Revised Date: 28/03/2017 Accepted Date: 06/04/2017

Access this article online

Quick Response Code:

Website: www.statperson.com

DOI: 06 April 2017

INTRODUCTION

With the early work of Hermann von Helmholtz and subsequent work of Georg von Bekesy on the mechanics of the basilar membrane and the traveling wave theory of cochlear stimulation, our understanding of cochlear physiology has grown considerably over the last century. Inner hair cells within the cochlea are responsible for transduction of sound energy into electrical signals carried by fibers of the auditory nerve, while outer hair cells are believed to play a role in altering this process of mechanoelectrical transduction. Much of our understanding of the pathophysiology of SNHL arises from studies of human temporal bone specimens, which allow correlation of these delicate inner ear structures with clinical characteristics of the affected individual. Pathophysiological correlates of SNHL include loss of hair cells as well as loss of spiral ganglion neurons and atrophy of the stria vascularis. In clinical practice, severe-

How to site this article: Rizwan Ahmad. Assessment and management of patients with severe-to-profound sensorineural hearing loss. *International Journal of Recent Trends in Science and Technology*. April 2017; 22(3): 157-162. http://www.statperson.com (accessed 14 April 2017).

to-profound SNHL may best be classified as either prelingual or postlingual, depending on whether the loss develops before or after the critical period of speech and language development in the brain, generally regarded as being between a and 7 years of age. Prelingual SNHL may be congenital, stemming from various genetic etiologies, developmental anomalies, intrauterine and perinatal infections such as cytomegalovirus (CMV), or neurodegenerative disorders. Additionally, acquired prelingual SNHL may occur as a result of prematurity, hypoxia, trauma, ototoxicity, hyperbilirubinemia, meningitis, or other systemic infections. Postlingual SNHL may be genetic in nature (delayed-onset), though it may also arise from ototoxic drug exposure, immune-mediated inner ear disease, trauma, noise exposure, infection (including meningitis), and aging. In many patients with severe-to-profound SNHL, an exact cause may not be identifiable. Although the incidence of children born with significant hearing loss is thought to be - 1 in 1.000, more than 30 million adult Americans have SNHL, of which as many as 3% may have severe-toprofound SNHL.

Clinical Findings: Prelingual Severe-to Profound Sensorineural Hearing Loss

Evaluation of prelingual children with suspected severe-to-profound SNHL begins with a comprehensive history and physical examination. Particular emphasis is placed on the pregnancy and birth histories for infants and young children suspected of having SNHL. Newborn hearing screening is now mandated in most states and has been successful in helping provide early identification of hearing loss. However, some hospitals may still not offer newborn hearing screening, families may refuse screening or miss screening opportunities, and false-negative tests are possible, highlighting the need for continued diligence by parents, families, teachers, and medical professionals. High-risk factors for congenital hearing impairment include a family history of congenital hearing loss or delayed-onset SNHL in childhood, a maternal history of infection during pregnancy (i.e., toxoplasmosis, rubella, CMV, syphilis, herpes, and human immunodeficiency virus [HIV]), severe hypoxia (low APGAR [appearance, pulse, grimace, activity, respiration I scores), prematurity, hyperbilirubinemia requiring phototherapy or exchange transfusion, and a need for neonatal intensive care unit admission. A history of maternal use of alcohol or illicit drugs, gestational diabetes or pregnancy-induced hyper-tension, and any other complications during pregnancy should also be sought. Additionally, a history of postnatal infection treated with potentially ototoxic drugs (e.g., gentamicin) or systemic infections, including meningitis or sepsis, should be elicited. A complete physical examination is also critical for identifying potential syndromic causes of SNHL as well as other associated anomalies in children. Special emphasis should be placed on the head and neck examination, with particular emphasis on craniofacial features. A basic neurological examination, including an assessment of vision, muscle tone, and developmental behavior, is also imperative. Although most genetic causes of SNHL are non-syndromic, autosomal recessive in nature, there are also several- autosomal-dominant conditions, many, of which may be discovered by a careful family history. Autosomal-dominant disorders associated with hearing loss include Waardenburg syndrome, Stickler syndrome, branchiootorenal syndrome, Treacher Collins syndrome, and neurofibromatosis type 2. Many of these syndromes have fairly classic features that are readily identifiable on physical examination, but additional studies may be indicated to further evaluate for the presence of associated abnormalities. Syndromic autosomal-recessive disorders associated with SNHL include Pend red syndrome, Usher syndrome, and jervell and Lange-Nielsen syndrome. Sex-linked syndromes associated with SNHL include Norrie disease, otopalatodigital syndrome, Wildervanck syndrome, and Alport syndrome.

MATERIAL AND METHODS

After the clinical history and physical examination, audiometric evaluation of a patient with suspected severe-to-profound SNHL is the next most important investigation. Following failure of a newborn hearing screening exam or if there is a suspicion of hearing loss in an infant, diagnostic audiometry should be initiated through the use of threshold auditory brainstem response (ABR) testing. Both air- and bone-conduction ABR testing should be performed to confirm a sensorineural (rather than conductive or mixed) hearing loss. Auditory steady-state response testing may also be used to help distinguish between severe and profound SNHL. In cases with absent ABRs, otoacoustic emission (OAE) testing should be performed to rule out the possibility of auditory neuropathy. For older children with suspected severe-to-profound SNHL, visual reinforcement audiometry, play audiometry, and standard pure-tone audiometry are used to help aid in the assessment of hearing loss. In the past, evaluation of children suspected of having severe-to-profound SNHL often involved a battery of blood tests and ancillary studies aimed at identifying a possible cause. Although a comprehensive history and physical examination can in fact provide a great deal of information about possible causes of prelingual SNHL, additional studies are occasionally necessary and still play an important role in many cases. Electroretinography is still recommended to assess for the presence of Usher syndrome if there is any suspicion of vestibular dys-

function or visual problems in a child with SNHL. Similarly, many clinicians believe electrocardiography is still a cost-effective test to perform in children with SNHL and any questionable history of syncope to evaluate for [ervell and Lange-Nielsen syndrome. Thyroid function testing is not useful in Pendred syndrome, though genetic testing may be useful if this is suspected. Genetic testing is also available for connexin-related SNHL, which is responsible for most cases of nonsyndromic genetic deafness. Imaging studies are also often employed in the assessment of patients with prelingual severe-toprofound SNHL. High-resolution computed tomographic (CT) scan of the temporal bone is a fairly low-cost, rapid test to examine the anatomy of the petrous bone. The detailed bony anatomy seen with a high-resolution CT scan allows for the identification of developmental anomalies of the inner ear. Specific radiographic inner ear abnormalities associated with SNHL include complete absence of the labyrinth (Michel deformity), common cavity or cystic deformity, hypoplastic abnormalities, and incomplete partition. Classic Mondini deformity includes an enlarged vestibular aqueduct, dilated vestibule, and incomplete partition of the cochlea. Enlargement of the vestibular aqueduct, narrowing of the internal auditory canal, and cochlear aperture abnormalities may also be seen with a high-resolution CT scan and may occur in isolation or as part of a larger syndrome. Magnetic resonance imaging (MRI) may also be employed in the evaluation of prelingual severe-toprofound SNHL. MRI has become the study of choice at some centers (and has superseded high-resolution CT scans), whereas others use it to complement the evaluation provided by the CT scan. High-resolution T2-weighted MRI sequences available today allow for careful examination of inner ear anatomy, including assessment of the shape, size, and patency of the cochlea and labyrinthine structures. These sequences also allow for assessment of the internal auditory canal and the cochleovestibular nerve complex. As a result of these sequences, MRI (unlike CT) is able to identify fibrosis within the cochlea or labyrinth, as well as cochlear nerve aplasia or deficiency. This is particularly important in the assessment of recent cases of meningitis-induced hearing loss, which may demonstrate fibrosis but not ossification of the cochlear lumen, and in children with profound SNHL who may lack a cochlear nerve. MRI also has the advantage of being radiation-free.

RESULTS

Clinical Findings: As with infants and children, evaluation of patients with postlingual SNHL should begin with a complete history and physical examination. In addition to the standard components of a clinical history, several specific questions should be asked when evaluating a patient with suspected severe-to-profound SNHL. A history of potential ototoxic medication use, such as intravenous antibiotics or diuretics, chemotherapeutic drugs, and certain pain medications, should be sought. A detailed noise history, a history of chronic ear infections, a history of direct head or ear trauma, and any prior ear, skull base, or intracranial surgery should also be questioned. A family history of hearing loss, a history of meningitis or other possible viral infections in childhood, autoimmune disorders, otosclerosis, Meniere disease, or other fluctuating or progressive hearing loss symptoms should also be routinely questioned. In children and adolescents, a perinatal history should be obtained because some congenital hearing losses may be progressive or delayed in onset. Physical examination should focus on the head and neck and neurotologic exam. Evidence of prior ear surgery or prior trauma to the head may be identified. Many autoimmune conditions may have other head and neck manifestations, including visual, nasal, and vestibular manifestations. Tuning fork examination should be performed to confirm an SNHL, rather than a conductive or mixed hearing loss.

Special Investigations: Standard pure-tone audiometry is used to help delineate the specific type, pattern, and degree of hearing loss in individuals with suspected hearing loss. Word-recognition testing is also included in the assessment of these patients. OAE testing may be helpful if there is a question of test reliability or to help rule out auditory neuropathy. ABR testing may be used similarly or if there is a suspicion for retrocochlear pathology. In certain situations, blood testing may be helpful. Thyroid studies, autoimmune markers (e.g., rheumatoid factor, antinuclear antibody [ANA], cytoplasmic antineutrophil cytoplasmic antibodies [c-ANCA], and erythrocyte sedimentation rate [ESR]); fluorescent treponemal antibody absorption (FTA-Abs) test (for syphilis), and human immunodeficiency virus (HIV) testing may be indicated if the clinical suspicion exists. In older children and adolescents, genetic screening may be indicated because certain diseases may be delayed in onset or progressive in nature. Imaging studies may be helpful if there is a history of trauma, ear surgery, meningitis, or ear infections. High-resolution (T scan of the temporal bone can identify cochlear or labyrinthine ossification but may miss fibrosis. MRI may be useful if there is a suspicion for internal auditory canal lesions and to assess cochlear patency in cases of recently diagnosed meningitis.

Management Nonsurgical Management and Candidacy: In patients with prelingual severe-to-profound SNHL, the educational environment in which

they are raised has a significant impact on their spoken speech and language skills. Three basic educational approaches have been described: (1) auditory-oral, (2) bilingual/bicultural, and (3) total communication (TC). All three have a common goal of developing functional language. The auditory-oral approach emphasizes only spoken language for communication. For children who are severely to profoundly hearing impaired, this strategy is usually selected in conjunction with cochlear implantation. The bilingual/bicultural strategy teaches American Sign Language (ASL) as the primary method of communication. Reading and writing the English language are taught secondarily. Advocates of this methodology contend that ASL is a natural and complete language that is totally visual and therefore accessible to the deaf child. Proponents of bilingual education align themselves with the deaf community, which recognizes deafness as a cultural difference rather than a disability. The success of this approach depends on ASL proficiency. ASL has an entirely different grammatical structure from Eng lish; therefore, the task of learning ASL for a normal hearing individual is challenging and limited to those who are closely associated with a deaf individual. The TC approach is the most common educational method used in the United States. TC employs all communication modalities, including speech, signs, gestures, finger spelling, and speech reading, reading, and writing. The objective is to develop a communication strategy that maximizes the residual auditory information a child can access. The most common implementation of TC incorporates simultaneous speech and signed English because ASL cannot be correlated with spoken language. Advocates of the TC school strongly believe that such a combined code will lead to better competence with the English language. Otolaryngologists, audiologists, speech therapists, and educators serve a vital function in helping families make decisions about the most appropriate communication strategy for an individual child. If a cochlear implant is to be considered as part of the child's rehabilitation, the earliest possible application with hearing aids must be strongly encouraged. Auditory-oral and TC education with a strong auraloral emphasis are suggested for families who want to emphasize spoken language for a child's future communication strategy. For any patient with severe-to-profound SNHL, nonsurgical management options are available for rehabilitation. Appropriately fitted hearing aids may provide some benefit to patients and should be initiated as early as possible after the hearing loss is identified. Speech reading, visual cues, written communication, cued speech, and ASL may be useful for some individuals. However, if there is a desire to develop spoken language (if prelingual SNHL) or restore auditory function (pre- and postlingual SNHL), application of the cochlear implant has become the preferred management option. The criteria used to determine cochlear implant candidacy have evolved fairly rapidly over the past several years. At present, pure-tone averages should exceed 70 dB sound pressure level, whereas wordrecognition scores should be less than 50% in the best-aided condition using the Hearing in Noise Test or comparable testing methods. As discussed, a trial of appropriate amplification should be undertaken before using a cochlear implant. Additional criteria include inner ear anatomy amenable to receiving a cochlear implant, a lack of medical contraindications, appropriate patient/family expectations and support, ear free of active infection, and age older than 12 months. As implant technology improves, indications for cochlear implantation will expand to include those with better residual hearing. Although CT and/or MRI may be used to aid in the evaluation of patients with severe-to-profound SNHL, one of these modalities is required to confirm the presence of an adequate cochlear lumen prior to cochlear implant surgery. Pre implant vestibular testing is also often performed in patients with suspected vestibular dysfunction or in elderly individuals and may aid in determining the side of implantation.

Surgical Management

Surgical Considerations

Candidates displaying significant labyrinthine ossification should undergo preoperative electrical stimulation testing using a round-window (evoked ABR) or promontory-stimulating paradigm to determine whether they perceive sound. Lack of response is not an absolute contraindication for implantation, but it may indicate limited auditory information transfer with an implant. Similarly, a history of temporal bone fracture, intracranial surgery, or head trauma causing profound deafness may also require preoperative electrical stimulation to determine whether the auditory nerve is intact. A diagnosis of otosclerosis should alert the surgeon to the possibility of distortion of cochlear anatomy with bony obliteration of the round window. Patients with otosclerosis may be more prone to facial nerve stimulation when the implant is activated due to demineralization of the otic capsule. A history of deafness following stapedectomy or iatrogenic labyrinthine injury could also result in intracochlear fibrosis that may be encountered during implantation. Intracochlear fibrosis is not seen on CT scans but may be identified with preoperative MRI, as previously discussed. Intracochlearfibrosis and/or ossification can be removed through a routine surgical approach if confined to the most basal portion of the basal turn. However, complete electrode insertion may be difficult in such cases.

Extensive ossification requires more radical surgery, including the use of split electrodes and alternative electrode placements. In patients with a mastoid cavity seeking a cochlear implant, ear canal closure with removal of all skin is recommended as a first stage. Silastic may be left within the cavity and over the region of the promontory to aid in identification of this area during the implant surgery.

Cochlear Implant Devices

Cochlear implant devices are continually undergoing modification and upgrades. However, the basic components of these devices are unchanged. Cochlear implants attempt to replace the transducer function of damaged inner ear hair cells. In most causes of neurosensory deafness, injury to the hair cells rather than auditory nerve fibers results in severe-to-profound SNHL. The basic components of a cochlear implant include the following:

1. A microphone to pick up auditory information
2. A speech processor that changes the mechanical acoustic sound energy into an electrical signal
3. A transmitter coil to send the information via radiofrequency through the skin
4. An implanted receiver/stimulator that interprets the electrical signal sent by the speech processor
5. An intracochlear electrode array that distributes the electrical sound information to the auditory nerve

Innovative methods of processing speech have been developed by many independent research programs and cochlear implant manufacturers. Most cochlear implants use a band-pass filter system to separate the acoustic signal into discrete frequency bands that can be delivered to the appropriate frequency regions of the cochlea, providing spectral information about the speech signal. Temporal and intensity cues are delivered by varying the rate of stimulation and the amount of stimulating current. Most implant systems use a nonsimultaneous stimulation paradigm that prevents stimulation of more than one channel at a time and eliminates channel interaction because it can induce excessive current loads from overlapping signals. Changes in speechprocessing strategies will continue as the technology advances. It is fortunate that new implant systems are being designed to enable adoption of a broad array of speech-processing software changes without requiring surgical hardware reimplantation.

Cochlear Implant Surgery

Standard cochlear implant surgery involves a post-auricular approach that accesses the middle ear through a mastoid-facial recess approach (posterior tympanotomy) with the use of facial nerve monitoring. Incision styles, methods of flap development, and techniques for securing the processor vary considerably. Depending upon surgeon preference, anatomical features, and preoperative audio met - ric factors, cochleostorny or direct round-window insertion techniques may be used. Various electrode designs are now available that allow the surgeon to tailor surgery for the patient's specific anatomy. Implants are normally placed in the scala tympani, though in rare situations, the scala vestibuli may be used. With children, special attention must be given to skull thickness and position ofthe facial nerve and mastoid cortex, but the remainder of the anatomy in

Cochlear Implant Outcomes

The mastoid, facial recess, and cochlea is similar to that of an adult. Patients with inner ear abnormalities (e.g., Mondini deformity, cystic cochlea, cochlear aperture deficiency) may require modifications to the standard surgical technique. Shorter, noncurved electrodes (with circumferential contacts) are often employed in such cases, and fluoroscopy may be used to ensure proper placement. Eustachian tube obliteration and packing of the middle ear may be required in these patients if perilymphatic gusher or cerebrospinal fluid leakage is encountered. Awareness of the potential complications of cochlear implant surgery is critical and should be thoroughly discussed during the informed consent process. Facial paralysis, implant extrusion, implant failure, and meningitis are serious complications that can occur following cochlear implant surgery. Preoperative vaccination against *Streptococcus pneumoniae* and early treatment of infections are important means of avoiding meningitis and device-related infections or failure. Although it is beyond the scope of this chapter to discuss the extensive literature that is available in terms of cochlear implant user outcomes, it is important to recognize that the use of cochlear implants has been transformational. Within the pediatric population, early use of cochlear implants has allowed language development equivalent to that of hearing children and has provided hearingimpaired children an opportunity to participate and thrive in mainstream educational settings. Certainly, there is great individual variability among cochlear implant users, and it is difficult to prognosticate expected outcomes for anyone potential candidate. Nevertheless, there is growing evidence that the average cochlear implant user will benefit greatly from the use of this device.

CONCLUSION

With advances in speech processing and electrode technology as well as improvement in patient outcomes, criteria for cochlear implant candidacy are continuing to broaden. Hybrid cochlear implants provide a means of delivering electrical and acoustic stimulation to a single

ear by inserting a shortened electrode into the more basal aspects of the cochlea. In combination with "soft" surgical techniques, this technology aims to preserve low-frequency residual hearing, which can subsequently be used in conjunction with "electrical" stimulation of the higher frequencies encoded along the basal turn, provided by the implant. Cochlear implants are also being explored in cases of single-sided deafness and tinnitus. Preliminary data are limited, but this technology may help improve listening for these individuals in a variety of situations, particularly when compared with the nonimplant and contralateral routing of sound (CROS)-aided conditions.

REFERENCE

1. Adunka OF. Teagle HFB. Zdanski C]. Buchman CA. Influence of an intraoperative perilymph gusher on cochlear implant performance in children with labyrinthine malformations. Otol Neurotol 2012;33(9):1489-1496
2. Arndt S. Aschendorff A. Laszig R. et al. Comparison of pseudobinaural hearing to real binaural hearing rehabilitation after cochlear implantation in patients with unilateral deafness and tinnitus. Otol Neurotol2011;32(1):39-47
3. Chau JK. Cho JJ. Fritz OK. Evidence-based practice: management of adult sensorineural hearing loss. Otolaryngol Clin North Am 2012;45(5):941-958
4. Eisenberg L. ed. Clinical Management of Children with Cochlear Implants. San Diego. CA: Plural Publishing; 2009
5. Niparko J. ed. Cochlear Implants: Principles and Practice. Philadelphia. PA: Lippincott Williams and Wilkins; 2009

Source of Support: None Declared
Conflict of Interest: None Declared