Newer Frontiers in Hearing Rehabilitation

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Introduction

To be aware of our surroundings is the most basic of animal instincts and essential for survival. Hearing forms an essential part of this awareness and is a basic sense that plays a vital part in the global development of an individual. Hearing loss cripples the ability not only to hear but also to develop speech and in turn leads to severe communication problems. Depending on the mechanism of hearing loss, there may be a problem in the conductive apparatus or the sensory organ and neural pathways. Most of the conductive hearing losses are correctible and never absolute. However it is the treatment of sensori-neural type of deafness that has evaded scientists over the years. The last couple of decades have seen inroads into this frontier and management has moved beyond conventional fitting of hearing aids and lip-reading. Hearing loss is one of the most frequent congenital anomalies with severe hearing loss occurring in up to 1-6 per thousand live births [1]. Armed Forces personnel are a vulnerable group for sensori-neural hearing loss (SNHL) with exposure to loud occupational environmental noise through out their service. Some of the newer developments in detection, rehabilitation of hearing impaired, recent research trends and future directions in this field are being presented in this communication.

Role of Immunisation

The initial step to deal with this handicap is to prevent it from happening in the first place. The preventive aspect of this horrendous disability is gaining increased importance as people realize the socio-economic costs of deaf mutism.

There are many infections that can lead to damage of the labyrinth and thus causing a sensorineural hearing loss. The role of Haemophilus influenzae in causing sensorineural hearing loss following neonatal meningitis is well known and studies estimate an incidence of up to 25% cases develop hearing handicap [2]. Meningococcal meningitis too is well known to cause hearing loss in the

adult population. The other common infections that lead to hearing loss are viral infections like mumps, measles, rubella, cytomegalovirus, herpes virus, Varicella zoster, and tubercular meningitis.

Most of these infections are preventable by appropriate vaccination. The Measles and BCG vaccines are already integrated into the national immunization program. Vaccines also exist for the other infections like Varicella zoster, CMV, mumps and rubella. Rubella is a common cause of hearing loss due to maternal infections or direct infection of the child. Vaccination for Haemophilus influenzae, mumps and rubella could also be included in the national program. The Armed Forces Medical Services have an efficient and strong delivery mechanism for vaccination and should take the lead in incorporating newer vaccines into the immunization schedule. Thus improved coverage of already available vaccines would be the most cost-effective way of reducing cases of hearing loss in children.

Universal Hearing Screening Programme

The consequences of missing the diagnosis of congenital hearing loss at an early age are grave. Inability to hear leads to inadequate feedback to the brain which fails to develop speech and results in a life of handicap. Whatever interventions are available, work best before any neural plasticity has set in. The best results are obtained when rehabilitative measures are taken by six months of age. To this end, a universal hearing screening programme can be of immense value. The need of identifying such cases is not new but lack of a reliable objective measurement prevented implementation of such a programme. With the advent of newer technology in the form of portable auditory brainstem evoked response (ABR) and oto-acoustic emissions (OAE), such a task is now simple. Most of the developed world today has statutory mechanisms in place which make it mandatory for hearing screening before three months of age. In fact a Joint Committee on Infant Hearing has

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issued comprehensive position statement including principles and guidelines for early hearing detection and intervention programmes in the United States [3]. In our country we do not have such a system and most cases unfortunately go undiagnosed even amongst the educated population. The average age of detection and hearing aid fitting in our country is about six years which is well beyond standard guidelines. An early assessment of these cases would have reduced if not eliminated the handicap they suffer today. The Armed Forces Medical Services have an inherent advantage in that all institutional births in our service hospitals can be easily screened. Those children identified as hearing impaired on screening need further assessment and close follow-up as some of cases do develop hearing subsequently. The role of testing with both ABR and OAE is important to reduce the number of false positives and false negative cases. There is also a need for including a systematic screening programme for children in the pre-school and primary school level.

Early Assisted Listening

After the disability has been diagnosed, it is important to counsel the parents and provide them the right inputs to enable them to make the right choice. Infants which do not develop hearing by six months should be enrolled for appropriate intervention. The aim of managing these cases is to provide earliest restoration of hearing input to the brain. The strategy depends not only on the age of diagnosis but more importantly on the available services and social support. Thus these interventions need to be tailor-made to the situation. All such infants need to be provided with an appropriate hearing aid and considered for cochlear implantation at the earliest. Recent evidence points to uncorrected hearing loss as a cause of cognitive decline in children [4] and it for these reasons that it becomes more imperative that every child with hearing loss be proactively fitted with hearing aids.

Bionic Ear

The Cochlear Implant or Bionic Ear as it now referred to, has added a new dimension in the management of hearing impaired. Hearing stimuli are directly passed on to the nerve endings bypassing the entire sound conducting apparatus. Improved performance due to advanced speech processing technology and electrode design has made cochlear implantation the cornerstone in the management of a deaf-mute child. Again the stress is on early implantation as the results are most gratifying in the younger age [5]. A well-timed cochlear implantation can make a deaf-mute handicapped child lead a fully useful and socially productive life. The Armed Forces Medical Services already has a fully working and established Cochlear Implant Programme that is

showing promising results. There is a potential for fruitful clinical research into improving the long term benefits of cochlear implantation and associated technologies.

What is New in Cochlear Implantation

The cochlear implant technology is also evolving and newer technical advances are being incorporated to optimize the benefit obtained. Newer materials have enabled the electrode array to bend and snugly fit around the modiolus, the spiral part of the cochlea that improves the surface contact with the surviving neuro-epithelial tissue. Research is also on incorporating newer electrode designs with irregular surface so that a larger surface area is available for stimulation thereby achieving stimulation of more number of ganglionic cells. Coating of the electrode with growth factors that stimulate ganglionic proliferation is also a topic of intense international research [6]. These so-called “therapeutic electrode array” implants promise to be the implants of tomorrow. The miniaturization of the cochlear implant has also taken place and totally implantable implants are no longer a dream. Only the power source poses a problem. At the behest of the President of India, DRDO and the AFMS have undertaken the task of producing an indigenous Cochlear Implant so as to make it affordable to the population.

Noise Induced Hearing Loss

Without doubt, Noise Induced Hearing loss (NIHL) is the single most important cause for preventable hearing loss in this world today. This has more relevance in the Armed Forces and one has to accept that it is practically impossible to reduce noise levels in our profession to safe enough levels for infinite exposure. Educating our clientele about adverse effects of noise and its prevention and the use of personal hearing protective devices are the major strategies against NIHL. Other important areas of newer research is the use of antioxidants in the treatment of noise trauma and role of “conditioning” or “toughening” of the ear by prior exposure to low intensity noise before exposure to damaging noise. Another new development in this field is a surgically implanted amplification device on the ossicular chain [7]. These electrically driven devices are placed on the incus and amplify the natural ossicular motions thereby delivering more sound energy to the oval window. An internal receiver is implanted in the skull and drives this device. In noise-induced deafness where hearing loss is not profound, these devices may help in rehabilitation. This eliminates the need of a hearing aid and problems of auditory feedback caused by hearing aids. Cochlear Implant remains a viable option in cases of severe to profound NIHL.

There is an immediate need to take the problem of NIHL as a serious public health problem at the national level and take appropriate steps on a priority basis to reduce the exposure to the susceptible population.

Beyond Cochlear Implant

What if there is no cochlea or the auditory nerves are absent or damaged on both sides. The answer lies in reaching higher up the hearing pathway and directly stimulating the cochlear nucleus. A device, called the Auditory Brainstem Implant is the first bionic device that directly stimulates the central nervous system and has shown some benefit. However experience with this device is limited and there is scope for research in this area. A lot of research has also been done on the possible uses of stem cell therapy in managing sensori-neural hearing loss [8]. Techniques like hair cell regeneration, gene transplantation in cochlea and role of embryonic growth factors show us the future directions and well may be the future treatment modalities of sensori-neural hearing loss.

Conclusion

The coming times may see newer advances change the face of managing sensori-neural hearing loss. The Armed Forces have to keep pace with the latest developments in the field and lead the field in the sub-continent. The frontiers of today would be accepted practices of tomorrow. It is very important to set-sail towards these frontiers and discover new lands. I implore the medical professionals of the Armed Forces

to take upon the challenging task of undertaking cutting-edge research in such fields so that the dream of “Let All May Hear” becomes a reality.

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