CHAPTER 1: INTRODUCTION

1.1. Background of the Study

(a) Hearing Loss – The Indian Perspective

Among the five special senses which humans possess, the sense of hearing hails to be the most important one, as it is crucial for the development of communication, which forms the basis of human civilization. Hearing loss at birth often remains undetected as a silent handicap until it ends up as a double tragedy, of deafness along with speech and language deprivation. ‘Deaf & Dumb’ individuals are considered to be a social stigma even in present day society living within a deaf world, with no means of verbal communication and thus leading a non-productive life.

As per the WHO report of 2010, nearly 2-3 per 1000 live births are found to have severe to profound hearing loss, making it the most common congenital abnormality to affect newborns worldwide. This scenario is even more pronounced in developing countries like the Indian subcontinent, especially with the problems of consanguinity and poor peri-natal care. India has a population of over 1 billion, of which an estimated 3 million children are affected by congenital hearing loss of varying degrees. Every year around 25,000 children are newly diagnosed with congenital severe to profound deafness, across the country. The above data from the National Program for the Prevention and Control of Deafness, published by Garg S et al, in 2011
emphasizes the gravity of the situation in India. However today, hearing loss is the only truly remediable congenital handicap, due to remarkable advances in biomedical engineering and surgical techniques.

**(b) Auditory Neural Prostheses**

The advent of auditory neural prostheses like the Cochlear Implant, have successfully broken the acoustic barrier, thus integrating children born with hearing loss into the normal society, providing them with vital communication skills to lead a highly productive life. The Human auditory system is unique in its organization, due to the phenomenon of tonotopicity (place-pitch organization) which gives it the opportunity to receive and integrate external electronic circuits (Laneau J & Wouters J, 2004). The Cochlear Implant is therefore considered as a monumental innovation of the twenty-first century, as it represents the most successful attempt by man, to interface a prosthetic device with the central nervous system, thereby re-establishing a lost special sense.

Today, Cochlear Implantation has been established worldwide as a successful time-tested technology for restoration of hearing in individuals with bilateral severe to profound cochlear hearing loss, un-aidable with conventional hearing aids. In India, around 12,000 Cochlear Implant surgeries have been performed till date, at various reputed centers across the country.
But, a very large population of more than one million potential implantees is still present in India, most of them being children with profound hearing loss. An early intervention with cochlear implantation is necessary to make a remarkable transition in the lives of these profoundly deaf children, bringing them out of the gloomy world of silence, onto the vibrant world of sound.

Cochlear implants are electronic devices designed to detect mechanical sound energy and convert it into electrical signals that can be delivered to the cochlear nerve, bypassing the damaged sensory hair cells of the cochlea. The cochlear implant comprises of an externally placed microphone, speech processor, a radiofrequency transmitter along with an internally placed receiver - stimulator over the mastoid bone and an electrode array within the cochlea. The acoustic signals are processed by the external speech processor into electrical signals and sent via the radiofrequency interface into the electrode array. The implant system preserves the tonotopic map of the cochlea and the auditory brain perceives these electrical impulses as sound signals.

The field of Cochlear implantation requires a dedicated team of professionals including CI surgeons, audiologists and auditory verbal habilitationists. The success of Cochlear Implantation is directly dependent upon its ability to address the issue of patient’s / family’s expectations and balance it with the eventual outcomes of auditory verbal habilitation.
Variables affecting the overall outcome of Cochlear Implantation are many, like the age at onset & duration of deafness, amplification history, pre-implant communication mode, the surgical expertise and challenges, type of cochlear implant & speech processor used, patient motivation, family support and habilitation facilities. Hence, it is paramount for the CI team to meticulously monitor the performance of the implantee over the period of auditory verbal habilitation and later on at follow ups, as & when necessary.

### 1.2. Lacunae in Knowledge

As cochlear implant technology continues to evolve today, the need arises for research in the various fields associated with cochlear implantation, in order to refine the standard protocols and methods, to comprehensively monitor the devices and to provide data on the performance of the implantees over time of implant use. Research has focused on improving electrode designs and speech processing strategies, in order to deliver a range of realistic sounds via the implant and provide a gratifying experience to the implantee in various acoustic environments.

The inclusion criteria for Cochlear Implantation, has thus expanded in recent times in various aspects, to include candidates ranging from post-lingual adults with partial high frequency hearing loss, onto children as young as six months of age with congenital profound hearing loss. Audiologists and
Habilitationists need to judiciously supervise the implant use especially in very young implantees or those with multiple handicaps and a poor CI performer needs to be identified at the earliest and appropriate corrective measures need to be taken. Such a poor CI user, when assessed for hearing skills may be found to have become less attentive to sound stimuli or may no longer hear sounds. He or she may further confuse sounds previously recognized or learned & may request repetition or clarifications from the habilitationist. There would also be depreciation in speech skills with decreased vocalizations, changes in pitch or loudness of voice, increased nasality, loss of intonation & changes in speech articulation.

Furthermore, the general behavioural responses also deteriorate with slower response times, change in demeanor and a less interactive, withdrawn nature. The plethora of above symptoms would be a cause of concern for the entire CI team & the implantees’ family. A multitude of Electrophysiological tests are available today to provide a helping hand to reconfirm the integrity of the implant in such clinical scenarios. Objective confirmation by these tests helps to alleviate the mortal fear of a device failure in the minds of the Implant team.

As many young children and those with multiple disabilities and syndromic associations are being implanted today, even experienced audiologists face the daunting task of programming such ‘Difficult to Measure
Auditory Percept (MAP)’ children using conventional methods, since it is often difficult to interpret behavioural responses without ample cooperation from them. In such cases, matching an effective MAP and appropriate MAP becomes the key issue in the habilitation program. Candidates with multiple handicaps may have cognitive problems, developmental delay, attention deficit etc, and behavioural responses may be inconsistent in such cases, since they vary depending upon their age, listening experience and cognitive abilities (Shallop et al, 1995; Spivak et al, 1994; Hodges et al, 2003).

In ‘Difficult to MAP’ scenarios, objective electrophysiological tests become paramount to pave the way forward and provide optimal current levels for setting in the MAP. Studies have shown that intra-operative and post-operative objective electrophysiological tests like Electrically Evoked Compound Action Potentials (ECAP), Electrically Evoked Stapedial Reflex Thresholds (ESRT) and Electrically Evoked Auditory Brainstem Response (EABR) thresholds correlate well with behavioural programming levels and these measurements may be used to ascertain an optimal behavioural MAP for the implantee (Hodges et al, 2003; Mason, 2004; Abbas et al, 2000).

Electrophysiological tests like ECAP and ESRT are usually performed to intra-operatively confirm the correct placement & functioning of electrodes within the cochlea and also for troubleshooting purposes, in case of a suspicion of a device malfunction. These tests have been declared to have a
good sensitivity rate of around 80 – 83% as per literature quoted by the different implant companies and hence have been marketed along with the Mapping software commercially. Thereby, when necessary, they are clinically applied by audiologists, in the post-operative period to ascertain the thresholds for setting an initial Map for an uncooperative subject or in a spectrum of ‘Difficult to MAP’ situations.

Audiologists do face a practical dilemma at times, when a candidate who had excellent ECAP / ESRT responses during surgery, does not show good behavioural responses while Mapping and eventually becomes a sub-optimal / poor CI user. On the contrary, there may also be instances wherein after an uneventful CI surgery, intra-operative electrophysiological responses are negative for reasons not yet fully understood, but the Implantee responds well at ‘Switch-on’, producing a normal behavioural Map and evolves into a good CI user. Hence, today it is widely accepted that intra-operative tests when successfully elicited, confirm device integrity on table, but may not reflect upon optimal implant performance later on during Mapping, habilitation and beyond.

Hence, in clinical practice, when a ‘Difficult to MAP’ scenario is anticipated or encountered, audiologists prefer to perform intra-operative and post-operative electrophysiological tests like ECAP measurements (NRI = Neural Response Imaging in Advanced Bionics Implants / NRT = Neural
Response Telemetry in Cochlear Nucleus Implants / ART = Auditory Response Telemetry in MedEl Implants) or ESRT (available in all the above Implant types), in order to get an idea on the optimal current level required for stimulation via the implant. They incorporate these current levels into the programming software to set a baseline MAP at ‘Switch-On’, and further redefine the levels thereon using psychophysical behavioural responses of the child. This method is quite successful for providing a working MAP for the child at ‘Switch-On’ and later-on fine-tuning the MAP is based on the child’s habilitation performance and psycho-acoustical feedbacks.

Sometimes in clinical practice, there have been situations where a child’s behavioural Mapping levels were found to be inappropriate or erroneous and hence the habilitation outcomes were sub-optimal (Brown et al, 2003; Gordon et al, 2004). Such children may return to the Audiologist for trouble-shooting and Re-Mapping, during which they need to rely upon objective electrophysiological responses of the auditory nerve, in order to accomplish the task of re-defining the MAP. Recent Mapping software, have provision for incorporating the electrophysiological current levels (tested intra-operatively or post-operatively) into the programming module for setting an ideal MAP (Basta et al, 2007). At times, such a method may not be very successful, due to inherent disparity between the electrophysiological current levels and the actual behavioural current levels which need to be set in the MAP. While ECAP thresholds help to identify the current levels required to
stimulate the auditory nerve, they may not evoke an optimal behavioural response from the child when set in the MAP. This disparity has been implicated to the variation in parameters like the stimulation rate and pulse duration, while measuring an ECAP and while programming a MAP (Davids T et al, 2008; Craddock et al, 2004; Kaplan-Neeman et al, 2004). A higher stimulation rate is used in Mapping for optimal processing of stimuli, while a lower stimulation rate is preferred while performing ECAP measurements, since accurate electrophysiological thresholds can thus be identified (Gordon KA, 2004; Davids T, 2008).

Research work done at various centers across the world, have shown that electrophysiological correlates of the auditory nerve are not identical, but vary with behavioural responses individually. Literature states that ECAP thresholds may be successfully recorded in approximately 80 - 83% of cases, but are not sensitive to identify accurate Mapping levels (Mason, 2004; Brown et al, 2003). ESRT is known to over-predict the optimal behavioural comfort levels during the initial period of habilitation (Spivak et al, 1994; Hodges et al, 2003; Stephan et al, 2000) and EABR though reliable, is found to be cumbersome, time-consuming and impractical to be done electrode-wise, in order to comprehensively program a cochlear implantee (Abbas, 2000; Brown, 2003; Shpak, 2004).
Hence, no single electrophysiological test has been found to have high
sensitivity and reliability for setting an ideal MAP and behavioural measures,
even if minimally available remain indispensable while programming the
cochlear implant (Gordon et al, 2004). This fact warrants further research in
the realm of cochlear implant electrophysiology, which the present study tries
to explore by combining the clinically available tests together as a test battery
and assess their clinical applications.

1.3. Need for the Present Study

In the Indian context, over the recent years, electrophysiology is more
widely discussed among Audiologists at National podiums, like the ‘Annual
Conference of the Cochlear Implant Group of India (CIGICON)’ and data is
emerging from various reputed implant centers across India, but there have
been no publications or research studies, longitudinally analyzing and
documenting the intriguing changes in electrophysiology among cochlear
implantees, during the habilitation period. Though, results from the western
world are widely read & accepted among our Indian professionals, indigenous
research data is yet to emerge, in support of the western literature.

In our clinical scenario, there has been a lack of normative data and
reference values, for electrophysiological tests and behavioural responses,
which may be used as a guideline for optimally programming cochlear
implantees. Thus, a lacuna remains till date in refining the set parameters of this western technology to ideally suit the Indian implantees. Chennai, being a pioneering city for cochlear implantation in India for more than a decade, with multiple reputed centers performing cochlear implants, and with a large population of cochlear implantees, does not have any previous data on electrophysiological studies for reference.

This practical fact triggered the need for this multi-centric research work, which was conceptualized to evaluate the clinical efficacy of a multi-modal electrophysiological test battery in predicting behavioural responses among cochlear implantees during their habilitation period.

1.4. (a) Study Hypothesis

The study hypothesizes that, multi-modal longitudinal correlations of various objective electrophysiological tests with subjective behavioural comfort levels recorded in a cohort of comparable cochlear implantees, would help to statistically predict reliable and optimal behavioural comfort levels (when unknown) for a member of the cohort, using the linear and multiple regression analysis methods, rather than using a single electrophysiological threshold for direct incorporation into the MAP, which has been the conventional method clinically followed, when a ‘Difficult to MAP’ situation is encountered.
1.4. (b) Anticipated Outcomes of the Study

This prospective clinical study was designed to explore the intriguing relationship between the objective electrophysiological responses of the auditory nerve and the subjective behavioural responses of the implantee, as obtained by psychophysical stimulation via the cochlear implant over time in the habilitation period. The results from this study would provide indigenous data and reference values for future research work in the Indian clinical scenario.

The anticipated outcomes from this study, was to document a longitudinal positive relationship between the two parameters compared, thereby establishing a protocol which may be followed as a guideline for successful programming among ‘Difficult to MAP’ cochlear implantees in future. This study may further help to refine the existing Mapping protocols and standardize the inclusion of electrophysiological tests for objective monitoring of the cochlear Implant in the habilitation period.