EARLY INTERVENTION – NO REHABILITATION?

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Early detection of deafness, followed by early intervention – will this mean, that there is no need for habilitation or rehabilitation any longer?
I will attempt to answer this question by reviewing the current situation and by drawing a future scenario.

The importance of early detection is generally accepted regardless of the kind of disease we are confronted with. More than 20 years ago the ophthalmologists already agreed that in case of a congenital cataract the opaque human lens had to be removed within the first two weeks of the babies life. Otherwise the risk of ambliopia (permanent vision impairment) was very high. Whether the appropriate intervention is to provide the baby with a contact lens or with an intra-ocular lens is still a hot topic.
The more conservative ophthalmic surgeons hold the view that one should wait with an IOL until the eye is fully developed and this takes approximately 18 years. Only then the secondary surgical intervention should happen. The more aggressive surgeons (mainly in the US) prefer to implant the IOL during the first surgical intervention, i.e. the cataract extraction. If need be the IOL can be replaced at a later point in life.
Experiments with kittens who received a black lens to make them temporarily blind clearly showed the long-term negative effect and this helped the ophthalmologist to decide for early intervention.

Unfortunately, it is not possible to deafen animals temporarily and to look for the long-term effects (ototoxic and surgical treatments result in permanent deafness) and therefore physiologists simply drew a parallel from the visual system.
They made the hypothesis that deafness is identified at a comparatively late point in time (sadly enough the average age of children who receive their first hearing aid in Germany is still 2years and 4 months!) and therefore the implantation of a Cochlear Implant could not have any significantly positive effect. The lack of conclusive results based on animal trials made it nearly impossible to identify the critical time window when the provision of a Cochlear Implant would be beneficial for deaf born children.
Nevertheless, scientists all over the world - like Djourno and Eyries in France in 1957and Zöllner and Keidel in Germany in 1963 - firmly believed in the concept of Cochlear Implant. The description of desirable features for

a C.I. published by Zöllner and Keidel is remarkably similar to the implementation many years later. Bill House – undoubtedly one of t h e pioneers in the field - had the courage to provide adult patients with his single channel device and presented for the first time at the World Congress in Venice in 1971. A. Chouard, who had previously worked with Djourno and Eyries developed his concept and started to perform surgeries in the seventies. Graeme Clark finally introduced the multi-channel CI concept and the first patient was operated in Melbourne in 1978.
As these post-lingually deafened adults showed positive results, otologists felt encouraged to broaden the indication and lower the age of potential candidates step by step. This approach, which we may call "trial and error" led to select also candidates, who turned out to be not really suitable. These were mainly deaf born adolescents, who did not fulfil the level of expectation, neither of their parents nor of the professionals.

At the time, when the first very small children received a Cochlear Implant, this decision was considered to be very courageous, some people even called it "criminal". The very impressive progress these children achieved in speech perception, speech production and language acquisition forced even the most critical sceptics to acknowledge: CI is a viable treatment for deaf born children. The validity for deafened children and adults was proven already and provided a sound basis for broadening the indication.

Personally, I remember the first only one and a half year old girl that Ernst Lehnhardt implanted in Hannover in 1988. The majority of his team protested and doubted that this will be a success. The young lady who's mother is German, father is Palestinian, lives in Dubai, speaks three languages (English, German and Arabic) fluently, attends normal school and enjoys music. Many more children, who received their CI at a very early point in time, show similar positive results. The average age of children who are considered to be suitable candidates is constantly decreasing and in Germany for example already 10% of the CI population is below the age of 2 years.

Average results in open set speech understanding have significantly improved over the years. This has led to further broadening the indications: According to the Food and Drug Administration adult candidates qualify when they achieve less than 40% open set speech understanding binaurally with well-fit hearing aids. In practice this varies between 30% and 50%!

Infants are considered to be candidates at the age of 12 months when they are deaf born and when they plateau in their development with well-fit hearing aids. Children post-meningitis should be implanted as soon as possible to avoid the risk of ossification of the cochlea. Indeed, the youngest child who has received a CI is only 4 months old.

There are many key factors that contribute to the results we can achieve nowadays. Some of them are patient inherent, like duration of deafness, degree of hearing loss (we have evidence that CI recipients with residual hearing have better results than recipients who are totally deaf), other disabilities or an anomaly of the cochlea that may prevent total insertion.

Other key success factors are definitely:

- The quality of the CI device
  It must be biocompatible, made of flexible material to follow the shape of the skull of an infant, have a removable magnet to allow for MRI, provide the platform for implementing a range of modern speech coding strategies and have the capability of recording the ECAP (Electrically Evoked Compound Action Potential) using Neural Response Telemetry.
  It must be reliable, i.e. have a very high cumulative survival rate, as many of the young recipients would hope to benefit from it for more than 70 years.
- A variety of speech coding strategies and stimulation modes must be available to enable the audiologist / engineer to provide the patient with the most suitable custom made programme. Clinical trials on a worldwide basis
  have clearly demonstrated that there is no “one fits it all best” strategy as recipients are individual and have their very individual needs.
- The surgical procedure needs to be constantly reviewed and improved.
  It seems to be obvious that with new candidates (very young infants and patients with residual hearing) as well as with the development of new devices (smaller and most recently modiolus hugging with a stylet) the surgical approach needs to be refined.
  In the future we may look at a combination of CI and Hearing Aid in the same ear and we may look at intra-cochlear-extra-luminar electrodes, we hope to have a totally implantable device.
  Again, there is an interesting parallel from ophthalmology, the cataract extraction. In the early times the lens was simply pushed back into the bulb
  then it was taken out with a loop and for this the cut into the cornea had to

be the size of the human lens. With phako-emulsification the cut could be much smaller as the lens was cut into little pieces and sucked out. The size of the cut was now determined by the diameter of the IOL, this was then reduced by the development of a foldable silicon lens. Continuous improvements in the development of the surgical technique and the design of the implant.
This is no different in CI surgery and we are aiming at soft surgery, less traumatic, less invasive following the medical principle "Nihil nocere".

- The skills of the audiologist/engineer are also very important, as we said before, to provide the patient with the Custom made Sound.
  With the very young infants, who are not cooperative, we are looking for tools that help to establish a first map that is providing sufficient auditory input without any risk of over-stimulation.
  Neural Response Telemetry has become a clinical tool (used in hundreds of clinics with thousands of adults and children) to predict Threshold and Comfortable Levels for the recipient's map. The T-NRT levels typically lie between the T- and C-levels on average at about 70% of the dynamic range.
  Fully automated fitting procedures, which we may envisage for the future, are definitely not indicated yet.
- (Re) habilitation, finally, is another very important success factor.
  Teachers, therapists and logopeds benefit from more experience with CI recipients and refine their methods as well. No doubt, auditory-verbal therapy is one of the most successful in helping deaf people to listen, to understand, to acquire speech and spoken language. In May this year, Warren Eastabrooks held a two days workshop in Wroclaw, some of you may have attended and hopefully took the message home, that habilitation is a key success factor to integrate a deaf child into the hearing world.

As we said before, there has been a substantial increase in the overall recognition abilities of CI users during the past 15 - 20 years. Despite this improvement the range in individual differences remain from a simple awareness of environmental sound to near 100 % open set speech recognition. (Curtis. W. Ponton, Lyon).
The question is: WHY? And we want to find out by applying objective measures to acquire knowledge about the effects of auditory deprivation on the plasticity and development of human central auditory pathways, the neural maturation in congenitally deaf children, the developmental plasticity in the auditory brainstem and midbrain in children with congenital hearing

loss, the potential application of electrophysiological indices in training and rehabilitation programmes (Customized Training for CI users).

These were the topics in a recent International Symposium and Workshop on "Objective Measures in Cochlear Implantation" in Lyon in March this year.

I would like to share with you a few results and conclusions, which I find of special interest.

We know that in a normal hearing infant the inter-peak latency between the waves Jewett I and V (the nucleus cochlearis and colliculus inferior) shows a significant shortening during the first 12 months after birth. (from 5,1 to 4,2 milliseconds). Afterwards it remains basically constant. This means that the maturation process is happening in the very early time of life. (Lehnhardt "Praxis der Audiometrie", Thieme Verlag, 2000, 8th edition, Page 27).

A similar maturation process has been proven by T. Van Den Abbeele et al (Paris) in congenitally deaf children with CI. Electrical Auditory Brainstem Potentials (EABR) were recorded in all children at the end of the surgical procedure of CI and after 9 – 12 months of daily use.

The value of the mean latency of wave V improved from 4,12 milliseconds to 4,07 milliseconds. The improvement for the congenitally deaf children is more significant than for the progressively deafened children. The conclusion is, that congenitally deaf children improve EABR after one year of daily use of the CI and this suggests that electrical stimulation induces a maturation of central auditory pathways. The authors believe that these results support the concept of early implantation in congenitally deaf children.

(Lyon Abstracts, page 29).

A different aspect - the time during which the human auditory system remains relatively intact ("non-degenerate") and/or highly plastic in congenitally deaf children - was investigated by Anu Sharma et al (USA). P1 and N1 cortical auditory evoked potentials were recorded in deaf children with CI. Children implanted after the age of 4 years had delayed cortical responses even after years of stimulation. Children implanted at the age of 3 years, in contrast, evidenced normal latency responses within months of electrical stimulation. These data suggest that the central auditory system does not suffer from degeneration for about 3 years despite the lack of normal auditory stimulation.

P1 and N1 latencies where retested several months to more than a year after onset of electrical stimulation. A large decrease in latencies

following 5 months of electrical stimulation was found in children implanted at ages between 2 and 6 years. Children implanted after the age of 11 years, however, did not demonstrate decreases in P1 and N1 latency. This seems to indicate, that too long a period of lack of stimulation may result in a loss of plasticity in the central auditory pathways.
In summary it appears that the degenerative effects of deprivation (lack of stimulation) of the central auditory system become apparent already after 3 years, whereas the loss of plasticity of the central auditory pathway (the possibility of re-occupation of the auditory parts in the brain) appears at a later point in time, probably after the age of 6 years. (Lyon Abstracts, page 36).

Gordon et al in Toronto presented their finding on the "Developmental plasticity in the auditory brainstem and midbrain in children with congenital hearing loss who use a CI". They wanted to find out whether the auditory brainstem and the midbrain show developmental plasticity in children with congenital severe to profound hearing loss using a CI. They recorded EABR in 35 children beginning with the first week of device activation extending over the fist year of implant use. They found that apically and basally evoked responses are consistently present at initial stages of implant use and show significant decreases in latency over time post-activation. Because the pathways are normally myelinated by one year of age the authors believe that the latency changes reflect a development occurring primarily at the synaptic level. Moreover, these results indicate the presence of plasticity in the system despite auditory deprivation in early development.
In a poster Gordon et al from Toronto described the "Normal sequence of auditory cortex development in children with congenital hearing loss who use a CI". They quote recent studies that have provided new insights into the sequence of normal development in the human auditory cortex using late auditory evoked potential recordings : Eggermont et al, 1998; Pang and Taylor, 1999; Ponton et al, 2000). Findings reported from a midline electrode (CZ) show that a positive wave (P1) occurring at approx. 100milliseconds post stimulus onset, found in children as young as 6 years of age, is gradually impinged upon by a negative wave (N1). The latency and amplitude of P1 decrease as the amplitude of the N1 increases. The underlying mechanism for this phenomenon remains unclear but the role of auditory stimulation may e investigated by

studying children who use a CI. The authors recorded late auditory evoked potentials in 16 children with congenital hearing loss with CI. They ranged in age from 3,8 to 15,5 years and had various durations of implant use. Their results show that the normal sequence of auditory development does occur in these children based on increasing chronological age rather than on duration of implant use. Responses from young children tended to show a prominent positive wave at 100 – 150 milliseconds while a negative wave at approx. 100 milliseconds was found in several older children. Gordon et al conclude, that the development in the auditory cortex is, in part, dependent upon age-elated changes occurring at this level.

As early as in 1997 (Acta Otolaryngol. Stockh) Eggermont already presented his studies on the effects of prolonged auditory deprivation in children in whom stimulation was restored by a cochlear implant. The latency of the P1 component of the late cortical potential was used as the indicator of auditory system maturation. For normal-hearing children there is a gradual evolution of evoked potential features that extends through adolescence with P1 latency becoming adult-like at about age 15. It appears that maturation of P1 latency in normal and implanted children occurs at the same rate, but the time to maturity in implanted subjects is delayed by an amount approximately equal to the duration of deafness.

This reminds us of what we said earlier about the maturation using wave V measurements.

Two years later Eggermont together with Ponton and Moore published in the Scandinavian Audiology Supplement about the fact, that prolonged deafness limits the auditory system developmental plasticity. They claim, that despite the tendency to implant children at an earlier age, little is known about how auditory deprivation and subsequent implant use affect the maturing central auditory system. Previously reported results indicate That stimulation of the auditory system by a CI is sufficient to restore at least some aspects of central auditory pathway maturation, as reflected by age-related changes in the auditory evoked potentials. Eggermont's and Ponton's analyses show that age-related changes in the Evoked Potentials may asymptote at levels different from those found in the adult norma-hearing population. They suggest that maturation of at least some aspects of central auditory system activity is limited by the onset and duration of the period of deafness prior to implantation.

I believe, this again advocates early implantation.

Ponton in Lyon now presented "The application of electrophysiological indices in training and rehabilitation programs for CI users".
So-called poor performers are mainly the pre-lingually deafened adults. They too normally perceive sounds (acoustic events), so therefore the problem in performance is not at the level of stimulus detection.
It is likely at the level of sound discrimination (which may reflect poor neural survival in the cochlea) and/or at higher levels of central auditory processing. The approach used by Ponton is the use of mismatch negativity as a measure of auditory discrimination. The MMN was used to guide and monitor discrimination training of sounds generated by stimulation of different electrode pairs along the cochlear implant's electrode array. Results from this combined electrophysiological and behavioural approach to training and rehabilitation not only demonstrated significant improvements in electrode discrimination, but an apparent generalization to enhanced levels of performance in syllable identification and open set speech recognition. This may also provide important insights into changes and plasticity of the central auditory system associated with CI use.

Sharon Phipps et al from Nottingham reported about the MMN as one of the two event-related potentials (the other is he P3), which can be used objectively to assess the ability to discriminate between different sounds in Lyon as well. She drew the same conclusions, i.e. it could be used to initiate rehabilitation of vulnerable children within the early years of implantation.

So what about the future?
Smaller implants, totally implantable, invisible thin film electrode arrays and in combination with a hearing aid for the babies as well as for the elderly with residual hearing?
Yes, most probably within the next 5 – 10 years.

Neural Growth Factors will stimulate the re-growth of nerve fibres (dendrites) and improve chances for excellent results especially for children, for whom the critical period for plasticity has passed?
Graeme Clark in the American Journal of Otology, Vol. 20, No 1, 1999 says: "Neurotrophin would be released from the CI electrode, pass to a

receptor site on the synapse between the auditory nerve fiber and cochlear nucleus cell, which would facilitate the release of transmitters across the synaptic cleft. These would activate a signal cascade of proteins in the cytoplasm that would reactivate the gene for the neurotrophin. This would also cause the neural sprouting required for the coding of sound to re-establish auditory plasticity. The neurotrophin produced in the cochlear nucleus would also propagate the higher auditory centes, establishing appropriate neural connections along the auditory pathways".
So, probably yes, this will be the future in the next 15 years.

Better reproduction of coding of sound to provide better perception of speech and other sounds in noise and make CI users enjoy music?
Very likely, progress will be ongoing.

Refined and less invasive surgical techniques to cause as little trauma to the delicate structures of the inner ear?
Definitely, surgeons are trying new methods and new instruments are under investigation.

Fully automated fitting procedures?
I do not believe that this is desirable. We do want tools to help us establish initial maps for small infants and for less co-operative children. We do want to save time and get suggestions from the software what kind of parameter is suitable for the individual patients. But we do not want fully automated procedures where the recipient is not heard any longer and does not get the level of attention from the clinical team he is expecting and hoping for.

No habilitation because all other so-called success factors are in place?
I do not believe that this is desirable either.
Firstly it is naïve to assume that we will always have the ideal candidate, the "perfect" surgeon, the optimal programming team. We will want to help also candidates who are not perfect, candidates with additional handicaps (30% of deaf children do suffer from an another impairment!) and in order to improve their chances to achieve satisfactory results we will need to develop more research based refined habilitation methods. As neonatal screening will hopefully become routine in the majority of countries in Europe in the very near future and very early professional intervention, including a CI will follow, we need to bear in mind the

effect on parents. They may not have enough time to understand the impact of the deafness of their child, they may not have time enough to grieve and therefore they may need more psychological support than parents of today who have at least a couple of months to accept the deafness as a fact, to make a conscious decision for CI and fully understand the need of continuous active support.