Esitetty (presented at): XXX Valtakunnalliset audiologian päivät, Kuopio 3.4.2009, Finland

The early intervention protocol for young hearing impaired children: what have we learned?

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Until not long ago to wake up the dead, to give the sight back to the blind or to let the deaf hear belonged to the mythical world of wonder. This is still the case for waking up of the dead, but not anymore for the deaf and hearing impaired patients. Nowadays, thanks to the modern otology, most of the deaf can get back most of their functional hearing.

With almost 200.000 implanted patients in the world, the cochlear implants (CI's) have become the most successful story in the ENT field during the last 3 decades. This success is due to enormous improvement of the achieved results that changed from mere assistance in lip-reading to open-set speech understanding in the majority of patients.

Concurrently to other med-tech devices, where improvements of the results are mostly effect of technological developments, the main source of improvement in CI's is our better understanding of the appropriate indications. In particular our understanding of the critical periods and the role of the time factor has been pivoting for achievement of good results with CI's. The time factor involves the decline in brain plasticity with age. This is due to the existence of a "critical time window", before the end of which the linguistic development must be (re-) initiated through auditory input to achieve normal aural/oral communication capabilities.

Through the years abundant literature has been gathered showing that in congenitally deaf children implantation before the age of 18 months allows for normal speech and language development in most children without concomitant pathologies. These deaf children follow the same timeline of speech development as the normal hearing children and are for example able to communicate by telephone with a familiar speaker (Category of Auditory Performance = 7) at the age of 2,5 years. These children have also more than 90% chance for the mainstream education. Implantation before the age of 3 years still allows for obtaining optimal performance in speech and language, but the development is significantly delayed. If the implantation has been performed after age of 3 years, the performance will most probably reach only the moderate level.

This does not mean that we must exclude older children from the CI programme, but that our expectations and counselling should be appropriately adjusted.

Early cochlear implantation programme in Antwerp

At our Department 50% of all paediatric cochlear implant candidates are effectively implanted before the age of 12 months and 90% before the age of 18 months. Such early implantation protocol would not be possible without the universal neonatal hearing screening (UNHS) program, that is available in Flanders from 1998. The UNHS program is based on automated ABR screening ("ALGO" device) and is highly successful. In Flanders the screening is performed at age 4 weeks by the well-baby organization ("Kind en Gezin") and from the very beginning the program was able to achieved the goal coverage above 95% (96,4% in 2007). "ALGO" is very effective in detection of the hearing losses, including the retrocochlear losses and neuropathy.

When a child is identified as potentially hard-of-hearing by the UNHS then referral is made to one of the national referral centres. The task of the referral centre is to confirm the hearing loss, its severity and to perform full aetiological diagnostics.

At the referral centre, after extensive anamnesis, audiological and ENT examinations are performed. We start with micro-otoscopy and 226Hz and 1000Hz tympanometry. For audiological diagnostics we use ABR (with registration of the cochlear microphonic potentials) and TEOAE's/DPOAE's. ABR is performed under general anaesthesia, after paracenthesis. We apply classical clicks via the headphones and also via the bone conductor, in selected cases frequency-specific ABR is performed. Preferably under the same anaesthesia high resolution imaging (CT and MRI) is done. Standard genetic screening for connexine-26 mutations is also performed together with a number of additional examinations looking for potential syndromic associations (kidney ultrasound and ECG). Laboratory data for maternal rubella and CMV infections are controlled. In Antwerp all children born to mothers with confirmed seroconversion during pregnancy are screened for CMV infection (urine culture within the first 2-3 weeks after birth). In the case of a positive culture in non-symptomatic children their hearing is followed on regular basis (every 6 months until the age of 3, later every year until the age of 6 years). In children older than 2-3 weeks with hearing loss and suspected CMV aetiology the PCR test of the viral DNA is performed in blood from the archived heel-prick cards ("Guthrie card") and confirmed by IgG and IgM serological tests in blood.

Currently the aetiological diagnosis can be made in approximately 70% children. The genetic hearing losses (syndromic and non-syndromic) comprise almost 50% of the children with hearing loss, of which the half is due to connexine-26 mutations. In the non-genetic group the most frequent causes of hearing loss are the peripartal complications, congenital CMV infections and anatomical malformations. Our goal is to finish the diagnostic work-up by the age of 3 months.

The severity of the hearing loss should be additionally confirmed by observation of behavioural reactions to sounds (BOA – Behavioural Observation Audiometry). This is usually done by the specialized hearing revalidation centres (see further), where the bilateral hearing aids will also be fitted at the age 4-6 months. During this period it is very important that the children become acquainted with and used to the test situation and to the audiologist, so that after a number of visits reliable behavioural reactions can be recorded and the hearing aids optimally fitted.

Behavioural confirmation of the hearing loss is very important for our decision making. We would never go for cochlear implantation in a child under 12 months based only on negative ABR testing. This rationale is based on our experience with children with dysmaturity of the auditory pathways in the first months after birth (especially in premature babies). In these children the early ABR's showed no reproducible traces up to 90 dBnHL, yet by the age 6-9 month the ABR results improved and became bilaterally completely normal. This is the most important reason for our reluctance to implant children aged under 7-8 month, before a reliable behavioural confirmation of the thresholds can be obtained. The only exception is the hearing loss being a sequel of bacterial labyrinthitis, where early (bilateral) implantation is necessary due to imminent fibrosis and/or ossification of the cochlear fluid spaces.

The next step is the evaluation of the hearing aid benefit. We must know whether these kids hear well enough with their hearing aids to guarantee the normal auditory and linguistic development. Adequate auditory input is prerequisite for the language development, but this "adequacy" is not easy to evaluate in preverbal children. Experience has shown us that mere detection of the sounds comprised in the speech signal is not enough to guarantee that the brain can process this input into meaningful linguistic information, thus generating language. The cochlea must also be in state to perform the spectral analysis of the incoming sounds since this is the basis for discrimination of the phoneme formants. Speech and language development requires further that the information from the cochlea reaches a sufficiently capable auditory brain. The brain must posses the intrinsic capability to process information (identification and interpretation stage) and to learn. Here not only the genome is important but also the environmental factors and the input of information are crucial.

We have to remember that a CI aims to replace only the cochlear (sensory) function and cannot replace the auditory brain. It can only feed acoustic information to the brain. Therefore dysfunction in the candidates for cochlear implantation should preferably be localized exclusively within the cochlea (fortunately this is the case in a vast majority of deaf children).

Many children can do quite well and develop linguistic skills with only very limited auditory input due to very good cognitive functions. Due to relatively good results on the auditory performance tests these children could potentially place

themselves beyond the CI indications while these are just these children who would benefit most from the CI's. On the other hand there are children with quite good cochlear function in whom poor speech and language development is due to the central/cognitive problems. Such children, when implanted, would also show poor result with a CI. The answer to this diagnostic problem would be to create a test that implies as little cognitive or linguistic skills as possible, but gives us as much information as possible about the cochlear function.

Another issue is that most audiometric performance test are based on speech understanding. Identification tests are available only for children older than 2-3 years and are completely impossible for children aged 7-8 months, when the evaluation of the adequate auditory input via the hearing aids has to be made. Waiting with evaluation of the hearing aids benefit until the age when the speech tests are possible to perform is the main reason for delayed cochlear implantations in a few European countries.

Therefore we have developed a phoneme discrimination test that is aimed at evaluation of only the cochlear discrimination function, does not presume linguistic skills and is feasible to be perform at the age 7-9 months. This test comprises 22 pairs of intensity-balanced phonemic contrasts typical for the Dutch language that are presented at 70dBSPL with intensity roving. The response is based on evaluation of the conditioned orientation or instrumentation reflexes. Our experience has shown that the children who show poor cochlear function on the phoneme discrimination tests in the aided condition are the best candidates for cochlear implantation.

The phoneme test is used not only to diagnose the cochlear function in candidates for CI, but also post implantation to follow up the rehabilitation progress. The consequent errors in phoneme discrimination supply analytical information useful for adjustment of the fitting parameters.

Last but not least issue in the selection of paediatric CI candidates is evaluation of the socio-economic factors that play a very important role in successful rehabilitation. What we need is an emotionally healthy, stable, caring, stimulating and consequent learning and rehabilitation environment. Issues of monolingual versus multilingual/multicultural and sedentary versus mobile social setting should be addressed too.

At this stage also a detailed revalidation plan is drafted (see further). Availability of such a plan is necessary for reimbursement of the CI's in Belgium.

Pre-operative otological / medical preparation

When the decision for CI has been taken, we take care that the surgery is performed in an optimal surgical field - a healthy middle ear. If, preoperatively,

otitis media with effusion is present we place a grommet 1 month before the planned CI surgery. The grommet may remain at place after implantation. Presence of the middle ear effusion at the day of planned CI surgery results in postponing of the implantation, on that day only a grommet is placed.

To prevent potentially fatal postoperative meningitis all children selected for CI receive vaccination against haemophilus influenzae B, pneumococcus and meningococcus C.

Importance of imaging

All CI candidates during the diagnostic course receive CT and MRI examinations. In children the examination is performed under general anaesthesia and, if possible, followed by diagnostic ABR/paracenthesis during the same session. The CT/MRI combination is indispensable to detect possible causes of congenital deafness and to evaluate the chances for successful cochlear implantation. Both examinations deliver complementary information and both are necessary to prove the existence of implantable “cochlea-cochlear nerve unit” and the absence of central lesions or concomitant pathology. For example when we are interested in patency of the cochlear fluid spaces, the CT can inform us about a potential ossification, but would be unable to predict presence of fibrotic tissue inside the cochlea. Concurrently, MRI would not be able to differentiate between fibrosis and ossification. CT allows for good diagnosis of the middle and inner ear malformations, pathologic course of the facial nerve, carotid artery, jugular bulb, etc. MRI is necessary for evaluation of the patency and the structure of the inner ear spaces as well as for confirmation the normal anatomy of the acoustic nerve and the auditory pathways. In our database we have recorded more than 100 ears with hypoplastic or aplastic cochleovestibular or cochlear nerve, including 5 children with bilateral total aplasia of the cochlear nerve. Pre-operative knowledge of such pathology is crucial for correct counselling and decision making in CI candidates. In doubtful cases of cochlear nerve hypoplasia or major inner ear malformations imaging could be accompanied by advanced electrophysiological workup, including E-ABR’s, that could potentially help in evaluation of the electrical functionality of the dysplastic cochlea-cochlear nerve unit.

Choice for the side of implantation

When the decision for cochlear implantation has been made, the side of implantation has to be chosen. This decision is made based predominantly on the results of medical imaging, that has to prove existence of an implantable cochlea-cochlear nerve unit (see above).

In small children the audiological factors play a lesser role in the choice of implantation side, because in most cases both sides show comparable thresholds and there is no issue of the duration of deafness, experience with unilateral hearing aids, history of chronic otitis media, etc. Yet the objective and behavioural audiological tests are standardly performed in order to exclude significant asymmetries that could influence the choice of the side of implantation. If one ear has significantly worse non-aided and aided auditory thresholds and/or phoneme discrimination, then the worse side is chosen for implantation.

The vestibular function is usually of no concern in small children, unless bilateral implantation is foreseen or there is unilateral cochleovestibular nerve dysplasia/aplasia. Testing vestibular function in small children with ENG (rotating chair and caloric testing) is possible but technically quite demanding.

If all other factors being equal, the right side is being chosen for implantation based on location of the speech and language centres in the contralateral hemisphere.

Bilateral CI's vs. bimodal stimulation

The benefit of unilateral early cochlear implantation (CI) in congenitally deaf children is well documented and generally accepted. Evidence has also accumulated that a second, contralateral CI can generate added benefit in children with congenital, bilateral profound hearing loss. In Antwerp we strongly support the concept of bilateral CI's in children and in adults. Our experience comprises above 70 bilaterally implanted patients.

Our philosophy of bilateral CI's is based on the belief that binaural hearing is very important and we have been phylogenetically provided with the second ear in order to survive the harsh competition which life is. At the current communication stage of social development optimal binaural hearing is crucial for instantaneous good speech understanding in difficult listening situations. Especially the congenitally deaf children need every bit of acoustic information they can get in order to shape their linguistic and cognitive capabilities according to their innate capacities. There are no doubts anymore about bilateral fitting of the hearing aids, functional operations in unilateral conductive deafness, etc. By the way, we also provide patients with 2 contact lenses, exchange both arthritic hips of knee joints…

On the other hand we still do not have any CI system that would allow for systematic preservation of the residual hearing. This means that when implanting both ears we have to sacrifice the last bits of analogue acoustic hearing the child might still benefit from. There is also an important issue of

possible bilateral vestibular hypofunction and persistent balance problems due to bilateral damage to the vestibular organs. Therefore, while strongly supporting the idea of bilateral CI, we remain very careful and prudent in our indications and always consider less invasive methods, such as bimodal stimulation (combination of CI and contralateral hearing aid). Approximately 30% of our paediatric CI population keeps using the contralateral conventional hearing aid.

This approach resulted in a sequential protocol for bilateral implantations at our Department. It was based on the outcome of a number of studies we performed on a cohort of congenitally deaf children, who received their first and second implants at various ages. The main conclusions of these studies were the following: 1.) There is a critical time window for the development of binaural processing. In bilaterally profoundly deaf children, the second implantation seems to give little or no added benefit for speech discrimination in quiet and in noise after the age of 10-12 years. 2.) In our sequentially implanted children population the second implant reaches a mean level of performance compared to the first implant 18 months after fitting. 3.) The second implant offers cues for binaural hearing based only on ILD (interaural level differences), but seems to offer no cues for binaural hearing based on ITD (interaural time differences).

Therefore, we offer early bilateral CI only to children with bilateral profound hearing loss, without any possible benefit from hearing aids. If the aided thresholds with a hearing aid stay below 60dBHL there is no chance at all for any binaural effects in bimodal stimulation and a child becomes the candidate for early bilateral CI.

On the other hand, in children with contralateral residual hearing, a hearing aid does not have to provide speech understanding by itself in order to give significant improvement of speech discrimination in noise when used in combination with a CI. Additionally, the bimodally fitted patients usually report a more natural sound quality and show better music appreciation. Since the critical time window for development of binaural hearing seems to be longer than in the case of unilateral implantations we most probably have more time to wait. Also the results of our research have shown that in children we do not have to be afraid that the second CI after sequential implantation would remain lagging and continue to give worse results than the first implant. Therefore waiting with implantation of the second ear until the age of 3-4 years we probably do not lose much, and at this age we can quite precisely evaluate the results of bimodal fitting in terms of binaural hearing. The binaural tests are feasible from the age of 3-4 years and comprise evaluation of the audiometric thresholds accompanied by speech tests in noise, localization tests, binaural masking differences, detection of binaural beats, etc. Questionnaires may additionally be used in order to evaluate the subjective benefit of bimodal stimulation.

At the age 3-4 years children also change their social environment, from mostly one-to-one communication within the family to the more demanding listening conditions within the peer group. At this moment their need for optimal hearing increases and then if bimodal stimulation is not sufficient we would offer bilateral CI's.

Availability of fully atraumatic CI electrodes, allowing for preservation of the residual hearing would obviously cause immense changes in the application and counselling of bilateral CI's.

ABI

Modern MRI is able to detect patients with anatomical malformations of the inner ears and the acoustic pathways. In cases of bilateral cochlear nerve aplasia, Michel's malformation of the inner ear and in most patients with major cochlear dysplasias, where no modiolar structures can be recognized, there is no chance for successful rehabilitation with cochlear implants. Such patients are potential candidates for brainstem implantations. In our patient database we have 5 patients with bilateral cochlear nerve aplasia but our own experience with brainstem implantations in paediatric population is very limited (1 patient). At this stage there exists no technique of pre-operative diagnostics that would allow us to predict the status of development of the cochlear nuclei in the brainstem. Limited success of ABI in patients with bilateral cochlear nerve aplasia in other centres (mostly only the signal function of the implant with no open-set speech understanding) raises suspicions about the physiological status and the correct function of the cochlear nuclei in these cases.

Combined ABI-CI

Recently we have implanted the first combined ABI-CI system in a patient with preserved cochlear branch after removal of vestibular schwannoma and progressive deafness in the contralateral ear. Such a hybrid ABI-CI system could provide added value in deaf children with bilaterally hypoplastic cochlear branches and/or bilateral inner ear malformations. With combined stimulation of the cochlea and the brainstem we can in only one surgery warrant that at least one system would work. CI should potentially give better speech discrimination, but if it would not work the ABI part should provide at least a moderate result.

Post-operative fitting

The child's brain is being "primed" by the very first setting of the speech processor and therefore no major deviations from the optimal map are allowed. Importance of correct CI fitting from the very first session cannot be overestimated. Therefore the fittings in children must be performed by experienced audiologists and preferably in the implantation centre itself. Localization of the fitting sessions at the implantation centre gives in our experience the highest chance for appropriate expertise based on sufficient number of cases. In our CI program the diagnostics of the hearing loss, implantation and the post-operative fittings are performed in the implantation centre. Revalidation is given in the vicinity of patient's domiciliation.

In paediatric fittings we very regularly use the information from the objective recording of compound action potentials (NRT, NRI, ART). The objective recordings are repeated at all fitting sessions in children. We also use the correlations between the postoperative C/M-levels and the electrode impedances.

The role of the rehabilitation centres

All hearing impaired children in Belgium are being followed and rehabilitated by dedicated rehabilitation centres. These centres participate at all levels of the diagnostic and treatment program. Already at the level of behavioural audiometric tests, the rehabilitation centres aid the clinical departments in performing these test in a reliable manner and in a child-friendly setting. They also perform or help in the fitting of hearing aids in paediatric population. The revalidation centres offer primary and secondary education for the deaf children as well as educational and familial support for the hard-of-hearing and implanted children, who follow the mainstream education. The hearing impaired children are also provided with life-long possibility for speech therapy. As mentioned above, a valid revalidation plan is prerequisite for reimbursement of CI's in Belgium. Accurate following of the revalidation plan by the patient is an additional criterion for potential bilateral implantation.

Conclusion

To summarize, we can say that a successful program for early intervention in young hearing impaired and deaf children requires a good functioning universal neonatal hearing screening programme, state-of-the-art medical and otological care, state-of-the-art audiology (fitting and evaluation), access to a well structured rehabilitation program and availability of the parent and family support structures. Last but not least, there must be also appropriate budget available for reimbursement of the CI's and the hearing aids.