Hearing loss in young children: challenges in assessment and intervention

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Discussion

In the past decades, important advancements have been made in the care of young children in general and in infants with hearing loss in particular. Newborn hearing screening (NHS) has led to earlier detection of hearing loss: the NHS made it possible to screen infants at just a few weeks old compared to the Compact Amsterdam Paedo-Audiometric Screener (CAPAS) which was administered at 9 to 11 months. This change gave rise to the development of other infant and family health care services that are now offered during and after the follow-up of the diagnosis. The current referral system that is used in the Netherlands when a infant fails a hearing screening is well-organised, and family support services are called in at an early stage. This referral of children at a younger age has created new diagnostic dilemmas and led to questions about the timing of interventions.

It is challenging to estimate a reliable hearing threshold in these very young children based on the available data, and implementing a timely intervention when one is needed. In addition, understanding the aetiology of the hearing loss can help in making decisions for interventions. It is also important for the prognosis and for treatment that may be needed for the child’s hearing or for the child in general.

The diagnostic follow-up for infants who fail a screening is influenced by three factors that affect the hearing threshold measurement: variables and uncertainties about the child’s overall development, variable middle ear pathology and any changes in the sensorineural hearing threshold. A full evaluation of a child’s hearing is carried out using various complementary audiological tests. There are no objective frequency- and ear-specific measurements that can quantify the severity of sensorineural hearing loss in middle ear pathology in children at this very young age.

In order to meet the above-mentioned changing and complex health care needs that arose after the implementation of the NHS, we set up a multidisciplinary team with an otolaryngologist, a paediatric audiologist and a neonatologist, with assistance from departments of clinical genetics, radiology and paediatric neurology. The otolaryngologist and paediatric audiologist play a central role in follow-up testing. Effective and well-organised collaboration between these two professionals, each with their own expertise and responsibilities, is essential when deciding on and evaluating hearing interventions. Multidisciplinary care implies attention for the ear and hearing evaluations, but also for the child’s overall development in a broader perspective. Similar multidisciplinary care has emerged in various other centres, some with a slightly different focus or with protocols for additional medical assessments.

In striving to gain more certainty about all the different variables related to children and hearing loss, there is the risk that we will expand the number of assessments even further, sometimes leading to a false sense of security and delays in the decision-making process. Given the complexity of this particular service for child hearing screening and the complexity of diagnostics and rehabilitation, it seems desirable to
centralise these services in centres staffed with teams of experts. On the other hand, it is essential to provide children with care close to home, which could be facilitated if the teams took on a coordinating role.

Another important issue is awareness of delayed auditory maturation. This phenomenon may be more universal than we now think, particularly in children born prematurely. In our small cohort of 14 children, the initial hearing threshold improved for 9 of them. Given advancements in obstetric and neonatal care, the population of children born extremely prematurely will continue to grow and we will be consulted more often for hearing diagnostics in this population. Awareness of delayed auditory maturation is particularly important by the decision for cochlear implantation, as the trend is to place implants in children at an increasingly younger age. We believe that it is preferable to use the child's age corrected for gestational age rather than the calendar age, especially for children born prematurely. This dissertation shows that monitoring a child up until about an age of 40 weeks, corrected for gestational age, seems sufficient to detect or rule out delayed maturation.

The child's overall development should be estimated in addition to his or her auditory maturation. However, a delay in overall, broader development is often not visible until later in a child's development. Especially in the beginning, it is not always clear whether a hearing disability is the cause of a delay or whether the lower score is caused by a delay in the child's overall development. Hearing loss also causes an additional disability in children who have an overall developmental delay.

The registration of otoacoustic emissions plays a large role in screening young children for hearing impairments. A positive result in a hearing screening would indicate sufficient hearing and would not constitute grounds for a referral for further testing. Since the middle ear has a dual impact on the detection of otoacoustic emissions (incoming stimulus and reverse propagation of emissions), middle ear pathology plays a large role in the hearing screening's outcome. Because middle ear pathology occurs frequently in children, it is expected that emissions are not detected due to the middle ear dysfunction. We have described a two-step scenario that would decrease the number of false-negative referrals when transiently evoked otoacoustic emission (TEOAE) are measured after pressure compensation.

Compensated TEOAE measurements could possibly also be used in hearing tests as a selection criterion for middle ear ventilation tubes in children. Conditioned play audiometry and the block test are not usually feasible until after the age of about 2.5 to 3 years. If a TEOAE can be recorded, with or without pressure compensation, we can prevent referrals to an audiological centre for paedo-audiometry. In this context, compensated TEOAE measurements could help improve the general ENT services.

In our last chapter, we examined the effect of middle ear pressure on TEOAEs based on a mathematical middle ear model. This showed that the mass factor also plays a role in addition to the compliance effect of the “tympanic-membrane and malleus-incus complex”. A mild form of middle ear pathology, such as a limited amount of fluid in the
middle ear, could explain a mass factor and allow for reduced mobility of the tympanic membrane. In line with this theoretical finding are the experiences that type A, and C tympanograms can detect effusion in the middle ear, which could correspond with the mass factor.

Future

This dissertation also clearly shows that the middle ear is a variable and difficult-to-quantify component in the diagnosis of hearing loss in children. The accuracy of the diagnosis of middle ear dysfunction and quantification of the conductive component of hearing loss has been a key focus point for decades. In the middle of the 20th century, tympanometry developed as a practical method for measurement using a 226 Hz probe tone. We have recently learned that a higher probe tone is more sensitive to middle ear changes in very young children. A 1000 Hz probe tone seems the most accurate in neonates; this shifts to a 226 Hz probe tone around the age of 9 months. Current tympanometers can measure both frequencies, so the best frequency can be chosen when testing. Although the prevalence of middle ear pathology is lower in neonates, it would be interesting to learn whether pressure compensated otoacoustic emission (OAE) measurements could also influence tympanometry in these very young children at either one or both probe tones.

The possibility of conductive hearing loss is a difficult and sometimes delaying factor in diagnosing children with severe hearing impairment and deafness, and in making a decision about cochlear implants. In the work-up, imaging under anaesthesia is often combined with auditory brainstem response (ABR) measurement, but the ABR is inadequate in cases of conductive hearing loss. Placing “diagnostic” middle ear ventilation tubes in this same setting is not feasible at an organisational level in our hospital, which often means a child will need to be placed under anaesthesia a second time. To quantify the conductive hearing loss in young children could add clinical value for this population.

In the search for the hearing levels by the current diagnostical audiological evaluation, we do not include the auditory cortex. It is still insufficiently clear which roles cortical auditory evoked potential and fMRI can play in the maturation of the auditory pathway and the decision regarding cochlear implants. Exploration of the auditory cortex will certainly raise new questions, but can also offer new possibilities for diagnosing children with a hearing impairment or deafness.