Dutch Cochlear Implant Group (CI-ON)
Consensus Protocol on Postmeningitis Hearing Evaluation and Treatment

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Objective: One of the most devastating sequelae of bacterial meningitis is profound hearing loss or even deafness. Although cochlear implantation is able to restore (some) hearing abilities, obliteration due to fibrosis and especially calcification of the cochlea in the postmeningitis period is limiting the success rate of an implantation. A national consensus assembled in a postmeningitis follow-up protocol has to increase awareness and thus the chances of an early detection and possible intervention when profound hearing loss occurs.

Setting: All cochlear implant (CI) centers of The Netherlands located in the 8 academic otorhinolaryngology and audiology departments of The Netherlands, gathered in the Dutch Cochlear Implant Group (CI-ON, Cochlear Implant Overleg Nederland).

Intervention: A protocol proposed by 3 centers was sent to all other CI centers in The Netherlands to review and agree on.

Main Outcome Measures: The CI centers agreed on the need for, and use of, the proposed protocol. Keystones of the protocol are treatment with dexamethasone before start of antibiotics, early magnetic resonance imaging and repeated audiological follow-up, and urgent referral to a CI center in all cases with greater than 30 dB SNHL.

Conclusion: The Cochlear Implant Centers in The Netherlands (CI-ON) have agreed on a protocolized follow-up after bacterial meningitis to increase the chances of an early detection and possible intervention should profound hearing loss occur.

Key Words: Algorithm—Audiology—Bacterial meningitis—Cochlear implant—Diagnostic protocol—Hearing loss—Magnetic resonance imaging—Netherlands.

is probably because of the interpretation of the communication problems in the recovery phase after an intensive care period. Postmeningitis concentration and word finding problems could be a plausible explanation, leaving the real cause, the hearing loss, undetected.

Cochlear implantation is nowadays the method of rehabilitation in profound and complete hearing loss. In postmeningitic patients with a cochlear implant, the results are usually fairly good. However, it is of extreme importance to have a patent cochlear lumen to insert the complete cochlear electrode array to obtain the best results (8–11). Meningitis can cause an immune reaction within the cochlea leading to fibrosis and finally to calcification of the cochlear turns (labyrinthitis ossificans), which makes implantation extremely difficult or even impossible. A good opportunity for hearing restoration could disappear within weeks; because of this obliterating process, an active postmeningitis hearing evaluation is not warranted (12,13).

These 2 main factors, a delay in speech and language development or an incomplete or impossible cochlear implant electrode insertion although that could have been avoided, have been the reason to create a national protocol on hearing evaluation and follow-up after bacterial meningitis. The Dutch Cochlear Implant Group (CI-ON, Cochlear Implant Overleg Nederland) presents “the Dutch consensus protocol on postmeningitis hearing evaluation and treatment” in an attempt to solve this problem and create more awareness for this urgency.

**METHOD**

In The Netherlands, the cochlear implant centers are located in the 8 academic otorhinolaryngology and audiology departments.

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**FIG. 1.** Dutch Cochlear Implant Group consensus protocol on postmeningitis hearing evaluation and treatment. Urgent indicates within 2 weeks. *Children need ABR.*

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Hearing assessment should be done as soon as possible (preferably before discharge from the hospital) in all survivors of meningitis, children as well as adults, irrespective of the existence of hearing loss before the first audiological tests. A protocol follow-up scheme of formal hearing tests should be followed in the following months because hearing loss can be progressive up to 6 to 12 months after the meningitis. Bacterial meningitis is more frequently found in children younger than 2 years. They are more prone to have meningitis because of their immature immune system. Also, this is the group who is right in the years in which hearing is critical for language development (6). Especially, in this group, hearing evaluation using otoacoustic emissions (OAE) (18) and auditory brainstem response (ABR) is necessary to obtain objective measures of hearing abilities after meningitis.

With this consensus protocol, we aimed for an easy accessible as well as a usable guidance for all involved professionals in every hospital. Although Streptococcus pneumoniae has a dominant role as a precipitant of SNHL and is the most important species causing obliteration of the cochlea (30%) (2,4,5), we think that the causative organism does not exclude the obligation to follow the same protocol because all bacterial species can cause a similar devastating effect. The protocol is therefore generalized regardless of the bacterial organism causing the meningitis to create a clear entry and standard care in each meningitis case. Moreover, because of the vaccination programs for Haemophilus influenzae (19) and S. pneumoniae (20), the number of meningitis cases is decreasing, making the routine of postmeningitic hearing follow-up more vulnerable.

One of the most important issues is to make sure that the clinician who is initially dealing with the patient with meningitis (mostly pediatricians, intensivists, or neurologists) is aware of the risk of hearing loss and the possible risk of obliteration. It is the responsibility of this clinician to refer these patients to an audiological center as soon as possible, preferably before discharge.

**Dexamethasone**

Administration of dexamethasone before antibiotic treatment in meningitis in adults (21,22) and children (23–25) will reduce the neurologic complications and risk of hearing loss. We therefore advocate dexamethasone in children 0.6 mg/kg per day in 4 doses for 4 days starting before the first antibiotic infusion and in adults at 10 mg in 4 doses for 4 days. As the lysis of bacteria in the cochlea is supposedly one of the triggers leading to the
intracochlear immune response, it seems important to start with the dexamethasone therapy at least 10 to 30 minutes before the first antibiotic administration (22,24).

First Hearing Evaluation

In case of a passed OAE, the follow-up should be frequently during the first months. The experience within the Dutch cochlear implant centers show that hearing deterioration is possible and most likely soon after the meningitis. Follow-up should be done in an audiological center, but if not feasible, at least OAE and a quick referral should be done in case of progression. After 1 year of follow-up, we have not observed any new cases of postmeningitis hearing deterioration and cochlear fibrosis, although we have seen a single case of severely fluctuating hearing loss (without ossification) leading to cochlear implant more than 10 years after meningitis. Some publications claim that the deterioration after 11 years still is due to the meningitis (26); nevertheless, our opinion is that these cases are extremely rare and the chances of cochlear fibrosis due to meningitis in these late cases are negligible.

In case of a failed OAE, the referral should be immediate, and objective measurements should be done within 2 weeks. In children, ABR is mandatory; in adults, ABR is merely in severe cases.

Still, an uncertain question is how fast the obliteration of the cochlea can be manifest. But according to animal experiments, case reports, and the experience in our group, it seems a matter of weeks (12,13,27). It is encouraged that when the OAE at discharge is failed within 3 weeks, a decision whether cochlear implantation is needed can be made.

To be able to make such a decision a cascade of actions should be completed. First of all a quick referral to a cochlear implant center is essential. In particular, a cochlear implant center because, in these cases, counseling, diagnostics, and expertise in a punctual and multidisciplinary approach is crucial. Also, to avoid patient-, or doctors delay, the correct counseling and knowledge of a specialized team is necessary, because the hearing loss seems in the eyes of the patient, the parents, or even the physicians around them not always to be an emergency case.

The cochlear implant center will evaluate all patients with a 30-dB SNHL or higher. In our experience, this is a safe and valuable cutoff point to decide between non-affected ears and affected ears. Thus, children or adults with a 30-dB hearing loss or higher should be considered “at risk” to have or to develop an irreversible SNHL, and children are prone to develop a problem in speech and language development. Although 30-dB hearing loss alone is not enough for cochlear implantation, signs of fibrosis on magnetic resonance imaging (MRI) and a 30- to 70-dB hearing loss should be very closely monitored via frequent hearing tests and MRI. In case of progression of hearing loss or fibrosis on the MRI scan, cochlear implantation is inevitable. Greater than 70-dB hearing loss and signs of labyrinthitis or fibrosis on the MRI (see next paragraph) are reasons to perform an urgent cochlear implantation. Below 30-dB, a follow-up policy is adequate; in the at-risk group, imaging and repeated hearing assessment are the keystones in the diagnostic workup.

Imaging

Magnetic resonance imaging is the superior type of imaging in postmeningitic cases because it shows abnormalities in the early stages of a labyrinthitis ossificans (28,29), which are well correlating with surgical findings (30). Computed tomography (CT) shows calcification commonly in the later stages (Fig. 2), but especially, an "open cochlea" has a poor correlation with the surgical findings intraoperatively (31). Still, CT is mandatory as anatomical roadmap in surgical cases. Inflammation within the cochlea is shown as an enhancement within the cochleae on gadolinium-enhanced T1-weighted MRI scan, as shown in Figure 2. This is a display of a pathologically increased perfusion within the cochlea and is associated with (the start of) fibrosis in the lumen of the cochlea. As soon as there is loss of fluid on a T2-weighted MRI scan, shown as a decrease in intensity (Fig. 2), there is a stage of fibrosis, frequently preceding ossification (28,29). In these cases, it is valuable to differentiate between the scala vestibuli and the scala tympani because the former is the optimal scala to insert but also the scala most frequently obliterated. In a later stage when fibrosis has filled the cochlea the T2-weighted MRI fluid, image of the cochlea could be gone, and in addition, a hypointense or an isointense figure of the cochlea in a nonenhanced T1-weighted MRI scan could be noticed. In this stage, it is almost impossible to completely insert the electrode array in the cochlea. Imaging is therefore an important step in the postmeningitic patient with hearing loss to optimize the rehabilitation and to arrange a good timing of a possible cochlear implantation. Achieving a complete cochlear electrode insertion is very important to obtain the best results in this type of hearing rehabilitation (8–11). Also, the time between the meningitis and the implantation is important because this will benefit the performance of the patient with a cochlear implant (32). Hence, MRI is the best reliable tool in the (preoperative) diagnosis and planning. It is advisable to plan MRI scan and computed tomographic scan in one single anesthesia in children who are likely to have surgery to minimize the burden of the narcosis and to fasten the diagnostic workup.

Hearing evaluation should be repeated to reaffirm the hearing status, in all cases and especially in the severe to profound SNHL cases. In these cases, one has to reassure that the whole team and the parents (or patient) are convinced of the severe or profound hearing loss and the need for a cochlear implant with all the pros and cons. The parents (or patient) have just overcome the shock of the life-threatening meningitis, and just within weeks, they are confronted with a decision about a life-changing operation.

Multiple scenarios are possible in case of mild to severe hearing loss, unilateral hearing loss, high-tone SNHL, mild to no enhancement, unilateral enhancement, and so on. The presented protocol will and cannot be all-embracing in this matter, but our CI-ON members agree that the best hearing
Preservation and rehabilitation option should be provided without loss of prospect. This requires a frequent and active diagnostic follow-up with repeated MRI and hearing evaluation or, in case of progression of hearing loss or cochlear patency, an immediate cochlear implant. Unilateral implantation with a close follow-up on the contralateral ear (with mild to moderate SNHL) has been done to ensure (electric) hearing rehabilitation without compromising the acoustic hearing abilities in the other ear.

The Dutch cochlear implant centers have agreed on a bilateral cochlear implantation in postmeningitic patients with (bilateral) severe to profound hearing loss, in accordance with the international consensus (33). In less severe cases, bimodal stimulation is advocated.

How to avoid and treat intraoperative difficulties regarding an obliterated cochlea (34,35), the use of different types of electrodes in postmeningitic cases (36–38), or the conversion and results of an auditory brainstem implant

**FIG. 2.** Changes in magnetic resonance and computed tomographic images in postmeningitis patients over time. Shown in the first column (A, D, G, J) are MR T1 images with contrast, in the second column (B, E, H, K) are MR T2 images (CISS sequence), and in the third column (C, F, I, L) are computed tomographic images, all in time sequence from top to bottom. Images are put in order to reconstruct the time sequence; not all images are of the same patient. Top row (A, B, C): Normal scans: normal magnetic resonance and computed tomographic images of the cochlea and labyrinth. Second row (D, E, F): “Enhancement phase”: increased uptake of contrast in the cochlea on T1 image (D), with no loss of fluid in the cochlea (E). Third row (G, H, I): “Loss of fluid phase”: still increased contrast uptake on T1 (G) but also decrease of fluid in the cochlea on T2 image (H). The scala tympani has clear loss of fluid, which correlates with an increase of fibrous tissue within this scala. Fourth row (J, K, L): “Ossification” and final stage: no active enhancement in the cochlea anymore on T1 image (J), fairly decreased amount of fluid in the cochlea on T2 image (K), and a calcification visible on the computed tomographic image (L).
in a severely ossified cochlea (39,40) are beyond the scope of this protocol but are well-adapted knowledge in The Netherlands.

CONCLUSION

The hearing of postmeningitic patients should be evaluated at discharge and followed up frequently. To have these patients immediately referred and to avoid the risk of incomplete cochlear implantation in cases of (bilateral) profound hearing loss, the Dutch Cochlear Implant Group has agreed on a flowchart protocol. This protocol is meant to be a tool for all medical and audiological specialists treating (post)meningitic patients.

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REFERENCES