European consensus statements on paediatric cochlear implantation

"Cochlear implant is one of the most successful neural prostheses developed to date"

World Report on Hearing, WHO 2021

Cochlear implants are electronic devices, especially useful when a conventional hearing aid has little or no benefit or cannot be used. Typically, these devices bypass the middle- and inner-ear structures to stimulate the auditory nerve directly and can give a deaf person a useful representation of sounds in the environment, making it possible to understand speech.

"Their use in children with severe degree of hearing loss has brought substantial benefits to those implanted, and when accompanied by proper rehabilitation they lead to significant improvement in audiological status, overall functioning and speech perception skills. Children with cochlear implants have greater likelihood of acquiring oral language, integrating into regular schools and being able to experience sounds along with better speech skills"

"Cochlear implants can also have a beneficial impact on learning and educational outcomes as well as the overall quality of life, though many factors other than implantation influence these results."

WHO World report on Hearing 2021 www.who.int/publications/i/item/world-report-on-hearing (page 98)

The main goal of European consensus statements is to raise awareness of cochlear implantation for children and to provide the best possible cochlear implant service for children with severe to profound hearing loss who are eligible for a cochlear implant. The document may be used by clinics to inform local authorities and healthcare providers about standard cochlear implant care which should be provided in European countries. The statements below are based on existing national guidelines on cochlear implantation in children amongst others of the United Kingdom, Belgium, Italy, France and Germany and have been agreed upon by a group of European experts and organizations representing patients and their families and cochlear implant advocacy groups.

Universal newborn hearing screening

Early detection and intervention of hearing loss greatly reduce the risk of delays in communication and spoken language development, educational challenges, and a lower quality of life. Therefore, governments should provide comprehensive newborn hearing screening programs and have an effective and prompt referral system to the centers of expertise. Data management and adequate prevalence monitoring should be warranted by the governments. A tracking system is necessary. Together with early diagnostics, a plan for rehabilitation should be made and early testing with hearing aids should be provided, if appropriate. If insufficient (or if etiology points towards CI) cochlear implantation should be indicated timely.

The Cochlear Implant program
The cochlear implant program ensures:

- the correct selection of candidate,
- timely, appropriate and safe surgery and programming,
- adequate and sufficient rehabilitation,
- close collaboration between specialists that make up the program,
- appropriate follow-up of the implanted patient,
- the maintenance of the implanted system,
- inclusion of patients and parents/caretakers
- shared decision making

The Cochlear Implant team
Cochlear implantation should be considered for children only after an assessment by a multidisciplinary team. This specifically dedicated team should master knowledge and be experienced in the field of otorhinolaryngology, audiology, radiology and specialized cochlear implant speech and language therapy necessary to provide a high-quality cochlear implant service. The team should be of sufficient size to provide sustainable care. The team should be within an institution with pediatricians and have access to geneticists, psychologists and educational specialists. The family and the local professionals working with the child should be considered part of the team, with liaison between them.

Indications for cochlear implantation
Cochlear implantation in children is indicated in cases of bilateral severe to profound sensorineural hearing loss when conventional hearing aids do not or are not expected to provide enough hearing for the development of speech recognition and appropriate spoken language development. If appropriate, as part of the assessment children should have had a valid trial of acoustic hearing aids for at least 3 months.

Assessment for cochlear implantation
Assessment for cochlear implantation involves the testing of hearing status with objective and/or psych-acoustic audiometry (depending on developmental stage of the child). Assessment for cochlear implantation in children should include the assessment of;

- hearing status
- communication skills
- general development
- medical condition
- high quality imaging
- vestibular investigation
- genetic tests
- family support
- educational support
- psycho-social development

Parents/carers should be able to meet other families of children with CI.

Bilateral cochlear implantation
All children with bilateral severe to profound hearing loss should be offered bilateral cochlear implants, preferably simultaneously. In cases where hearing loss is progressive and/or asymmetrical, a bimodal fitting (HA and CI) may be advised. Sequential bilateral cochlear implantation may be considered if the expected benefit of a second cochlear implant would be more than that of the conventional hearing aid.

Discussions of expectations
Cochlear implantation has been proven to be a safe, effective and cost-effective intervention for bilateral severe to profound hearing loss in children. Counseling parents about the CI journey, the contribution they will have to make, and about the likely outcome of the intervention is vital. Setting realistic expectations helps create the best possible conditions for the success of cochlear implantation. Parents of children with additional disabilities or specific medical conditions like a hypoplastic cochlear nerve or a cochlear malformation should be informed that CI results may be limited. At young age additional needs may still be unknown, so it is important to monitor the development of the children well during the early years after implantation. In case a genetic diagnosis has been made the results should be fully discussed which may provide helpful information about the child's likely development.

Surgery
In children with congenital bilateral severe to profound hearing loss, performing cochlear implantation as early as possible provides the best results in terms of benefit. It is recommended that CI surgery should be performed by experienced surgeons and so far as possible save external, middle and inner ear structures (with or without preoperative residual hearing). The possible risks of surgery should be explained to parents/carers and they should be provided with information to take away about how to look after their child after surgery.

Activation and rehabilitation
The external parts of the system (processor etc) should be fitted and activated as soon as possible by experienced audiologists, with the parents/carers present. Fitting the processor in children should be performed in specialized rehabilitation centers by experienced fitting specialists, either at the implantation center or in close collaboration with it. Parents/carers should be involved in the choice of the brand of the cochlear implant and the fitting. The management of the equipment should be explained, including how to connect assistive listening devices or on how to stream audio sources to the sound processor. The team's knowledge and experience in conventional hearing aid fitting is needed for optimal bimodal fitting. Rehabilitation should consider combining acoustic and electrical stimulation where there is low-frequency residual hearing. Trained rehabilitation specialists should be involved in the rehabilitation process to promote listening and complex spoken language development and to monitor the child's progress. They should liaise with the child's local educational or early intervention teams.

Ongoing support
Cochlear implantation is a life-long intervention. Cochlear implant recipients have the right to continuous support and access to functional sound processors as well as access to spare

parts, repairs and regular upgrades. It is recommended to assess the resources, difficulties and needs of the family environment of children with cochlear implants and provide aftercare accordingly. The rehabilitation center should provide advice and guidance for the child's local educational staff and those caring for them. Following cochlear implantation, children need the opportunity to hear rich spoken language, with fully functional technology at home, day care and school. Their development and wellbeing should be regularly monitored by the rehabilitation center, direct or indirect and possibly remote, to ensure that progress continues, and to monitor any issues with equipment, the environment or the child's development.

European consensus statements on paediatric cochlear implantation by (in alphabetic order);

Sue Archbold, Antje Aschendorff, Manohar Bance, Caglar Batman, Eliana Cristofari, Helen Cullington, Aarno Dietz, Tobias Fischer, Jozsef Geza Kiss, Thomas Lenarz, Natalie Loundon, Manuel Manrique, Emmanuel Mylanus, Thomas Nikolopoulos, Luminita Radulescu, Leo De Raeve, Angel Ramos, Pjotr Skarzynski, Per Caye-Thomasen, Franco Trabalzini, Line Troost, Robert Trotic, Eric Truy, Nicolas Verhaert, Jantien Vroegop, Astrid van Wieringen, Andrzej Zarowski