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Professor Wagenfeld has been Chairman of the largely otology- and audiology-orientated Department of Otorhinolaryngology at Stellenbosch University and Tygerberg Hospital from 1981 to 1988, during which time the cochlear implant programme was established. Subsequent to leaving the full-time academic post he has continued as Director of the Cochlear Implant Programme while in private practice in Somerset West. His current fields of interest are otology and audiology.

Paediatric cochlear implantation

Congenital hearing loss, by its impact on spoken language acquisition, has far-reaching implications for intellectual and social development, literacy, educational attainment, employment and quality of life.¹

For centuries profoundly deaf children were largely isolated from communication and education. Today cochlear implant (CI) technology has changed the lives of deaf children by providing them with the opportunity of hearing (with CIs) and thus enabling them to develop speech and language.

The first paediatric CI was done at the House Ear Institute in Los Angeles in 1980. Initial concerns about device infection from otitis media and electrode problems caused by head growth were soon proven to be essentially irrelevant.² In 1988 the first cochlear implantation was done in South Africa at the University of Stellenbosch-Tygerberg Hospital Cochlear Implant Unit. Since then 260 children have received CIs in South Africa.

Children who undergo implantation early in life, followed by a period of appropriate rehabilitation, can achieve speech and language skills exceeding those seen in profoundly deaf children with hearing aids. However, there is considerable variability in performance between individual children.² Both congenitally and postlingually deaf children can develop hearing and oral language using CIs.

TECHNOLOGY

Current CI systems consist of internal and external components (Figs 1 and 2).

The internal portion consists of a multi-channel electrode array that is implanted in the cochlea and is attached to a receiver/stimulator, an antenna and a magnet

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attached to the external headset.

Multichannel implants take advantage of the tonotopic organisation of the cochlea.

The external component consists of a microphone, microprocessor-based speech processor, connecting cables and transmitting coil with external magnet. External processors are either body worn or 'ear level'.

The microphone converts the incoming sound into electrical signals which are then sent to the speech processor where they are analysed and digitalised. The resulting coded signals are sent to the transmitting coil from where they are sent via radio frequency to the internal components. The coded signals contain information as to which electrodes to stimulate,



Fig. 1. Diagrammatic representation of internal and external components. 1: Nucleus 24 Contour implant; 2: receiver/stimulator; 3: Contour electrode array; 4: ESPrIt 3 G speech processor.

and the intensity level required to generate the appropriate sound sensations. The electrodes stimulate the nerve fibres via a controlled electrical current which is recognised by the brain as sound. Electrical stimulation of the auditory system is effective, because almost all sensorineural hearing loss is caused by hair cell dysfunction, while the auditory nerve itself remains responsive to stimulation and can conduct impulses carrying auditory information to the brain.²



Fig. 2. Diagrammatic representation of the Nucleus 3 cochlear implant system.

PAEDIATRIC CANDIDACY GUIDELINES

The clinical population considered for cochlear implantation has evolved to include children younger than 12 months, children with significant cochlear abnormalities, and children with multiple handicaps. Given these expansions, it is more important than ever that each case be considered

individually by an experienced CI team. The team should consist of otorhinolaryngologists, audiologists, speech therapists, educational specialists, physio- and occupational therapists, psychologists and social workers.

Degree of hearing loss and aided performance

Quantification of performance takes into consideration the age of the child, the degree of the hearing loss and the type and the outcome of intervention, as shown in Table I.

The aided audiogram can be used as a guideline to refer children for CI evaluation. As shown in Fig. 3, a referral can be made when the aided thresholds fall within the blue shaded area.

A 6-month trial using well-fitted hearing aids is necessary to determine the expected development of auditory skills. This trial can be waived in the case of acquired deafness due to meningitis when there are indications that imminent ossification of the cochlea will make implantation difficult or impossible.

Age at implantation

For congenitally deaf children, age at implantation is a prognostic variable. The earliest implantation leads to the most normal developmental patterns of auditory and communicative skill for congenitally and prelingually deaf children. Sensitive periods of neural development exist during the first 3 years of life and are critical for the establishment of auditory mechanisms, including speech understanding and language.^{2,3} Early auditory deprivation can lead to reallocation of perceptual resources in the auditory cortex.¹ Conversely, electrical stimulation with ongoing use of the implant can result in neural survival and developmental changes in the central auditory system.⁴

In general, congenitally deaf children who undergo implantation as adolescents do not demonstrate the open-set speech perception abilities seen in younger children who use CIs. However, children with progressive hearing loss who undergo implantation at a stage when the hearing loss has become profound often perform well on speech perception tasks. Early and continuous use of residual hearing by such children, plus use of spoken language, provides them with an advantage in processing auditory information from a CI.⁵

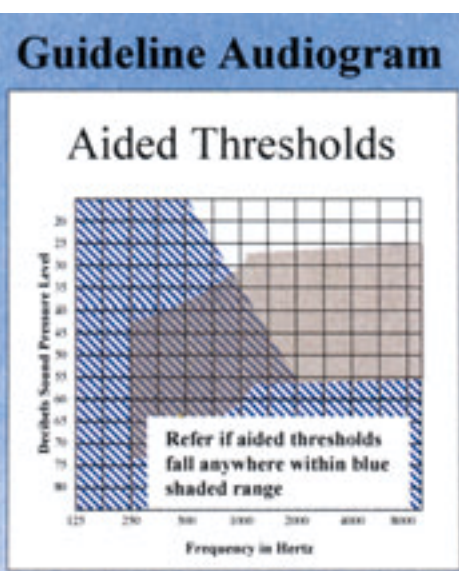


Fig. 3. Referral guideline audiogram.

Ossification

Ossification of the cochlea is likely to develop in children deafened by meningitis, and can make implantation technically more difficult and increase the likelihood of only partial insertion of the electrode array. Approximately 1 in 10 - 20 paediatric CI candidates have some cochlear ossification, but in nearly 90% of these the ossification is limited to a short segment adjacent to the round window membrane.² This can be removed via the cochleostomy. In these cases speech perception results are similar to those with patent cochleas. Evidence of ossification can be seen as early as 2 months following

Table 1. Paediatric candidacy requirements

Age (years)	Hearing level (dB)	Speech perception score	Other
1-2	Profound ≥ 90 dB	—	Lack of progress in development of auditory skills with hearing aids
2-5	Severe to profound ≥ 70 dB	< 30% (open-set word discrimination)	Lack of progress in development of auditory skills with hearing aids
5-18	Severe to profound ≥ 70 dB	< 30% (open-set word discrimination)	Lack of progress in development of auditory skills with hearing aids

Open-set word discrimination = word discrimination through hearing alone, without lip-reading.

meningitis. There may therefore be a time window following meningitis during which complete electrode insertion can be performed, after which it could become impossible.³ Early referral and evaluation for implantation is vital in children with postmeningitic deafness.

Malformations of the cochleae

Labyrinthine malformations are no longer considered contraindications to cochlear implantation. Mondini dysplasia and its variants and large vestibular aqueducts are associated with excellent results from implantation. Technical difficulties associated with cochlear malformations are congenital anomalies of the facial nerve and management of the cerebrospinal fluid gusher.^{2,3} Cochlear aplasia and an absence of the cochlear nerve, however, are contraindications to cochlear implantation.

Other handicapping conditions

As many as one-third of children with hearing loss may have other handicapping conditions or developmental delays.⁶ Some children with cognitive and/or motor developmental delays, learning difficulties, cerebral palsy, sensory integration problems, blindness and autism can be considered as CI candidates. The literature shows that CIs benefit children with hearing impairment and additional handicaps, but that progress is like-

ly to be slower and more inconsistent than in their more able-bodied peers.^{3,6}

Appropriate expectations must be set at home and at school for each child's progress after implantation. Infants and toddlers with auditory neuropathy present a challenge to implant teams because their physiological and behavioural auditory function may be changing at the time of deciding about cochlear implantation. The vast majority of CI children with auditory neuropathy responded favourably to electrical stimulation.⁷

Criteria for cochlear implantation in developing countries

No highly technical device should be implanted in a child until a lifetime commitment can be given, not only to the maintenance of the device, but also to the sociological and educational implications. This becomes even more compelling in the South African society where resources are severely limited and competition for funds is intense. Children being considered for cochlear implantation must have adequate family support, and parents must be gainfully employed. Accessible, compulsory, and appropriate educational and audiological facilities must be available.⁸

Family expectations

It is essential that families understand that the surgery is just the

beginning of a long-term educational and supplementary therapeutic process that requires a commitment from the whole family. This is necessary for optimal function of the implant.

CANDIDATE EVALUATION

The primary aim of the preoperative evaluation is to determine whether the patient is medically, audiological and psychologically suitable for cochlear implantation. Assessment of parental expectations, support and commitment to therapy and the availability of appropriate educational facilities are crucial factors in paediatric candidate selection. It is important that the team establish a long-term relationship with the family to support the child's auditory and spoken language development.

Medical and surgical evaluation

A complete medical and ENT evaluation is performed, to attempt to identify the aetiology of the hearing loss, and to determine whether there are other medical factors which may influence the patient's suitability for surgery and rehabilitation. Computerised tomography (CT) scans of the temporal bone allow for assessment of the patency of the cochlea, mastoid aeration, facial nerve position, and middle ear status. Magnetic resonance imaging (MRI) is particularly useful for

assessing the patency of the cochlea. It may show evidence of fibrosis preceding neo-ossification of the cochlea following meningitis. It also demonstrates the neural structures, such as the integrity of the cochlear nerve and its central connections. Imaging aids in assessing the feasibility of the procedure, and is useful in selecting the appropriate ear for surgery.

Audiological evaluation

The primary aim of the preoperative audiological evaluation is to determine the type and severity of the hearing loss. In very young children objective measures such as auditory evoked potentials and otoacoustic emissions can help to determine auditory thresholds. Appropriate hearing aids should already have been fitted and the length of time the candidate has used the amplification should be considered. Candidates without previous hearing aid experience should have a hearing aid trial of 3 - 6 months.

Ongoing diagnostic therapy is essential in the assessment of functional benefit from amplification in very young children. The Infant-Toddler Meaningful Integration Scale (IT-MAIS) can be used to obtain this information.⁹

In general, the aetiology of deafness does not appear to impact on speech perception performance in children.

Speech and language evaluation

Assessment of speech, language and cognitive abilities before and after implantation is required to monitor progress, and to determine whether additional developmental disorders could influence

the child's auditory development. For older children considering cochlear implantation, language development should be assessed along with the current use of residual hearing for speech perception. A detailed history of the progression of hearing loss is also needed.

SURGERY

Surgery to insert a CI usually requires 1½ - 3 hours and an overnight stay in hospital. The surgery varies according to the design of the particular device being implanted. The cochlea is full size at birth and there is no anatomical difficulty with electrode insertion in very young children.² CIs are designed to allow implantation in 6-month-old infants, and the surgical techniques used in infants and children do not differ in principle from those used in adults. There are minor adaptations to accommodate age-related aspects of head growth, thickness of skull, and also the tendency to otitis media in very young children. The incidence of otitis media does not increase with implantation, and its treatment with oral antibiotics, and occasionally ventilation tubes, is safe and effective.

DEVICE PROGRAMMING

Programming CIs in young hearing-impaired children with limited language abilities and limited sound experience continues to be a major challenge for paediatric audiologists. Approximately 3 - 4 weeks after surgery the activation of the implant begins. The minimum and maximum electrical levels for hearing, the softest (T-levels) and comfortable listening levels (C-levels), are determined for each electrode by means of conventional conditioning techniques depending on the age of the child.

Where children cannot indicate the T- and C- levels, an objective pro-

cedure such as neural response telemetry (NRT) can be helpful. NRT is an objective procedure of recording the electrically evoked compound action potential of the peripheral auditory nerves.¹⁰ Long-term follow-up programming is required to ensure the most effective stimulation of the electrodes. New technology for trouble-shooting allows parents and teachers to assess the functioning of the external device.

OUTCOMES

It has been well established that CIs are reliable and effective devices for significantly improving access to sound for children with severe-to-profound hearing loss.² There is however great variability in outcomes, which is thought to be primarily related to patient factors. In general, the aetiology of deafness does not appear to impact on speech perception performance in children. Variables significantly affecting outcomes are: age at onset of deafness, age at implantation, amount of residual hearing before implantation, duration of implant use, and educational setting.^{2,11,12}

Age at implantation

Studies have shown that young congenitally deaf children who undergo cochlear implantation have the ability to learn language at rates comparable with those of their hearing peers.² In a retrospective study, the rate of language development was compared between groups of children on the basis of the age at which they received their CIs. Children implanted before 30 months of age had spoken language skills within 12 months of their chronological age in comparison with children implanted after that age. Children implanted at 18 months and younger had spoken language skills within 6 months of their chronological age.¹² In another study, the

MAIN TOPIC

rate of speech perception development for congenitally deaf children implanted before 24 months of age resulted in earlier open-set speech discrimination compared with congenitally deaf children implanted between 2 and 3 years of age.⁸

Duration of implant use

One of the most consistent findings in the literature is that the speech perception abilities of children with CIs improve with increased device experience. A meta-analysis of published paediatric CI performance data indicated that:

- more than half of the implanted subjects obtained open-set speech understanding within 2 years of implantation
- earlier implantation tended to be associated with a greater trajectory of gains in speech perception
- subjects did not demonstrate a plateau in performance over time
- differences between children with congenital and acquired deafness diminished over time.¹³

Residual hearing

Several studies have shown that the amount of unaided residual hearing pre-implant was an independent predictor of postoperative speech perception performance.⁵ Increased auditory experience before implantation facilitated the development of speech perception post-implant. Children should be considered for implantation if their speech perception performance is less than that obtained by the average paediatric CI recipient.¹ Children with unilateral CIs are encouraged to use a hearing aid in the non-implanted ear for possible improved sound localisation and improved speech perception in noise.¹⁴

Educational environment

The maximum benefit in both speech perception and language development is achieved when a child is enrolled in an auditory-

orally based, and family-centered therapy programme. Children with CIs in oral communication programmes developed language at a faster rate than children in manually based programmes.¹¹ Cochlear implantation accompanied by aural rehabilitation increases access to acoustic information of spoken language. This leads to higher rates of placement in mainstream schools and lower dependence on special education.

Children implanted prior to educational placement were significantly more likely to succeed in mainstream schools than those implanted after educational placement.

Children implanted prior to educational placement were significantly more likely to succeed in mainstream schools than those implanted after educational placement. Significantly more profoundly deaf children with CIs were attending mainstream schools when compared with those with hearing aids. Deaf young adults not educated in mainstream elementary and post-secondary school are less likely to pursue secondary education and are more likely to be underemployed or unemployed.¹⁴

Parents

For most parents, the primary motivation for getting a CI for their child is to help their child to learn to talk, to understand speech and participate in the family social environment and the world at large.¹¹ Parents have reported many benefits of cochlear implantation, including increased self-esteem, reduced isolation, closer relation-

ships with siblings and peers, improved language, more intelligible speech, and the ability to use the telephone. A universal reaction of the parents was that these observable benefits of the implant have resulted in brighter prospects for the child's future.¹

References available on request.

IN A NUTSHELL

Cochlear implants provide children with severe-to-profound hearing loss greater access to sound and consequently makes it easier for them to learn to talk.

Referral guidelines have changed to include:

- bilateral severe-to-profound or moderate-to-profound hearing loss
- no minimum age for referral
- restricted or no useful benefit from hearing aids
- children with additional handicaps.

Careful evaluation over time, including hearing aid trial, remains paramount.

Congenitally deaf children who undergo implantation before 2 years of age show greater benefit than children who are implanted after 3 years of age.

Criteria for paediatric CI candidacy include placement of the child in an educational environment that encourages the development of auditory and oral language skills.

Children in oral educational programmes benefit more from a CI than children in total communication or sign language programmes.

Benefits for congenitally deaf adolescents may in general be more limited. However, prospective patients should be considered on an individual basis.

Involvement of parents in rehabilitation is essential.

Dynamic technological advancements in implant design and speech coding strategies continue to improve outcomes with CIs.

Life-long commitment of the implant team to the child and the family must be guaranteed.