

19 Candidacy Considerations and Other Medical and Surgical Issues for Cochlear Implantation in Children

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19.1 Candidacy

The cochlear implant candidacy is becoming wider as children with more and more residual hearing are now considered. The children considered for a cochlear implant can be classifed into a pre-lingual category when the hearing loss has occurred before the child has been able to hear and learn to copy speech; a peri-lingual category, when the child has been deafened during the initial acquisition of speech; and a post-lingual category when the hearing loss has occurred after the child has acquired speech. Sadly, the peri-lingual category will lose their ability to speak if no intervention occurs. Recent studies [\[1](#page-14-0)] suggest that cochlear implant surgery for pre-lingually hearing impaired children should be undertaken under the age of 1 year to achieve the optimal speech and language outcome.

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19.1.1 Medical Considerations: Discovering the Cause of the Hearing Loss

The cause of the hearing loss should be determined where possible, as this may infuence the surgical technique and the cochlear implant outcome. Knowing the cause of the hearing loss is essential for counselling the parents and deciding on the appropriate post-implant training.

19.1.1.1 Intrauterine Causes: Intrauterine Infection, Ototoxic Medications and Genetics

Genetic hearing loss can be subdivided into syndromic and non-syndromic causes. The syndromic causes may be linked to other disabilities such as loss of vision, kidney failure, developmental delay or heart disease that need evaluation prior to any surgical intervention. Often there are facial clues such as the white forelock, different coloured eyes and a wide space between the eyes seen in Waardenburg syndrome [[2\]](#page-14-1). Jervill Lange Nielson syndrome is a profound hearing loss associated with a prolonged QT interval on the electrocardiogram that results in faints and loss of consciousness and causes sudden death in half the sufferers before the age of 15 years [\[3](#page-14-2)].

The commonest non-syndromic cause is due to Connexin 26 [[4\]](#page-14-3). A genetic defect that affects the intracellular gaps and causes loss of electrical ionic charges within the cochlea. There are many other genetic causes of hearing loss. One example is 'large (or enlarged) vestibular aqueduct syndrome' (LVAS), which is often associated with Pendred syndrome [[5\]](#page-14-4) (Fig. [19.1a, b\)](#page-1-0). The hearing loss may deteriorate during childhood. These children usually experience transient dizziness during the recovery after cochlear implant surgery.

Viral illnesses during the frst trimester of pregnancy can cause hearing loss, such as rubella [[6\]](#page-14-5) and CMV [\[7](#page-14-6)]. Rubella may be associated with eye and cardiac problems. Fortunately, rubella deafness is rare now in developed countries where vaccination occurs. Intra-uterine CMV may give no hearing loss or varying levels of hearing loss up to profound hearing loss, which can be unilateral or bilateral. It may be associated with other conditions such as developmental delay. If CMV can be detected by neonatal (PCR) screening and, if the appropriate treatment (Valganciclovir) can be given within the frst 21 days a profound sensorineural hearing loss may be prevented [[8\]](#page-14-7).

Ototoxic drugs, especially during the frst trimester may also cause hearing loss as the mother may have been unaware she was pregnant. In some countries, antibiotics such as gentamicin are frequently prescribed.

19.1.1.2 Birth Events: Prematurity, Hypoxia and Jaundice

Prematurity (below 32 weeks gestational age) can be associated with hypoxia at birth and this is a major cause of perinatal hearing loss. Hypoxia is common in premature infants.

Hypoxia can cause a form of auditory neuropathy spectrum disorder (ANSD), which causes a loss of inner hair cells with survival of outer hair cells [[9\]](#page-14-8). Using otoacoustic emissions as a screening tool to detect congenital hearing loss can lead to false reassurance and, later, the children may have diffculty perceiving speech using conventional hearing aids [[10\]](#page-14-9). In the past, this was attributed to 'central auditory dysfunction (CAD)' and it was initially believed that a cochlear implant would be unhelpful. Fortunately, ANSD associated with birth hypoxia offers a favourable outcome using a cochlear implant. Unfortunately, other forms of ANSD may not be so amenable. Genetic forms of ANSD can be divided into pre- and post-synaptic causes [\[11](#page-14-10)]. Post-synaptic causes are often associated with abnormalities of the cochlear nerve and other disabilities such as visual loss.

Perinatal jaundice is common and usually settles within a few days. Kernicterus occurs with excessive bilirubin levels. A common cause is a mismatch of the Rhesus factor between the mother's blood and the neonate's blood [[12\]](#page-14-11). Deafness due to kernicterus occurs because the cochlea flls with bile pigments causing loss of hair cells. Neurological problems, such as cerebral palsy, can also arise because the bile pigments damage nuclei within the brainstem.

19.1.1.3 Post-natal Causes: Meningitis, Trauma and Ototoxic Medications

Severe or profound hearing loss can occur during infancy and childhood. Meningitis causes cochlear damage when the organism passes up the cochlear aqueduct to reach the inner ear. The infammatory debris (pus) may become ossifed and block the scala tympani. The cochlear aqueduct closes in most humans later in life and the threat of cochlear damage is lessened. Ossifcation within the cochlea prevents

Fig. 19.2 Obliteration of the basal cochlear turn due to osteogenesis as shown by CT. (The CT is reversed to show bone as a black image)

electrode insertion (Fig. [19.2\)](#page-3-0). So meningitis in infants is treated as an emergency and cochlear implantation should not be delayed once the hearing loss has been verifed [[13\]](#page-15-0). Other viral illnesses causing encephalitis such as measles, mumps and rubella can have a similar effect.

Other causes of post-natal hearing loss include trauma, ototoxic agents, severe middle ear infections and cholesteatoma. Large vestibular aqueduct syndrome (LVAS) may only become evident during childhood with stepwise drops in hearing, particularly after head injuries.

19.1.2 Medical Considerations: Laboratory Investigations

Some investigations should always be undertaken prior to surgery. Some conditions may require special investigation. If the investigations show there would be an adverse outcome, the parents or guardians need to be informed and counselled.

Electrocardiogram (ECG) This test is done routinely for children with severe or profound congenital hearing loss, especially if there is any history of falls or blackouts to exclude Jervill Lange Nielson syndrome.

Blood tests may have been performed as part of the investigation for the initial hearing loss. For cochlear implant surgery routine blood tests are not normally required, unless there is a family history of excessive bleeding or bruising, tests to check platelet and coagulation factors may be required.

Genetic testing can be undertaken. Tests for connexin 26 and 30 are now easily available in Australia but tests for other genes such as the Pendred gene are more complex to obtain. It is anticipated that broader genetic testing will become more readily available for hearing loss.

19.1.2.1 Radiology: CT Scan and MRI

Computerised tomography (CT) is often avoided because of radiation concerns as most abnormalities can be detected on magnetic resonance imaging (MRI).

MRI has a defnite role and is always required. The MRI scan shows the fuid space of the cochlea, vestibule and semi-circular canals, allowing identifcation of any abnormal anatomy. After meningitis, the patency of the cochlear turns can be determined (Fig. [19.1a, b\)](#page-1-0). LVAS can be detected (Fig. [19.2\)](#page-3-0). Mondini and Michel abnormalities are evident on MRI. The vestibular labyrinth is abnormal in some ears affected, for example after intra-uterine rubella, and this can make identifying the entrance to the basal coil diffcult to locate. Inner ear abnormalities may be associated with an abnormality in the course of the facial nerve; in these cases, a CT scan can be helpful.

Most importantly, the contents of the internal acoustic meatus (IAM) can be seen on a transcanal view. Normally four separate nerves can be seen (Fig. [19.3a\)](#page-4-0). In Fig. [19.3b,](#page-4-0) only the facial nerve is present and the cochlear nerve is not visualised. Even, if only three or two nerves are counted, it is possible that cochlear nerve elements are absent or hypoplastic and may have fused with another nerve or are too small to see on the MRI scan resolution.

Fig. 19.3 (**a**) A MRI transcanal view of the contents of the internal acoustic meatus showing the presence of all four nerves (cochlear nerve, facial nerve, superior vestibular nerve and inferior vestibular nerve). (**b**) A MRI transcanal view showing absence of the cochlear nerve in its normal position

19.1.2.2 Electrophysiology: Electrocochleography and Electric Auditory Brainstem Potentials

The introduction of neonatal hearing screening and follow-up diagnostic auditory brainstem responses (ABR) enables children to be identifed early and accurately within a few months of their birth.

In some cases, further electrophysiological testing is used to determine candidacy for cochlear implant surgery and can be performed with an audiologist or biomedical engineer. A surgeon inserts the electrode through the tympanic membrane to perform transtympanic electrocochleography (TTECochG) and transtympanic electric auditory brainstem potentials (TTEABR).

Transtympanic electrocochleography has become virtually redundant for the majority of paediatric candidates, as it can be replaced by auditory brainstem responses (ABR). It still has a role for cochlear nerve dysplasia, for older children who cannot be tested except under general anaesthetic, and those who have otitis media with effusion as well as signifcant hearing loss. The ECochG is quick to obtain and does not need any masking of the contralateral ear. The author utilises a 'golf club' electrode, which is inserted through a posteriorly placed myringotomy incision under direct vision [[14\]](#page-15-1). Although it is very rare for a needle electrode to perforate an abnormal round window, the 'golf club' has a rounded surface making any round window damage unlikely (Fig. [19.4](#page-5-0)). The input impedance of a 'golf club' electrode is far less than a needle and allows larger recordings, which are not vulnerable to electrical interference within operating suites.

Fig. 19.4 The golf club electrode positioned in the round window niche

Transtympanic electric auditory brainstem response (TTEABR) testing can indicate if there are any useable cochlear nerve fbres (Fig. [19.5](#page-6-0)). TTEABRs are diffcult to obtain and require a 'golf club' electrode or a silver ball electrode placed accurately in the round window niche [[14\]](#page-15-1) (Fig. [19.4](#page-5-0)). The output of a cochlear implant is used to stimulate the cochlea via the cochlear implant software, which is synchronised to the recording equipment. The stimulating electrical charge must be delivered directly into the basal coil of the cochlea without diffusion into the middle ear tissues. The presence of otitis media with effusion makes TTEABR recording diffcult.

TTEABR is undertaken when the cochlear nerve cannot be identifed using transcanal MRI. In these cases, it is not known if the dysplastic cochlear nerve is entirely absent, or whether the nerve is hypoplastic or fused with an adjacent nerve. A recent consensus stated that a conventional cochlear implant should be offered frst when a cochlea is present, and the auditory brainstem implant be offered only if the cochlear implant fails to provide adequate stimulation. A positive or negative TTEABR provides extra information when counselling the carers.

19.1.2.3 Vaccinations: Rubella and Meningitis

Rubella Intrauterine rubella was one of the commonest causes of congenital deafness until the measles, mumps and rubella (MMR) vaccine was introduced in the 1980s [[6\]](#page-14-5). Rubella; caused eye and heart problems in addition to hearing loss. The hearing loss was often associated with abnormal development of the otic capsule.

Meningitis Three types of meningitis occur: bacterial meningitis, viral meningitis and aseptic meningitis [\[15](#page-15-2)]. There is no vaccine for viral meningitis. Bacterial meningitis is the most serious illness and it is most often due to pneumococcus. There are several different strains of pneumococcal meningitis. Prevenar® vaccine is given in two doses initially. Pneumovax® vaccine is usually given after the age of 2 years and should not be given with Prevenar® vaccine but several weeks later.

19.1.3 Age Considerations: Auditory Plasticity

Age It has now been shown convincingly that the optimal speech and language outcome occurs when a child receives a cochlear implant when under 2 years of age [\[1](#page-14-0), [16–](#page-15-3)[18\]](#page-15-4). Profoundly deaf children who receive a cochlear implant after the age of 8 years have a far less favourable outcome, with little language understanding. This is due to the process of auditory plasticity.

Neural plasticity is the ability of the brain to develop certain neural pathways whilst the brain has the ability to change. The more essential the task, the more critical the time period for the development. For example, a foal should stand next to its mother within a few hours of birth. If the foal cannot stand within a day, the foal will never develop the ability to walk as the brain no longer has the ability to learn the task (loss of plasticity). In humans, both audition and vision have a critical time period for development after birth. Auditory plasticity is the ability to learn to hear, understand speech and this leads to the ability to produce speech. A baby listens and learns to babble using the sounds of speech soon after birth. By the age of 2 years, most children have developed some meaningful speech.

Auditory plasticity declines as the child ages. For a profoundly deaf child, if there has been no audition even using hearing aids, only limited beneft from a cochlear implant can be expected if the child receives a cochlear implant after the age of 6 years. Various studies have shown the beneft of early cochlear implantation and an ongoing study in Australia is suggesting that the optimal time for implant surgery is below 1 year of age [[1\]](#page-14-0).

Similarly, auditory plasticity affects the ability to utilise bilateral cochlear implants and to develop directional hearing and the benefts of binaural hearing. A teenager who has successfully received a cochlear implant in early childhood and then receives the contralateral cochlear implant when older, may have less speech discrimination from the second implant than from their frst implant and is unlikely to develop directional hearing.

19.1.4 Surgical Considerations: Age, Blood Volume, Incision, Mastoid, Skull and Skull Abnormalities and Osteogenesis

Surgery for infants, especially if under the age of 1 year, requires careful evaluation and surgical expertise. Blood loss is to be avoided, especially if bilateral simultaneous surgery is performed. The average blood volume of a 7 kg infant is 560 ml (approximately weight in kg multiplied by 80). Underweight or frail infants may need to defer surgery until they are sufficiently robust.

Large incisions are more likely to become infected and to suffer keloid formation. A small postaural incision is better because the skin is elastic in young children [\[18](#page-15-4), [19\]](#page-15-5). A pocket can be made, posteriorly, to hold the receiver snugly. The modern cochlear implant packages are thin, but the parents should be informed that the packages of the implants can be prominent in infants for several months after the surgery. Antibiotics are given prophylactically after induction and continued for 5 days after the surgery.

The development of the cochlea, middle ear and mastoid affect the surgery in infants. Fortunately, the cochlea is fully developed during the frst trimester of pregnancy. The middle ear, ossicles and the antrum are at adult proportions but the mastoid bone develops considerably during the frst few years of life [[20\]](#page-15-6). The mastoid bone expansion is about 0.6–0.9 cm/year in length and width and 0.4 cm/year in depth in the first year, followed by half that rate until the age of 607 years. At puberty, there was a slower growth to reach adult size.

At surgery, there is limited access through the mastoid and this contain haematopoietic marrow that can bleed and needs special attention. Once the limited mastoid space is secured, the posterior tympanotomy and middle ear structures are viewed as in adult surgeries (Video 19.1), however, in infants, the petrous bone is slightly superiorly rotated, and the round window will be found slightly higher. The horizontal facial nerve is often seen from the antrum and even the stapes superstructure may be seen through the aditus in young children (Fig. [19.6\)](#page-8-0).

The thickness of the temporal bone may be only a few millimetres in infants under 1 year of age. A bony well is drilled to anchor the receiver-stimulator, but this is often shallower in a small infant compared with an adult. There is signifcant posterior growth of the temporal bone during early childhood [\[21](#page-15-7)]. The electrode lead wire can be subject to 2 cm of growth. All current cochlear implants have an expandable electrode lead which can lengthen to accommodate this growth. The bony bridge remaining above the tympanotomy does not alter signifcantly and can be utilised to fxate the medial end of the array.

19.2 Osteogenesis

The formation of bone within the cochlea occurs after meningitis in young children, but can also occur in adults, particularly with pneumococcal meningitis. The cochlear aqueduct is patent in the frst few years of human life and infection from the cerebrospinal fuid can pass into the basal turn of the cochlea. The exudate can ossify within a few months. Initially, the soft new bone can be scooped out but after time it hardens and it may not be possible to insert a cochlear implant array. It may be possible to insert the array through the scala vestibule, in front of the oval window. There are also some special arrays provided by the manufacturers that are inserted through channels superior to the promontory and inferior to the promontory.

Urgent referral to a cochlear implant programme is essential if a child has severe or profound hearing loss following meningitis, as early surgery may ensure a cochlear implant can be placed.

19.3 Soft Surgery

Ears that have some low-frequency residual hearing can be implanted in the hope that the low-frequency hearing can be retained, and a 'hybrid device' can be utilised.

The hair cells are in the scala media and the implant array is inserted into the scala tympani so inserting the fne array slowly and gently can prevent displacing the basilar membrane and causing trauma to the hair cells. If the array is inserted too quickly, it can send the perilymph surging down the cochlea like a tsunami causing possible trauma. Often steroids are given to limit any infammatory response.

19.4 Surgery in Presence of Skull Abnormalities: CHARGE and Other Dysplasias

The letters in CHARGE syndrome stand for: Coloboma of the eye, Heart defects, Atresia of the choanae, Retardation of growth and development, Genital hypoplasia and Ear abnormalities and deafness. The diagnosis is via major and minor features, and the majority of children have a chromosomal abnormality. In these children, the cochlea may be malformed (small and hypoplastic) with absent lateral semicircular canals causing the cochlea to rotate posteriorly [[22\]](#page-15-8). Atresia or hypoplasia of the oval and round windows may occur. It can be diffcult to locate the round window and the basal cochlear coil. CT-guided image surgery may be necessary to insert the array.

The cochlear nerve may be hypoplastic or absent. A trans-canal MRI view is essential and TTEABR may be helpful in reaching a decision to use a conventional cochlear implant or to consider a brainstem cochlear implant. Sometimes a favourable outcome occurs despite a seemingly compromised cochlear nerve. [[23\]](#page-15-9) Initially, the brainstem cochlear implants were used only for neurofbromatosis type two (NF2) patients and the results were often disappointing. Ten years ago, Collettti began using brainstem implants in non-NF2 cases including children with often encouraging results [\[24](#page-15-10)]. The surgical team must include an electrical neurophysiologist to help correctly place the electrodes in the brainstem and to avoid any unwanted stimulation of adjacent nuclei.

19.5 Further Surgical Considerations and Risks

Both the surgeon and the anaesthetist usually undertake the normal immediate preoperative checks. The surgeon sees the child and the parents prior to the surgery date. The surgeon should enquire about previous anaesthetics and if there were any complications. Any previous anaesthetic diffculties, any allergies and any recent or present infection should be noted.

The surgeon and the anaesthetist have to inform the parents of any potential risks.

19.6 Immediate Possible Complications of Surgery: Facial Nerve Palsy and Misplaced Electrode Array

The risk of damaging the facial nerve when undertaking the posterior tympanotomy is no greater than in adult surgeries. The mastoid cavity can be smaller, but the risk is very minimal with a well-trained surgeon. The risk of a delayed facial palsy in children is less than in older people as avascular changes are uncommon.

The electrode array can be misplaced, particularly in children with abnormal anatomy. Misplacements into a hypotympanic cell, the vestibule, semicircular canals or the internal auditory meatus are possible. These problems can be detected using intraoperative electrophysiological tests. [\[25](#page-15-11)] Neural Response Telemetry (Cochlear®) or the equivalent tests using other manufacturer's devices, show the neural responses and that the electrodes are correctly placed. If there is no response then intraoperative radiology should be requested.

Intraoperative cochlear implant evoked EABR testing does require a dedicated electrical neurophysiologist or audiologist and provides a robust means of testing the function of each electrode. This is most helpful in children with cochlear nerve dysplasia, as the mapping parameters can be adjusted to obtain an optimal cochlear implant evoked ABR response.

19.7 MRI After Cochlear Implantation

The cochlear implant receiver-stimulator package contains a magnet, which can be dragged or even displaced during a MRI examination. Furthermore, the MRI can heat long electrode leads causing tissue damage. Initially, the solution was to explant the magnet, perform the 1.5-Tesla MRI and then reimplant the magnet after the examination. An incision in the scalp is made behind the stimulator package and the magnet is removed and then replaced after the MRI has been performed. As this requires surgery, there is always a risk of damaging the device or infection. The next solution was for 1.5 Tesla MRI compatible cochlear implant devices, when a very tight bandage was wrapped around the head to prevent the magnet from moving. This can be painful and unfortunately, there are case reports of the magnet being displaced from the package and even tearing the retaining sialastic [[26](#page-15-12)].

Most manufacturers now offer a 3.0-Tesla MRI compatible device. At time of writing, the limit seems to be 3.0 Teslas and the head shadow effect is still evident. MedEl was the frst to provide a magnet that could align itself to the MRI. The other manufacturers are now offering similar solutions.

19.8 Delayed Complications of Surgery [\[27](#page-15-13)]**: Infection, Cholesteatoma, Displacement of the Receiver-Stimulator Package, Extrusion of Electrodes, Device Extrusion, Electrode Malfunction and Failure of the Magnet to Keep Contact**

Post-operative infection can occur some months or even a year after the surgery. If an infection occurs around the receiver-stimulator package, antibiotics may not be able to control the infection if a bioflm is present. A broad-spectrum antibiotic should be prescribed, and often given intravenously and continued orally for several weeks. If the infection appears to have settled but when the antibiotics are ceased, if the infection recurs a few weeks later, there is no option but to remove the package and wait for resolution of the infection in the wound before inserting another cochlear implant. The electrode array can be left in the cochlea by cutting the array close to the round window/cochleostomy but the package and leads must be removed. Leaving the array in the cochlea helps to identify the position of insertion and also lessens the risk of fbrosis closing the cochlea or infection entering the cochlea and causing osteogenesis.

Cholesteatoma formation within the implanted ear is uncommon. If this occurs, surgery is needed. Depending on the size of the cholesteatoma, it may be necessary to remove the cochlear implant, and a blind sac closure of the external meatus is performed. Most surgeons stage the procedure and wait for 3 months before reexploring the ear to ensure the cholesteatoma is completely eradicated. Occasionally,

when the cholesteatoma is caught early, only local surgery is required and it is possible to preserve the cochlear implant.

Displacement of the stimulator receiver package is rare. If the device was placed in a tight periosteal pocket, then it can only displace towards the external meatus. This can be prevented by drilling a socket in the skull bone or by placing a tie around where the electrode array leaves the package. If the head coil can still be placed, then there is no need for surgery to reposition the package unless it is too close to the pinna. If surgery is undertaken, then care should be taken not to pull the array out from the cochlea.

Extrusion of electrodes can occur if the implant was performed at an early age and there has been considerable skull growth during childhood or following infection or a signifcant knock to the head. The problem can be detected on device programming (mapping) and on radiology. Often the array can be reinserted if picked up early. Sometimes the existing cochlear implant array cannot be reinserted, or shows evidence of damage after reinsertion so it may be necessary to reinsert another device. The new array can usually be reinserted into the scala tympani, but if there is considerable fbrosis it may still be possible to insert it through the scala vestibuli.

Electrode malfunction is not uncommon. All the leading brands have had to recall devices when a manufacturing fault occurs. The companies have been exemplary in admitting the fault and offering assistance to replace the faulty devices.

Malfunction of a few of the electrodes occurs in some of the devices, especially after several years. In Cochlear® devices, up to 5 of the 22 electrodes can be faulty without signifcant impact on the recipient's hearing, depending on the location of the faulty electrodes. The ultimate decision to replace the device is not dependent on the number of faulty electrodes but on whether or not the hearing has been compromised.

Failure of the magnet to keep contact occurs due to thickening of the scalp or very strong sturdy hair. For obese children, it is better to place the receiver/stimulator package upright above the ear where the scalp remains thinner. In some children of African descent, the hair is very strong and a small circle of hair has to be shaved away to allow the head coil to stay in contact.

19.9 Pain After Cochlear Implantation

Cochlear implant surgery is not very painful, on average children use paracetamol for 2 days following surgery [[28\]](#page-15-14). Long-term pain around the site of the stimulatorreceiver package following surgery is rare. It is necessary to exclude an infection. The area should be palpated to see if there is any swelling and the tympanic membrane is checked for any sign of otitis media. An ultrasound examination can show if there is fuid (haematoma or seroma) around the package. If the magnet is too strong, it may cause reddening of the skin but rarely causes any signifcant pain.

Skin erosion due to the magnet is rare in children but can be a problem in elderly patients.

Sadly, some congenitally deaf recipients may have received the implant at an older age and have not been able to distinguish useable sounds and wish to be disassociated with a cochlear implant. Similarly, we have seen a few older teenagers, develop a sensation of pain associated with a previously fne, functioning device. Investigations may show no cause, however, sometimes there is co-existing anxiety or depression. Some have had the device removed due to their symptoms of pain and have felt that their pain was relieved.

19.10 Loss of Residual Hearing

If residual low frequency has been retained after surgery, a 'hybrid device' can be used which combines a cochlear implant and a hearing aid. Sadly, often the residual hearing slowly declines over time and the hearing aid component may is no longer be viable. In some ears, the residual hearing is lost suddenly about 2–3 months after the surgery. Various explanations have been offered. One possibility is that the vein of the modiolus, which drains venous blood from the hair cells becomes obstructed as fbrosis occurs around the electrode array. Even after successfully retaining some low-frequency hearing, some patients may elect to just use the cochlear implant, rather than a hybrid device, as they fnd the sound provided by the cochlear implant is clearer. For this reason, a full-length electrode array is preferable to a shortened array.

19.11 Removal of a Malfunctioning Cochlear Implant: Indications and Outcome

19.11.1 Indications for Removal of the Cochlear Implant, Outcome After Reinsertion

Indications for removal, commonest reasons are device malfunction or infection (especially if cholesteatoma has occurred). Less common reasons are device or electrode extrusion, pain, or just the wish of the recipient. Details of these complications have previously been discussed.

Outcome after reinsertion, it appears that there is little change in the performance of the cochlear implant after reinsertion [\[29](#page-15-15)]. On removing the array, the exact site of reinsertion should be noted, as there have been cases when the array is inserted into a false tract. The insertion can be checked by electrically using neural responses such as NRT or ART. It may be diffcult to remove the array if there is osteogenesis and the array can be snapped if excess traction is applied. In such cases, it is often possible to reinsert into the scala vestibuli.

19.12 Conclusions

Paediatric cochlear implants are a modern success story, allowing children with severe or profound hearing loss to learn to listen, hear and speak. For the majority of children, timely provision of a cochlear implant allows the child to have suffcient hearing and language to go to a regular school. This chapter covers aspects that the surgeon in the cochlear implant team may encounter. The surgeon is not only involved in the surgery, but also in the pre-operative candidacy evaluation and in the long-term follow-up of the recipients.

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