

CHAPTER 8.

HEARING TECHNOLOGY

1. HISTORICAL



Figure 1: Antique hearing horns. A great variety of these were available, many relying on disguises to minimise the embarrassment of an evident disability.

In the last hundred years there has been major advances in communication technologies. Management has not only advanced from the hearing horn to the cochlear implant (the only complete replacement of a human sensory organ by technology) but this is now supplemented by complex aid electronics and also additional implantable technologies that stimulate hearing by other means.

2. HEARING AIDS

These devices have undergone continual refinement in size, function and cosmetic aspects. A range of models is available, mostly ear-level designs, the older body-level devices being used mainly for very severe losses that require greater power.



Figure 2: Early electric hearing aids. Given the constraints of the technology, device and battery bulk were considerable for the wearer.

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Air Conduction (AC) Aids

- Behind the Ear (BTE)
- In the Ear (ITE)
 - Mould fitting
 - Bell tip
 - Deep insertion

Bone Conduction (BC) Aids

- Head clip
- Head band

Advances in digital technology have generated overall aid design refinement. The devices are small and lighter. Multiple microphones provide better pick-up. Complex circuitry permits a range of programs, e.g. for quiet or noisy surroundings. The amplification is tailored to complement the losses noted on the evaluation audiology. Batteries are smaller and more powerful. The BTE models often use fine, less visible catheters with a small “bell” tip inserted into the EAC that is more comfortable and less occlusive.

In addition, there has been a ground shift in attitudes to the cosmetic aspects. Whereas in the past, considerable efforts were made to disguise hearing horns, and aids were for many year supplied in flesh-coloured tones, the modern aid is likely to display a trendy titanium finish or other creative design. Similar approaches are seen in the cochlear implants, particularly amongst younger users.

Air Conduction Aids

Given their miniaturisation and improved complexity of abilities, AC aids are generally the model of choice. The choice of BTE vs. ITE is less clear, made on a variety of considerations,

including power, cosmesis, comfort, or occlusion aspects, amongst others.



Figure 3: A modern BTE AC aid; trendy non-cosmetic finish.

More powerful BTE devices and the ITE models require a mould fitted to the individual EAC. A poor fit results in “feedback howl” as the aid’s amplified sound restimulates the microphone causing an ascending electronic noise. The moulds may cause perspiration, plug the ear, become blocked by wax, or cause noise during eating.

The “bell” tip devices avoid many of these difficulties.



Figure 4: A BTE AC aid for very severe hearing loss, using a custom mould to minimise feedback howl.

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The “Lyric” model is a device fully set deeper into the external canal, retained in a soft coating left in situ continually. This requires a wax-free EAC, sitting by a specialised audiologist with a microscope, and occasional battery recharge under audiology auspices.



Figure 5: A Lyric deep insertion ITE aid in situ. The nylon loops facilitate removal for recharging or other purposes.



Figure 6: Cable CROS aid, for unilateral profound SND. Later models use Bluetooth.

Bone Conduction Aids

Some ears cannot utilise air conduction aids for a variety of reasons: EAC atresia, wax, exostoses, or chronic infection; occlusion, mastication noise,

or perspiration due to blockage; feedback howl, manual disability or cosmesis.

BC aids avoid the EAC by providing sound by skull vibration. This stimulates the cochlear hair cells similar to normal sound transmission, and is highly effective. The vibration is produced from a buzzer placed on the hard bone of the mastoid, behind the ear. The vibrator is kept in place by either a head clip or headband. The disadvantages of the method are pressure effects over the vibrator site that may cause skin ache, and the more overt cosmetic appearance of the head clip/headband.



Figure 7: Head clip BC aid. Useful for deafness when the EAC is aid-intolerant, but often uncomfortable.



Figure 8: “Softband” BC aid, used for infants with conductive loss, e.g. canal atresia.

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3. IMPLANTABLE TECHNOLOGY

In recent decades, surgeons and technology companies have looked to restoring hearing with implantable or semi-implantable devices, with major successes. Passive devices, essentially ossicular replacement devices (stapedectomy pistons, PORPs and TORPS) are described in the chronic ear surgery section. Active technological implants are driven by power packs, usually extracorporeal at this time.

Classification of Active Hearing Implants

- Cochlear Implants (CI)
- Active Middle Ear Implants
- Floating mass transducer
- Driver models
- Bone conduction implants
- Bone Anchored Hearing Aids (BAHA)
- Active Bone Conduction Implants

3.1. COCHLEAR IMPLANTS

In a period of some thirty years, cochlear implantation (CI) has advanced from the early clinical phase to become the only current example of a complete body sensory system being replaced by technology. In that time literally hundreds of thousands of patients have been delivered from profound deafness, to achieve near normal communication.



Figure 9: Cochlear implant processor. CIs have delivered lifetime benefits for the deaf child.

CIs have two components, the implant itself, placed under the skin above and behind the ear, and an external processor, similar to a BTE aid with batteries and an attached induction coil. The processor receives sound and converts this into coded electrical stimuli. These signals and power is then induced in the implant via a complementary coil. The two coils being held in position by opposing magnets.

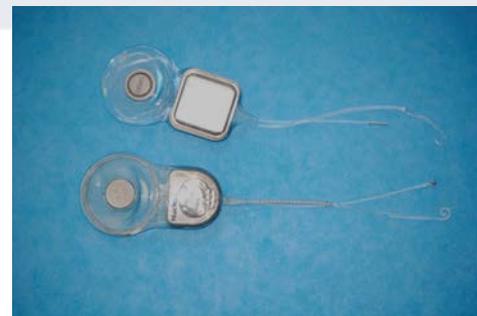


Figure 10: Cochlear implants. The induction coils to the left receive electronic data, which is processed and enters the cochlea via the electrode arrays on the right.

Implantation has been a particular success for the profoundly deaf child, who was once capable of only limited signing communication to a small section of the community, and is now capable of reaching the highest levels of education to which the individual circumstances permit.

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Implantation has considerably different clinical implications for adults and children; the two groups are therefore discussed separately.

3.1.2. PAEDIATRIC COCHLEAR IMPLANTATION (PCI)

The Impact of Deafness

The normal development of hearing and speech is dependant upon development of the auditory cortex. It is this development that cochlear implantation essentially addresses.

The auditory cortex undergoes substantial development of its size, synaptic connections and other CNS organisation during the first two years of life. This development is dependant upon auditory stimulus from the ear progressing normally. If this stimulus fails, the cortex remains under-developed, and in time the neurones present become diverted to the other adjacent sensory areas, such that a never-stimulated cortex in the adult is an atrophied example of the normal, incapable of adequate response to sound. In addition, “cross-plasticity” changes may produce aberrant responses upon stimulus, e.g. tactile sensations.

If one ear is deaf, the better ear can largely stimulate both cortical structures, but the ability of the deaf ear to respond to PCI will gradually reduce with time.

There is thus a compelling impetus to implant the profoundly deaf child as early as practical, preferably in both ears. Many surgeons will endeavour to operate, perhaps bilaterally, at six months, if there are no contraindications to surgery at this age.

Presentation

Deafness in children may occur in isolation, or may be a result of more generalised medical problems. Many of the problems in otherwise normal children are genetic; growing numbers are now being recognised as particular genome mutations. Others may be part of recognisable clinical situations; especially syndromal patterns. A sizable minority however, continue to defy diagnosis. The task for the clinicians is to identify the overall spectrum of conditions, their pathological manifestations, and the implications for successful PCI.

Investigation

- i. Audiology: Initial and ongoing audiology monitors the child’s hearing ability as closely as possible from birth to the lead-up to PCI. Initially, objective assessments (ABR, ECoG, OAEs, and possibly transtympanic EABR) assess for any residual responses. Later, around six months age, VROAs and other pure tone assessments may be employed on older children.
- ii. CT and MRI scans are routine, to identify labyrinthine, VIII, or CNS pathology.
- iii. Developmental or other specialised paediatric expertise may be sought, particularly neurological, if there is evidence of CNS pathology.
- iv. The team psychologist assesses not only the child’s abilities, but also the motivations, expectations, or other aspects of the family situation and participation. This is of major importance for successful outcomes. A social worker may also be engaged for these purposes.

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Prognostication

It is important for the team to accurately assess the child's prognosis. Successful PCI may be defined as the outcome having met or exceeded the family's reasonable expectations, fully counselled to them before surgery, and the PCI process being performed to their satisfaction. Thus, the prognostication is the key, as this sets both the team's and the family's expectations.

To achieve this, the team must be aware of any potential threats. Such threats should be identified as to their physical location, their likelihood of effect, and the severity of impact on the child's ability. Adverse factors will affect abilities in three ways:

- i. The ability to **achieve CI stimulus** of the cochlea: This covers the availability of PCI, the family's compliance with the rigours of the process, the child's acceptance of the device, any intervening family or social disruptions (e.g. adolescence).
- ii. The ability to **transmit** such stimulus to the brain: Adequate electrode siting (e.g. congenital or meningitic labyrinthine abnormalities), VIIIth nerve status.
- iii. The ability of the brain to **process** the data received, whether neurological pathology, or a dysmature cortex, due to delayed PCI.

Evaluation of PCI candidature necessitates a team that can coordinate the evaluation of the child with a view to CI, often at a tender age. The team will focus on the areas that impact on the situation: six domains that represent the child's ability to respond to PCI:

- i. Status of the **cortical maturity**: The child's age, previous sound exposure (if deafened later in life), degree of deafness, prior aiding.

- ii. **Otological** aspects: The anatomy of the cochlea and VIIIth nerve, effects of meningitis (labyrinthitis ossificans), auditory dyssynchrony, or neuropathy of the VIIIth nerve
- iii. **Neurological** status: Assessment for damage resulting from CMV viral infection, cerebral palsy, hypoxia, kernicterus etc.
- iv. **General Medical** status: presence of medical conditions that may interfere, with PCI, e.g. CHARGE, renal failure, malignancy.
- v. **Psychological** status: Severe intellect difficulties, autism, ADHD.
- vi. **Family and Social**: Assessment of family motivation and any social problems.

Each of the six domains above is assessed for individual adverse factors. Within each domain, each factor is evaluated by likelihood and severity, 1-4, good to poor. The cumulative effect is assessed and the domain rated similarly. The collective domains are then rated for the overall effect, again similarly, 1-4, this final rating being the prognosis.

The team assesses the outcome, and counsels the family re this and the surgical process.

Surgery

PCI is now refined to the point that in expert hands a bilateral implantation on a six month old infant is a safe procedure. The optimal surgery employs a minimal postaural incision, small in size, such that parental distress is minimal.

The implant body is embedded in a pocket created between the pericranium and skull, the electrodes passing into the cochlea via a small mastoidotomy, the surgery lasting 30 minutes in routine cases.

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Figure 11: Minimal exposure cochlear implantation. The implant is sited in a pericranial pocket, with the electrode array passing into the cochlea via a small mastoidotomy.



Figure 12: CI surgery, closure site. Minimally traumatic surgery permits implantation at the earliest optimal age.

In a minority of cases, useful hearing in the lower frequencies may remain, prior to surgery. In these instances, hybrid devices are employed. Finer electrode arrays seek to preserve hearing, and postoperatively the CI stimulus is combined with conventional hearing aiding to provide a fuller auditory experience.

Postoperatively, a head bandage is applied for a week to enhance implant stability.

Habilitation

In children, the impetus is to develop listening, speech and language in line with that of the normally hearing child. The implant child falls into one of three broad categories. If the child is profoundly deaf from birth, early implantation is imperative to duplicate the normal cortical development process that permits the appreciation and application of hearing. Profoundly deaf children implanted later (i.e. prelingual) are at risk of slower progress perhaps with lesser outcomes, this situation worsening with longer delays. In other cases, the aiding of prior partial deafness may or may not suffice to achieve these ends.

However, children who have experienced useful hearing before deafness and are postlingual (speaking clear language), generally achieve well, as the auditory cortical development has been achieved.

Habilitation may involve auditory-verbal techniques where the family input is the key to the child's development of listening, hearing, and speech production. This relies heavily on a motivated family. Auditory-oral methods include visual cues. Signing techniques (hand gesture communication) are useful, but are now superseded by the first two methods, that provide the best social communication skills in mainstream society.

Complications

Surgical problems are now fortunately limited, due to the refinement of technique. Infection is very uncommon. Occasional implant or electrode failure or displacements are encountered, but replacement of a faulty implant is usually uneventful. The main surgical difficulties relate to a malformed or previously diseased cochlea;

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these are usually detected preoperatively and the family warned accordingly, provided the above evaluation is thorough.

Subsequent problems remain a risk. Poor parental motivation or unforeseen social, family, or other problems may remain undetected until after the event. Long term electronic problems or trauma to the implant may occur.

Unilateral Paediatric Cochlear Implantation

CI is now readily considered for the child with unilateral deafness, as children adapt well to the procedure. The child with a unilateral loss is more handicapped than previously realised, socially, during schooling, and later, economically. CI is therefore now recognised as optimal management, for those children who are otherwise suitable for CI. Management is much the same as for bilateral CI, but with generally more rapid and reliable longer term progress, given the better auditory experience by virtue of the unaffected ear. Lesser effectiveness remains a concern for those children considered later in life for these unilateral procedures.

3.1.3. ADULT COCHLEAR IMPLANTATION

Adult CI procedures differ from the paediatric in several ways. Generally, the patients are postlingual, or have well-developed speech (CI for adult pre-lingual patients carries a very guarded prognosis). A long adaptation to deafness may have occurred, giving lip-reading or other

additional skills. Few have cochlear damage or distorting disease. Motivation may be strong, especially in the aged or recently deafened. Many are familiar with the concept of hearing technology.

However, if deafness has been present for extended periods, outcomes will be affected in some cases. Strong support by associated family or friends for postoperative adaptation to the CI is desirable. Adults as a rule are not as supple as the young in adaptation to CI.

Presentation

Although the concept of adult CI has perhaps been less publicised than the paediatric, acceptance of this modality has steadily advanced such that the severely afflicted tend to be more readily referred for evaluation. This has been the case particularly for the aged deafened patient, where isolation has increased with the loss of friends, then from the family circle; CI has proven to be "God's gift" for this group.

Total adult deafness forms a lesser group than in the paediatric cases; most have hearing but are struggling with even the best aids. Hence, the thrust of evaluation commonly centres on speech discrimination. CI is appropriate when the patient's communication ability cannot cope even with the most advanced aiding.

However, evaluation, as with children, remains a thorough task. Aspects examined particularly include age effects (especially cognition and senility), motivation, postoperative support, and the duration of deafness. Psychological assessments are required to ensure that the patient's (and family/carer's) expectations and motivation are appropriate. In some cases social work support or input is relevant.

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Surgery

The adult surgical process and complications differ little from the paediatric. In adults however, bilateral surgery is best avoided. Adults differ from children with respect to the affect of surgery on vestibular function. This is rarely troublesome for the child, but bilateral surgery on an adult risks severe disequilibrium, which may remain uncompensated – a potentially disastrous outcome. Fortunately, vestibular problems are generally benign in CI, as the balance apparatus is damaged along with the hearing, such that CI produces relatively little effect, usually rapidly compensated. Bilateral adult CI is therefore usually sequential rather than concurrent.

As with children, in practiced hands the surgery is normally uneventful. The aged group may be more prone to wound repair delays, but these are not usually troublesome.

Rehabilitation

As above, adult patients are postlingual, all have developed speech patterns. However, considerable adaptation for best outcomes is necessary and best undertaken using a thorough program of auditory exercises. Nonetheless, the great majority do well, with many being very difficult to notice as deaf, and many achieving good telephone ability.

CI in Adult Unilateral Deafness

CI may be used in adults who are profoundly deaf in one ear, but the results are less certain than when this modality is employed in children; a greater proportion of cases may be disappointed. The surgery is best applied in recently deafened individuals, although many longer term cases will benefit.

The alternative is the use of CROS/BiCROS aids, or alternatively, active bone conduction implants if the contralateral ear has excellent hearing. Both these methods deliver clearer speech reception on the deafened side, eliminating “head shadow”, but not direction finding, stereo, or bilateral summation. The CI, whilst delivering these latter benefits, provides speech reproduction that usually does not match the BC/CROS methods. The appropriate benefits and the disadvantages thus remain an individual assessment.

Not uncommonly, adults struggle for many years before coming aware of the benefits of CI. Fortunately, this is becoming less common as the effectiveness of the surgery becomes more widely appreciated.

3.2 ACTIVE MIDDLE EAR IMPLANTS

These devices act by applying direct powered vibration onto the ossicular chain. They are thus valid only when this is practical and are not applicable in a range of congenital deformities when the middle ear cleft is disorganised. As an alternative, the vibration may be applied to the round window, if possible or preferable.

Models

- a) Floating Mass Transducer:
 - i. Med El Vibrant Soundbridge (VSB)
- b) Driver Devices
 - i. Esteem (Envoy Medical)
 - ii. Carina (Otologics)
 - iii. DACS (Cochlear)

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a) Med El Vibrant Soundbridge

The VSB utilises a unique floating mass transducer (a tiny barrel-like electromagnetic vibrator) that is clipped to the incus. It can also be sited against the round window, or adapted to fit onto ossicular replacement prostheses to enhance the performance of these passive devices. The device is powered by an external "Amade" processor, which induces implant power via complementary induction coils.

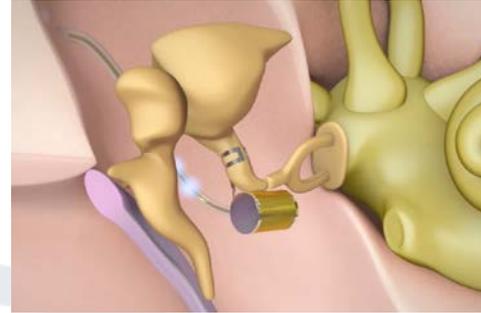


Figure 14: Soundbridge in situ, clipped to the long process of the incus. Vibration of the floating mass transducer produces stimulus of the cochlear hair cells, recognised as sound.



Figure 13. Vibrant Soundbridge active middle ear implant. The device functions via the tiny floating mass transducer at the tip of the cable, right.

VSBs are used in moderate conductive and sensorineural losses, or in moderate mixed losses. The surgery utilises a limited cochlear implant Keyhole incision, as above, with the body of the device stabilised in a pericranial pocket. Once in place, the device acts similarly to an aid, but with the Amade on the scalp above and behind the ear.

b) Driver Devices

The Esteem, Carina and DACS (Direct acoustic cochlear simulating) devices. These implants have an induction coil and electronic component that is fixed to the skull. They are attached to, direct, and power tiny vibrating rods ("drivers") that induce sound via the chain or round window.



Figure 15: Envoy Esteem implantable driver-type hearing device. The device requires dislocation of the incus off the stapes to prevent feedback howl.

The devices are suitable for moderate sensorineural, mixed or conductive losses, but are less commonly used at this time, being relatively complex to implant and utilise.

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3.3. BONE CONDUCTION IMPLANTS

Active bone conduction implants are fixed to the skull, creating sound by directly vibrating the cochlea.

Models

- a) Bonebridge (Med El)
- b) BAHA (Cochlear)

a) Med El Bonebridge:

In this device the vibrating component is in the implanted section itself, powered by an external processor that is similar to the VSB processor, above, via induction coils.

The vibrating cylinder is fixed in a bony well created in the mastoid. Bonebridge devices are implanted via a modified CI Keyhole technique, and are stable once in position.



Figure 16: Bonebridge active bone conduction implant. The vibrating cylinder (right) is fixed to the mastoid, the vibration stimulating the cochlea to produce sound.

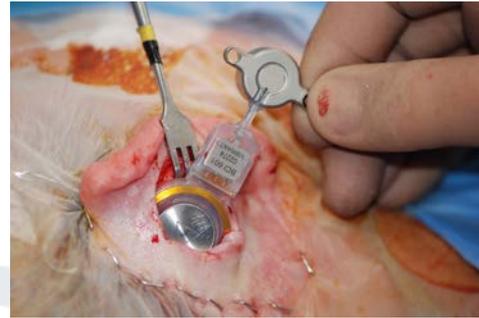


Figure 17: Implantation of a Bonebridge device using techniques similar to the CI methods above.

b) Cochlear BAHA

BAHA devices act via a titanium pedestal fixed to the skull behind the ear, either via a percutaneous (through the skin) titanium screw. Earlier models utilised a percutaneous pedestal. The later Attract version uses a transcutaneous (intact skin) magnetic attraction. The device is powered via an external vibrator, mounted either on the pedestal, or on the external magnet (in the Attract).

The surgery employed depends on the implant type. Percutaneous BAHAs use a linear vertical incision, but magnetic BAHAs require a larger scalp C-incision and head shave.



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(b)

Figure 18 (a): Percutaneous BAHA pedestal. Prone to local reactions, but valuable for hearing when other techniques are inappropriate. (b) Processor in situ.

supplied by the BC implants. If the contralateral thresholds are below 20db, a cochlear implant is required.

Once implanted, the Bonebridge and transcutaneous (magnetic) BAHAs have few problems, but the percutaneous BAHA has a frequent occurrence of pedestal-related difficulties.

Indications

Bone conduction devices have a variety of uses. They are particularly useful for if the patient cannot use a conventional aid, the situations described above for BC aids,

- i. The implants apply to moderate sensorineural losses, severe conductive loss, or severe mixed losses where the SND component is moderate. They have particular use in severe conductive losses, when aid use is uncomfortable and the discrimination is reduced by the extent of amplification used. The BC stimulation modality bypasses the conductive component, thus only amplification of any sensorineural loss is needed.
- ii. A second major use is in unilateral SND. When the contralateral ear has excellent sensorineural reserves, the BC implants (sited over the deafened ear) can eliminate the "head shadow" on the deafened side by stimulating the better cochlea, restoring communication on the deafened side, but without binaural effect, direction finding or stereo ability. To achieve these functions, a cochlear implant would be required, with perhaps lesser hearing quality than that

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