Implantable hearing aids in Children

Abstract
This review article gives an overview of the current implantable hearing aid devices which are available in clinical practice for children. Although hearing aids remain the frontline option for children with hearing loss, there are now other alternatives to rehabilitate hearing in children where hearing aids are not applicable. This article summarizes current views on cochlear implants, bone anchored hearing aids, middle ear implants and auditory brainstem implants in the paediatric population.

Keywords
Implantable devices, Cochlear implants, Middle ear implants, Bone anchored hearing aid, ABI.

Introduction
Hearing loss in children is common. Permanent hearing loss affects approximately 1 per 1,000 live births. Over the last decade there have been great advances in the technology available to rehabilitate hearing. Although conventional hearing aids remain frontline options, there is an expanding pediatric population where conventional hearing aids are either not applicable or provide insufficient gain for adequate communication. This review summarizes the various implants available in clinical practice as an alternative to conventional hearing aids.

Cochlear Implants
Cochlear implants are an established intervention to restore hearing and speech perception in children with profound to severe sensorineural hearing loss. A cochlear implant consists of an external component which contains the microphone and a sound processor. The internal component consists of a receiver stimulator package and an electrode array. The receiver stimulator receives data from the internal component and transmits the data to the electrode. The electrode is surgically positioned in to the cochlea and directly stimulates the cochlear nerve. Most children can expect to perform well with a cochlear implant with marked improvements in their quality of life.

Selection Criteria
In the UK, NICE have advised that all children with profound to severe hearing loss should have a thorough assessment by a multidisciplinary team prior to being offered a cochlear implant. The recommendations are summarised below:

- Severe to profound deafness is defined as hearing only sounds that are louder than 90 dB HL at frequencies of 2 and 4 kHz without acoustic hearing aids.
- Children should have had a valid trial of an acoustic hearing aid for at least 3 months (unless contraindicated or inappropriate).
- Simultaneous bilateral cochlear implantation is recommended as an option for children with severe to profound deafness who do not receive adequate benefit from acoustic hearing aids.
- Adequate benefit from acoustic hearing aids is defined for children as, speech, language and listening skills appropriate to age, developmental stage and cognitive ability.

A systematic review was recently conducted in an attempt to determine preoperative factors that may contribute towards a favourable outcome following a cochlear implant in children. Several factors that may contribute towards determining outcome have been suggested, however, only 4 of these factors are supported by appropriate evidence to act as predictors of outcome following a cochlear implant in children.

Predictors of outcome:
- Age at implantation
- Hearing loss related to GJB2 gene
- Inner ear malformation
- Meningitis

Other factors that may contribute:
- Duration of implant use
- Mode of communication (e.g. sign language +/- oral)
- Cognitive and information processing skills

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Predictors of outcome

Age at implantation
The age at which children are implanted has a critical impact on outcome. For children with prelingual hearing loss, outcomes tend to be better if children are implanted early (18-24 months) however; children who are implanted later may also get satisfactory outcomes if they have benefited from a hearing aid prior to a cochlear implant. Evidence suggests that if the auditory pathways are not stimulated early, the higher auditory centres may become recruited by the other sensory modalities as part of their own and this process may be irreversible. Hence any future stimulation of the higher auditory regions by a cochlear implant is likely to be associated with poor outcomes. For children with postlingual hearing loss (loss of hearing after acquisition of speech and language), favourable outcomes were associated with earlier implantation although some benefit was evident for children who received their implant several years after the hearing loss.

GJB2
Defects in the GJB2 gene are responsible for one the most prevalent causes of congenital hearing loss. The GJB2 gene encodes connexion 26 which is a gap junction protein. Connexin plays a key role in maintaining cochlear hair cell haemostasis and abnormalities of this protein are responsible for sensorineural hearing loss. However children with hearing loss associated with the GJB2 gene perform very well with cochlear implants.

Inner Ear malformations
Outcomes following cochlear implantation are highly positive for children who have milder forms of inner ear malformations such as widened vestibular aqueducts or mild mondini’s. Children with more severe inner ear malformations such as severe Mondini’s malformation can also expect to do well however the progress is often much slower.

Meningitis
One potential sequelae of meningitis is sensorineural hearing loss which may occur within a few days or even several years later. The pathogenesis of hearing loss following meningitis is related to obliteration of the cochlea and is summarised in to 3 stages: acute, fibrous and ossification. The acute stage is characterised by infiltration of the perilymphatic space with purulent material. In the fibrous stage, there is proliferation of fibroblasts and angiogenesis within the perilymphatic space. The final stage of ossification involves bone formation typically first seen in the basal turn of the cochlea.

One of the proposed mechanisms responsible for initial involvement of the basal turn of the cochlea is thought to be due to spread of infection from the meninges to the cochlear aqueduct which opens in to the basal turn close to the round window.

It is vital to fast track these patients for cochlear implant surgery to maximise the chances of achieving full implant electrode insertion in to the cochlea. Children who receive implants early often achieve good outcomes.

Unilateral Vs Bilateral
It is now widely accepted that children with a unilateral cochlear implant who met the criteria for implantation develop improved speech and language acquisition compared with no hearing device or using a hearing aid. In addition the implanted children acquire marked improvements in their social, emotional and academic development. However children with unilateral implants have difficulty with speech perception in noisy environments. Recent literature supports increased advantages of bilateral cochlear implants compared to a unilateral cochlear implant. Children with bilateral implants have superior hearing and speech perception in both quiet and noisy environments compared with unilateral implant recipient. In addition the bilateral implant group demonstrate improved sound localisation by eliminating the head shadow effect.

Simultaneous vs Sequential
There has been an increasing trend towards bilateral cochlear implants in the paediatric population. This has stemmed from the perceived benefits from bilateral implants and the approval by NICE. Simultaneous implantation involves inserting a cochlear implant in both cochleae under the same anaesthetic. Sequential implantation describes insertion of a cochlear implant in one cochlea followed by a delay before implanting the opposite cochlea. Most authors agree that outcomes following bilateral cochlear implants in children are greater for children who receive simultaneous implants or where the time interval between the 1st and sequential implant is short i.e. 24 months or less.

Recent work by Papsin suggests that children who have more than a 2 year gap between their sequential implant have poorer speech perception and are not as accurate in localising sound compared with children who receive their sequential implant within 12 months. These assessments were performed at 12 months after the sequential implant hence, longer term follow up will determine whether the advantage of a short gap between implants is sustained in the long run.

Furthermore outcome following sequential implantation is associated with a more favourable outcome if the 2nd implant is received before the age of 6. Speech perception and quality of life scores resulting from the sequential implant were much lower for children who received their 2nd implant after their 6th birthday.

In the short term these children demonstrate improved auditory perception, language skills and electrophysiological responses as measured using auditory brainstem responses compared with children who receive their sequential implant much later. Whether this improved language is maintained in the long term is yet to be determined. There is now increasing evidence to suggest that children who receive a sequential implant several years after their 1st implant may not benefit from the 2nd implant and may even suffer an adverse outcome.

The United Kingdom National Paediatric Bilateral Audit is currently analysing data on children with bilateral cochlear implants who have been implanted simultaneously or sequentially. The results of this data should hopefully clarify some of the issues discussed earlier.
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Post Implant Complications
The main group of complications may be broadly grouped in to infections and device failure.

Infection

Wound/implant infection
The incidence of wound infection post-surgery has been reported to be between 1-12%. Although the vast majority of these infections are successfully treated with systemic antibiotics, in children there is a higher incidence of wound dehiscence or skin flap necrosis resulting in extrusion of the implant.12 The American Academy of Paediatrics advise broad spectrum antimicrobial therapy with activity against methicillin-susceptible and methicillin-resistant Staphylococcus aureus for all suspected cases of wound or implant infection.13

Acute Otitis Media
Clinicians are now increasingly faced with children who have cochlear implants and present with acute otitis media (AOM).12 These children require urgent systemic antibiotics. Meningitis is a potential complication following acute otitis media in children with implants. Oral antibiotics may be considered in the presence of a clinically mild infection. For children who fail to respond to intravenous antibiotics, the AAP advise a myringotomy and culture of the middle ear fluid +/- insertion of a ventilation tube although this is not common practice in the UK.

Meningitis
Meningitis is a potentially catastrophic complication following a cochlear implant. The reported incidence is between 16 – 30 times higher in implanted children.13 The predominant organism responsible for meningitis in children with cochlear implants is Streptococcal Pneumoniae.13 In most cases of implanted children who present with meningitis, AOM is often the preceding cause.12 Bacteria can infiltrate the inner ear and subsequently the CSF by entering the cochleostomy. Other risk factors for meningitis include children with inner ear malformations and children who have a cochlear implant associated with a positioner (wedge which stabilises the implant electrode in the most optimum position for the implant to work). Cochlear implants with positioners are not used in clinical practice now. Currently there are no studies comparing meningitis rates in children with congenital hearing loss and inner ear malformations to a similar group of children who in addition have cochlear implants. Hence, in children with inner ear malformations we have yet to determine whether the higher rates of meningitis are related to the inner ear malformation or the cochlear implant. AAP recommend prevention of meningitis by ensuring children are fully immunised against Streptococcal Pneumoniae and Haemophilus Influenzae type B. Both the vaccines for Streptococcal pneumonia and Haemophilus influenza are part of the routine immunisations schedule offered to all children in the UK.

Device Failure
Device failure is one of the primary reasons for revision surgery.6 Other than technical failure of the implant hardware, head trauma is a significant cause of device failure or device extrusion in up to 42% of implanted children who are diagnosed with a malfunctioning implant.14 Although outcome following revision surgery following device failure is favourable, a small proportion of children may experience a significant deterioration in their performance compared with their first implant.14 Revision rates for device failure or other causes vary between 11-13%.6,14 There is a small risk of a permanent facial palsy of 1% or less with the procedure.6,4

Long term Outcomes
It is now apparent that children with cochlear implants demonstrate continued improvements in their speech performance with time. 10 year follow up studies have revealed that although some children plateau at about 5 years, there is a significant proportion that are able to progress to conversing over the telephone during 5-10 years of using their implant. Interestingly the vast majority of these children where either working or were in full time education.6

Recent Advances
There has been a recent trend towards hearing preservation surgery. These children have normal or mild hearing losses in the low frequencies and profound/severe hearing loss in the high frequency range. This pattern of hearing loss is challenging to manage with a hearing aid alone.

Several authors have reported the preservation of low frequency hearing following cochlear implant surgery in the ipsilateral ear. Hearing preservation rates vary between 45-90%.15 Children with cochlear implants who also retain natural residual hearing in the implanted ear reveal enhanced speech perception compared to children with cochlear implants and no preserved hearing.15 In addition the hearing preservation group demonstrate better appreciation for pitch and are better at discriminating voice. It is possible to achieve hearing preservation in children by meticulous surgical techniques and appropriately designed implant electrodes.16

In cases where hearing preservation surgery is unsuccessful or the hearing loss occurs as a result of other aetiology, low frequency hearing may be established by fully activating the implant electrode.

Bone anchored hearing aids
The bone anchored hearing aid (BAHA) is now widely accepted as an appropriate intervention for children with bilateral conductive hearing loss. The BAHA consists of 3 parts: a titanium fixture, a percutaneous abutment and a detachable sound processor. The percutaneous abutment acts as a vibrational transducer and is able to transmit sound to the cochlea by bone conduction through the titanium fixture which is implanted in to the temporal bone. This trans-osseous conduction of sound to the cochlea is much more effective than trans-cutaneous transmission of sound provided by the conventional bone conduction hearing aids. In addition the BAHA system offers better gain at high frequency and less distortion of sound compared with the conventional bone conductor hearing aid.1 Furthermore the BAHA does not require any adjustments for fluctuating conductive hearing loss.1

Clinical Criteria
The clinical indications may be summarised in Figure 1.
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BAHA/BAHA soft band

Insertion of the titanium fixture in to the temporal bone requires the bone thickness to be at least 3mm for the fixture to remain stable. The desired skull thickness is often only achieved in children aged 3-4 years. Hence, most centres avoid inserting Bahas in children younger than 3. The alternative option for the younger child is a Baha soft band. The Baha soft band consists of a Baha sound processor that clips onto a soft elastic headband. The auditory thresholds obtained with a Baha soft band are between 25-30dBHL. Hence the Baha soft band is appropriate for adequate speech and language development in very early childhood. However it is advisable to convert to a bone anchored system as soon as possible as speech and language scores with a baha soft band are inferior to those obtained using a bone anchored system from ages 3-6 years and beyond.

Quality of Life

There is a marked improvement in the quality of life scores for children with bilateral conductive hearing loss who receive a baha. Furthermore, it has been reported that most paediatric centres report a compliance rate up to 96%. Although the baha is an option for unilateral conductive or sensorineural hearing loss, the early use of baha for young children in this group is controversial. Bilateral baha is an option for children with bilateral conductive hearing loss and unlike cochlear implant surgery, a prolonged gap between the sequential baha’s is not associated with an adverse outcome.

Middle Ear Implants

Middle ear implants (MEI) are now an additional option for rehabilitating hearing in children. MEI’s may be considered in children who have moderate to severe sensorineural hearing loss or mixed/conductive hearing losses where hearing aids have been unsatisfactory. There are 2 main types of MEI’s: piezoelectric or electromagnetic which are summarised in Table 1. Both implant types essentially rely on the conversion of sound in to electrical energy which then causes vibration of the middle ear structures using either piezoelectric crystals or an internal magnet. These vibrations are then transmitted to the cochlea.

Table 1

<table>
<thead>
<tr>
<th>MEI</th>
<th>Piezoelectric</th>
<th>Electromagnetic</th>
<th>Totally Implantable</th>
</tr>
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<tbody>
<tr>
<td>Vibrant Soundbridge</td>
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<td>No</td>
</tr>
<tr>
<td>Soundtec</td>
<td>no</td>
<td>Yes</td>
<td>No</td>
</tr>
<tr>
<td>Otologics middle ear transducer</td>
<td>no</td>
<td>Yes</td>
<td>No</td>
</tr>
<tr>
<td>Otologics carina</td>
<td>no</td>
<td>Yes</td>
<td>Yes</td>
</tr>
<tr>
<td>Esteem Envoy</td>
<td>yes</td>
<td>No</td>
<td>Yes</td>
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</tbody>
</table>
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The most commonly used device in clinical practice that is also licensed for children is the Vibrant Sound Bridge middle ear implant. The VSBS consists of an external audio–processor, a receiver package and a floating mass transducer (FMT) which contains the magnet. These implants operate by converting sound into electrical energy picked up from the microphone in the audio–processor. This electric current sets up a magnetic field around the FMT causing it to vibrate. The FMT is attached to either the incus or stapes and if these are not available then the FMT may be anchored to the round or oval window.

Audiological Criteria
To benefit from a MEI for sensorineural hearing loss, the auditory thresholds should not exceed 65–70dBHL and speech discrimination scores should be above 50%. A cochlear implant would be a more suitable alternative in cases where speech discrimination scores are lower.

MEI’s are also suitable for mixed and conductive hearing losses and there is significant overlap between the auditory thresholds for MEI and Baha.

MEI vs Conventional hearing aids
MEI’s have primarily been used in adults with a few case reports in children in the UK. All papers published to date only contain a handful of patients with short follow ups; hence; it is very difficult to draw definitive conclusions about the benefits of MEI’s. MEI’s are however an appropriate option in subjects who are conventional hearing aid candidates but are unable to use the hearing aid as a result of ear mould allergies, discomfort, occlusion effect and recurrent infections. A recent systematic review suggested that MEI’s are comparable to conventional hearing aids for speech perception in quiet. However, speech perception in noise and the sound quality produced by the MEI are possibly superior to conventional hearing aids.

Complications
The majority of the reported complications are small and mostly include injury to the chorda tympani nerve, tinnitus, headache, infection and otalgia. In general there was no change in residual hearing following MEI surgery except in a minority of cases and a revision rate of 15% has been quoted in 1 study. Some authors have commented on the reduced mobility of the stapes when the FMT is attached to it however, this does not seem to effect the hearing outcome.

Auditory Brainstem Implants
The final implantable hearing aid which deserves a brief mention is the Auditory Brainstem Implant (ABI). The American Academy of Audiology has outlined the 2 main groups in addition to children with NF2 who may be considered for an ABI. These include children with prelingual hearing loss and children with postlingual hearing loss (Table 1).

Table 1.

<table>
<thead>
<tr>
<th>Prelingual hearing loss causes</th>
<th>Postlingual hearing loss causes</th>
</tr>
</thead>
<tbody>
<tr>
<td>Inner ear malformation</td>
<td>Cochlea ossification from Meningitis</td>
</tr>
<tr>
<td>Cochlear nerve hypoplasia/aplasia</td>
<td>Trauma to cochlear nerve</td>
</tr>
<tr>
<td>Cochlea ossification from Meningitis</td>
<td>Cochlear destruction from otosclerosis</td>
</tr>
<tr>
<td>Unmanageable facial stimulation from cochlear implant</td>
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</table>

The ABI device is similar to a cochlear implant except that instead of placing the electrode in to the cochlea, it is positioned across the cochlear nucleus in the brainstem. Generally, the outcomes following an ABI are much less favourable and more unpredictable compared with those following a cochlear implant. However a recent report suggests that a small number of children who have had an ABI can achieve good speech performance that is comparable to age matched children with cochlear implants. Longer term studies are necessary to determine long term outcomes and factors that may have a role in selecting appropriate candidates.

At present auditory neuropathy is a contraindication for an ABI. Authors of the consensus document on ABIs in children recommend 12 months as the minimal age for inserting an ABI.

Conclusion
A range of devices are now available to improve hearing in children when conventional hearing aids are not suitable. With advancing technology and surgical techniques, the outcomes are improving and the criteria for patient selection is expanding.

Hearing preservation surgery offers the prospect of improved pitch discrimination for children in whom the residual low frequency hearing has been preserved. The preserved hearing contributes towards improved music and voice discrimination and possible improved speech perception for tonal languages.

Most implantable devices that are available in clinical practice are semi implantable. The future goal would be to provide a totally implantable device that excels in hearing also allows children to hear while participating in water activities. Any child with hearing loss who is unable to tolerate hearing aids should be referred to specialist centres for consideration of other hearing rehabilitation options. The child should be managed in a multidisciplinary setting where there are expertise in assessing the child and offering the appropriate implant. It is vital that the child and family are fully informed of all the available options and are appropriately counselled prior to proceeding with implant surgery. This will provide the child and family with realistic expectations and minimise potential misunderstandings.

Conflict of Interest
All authors have no conflict of interest to declare. No extraneous funding was obtained.
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References