Preschool Hearing Screening

- Only Important Heads
- Screening is a one time process whereas surveillance is an ongoing process.

**RISK FACTORS**

**Congenital permanent bilateral hearing loss**
- An illness or condition requiring admission of 48 hours or greater to a NICU.
- Stigmata or other findings associated with a syndrome known to include a sensorineural and/or conductive hearing loss.
- Family history of permanent childhood sensorineural hearing loss.
- Craniofacial anomalies, including those with morphological abnormalities of the pinna and ear canal.
- In utero infection such as cytomegalovirus, herpes, toxoplasmosis or rubella.

**Acquired and late-onset permanent bilateral hearing loss**
- Trauma and Post Meningitic

**Unilateral permanent hearing loss**
- Rare

**Auditory neuropathy/dys-synchrony**
- Outer hair cells normal and thus Normal OAE but abnormal BERA. Fault may be in inner hair cell, Spiral ganglion, 8th nerve etc.
- NICU patients admitted more than 48 hrs may have this.

**Temporary childhood hearing loss**
- OME
- Point prevalence of OME is 20% and period prevalence of 80%.

**NEWBORN HEARING SCREENING**

**Current screening tests available**
- The tests used for newborn hearing screening are currently one or more of:
  - Automated otoacoustic emissions (AOAE);
  - Transient evoked otoacoustic emissions (TEOAE);
  - Distortion product otoacoustic emissions (DPOAE);
  - Automated auditory brainstem response (AABR or BERA).
- TEOAE reflect the activity in the outer hair cells of the cochlea, while AABR waveforms are affected by cochlear and neural lesions.
It is important to note that a newborn hearing screening programme protocol that regards a clear OAE result as a screen pass will miss neonates with auditory neuropathy, since the latter will manifest with OAE present, but ABR absent.

**Cut-off level and case definition**

- > 40 dB average hearing loss is considered significant

**'At-risk' screening**

- Babies admitted to a neonatal intensive care unit for more than 48 hours were 10.2 times more likely to have a permanent hearing loss.
- 14 fold increased prevalence if Family History of hearing loss present.
- Craniofacial Anomalies.
- Syndromic Association
- Congenital infections like toxoplasma, rubella, CMV, herpes.

**WHY SURVEILLANCE**

- (1) Newborn hearing screens will not find all those with preschool permanent hearing loss, because of late onset and progressive hearing loss
- (2) Any screening programme will miss an irreducible minimum of true cases
- (3) Surveillance is a more justifiable approach to identification of children with persistent OME than a screening programme.

**Hearing tests In children**

**ELECTROPHYSIOLOGICAL TESTING**

**Key developmental age: 0-6 months, up to adult if appropriate**

- EPT includes OAE i.e. AOAE, TEOAE, DPOAE and also BERA (or CERA or AABE or ABER.)
- They can also prove of value in children of any age, when behavioural testing has failed to produce reliable results, in particular, those with severe learning or communication difficulties.
- Electrophysiological techniques are also used to confirm hearing thresholds, for example in children with profound hearing loss, prior to fitting of high power hearing aids or cochlear implantation or where nonorganic factors are suspected.

**BEHAVIOURAL OBSERVATION AUDIOMETRY (BOA)**

- Key developmental age: 0-6 months
- In behavioural observation audiometry (BOA), changes in activity are observed in response to a sound stimulus.
- Response behaviours in infants up to four months might include eye widening, eye blink (auropalpebral reflex), arousal from sleep, startle or shudder of the body or definite movement of the arms, legs or body.
From four to seven months, lateral inclination of the head towards the sound or a listening attitude or stilling may be observed. The test is usually performed with the child cradled in the parent's lap.

The sound stimulus is presented for < 2 seconds, in a horizontal plane, 15 cm from the child's ear, out of peripheral vision.

Reliability as a diagnostic test is obviously a concern.

**THE DISTRACTION TEST**

**Key developmental age: 6-18 months**

- The test is based on the principle that the normal response observed when sound is presented to a baby is a head turn to locate the source of sound.
- Habituation is increasingly likely to occur after 12 months, although the technique may prove to be useful in older children with learning or communication difficulties where other methods have been unsuccessful.

**TEST METHOD**

- The test should be performed in a tidy, uncluttered, suitably sized room (recommended minimum >16 m²) with ambient noise levels <30 dB.
- The child sits on the parent's knee, facing forward and erect, lightly supported around the waist.
- The distractor directs the attention of the child to a simple activity usually performed on a low table. Suitable examples include spinning a brightly coloured object, using finger puppets or gently pushing a miniature car.
- The sound stimulus is presented by the second tester, half a second after the item is covered. The distractor observes the child's response.
- Stimulus should continue up to 10 seconds if there is not an immediate response.
- The stimulus
  - A wide range of sound stimuli can be used to elicit a response, including voice, musical toys, 'everyday sounds', narrow band noise and warble tones.
  - Initially, a sound stimulus is presented which is anticipated to be likely to be suprathreshold, for example at 70 dB(A) in a child with probable normal hearing. A sound is then presented at the anticipated minimal response threshold, for example, 30 dB(A).

**No sound control trials**

- Some children exhibit checking or searching behaviour or simply turn towards their parents for reassurance during the test.
- Appropriate to introduce control trials in which the test is performed in the usual manner by both testers, but no sound is generated. If the child appears to make a response, then it must be assessed how this compares to the responses observed to sound trials.
Response Recording

- The response threshold is regarded as the quietest level at which two out of three clear responses were recorded for each particular sound stimulus.

VISUAL REINFORCEMENT AUDIOMETRY

Key developmental age: 6-36 months

- VRA generates more auditory evoked head turns than an unrewarded sound stimulus and delays habituation so that the test was more likely to be completed.

The test arrangement

- For sound field testing, the speakers may be placed at 45, 60 or 90° from the child and at the same height as the child's head at a distance of at least 1 m from the ear.
- Visual reinforcers are generally placed adjacent to or above each speaker. The reinforcer acts as a reward and therefore increasing the attractiveness or appeal to the child.

PERFORMANCE TESTING

Key developmental age: 2-5 years

- The performance test was first described by Ewing and Ewing as a transitional technique suitable for children from 2.5 years and in some cases younger, until cooperation with pure tone audiometry can be achieved.
- It follows the simple principle that the child is conditioned to wait for a sound and then to respond with a play activity.
- The child should be seated on a low chair adjacent to the parent in an uncluttered room with low levels of ambient noise. A toy is placed on a low table in front of the child. Toys which involve a simple repetitive activity are most suitable, such as the classic 'men in a boat', balls on sticks, knocking down skittles or pegs in a board. The conditioning sequence starts with the tester engaging the child's attention by holding the response item, e.g. the wooden man, poised waiting in front of the child. After a few seconds, a suprathreshold sound stimulus is presented and the tester responds by an appropriate activity, e.g. placing the man in the boat. This sequence is repeated several times often supported by gestures such as a stop sign using the palm of the hand and a cupped hand to the ear to indicate listening. This has the advantage of avoiding dependence on spoken language. The child is
then offered the response item and guided to wait and perform the task as shown. Vocal praise should be used to reinforce a correct response.

- The number of repetitions required to successfully condition the child will depend on their age, developmental status, willingness to co-operate and in particular their ability to inhibit the response until the signal is detected.
- Examples might include knocking the man off the table, flicking a small ball into a goal, or even poking the tongue out at the tester.

**PURE TONE AUDIOMETRY**

**Key developmental age: 3 years onwards**

- The recommended procedures for pure tone audiometry based on the Hugson and Westlake descending/ascending technique using 10/5 dB steps should be flexibility adapted.

**The co-operative test (18 to 39 months)**

- Different simple instructions, e.g. having been handed a small toy, asked to 'give it to Mummy' or 'give it to teddy' or 'give it to baby'.

**Toy discrimination tests (30 months onwards)**

- This employs seven pairs of similar sounding nouns such as /cup/ and /duck/ or /tree/ and /key/, each represented by a small easily recognizable toy, placed on a table in front of the child in a quiet room. Using live voice, the child is asked to 'show me the ... spoon', etc.

**Consonant discrimination tests**

**LINGUISTIC AGE: OVER 6 YEARS**
Investigation and management of the deaf child

SYNDROMIC HEARING LOSS IN

- Usher syndrome ➔ SNHL + retinitis pigmentosa
- Warderbergs Syndrome ➔ White forelock heterochromia iris broad nasal bridge, hypertelorism mod - profound SNHL
- Brachio oto renal syndrome ➔ Branchial cyst preauricular pit, tag or microtia mild to profound con/ mixed/ SNHL absent / Hypoplastic kidney
- Pendred Syndrome ➔ congenital B/L SNHL goitre ± Hypothyroidism.
- Treacher Collins syndrome ➔ shunken middle face, prominant nose micrognathia cleft palate down slanting eyes lower eyelid coloboma microtia, CHL.

- Sensorineural hearing loss as a result of mumps is mostly unilateral, although bilateral loss has been described.

INVESTIGATIONS

- It is recommended that core investigations are offered to the parents of all children with newly diagnosed bilateral sensorineural hearing loss and thresholds over 70 dB in the better ear averaged across 500, 1000, 2000 and 4000 Hz.
- Pediatric History (Intrapartum and postpartum history of infections, drugs etc), Family History, Clinical Examination, Audiology, Imaging (MRI inner ear and CT of temporal bones) ECG

Causes of hearing impairment

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<td>X-linked 1%</td>
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<td>Measles</td>
<td>AIDS</td>
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| Ototoxic drugs | Trauma | Neoplastic disease |

Suggested additional investigations

- Haematological and biochemical tests
- Serological test for congenital rubella and cytomegalovirus infections
- Thyroid tests
- Immunology tests
- Metabolic screen blood and urinalysis
- Renal ultrasound
- Chromosomal analysis
- Urine for microscopic haematuria, Connexin 26 and 30 mutations assay, Ophthalmic assessment, Referral to clinical geneticist, Vestibular investigations.
- Mitochondrial inheritance only via mother who may or may not have symptoms

**HEARING AIDS**

- **Behind the ear (BTE)** air conduction hearing aids with a soft mould, replaced at regular intervals as the child grows, are most commonly used.
- Older children, where the ear canal volume is greater, may be able to use in the ear (ITE) hearing aids.
- **BAHA** While waiting for the skull to mature sufficiently to enable fitting of a bone anchored hearing aid (BAHA ®), a Softband consisting of a microphone mounted on an elastic headband placed around the head may be used.
- Children with bilateral hearing loss should be fitted with bilateral aids unless there is a definite medical contraindication, e.g. infected ears.
- **Cochlear implantation** has transformed the rehabilitation of severe and profoundly deaf children in healthcare systems where it is available.
- Communication choices used are
  2. Those approaches using speech and sign (total or simultaneous communication).
  3. Sign bilingualism.

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**Paediatric cochlear implantation**

- Hair cell loss is the principal cause of sensorineural deafness. Sufficient neural elements usually survive and are available for electrical stimulation enabling meaningful activation of the auditory cortex. A cochlear implant is a prosthetic device which replaces the transducer function of damaged hair cells and provide this electrical stimulation.
- Almost all children with severe to profound sensorineural hearing loss may now be considered for implantation.

**DEVICES**

- **External Device** = Microphone + Microprocessor + Transmitter
- **Internal Device** = Receiver + Electrode Array
- The microphone is connected to a speech processor, which digitally processes and then encodes the auditory signal. The signal is then transmitted transcutaneously to the implanted receiver/stimulator, which stimulates the surviving neural elements in the modiolus. This enables selective stimulation of the relevant segments of the surviving ganglion cell population.
TIMING OF IMPLANTATION

Prelingual deafness

- Depending on the time of onset, deafness can be prelingual or post-lingual, i.e. occurring before or after the development of normal speech and language.
- The existence of a critical period for language development during the first five years of life is well established.
- Early implantation, probably before the age of two and possibly significantly earlier, is required to maximize the potential for normal language development in prelingually deaf children.

EARLY IMPLANTATION

- The National Institutes of Health (NIH) advocated implantation in children after the age of two years.
- Now done < 2yrs of age.
- Implantation below the age of one year usually requires special circumstances, such as rapid obliteration of the cochlea following meningitis.

POST-LINGUAL CANDIDATES

- Post-lingual deafness in children is most frequently a consequence of meningitis or head injury. Although the duration of deafness is a factor in speech perception outcome after cochlear implantation.

ASSESSMENT

AUDIOLOGICAL TESTS

- BERA, ECoG, Behavioural testing (Imp. As for full hearing pathway).
- Post lingual – PTA, Speech discrimination tests important.
- Vestibular testing is not routinely performed on children.

IMAGING

- High-resolution computed tomography (CT) scanning shows the bony morphology of the cochlea and labyrinth.
- Congenital abnormalities include the Mondini dysplasia, common cavity deformity or a large vestibular aqueduct.
- The internal auditory meatus may be narrow and raise the possibility of auditory nerve aplasia.
- In meningitis ➔ Ossification is usually found in the basal turn of the cochlea near the round window.
- MRI scans may demonstrate the presence of an auditory nerve and better to detect cochlear ossification also.

PREOPERATIVE COUNSELLING
This will include explanation of the benefits and risks of surgery, variability of outcome and the requirement for intensive and prolonged input from the implant centre team and local professionals.

**SURGERY**

**TECHNIQUE**

The surgery involves several steps.

**INCISION AND FLAP DESIGN**

Some version of what resembles a standard post-aural mastoidectomy incision.

**BONY APPROACH TO COCHLEA.**

A standard mastoidectomy is followed by a posterior tympanotomy preserving the chorda tympani. The round window region is visualized through a posterior tympanotomy. The scala tympani is entered via a cochleostomy anterior to the round window niche. Following electrode insertion, any space between the electrode and the cochleostomy can be sealed with soft tissue, usually fascia or muscle.

**BONY RECESS FOR AND FIXATION OF RECEIVER/STIMULATOR.**

A device-specific contoured bony recess is fashioned for the receiver/stimulator package. In children this almost always involves exposure of the dura.

Most surgeons prefer to preserve a bony island over the dura if possible. Fixation of the device can be with either absorbable or nonabsorbable sutures. Bone cement is also used for electrode fixation at the mastoid rim or posterior tympanotomy.

**CLOSURE AND POSTOPERATIVE CARE.**

Drains are rarely required.

Few patients – antibiotic coverage.

Many patients have some form of imaging of the electrode array (most frequently via a modified Stenvers view) to confirm satisfactory position prior to discharge.

**COMPLICATIONS**

Facial nerve injury due to non auditory stimulation by electrodes – mostly post meningitic patients.

Dural Damage

CSF leaks

Meningitis

Device Extrusion

Wound Infections

Device Failures

**SPECIAL SURGICAL CIRCUMSTANCES**
COCHLEAR ANATOMICAL ABNORMALITIES

Implantation of the malformed cochlea is technically feasible.

Patients with an enlarged vestibular aqueduct or enlarged vestibule may have 'gushers' intraoperatively.

OBLITERATED COCHLEA

Ossification develops as a consequence of meningitis, temporal bone trauma and some autoimmune disorders.

The basal turn of the scala tympani is most frequently affected due to the connection between the subarachnoid space with the scala tympani via the cochlear aqueduct. Scala vestibuli ossification is less common and may take place later in the ossification process.

Ossification was initially thought to be a contraindication to cochlear implantation, but various surgical procedures have been developed to allow at least partial implantation in those cochleas with significant degrees of ossification.

Scala vestibuli insertion was an acceptable alternative to scala tympani insertion in partially obliterated cases.

A dual/split array is available; one electrode is inserted into an inferior tunnel drilled along the basal turn and the other electrode is inserted into a separate cochleostomy drilled into the superior limb of the basal turn.

CHRONIC SUPPURATIVE OTITIS MEDIA AND OPEN MASTOID CAVITIES

Patients with mastoid cavities were initially considered unsuitable for implantation. However, increasing experience has led most centers to offer surgery to these patients.

One or Two staged procedures.

In the two stage procedure, all squamous epithelium is removed from the cavity, which is then obliterated behind a blind sac closure of the external canal. After an interval, the cochlear implant device is inserted at a second-stage operation.

POSTOPERATIVE HABILITATION

This is a multidisciplinary effort involving audiologists, speech and language therapist and specialist educators.

It begins with programming the device with stimulation parameters for the electrode array which will provide auditory perception across the speech range.

CONTROVERSIES IN MANAGEMENT

Implantation in older children and adolescents

Adults demonstrated improvement in mean scores for word and sentence recognition, although the improvement was not as significant as in children implanted at a younger age.

BILATERAL COCHLEAR IMPLANTATION
In the last few years, experience has grown with bilateral implantation and studies in adults report the benefits binaural hearing should provide, including sound localization and enhanced speech recognition in background noise.

**MRI FOLLOWING COCHLEAR IMPLANTATION**

The devices have a removable magnet; a small incision over the posterior half of the receiver/stimulator package will allow the implanted magnet to be removed to enable MRI to be performed.

A recent study shows that it is possible to perform an MRI scan provided the scanner is equipped with a 1-Tesla magnet.

**Congenital middle ear abnormalities In children**

- A variety of nonossicular congenital middle ear abnormalities may also be associated with ossicular deformities.
- They include:
  - Persistent stapedial artery.
  - Anomalous course of the facial nerve
  - Congenital perilymphatic fistula
  - High jugular bulb
  - Aberrant internal carotid artery.

**DEFINITION OF CONGENITAL OSSICULAR ABNORMALITIES**

Congenital ossicular fixation and defect is defined as a malformation affecting the ossicular chain, present at birth, which leads to a dysfunction of the ossicular mechanism due to immobility or discontinuity of the ossicular chain.

**INVESTIGATION OF CONGENITAL OSSICULAR ABNORMALITIES**

**AUDIOMETRY**

There is an average threshold of approximately 50 dB, producing a flat air conduction...
line, with no low frequency bias as with otitis media.

- There is an average air-bone gap of 35 dB
- Tympanometry (As type curve) usually demonstrates a normal middle ear pressure with reduced compliance due to fixation of the ossicular chain.

**IMAGING**

- High resolution computed tomography (CT) scanning remains the primary imaging modality though imaging (MRI) studies may demonstrate associated labyrinthine and internal auditory meatal abnormalities.
- CT virtual endoscopy may offer a further mode of presenting the images for preoperative surgical planning.

**EXPLORATORY SURGERY**

- The diagnosis may only be made during a tympanotomy, though an interesting alternative to lifting the tympanic membrane is to attempt to visualize the ossicles via Eustachian tube and middle ear endoscopy.

**PRINCIPLES OF MANAGEMENT**

**In the presence of a bilateral moderate hearing loss**

- Air Conduction Hearing Aid
- BAHA
- Unilateral cases are less well defined in terms of best management. In the presence of ipsilateral tinnitus, amplification may act as a tinnitus masker.
- A hearing aid may improve the patient's hearing performance in background noise and optimize sound localization.
- When the diagnosis has been made in childhood, consideration for surgery should be preceded by an adequate trial of amplification. Whatever middle ear surgery is contemplated, but particularly with stapedotomy, there is a significantly higher risk of delayed sensorineural hearing loss due to sporadic episodes of acute otitis media in children under ten years of age.

**MANAGEMENT OF SPECIFIC CONGENITAL OSSICULAR ABNORMALITIES**

**Isolated stapes ankylosis**

- Crus being absent (monocrural) or there being no recognizable crura but instead a single strut (monopodial).
- Congenital stapes ankylosis at the level of the footplate was first described by Shambaugh.
- He emphasized the clinical contrast to otosclerosis, in particular that the margins of the congenitally fixed footplate and the annular ligament are difficult to visualize since the footplate bone blends into the bone of the surrounding otic capsule.
SURGERY FOR CONGENITAL STAPES FOOTPLATE FIXATION

- Preoperative scanning may demonstrate labyrinthine dysplasia
- In particular, a dilated fundus of the internal auditory meatus (IAM) should be sought.
- The dilation of the IAM is associated with a defect allowing communication of cerebrospinal fluid (CSF) with the labyrinth
- The surgical technique is similar to otosclerotic stapes ankylosis. A common feature of congenital fixation is the presence of thick anterior and posterior crura. To reduce the risk of inner ear damage these may be vapourized with a KTP laser.

SURGERY FOR STAPES SUPRASTRUCTURE FIXATIONS

- Removal of the bony bar can be achieved using a CO2, argon, KTP or erbium laser or with a micro drill.

CONGENITAL APLASIA OR SEVERE DYSPLASIA OF THE OVAL AND ROUND WINDOWS

- Auditory rehabilitation with hearing aids or a BAHA may be most appropriate.
- A neo-oval window operation, in which a de novo entrance into the labyrinth is either drilled on the promontorial or even on the rostral side of the Fallopian canal, has a high risk of inner ear damage.

NONOSSICULAR CONGENITAL MIDDLE EAR ABNORMALITIES

PERSISTENT STAPEDIAL ARTERY

- The stapes forms around the stapedial artery, leading to formation of the obturator foramen.
- By ten weeks of development the artery atrophies, leaving a patent foramen underneath the arch of the stapes.
- If persistent, it arises from the petrous internal carotid artery (ICA) traverses Jacobsen's canal for a short segment exits at the promontory passes through the stapes obturator foramen and enters the Fallopian canal close to the cochleariform process.
- The persistent stapedial artery (PSA) passes anteriorly, exiting the canal at the geniculate ganglion and passes into the extradural space of the middle cranial fossa, where it gives rise to the middle meningeal artery.
- In the presence of a PSA, the foramen spinosum is usually absent and the ICA may have an aberrant course due to its collateral formation secondary to a segmental agenesis of the ICA.
- Asymptomatic or the cause of pulsatile tinnitus and has no relationship to conductive hearing loss.
- Traditional teaching has been that the presence of the artery is an absolute contraindication to stapedectomy.

ANOMALOUS COURSE OF THE FACIAL NERVE

- An abnormal course is particularly common with microtia or with dysplasia of the oval and round windows.
- Rohrt and Lorentzen classified facial nerve displacement in the middle ear into four groups:
  1. Facial nerve partially obliterates the stapes footplate
  2. Bifurcation of the facial nerve
3. Facial nerve rests on the footplate with deformed stapes or oval window
4. Facial nerve rests on the promontory

CONGENITAL PERILYMPHATIC FISTULA

- It may be associated with:
  - micro fissures around the oval and round windows
  - labyrinthine or IAM dysplasia.

HIGH JUGULAR BULB

- The jugular bulb can be defined as 'high' if it reaches the level of the inferior bony annulus and is often covered by thin bone or is dehiscent.
- There may be a more direct association with a conductive hearing loss due to interference with the ossicles, contact with the tympanic membrane and obstruction of the round window niche.
- Major surgery to occlude or re-route an HJB is unlikely to be justified by symptoms alone.

ABERRANT INTERNAL CAROTID ARTERY

- This may be associated with other vascular abnormalities such as a PSA and likewise present as a vascular middle ear mass.
- Associated symptoms include pulsatile tinnitus, which may be objective, and hearing loss.
- In approximately 20 percent of cases it is bilateral
- Brisk bleeding, hemiparesis, aphasia, deafness, Horner syndrome and intractable vertigo may result if the vessel is unintentionally injured.