

## THE SEVERE NERVE DEAFNESS CHILD: ASSESSMENT AND MANAGEMENT

The Problem of the Deaf Child:



The Deaf Child: Unresponsive and in need of early hearing care.

In communities with neonatal hearing screening testing in place, many congenitally severely deaf children are identified at birth. A smaller group are missed by this screening, or develop deafness subsequently, the loss being noticed by the family, albeit sometimes with considerable delay. Identification of the cause may require substantial investigation.

Early identification of the profoundly deaf child is essential to ensure normal development of the auditory cortex. This section of the brain depends on auditory stimulation during the first two years of life for optimal function, as this stimulus promotes the synaptic complexity of the structure, and hence the brain's hearing capacity. The effects of delayed stimulus may be overcome by early intervention, but the longer this is delayed, the less likely a full recovery, and the longer the time to achieve this.

Delayed intervention beyond 2-3 years risks the development of cross-modal plasticity effects. The surrounding sensory cortical structures tend to gradually re-align the auditory neurones to alternative tasks, leading in the extreme case where an under-developed cortex produces aberrant sensations, e.g. tactile, if stimulated by sound impulses.

Therefore, identification and management of the severely congenitally deaf infant is a matter of some urgency. For the profoundly deaf child at birth, bilateral simultaneous cochlear implantation (CI) at six months, using least traumatic surgery, is highly desirable, and in the usual case will deliver excellent hearing and speech outcomes at an early stage.

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However, in some cases, other adverse factors may intrude. Ear disease, neurological, general medical, psychological, family, or social aspects may be present and impede a successful outcome. It is the role of the CI team to anticipate, identify and minimise these threats, preferably well before surgery.

### Neonatal Hearing Screening:

Universal neonatal screening for deafness in developed communities has been a major advance for identification of congenital deafness. Testing uses automated auditory brainstem response (ABR) techniques, perhaps combined with other objective methods (otoacoustic emissions [OAEs], tympanometry, reflex testing). Children not achieving a “pass” are referred for further testing and observation.

Suspect cases are then referred to an otologist for evaluation.

### Medical Evaluation:

Assessment of the infant’s medical status may begin with the otologist, or in other cases with a neonatal paediatrician, especially if other conditions are evident at birth (prematurity, jaundice, syndromal changes, etc.). Assessments will include the otological status, general medical aspects, plus other medical specialties (e.g. neurology, genetics) as required. The otologist will liaise with the audiology staff for further testing and will arrange a hearing aid trial. Early intervention hearing and speech development is organised for ongoing therapy.

Should the loss prove partial, aiding and auditory-verbal or alternative communication therapy is appropriate, but if the losses are severe or progressive, the family is counselled re the possible need for CI at an early stage.

### CI Evaluation:

Approaching the end of the first six months, the child’s responses to the aids and testing give a clearer picture of ability, and the likely need for CI surgery. A CT and MRI scan are arranged, if not previously undertaken. Sedation or general anaesthesia may be needed. CT scans evaluate the labyrinthine and other temporal bone structures, the MRI the inner ear, cranial nerves and central nervous system (CNS).

Developmental paediatric, psychological and family assessments are finalised and a CI team meeting assesses the case with a view to prognosis and pre-operative family counselling. Establishing realistic family expectations and achieving outcomes at those levels is the key to successful paediatric CI surgery.

(at Queensland Otology, Dr Jane Black is a leading expert in this field).

Pre-operative surgical counselling emphasises these aspects plus any surgical risk factors.

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## Prognostication:

In the absence of evident drawbacks, current paediatric cochlear implantation (PCI) techniques should succeed in restoring excellent communication, unless unexpected surgical or subsequent events intervene. When perceived adverse factors are present, the likely outcome of PCI is gauged via an evaluation of their threats to the process. Prognostication follows clear lines.

Individual threats are initially identified by their site and action on the auditory pathway. They can be classified as primary (auditory cortex or CNS damage), secondary (failure to transmit CI stimulus through the cochlea or VIII n), or tertiary (interruption of CI activation). The adverse factors are then assessed as to their effect on a domain of ability: cortical ability, neurological, otological, general medical, psychological, and family/social. The individual domain effect is derived from the worst threat, or the combined effect of multiple factors.

The prognosis is then gained by judging the combined domain overview, the prognosis being derived from a single worst affected domain or the cumulative effect of multiple domains' problems.

Performed thoroughly and professionally, the pre-operative evaluation should allow a full and accurate picture of the situation for the child's family. Whilst unforeseen surgical or subsequent events may intrude, an expert team should achieve a successful outcome in the great majority of cases.

## Surgical management:

Some causes of deafness are correctable back to the normal hearing situation by surgery. This is dealt with in other articles. Nerve deafness, however, requires either aiding, where possible, or implantation if the aiding is ineffective/inappropriate.

Severe or profound sensorineural deafness is managed, where indicated, by cochlear implantation. This is performed via small incisions on the rear of the ear. Bilateral implantation is possible at six months age, as many of these cases can be undertaken in little over an hour in skilled hands. The child remains in hospital overnight, but a head bandage is retained for a week whilst the implant stabilises in position. Switch-on takes place 10-14 days after surgery.

## Summary:

The deaf child is a major developmental concern that is best managed by awareness, thorough evaluation, and expert management. Other factors not intervening, excellent speech and hearing should be the normal achievement.

## More information:

[Paediatric Cochlear Implantation](#)

[Cochlear Implants](#)

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