

Cochlear Implantation in Congenital Cochlear Abnormalities

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Summary

Many children have benefited from cochlear implant device including those with congenital malformation of the inner ear. The results reported in children with malformed cochlea are very encouraging. We describe 2 cases of Mondini's malformation with severe sensorineural hearing loss. Cochlear implantation was performed and both of them underwent post-implantation speech rehabilitation. Post-implantation, both of them were noted to respond to external sound. But the second case developed facial twitching a few months after the device was switched on. It is important to evaluate the severity of the inner ear deformity and the other associated anomalies in pre-implantation radiological assessment in order to identify the problem that may complicate the surgery and subsequent patient management.

Key Words: Cochlear implant, Mondini's dysplasia, Radiological assessment

Case 1

A 3 year old Malay girl, was referred to ORL Clinic at the National University Hospital Malaysia with bilateral profound sensorineural hearing loss, with delayed speech development. Otherwise, she had normal developmental milestone.

On examination both external auditory canals were normal with intact tympanic membranes. There were no obvious stigmata to suggest any known syndromic features. There were no cardiologic, urologic or haematologic anomalies identified.

Auditory brainstem response and free field distraction test demonstrated bilateral profound sensorineural hearing loss. The child was fitted with hearing aid for almost 6 months but there was no improvement in her hearing. She was further evaluated for possible cochlear implantation where a computed tomographic (CT) scan of the temporal bone was performed.

Magnetic resonance imaging (MRI) T2-weighted in axial and coronal views (Fig. 1) revealed absence of right cochlear and semicircular canal. This is suggestive of Michel's deformity with small caliber of the IAM and presence of single nerve within. The left ear revealed large featureless cochlear with absence of interscalar septum which is suggestive of Mondini's dysplasia, there was good caliber of left IAM with the presence of double nerve within it.

After adequate discussion with the parents on the benefit and limitation of cochlear implantation in children with cochlear abnormalities, they agreed for the implantation in the left ear. She underwent a left cochlear implantation on 3rd May 2001. The cochlea was assessed via transmastoid facial recess approach. The lips of the round window niche were identified and cochleostomy performed. The electrodes array were inserted without difficulty. There was no perilymph leak or gusher. The fenestration was covered with subcutaneous fat. The wound was closed

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routinely and mastoid dressing applied for 3 days. The postoperative X-ray showed all the electrode arrays were within the cochlea.

Switch on and mapping of cochlear implant were started 1 month later. She was seen weekly by the audiologist and speech therapist for rehabilitation. Five months post-implantation the parents noted that their child started to respond to loud environmental sound.

Case 2

A 2 year old Punjabi girl, was diagnosed to have profound sensorineural loss at the age of 1 year. There were no history of head or ear trauma or chronic ear discharge. She was a full-term baby delivered via spontaneous vaginal delivery, with birth weight of 3.2 kg.

Examination revealed an active child who did not have any syndromic facial features. Both the external ear canals and the tympanic membranes were normal. Auditory brainstem response demonstrated a profound sensorineural hearing loss. Otherwise the child had normal developmental milestone. Cochlear implant was suggested to the parents as a method of hearing rehabilitation after hearing aid trial did not show any benefit. A high resolution CT scan and MRI (Fig. 3) of the temporal bone was performed. It revealed common cavity of both cochlea. However, there was presence of vestibulocochlear nerve and of normal caliber. After exhaustive discussion on the suitability of the child for cochlear implantation, both parents and

the cochlear team agreed to proceed with cochlear implantation.

Cochlear implantation was performed in October 1999. The surgery was performed under prophylactic antibiotic coverage. A standard cortical mastoidectomy with posterior tympanotomy was performed to assess the middle ear. Intra-operatively it was noted that there was absence of round window and promontory. Cochleostomy was initially attempted at the area anterior to the stapes. However, the bone on that side was solid. Intraoperative review of the CT scan suggested an area of common cavity superoposterior to the mesotympanum. A bulge was found near the attic region. A fenestration was successfully made over the bulge, which was the common cavity. There was no evidence of perilymph gush. The electrode was inserted as deep as possible into the common cavity. The cochleostomy was packed with fat.

The child made an unremarkable recovery. Rehabilitation was started 1 month post-surgery. The progress was much slower than expected. Although the child was responding to loud sound, there was no speech development even after 1 year of rehabilitation. One year 6 months after surgery the child developed facial twitching involving the upper eyelid when switching on the implant. CT scan of the temporal bone revealed that the tip of the electrode was adjacent to the fundal part of the internal auditory meatus. The audiologist switch off 2 corresponding distal electrodes which ceased the twitching. The child continued to use the cochlear implant despite making slow progress on rehabilitation.

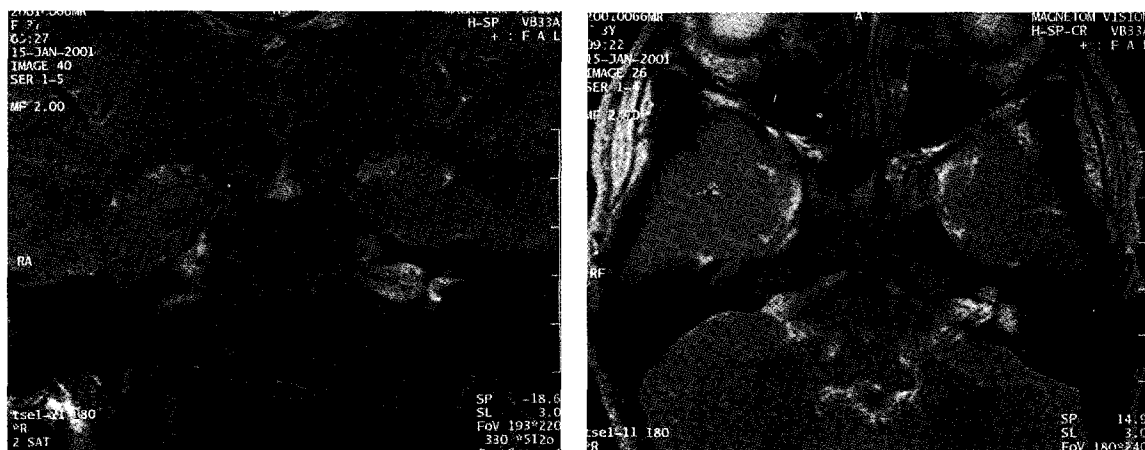


Fig. 1: MRI T2-weighted in coronal and axial views

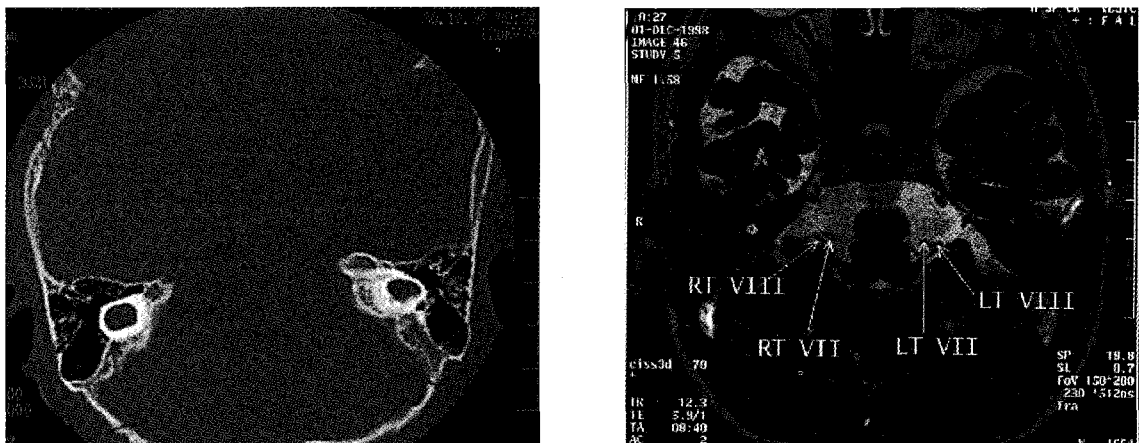


Fig. 2: On the right showing a common cavity of both cochlea on the CT scan, on the left the MRI showing the presence of both vestibulocochlear and facial nerve bilaterally.

Discussion

Patient with cochlear malformation can benefit from cochlear implantation¹. However, the various degree of deformities may pose problems during surgery. Jackler et al² have proposed a classification system for the congenitally malformed inner ear based on the theory that a variety of the anomalies result from arrested development at different stages of embryogenesis.

Incomplete partition of the inner ear is the most commonly encountered making up 55% of the cases². This is the mildest malformation that have been described by Mondini. In this deformity, the cochlea possess only 1½ turns and has incomplete partitioning by a deficient interscalar septum and osseous spiral lamina. This result in confluency of the apical and middle turns of the cochlea. The left ear in Case 1 was a Mondini's deformity.

The second most common abnormality described is common cavity which made up 26% of the study by Jackler et al². In this deformity, there is confluency of the cochlea and vestibule into a common rudimentary cavity that usually lacks an internal architecture. Case 2 has common cavity dysplasia in both ears.

Complete labyrinthine aplasia, also known as Michel deformity, is the rarest among the inner ear malformations. This abnormality would be an absolute contraindication for cochlear implantation on the affected side. Case 1 had Michel deformity on the right side.

In general, inner ear deformities are no longer an absolute contraindication to cochlear implantation. However, the more severe the deformity the more challenging the surgery². The postoperative results are also less predictable. Therefore, detailed preoperative evaluation is important in order to get a good surgical result without any postoperative complication.

High resolution CT scanning with thin section in both axial and coronal planes should be performed in pre-implantation candidate. This will provide the diagnosis of the congenital malformation, the extent of the deformity and also other associated anomalies. Furthermore, the landmarks for performing mastoidectomy and facial nerve may be abnormal or absent in patients with cochlear abnormality. These findings may help the otologist to anticipate any surgical problem during the implantation and also help in assessing which ear would be easier for implantation.

Apart from CT scan, patients with a deformed inner ear or narrow internal auditory canal should undergo MRI. MRI would be able to identify the non-osseous partitioning of the malformed cochlea. It can show the soft tissue densities in the cochlear coils and show presence of cochlear fluid to rule out any luminal obstruction. It is also vital to identify the neural structures contained within the internal auditory meatus. Narrow (1 to 2 mm) canal may indicate an absence of the cochlear nerve. The failure to identify the cochlear nerve by high resolution MRI would represent an absolute contraindication for cochlear implantation¹.

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The most important structure from a surgical point of view is the facial nerve. In congenitally deaf children Balkany et al³ have stated that there is greater risk for the facial nerve to follow an aberrant course within the temporal bone. Preoperative imaging can provide valuable information in identifying the position of any possible aberrant facial nerve that may be associated with cochlear malformations. The abnormalities of the facial nerve depend at which stage it is being arrested during the embryologic life. If it is arrested early in life, before the development of labyrinth, then it usually lies anterior to the primitive otic sac. If the development is arrested after the cochlea has formed, the tract is found at its usual situation above and lateral to the cochlea. The most common abnormality recorded at surgery is the facial nerve being exposed and it may overhang in the tympanic cavity⁴.

In congenital cochlear anomalies, it is also important to observe for a deficiency of bone between the lateral end of IAM and the basal turn of the cochlea. If there is deficiency, the electrode array may be introduced into the IAM and may cause facial nerve stimulation. This complication was seen in the second case presented above. She presented with this problem a few months after the implantation. This could be due to migration of the electrode into the fundal part of the IAM. However, this complication was easily overcome by eliminating the specific electrodes.

An abnormal connection between the perilymphatic space and the subarachnoid space may exist in the malformed cochlea and this may present as a spontaneous fistula into the middle ear or a gusher of the cerebrospinal fluid (CSF) during fenestration of the inner ear. This abnormality may also cause the patient to develop meningitis after the implantation. Page et al⁵ reported a case of delayed CSF otorrhoea with resulting meningitis after cochlear implantation in a child with a Mondini malformation. They advised a great and meticulous care to seal the cochleostomy in children with inner ear malformation to prevent this complication.

In the first reported case, she showed a quite good results of rehabilitation, within 3 months post-implantation she was responding to the external environmental sound and started to develop speech. Whereas in the second case, despite undergoing rehabilitation for the past 1 year and responding to loud environmental sound, her progress was very slow and speech development was very poor.

A child with profound hearing deafness secondary to congenital inner anomalies is not totally contraindicated for cochlear implantation. But preoperative evaluation with CT scanning and MRI are very important in order to anticipate any abnormality which have been described previously because the surgery is more challenging. The results of rehabilitation may be satisfactory. The most important factor is to provide realistic expectation for the parents.

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