

# INTERNATIONAL MEDICAL REVIEW ON DOWN'S SYNDROME

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## CASE REPORT

# Down's syndrome with abnormal inner ear: Is it suitable for cochlear implantation?

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### KEYWORDS

Down's syndrome;  
Hearing loss;  
Cochlear implant;  
Large vestibular  
aqueduct syndrome

### Abstract

Hearing loss is a common problem in Down's syndrome (DS). The majority of this population, up to 80% are suffering from a conductive type hearing loss, whereas estimating 4-20% are due to sensorineural hearing loss. Over the years, the treatment of profound sensorineural hearing loss has been changed since the introduction of cochlear implants. We report a case of a 4 years and 5 months old child with DS and low Intelligence Quotient that had been referred to our centre for cochlear implants. In view of late referral and multiple additional handicaps, with addition of having Large Vestibular Aqueduct Syndrome (LVAS), bilateral incomplete partition of cochlear Type II and abnormal periventricular white matter, she had been rejected for cochlear implantation.

### PALABRAS CLAVE

Síndrome de Down;  
Hipoacusia;  
Implante coclear;  
Síndrome del  
acuoducto vestibular  
dilatado

### Síndrome de Down con oído interno anómalo: ¿es apto para un implante coclear?

### Resumen

La hipoacusia es un problema frecuente en el síndrome de Down (SD). La mayoría de esta población, hasta un 80% sufre hipoacusia conductiva, mientras que el 4-20% según las estimaciones, corresponde a hipoacusia neurosensorial. A lo largo de los años, el tratamiento de la hipoacusia neurosensorial profunda ha cambiado desde la introducción de los implantes cocleares. Presentamos el caso de una niña de 4 años y 5 meses de edad con SD y un bajo cociente intelectual, que fue remitida a nuestro centro para ser sometida a implantes cocleares. En vista de la derivación tardía y las múltiples discapacidades adicionales, además de la presencia de síndrome del acuoducto vestibular dilatado (SAVD), partición incompleta bilateral coclear de tipo II y sustancia blanca periventricular anómala, no se consideró adecuado el implante coclear.

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## Introduction

Down's syndrome (DS) is the most common genetic disorder, occurring in approximately 1:800 live births. Children with DS have altered head and neck structure that results in increased otologic, upper airway, and sinonasal disease. Between 38% and 78% of peoples with DS have abnormalities of the external, middle and inner ear have been described, which contribute to the hearing loss in these individuals<sup>1</sup>. Out of this, over 80% of the hearing loss is conductive and this is due to otitis media with effusion, therefore amenable to medical and surgical intervention<sup>2</sup>. However, 4 to 20% of hearing loss in this population is due to sensorineural hearing loss<sup>3</sup>. It was initially thought that individuals with additional disabilities and learning disabilities were not suitable candidates for implantation, but with a growing body of knowledge and good results, inclusion criteria are expanding and increasing numbers of such candidates have been implanted. Many of these individuals, especially those implanted at a young age, do remarkably well due to preservation of the spiral ganglion and successful post operative habilitation.

## Clinical presentation

A child with DS and global developmental delay was referred to our centre at 4 years and 5 months of age for audiological assessment as a potential candidate for cochlear implant. The child was diagnosed to have prelingual bilateral hearing loss at 4 months of age and bilateral middle ear effusion. Auditory brainstem recordings confirmed a profound sensorineural hearing loss on the right and moderate to severe hearing loss on the left ear when she was 5 months of age. However, the myringotomy with ventilation tube insertion was performed only at the age of 1 year 4 months old and postoperatively, she has been fitted with hearing aid binaurally. However the usage of hearing aid was inconsistent until 4 years old. At the age of 2 years, serial of re-programming and optimization of hearing aid were performed, however the result of aided response evaluation

showed the hearing aid was under amplification. She had a trial of consistent hearing aid for about 5 months, however there was no benefit. She was then referred for consideration of cochlear implant.

High Resolution Computed Tomography (HRCT) imaging of the temporal bone performed at 4 years and 9 months of age revealed a large vestibular aqueducts bilaterally (fig. 1A) and bilateral incomplete partition of cochlear Type II (fig. 2). There was also fluid within mastoid air cells, both middle ears and both epitympanic spaces.

The magnetic resonance imaging (MRI) demonstrated an enlarged endolymphatic sacs bilaterally (fig. 1B) with normal 7<sup>th</sup> and 8<sup>th</sup> nerves, internal auditory canal, vestibules and semicircular canal. There are multiple dilated periventricular region in both temporal and parietal lobes likely represents incomplete myelination and steep straight sinus with absent sagittal suture (not well demonstrated in MRI) suggestive of brachicephaly (fig. 3).

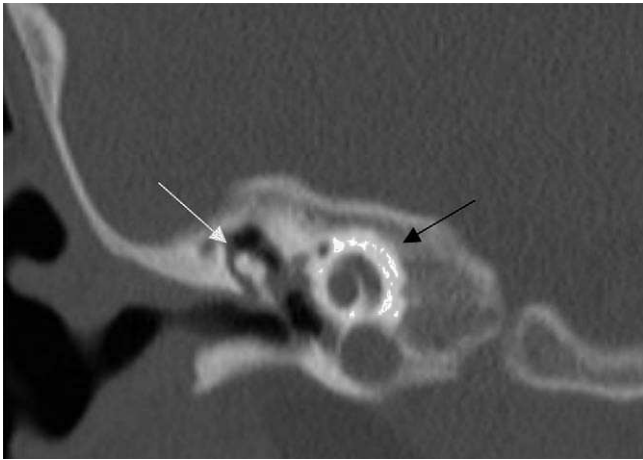
## Discussion

Cochlear implantation is the treatment frequency of severe to profound sensorineural hearing loss. More candidates had been implanted at younger age with good capacity to develop language at a rate equal to that of their hearing peers. Previously due to limited studies on outcomes for this implantation procedure, the candidacy criteria were very strict. However, recent serial studies have shown good outcome from this invasive procedure, and the indications for implantation have gradually been revised<sup>4</sup>. Now there are more implant devices licensed for use in children as young as 12 months and in additional and learning disabilities<sup>4</sup>. The children with additional disabilities can potentially broaden their communication skills, and make progress, though possibly at a slower pace than children without additional disabilities.

Patient with DS and hearing loss posed a major challenge to the successful use of hearing aids and other rehabilitative devices including cochlear implants. They can have multiple additional handicaps, including learning and communication dif-



**Figure 1** Axial high resolution computed tomography temporal bone (A) showing dilated vestibular aqueduct on both sides (arrows) and axial magnetic resonance imaging T2WI (B) demonstrating enlarged endolymphatic sac bilaterally (arrows).



**Figure 2** Coronal high resolution computed tomography temporal bone showing the fusion of the apical and the middle turn of the cochlea (black arrow) and incudomalleolar complex is normal (white arrow).



**Figure 3** Sagittal T1WI showing steep straight sinus (arrow) with absent sagittal suture (not well in demonstrated magnetic resonance imaging) suggestive of brachicephaly.

ficulties. This group of children have been shown to have an effect on subsequent language development and performance post-implantation, with outcomes below those of implanted children without additional disabilities. A recent survey of The Cochlear Implant Programmes in DS by the British Cochlear Group (BCIG) in 2010<sup>4</sup>, four with DS children have received implants. They reported that all children remain implant users 12 months to 4 years post-implantation with a significant improvement seen as early as from 9 months post-implantation in terms of communication and behavioural outcomes<sup>4</sup>.

This case reported a DS's child with global developmental delay and pre-lingual congenital sensorineural hearing loss. She initially had inconsistent use of her hearing aid. By 20 months, the equipment was being worn more consistently with optimal fitted hearing aid. However the child was not having benefit from the hearing aid, therefore she was referred for cochlear implantation.

Several factors were identified which cochlear implant was not an appropriate intervention for this child. She had late referred to our centre for cochlear implant (at 4 years

5 months old), in which ideal age for referral as early as 3 months old. Susan Willey et al. in 2009<sup>5</sup> reported that the possible factors of delayed in referral were multi-disciplinary process when deciding whether a child should be referred for an implant, such as degree of hearing loss, marital status of parents, type of insurance, and living in area where income is below the average. In addition to that, an audiologist's ability to determine possible audiological candidates for referral and managing otolaryngologist who focused on otology were more likely to be referred early compared to children managed by an otolaryngologist who had a wider range of interests<sup>5</sup>.

The other concern about the child is having abnormal inner ear structures and abnormal brain parenchyma. She was rejected for cochlear implantation as HRCT showed bilateral incomplete partition of cochlear Type II. She also has other otological abnormalities, which is LVAS. She is at risk of perilymph gusher intra-operatively and at risk of meningitis post-operatively. However, few reports of several studies have showed benefit with speech recognition to varying degrees from implantation in patients with LVAS and can be offered as an eventual treatment for hearing loss in these patients<sup>6</sup>. In addition, Asma et al. in recent series in 2010<sup>7</sup>, had advocate this group of child should be implanted earlier after discussing pros and cons with parents as they found out duration of profound hearing loss and residual hearing appear to be critical factors in determining implants success.

Furthermore, she was suffering from otitis media with effusion (OME), this raise the issue in the candidacy of cochlear implant. Schwartz & Schwartz in 1978<sup>8</sup> in their study of 38 children's (mean age, 3.1 years) with DS, reported that more than 60% of the series demonstrated otoscopic and acoustic impedance evidence of middle ear effusion. It is postulated that the OME is secondary to atypical head and neck anatomy, including macroglossia, hypoplastic nasal bones, oropharynx and nasopharynx that are narrower volume. In addition, Eustachian tubes are smaller in diameter and at a less acute angle to the hard palate.

There was concern that implantation in the situation of the otitis media prone ear would lead to increased rates of complications, particularly the risk of infection spreading from the middle ear intracranially through the channel created by the cochlear implant. However, Hans et al. and BCIG in their survey in 2010<sup>4</sup> reported all their patients had OME, no intra-operative or post-operative surgical complications were encountered.

Apart from otological abnormalities, she also has global developmental delay and her MRI showed multiple dilated periventricular region in both temporal and parietal lobes likely represents incomplete myelination and features suggestive of brachicephaly. With all the reasons discussed earlier, she has very limited benefit of cochlear implantation, and University Kebangsaan Malaysia cochlear implant committee decided to reject her from the programme. She will learn later for sign language.

As conclusion, DS babies with hearing loss should be encourage to have consistent audiological followed up and having hearing aid intervention. We would encourage clinicians caring for these children and their families to consider referral for assessment by a Cochlear Implant Programme as early as 6 months of age.

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