Incomplete partition type III, computed tomography features and cochlear implantation complications

Raya Salim Al-Busaidi¹, Salwa Jaffar Habib¹*, Ammar Mohsin Al-Lawati², Khalid M.W. Tahhan¹ and Yousuf Ali Al-Saidi¹

¹Department of Radiology, Al Nahdha Hospital, Muscat, Oman
²Department of Ear, Nose and throat, Al Nahdha Hospital, Muscat, Oman.

Received: 3 November 2019
Accepted: 3 May 2020
*Corresponding author: umnufnuf@yahoo.com
DOI 10.5001/omj.2021.34

Abstract
Cochlear incomplete partition type III anomaly (IP-III), also referred as X-linked deafness type 2 or X-linked stapes gusher syndrome, is a rare congenital cause of severe progressive mixed hearing loss affecting mainly male patients. It has characteristic radiological features, consisting of, a corkscrew appearance of the cochlea due to dilatation of the internal auditory canal (IAC) and the direct communication between the IAC and the cochlea secondary to the absent bony plate between the two cavities (lamina cribrosa). Preoperative diagnosis of IP-III facilitates the surgeon in avoiding unnecessary stapes surgery and possible complications of cochlear implantation, such as perilymph gushing during cochleostomy and misplacement of the electrode into the internal auditory meatus, which can lead to further hearing loss. Furthermore, the preoperative HRCT images facilitate the surgeon in sizing and selecting of the most appropriate implant type for the procedure.

Key words: Incomplete partition type-III, X-linked deafness, stapes gusher syndrome, inner ear anomaly, corkscrew cochlea, internal auditory canal (IAC), Cochlear implantation, internal auditory meatus (IAM).
Introduction

In this paper, we review a male child who presented with severe bilateral hearing loss. Preoperative HRCT evaluation facilitated the initial diagnosis of the disease, which revealed typical findings of cochlear IP-III, surgical planning and cochlear implant selection, in order to avoid possible complications. The child under-went cochlear implantation, which resulted in gushing and misplacement of the electrodes into the IAC as post-operative complications. Post-operative imaging was used to determine the position of the implant and to assess the complications. The child’s post-operative x-ray revealed misplacement of the cochlear implant (figure 1), the extent of which was further assessed by a HRCT scan for preplanning the revision surgery and electrode selection. Following the revision surgery, a further HRCT scan confirmed proper implantation and ruled out any further complications.

![Figure 1: post-operative x-ray demonstrating the misplaced cochlear implant electrode in the right IAM (arrow)](image)

Case report

A 10 months old male infant presented for the first time with his parents expressing concerns about his poor response to sound stimuli, by not turning towards soft or loud sound sources. The child’s past medical and surgical history was none contributory with an uneventful antenatal period followed by a normal full-term delivery. As reported by the mother, he underwent an otoacoustic emissions (OAE) test at time of birth and passed. No family history of hearing loss was reported. Physical examination revealed profound sensorineural hearing loss bilaterally. The Auditory Brainstem response (ABR) test showed profound hearing loss at 85 decibel above normal adult hearing level (dBnHL) on the left and 90 dBnHL on the right.
Radiological imaging was performed to assess inner ear structures and to determine the selection criteria for surgical intervention.

Temporal bone High resolution CT (HRCT) was performed with a slice thickness of 0.8 mm. This revealed, typical bilateral bony abnormalities affecting the inner ear structures but sparing the middle and external ear structures bilaterally. These abnormalities included, symmetrical corkscrew appearance of the cochlea and absent bony separation between the basal turn of the cochlea and the IAC; lamina cribrosa (figure 2, b). Both the IAC were bulbous in appearance with diameters of 7 mm on the left and 6.8 mm on the right (figure 2, a) normal diameter ranges from 2-8 mm with an average of 4 mm\(^1\). In addition, poorly formed modiolus and interscalar septa were noted. The facial nerve canal and vestibular aqueduct were prominent but were within normal limits\(^{1,2}\). The semicircular canals were preserved. These CT features are considered characteristic of IP-III.

The child was initiated on hearing aids, as per his parent’s preference, although cochlear implantation was advised. No improvement was noted following 18 months of regular follow ups and speech therapy after which the family consented to proceed with cochlear implantation.
A perimodiolar electrode was selected following discussion with the otology committee, accounting for the cochlear malformation and the short area of the basal turn, to achieve full electrode insertion. Cochlear implantation was performed on the right side at the age of three years.

During surgery (figure 4), the expected perilymph/CSF gushing occurred on exposing the round window, which gradually stopped after five minutes. The electrode of the implant was inserted and then sealed with periosteum to prevent fluid leak.

Postoperative audiological tests showed some positive result in hearing; impedance and field telemetry (IFT) was detected, neural response telemetry (NRT) was satisfactory, speech recognition threshold (SRT) was seen in two channels.

However, abnormalities were identified on post-operative imaging. The X-ray revealed an abnormal straight course of the electrode (figure 1) and a HRCT scan confirmed the mispositioning of the electrode, the tip of which was reaching and hooking medially around the internal auditory meatus (figure 5).
The patient was taken back to surgery within 48 hours. The mispositioned cochlear implant was removed and a new slim straight cochlear implant was re-implanted with great difficulty due to significant bleeding and perilymph / CSF fluid leak. Complete insertion could not be achieved; only 10 out of 24 channels could be inserted. The electrode position was confirmed by HRCT scanning immediately after surgery while the patient is still intubated (figure 6) and audiological NRT test demonstrated a satisfactory response.

![Image](image.png)

Figure 4: Posterior tympanotomy, full insertion of the electrode in the basal turn of the right cochlea round window during the first operation.

The post-operative recovery period was uneventful and follow up audiological NRT testing demonstrated a normal response, representing a significant improvement compared to the initial test.

The child is under regular follow up and at five years of age demonstrated normal attenuation of language milestones. No genetic testing was done for him.
Genetic causes account for 50% of hearing loss. 70% of these are nonsyndromic and less than 5% of the nonsyndromic hearing loss are X-linked. So far, five genes have been identified for X-linked nonsyndromic hearing loss³. Cochlear IP-III is the rarest among the X-linked nonsyndromic causes of hearing loss. It is an X-linked recessive disorder caused by mutation of the POU3F4 gene located on chromosome X⁴. It was first described genetically and clinically in 1971 by Nance et al⁴,⁵. It is characterized by bilateral progressive mixed hearing loss, congenital fixation of the stapes, and a perilymph gusher (flow of perilymph/CSF into the middle ear when a stapedectomy or cochleostomy is attempted)⁵

Discussion

Figure 5: a) Axial plane of IAM CT post-surgery showing the cochlear implant electrode passing through the IAM. b) MIP reconstruction demonstrating the full course of the electrode from the subcutaneous devise till the tip of the electrode reaching the intracranial fossa.

Figure 6: Coronal section of right IAM post repositioning of the electrode. Electrode is in place reaching the cochlear cavity.
Inner ear anomalies have been reported in 20% of patients with congenital hearing loss\textsuperscript{3}. According to the current literature, inner ear malformations are classified into eight distinct groups\textsuperscript{8}. IP-III anomaly represents around 4% of them\textsuperscript{3}. Recognition of these abnormalities can prevent unnecessary surgery which can lead to perilymph gushing and worsening of hearing loss. High resolution computed tomography (HRCT) is the modality of choice for assessing congenital ear malformations with recommended section thickness of 0.4-0.7 mm\textsuperscript{1}. It mainly illustrates the bony structures of the middle and internal ear. MRI is used preoperatively to assess the membranous labyrinth and the presence of normal nerves\textsuperscript{5}.

X-linked anomaly radiological features were first described in early 1970s by polytomography sections which demonstrated widened IAC and absent bony plate separating IAC from the cochlea (lamina cribrosa)\textsuperscript{9,10}. Phlebs et al. was first to report these anomalies on CT scan. He reported dilated bulbous IAC, incomplete separation of the cochlear basal turn and IAC, and dilated intratemporal parts of the facial canal\textsuperscript{6}. In addition to these findings, Talbot and Wilson subsequently described the absence of bony modiolus and abnormal vestibular aqueducts\textsuperscript{11}. Other features have been reported later in some patients including structural abnormalities in the vestibule and the semicircular canals, Small or atretic oval and round window\textsuperscript{12}, thickened stapes footplate and hypertrophic bills bar; a bony landmark that divides the superior compartment of the internal acoustic meatus into an anterior and posterior compartment\textsuperscript{13}.

The presence of bulbous IAC can be a normal variant in some individuals, after excluding a tumor\textsuperscript{6}. However, when it coexists with the absent osseous separation between the lateral end of the IAC and basal turn of the cochlea, the direct communication between the two cavities forms what is called a corkscrew cochlea, a classical sign for cochlear IP-III\textsuperscript{14}. Moreover, this connection causes direct communication between cerebrospinal fluid and perilymph which increases the pressure in the cochlea and leads to gushing when the cochlea is opened during surgery\textsuperscript{12}.

Despite the distorted architecture of the inner ear in these patients, the cochlea’s outer dimensions are normal, making cochlear implantation possible. In fact, it is the only means in which hearing can be improved in these patients. However, surgeons should be careful as these patients are at greater risk to develop complications. In addition to the commonly
known complications of cochlear implantation, such as device failure and facial nerve stimulation, these patients can develop complications attributed to the absent bony septum separating the cochlear cavity from the IAC. The two main complications are CSF gushing into the middle ear during cochleostomy and misplacement of the cochlear implant electrode into the IAC. To prevent misplacement, position monitoring with intraoperative imaging is advised\textsuperscript{15}. Also, careful selection of the electrode type can minimize misplacement in these patients. Meningitis is also another possible complication. Covering the patient with vaccination and firm sealing of the cochleostomy to stop CSF leaks can prevent this risk\textsuperscript{16}.

In conclusion, IP-III is one of the rarest inner ear anomalies. However, radiological features are typical for diagnosis. This should direct surgeons to avoid possible complications during stapes manipulation and cochlear implantation surgeries.

**Conflict of interest:**
The authors declare no conflicts of interest

**References:**
12. Incesulu A · Adapinar B · Kecik C. Cochlear implantation in cases with incomplete partition type III (X-linked anomaly). Eur Arch Otorhinolaryngology 2008; 265:1425-1430