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SURGICAL DILEMMA DURING COCHLEAR IMPLANTATION IN MONDINI DEFORMITY

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ABSTRACT

Enlarged Vestibular Aqueduct (EVA), a component of Mondini deformity, is the most common Inner Ear Malformation (IEM), causing congenital sensorineural hearing loss in children, often bilaterally. Cochlear implantation in a case of Mondini deformity poses a great risk of CSF gusher or ooze intra-operatively, primarily due to the Enlarged Vestibular Aqueduct (EVA). A 7-year-old boy, receiving limited benefits from hearing aid usage, underwent right sided cochlear implantation. High resolution CT scans confirmed the presence of Mondini deformity with EVA. During the surgery, extreme caution was maintained while drilling the round window niche to prevent any inadvertent opening of the membrane. The round window membrane showed frank pulsations after exposure, indicating high CSF pressure. Hence, additional surgical preparation was taken to tackle the leak. In anticipation of a perilymph gusher/ooze, an additional amount of temporalis fascia, periosteum, subcutaneous and muscle tissue was collected and preserved in saline. Tissel glue was also kept on reserve. However, on gently opening the membrane, absolutely no CSF gusher/ooze was encountered throughout the period of insertion. So, this case highlights the surgical dilemma faced during cochlear implantation in such cases with IEM.

KEYWORDS

Enlarged Vestibular Aqueduct (EVA), CSF gusher, Cochlear implantation, Inner Ear Malformation (IEM)

INTRODUCTION:

Cochlear implantation is the treatment of choice for patients with congenital, bilateral profound sensorineural hearing loss (SNHL), receiving minimal benefit from hearing aids. Children with Inner Ear Malformations (IEMs) are prone to progressive hearing loss and are undergoing cochlear implantation increasingly over the past few years. Enlarged Vestibular Aqueduct (EVA), a component of Mondini deformity, is the most common IEM, causing hearing loss in children, often bilaterally.(1) However, operating on such children poses a greater risk of cerebrospinal fluid gusher or ooze, facial nerve injuries, difficulty in localizing the round window or erroneous electrode insertion.(2),(3) Limited studies highlight the surgical experience of implantation in these children. The purpose of this report was to evaluate the surgical dilemma faced during implantation in such a case.

CASE PRESENTATION:

A 7-year-old boy from rural Eastern India, presented with congenital bilateral profound SNHL. The child had no improvement with hearing aids since the age of 2 years. Due to financial constraints, the child could not undergo cochlear implantation earlier. On examination, bilateral tympanic membranes were intact and mobile. Other ENT parameters were within normal limits and development was appropriate for age. Child had average intelligence with Intelligence Quotient (IQ): (90+/-5), Social Quotient (SQ): (85+/-5).

Audiological assessment with Brainstem evoked auditory response (BERA) showed bilateral profound SNHL. Tympanometry showed bilateral 'A' type curves and aided audiometry had poor responses on both the sides with curves below the speech banana.

High resolution CT (HRCT) scan of temporal bones revealed bilateral enlarged vestibular aqueducts with Incomplete Partition-II (IP-II) deformity of cochlea - a Mondini deformity.

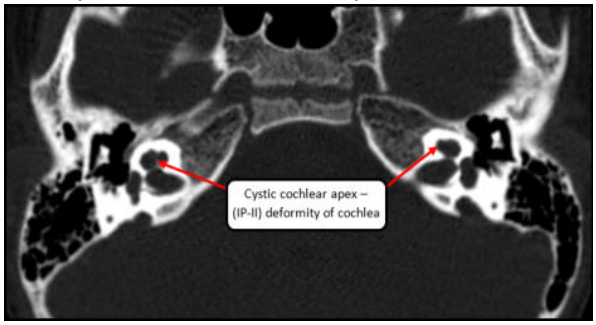

Figure 1: HRCT scan of temporal bones of child showing bilateral IP-II deformity of cochlea

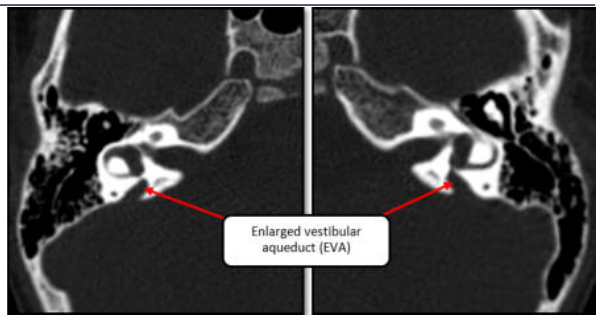

Figure 2: HRCT scan of temporal bones of child showing bilateral Enlarged Vestibular Aqueduct (EVA)

MRI of the temporal region showed a normal vestibulo-cochlear nerve bundle, enlarged vestibules and vestibular aqueducts with a cystic cochlear apex.

On December 2024, cochlear implantation was performed on the right, under general anesthesia by the transmastoid, posterior tympanotomy approach. Advanced Bionics (AB) HiRes Ultra Slim J implant was used. In anticipation of a perilymph gusher/ooze, an additional amount of temporalis fascia, periosteum, subcutaneous and muscle tissue, was collected and preserved in saline. Tissel glue was also kept on reserve. Extra caution was maintained while drilling the operculum around the round window niche, to avoid inadvertent damage and accidental opening of the round window membrane. On exposure of the complete round window membrane, frank pulsations were noticed over the membrane, indicating a high CSF pressure. These pulsations posed a warning of a CSF gusher on opening the membrane. Hence, the surgeon decided to make a soft opening into the cochlea, towards the end of the surgery.

After drilling of the implant bed, the implant was taken for insertion. The round window membrane was carefully and gently opened, on the antero-inferior aspect using a curved needle. However, absolutely no CSF leak/ooze was encountered. The basilar membrane was also seen to be pulsating inside the cochlea. Fortunately, uneventful, and complete insertion of the electrode was done through the round window approach. The opening was sealed with soft tissue collected earlier. Post-operatively, there was no CSF leak or other complications.

Child is currently undergoing auditory-verbal therapy and showing good outcomes with development of hearing and speech. Child is attending school as well and can communicate better with peers and teachers.

DISCUSSION:

Mondini malformation refers to the association of an enlarged vestibular aqueduct (EVA), enlarged vestibule and incomplete partition type-II (IP-II) deformity in cochlea (cystic cochlear apex).(1) During embryogenesis, the vestibular aqueduct begins as a long and narrow vestibular diverticulum. Any defect before the diverticulum starts to narrow, results in an EVA. (4) On HRCT Temporal bones, a vestibular aqueduct is considered to be enlarged when the antero-posterior diameter, at the midpoint of the aqueduct is greater than or equal to 1.5mm.(5)

Cochlear implantation when associated with IEMs, poses a greater risk of peri-operative complications. (6) Perilymph leaks/gushers are the most common complication encountered, with up to 50% incidence in patients with IEMs.(3),(7)

A 2016 study, showed CSF gusher in 14.2% patients, all with either Mondini malformation or EVA.(3) However, Harker et al. reported no gusher during the implantation on 5 children with EVA. Bent et al. reported minimal perilymph pulsating oozing in 50% of cases with EVA.(8) Myamoto et al. experienced CSF leak in 21.7% patients with EVA, but there were no difficulties in inserting electrodes.(4)

Thus, CSF gusher/ooze, though a common complication in cochlear implantation of Mondini deformity, is not always present, even in the presence of pulsating round window/basilar membranes.

CONCLUSION:

CSF leak (gusher/ooze) may be absent intra-operatively during the cochlear implantation of patients with Mondini deformity. If present, it can be managed easily with sealing of the cochleostomy with temporalis fascia/ soft tissue. Thus, necessary caution should be maintained throughout for an uneventful procedure.

Funding: This research received no external funding.

Conflict Of Interest: There are no conflicts of interest related to this research.

Ethical Approval: The study was conducted in compliance with ethical standards. We ensured that all aspects of the research were conducted ethically and with respect for the rights and well-being of the participants.

Informed Consent: Informed consent was obtained from the study participant.

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