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Introduction

Hearing loss is the most common congenital and acquired sensory deficit among children. Sensorineural hearing loss (SNHL) occurs when sound is not efficiently transduced into electric potentials or when transmission of these signals to higher-order auditory centres is disrupted. Small subset of paediatric patients with severe to profound SNHL loss that will not benefit from the Cochlear Implant (CI) due to a small or absent cochlea or auditory nerve or scarring of the inner ear due to infection or trauma, will benefit from Auditory Brainstem Implant surgery (ABI) [1]. The ABI differs from the CI as it bypasses the cochlea and cochlear nerve to directly stimulate second order neurons of the auditory pathways in the brainstem called the cochlear nucleus

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speech and language development, were assessed for etiology of deafness and underwent electrophysiological evaluation for hearing and also radiological evaluation of inner ear (high resolution CT and 3T MRI). Patients with profound retro cochlear SNHL were diagnosed and the parents of the children with retro cochlear pathology, were counselled for ABI surgery, duration, likely complications, post-operative switch on, audio visual therapy for 2 years, expected outcomes, overall cost and available government schemes for surgery. An informed written consent from parents for surgery was taken. All required investigations were done and the child was prepared for surgery a er comprehensive evaluation and opinions from neurosurgeon, paediatrician, ophthalmologist, cardiologist, anaesthesiologist, speech pathologist, clinical psychologist and occupational therapist. Surgical team comprised of ENT surgeon, Neurosurgeon, Neuro-anesthesiologist, Audiologist. Intra-operative anatomical ndings and variants from normal anatomy were noted. Electromyography monitoring of 7 and 9 nerves and intra OP monitoring of evoked auditory brainstem response was done. Subjective grading of cerebellar occulus and electrode insertion di culty for di erent grades of occulus was assessed intra-operatively. e same data was retrieved from previous medical records, of who had undergone ABI surgery, before the study has started.

Surgery is done by retrosigmoid approach (Figure 1). A er, sub periosteal dissection, 3×3 cm craniotomy is done and extended to visualize the transverse sigmoid sinus junction. ABI receiver stimulator coil bed was drilled with tie-down holes (Figure 2). Dura was inccised and re ected to visualize cerebellum. e later was retracted inferio-medially and cerebral spinal uid (CSF) was released. Flocculus was identi ed and graded depending on visibility of choroid plexus and root entry zone of 7^{th},8^{th} nerve and lower cranial nerves. A er arachnoid dissection, foramen of Luschka was identi ed. When in doubt, location of lateral recess was further con rmed by noting for the egress of CSF during valsalva manoeuvre. Dummy electrode was negotiated through the foramen of Luschka to oor of the 4^{th} ventricle to check optimal positioning of electrodes, followed by placing the permanent electrode through the same route, a er xing receiver stimulator coil (Figure 3).

Prior to placement of electrode, its dacron mesh was trimmed in all our pediatric ABI candidates. is was done because of smaller

Intra-operative EABR was measured. EABR in ABI lacks waves I and II waves. Positive auditory responses are multi-peak waveforms generated in rst 2-4 min of onset of stimulus, while larger waves with longer amplitude are generally non-auditory responses. A er optimal placement, electrodes were secured with surgicel. Dura was closed in water tight manner and craniotomy defect covered with gel foam and bone sandwich. Receiver stimulator area was closed with palva ap and skin wound was closed in layers followed by mastoid dressing.

During the surgery, the following anatomical landmarks were very helpful in guiding the electrode towards the cochlear nucleus, which is not under direct vision.

Surgery-anatomical landmarks

1. Choroid plexus marks the entrance of the lateral recess and the taenia traverses the roof of the lateral recess [8].

2. Flocculus of cerebellum which forms an operculum covering the entry to lateral recess of fourth ventricle [9].

3. Exits of cranial nerves 7, 8 and 9 form a triangle which help in identi cation of the entrance of the lateral recess of fourth ventricle (~ 5×6 mm) [10].

4. Straight vein, running from 9th 10th

Results

Total of 24 children underwent ABI from 2006 till March 2017. 12 implantees were male and rest were female. e mean age at implantation was 4.16 years. Out of the 24 implantees, 33% of the cases had bilateral absent cochlear nerve, 25% cases had Michle's aplasia and remaining cases had etiology as shown in Figure 6.

Excessive CSF leak, with no anatomical variant also made positioning of electrode di cult. 2×2 table shows, normal surgical anatomy and variant anatomy cases which had di culty in electrode insertion. Chi square test was applied to Table 1 and the p value was signi cant, stating that anatomical variants encountered during ABI surgery made the pID 339 fsit my and a-5.9 (a)19 (pID 33e)-5 (c)-6.9 (t)-5 (r)13 (o)-9 (de)0.5 (c)-7 (u)-612 (t 12 (t.)0.5 T EMC /S

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