



Case Report

Contralateral non-auditory stimulation in auditory brainstem implantation: A case report

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1. Introduction

Auditory brainstem implants (ABIs) are neuroprosthetic devices that stimulate the cochlear nuclei without any connection between the inner ear and brainstem [1,2]. The first pediatric ABI surgery was performed in 2000 on a prelingually deafened child with common cavity and cochlear nerve aplasia [3]. More than one thousand ABIs have been placed in adults and children around the world since the late 1970s [4].

After ABI surgery, it is possible to encounter non-auditory stimulation due to the stimulation of the cranial nerves (CN), such as the CN V, VII, IX, and X. To monitor the possible non-auditory stimulation, it is important to perform electrical auditory brainstem response (ABR) testing during the surgery to ensure for appropriate electrode placement [1].

In ABI programming, the comfortable electrical stimulation levels should be measured for each electrode, one by one. The most comfortable loudness levels can be adjusted according to the behavioral responses. It is possible to increase the stimulation levels until non-auditory stimulation are seen. The most common non-auditory stimulation types are tactile stimulation, dizziness, facial twitching, dysgeusia, nausea, and shoulder contraction [2,4,5]. Non-auditory stimulation clearly show the activation of the anatomical regions near the cochlear nucleus [6] and can be seen in 42–92.3% of multichannel implant users [7,8]. Most of the non-auditory stimulation were seen on

the ipsilateral side, Nevison et al. mentioned contralateral non auditory stimulation in NF2 patients [7].

Pontocerebellar hypoplasia is an inherited progressive neurodegenerative disorder with fetal onset associated with the hypoplasia/atrophy of cerebellum and pons, and other symptoms such as developmental delay [9]. Although the hearing loss was not commonly seen in pontocerebellar hypoplasia, the first case report with a combination of pontocerebellar hypoplasia and sensorineural hearing loss was presented in 1997 [10].

So far, 119 children with inner ear malformations have received ABIs at Hacettepe University. In this case report, we present a two-year-old female with an ABI who experienced side effects with the stimulation of the ABI on the contralateral side. To our best knowledge, this is the first report of contralateral non-auditory stimulation in a child with an ABI.

2. Case presentation

2.1. Demographic evaluation

A female patient was born at 37 gestational weeks and was delivered spontaneously. Her birth weight was 2600 g, and she was immediately put in an incubator. While she was in the incubator, she had hyperbilirubinemia, which increased to 20 mg/dl. Due to the

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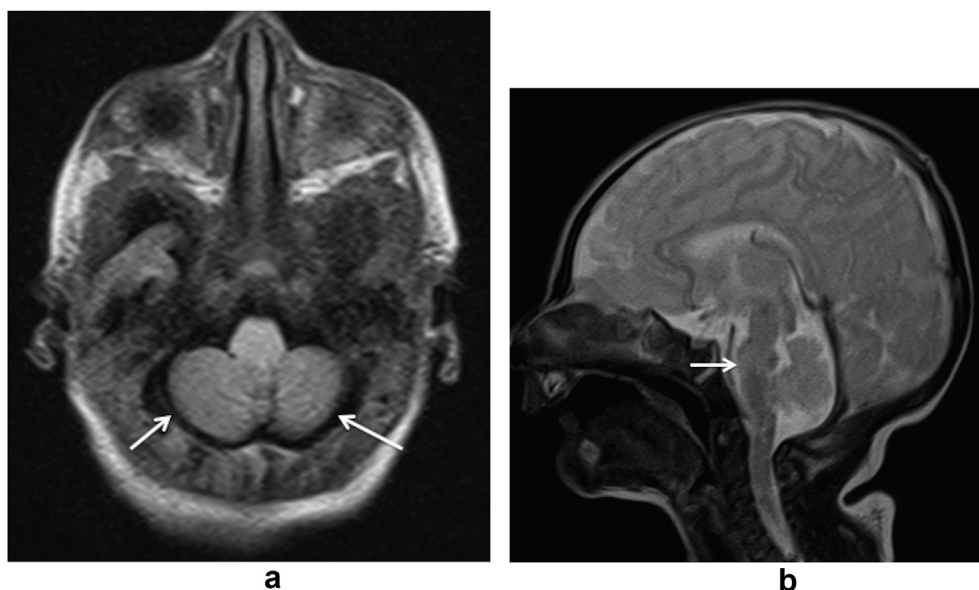


Fig. 1. A. Brain MRI demonstrating small posterior fossa with hypoplasia of cerebellum (arrows). B. On mid-sagittal brain MR images, marked hypoplasia of ventral pons could be seen (arrow).

hyperbilirubinemia, phototherapy was indicated. There was no additional risk (such as high fever, head trauma, or ear infection) in the postnatal period. Her eye examination was normal. She was diagnosed with pontocerebellar hypoplasia based on radiological evaluation. Her posterior fossa was small with hypoplasia of both cerebellar hemispheres and vermis (Fig. 1). Also, she had a small pons with a decreased pontine bulge on mid-sagittal images (hypoplastic ventral pons). ‘Thin pons sign’ is important in establishing the diagnosis [11]. There was no history of surgery due to other health problems. There was no family history of a genetic disorder or hearing loss. The informed consent was obtained from the parents.

2.2. Preoperative audiological and radiological evaluation

She failed the neonatal hearing screening bilaterally. Bilateral profound sensorineural hearing loss was identified at the age of 13 months in another center. She was referred to our clinic for an audiological follow-up. The automated ABR and otoacoustic emissions were negative on both sides in the first audiological evaluation session in March 2017. The threshold ABR was planned. The ABRs (click/27.5/s rate) showed no response at the level of 99 dBnHL on the left ear, whereas wave V was observed at the level of 90 dBnHL on the right side (Fig. 2). The female was also tested with insert earphones to observe behavioral responses. Behavioral testing suggested better responses on the right ear, and no response was observed on the left ear except vibrotactile stimuli at low frequencies in April 2017 (Fig. 3). Bilateral hearing aids were recommended and an auditory rehabilitation program was planned during the follow-up. The radiological assessment showed bilateral narrow internal auditory canals with bilateral cochlear nerve aplasia (Fig. 4). Cochlear apertures were of normal size.

2.3. Preoperative auditory perception evaluation

The auditory perception test was administered by the same audiologist, with a live voice. The language assessment test aimed at determining the verbal communication skills. All the test items were presented in an auditory-verbal condition. In her initial assessment, the female had no behavioral response to the live voice. She could make sounds such as/m/. Her development was evaluated with the Denver Developmental Screening Test. She was delayed in all four areas of development (fine motor, gross motor, social, and language). She

started to hold herself up at eight months old. She could hold her head up when placed in a sitting position at 12 months old. She had difficulty swallowing and had recently developed a sucking reflex.

The Meaningful Auditory Integration Scale (MAIS) which is a parent-reported questionnaire was used to assess listening skills in children with hearing loss [12]. Each item was rated by parents and scored from 0 to 4 (0 = never, 1 = rarely, 2 = occasionally, 3 = frequently, and 4 = always), with a total score ranging between zero and 40. Language skills were assessed with the Test of Early Language Development—3 (TELD-3) [13]. The test was administered in auditory-verbal condition, and it included two subtests: receptive language and expressive language. TELD-3 assesses language development in children between the ages of two and seven years, eleven months. TELD-3's norm values were collected from typically developing Turkish children. Unfortunately, the norms scores for children with hearing impairment are not presented in the test manual. In this reason, we share the age-equivalent comparison of the test results. All auditory perception and language tests were administered only ABI condition and verbally. Because she did not have adequate experience with her CI.

2.4. Auditory brainstem surgery

The ABI was performed on the left ear when the patient was 24 months of age (Med-EL Coop., Innsbruck, Austria). The intraoperative monitorization of the cranial nerves during the ABI surgery were not performed. Electrical evoked auditory brainstem responses (E-ABR) were able to elicit good responses on the left ear during the left ABI surgery (Fig. 5). In the measurement of three different electrodes (E1-E5-E11), single peak response was observed at the amplitude of 400 qu with 60 μ sec pulse duration.

2.5. Initial stimulation of the ABI

During the initial stimulation of the ABI, facial nerve stimulation was observed on the contralateral side during the telemetry measurement. The electrodes were stimulated one by one, and non-auditory sensation was seen on nine electrodes. Ipsilateral facial nerve stimulation (FNS) was seen in four electrodes (E1, E2, E5, and E9) in the Medel ABI plaque. Five electrodes (E3, E4, E6, E8, and E11) caused contralateral FNS. The remaining electrodes (E7, E10, and E12) were stimulated to give an auditory response at a 8–10 qu charge levels with

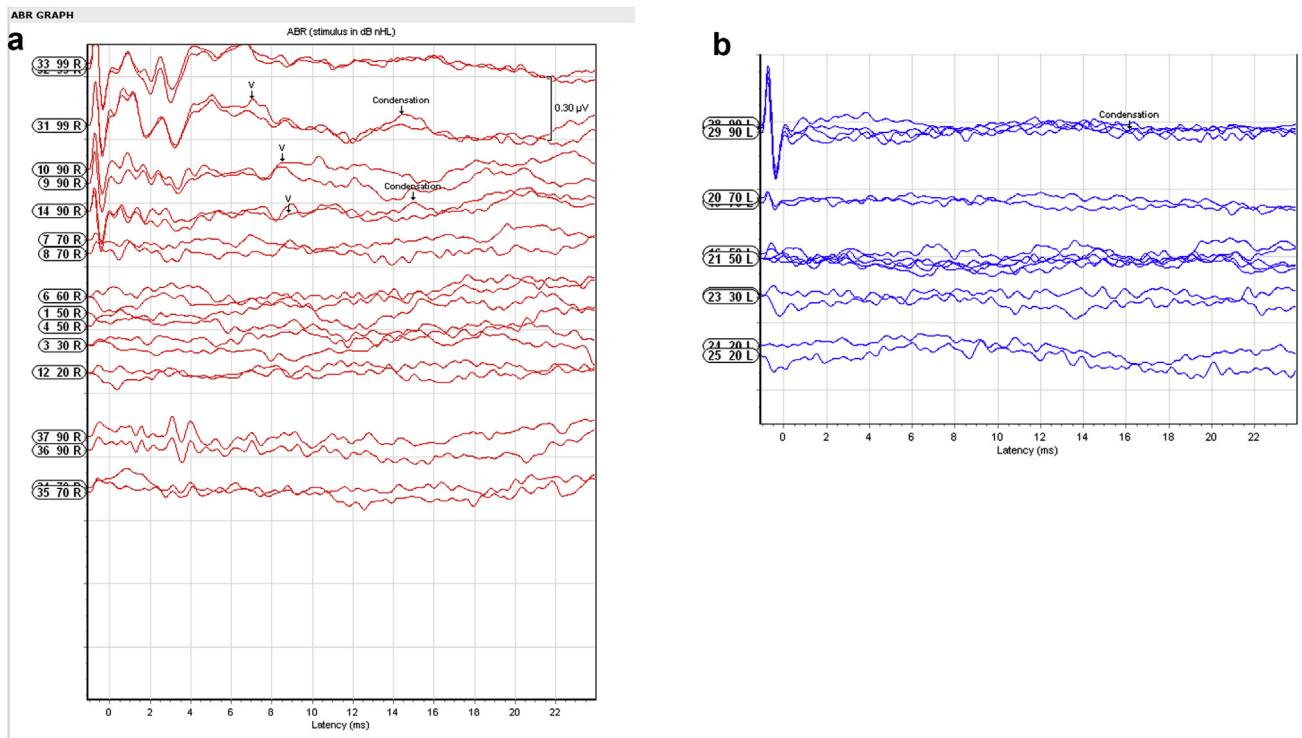


Fig. 2. Auditory brainstem responses in preoperative audiological evaluation A. Right ear B. Left ear.

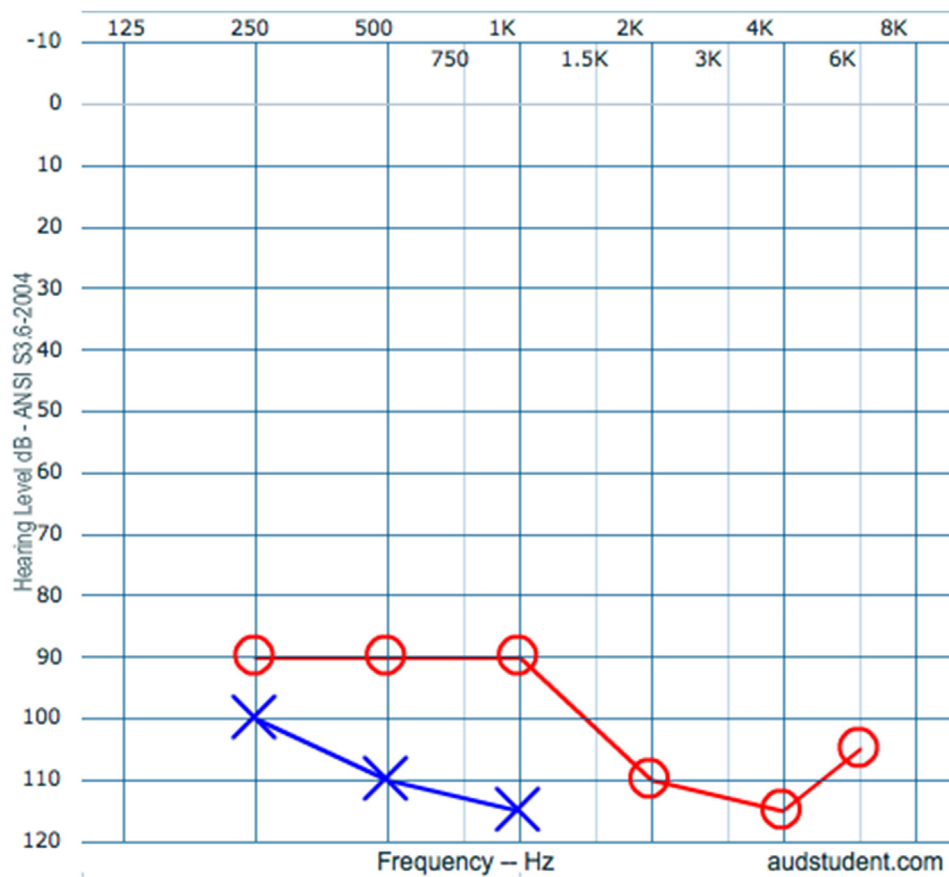


Fig. 3. Behavioral testing with insert earphones before surgery.

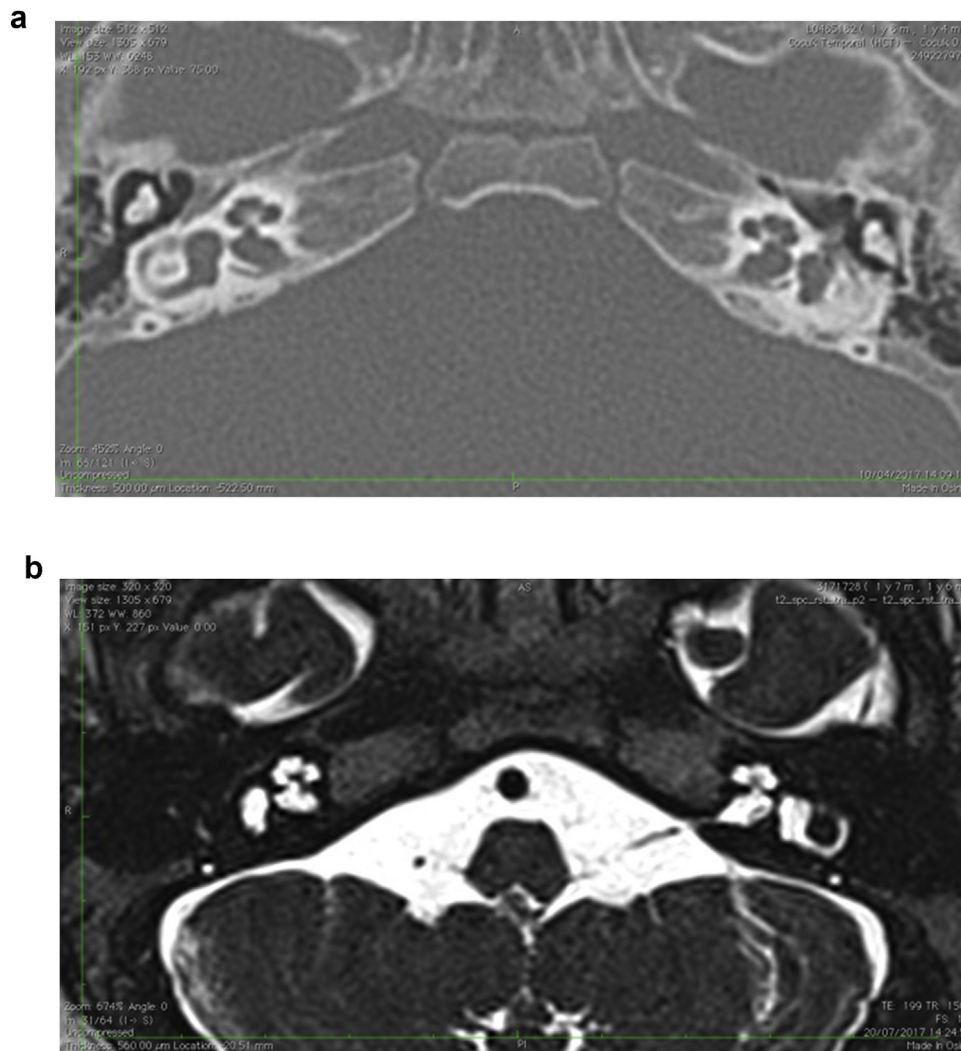


Fig. 4. A. Temporal CT showing normal cochlea and cochlear aperture. Internal auditory canal shows marked narrowing on the medial part. B. Temporal MRI showing bilateral absent cochlear nerve and narrow internal auditory canal.

7–12 μ sec pulse duration. Only three electrodes could be activated in the initial stimulation. The schematic view of the ABI electrode array was given in Fig. 6. In the first month follow-up visit, all the electrodes were checked for non-auditory stimulation, and none of the deactivated electrodes could be activated. In case of three activated electrodes, she started to use the sound processor regularly and started to recognize the environmental sound after the first month. An auditory perception assessment was performed 20 days after initial stimulation. Her sound awareness started to improve from environmental sounds to speech sounds at that time. She used her implant regularly. She started to use sign language. The sign language helped her to develop receptive language development.

2.6. Six month follow-up visit with the ABI

In the six month follow-up visit, her auditory responses with the ABI were determined as 65 dB and 50 dB for/ba/and/sh/speech stimuli, respectively. Her Ling Six Sound Test was increased from 3 to 5. Her auditory perception skills were improved through speech sound discriminations. Parallel to her auditory perception skills, her language performance was improving. Her performance in all areas improved steadily.

2.7. Cochlear implant surgery

Although her language development showed improvement with the left ABI, the family reported good responses to the environmental sounds with hearing aids in the right ear in daily life during the follow-up. Together with the family, cochlear implantation (CI) was recommended for the right ear to provide bimodal stimulation.

2.8. First year follow-up visit with ABI

Behavioral responses of the patient with the left ABI in the first-year follow-up visit are given in Fig. 7. She started to discriminate between Ling's Sounds, except the high-frequency sound/s/. The parents' observation was similar to the clinical findings. The MAIS scores indicated that she used her implant regularly and started to tell the difference between whether there was a sound or not. She started to be aware of the new sounds around her, and she asked with sign language. She preferred total communication, and her verbal language development also continued to improve. Her expressive language level reached a single-word production level. The auditory perception and language development of the case before and after ABI surgery was given in Table 1. The outcomes with cochlear implantation were not shared in this paper due to the limited duration of the CI use.

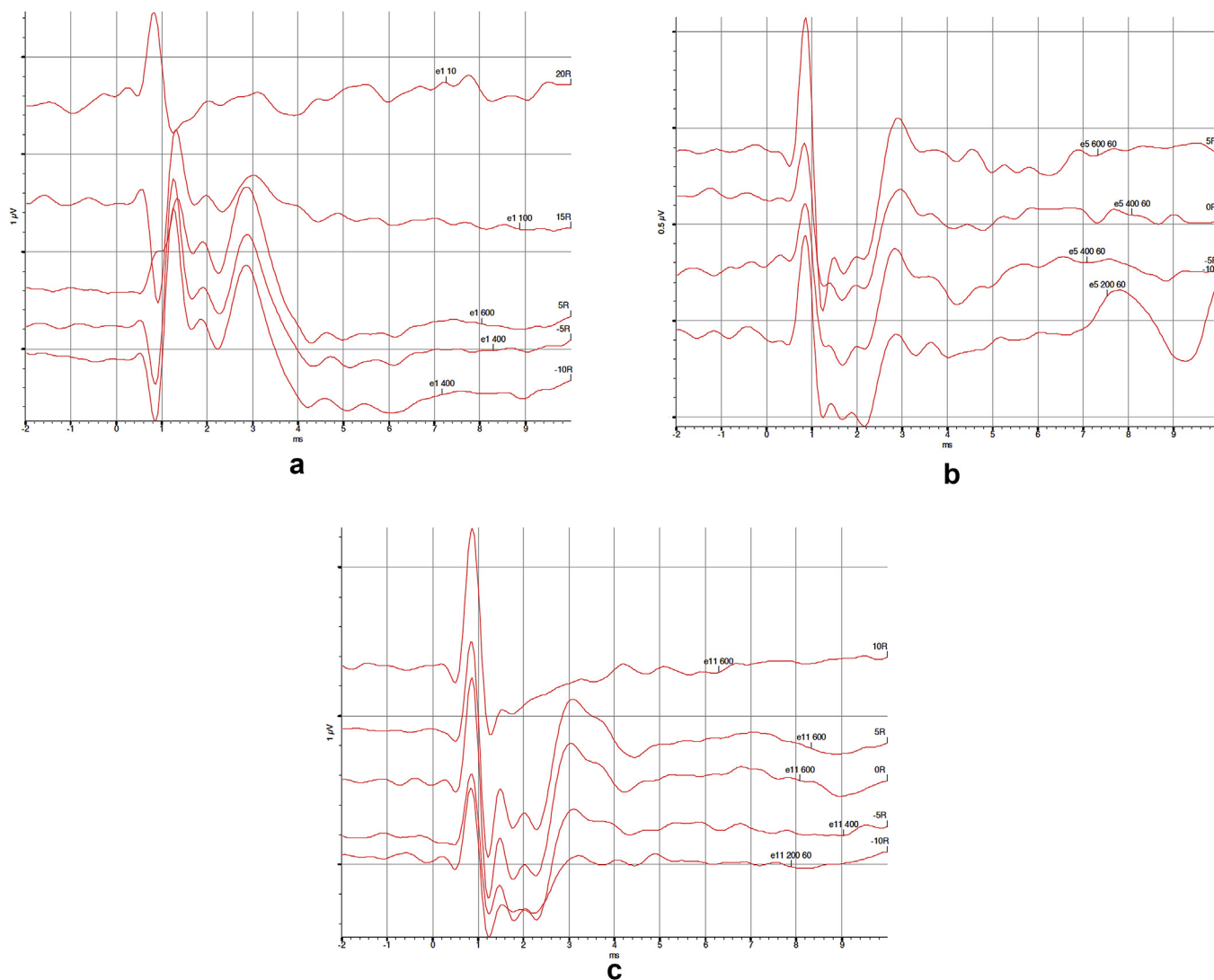


Fig. 5. eABR waveforms with ABI from the electrodes A. E1, B. E5, C. E11.

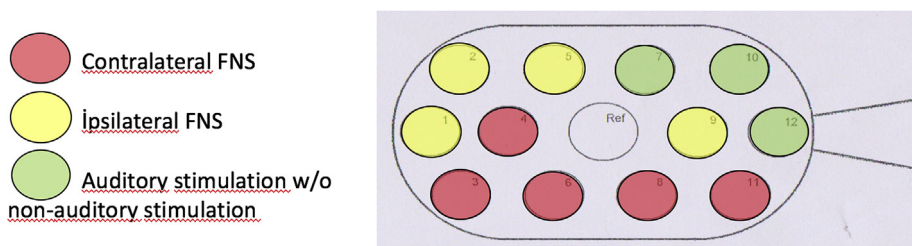


Fig. 6. Schematic view of the ABI plaque and observed non-auditory/auditory stimulation on each electrode.

3. Discussion

The patient presented here demonstrates a case of contralateral non-auditory stimulation related to the stimulation of the ABI. In the literature, many studies reported non-auditory stimulation after ABI surgery [1,2,5–8,14]. Only in the study of the Nevison et al., contralateral non-auditory stimulation was reported in two NF2 patients on the arm and ear [7]. None of these studies gave any information about the stimulation of the regions near the cochlear nucleus on the contralateral side in children. In this case, possible reason for the contralateral stimulation may be pontocerebellar hypoplasia. Due to the hypoplastic structure of the brainstem, cranial nerve nuclei on either

side of the brainstem maybe situated abnormally closer to each other. Therefore, the electrical stimulation may affect the other structures located in the contralateral pathways. The main observed non-auditory side effect was facial nerve stimulation, indicating facial twitching, movement of the wings of the nose, and eye blinking. The possible reason of the facial nerve stimulation in the present case may be the proximity of cochlear and facial nuclei. Due to the limited increase of the charge levels, it was not possible to observe non-auditory stimulation of the other cranial nerves. If the current level could be increased, perhaps we could see non-auditory stimulation in other cranial nerve nuclei. It is difficult to explain not only the non auditory stimulation in a specific region of the ABI plaque but also auditory responses

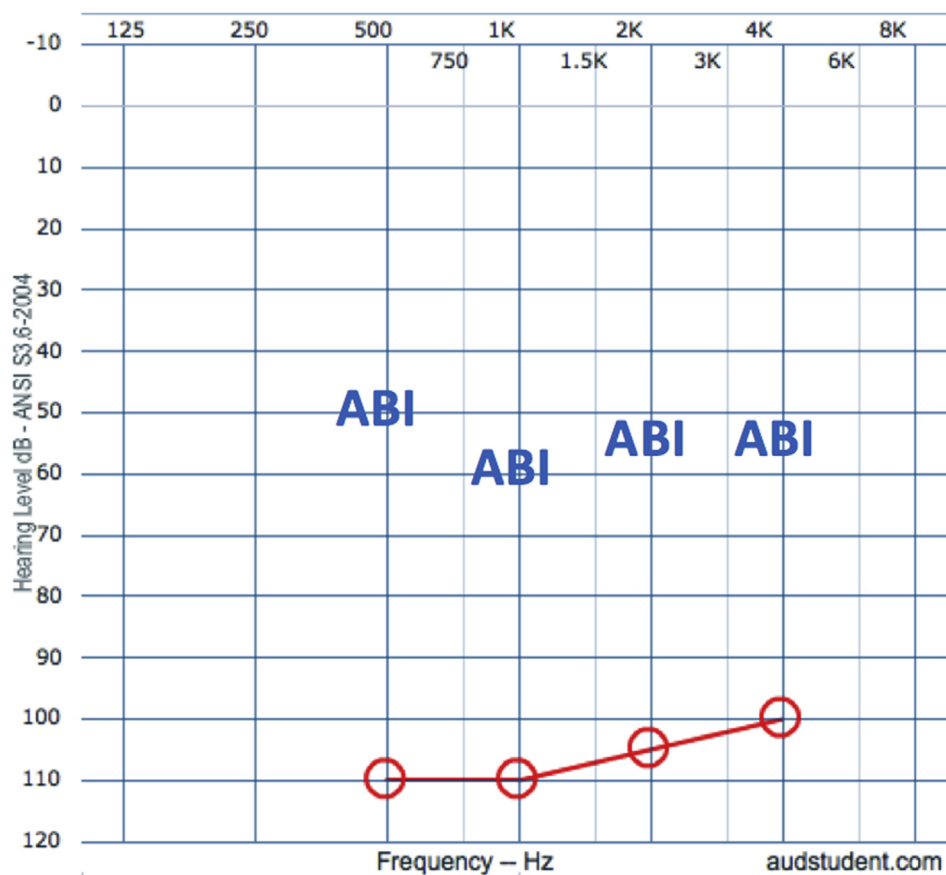


Fig. 7. Behavioral responses with ABI in first year follow-up visit.

at the very low stimulation levels (at a 8–10 qu charge levels with 7–12 µsec pulse duration) in three electrodes. In case of the increase of the most comfortable levels of these electrodes, facial nerve stimulation was also observed in the ipsilateral side. Due to the good auditory responses, the most comfortable levels were set at the levels without facial nerve stimulation.

Depending on the placement of the ABI electrode, non-auditory stimulation may occur with the stimulation of the different cranial nerves. For instance, in the case of a low placement, a sense of tingling or constriction in the throat can be observed, related to the stimulation of the glossopharyngeal nerve [5]. In that study, Toh and Luxford reported facial twitching caused by the low placement of the ABI electrode [5]. In our case, the reason for the stimulation of the facial nerve may be the high placement of the ABI electrode.

Although the response was obtained in the intraoperative measurement, a sufficient number of electrodes could not be activated due to non-auditory stimulation in the first activation. The response from the ABI electrode was only from the outer part of the plate electrode. If we think of the possibility of the migration of the electrode out of the recess, there would be audiological response only from the tip of the electrode. Therefore, it is not possible to explain these findings with the

possibility of the migration of the electrode. The presence of significant improvement in the auditory development and only auditory response from the outer part of the electrode plaque in this case remove the idea that these findings may have happened due to a displacement of the ABI plaque in the postoperative period before the activation.

Anwar et al. reported the intraoperative E-ABR results in adult NF2 patients with an ABI. The results of the study indicated that each E-ABR waveforms should have a stimulus artifact within the first 0.4 ms, and the peaks after 4 ms show the non-auditory stimulation without any stimulation of the auditory pathway. It was recommended to not use the electrodes with non-auditory stimulation while measuring E-ABR during the actual device stimulation [15]. In the intraoperative testing of our case, two peak responses were observed without any non-auditory stimulation in the EABR measurement in E1. On the other hand, one-peak response was observed at 3 ms in the waveforms of E5 and E11. It was possible that an eABR response was found in the intraoperative testing, but at high charges that cannot be achieved post-operatively because they activate non-auditory responses at very low charges. During the intraoperative measurement, any non-auditory responses were not identified after 4 m.s. Even not having late potentials during intraoperative electrical ABR, our findings suggest that other

Table 1
Auditory perception and language development of the case before and after ABI surgery.

	Preoperative Evaluation	Initial Stimulation of ABI	6th Month Follow-Up Visit	1st Year Follow-Up Visit
Auditory perception skills				
Ling's Six Sound Test	0/6	3/6	5/6	5/6
IT/MAIS total scores	0/40	3/40	6/40	10/40
Language development				
Receptive Language (age equivalent score)	0–6 months	6–12 months	12–17 months	17–24 months
Expressive Language (age equivalent score)	0–6 months	0–6 months	6–12 months	12–17 months

structures would have been stimulated in that region. As we directly observe the cranial nerves, we routinely do not monitor the cranial nerves during ABI surgery in non-tumor patients. In case of ABI surgery in patients with tumors (such as neurofibromatosis type II), we routinely monitor the cranial nerves because there is a tumor and it is difficult to identify the nerves in case of a tumor without monitorization. Even if monitoring had been performed during surgery, it would not be possible to observe the contralateral non-auditory stimulation due to the routine monitorization on the ipsilateral side.

In Shah et al.'s paper, the authors state that severe neurodevelopmental delay, central hearing loss, and brainstem deformities are contraindications for ABIs [16]. In our case, pontocerebellar hypoplasia and developmental motor delay were diagnosed. The possible reason for the contralateral non-auditory stimulation may be this brainstem pathology. In the first ABI consensus statement reported by Sennaroglu et al., the authors found that the presence of additional handicaps diminished the success of the ABI in children [17]. Our case was diagnosed with developmental motor delays in the follow-up of the ergotherapy, physiotherapy, and neurology. Despite additional handicaps and severe non-auditory stimulation, our case benefits from the ABI, and her auditory perception abilities and speech development improved after ABI surgery.

Despite the low stimulation levels, facial nerve stimulation was observed during the telemetry measurement in our case. It is important to start increasing the most comfortable levels sequentially for each electrode, until reaching the levels of the telemetry (nearly 5k Ω in Med-El devices) in the initial activation of the ABI. Otherwise, it is a high possibility to experience non-auditory stimulation during the telemetry. After the achievement of the current telemetry levels without any undesired non-auditory stimulation, telemetry measurement can be performed safely. After the telemetry measurement, the programming session can continue by adjusting thresholds with behavioral methods. While setting the uncomfortable levels, it is essential to observe possible non-auditory stimulation until the expected dynamic range is reached. In order to prevent any possible non auditory stimulation, it is recommended to decrease the stimulation levels for all electrodes simultaneously before activating the sound processor into a live voice. Activating all the possible electrodes can be annoying for the child. After observing no side effects, it is advised to increase the first planned levels [18].

Programming techniques in ABIs may change between different clinics because of the differences in training, the availability of the equipment, and the patient demographics [4]. In pediatric ABI users, it is preferred to adjust the thresholds automatically (15–30%) in our clinical practice, because most of the young children show supra-threshold responses to the sound. It is important to plan the ABI programming sessions together with two pediatric audiologists in children with an ABI. One audiologist should be close to the patient to observe not only auditory responses but also possible non-auditory stimulation, while the other can set the stimulation levels. Thus, the programming can be performed more accurately and safely.

In the previous papers, many modifications were recommended for non-auditory stimulation, such as increasing the duration of the stimulus pulse, deactivating the electrode, switching the reference electrode, or changing the stimulation mode [1,5,19]. During the follow-up visits, increasing the minimum duration levels and triphasic stimulation was tried to eliminate the non-auditory stimulation; however, none of these modifications could solve the problem. Consequently, all electrodes with non-auditory stimulation were deactivated.

Goffi-Gomez et al. reported the map parameters of children with an ABI and mentioned that the number and position of the active electrodes were not directly related to the outcomes [19]. Similarly, it was possible to improve the auditory skills and speech development in our case one year after surgery, despite three active electrodes. This finding was similar to the consensus paper by the Sennaroglu et al. It was reported that children may benefit from an ABI in view of the

improvement in the auditory skills and language development, but it is hard to develop speech like that of children with cochlear implants due to the complex inner ear malformations. Children with an ABI need to be included in intensive auditory rehabilitation programs with substantial family support, using both sign language and lip reading [20]. In our case, the mother takes care of the patient, and they communicate using all communication methods, such as auditory verbal, baby sign language, and lip reading. With the substantial support of the family, her communication abilities developed surprisingly well in one year of ABI use.

In the present case, auditory comprehension must be improved with ABI because all auditory perception tests were administered only auditory condition. On the other hand, language development is a more complex skill which includes semantics, syntax, and lexical development. We believed that sign language helps her to improve her receptive language skills. She started to combine the visual and auditory cues more efficiently. In our clinical observation and feedback from family were verified our findings in her daily life that her comprehension was improved with her ABI. Also, our initial test results were administered with her hearing aids and she did not show any improvement in her auditory perception and language skills. One of the limitations of our case was no additional information or outcomes with cochlear implantation. Because at the time of our case was written, her experience with CI was not enough to share the outcomes.

4. Conclusions

In view of the present findings, it is possible to encounter contralateral non-auditory stimulation with the stimulation of the cranial nerves near the cochlear nucleus in children with pontocerebellar hypoplasia. In children with an ABI, it is possible to provide auditory information to improve auditory skills and develop speech, despite additional handicaps. In the case of cochlear nerve aplasia, audiological evaluation plays an important role to make a decision for CI.

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Conflicts of interest

All authors declare no conflict of interest in relation to the attached article.

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