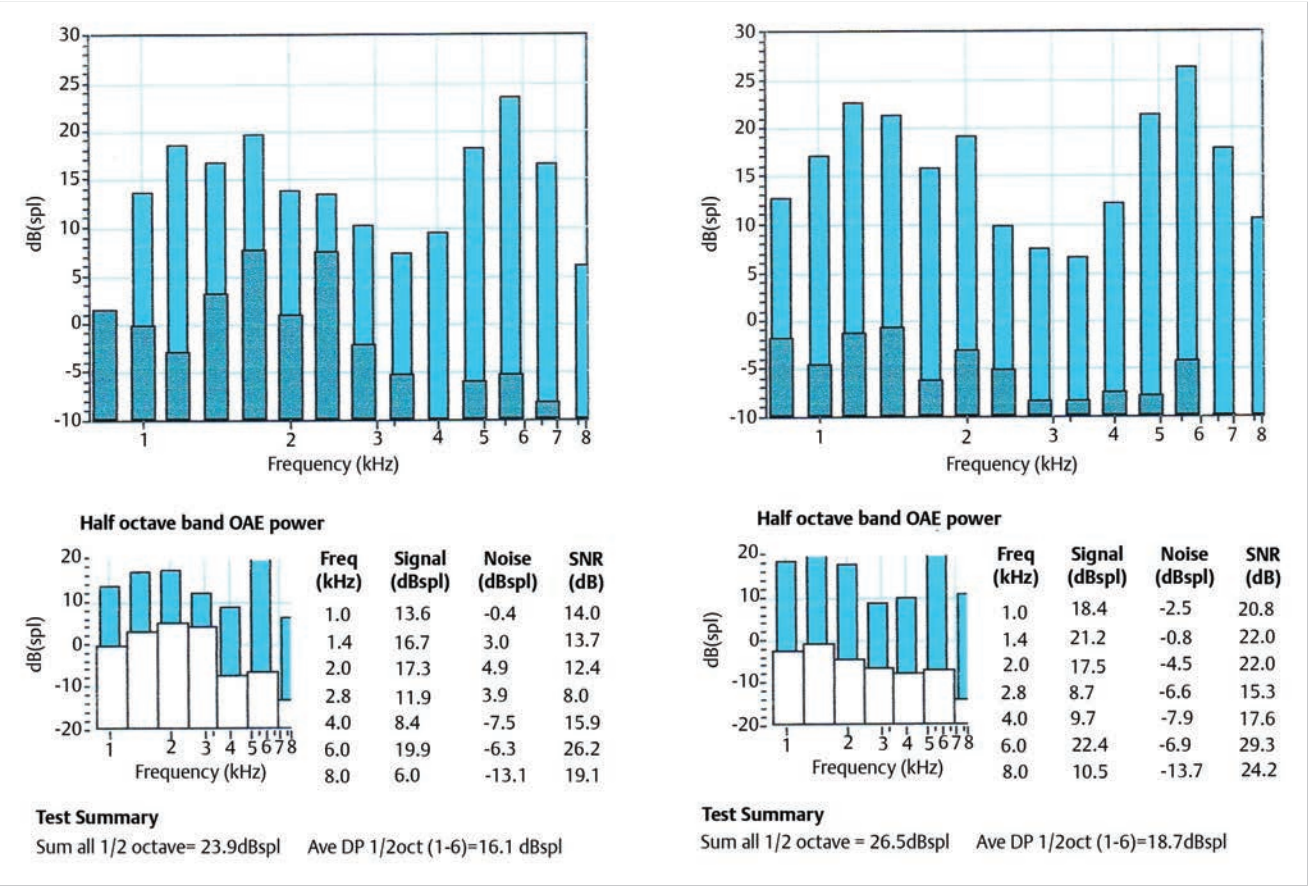

Fig. 2.1 Results of distortion product otoacoustic emission (DPOAE) testing at follow-up appointment.

Table 2.2 Soundfield speech detection and warble-tone thresholds obtained at follow-up appointment (6 months of age)

	SDT	500 Hz	1,000 Hz	2,000 Hz	4,000 Hz
Soundfield	20	20	20	20	20

Note: Testing was not attempted at presentation levels below 20 dB HL. SDT, speech detection threshold.

However, ear-specific behavioral testing, using ear-insert earphones, should be performed at the next test session.

3. **Is there still any reason to be concerned that Marissa may have a disorder of hearing?**
Our test results suggest that Marissa has hearing sensitivity within normal limits at 6 months of age. The reason for her referral from the newborn hearing screening program is not indicated, but it may have reflected a transient conductive hearing loss due to vernix in the ear canals or other environmental obstacle to obtaining a successful hearing screening result. However, coupled with the family history of childhood hearing loss, there is reason for Marissa's parents, doctor, and audiologist to remain vigilant about changes in her hearing and development of speech and language. The results reported here do not rule out progressive postnatally developing sensorineural hearing loss, auditory processing disorder, or other subtle pathology.

2.8 Recommended Treatment

Hearing evaluations every 6 months until 3 years of age were recommended, or sooner if parents noted a decrease in response to sounds. Genetic evaluation was recommended due to the positive family history of hearing loss.

2.9 Outcome

Marissa's parents were relieved to know that, at this time, Marissa seems to be responding well to sound. Her parents received information on speech-language and hearing milestones, and plan to follow up with hearing evaluations.

Suggested Reading

[1] American Speech-Language-Hearing Association. Newborn Hearing Screening (Practice Portal). Available at: www.asha.org/Practice-Portal/Professional-Issues/Newborn-Hearing-Screening. Accessed 2016

[2] American Academy of Pediatrics, Joint Committee on Infant Hearing. Year 2007 position statement: principles and guidelines for early hearing detection and intervention programs. Pediatrics. 2007; 120(4):898–921

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3 Probable Enlarged Vestibular Aqueduct and Hearing Loss in a 6-Year-Old Child

Jessica Sullivan and Homira Osman

3.1 Clinical History and Description

Luis was diagnosed with profound sensorineural hearing loss in his right ear at birth via brainstem auditory evoked response testing. Distortion product otoacoustic emissions were absent from 750 to 4,000 Hz for the right ear. The left ear demonstrated normal hearing at birth. His parents reported that his hearing was evaluated approximately every 6 months consistently from the time of diagnosis at an outside facility (▶ Fig. 3.1). Luis was not fitted with any type of amplification given the severity of hearing loss in the right ear. Luis's parents did not enroll him in any early intervention services as they felt he was demonstrating consistent responses to their voice and to sounds in the environment. At the age of 4 years, Luis's parents began to suspect a change in his hearing. On the date of this exam, case history revealed normal birth history and developmental milestones, including speech and language. His parents reported no permanent childhood hearing loss in their family. An audiologic evaluation revealed that the hearing in the left ear had dropped to a moderate to moderately severe sensorineural hearing loss (▶ Fig. 3.2). The hearing in the right ear was stable as a profound sensorineural hearing loss. His pure-tone average (PTA) of 60 dB HL (hearing level) in the left ear was within 5 dB of his speech reception threshold (SRT). An SRT was unable to be obtained at the equipment limits for the right ear. Tympanograms were type A_S bilaterally, showing reduced compliance of the eardrum mobility bilaterally.

Luis was referred for close monitoring of his hearing. Audiologic thresholds at the 1- and 3-month follow-up appointments were consistent with the thresholds on audiogram 2. His parents reported no fluctuations in hearing or dizziness. Luis was fitted with a digital behind-the-ear hearing aid for the left ear. Initially, he did not wear his hearing aid consistently, but eventually he was wearing his hearing aid for close to 9 hours per day. According to his audiologist, the hearing aid was fitted to prescribed targets using real ear measures. An aided word recognition score of 68% using the NU-CHIPS (Northwestern University-Children's Perception of Speech) recorded test materials was obtained, but no aided thresholds were measured. Additionally, Luis began to use a remote microphone (RM) system at school, and his mother reported benefit from the RM system when it was used. Luis has been using successfully a digital behind-the-ear hearing aid since that time.

At the age of 5 years, Luis described spinning vertigo, usually in the morning before he got out of bed. He reported the spinning vertigo to occur as frequently as twice a month. His parents reported that Luis easily became sick in the car. Luis was referred to a multidisciplinary clinic to explore the etiology of his hearing loss. As part of this clinic, a computed tomography (CT) scan of the temporal bone was ordered.

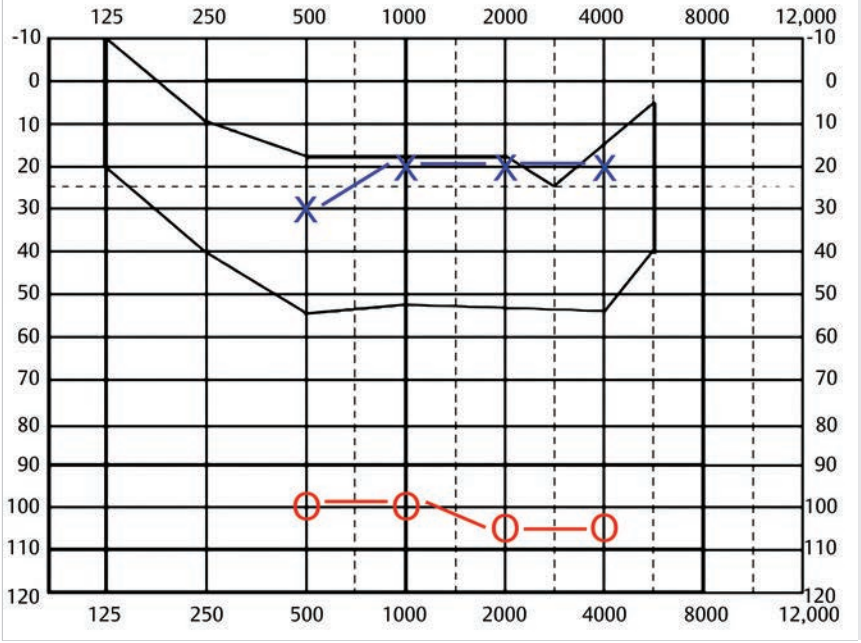

Fig. 3.1 Results at the diagnostic brainstem auditory evoked response test (age 5 weeks). O, right ear air conduction; X, left ear air conduction.

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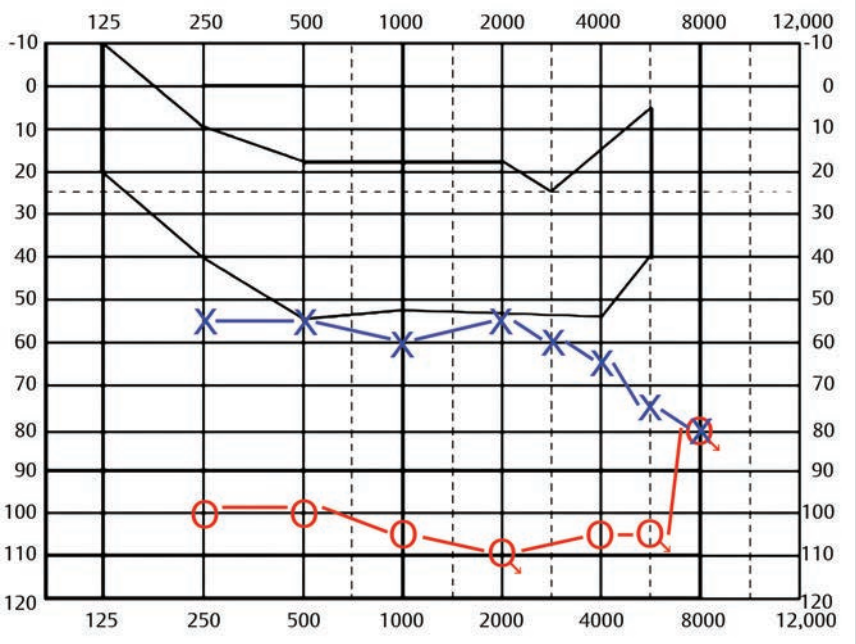

Fig. 3.2 Pure-tone test results (age 4.2 years). O, right ear air conduction; X, left ear air conduction; Arrows, no response.

3.1.1 Question: What is the probable diagnosis? What findings led you to this conclusion?

A CT scan of his temporal bones indicated enlarged vestibular aqueducts for both ears, with the right (4.0 mm) being larger than the left (1.6 mm). Following a discussion with the physician and audiologist, Luis's family was given information about cochlear implants.

3.2 Cochlear Implant Candidacy Evaluations

At this initial appointment through the cochlear implant program, Luis received a complete hearing evaluation. Otoscopy revealed clear ear canals bilaterally. No vestibular episodes were reported. Tympanograms were type A bilaterally, showing no evidence of middle ear pathology. Pure-tone thresholds had worsened since his previous testing approximately 6 months before. Previous testing yielded moderate to moderately severe sensorineural hearing loss, and current testing was consistent with a severe to profound sensorineural hearing loss (▶ Fig. 3.3). Programming changes were made to Luis's hearing aid. The hearing aid was fitted to prescribed targets using real ear measures, but the output was slightly lower than the targets for the mid- to high-frequency range. It was recommended that Luis return for an aided audiologic evaluation.

Aided warble thresholds were obtained (▶ Fig. 3.4). Aided word recognition (NU-CHIPS, recorded) performance was 16% correct in the left ear with a 45 dB HL presentation level. This is a significant decrease from the last evaluation (▶ Fig. 3.2). A few weeks following this appointment, Luis's family met with an otologist and aural habilitation specialist.

3.3 Outcome

At present, Luis, age 6 years, continues to use the hearing aid in his left ear and an RM system at school. Luis's parents are very interested in cochlear implants and are awaiting a decision from the cochlear implant program team. His parents would like for him to be implanted in both ears prior to the new school year. They have started services with an aural habilitation specialist at the hospital.

3.4 Questions for the Reader

1. How prevalent is enlarged vestibular aqueduct (EVA) disease in children?
2. Is Luis a good candidate for a cochlear implant?
3. Would you recommend one or two cochlear implants in this case?
4. What would an aural habilitation plan consist of postimplant?
5. What other recommendations would you suggest for this family?

3.5 Discussion of Questions

1. **How prevalent is EVA disease in children?**
   It is estimated that 5 to 15% of children with a sensorineural hearing loss have EVA. Most children with EVA will have some degree of hearing loss and a small portion of them will also have vestibular function issues. Alemi and Chan[1] conducted a systematic review of 23 studies and found that progressive sensorineural hearing loss was found in 39.6% of ears (1,115 with enlarged vestibular aqueduct) and only 12% were associated with head trauma. In addition, EVA has been associated with a genetic disorder (Pendred's syndrome) that

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12 Transient Auditory Neuropathy Spectrum Disorder or Delayed Auditory Maturation in a Well Baby

Andi Seibold

12.1 Clinical History

Jacob was born a healthy, full-term, 9-pound baby. The pregnancy and birth were unremarkable with no complications warranting neonatal intensive care. Before discharge from the hospital, Jacob referred on his newborn hearing screening in the left ear four times.

Jacob was seen at 4 weeks of age for diagnostic auditory brainstem response (ABR) testing at an outpatient pediatric audiology clinic. After obtaining a full case history, no family history of hearing loss was reported, and his mother believed he startled to sounds. Upon testing, there were no synchronous responses to click or toneburst stimuli in either ear under insert earphones. A low-amplitude wave V with poor morphology was identified but did not follow a normal latency-intensity function with a decrease in intensity to clicks, 4,000-Hz or 500-Hz toneburst stimuli (▶ Fig. 12.1). It was also noted that wave III had a larger amplitude in relation to waves I and V. Responses were recorded down to approximately 65 to 75 dB eHL in both ears, although this was not an estimate of hearing sensitivity in light of the abnormal ABR.

A large cochlear microphonic was noted to click stimuli at 95 dB nHL in both ears to both rarefaction and condensation stimuli. The mirror image of the preneural response was noted out to 2.6 ms in the right ear and 2.9 ms in the left ear. An alternating run showed a smoothing and near-cancellation of the cochlear microphonic, which was to be expected.

Transient-evoked otoacoustic emissions (TEOAEs) with normal amplitudes were recorded at four frequencies in the left ear and three frequencies in the right ear, indicating normal cochlear function at least through the level of the outer hair cells. Tympanometry was performed using a 1,000-Hz probe tone and showed mobile tympanic membranes. Knowing this, acoustic reflexes were performed and were found to be elevated (100–105 dB HL) at 500, 1,000, and 2,000 Hz and absent at 4,000 Hz, bilaterally. A confirmation ABR was done at 8 weeks and yielded the same results.

12.1.1 Question: What is the probable diagnosis? What diagnostic findings led you to this diagnosis?

Following the testing, a diagnosis of auditory neuropathy spectrum disorder (ANSD) was given. This diagnosis was made in the presence of the following: poor morphology/near-absence of wave V at high intensities to click; abnormal latency-intensity shift to tonebursts on the auditory evoked test; a large cochlear microphonic to rarefaction and condensation clicks; present OAEs bilaterally; and elevated to absent acoustic reflexes.

12.2 Diagnosis and Treatment

A referral was made to a pediatric otologist who also specialized in cochlear implantation. To determine how Jacob was using his hearing, behavioral observation audiometry (BOA) was performed, specifically using an approach to watch suckling reflexes during a feeding.[1] Two pediatric audiologists trained in this technique performed the test.

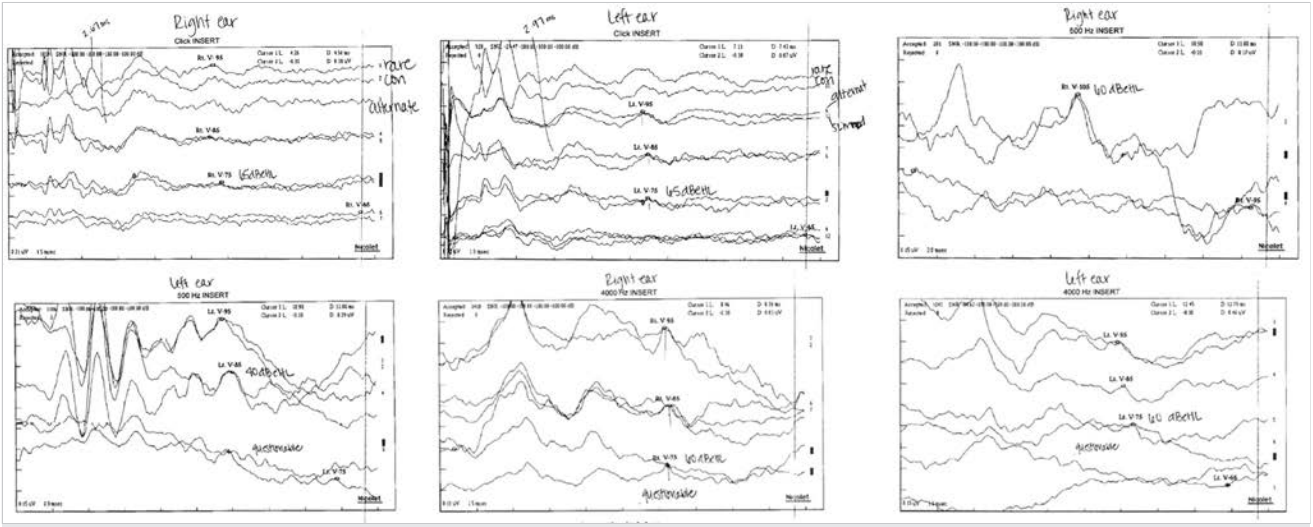

Fig. 12.1 Diagnostic auditory brainstem response at 4 weeks of age; mirror-image preneural waveform to rarefaction and condensation was observed well beyond 2 ms bilaterally, with poor wave V morphology. Questionable waveforms were marked as a note to the audiologist and otologist, and were

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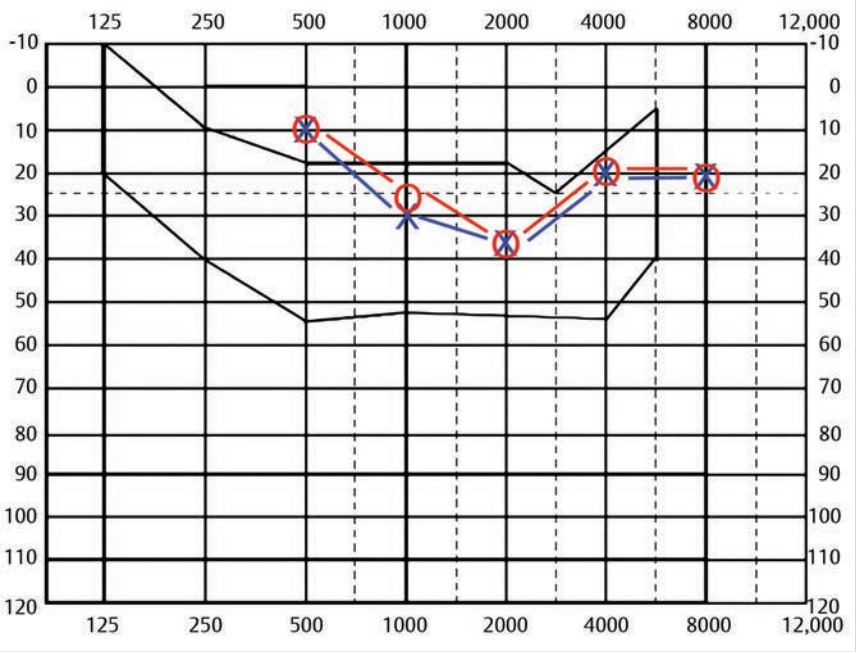

Fig. 12.2 Behavioral hearing thresholds at 12 weeks using BOA. Speech awareness threshold (dB HL): right, 10; left, 20. O, right ear air conduction; X, left ear air conduction.

Table 12.1 Aided speech awareness threshold to warbled pure tones

	500 Hz	1,000 Hz	2,000 Hz	4,000 Hz
Binaural aided (dB HL)	10	15	15	20

Jacob was 12 weeks old at the time of his first booth test; 1,000-Hz tympanometry revealed mobile tympanic membranes, bilaterally, and elevated acoustic reflexes. Warbled pure tones and narrowband noise were used as test stimuli. Responses to 500, 1,000, 2,000, 4,000, and 8,000 Hz were obtained in both ears using insert earphones in a mild cookie-bite configuration (▶ Fig. 12.2). A speech awareness threshold (SAT) by BOA was also obtained at 10 dB in the right and 20 dB in the left.

Due to a mild hearing impairment, amplification was recommended as a first step in treatment to the family, and this recommendation was supported by his otologist. Jacob was fitted with low-gain hearing aids at 13 weeks, according to Desired Sensation Level 5.0 targets and a real-ear-to-coupler difference (RECD) measure obtained in the right ear; due to inability to obtain RECD on the left ear, the right ear values were also applied to his left ear. He was then scheduled for an initial evaluation by an auditory-verbal therapist who would help monitor his progress over time.

12.3 Outcome

Jacob was seen every 2 to 3 months for the first year of his life and had both aided and unaided testing using BOA and insert earphones to track his progress. His first aided testing showed responses to warbled tones in the normal range, with an SAT obtained binaurally at 15 dB HL (▶ Table 12.1). Ling sounds were not tested due to rapid patient fatigue.

A magnetic resonance imaging of the internal auditory canals at 3 months of age yielded no abnormal findings.

Jacob had a sedated ABR at 6 months to reevaluate the neural integrity of his auditory system (▶ Fig. 12.3). This test confirmed poor wave morphology, but showed slight improvements since his first diagnostic ABR. Additionally, a present cochlear microphonic with reversal noted out to 2.4 ms was still noted with a large wave III in relation to waves I and V in both ears. Thresholds to alternating click stimuli were obtained down to 15 dB eHL, bilaterally, which was an improvement from the 65 dB eHL thresholds previously obtained. Toneburst testing was recorded down to thresholds with better morphology than his previous ABR at 4 weeks. In light of the polarity reversals and poor morphology, behavioral test results were still used to estimate Jacob's hearing.

To evaluate auditory maturation in the presence of abnormal ABRs, behavioral hearing loss, and hearing aid use, evoked P1 cortical testing was performed within a week of his 6-month ABR in the aided condition. Results were in the normal range, indicating adequate stimulation for auditory maturation (▶ Fig. 12.4).[2]

At 7 months, visual reinforcement audiometry (VRA) was attempted but Jacob could not reliably be conditioned. BOA still showed a mild hearing loss with Type A tympanograms, but acoustic reflexes were present at 90 dB at all frequencies, which was an improvement over his testing at 2 months of age (▶ Table 12.2). It is important to note that present acoustic reflexes at 90 dB are fairly uncommon for children with ANSD.[3]

At 9 months, Jacob responded to speech and tones reliably in the normal hearing range in both ears using VRA (▶ Fig. 12.5). A sedated ABR was scheduled at 1 year of age.

ABR responses showed synchronous, neural responses with waves I, III, and V at normal latencies down to 15 to 20 dB eHL in both ears for click and toneburst stimuli (▶ Fig. 12.6). His

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19 Hearing and Vestibular Loss in a 9-Month-Old Child

Kristen Janky

19.1 Clinical History and Description

The patient is a 9-month, 3-week-old female patient who is currently being followed for unilateral, profound, sensorineural hearing loss in the right ear. The patient's family wanted to determine the underlying etiology of the hearing loss; therefore, she was seen for interdisciplinary assessment including audiologic, vestibular, ophthalmology, ear, nose, and throat (ENT), and genetics evaluations.

The patient was born full-term without any complications. At age 9 months, she is reportedly meeting all gross motor milestones in that she is sitting independently and demonstrates good head control. She has history of middle ear effusion.

19.2 Audiologic Findings

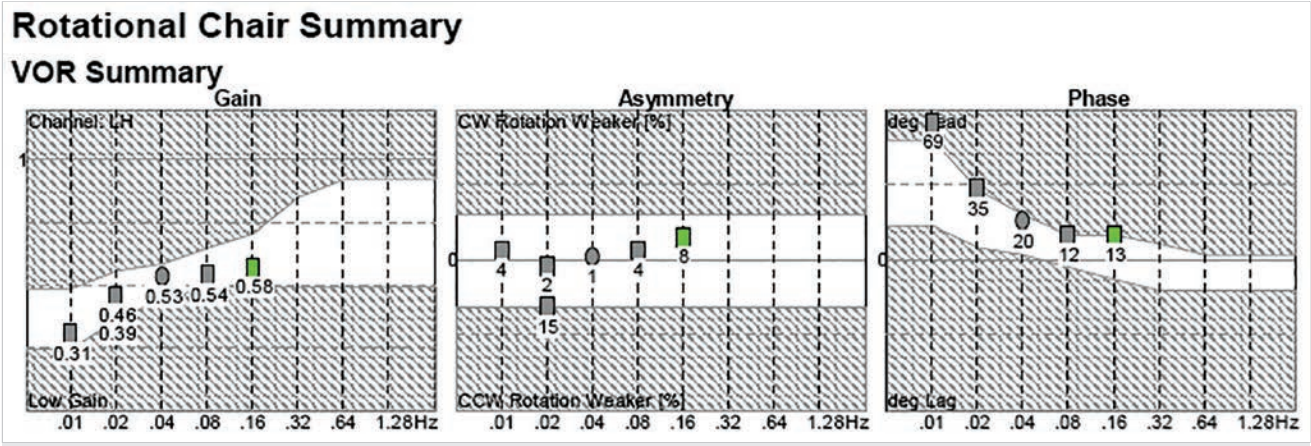
The patient did not pass her newborn hearing screen in either ear. At her rescreen, age 8 days, she passed in her left ear, but not in her right ear. Confirmatory auditory brainstem response (ABR) and distortion product otoacoustic emission (DPOAE) testing were completed at age 2 months. Tympanometry with use of a 1,000-Hz probe tone was normal in both ears prior to ABR and DPOAE testing. Responses to tone-burst ABR suggested normal hearing sensitivity in the left ear with DPOAEs present at normal absolute amplitudes from 1 to 8 kHz and profound hearing loss in the right ear with absent DPOAEs from 1 to 8 kHz. Interpeak latencies were normal, which ruled out neuropathy affecting the auditory brainstem pathways for the left ear, but it could not be ruled out for the right ear due to the degree of hearing loss. The normal tympanograms ruled out any conductive component to the hearing loss. Use of an auditory osseointegrated device (AOD) on a soft band as well as interdisciplinary evaluation, including audiologic, vestibular, ophthalmology, ENT, and genetics evaluations, was recommended, which were all completed at age 9 months.

Conventional audiometric assessment using visual reinforcement techniques was attempted at age 9 months, albeit with poor reliability. Tympanograms were consistent with middle ear effusion bilaterally. Use of an AOD on a soft band was again recommended; however, the child's mother declined to follow up on this recommendation.

19.3 Vestibular Findings

Rotary chair testing was completed at age 9 months to assess for vestibular loss as part of the interdisciplinary evaluation. The vestibulo-ocular reflex was monitored during rotation using electrodes with the patient seated on her parent's lap. Rotary chair findings are shown in ▶ Fig. 19.1. Given the abnormal phase lead at 0.01 and borderline phase lead at 0.02 Hz, these findings do not rule out unilateral peripheral vestibular system involvement. Because rotary chair is a test of overall responsiveness of the vestibular system, it does not localize to the right or left ear. However, due to the profound hearing loss in the right ear, vestibular function would most likely be affected in the right ear.

Additional testing to confirm a unilateral vestibular weakness could not be completed for this patient. Typically, in addition to rotary chair testing, the cervical vestibular evoked myogenic potential (VEMP) is evaluated in children. The cervical VEMP is an assessment of the inferior portion of the vestibular nerve and saccule and, therefore, complements the rotary chair test, which is an assessment of the horizontal semicircular canals and superior branch of the vestibular nerve. Cervical VEMP testing with air-conducted stimuli can be completed in the presence of sensorineural hearing loss as it is strictly a test of vestibular function; however, cervical VEMP testing requires normal middle ear function as conductive hearing loss attenuates the intensity of sound getting to the vestibular system. Therefore, due to the presence of abnormal tympanograms, and subsequent conductive hearing loss, cervical VEMP testing could not be completed for this patient.



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19.4 Other Findings

19.4.1 Genetics

Physical exam was not suggestive of any type of syndromic hearing loss; therefore, genetic testing was not recommended.

19.4.2 ENT

Physical exam was normal with the exception of possible bilateral effusion. In children with unilateral sensorineural hearing loss, the most common cause of hearing loss is a form of inner ear anomaly; therefore, magnetic resonance imaging (MRI), including the brainstem, with and without contrast was recommended after the patient's first birthday.

19.4.3 MRI

At age 12 months, MRI of the brain and internal auditory canals with and without contrast under sedation was completed (▶ Fig. 19.2). On the right, the eighth cranial nerve and superior portion of the vestibular nerve were visualized; however, the cochlear nerve and inferior division of the vestibular nerve were markedly hypoplastic or absent (▶ Fig. 19.2b,c). The MRI was otherwise unremarkable for the left ear (▶ Fig. 19.2a–c).

The rotary chair phase lead in the present patient would suggest that the superior portion of the vestibular nerve, while visualized on MRI, may still be affected and that cervical VEMP would likely have been absent.

19.4.4 Ophthalmology

The patient was noted to have a moderate amount of far-sightedness with astigmatism. Prescription lenses were recommended.

19.5 Questions for the Reader

1. In children with hearing loss, who is at risk for also having vestibular loss?
2. Which tests of vestibular function are typically completed in children?
3. When should a child be referred for vestibular testing?
4. What are the functional consequences of vestibular loss in children?
5. If a child with hearing loss is diagnosed with vestibular loss, what are some additional recommendations?

19.6 Discussion of Questions

1. **In children with hearing loss, who is at risk for also having vestibular loss?**
The presence of hearing loss puts children at risk for also having vestibular loss[1]; however, not all children with hearing loss will have vestibular loss. Vestibular loss is more likely to occur in children with greater severity of hearing loss,[2,3] which is why it is not surprising that in children who receive a cochlear implant, approximately 50% demonstrate some degree of vestibular loss, with 30% having bilateral vestibular loss.[4,5]

Vestibular loss is also more likely to occur with specific etiologies of hearing loss. Etiologies of hearing loss that are associated with vestibular loss include cytomegalovirus, meningitis, Usher's syndrome, Pendred's syndrome, enlarged vestibular aqueduct syndrome and other inner ear malformations, Waardenburg's syndrome, auditory neuropathy, connexin mutations (GJB2), measles, mumps, and ototoxicity, among others.[6,7,8,9,10,11,12]

2. **Which tests of vestibular function are typically completed in children?**
Vestibular testing includes traditional videonystagmography with caloric testing, video head impulse testing (vHIT), ocular and cervical VEMPs, rotary chair, and computerized dynamic posturography (CDP). Each of these exams provides complementary information regarding vestibular function; however, not all exams are appropriate for children, particularly children younger than 5 years. Therefore, the age and ability of the child are important considerations when choosing which tests of vestibular function are appropriate.

Additionally, it is important to consider the information provided by each vestibular test when choosing which tests of vestibular function are appropriate. The vestibular system is made up of five rate sensors: three semicircular canals (horizontal, posterior, and anterior canals) and two otolith organs

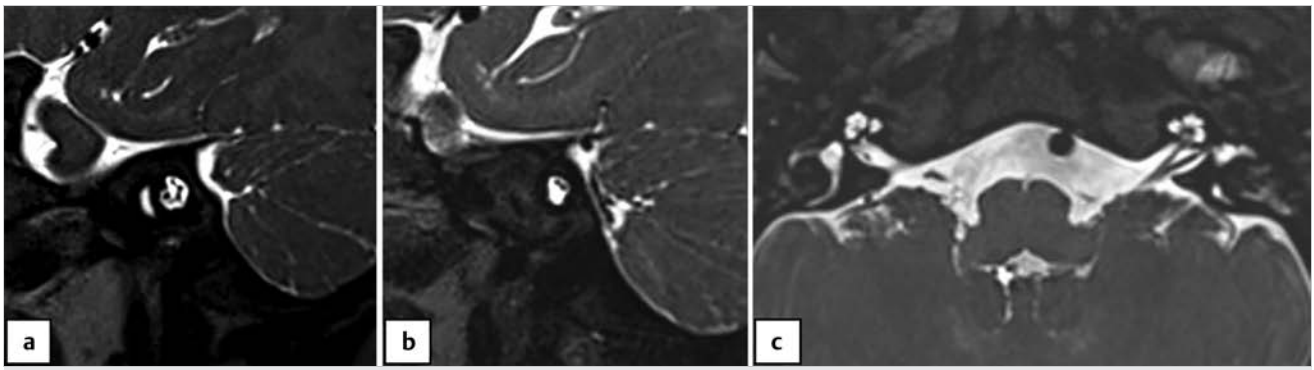

Fig. 19.2 **(a)** MRI, *left* sagittal cut of the internal auditory canal showing normal facial and vestibulocochlear nerve branches; **(b)** MRI, *right* sagittal cut of the internal auditory canal, showing facial and probable superior branch of the vestibular nerve with no visualization of the cochlear and inferior vestibular nerve branches; and **(c)** MRI, axial cut, showing the internal auditory canal. Two nerve branches (darker lines) can be seen on the left (one going to the cochlea, the other going to the vestibular system) and almost no evidence of innervation on the right.

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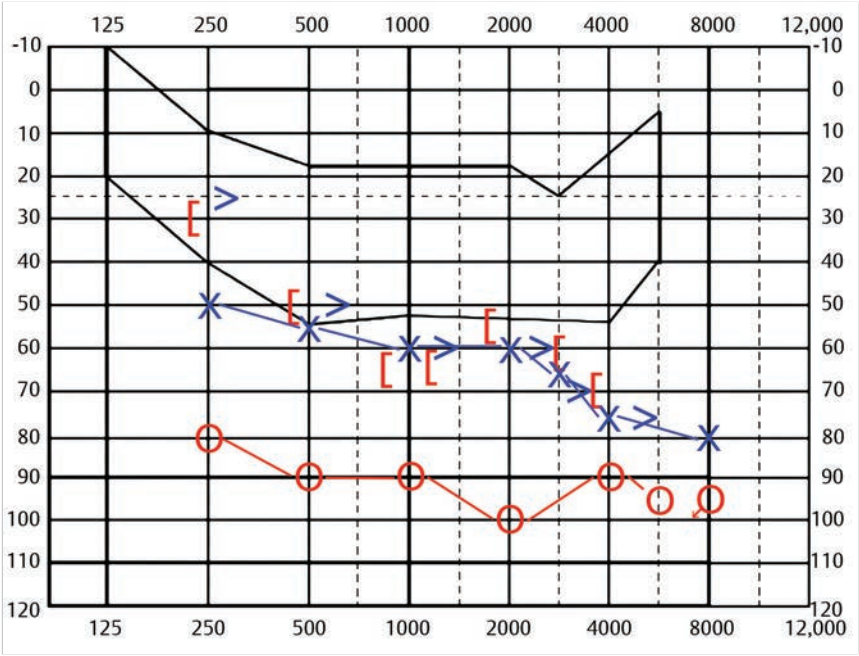

Fig. 24.2 Audiogram from June 2013. O, right ear air conduction; X, left ear air conduction; >, left ear bone conduction; [, right ear bone conduction masked.

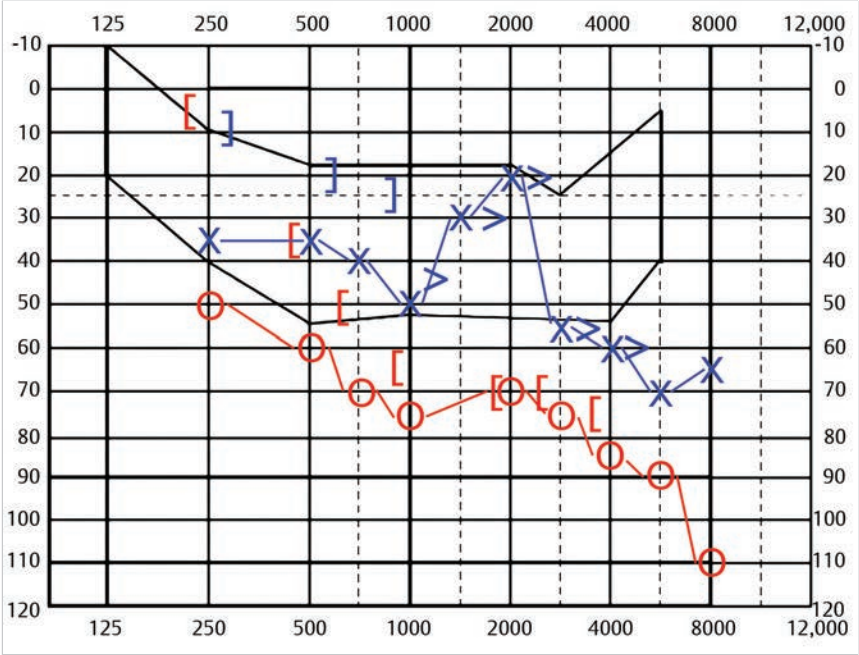

Fig. 24.3 Audiogram from August 2013. O, right ear air conduction; X, left ear air conduction; >, left ear bone conduction; [, right ear bone conduction masked;], left ear bone conduction masked.

the right ear hearing would have ever recovered. Waiting to see if the hearing recovered would have delayed her brain access to auditory information at the time.

2. **Why did Sarah adjust so quickly to the CI? What may have happened if surgery was delayed?**
   Sarah had normally developing speech and was doing well academically, so it is very likely she had normal hearing in her early years, and therefore she relied on audition for language access and her brain had been developed with auditory information. She had been quickly introduced to hearing aids, and therefore, her auditory system had been flooded with sound, keeping the brain pathways active. Her auditory system quickly adjusted from acoustic sound with brain had been developed with auditory information, and the drop in hearing happened fairly quickly. If surgery was delayed and her hearing remained in the profound range, her brain may not have received enough auditory input from hearing aids. This lack of auditory information may have resulted in auditory neural deprivation and may have had negative effects on Sarah's ability to interpret auditory information from the CI.
3. **Would a CI be a good choice for the left ear?**
   At this time, the left ear is not a candidate for a CI. The left ear hearing will continue to be monitored. However, if the hearing sensitivity of the left ear hearing decreases and/or continues to fluctuate, a CI may be considered if

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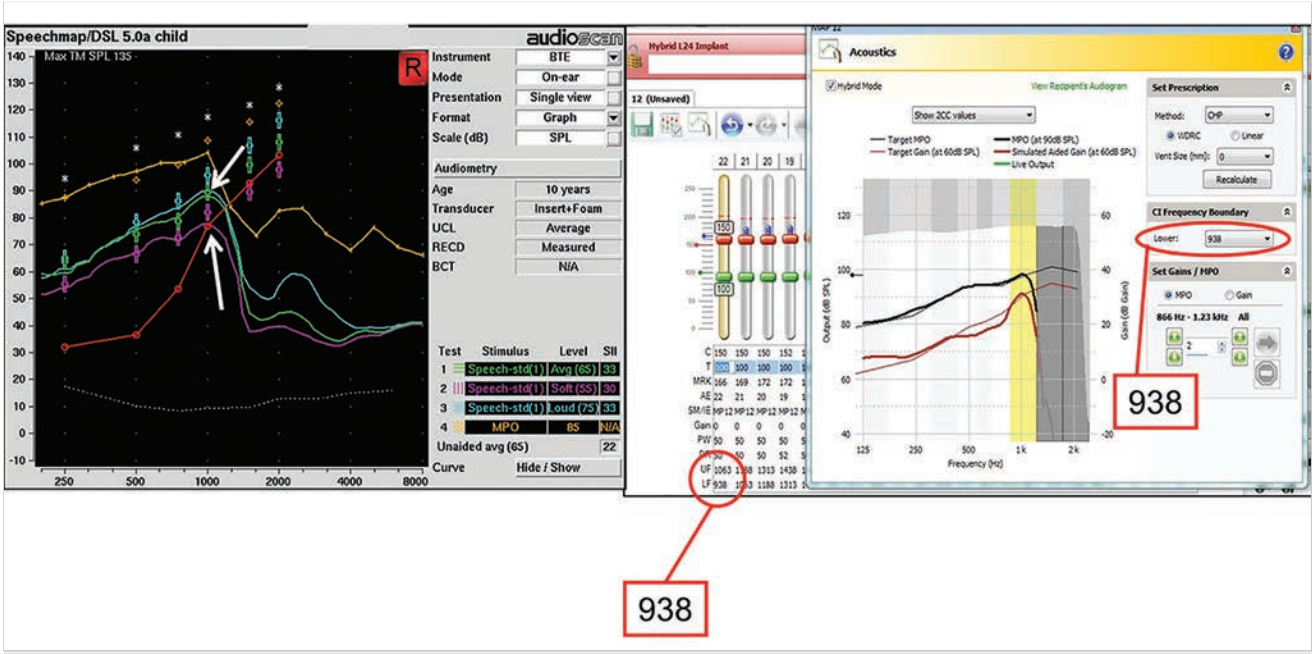

Fig. 42.6 Example of how the audio signal is allocated for acoustical and electrical stimulation.

Table 42.3 Aided speech recognition results obtained after Jayden had used his Cochlear Nucleus hybrid implant for 3 months

Tests	Prehybrid: right HA	Prehybrid: bilateral HAs	1 mo posthybrid: hybrid only	1 mo posthybrid: hybrid + left HA	3 mo posthybrid: hybrid only	3 mo posthybrid: hybrid + left HA
PBK-50 (60 dBA)	20%	36%	60%	64%	80%	76%
BKB-SIN (60 dBA)	14.5 dB	13 dB	7.5 dB	9 dB	5 dB	4 dB
CNC words (60 dBA)					92%	88%
AzBio sentences (60 dBA)					68%	77%

HA, hearing aid; CNC, consonant-nucleus-consonant

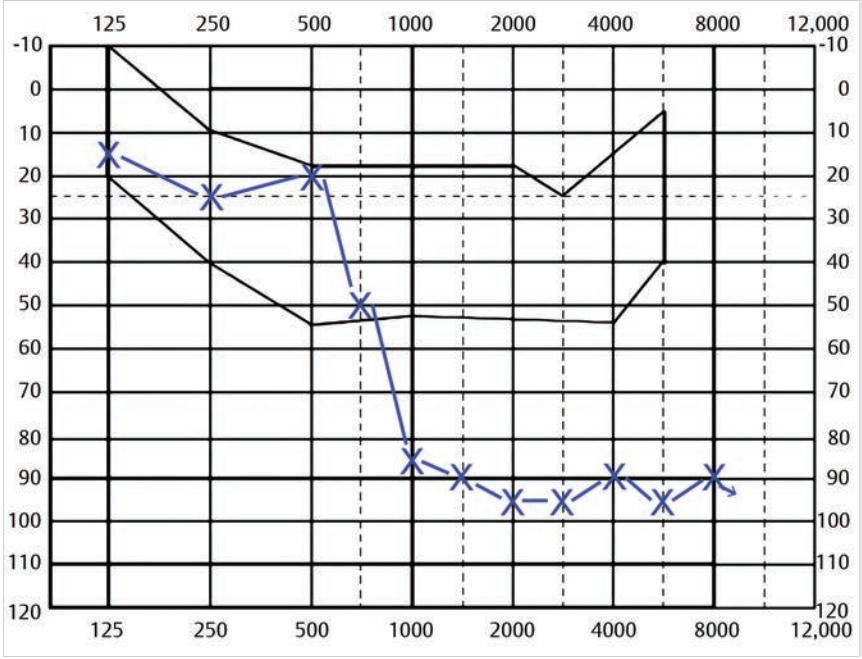

Fig. 42.7 Audiogram obtained after Jayden received a Cochlear Nucleus Hybrid implant for his left ear. X, left ear air conduction; Arrows, no response.

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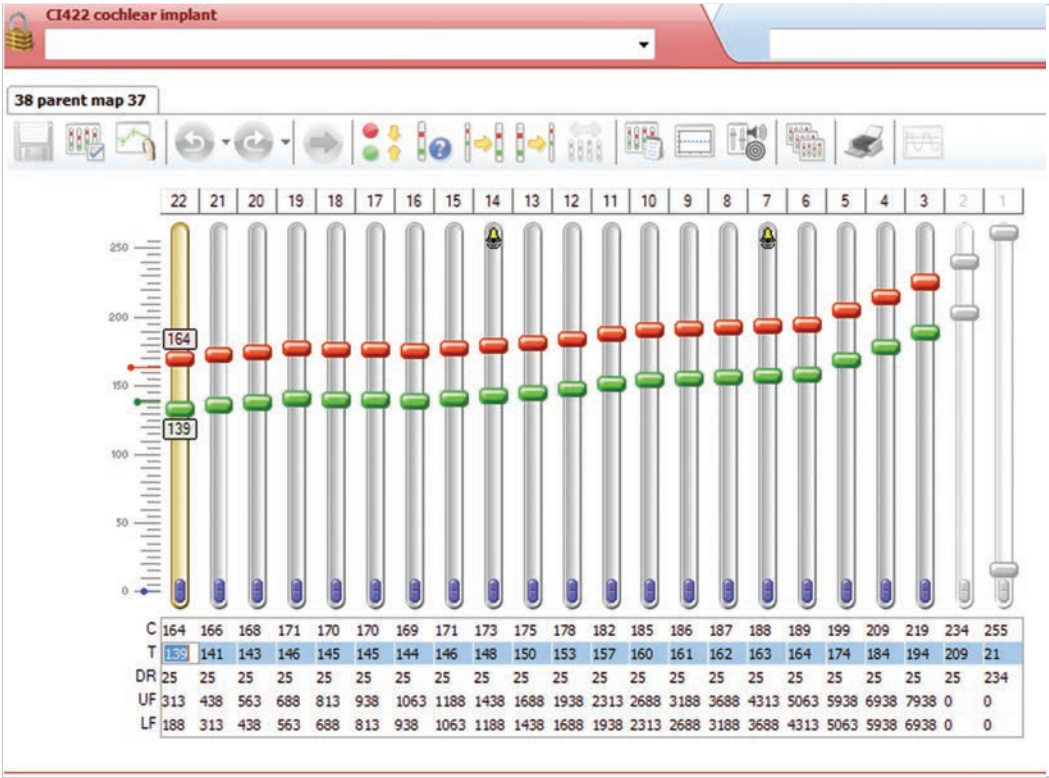

Fig. 51.1 Map programming characteristics for the right cochlear implant ear. Map T and C levels for active electrodes 3 to 22 are shown in green and red, respectively, and noted in table below map. The upper and lower frequency boundary assignments are listed in the table for electrodes 3 to 22. C, comfort level; T, threshold level; DR, dynamic range; UF, upper frequency; LF, lower frequency.

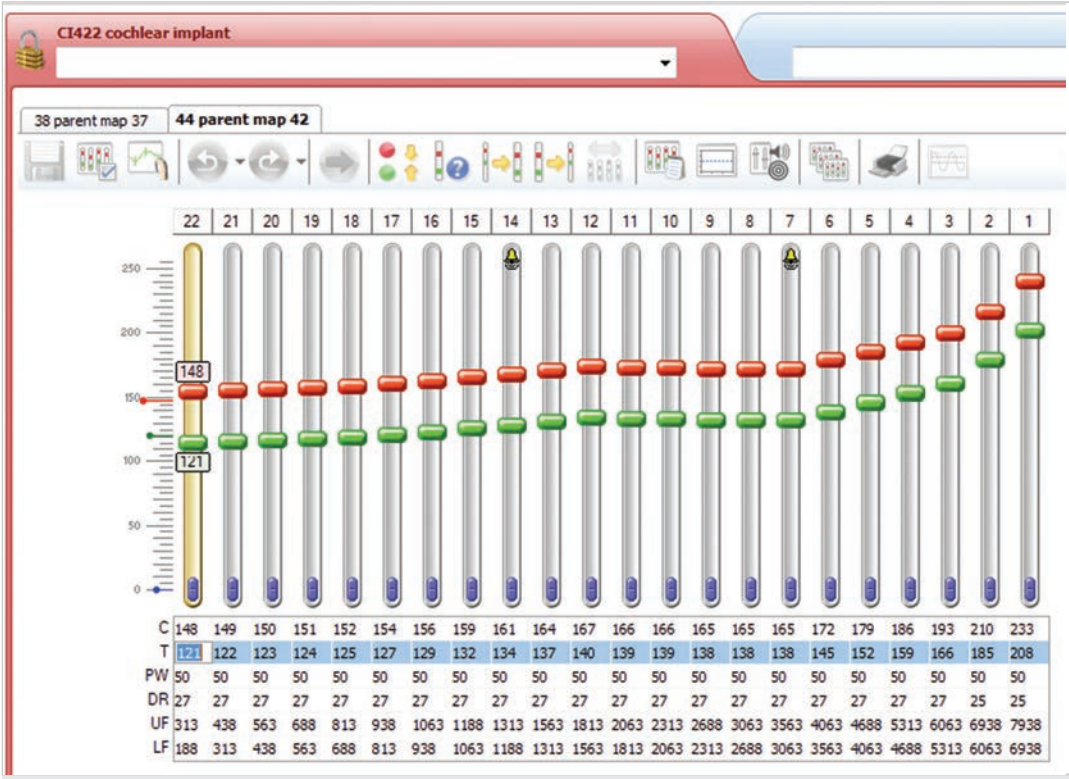

Fig. 51.2 Map programming characteristics for the right cochlear implant ear at 6 weeks post activation. Map T and C levels for active electrodes 1 to 22 are shown in green and red, respectively, and noted in table below map. The upper and lower frequency boundary assignments are listed in the

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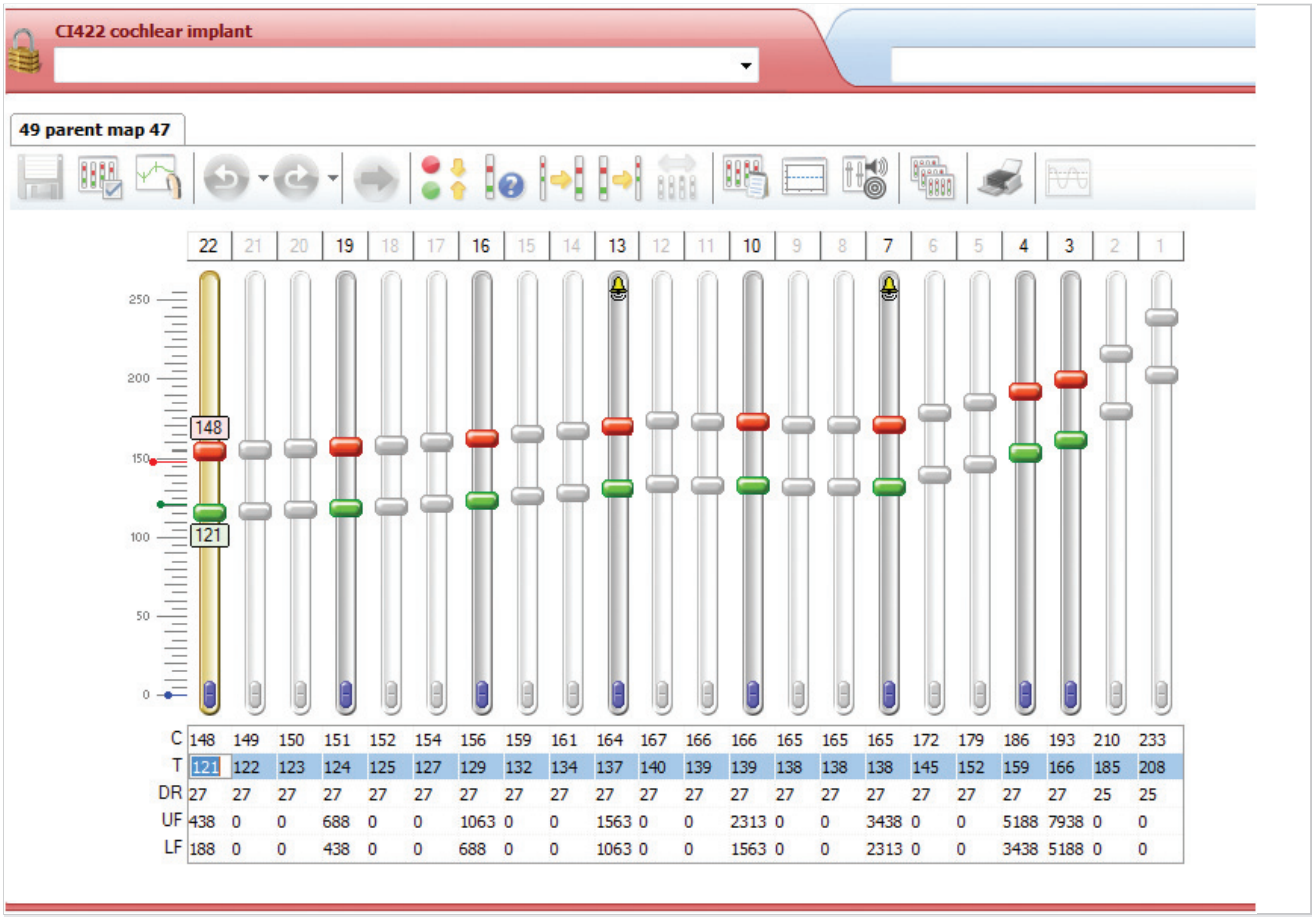

Fig. 51.3 Map programming characteristics for the experimental map for the right cochlear implant. Map T and C levels for active electrodes 3, 4, 7, 10,13, 16, 19, and 22 are shown in green and red, respectively, and noted in table below map. The upper and lower frequency boundary assignments are listed in the table for active electrodes. C, comfort level; T, threshold level; DR, dynamic range; UF, upper frequency; LF, lower frequency.

trial ensued with the experimental map at the right CI. At the end of the 2-week trial, speech perception testing was assessed using the Consonant-Nucleus-Consonant (CNC) and Lexical Neighborhood Test (LNT) word lists, presented at 60-dB SPL (▶ Table 51.4).

No significant difference in speech perception was noted between the different conditions following the 2-week trial with the experimental map at the right CI. A 2-week trial with the full map ensued. When the full map was loaded, RW was able to identify 100% Ling sounds with both the experimental and full map. This was not the case previously. After a 2-week trial with the full map, speech perception was retested.

Results indicate a doubling of score on the LNT at the right CI alone with the full map post 2-week trial (▶ Table 51.5).

Table 51.4 Speech perception, post 2-week trial with experimental map at right cochlear implant (CI)

Condition	CNC words	CNC phonemes	LNT words	LNT phonemes
Left CI alone	48%	77%		
Both CIs (Right = experimental map)	48%	75%		
Both CIs (Right = full map)	44%	73%		
Right CI, experimental map			14%	35%
Right CI, full map			16%	45%

Abbreviations: CNC, consonant-nucleus-consonant; LNT, lexical neighborhood test.

51.3 Questions for the Reader

1. Why was RW remapped at 6 weeks post initial activation? Explain rationale.
2. What other programming changes could have been made to influence compliance levels?
3. Why is speech perception testing essential for this case?

51.4 Discussion of Questions

1. **Why was RW remapped at 6 weeks post initial activation? Explain rationale.**
   A mapping adjustment was needed for two reasons. First,

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Table 51.5 Speech perception, post–2-week trial with full map at right cochlear implant (CI)

Condition	CNC words	CNC phonemes	LNT words	LNT phonemes
Both CIs (Right = full map)	50%	79%		
Right CI, full map			34%	53%

Abbreviations: CNC, consonant nucleus consonant; LNT, lexical neighborhood test.

adjustment. Second, remapping was warranted because C levels closely approached voltage compliance limitations. Approaching or exceeding voltage compliance levels can create a distorted signal because increases in current may not be able to be delivered by the implant at some electrodes, and channel interaction is also more likely.

2. **What other programming changes could have been made to influence compliance levels?**
   In this case, pulse width was widened. Another change that may affect voltage compliance limitations is the use of alternative battery options (e.g., rechargeable batteries may provide more voltage capacity than disposable zinc-air batteries).

3. **Why is speech perception testing essential for this case?**
   While important, aided detection only indicates audibility. Speech perception helps us understand the clarity of RW's signal. Several layers of speech perception were used throughout the case from detection of Ling sounds to recorded CNCs. Speech perception was also used as the outcome measure to compare the experimental and full map.

51.5 Final Diagnosis and Recommended Treatment

Based on improved speech perception scores, a full map was recommended. It is possible less information was necessary initially, but given time, introducing more information no longer compromised the signal or clarity. It is also possible that RW needed more time to acclimate to the new program containing a stimulus with a longer pulse width. Implant detection was verified with good audibility using the full map at the right CI (▶ Table 51.6).

Table 51.6 Implant thresholds, post 2-week trial with full map at right cochlear implant (CI)

Condition	250 Hz	500 Hz	1,000 Hz	2,000 Hz	4,000 Hz	6,000 Hz
Right CI thresholds	25 dB	25 dB	25 dB	30 dB	30 dB	25 dB

Table 51.7 Speech perception, 10 months post initial activation

Condition	CNC words (%)	CNC phonemes (%)
Both Cis	80	90
Right CI	72	89
Left CI	68	88

Abbreviations: CI, cochlear implant; CNC, consonant nucleus consonant.

51.6 Outcome

RW has since used a full map at her right CI. Speech perception completed 10 months post initial activation at the right CI indicates continued improvement (▶ Table 51.7).

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52 A Case of Pediatric Hyperacusis

Jenne Tunnell

52.1 Clinical History and Description

DR was a 5-year-old boy referred to audiology by his psychologist to evaluate and treat for hyperacusis. At his initial visit, his mother rated his perception of hyperacusis as 4/10 on a severity scale (10 being the most severe) (▶ Fig. 52.1). He had no medical, neurological, or traumatic history that would predispose him to hyperacusis. His sleep patterns were age-appropriate, and his mother denied a predisposition to anxiety or depression. He wore hearing protection (plugs/muffs) when exposed to dangerously loud noise levels, but not otherwise. His reactions to moderate- to high-level sounds varied; he might withdraw from the situation, refuse to participate, cover his ears, and sometimes he would cry. It would take him 5 to 60 minutes to recover from adverse sound exposure.

The following is a list of sounds or types of sounds that were bothersome to DR (in order of severity, starting with the most severe): fire trucks, fire alarms, motorcycles, gunshots, monster trucks, semi-trucks, and loud voices. DR's mother reported that she stayed at home so DR did not have to attend daycare. She reported that the home was quiet and that the TV volume is low. DR reportedly did not tolerate noisy children or adults with loud voices.

DR's mother also reported that he had none of the following characteristics or traits: high-risk indicators for hearing loss, history of otitis media or any chronic ear or upper respiratory disease, predisposing medical or neurologic history, or exposure to loud noise or ototoxic agents.

52.2 Initial Evaluation

At the first assessment, conditioned play audiometry was performed under insert earphones. Results were repeatable and reliable and showed hearing within normal limits bilaterally (▶ Fig. 52.2). There was a slight asymmetry in the high frequencies as the right ear responses were as low as –10 dB at 6,000 Hz. Speech reception thresholds were 5 dB for each ear. DR's most comfortable listening level for speech was 35 dB HL for each ear. Acoustic immittance measures showed normal type A tympanograms, suggesting intact and appropriately mobile tympanic membranes. DR's mother denied any significant history of recurring otitis media.

Frequency-specific loudness discomfort levels (LDLs) were measured using Jastreboff's recommended measurement protocol,[1] which calls for the clinician to present a tone at the child's hearing threshold and increase the presentation level in 5-dB increments. This procedure is completed with pure tones at octave frequencies from 250 to 8,000 Hz. Jastreboff recommends measuring each frequency twice, and taking the second measurement, as this is likely the more accurate of the two. He stresses the importance of ensuring that the children understand that they have the control to stop the testing at any moment. The clinician established trust with DR prior to testing, and they practiced "stop" behaviors so that DR understood that he was in control of how loud the sounds would ultimately become. DR's responses are shown in ▶ Table 52.1.

52.3 Interpretation and Counseling

DR exhibited abnormal sensitivity to sound. Normal comfort levels for speech should occur at 60 to 70 dB SPL. Typically, children allow 90 dB HL or louder for frequency-specific LDLs. The test results were explained to DR's mother, and she was given a copy of the findings. She was counseled about hyperacusis and sound therapy as well as habituation exercises that should be done in tandem with behavioral health visits. She was provided with information about web sites with links to downloadable sound therapy MP3 s. An appointment was made for follow-up for further evaluation and to establish a plan of care.

52.4 Treatment

52.4.1 What's Been Done Before?

Stiegler and Davis[2] recommended that, when treating children with hyperacusis, simultaneous supports should be provided by caregivers to show that the sounds and environment are safe for the child. Koegel et al[3] successfully treated children with autism with a similar "successive approximation" approach, exposing children first to sounds in the distance and gradually bringing them closer.

52.4.2 Treatment Plan

▶ Fig. 52.3 outlines the treatment plan for DR, which was given to his mother at the first session.

52.5 Additional Testing

52.5.1 Session #2: Outcomes and Further Assessment

During the second session, the treatment plan was reviewed with DR's mother, and she reported that she had not had a chance to give the sound aversion diary to his teacher yet. She also reported that she did not feel it was necessary to follow up with the psychologist at present. She reported that all providers now had a good understanding of hyperacusis. She did keep a journal of aversion behaviors for a typical week and reported that the only noticeable aversion behavior, other than loud voices, was a church program where the preschool children at the church were putting on a very noisy Christmas show (sample journal entry provided in ▶ Fig. 52.4). At that point, DR wanted to leave the church; she reacted by comforting him, reminding him why they were there, and telling him that he had to stay. DR had been lis-

comfortable listening level using the iPad with headphones. He

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