Case of Recurrent, Reversible, Sudden Sensorineural Hearing Loss in a Child

Michael P. Gorga*
Patricia G. Stelmachowicz*
Steven M. Barlow†
Patrick E. Brookhouser*

Abstract

This paper describes audiologic, electrophysiologic, and medical test results for a now 10-year-old girl who has had 45 episodes of reversible, sudden sensorineural hearing loss over the last 8 years. Episodes have lasted from 6 to 72 hours and often have been accompanied by a mild illness. Acoustic immittance measures have been consistent with normal middle-ear function with the exception of absent ipsilateral and contralateral acoustic reflexes. Mechanically evoked perioral reflex activity was markedly asymmetric following lower lip stimulation. The asymmetry of R1 activation between right and left side lower lip inputs raises questions about the integrity of central connections within the brain stem, including inter-nuncial pathways coursing between trigeminal sensory relay nuclei and the facial motor nucleus. An electrocochleographic evaluation revealed cochlear microphonic but absent or markedly abnormal whole nerve action potentials. Auditory brainstem responses (ABR) have been either absent or poorly formed and significantly delayed, regardless of hearing sensitivity. Middle and late auditory evoked potentials were essentially normal. Both transient-evoked and distortion-product otoacoustic emissions were present regardless of peripheral auditory sensitivity. All medical tests have been essentially normal. Although no definitive diagnosis has been reached, beta blockers have been used with some success. Taken together, these data document a very unusual case of fluctuating hearing loss. The electrocochleographic and otoacoustic emission data suggest that the outer hair cells are functioning normally and that the loss is not cochlear in origin. The ABR evaluation, mechanically evoked trigemino-facial cutaneous reflex, and acoustic reflex data point to a neural, perhaps brainstem origin; however, all other tests have failed to reveal any abnormality or underlying mechanism to explain the unusual pattern of hearing loss.

Key Words: Auditory brainstem response (ABR), auditory late potentials (ALRs), electrocochleography, middle latency response (MLR), otoacoustic emissions, perioral reflex, reversible sensorineural hearing loss, sudden sensorineural hearing loss

This paper describes audiologic, electrophysiologic, and medical test results for a young girl with complaints of sudden hearing loss, first noted when she was approximately 2 1/2 years old. Over the past several years, we have documented this hearing loss in terms of the number of episodes, its severity, reversibility, the time course over which recovery to normal hearing occurs, the pattern of recovery for both speech and pure tones, and the presence of any other symptoms associated with the episode. Although sudden hearing loss is unusual in a child, its complete reversibility is extremely rare. In the discussion to follow, we will describe our observations with this child, including test results from a variety of different measures. While we have ruled out many causes for her symptoms of hearing loss, the underlying mechanisms remain undetermined.

GENERAL BACKGROUND

This patient first came to our clinic when she was 4 years old. Her mother, an excellent
historian, reported that the patient had suffered six episodes of intermittent sudden hearing loss over the prior 2 years; however, only one of these episodes was documented audiometrically. The audiometric data for this episode, obtained at another facility, revealed a profound sensorineural hearing loss. Her mother reported that complete recovery occurred within approximately 24 hours of onset. No other exceptional events were reported to have occurred concurrently with these episodes, with the possible exception of a low-grade fever.

Prior to her initial visit at the Boys Town National Research Hospital, the patient had undergone bilateral middle-ear explorations in search of perilymphatic fistulas. Although negative, prophylactic connective tissue grafting of labyrinthine windows was performed. Two months after the surgery, another episode occurred, this time accompanied by a headache. At that time, a variety of tests were performed, again at another facility. Reportedly, there was no high-level click-evoked auditory brainstem response (ABR) in the left ear and only a questionable wave I following high-level stimulation of the right ear. Tympanometric findings were normal but both ipsilateral and contralateral acoustic reflexes were absent bilaterally. Routine blood tests, a neurologic evaluation, and a computed tomography (CT) scan (without contrast) were reported as normal.

**AUDIOMETRIC FINDINGS**

When first seen by us for an audiologic evaluation, test results were obtained using conditioned play audiometric techniques and were felt to be of good reliability. Findings revealed essentially normal hearing sensitivity and normal speech-recognition ability bilaterally. There was no evidence of decreased speech recognition at high intensities. Immittance measures revealed normal middle-ear function with the exception of absent ipsilateral and contralateral acoustic reflexes for both ears.

Approximately 2 months later, the patient's parents noticed that she did not appear to respond normally to sound upon awakening in the morning. She also exhibited a low-grade fever (101° F) and complained of a mild headache localized at the back of the head. As shown in Figure 1, audiologic results revealed a severe-to-profound, bilateral, sensorineural hearing loss. Due to the degree and configuration of hearing loss, speech-recognition measures were not obtained. Acoustic immittance measures were consistent with normal middle-ear function with the exception of absent acoustic reflexes, as noted previously. The patient was hospitalized at this time and a series of medical tests were performed. During a routine check of her vital signs (approximately 21 hours after onset of symptoms), a nurse noted that she appeared to be responding more readily to sound. Hearing was re-evaluated 4 hours later and found to be well within normal limits for both pure tones and open-set speech materials (Phonetically Balanced Kindergarten List, PBK-50). Hearing was re-evaluated 8 hours later and again was found to be completely normal.

Following another episode 18 days later, the patient's mother was given a portable audiometer and the necessary instructions to monitor hearing sensitivity at home on a daily basis. The intent was to determine if smaller fluctuations in hearing sensitivity occurred between the more marked episodes that had been documented clinically. Hearing was screened daily at a level of 15 dB HL (ANSI, 1989) at octave frequencies from 500 to 8000 Hz for a period of 2 months. With the exception of the one episode described below, the patient demonstrated no changes in hearing sensitivity at home on a daily basis.

![Figure 1](image_url)
At 4 1/2 years of age, the patient's mother called in the afternoon to indicate that another episode possibly was in progress. On the previous evening, the patient experienced difficulty hearing subjectively but had passed the pure-tone audiometric screening performed at home by her mother. This particular episode was well documented in terms of the time course of events, as shown in Figure 2. At 2:30 PM, hearing sensitivity was found to be within normal limits for pure tones but speech-recognition ability was markedly impaired bilaterally. She was re-evaluated at 4:00 PM, at which time her pure-tone hearing sensitivity was still in the normal to borderline-normal range. However, her performance on the same open-set speech-recognition test had deteriorated further. A closed-set test was administered (Word in Picture Identification, WIPI), for which test scores of 56 percent and 68 percent were observed for the right and left ears, respectively. Three hours later, responses to pure-tone stimuli revealed a mild sensorineural hearing loss, but due to inconsistent response patterns, test reliability was judged to be poor. Speech-recognition ability for closed-set materials was essentially nonfunctional. By 8:00 AM on the following morning, pure-tone hearing sensitivity had decreased into the profound hearing loss range, but by 11:30 AM, both hearing sensitivity and speech-recognition ability began to improve. By 8:00 AM of the next day (roughly 48-60 hours from when symptoms first appeared), hearing sensitivity for both pure tones and open-set speech materials had returned to within normal limits.

Since 4 years of age, this patient has had approximately 45 episodes of recurrent, reversible sensorineural hearing loss. The duration of the episodes has ranged from as short as 6 hours to as long as 72 hours, with an average duration of 30 hours. Tympanograms have always been indicative of normal middle-ear function, but acoustic reflexes have consistently been absent for both contralateral and ipsilateral stimulation. In most instances, the episodes have been accompanied by some type of mild illness, such as a headache, gastrointestinal disturbance, documented strep infection, chicken pox, sore throat, or hives. Often, a low-grade fever and headache have been present during an
episode. There have been no reports of dizziness or blurred vision and vestibular findings have always been normal.

TRIGEMINAL AND FACIAL NERVE TESTING

The integrity of the child's trigeminal and facial nerve pathways was assessed using the perioral reflex modulation paradigm (Barlow, 1991; Barlow et al, 1993). As shown in Figure 3, a servo-controlled linear motor was used to present mechanical inputs to the lip vermillion (1400 microns displacement delivered at 6 taps per second). Mechanically evoked electromyographic (EMG) responses were sampled from bilateral sites overlying the orbicularis oris superior (upper lip) and the orbicularis oris inferior (lower lip) using 4-mm diameter Ag/AgCl surface electrodes.

Mechanical inputs delivered to either the left or right half of the upper lip vermillion were effective in driving the short latency (12.7–13.5 msec) R1 component of the perioral reflex as shown in the integrated and signal-averaged (N = 64 sweeps) EMG records for the upper lip recording sites (Fig. 4). As expected, greater R1 activity was found ipsilateral to the stimulus site and was replicable from the first to the second stimulus set. Low-level R1 activation was apparent in both lower lip recording sites.

In contrast to upper lip stimulation, an unexpected asymmetry in the pattern of R1 activation was found following lower lip stimulation. As shown in Figure 5, mechanical inputs delivered to the right half of the lower lip vermilion were ineffective in driving the

Figure 3 Line drawing of the mechanical stimulator in relation to the lip vermillion. The linear motor, operating under a position servo, was suspended in from the perioral region by a Zeiss articulating microscope arm. The skin contactor of the stimulator was placed against the vermilion skin of the lower lip. Approximate electrode locations are shown for orbicularis oris superior (OOS) and orbicularis oris inferior (OOI).
short latency reflex component at the lower lip (right) recording site. On the other hand, inputs to the left half of the lower lip yielded replicable R1 responses at the ipsilateral lip recording site. This pattern of R1 asymmetry was replicated four times over a period of 2 years. The peculiar asymmetry in R1 activation between right and left side lower lip inputs raises questions about the integrity of central connections within the brain stem mediating information flow from trigeminal afferents to the facial motor nucleus.

These data are particularly interesting in light of the fact that voluntary activation of facial muscles during speech yields bilateral and symmetric EMG interference patterns in this patient. Thus, it appears that descending inputs to the facial motor nucleus by way of corticobulbar projections yield balanced activation of the circumoral musculature during speech and smiling. The results of the perioral reflex studies are consistent with the notion that access to the circumoral musculature by way of the trigeminal system is degraded with likely involvement of brainstem connections between trigeminal sensory relays and the facial motor nucleus.

**AUDITORY ELECTROPHYSIOLOGIC AND ACOUSTICAL MEASURES**

**Auditory Evoked Responses**

An ABR evaluation was first performed during an episode when profound hearing loss was present. Clicks were presented at a rate of 13 per second and at a level of 80 dB nHL (116 dB peak SPL). Not surprisingly, no replicable response was observed from either ear, which is consistent with the magnitude of the hearing loss that was present at the time of the test.

Figure 6 shows examples of auditory electrophysiologic response waveforms when hearing was completely normal. The top row of panels represents ABR recordings, the middle row shows middle latency responses (MLRs), and the bottom row shows auditory late potentials (ALRs). Data for left and right ears are shown in the left and right columns, respectively. For ABR recordings, stimulus and recording conditions were identical to those used when the child was symptomatic.

An extremely abnormal ABR was evident when stimulating either ear with high-level clicks. In the left ear, wave I and possibly wave
V were observed, with a prolonged I–V interval. No repeatable components could be measured for the right ear. These findings are surprising in view of the fact that normal hearing was present at the time of the test. Similar results have been obtained on numerous other occasions, with only slight variations in the pattern of abnormal responses observed. More often than not, we have been unable to measure wave V in either ear, even in response to high-level clicks.

The presence of poorly formed ABR waveforms or their complete absence in the presence of normal hearing represents an abnormal finding and would suggest neuropathy affecting auditory brainstem pathways bilaterally. The bilateral nature of these abnormalities further suggests that the underlying process(es) is diffuse. However, radiologic evaluations (including CT scans and magnetic resonance imaging [MRI]) have revealed no abnormalities (see below for more details).

The middle row shows MLRs, recorded with a response bandwidth of 10 to 1500 Hz. These responses were elicited with clicks presented at a rate of 9 per second at an intensity of 60 dB nHL. These responses appear to be essentially normal (see Hall, 1992 for a review). The bottom row shows recordings of ALRs. These data were obtained with a response bandwidth of 1 to 30 Hz and were elicited with 80 dB nHL clicks presented at a rate of 0.9 per second. These responses also appear to be within the normal range (see Hall, 1992 for a review). Assuming that both MLRs and ALRs are generated at higher levels in the auditory nervous system, these electrophysiologic data would argue that the dysfunction is localized to the brain stem. These data also indicate that abnormal ABRs do not necessarily preclude the generation of MLRs and ALRs, suggesting some independence among these responses.

**Electrocochleography**

At a time when hearing was completely normal and no ABRs were observed, an electrocochleographic evaluation was performed using a noninvasive tympanic membrane electrode (Lilly and Black, 1989). Clicks, presented at a rate of 8 per second, were used as eliciting stimuli. Responses were filtered between 1 and 1500 Hz. Figure 7 shows examples of response waveforms for each ear. Responses were observed that appeared to be cochlear microphonic (CM) and not stimulus artifact, based upon two observations. Since stimuli were delivered via insert
earphones, there is a 0.9-msec acoustic delay between the electrical signal and the arrival of the acoustic signal in the ear canal. If these waveforms represented electrical artifact, they should occur without delay, which was not the case. We do not believe that the responses on the left side were neural in origin because they reversed with stimulus polarity. This polarity sensitivity would not be expected if the recorded waveform represented neural events; however, it is entirely expected of a response that follows the stimulus waveform, such as CM. As a result, we believe that these responses suggest that hair cell function is present on both sides. Complete cancellation was not observed on the right side when responses to rarefaction and condensation clicks were added, suggesting that perhaps a neural response was present. However, response morphology is not normal. In addition, these latencies would be extremely long for wave I at this intensity, yet the entire response is over within 4.1 msec (after taking into account the 0.9-msec acoustic delay introduced by the insert earphones) long before wave V is expected. In fact, nothing resembling later components (i.e., wave V) was observed.

**Otoacoustic Emissions**

While the observation of cochlear microphonics bilaterally supports the presence of hair cell function, otoacoustic emissions (OAEs) also can be used to determine whether outer hair cells are functioning normally. OAEs are thought to represent a byproduct of normally occurring nonlinear mechanical properties within the cochlea. These nonlinear properties are highly dependent on the presence of normal outer hair cells. In the presence of even mild hearing loss (presumably accompanied by outer hair cell damage), these responses disappear (e.g., Kemp et al, 1986; Lonsbury-Martin et al, 1991; Probst et al, 1991). Thus, it would appear that normal functioning outer hair cells (and, therefore, normal hearing) would be prerequisites for the observations of these responses.

Figure 8 provides examples of distortion product otoacoustic emissions (DPOAEs) when hearing was completely normal (left column) and when severe-to-profound hearing loss was present (right column). The audiograms are represented in the top row of panels. The details of these measurements are described elsewhere (Gorga et al, 1993a) and will be reviewed here only briefly. DPOAEs were measured using the CUBDIS system, which runs software (Allen, 1990) on an Ariel DSP+16 signal-processing board and uses associated hardware provided by Etymotic Research (ER-10B low-noise microphone system, ER-2A insert earphones). DPOAEs were elicited with primary frequencies swept downward from 8000 to 500 Hz (3 points per octave), primary frequency ratios (f2/f1) of 1.2, and primary levels of 65 and 50 dB SPL for f1 (the lower frequency primary) and f2 (the higher frequency primary), respectively. The shaded area represents the range of normal values obtained from 80 subjects with audiometric thresholds of 20 dB HL (ANSI, 1989) for octave frequencies from 250 to 8000 Hz. The middle row of panels provides estimates of both response and noise amplitudes (in dB SPL) and the bottom row of panels provides the ratio of these two numbers (DPOAE/noise) in dB. Clearly, the patient's cochleae produced DPOAEs that were well within the range observed in subjects with normal hearing. This pattern was observed, regardless of whether hearing was normal (left column) or severely impaired (right column).

Figure 9 shows transiently evoked OAEs (TEOAEs) for this child obtained at the same times that the DPOAEs (shown in Fig. 8) were measured. Thus, the TEOAE data in the left column were obtained when hearing was normal and the data in the right column were obtained when hearing was impaired. These TEOAEs were measured using the ILO88 Otodynamic Analyzer, operating at its default settings. The details of these measurements can be found elsewhere (Gorga et al, 1993b; Prieve et al, 1993) and again will be reviewed only briefly. TEOAEs were obtained using the "nonlinear paradigm" in which responses are measured to sets of four clicks, with the first three clicks presented at about 70 dB SPL and of
Figure 8  Audiometric and DPOAE data when the child's hearing was normal (left column) and when hearing loss was present (right column). Audiometric data are provided in the top row of panels. DPOAE and noise amplitudes are shown in the middle panels for both left and right ears. DPOAE/noise are shown in the bottom panels. Shaded areas represent the range of values observed in 80 normal-hearing subjects, defined as audiometric thresholds of 20 dB HL or less for octave frequencies from 250 to 8000 Hz.

Figure 9  TEOAEs obtained when hearing was normal (left column) and when hearing was impaired (right column), as shown in Fig. 8. TEOAEs were analyzed as broadband responses (BB) and within octave bands centered at frequencies from 500 to 4000 Hz. Top row: TEOAE and noise amplitudes for left and right ears; middle row: TEOAE/noise; bottom row: reproducibility. Shaded areas represent the range of values obtained from 80 normal-hearing subjects, defined as audiometric thresholds of 20 dB HL or less for octave frequencies from 250 to 8000 Hz.

MEDICAL TESTS

Evaluation

Otoneurologic, allergy, neurologic, and genetic evaluations failed to reveal any obvious etiology for the hearing loss. Both CT and MRI scans with contrast of the temporal bones and posterior cranial fossa were interpreted as normal. An EEG was also interpreted as negative. Laboratory evaluations that were either negative or within normal ranges included complete blood count, FTA-abs, creatinine, lipids, urine analysis, serum electrolytes, blood glucose, and thyroid studies. Quantitative immunoglobulins were normal except for a slightly depressed IgG. Baseline and convalescent viral serology, as well as pharyngeal
viral cultures obtained during an acute exacerbation of her hearing loss, were negative. A lumbar puncture was performed and myelin basic protein was among the normal findings. Oligoclonal banding was read as equivocal. The patient refused an additional lumbar puncture and this finding appears to have had no clinical significance in view of the negative MRI and the subsequent observation of the course of the disease during the intervening seven years. No evidence of systemic autoimmune disorders was found, and laboratory evaluations for possible immune-mediated inner ear disease at two outside laboratories proved negative.

Treatment

Although no definitive diagnosis has been reached, the assumption has been that the underlying problem is vascular in nature and may be associated with migraine headaches. The patient has been treated with a beta-adrenergic receptor blocking agent for the past 3 1/2 years. In the year prior to this treatment, she experienced 13 episodes with an average duration of 35 hours. Since treatment began, the number of episodes has dropped to 6 per year, with an average duration of 24 hours. Although these findings may lend some credibility to the diagnosis of migraine, it remains to be determined if this diagnosis is correct.

In summary, medical test results did not reveal any obvious cause for this patient’s auditory symptoms. In view of the bilateral nature of the hearing loss, the lack of apparent involvement of other systems is surprising.

SUMMARY

There have been reports of children having abnormal electrophysiologic responses that are unexpected, given their pure-tone sensitivity (Worthington and Peters, 1980). These children, however, had other auditory perceptual problems. Similarly, Stach et al (1993) reported on a child who had abnormal ABRs but normal emissions. However, under many of the stimulus conditions shown for this child, more normal ABR waveforms were observed, including wave V at least at the highest test intensities (see for example, the right ear data in Figs. 3A and 3B, Stach et al, 1993). Furthermore, this child had other neurologic problems, which were associated with severe hydrocephaly. The child described in the present paper is unique in that she is developmentally and neurologically normal in all other regards. This child has recurrent, reversible episodes of sudden sensorineural hearing loss of unknown etiology. The relatively short time course of episodes, the frequency of their occurrence, and the complete reversibility of the hearing loss are extremely unusual. Invariably, her ability to understand speech decreases prior to the time when pure-tone sensitivity deteriorates. Pure-tone sensitivity also recovers more quickly than her ability to understand speech.

Tests of cochlear function (regardless of auditory status), such as TEOAEs, DPOAEs, and electrocochleography, consistently have been normal, thus suggesting that the source of the hearing loss is not peripheral. In contrast, those tests that presumably assess brainstem levels of the nervous system have been abnormal, even when she is symptom free. Acoustic reflexes, ABRs, and mechanically evoked trigemino-facial reflexes all fall outside normal limits, even when hearing is completely normal. These findings point to the brain stem as the most likely site of lesion. Responses thought to assess higher levels in the auditory nervous system (i.e., MLR, ALR) appear normal.

The child has been treated under the assumption that the source of the dysfunction is vascular (i.e., vasospastic), with some success. If this assumption is true, however, it would appear that the vascular disturbance is diffuse (both sides are affected), it does not affect cochlear blood supply (if it did, one would expect that the hearing loss would not be completely reversible and OAEs would be depressed during an episode), and some of the damage in the brain stem is likely permanent (ABRs are abnormal and acoustic reflexes are absent, even when the patient is symptom free). We are left, therefore, with a perplexing case for which no completely satisfactory diagnosis has been reached. On the other hand, the combined data from audiometric, acoustic, electrophysiologic, and medical evaluations provide a relatively complete description of where in the nervous system the disturbance is occurring.

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REFERENCES


