Considerations for Cochlear Implantation of Children with Sudden, Fluctuating Hearing Loss

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Abstract

The histories of two pediatric patients who received cochlear implants with subsequent partial recovery of hearing in the nonimplanted ear are reviewed. One child had a sudden bilateral hearing loss, presumably secondary to autoimmune ear disease. The other child had a bilateral progressive hearing loss diagnosed as large vestibular aqueduct syndrome (LVAS). The rationale for the timing of the surgical implantation is discussed. Retrospectively, recovery of hearing in the nonimplanted ear suggests the possibility that the implant could have been delayed or eliminated as a treatment option, and that wearable hearing aids may have been appropriate. A number of factors, however, suggest the decision to implant was appropriate. Issues involved in the decision-making process of when to implant are presented and discussed.

Key Words: Autoimmune ear disease, cochlear implant, Cogan's syndrome, contralateral hearing aids, large vestibular aqueduct syndrome, Pendred syndrome, sudden hearing loss

Abbreviations: ABR = auditory brainstem response; AIED = autoimmune ear disease; CT = computerized tomography; ESR = erythrocyte sedimentation rate; FDA = Food and Drug Administration; LVAS = large vestibular aqueduct syndrome; MRI = magnetic resonance imaging; NICU = neonatal intensive care unit; NU-CHIPS = Northwestern University-Children's Perception of Speech Test; PBK-50 = Phonetically Balanced Kindergarten Test; WIPI = Word Intelligibility by Picture Identification (Test); WRS = word-recognition score

Sumario

Se revisan las historias de dos pacientes pediátricos quienes recibieron implantes cocleares, con una parcial recuperación posterior de la audición en el oído no implantado. Un niño tuvo una hipoacusia súbita bilateral, presumiblemente secundaria a una enfermedad autoinmune del oído. El otro niño tenía una pérdida auditiva progresiva bilateral diagnosticada como un síndrome de acueducto vestibular grande (LVAS). Se discuten las razones sobre la decisión de cuándo realizar la cirugía de implante. Retrospectivamente, la recuperación de la audición en el oído no implantado sugiere la posibilidad de que el implante pudiera haberse postergado o cancelado como una opción terapéutica, y que hubiera sido apropiada la adaptación de un auxiliar auditivo convencional. Un número de factores, sin embargo, sugieren que la decisión de implantar fue apropiada. Se presentan y discuten aspectos involucrados en el proceso de toma de decisiones sobre cuándo implantar.
There has been an exponential growth in the number of cochlear implants. Worldwide, there are now approximately 60,000 individuals wearing cochlear implants. This growth is justified by improvements in the quality of the devices, programming strategies, and surgical procedures. As a consequence, the criteria for implantation have been expanded for adults and children. For example, implant candidacy has been expanded to include children as young as three months and adults possessing significant residual hearing, particularly in the low frequencies (Zeng, 2004). Food and Drug Administration (FDA) regulations now specify that for children under age two, the magnitude of bilateral hearing loss must be profound, while for children between two and eighteen years, the degree of bilateral hearing loss must be severe to profound (70 dB HL or worse). Prior to implantation, amplification must be tried for a minimum of three to six months. The FDA regulations, however, do not specify when or if a child with fluctuating hearing loss should be implanted. Delaying implantation has the potential for allowing neural plastic and behavioral changes that may make subsequent restoration and development of speech and language skills more problematic. Furthermore, children (and adults) with deteriorating or fluctuating hearing loss may experience emotional problems such as depression, frustration, and anger. On the other hand, an overly rapid decision to implant could prove to be premature if hearing recovers sufficiently to the degree that acoustic amplification would have been a more appropriate treatment option. This is not to imply that acoustic amplification is necessarily superior to cochlear implantation. It is not always possible, however, to reliably or accurately predict postsurgical performance with a cochlear implant. Furthermore, full, as opposed to hybrid, implantation (short electrode cochlear implant in addition to acoustical amplification) greatly minimizes the likelihood of functional usage of that ear, pending scientific advancements in cochlear restoration.

In this paper, the authors present case histories of two children who received cochlear implants with subsequent partial recovery of hearing in the nonimplanted ear. These cases, while having different etiologies, raise a number of questions about the decision-making process that audiologists and surgeons should contemplate regarding the time course and consideration of cochlear implantation.

Please note that for both cases, only a portion of the testing was completed at the University of California, San Francisco. Most of the early data were obtained at the facilities at which the cases were initially presented. Therefore, not all of the procedures that may have contributed to the overall interpretation of the case are represented in this manuscript.

CASE 1

A four-year-old child (JC) presented to the otolaryngology clinic in late April 2002, three weeks post initial onset of an acute subjective hearing loss. Prior to the sudden decrease in her hearing, she had normal hearing and was developing speech and language at the expected pace. Her past medical history was significant for prematurity for which she spent one week in the neonatal intensive care unit (NICU) on nutritional supplementation. She did not receive any antibiotics or other medications during her NICU stay. She had normal developmental milestones, and her immunizations were current. She was not
on any medications and had no known allergies. Her family history was negative for hearing loss, and she had not traveled outside the United States. She had several hospitalizations related to juvenile rheumatoid arthritis, an autoimmune disorder that affects approximately 75,000 children in the United States. Her physical exam was significant for mildly retracted tympanic membranes and left scleral injection. She had a nonfocal neurologic exam, meaning cranial nerves II through XII (except VIII) were found to be intact. Ophthalmology reports indicated she had a recent bout of anterior uveitis. This ocular pathology is commonly found in children with Cogan’s Syndrome. She did not have interstitial keratitis, however, an even more frequent finding in Cogan’s Syndrome.

The initial audiogram indicated a profound hearing loss in the right ear, and a severe to profound loss in the left ear. Word-recognition scores (WRS) were 0% bilaterally. Tympanograms showed -280 daPa middle ear pressure in the right ear, and -320 daPa in the left ear. Acoustic reflexes and transient otoacoustic emissions were absent. The child was cooperative, and the results were considered reliable and valid. Due to the suddenness of her hearing loss, she was initially placed on prednisone at 1 mg/kg/day. An audiogram performed approximately one week following the beginning of the prednisone regimen revealed further decrease in hearing. Serial unaided audiograms and word-recognition scores from May 2002 to August 2004 are shown in Figure 1 (right ear) and Figure 2 (left ear). Please note that to prevent clutter, not all the hearing test results are shown in these figures. She also had difficulty tolerating the medication, and since her hearing was worsening, it was decided that stopping the steroids was prudent.

In addition to the audiologic testing, hematologic results indicated an elevated sedimentation rate of 80 and a positive rheumatoid factor of 29. Additional blood tests ruled out Russell’s viper venom, coagulation factors, von Willebrand’s factor, Lyme disease, and Coccioidiodes as possible causes of her hearing loss. ANA (antinuclear antibody) and ANCA (anticytoplasmic antibody) were also normal, along with liver enzymes. Lumbar puncture also showed normal CSF (cerebrospinal fluid) protein, glucose, and white and red blood cell counts. An MRI (magnetic resonance imaging) was performed to rule out inner ear malformations such as large vestibular aqueduct syndrome (LVAS). Given the patient’s history of juvenile rheumatoid arthritis, the results of the hematologic workup, and the negative MRI

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**Figure 1.** JC right ear thresholds and word-recognition scores (WRS).

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**Figure 2.** JC left ear thresholds and word-recognition scores.
findings, the etiology of her hearing loss was diagnosed as autoimmune inner ear disease (AIED). Since there are no known diagnostic tests for Cogan’s Syndrome (Olfat and Al-Mayouf, 2001), it was not possible to rule this out; however, the treatment plan would not have differed, regardless of the definitive diagnosis.

JC returned for additional audiologic testing in late May 2002. Pure-tone thresholds improved by approximately 20 dB relative to the initial test results, and tympanograms were normal bilaterally. Word-recognition scores, however, remained at 0%. She was fit with a loaner postauricular hearing aid on her left ear. With the hearing aid in place, her aided thresholds improved by 30–40 dB to 35–55 dB HL (500–4000 Hz). One week after receiving the hearing aid, an MRI showed abnormal hyperintensity of the cochlea, vestibule, and semicircular canals, possibly indicative of labyrinthitis and AIED. In early June, her hearing was retested, and results again showed a profound loss in the right ear, and severe-to-profound loss in the left ear. There was negative middle ear pressure in the left ear, but the tympanogram was normal in the right ear. Thus, the loss was considered sensorineural, at least on the right side. In addition, the audiologist, as well as the child’s mother, reported a noticeable decline in the child’s expressive speech intelligibility. On this date, a referral to the cochlear implant division was initiated. A second MRI was performed in late June because JC was experiencing headaches, nausea, and vomiting. Imaging indicated enhancement of both internal auditory canals as well as cranial nerves III, V, VI, VII, and VIII, suggesting a viral or autoimmune disorder. Steroids were again administered. Two weeks after the onset of this steroid regimen, a follow-up MRI appeared significantly improved with only faint enhancement of the cochleas and no enhancement of the cranial nerves.

In late August, hearing test results indicated a reversal of threshold acuity of the ears with profound hearing loss for the left ear and severe hearing loss in the right ear. In October 2002, six months after the onset of the sudden hearing loss, the decision was made to implant the patient’s left ear, presumably because of the greater degree of hearing loss in that ear, with a Nucleus 24C device. At that time, she was wearing a loaner hearing aid on her right ear switched from the left (because of the reversal in hearing), with aided thresholds from 35–50 dB HL and aided word-recognition score of 32% for PBK-50 (Phonetically Balanced Kindergarten Test) words presented at 55 dB HL. She was unable to correctly identify any PBK-50 words under earphones. The Nucleus 24C was mapped in January 2003, and an ACE (Advanced Combination Encoder) strategy with a 900 Hz rate and 8 maxima was employed. The child was then enrolled in an oral school for the deaf. The audiologist’s report indicated that JC’s speech and listening skills continued to improve. Aided thresholds (Figure 3) using the cochlear implant alone ranged from 35–40 dB from 750 Hz through 6000 Hz and her aided word-recognition score was 66% using the closed-set WIPI (Word Intelligibility by Picture Identification [Test]). In May 2003, aided word recognition improved to 100% for the WIPI and 82% for an open set (PBK-50s) at 55 dB HL without visual cues. In December 2003, audiometric earphone tests on her right (nonimplanted ear) were performed. Thresholds remained in the severe-to-profound range while word recognition remained at 0% for the WIPI at 105 dB HL.

JC was retested in March 2004 on the day she was to be fit with a new hearing aid for her right ear. Her unaided soundfield thresholds ranged from 70–75 dB from 500 through 4000 Hz. This suggests that her right ear had improved by approximately 20 dB in the last four months. In addition, with the new hearing aid (cochlear implant turned off), her aided soundfield thresholds (Figure 3) were 25–35 dB HL at 500–4000 Hz. Her aided word recognition for open-set words, presented at 55 dB HL, however, remained at 0%.

Two weeks following this test, word recognition was evaluated in quiet and noise (+10 dB S/N) using PBK-50 word lists presented at 55 dB HL when wearing the hearing aid (right ear) and the Nucleus 24 cochlear implant (left ear). Word-recognition scores had dramatically improved to 92% and 88%, respectively, for quiet and noise. In addition, word-recognition testing was performed with the hearing aid alone with a score of 60% and the cochlear implant alone with a score of 92%. This suggests that her improved auditory skills were primarily from the cochlear implant, but for the first time in nearly two years, the nonimplanted right ear
appeared to be making a contribution. Aided word-recognition data are shown in Figure 3. This increased contribution was confirmed three months later in June when earphone pure-tone testing of the right ear revealed hearing thresholds in the 50–65 dB HL range through 4000 Hz and word recognition of 88% at 90 dB HL using the WIPI. In August 2004, pure-tone thresholds remained stable, and the word-recognition score for closed-set PBK-50s presented at 95 dB HL remained good at 84%. At the most recent testing performed in January 2005, hearing and word-recognition scores had remained stable. An FM system was recommended to interface with her cochlear implant and hearing aid. When asked what she would prefer if she had to choose between wearing either the cochlear implant or the hearing aid or both, JC preferred both. However, she wears only the hearing aid on weekends and indicated that if she had to choose between the devices, she would choose the hearing aid over the implant because she “heard people easier.”

**CASE 2**

KI, born in 1990, first presented to an audiologist at age two and a half because of delayed speech and language development. She had no history of prior ear infections; her mother's pregnancy was unremarkable; and she was born at full term without complications from a Caesarian section. There was no history of hearing loss in the family. It was reported she was difficult to test, but behavioral testing indicated soundfield speech awareness and responses to warbled pure tones at approximately 50 dB HL. Auditory brainstem response (ABR) testing revealed click thresholds at 80 dB nHL. Transient otoacoustic emissions were absent bilaterally. Conditioned play audiometry performed six months after the initial hearing test indicated a bilateral moderate mixed loss at 250–2000 Hz (higher frequencies were not assessed). Tympanograms were shallow, but the otologic examination did not reveal middle ear infection.

A more comprehensive audiometric test was obtained two months later (September, 1993), and results indicated a bilaterally asymmetrical loss, moderate to profound in the right ear, and severe to profound in the left ear. Serial audiograms are shown for the right ear (Figure 4) and left ear (Figure 5). These high-frequency thresholds were in agreement with the earlier ABR data. Significant air-bone gaps were found in the low frequencies (unmasked bone-conduction thresholds were 5 dB at 250 Hz, 20 dB at 500 Hz, and 65 dB at 1000 Hz). Tympanograms revealed significant negative middle ear pressure, bilaterally. Ventilating tubes were placed bilaterally. Another three months passed, and KI was fit with postauricular hearing aids. Aided soundfield thresholds ranged from 25–55 dB HL at 500 through 4000 Hz (Figure 6). Low-frequency thresholds (Figures 4 and 5) were improved relative to the previous test, but high-frequency thresholds remained in the profound range. Her low-frequency hearing continued to fluctuate over the next year, and tympanostomy tubes were placed in each ear. She was enrolled in a preschool program for hearing-impaired children. By age five, word-recognition scores for the right and left ears of 60% and 78%, respectively (Figures 4 and 5), were obtained using the NU-CHIPS
Perception of Speech Test). The hearing loss stabilized over the next several years, revealing an asymmetrical loss through 750 Hz (mild in the right ear, moderate in the left ear) precipitously sloping to a severe-to-profound loss beyond 750 Hz. Tympanograms were normal once the tympanostomy tubes were exuded. The aided word-recognition score on the WIPI was 68% at 50 dB HL (Figure 6).

In February 2000, word-recognition testing revealed a significant decrease bilaterally (40% and 52% for right and left ears, respectively) using PBK-50s. Pure-tone thresholds remained stable, and aided soundfield thresholds (now with digital hearing aids) were 25–35 dB HL through 3000 Hz and 50 dB HL at 4000 Hz. Of course, these aided soundfield thresholds may have overestimated overall usable gain because their low-compression kneepoints (approximately 20 dB SPL) provided for increased gain at lower input levels. Aided word recognition, using PBK-50 word lists presented at 50 dB HL, was 44%. Although she was provided minimal initial otolaryngologic workups, at this point she was referred to a pediatric otologist, who ordered laboratory testing, ophthalmologic consultation, and a temporal bone CT (computerized tomography) scan to rule out syndromic forms of hearing loss. Radiologic and laboratory findings revealed bilateral enlarged vestibular aqueducts and Pendred syndrome.

In March 2001, word-recognition scores fell to 36% and 44% in the right and left ears,

Figure 4. KI right ear thresholds (air conduction—solid lines; bone conduction—dashed line) and word-recognition scores.

Figure 5. KI left ear thresholds (air conduction—solid lines; bone conduction—dashed lines) and word-recognition scores.

Figure 6. KI aided threshold data for hearing aid (solid lines) with word-recognition scores and cochlear implant (dashed lines) with aided word-recognition scores. CI = cochlear implant; HA = hearing aid; L = left; R = right.
respectively, and her pure-tone thresholds also decreased. She was referred for an evaluation for candidacy for cochlear implantation. The audiologist reported, “Pediatric candidacy for cochlear implantation requires a profound hearing loss bilaterally with a lack of benefit from conventional amplification. Since KI continues to receive benefit from her hearing aids, she does not qualify for cochlear implantation. However, her particular etiology results in progressively worse hearing over time. Therefore, it is likely that at some point in time she will qualify for implantation.” In October 2002, the audiologist’s recommendation proved prophetic. KI’s mother reported her daughter’s hearing seemed to have been fluctuating considerably for the past month, despite the fact there were no recent illnesses or head trauma. Pure-tone testing confirmed that her thresholds decreased significantly bilaterally (see Figures 4 and 5) and now ranged from 80–105 dB HL through 4000 Hz. Speech reception thresholds (SRT) were 105 dB HL bilaterally (as opposed to the 50–60 dB HL previously measured in 2001). Word-recognition scores using PBK-50s were 24% and 16% for the right and left ears, respectively. There were no responses to bone-conduction testing. This was the first time low-frequency bone-conduction thresholds showed such significant deficits. Tympanograms were normal. Aided soundfield thresholds are displayed in Figure 6. She was subsequently referred back for another evaluation of cochlear implantation candidacy, for which she now qualified. Six months later, in April 2003, at age 13, she received a Clarion CII 16 channel implant for her right ear, which was subsequently programmed with Clarion’s “HiResolution” strategy. Aided soundfield thresholds using the cochlear implant are shown in Figure 6.

Nine months later (January 2004), KI returned to her audiologist for routine audiometric testing. Unaided thresholds for her left ear improved to 2001 levels (moderate through 1000 Hz sloping to severe to profound through 4000 Hz (see Figure 5) with 48% word recognition. Aided soundfield thresholds with the cochlear implant indicated an SRT of 25 dB HL and warble tone thresholds from 30–40 dB HL. By comparison, Figure 6 depicts thresholds obtained using the left ear hearing aid (Widex P38) ranging from 15–35 dB HL. In June 2004, thresholds for the left ear remained stable.

KI continues to wear the hearing aid and cochlear implant (coupled to an FM system at school) with success. When asked what she would prefer if she had to choose between wearing the cochlear implant or hearing aid or both, she preferred both, but indicated that if she had to choose between the devices, she would definitely choose the hearing aid over the implant as it sounded less distorted and provided her with better speech understanding.

**DISCUSSION**

Both children continue to wear a cochlear implant in one ear and a hearing aid in the other ear. Ching et al (2001) indicated that contralateral hearing aid fittings can provide benefits in speech perception, localization, and speech production. Indeed, both children prefer wearing both devices over either one alone. In both cases, the nonimplanted ear recovered hearing sufficiently to allow for good use of acoustic amplification. One cannot rule out the possibility that the recovery of hearing in the nonimplanted ear would not have also occurred in the implanted ear. However, there is a strong likelihood since the onset of the loss or progression was simultaneous, that recovery would have been bilateral. Since the damage to the cochlea is irreversible when a fully implanted cochlear implant is inserted, the decision to implant must be carefully considered. This is particularly the case if the hearing loss is fluctuating. A number of factors should be analyzed in deciding whether a sudden, fluctuating, or progressive loss of hearing warrants cochlear implantation or whether it would be prudent to delay.

The first factor to consider is the onset of the hearing loss. For Case 1 (JC) there was a sudden-onset hearing loss. Mattox and Simmons (1977) reported a 65% spontaneous recovery rate to “functional hearing levels.” Byl (1984) reported a recovery rate of about 69%. Patients that recover a significant portion of their hearing within the first two weeks following the onset of a sudden hearing loss have a better prognosis than those who do not recover in the early stages (Ito et al, 2002). This did not happen in JC’s case despite the use of steroids. Wilson (1993) points out that it is difficult to accurately predict the
outcome of sudden sensorineural hearing loss given the low incidence and unpredictability of its natural course. Also, there are problems evaluating recovery statistics, especially for treatment protocols, because of the significant percent of patients that recover spontaneously.

There are, however, some factors that have been linked with prognosis. Patients who seek medical treatment within 7–10 days after onset of the hearing loss do better than those waiting 30 or more days. JC first reported to ENT three weeks post-onset. The severity of the initial hearing loss has some prognostic significance, in that the severity of the hearing loss is inversely proportional to the rate of recovery. Her sudden hearing loss was severe to profound. Patients with rising audiometric configurations or midfrequency loss do better than those with high-frequency or sloping hearing loss (Wilson, 1993). Her audiometric configuration was not rising. Elevated ESR (erythrocyte sedimentation rate) rates above 25 mm/hour additionally predict a poor prognosis. The hearing status of the opposite ear also plays a role in determining prognosis. Patients with a normal audiogram in the opposite ear perform better than those with hearing loss in the opposite ear. Both of JC’s ears were initially affected. Age does not appear to play a significant role in recovery from sudden SNHL except that those younger than 15 years or older than 60 years experience poorer recovery. Thus, all of these signs seem to indicate a poor prognosis for recovery. Moreover, significant recovery of hearing more than two years post-onset is highly unusual. For KI, the onset of the hearing loss was not sudden. Fluctuations in threshold occurred over the years following a period of relative stability that preceded the sudden change propelling her into candidacy for cochlear implantation.

The next factor to consider is etiology. For JC, it was believed the hearing loss was related to autoimmune inner ear disease. The differential diagnosis of this patient’s hearing loss includes juvenile rheumatoid arthritis with inner ear involvement, and atypical Cogan’s syndrome. Although McCabe (1979) first reported AIED in adults, little is known about the diagnosis and management of AIED in the pediatric population, other than that it is quite rare. When considering AIED as a possible etiology, the major features include a rapidly progressive bilateral hearing loss secondary to either systemic disease or organ-specific disease when there is reactivity against a self-antigen found in the inner ear (Ndiaye et al, 2002). The pathogenesis of immune-mediated sensorineural deafness and vestibular dysfunction is unclear but is presumed to include vasculitis of vessels supplying the inner ear, autoantibodies directed against inner ear antigenic epitopes, or cross-reacting antibodies. Autoimmune hearing loss implies that inner ear proteins are recognized immunologically as foreign or nonself. Ndiaye et al (2002) reported three cases of pediatric Cogan’s syndrome that share some features with this case. Examples include ophthalmologic and rheumatologic involvement with sensorineural hearing loss. Veldman et al (1984) reported a case of sudden onset bilateral SNHL without vestibular symptoms in a 14-year-old girl. She, like JC, also complained of fatigue, arthralgia, and malaise, and was found by an indirect granulocyte phagocytosis test and C1q binding assay to have circulating immune complexes that later disappeared after response to steroid treatment. Cogan’s syndrome (CS) is an autoimmune disease of the cornea and vestibuloauditory apparatus that was first described by Cogan, an ophthalmologist, in the 1940s. It occurs primarily in young adults (average age of onset 22–29 years) and typically presents with interstitial keratitis (IK) and Meniere’s-like attacks of vertigo, ataxia, tinnitus, nausea, vomiting, and hearing loss that develop within several months of each other. Hearing fluctuation in CS coincides with disease exacerbations and remissions. It often culminates in deafness.

The cause of CS is unknown. Temporal bone histopathologic studies done at autopsy of patients with CS are characterized by chronic inflammation including infiltration of the spiral ligament with lymphocytes and plasma cells, endolymphatic hydrops, degenerative changes in the organ of Corti, and demyelination and atrophy of the vestibular and cochlear branches of cranial nerve VIII. There are no criteria currently established for the diagnosis of CS. The general thought is that diagnosis requires clinical signs of both eye and inner ear inflammation. Comprehensive workup includes an audiogram and laboratory tests including CBC (complete blood count), ESR,
and RPR (rapid plasma regain). Imaging including MRI and/or CT should be done primarily to rule out cerebellar-pontine angle tumors and other disorders. MRI with gadolinium may show enhancement of vestibular and cochlear structures. The cornerstone of therapy for sudden hearing loss, AIED, or CS is rapid deployment of steroids. Most authors suggest using prednisone 1 mg/kg for two to four weeks with a subsequent rapid taper for cases of complete resolution and slow taper for those with incomplete response. The best outcome is in patients in whom therapy begins shortly after the onset of symptoms. This did not prove helpful in JC’s case. Given the fact that JC’s nonimplanted ear did not show recovery of hearing for nearly two years postsurgery and KI’s nonimplanted ear did not show recovery of hearing for nearly a year postsurgery, there was no reason to expect hearing was going to recover. However, one must be cognizant of the fact that significant fluctuations in hearing can continue throughout the course of the disease. Hurley and Sells (1997) reported on one patient with AIED whose hearing had substantially improved 14 months postonset, only to decrease again 23 months postonset.

The etiology responsible for KI’s hearing loss was deemed to be Pendred syndrome, LVAS, or both. Pendred syndrome is an autosomal recessive inherited disorder. Obligatory features are profound deafness in childhood and defective organic binding of iodine in the thyroid gland. Pendred syndrome is characterized by sensorineural hearing loss, goiter, and an abnormal TSH (thyroid stimulating hormone) or perchlorate discharge test. It is possible that the widened vestibular aqueduct is responsible for the increase in hearing impairment. Aside from the branchio-otorenal syndrome, Pendred syndrome is the only other known genetic disorder with a widened vestibular aqueduct. Valvassori and Clemis (1978) indicated the loss was progressive in 46–65% of cases. It has long been recognized that there is an association between sensorineural hearing loss and enlarged vestibular aqueducts as demonstrated on histological studies. In 1978, Valvassori and Clemis demonstrated enlarged aqueducts radiologically for the first time on inner ear tomography. They coined the term “large vestibular aqueduct syndrome” to describe the condition. The vestibular aqueduct was defined to be enlarged if the diameter was greater than 1.5 mm. Bilateral involvement is twice as common as unilateral involvement. There is a female predominance. The hearing loss is acquired rather than congenital and typically progresses as a stepwise decrement often triggered by minor head trauma. In KI’s case, however, it is uncertain whether the hearing loss was congenital since it was first discovered at 30 months of age.

It is hypothesized by Tan (1999) and Levenson et al (1989) that the enlarged patent endolymphatic duct places the endolymphatic circulation at risk of reflux from the hyperosmolar endolymphatic sac content. Sudden fluctuation in cerebrospinal fluid pressure (e.g., during minor trauma to the head) could compress the dural envelope surrounding the lymphatic sac. This could forcibly push the hyperosmolar fluid within the endolymphatic sac through the dilated duct and into the endolymphatic circulation. The hyperosmolar content would damage the neuroepithelium of the cochlea, leading to hearing loss. The abnormal transmission of cerebrospinal fluid pressure fluctuations to the inner ear via the wide vestibular aqueduct could also be a contributor to hearing loss. Normally, the inner ear is guarded from any rapid intracranial pressure changes by the narrowness of the vestibular and cochlear aqueducts. When the vestibular aqueduct is enlarged while the cochlear aqueduct is normal in size, any rapid fluctuation in cerebrospinal fluid pressure, as in the case of head trauma, might create transient force imbalances across the cochlear partition. As a result, shearing forces could be created causing damage to the membranous labyrinth. Arcand et al (1991) found an incidence of LVAS of 12% of the children referred for audiological assessment of sensorineural hearing loss. Levenson et al (1989) summarized the findings of 12 children with LVAS. They report that the hearing loss associated with LVAS is acquired and progressive. Valvassori and Clemis (1978) indicated the loss was progressive in 46–65% of cases.

The hearing loss caused by LVAS is primarily a sensorineural hearing loss. However, because other abnormalities are
often present, there may be a conductive component as well. Govaerts et al (1999) indicated that 90% of their sample had mixed hearing loss, although the sensorineural hearing loss was the most predominant. They stated that the conductive component can easily be misinterpreted as a middle ear ventilation problem or a type of otosclerosis. This may have been the case in the early diagnosis for KI, as the frequent negative middle ear pressure did imply eustachian tube dysfunction. Bilateral LVAS is much more common than unilateral LVAS. As stated earlier, patients with LVAS experience sudden hearing loss following a minor head injury or other activity that causes increased intracranial pressure (increased cerebrospinal fluid pressure). Also, sudden hearing loss can follow a minor illness such as a common cold, strenuous exercise, or a sudden change in barometric pressure. KI’s mother did not report any such precipitating factors. After episodes of sudden hearing loss, hearing may recover to the previous level, or much more commonly, it may recover partially to a new “normal.” Reardon et al (2000) assessed 57 subjects referred with radiological evidence of vestibular aqueduct enlargement by history, clinical examination, perchlorate discharge test, and molecular analysis of the PDS locus. Forty-one patients (72%) had unequivocal evidence of Pendred syndrome. The finding of a single heterozygous mutation at the PDS gene was strongly suggestive of a critical role for pendrin, the protein product of the PDS gene, in the generation of enlarged vestibular aqueducts in 86% of their patients with this radiological malformation. They indicated that enlargement of the vestibular aqueduct should be considered as the most likely presentation of Pendred syndrome. Jackler and DeLaCruz (1989) reported an average deterioration of 25 dB over a six-year period. The authors found no reports of permanently recovered hearing over time with LVAS. Thus, the recovery of hearing for KI was not expected. It was reported by Miyamoto et al (2002) that cochlear implants generally work well in children with LVAS. So, in both of these cases, the etiology did not suggest that recovery of hearing was likely.

A third factor the surgeon and audiologist must consider when formulating a treatment plan is the ramification of delaying implantation. It would appear to be a tragic error if a premature decision is made to implant a child, thereby typically rendering the surgical ear void of residual hearing, only to find out later that there was a possibility hearing may have recovered to the extent that hearing aids could have been beneficial. Of course, this is why the poorer ear is normally chosen as the one to be implanted in cases of asymmetrical hearing loss, thereby still allowing for potential usage of acoustic amplification by the nonimplanted ear. Moreover, unlike meningitis, neither AIED or LVAS is impacted by ossification issues. As stated earlier, however, delaying implantation has the potential of allowing for neural plastic and behavioral changes that may make subsequent restoration and development of speech and language skills more problematic. The partial recovery of JC’s hearing occurred more than a year postimplant. Considering her reported deteriorating speech and language skills, delays in implantation would not have seemed reasonable. Similarly, the authors could find no data suggesting that hearing will permanently improve following progressive deteriorations from AIED or LVAS. In addition, children (and adults) with deteriorating or fluctuating hearing loss may experience emotional problems such as depression, frustration, and anger. Lustig et al (2003) have suggested that fluctuating disorders such as Meniere’s syndrome, cochlear implantation may be an effective means of treating unstable hearing impairments. The stability of the cochlear implant thresholds, in contrast to the hearing aid thresholds, is illustrated in Figures 3 and 6. The hearing aid threshold fluctuations are directly related to the changes in hearing. The differences in cochlear implant thresholds for KI in Figure 6 are related to changes in programming. It appears intuitive to try to provide a child a stable auditory status. In addition, the potential benefits afforded by the use of a contralateral hearing aid (Ching et al, 2001) in conjunction with a cochlear implant would provide some degree of assurance that residual hearing can be maximized regardless of the rate of recovery in the nonimplanted ear.

CONCLUSIONS

The paper is not intended to criticize the decision-making process that went into either of these treatment plans. Instead, the
purpose is to raise awareness that partial hearing can return in certain cases; therefore, careful consideration of multiple issues should be given prior to undertaking a surgical procedure, the effects of which cannot be undone. Adopting an excessively conservative approach, however, could result in delays that negatively impact the child both developmentally and emotionally. Perhaps the continued advancement of hybrid implants will prove to be preferable for patients with fluctuating losses since it would leave the residual low-frequency hearing intact.

REFERENCES


