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Catastrophic Progressive Hearing Loss in Childhood

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Abstract

We present a report on a 5-year-old child with a complex medical and audiologic history who exhibited catastrophic progression in hearing loss. Hearing loss was initially attributed to bacterial meningitis at age 3 months; progression was apparently related to perilymph fistula at age 8 years. Etiologies associated with progressive hearing loss in children as well as signs of progression and monitoring protocols for children at risk for progressive hearing loss are discussed.

Key Words: Cytomegalovirus, hearing loss, meningitis

Abbreviations: ABR = auditory brainstem response, CMV = cytomegalovirus, JCIH = Joint Committee on Infant Hearing, LVA = large vestibular aqueduct, PPHN = persistent pulmonary hypertension of the newborn, TCH = The Children's Hospital

Decrease in hearing sensitivity is one of the most feared complications for children with sensorineural hearing loss. Hearing loss may fluctuate or progress for a variety of reasons including development of middle-ear disease, change in overall health status, exposure to ototoxic drugs or noise, or for unknown reasons apparently related to the natural history of the underlying condition. Whatever the cause, progression in hearing loss complicates audiologic and medical/surgical management and educational planning for the child.

The precise prevalence of progressive sensorineural hearing loss in children is unknown. Previous studies suggest that as few as 6 percent to more than 20 percent of children with sensorineural hearing loss exhibit progressive hearing loss (Parving, 1988; Brookhouser et al, 1994). In many cases, hearing loss does not progress gradually and symmetrically over time. Rather, fluctuations in hearing sensitivity and asymmetric changes occur in many children whose hearing loss ultimately progresses 10 dB or more (Brookhouser et al, 1994).

Late-onset and/or progressive sensorineural hearing loss is associated with several well-defined etiologies of childhood hearing loss including hereditary hearing loss (Brookhouser et al, 1994; Gorlin et al, 1995; Angeli et al, 1999), infectious disease (Dahle et al, 1979; Brookhouser et al, 1988; Williamson et al, 1992; Huygen and Admiraal, 1996; Woolley et al, 1999), anatomic malformation (Grundfast and Bluestone, 1978; Parnes and McCabe, 1987; Levenson et al, 1989; Davis, 1992; Zalzal et al, 1995; Shetty et al, 1997), perinatal high-risk factors and treatment (Sell et al, 1985; Naulty et al, 1986; Nield et al, 1986; Hendricks-Munoz and Walton, 1988; Salamy et al, 1989; Walton and Hendricks-Munoz, 1991), and ototoxic drugs (Walker et al, 1989; Pasic and Dobie, 1991; Weatherly et al, 1991; Parsons et al, 1998). The Joint Committee on Infant Hearing (JCIH, 1994) recommends identifying infants at risk for delayed-onset hearing loss and monitoring those infants on a regular basis to detect changes in hearing sensitivity.

We provide audiologic management for a child with a complex medical and audiologic history who developed a sudden, catastrophic decrease in auditory thresholds. As the following case report illustrates, sudden decrease in auditory thresholds is an otologic and audiologic emergency that requires immediate

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attention by parents, teachers/therapists, audiologists, pediatricians/family physicians, and otolaryngologists.

CASE REPORT

TC, a developmentally delayed boy, has received audiologic management at The Children's Hospital (TCH) since 1994 (age 5 years). Per his mother's report, TC's prenatal and birth history were unremarkable, and there was no family history of childhood hearing loss. At age 3 months, TC was hospitalized in another state with bacterial meningitis. Sequelae of the meningitis included a stroke, seizure disorder, and significant speech and language and developmental delays. According to his mother, TC received an auditory brainstem response (ABR) evaluation prior to discharge that showed normal hearing in his left ear and a profound hearing loss in his right ear. No other information about TC's audiologic care in the other state was available. At the time of his initial audiologic assessment at TCH, TC was enrolled in speech-language therapy as well as early childhood special education.

TC's behavioral audiogram (Fig. 1), obtained in January 1995 (age 5 years, 11 months), shows mild sensorineural hearing loss at 250 through 1 kHz, normal sensitivity at 2 and 4 kHz, and a moderate sensorineural hearing loss at 6 and 8 kHz in the left ear. Masked thresholds could not be obtained from TC's right ear; however, a previous ABR completed at TCH revealed no response from that ear to 90 dB nHL clicks. At that time, TC was fitted with a loaner mild-gain hearing aid in his left ear.

Follow-up audiometric evaluations are summarized in Table 1. An audiogram completed in the public schools in July 1995 (age 6 years, 5 months) showed stable hearing thresholds (± 5 dB) in both ears. In September 1996 (age 7 years, 6 months), TC returned to TCH for follow-up assessment and demonstrated stable audiometric thresholds (± 5 dB) except at 4 kHz in his left ear, where sensitivity decreased 25 dB (20–45 dB HL). TC's mother and his educational program (private school for children with spe-

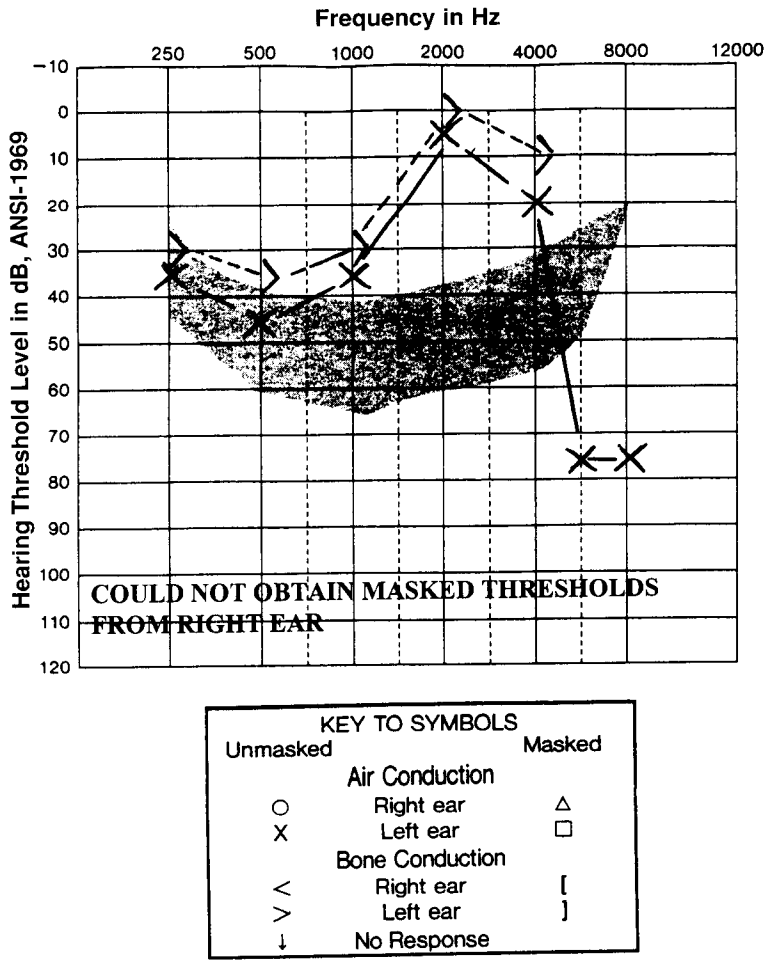

Figure 1 Behavioral pure-tone audiogram obtained from TC, a developmentally delayed hearing-impaired boy, in January 1995 (age 5 years, 11 months), showing mild sensorineural hearing loss at 250 through 1 kHz, normal hearing sensitivity at 2 and 4 kHz, and moderate sensorineural hearing loss at 6 and 8 kHz in the left ear. Masked thresholds could not be obtained from TC's right ear.

Table 1 Follow-up Audiometric Evaluations for TC, a Developmentally Delayed Hearing-Impaired Boy

Date (Location)	*Age*	*Results*
Jan 1995 (TCH)	5 yr, 11 mo	Baseline (see Fig. 1). In left ear, mild sensorineural hearing loss from 250 through 1 kHz, normal hearing at 2 and 4 kHz, moderate sensorineural hearing loss at 6 and 8 kHz. Could not obtain masked thresholds from right ear.
July 1995 (school)	6 yr, 5 mo	No change in left ear. All thresholds ± 5 dB of thresholds obtained January 1995. Could not obtain masked thresholds from right ear.
Sept 1996 (TCH)	7 yr, 6 mo	No change in left ear from 250 through 2 kHz or at 8 kHz. 25-dB decrease in sensitivity at 4 kHz. Could not obtain masked thresholds from right ear.
Apr 1997 (TCH)	8 yr, 1 mo	Significant decrease in left ear sensitivity from 250 through 8 kHz; profound sensitivity loss bilaterally (see Fig. 2).

TCH = The Children's Hospital.

cial educational needs) were advised of the change in hearing sensitivity at 4 kHz in the left ear and encouraged to obtain follow-up within 6 months. At TC's next appointment in April 1997 (age 8 years, 1 month), his mother reported a 3-week history of decreased auditory responsiveness and refusal to wear his hearing aid. Behavioral test results (Fig. 2) showed a profound sensorineural hearing loss bilaterally. Acoustic immittance results revealed normal tympanograms, normal physical volume measures, and absent acoustic reflexes in both ears.

TC was immediately referred for otologic assessment and intervention. The otolaryngologist reported that physical examination revealed a positive fistula test on both sides

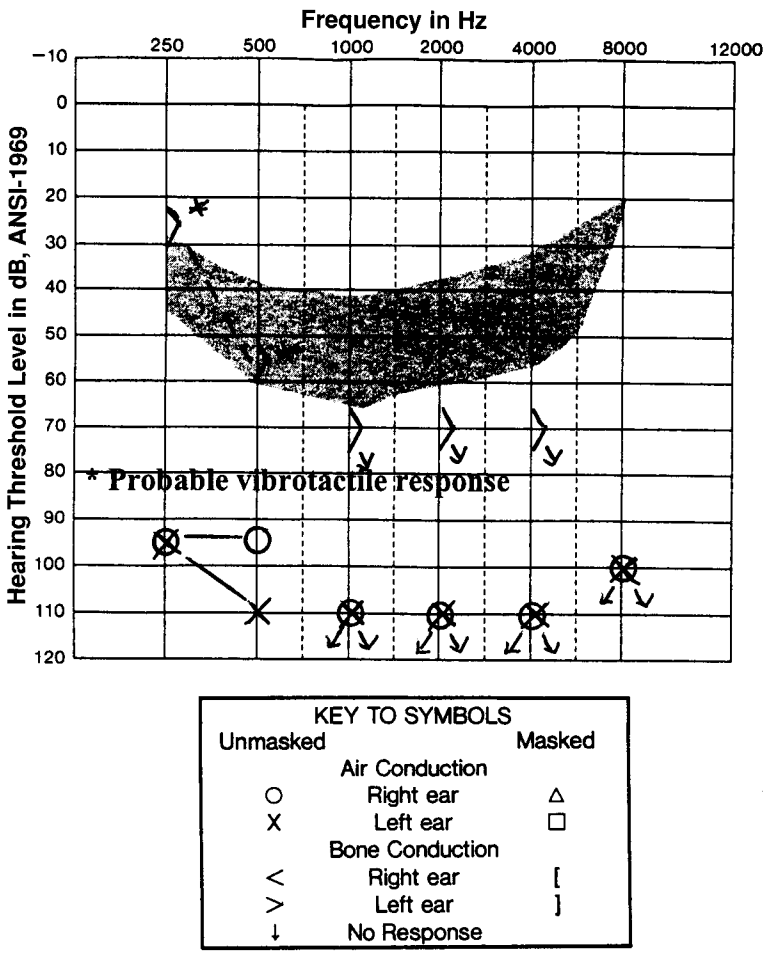

Figure 2. Behavioral pure-tone audiogram obtained from TC in April 1997 (age 8 years, 1 month), showing a profound sensorineural hearing loss bilaterally.

with left-beating nystagmus. There was no history of dizziness, and computed tomography scan revealed no structural abnormality. Exploratory tympanotomy was performed bilaterally 6 days after his audiologic evaluation. At surgery, the otolaryngologist reported that "...on both sides, the round window was vastly enlarged to about four times normal size...with an active leak...from the right side only..." There was evidence of a fistula on the left side as well, and both round windows were patched with a fascia graft.

TC received follow-up audiologic evaluation and management services in June 1997 (age 8 years, 3 months) and March 1998 (age 9 years, 1 month). Results of behavioral audiologic evaluation continued to show profound hearing loss bilaterally. Otoacoustic emissions were absent in both ears. Acoustic immittance results were consistent with normal middle-ear function.

TC now wears binaural hearing aids from which he receives limited benefit. Although he detects frequency-specific signals at approximately 45 to 75 dB HL, he is unable to discriminate spondaic words without visual cues. His mother has declined evaluation for cochlear implantation at TCH. He is enrolled in a special education total communication program.

COMMENT

Parents and care givers of any child with an etiology of hearing loss known or suspected to be associated with fluctuation and/or progression should be alerted to monitor the child for progression in hearing loss. In the case described above, the presumptive etiology of hearing loss was bacterial meningitis, although the catastrophic drop in hearing sensitivity was apparently related to perilymph fistula. Because there was incomplete audiologic information from age 3 months (postmeningitis) to age 5 years (audiologic management at TCH), we were unable to predict stability of TC's hearing thresholds prior to his behavioral evaluation at age 5 years, 11 months. TC's mother reported that an ABR completed after treatment for meningitis (age 3 months) revealed a normal response from his left ear and no response from his right ear. Although TC's behavioral audiogram (age 5 years, 11 months) showed significant sensorineural hearing loss in his left ear, the "normal" ABR obtained from that ear is not incompatible with these later behavioral results. TC's normal hearing sensitivity at 2 and 4 kHz in his left ear could have yielded in a normal click-evoked ABR from that ear despite significant low- and high-frequency sensorineural hearing loss (Hyde, 1985; Laukli and Mair, 1985). Follow-up audiologic evaluation using frequency-specific ABR or age-appropriate conditioned behavioral tests might have revealed TC's significant low-frequency hearing loss. Based on no response to ABR from TC's right ear, we might presume that severe-to-profound hearing loss in that ear was present at least since the meningitis. Thus, at the time of TC's behavioral assessment at age 5 years, 11 months, there was no specific audiometric information to indicate that his hearing loss had progressed since treatment for meningitis.

According to his mother, TC exhibited only minimal signs of change in hearing sensitivity and auditory function. His family identified reduced auditory responsiveness and refusal to wear his hearing aid as the only signs suggesting a change in hearing sensitivity. Despite presence of a perilymph fistula, TC did not exhibit dizziness or other behaviors consistent with vestibular dysfunction. TC's limited speech and language skills as well as his significant developmental delays precluded accurate self-report of change in hearing sensitivity. Although the reported behavioral changes were observed over a 3-week period, we do not know if TC experienced a gradual or sudden change in hearing sensitivity.

Based on the presumed etiology of hearing loss, marked interaural asymmetry, and presence of significant developmental delays, TC's audiologist recommended audiologic monitoring on a 6-month follow-up schedule. Despite this recommendation, we were unable to define the time course of change in hearing sensitivity or to initiate otologic treatment that conserved residual hearing function. In the absence of longitudinal audiologic information, we were unable to predict potential progression and to initiate a more sensitive follow-up schedule.

TC's case suggests three important issues concerning progressive hearing loss in children. First, this case demonstrates the importance of determining etiology of childhood hearing loss whenever possible. Second, this case highlights the need to describe the behavioral manifestations of fluctuation and/or progression of hearing loss in children, especially very young or developmentally delayed children. Third, TC's case suggests the need to define recommended monitoring protocols for children at risk for progressive hearing loss.

Etiologies of Progressive Hearing Loss in Children

In many cases, the etiology of hearing loss in children with progressive hearing loss cannot be definitively ascertained (Parving, 1988; Brookhouser et al, 1994). In those cases where etiology can be determined, four principal conditions are implicated in progressive hearing loss in children: (1) hereditary hearing loss, (2) infectious disease, (3) anatomic abnormality, and (4) perinatal high-risk factors and associated medical treatments.

Brookhouser et al (1994) evaluated 229 children who demonstrated progressive and/or fluctuating sensorineural hearing loss. Hereditary hearing loss was present in 14 percent of children, and an additional 7 percent of children had a positive family history of hearing loss without a discernible pattern of inheritance. Parving (1988) followed 138 children for progressive hearing loss, only 6 percent of whom demonstrated progression over the 5-year study period. In her sample, 48 percent of the children demonstrated either syndromal (12%) or nonsyndromal (36%) inherited hearing loss. Syndromal etiologies associated with progressive hearing loss include Alport's syndrome, Waardenburg's syndrome type II, and Alström syndrome (Gorlin et al, 1995). In addition, autosomal dominant, autosomal recessive, and x-linked nonsyndromal progressive hearing losses have been reported (Gorlin et al, 1995). Recently, mitochondrial inheritance of nonsyndromic progressive hearing loss in a single family has been described (Angeli et al, 1999).

In one of the first reports on etiology-specific progressive hearing loss in children, Dahle et al (1979) described the relationship between congenital symptomatic cytomegalovirus (CMV) and progressive hearing loss. Subsequent reports (Pass et al, 1980; Williamson et al, 1982, 1990, 1992) confirmed these findings and further demonstrated the presence of late-onset, progressive hearing loss in infants with asymptomatic congenital CMV. Because congenital CMV is estimated to occur in 0.4 percent to 2.3 percent of all live births, and because 90 percent of infants with congenital CMV are asymptomatic at birth (Williamson et al, 1992), tens of thousands of infants are born in the United States each year with the undetected potential to develop late-onset, progressive, sensorineural hearing loss.

Children with postmeningitic sensorineural hearing loss may exhibit fluctuation or progression in hearing loss for many years after recovery from meningitis (Brookhouser et al, 1988; Woolley et al, 1999). Even children with initially severe-to-profound hearing loss may exhibit further decrease in auditory thresholds resulting in significant decrease in hearing aid benefit (Brookhouser et al, 1988).

Anatomic malformations are a well-known cause of progressive hearing loss. Mondini's abnormality, characterized by bony and membranous labyrinthine abnormalities, is commonly associated with progressive sensorineural hearing loss (Mitchell and Rubin, 1985; Parnes and McCabe, 1987; Shetty et al, 1997). Large vestibular aqueduct (LVA) syndrome, characterized by an enlarged endolymphatic duct and sac detectable by magnetic resonance imaging (Hirsch et al, 1992), may be significantly underdiagnosed in children with sensorineural hearing loss (Jackler and De La Cruz, 1989). LVA typically results in sensorineural hearing loss in the affected ear, which may progress to profound deafness (Jackler and De La Cruz, 1989; Levenson et al, 1989; Arcand et al, 1991; Zalzal et al, 1995; Au and Gibson, 1999; Harker et al, 1999). Perilymph fistula, characterized by an abnormal communication between the inner and middle ear most commonly at the round window, may be associated with Mondini's or other malformation, barotrauma, or head injury and may occur spontaneously in some children (Grundfast and Bluestone, 1978; Parnes and McCabe, 1987; Davis, 1992). In some cases, surgical intervention to repair the fistula has been successful in halting progression in hearing loss (Parnes and McCabe, 1987; Davis, 1992).

Medical treatments that result in progressive hearing loss are typically life-saving interventions for congenital or acquired disorders. Numerous investigators have reported the association between persistent fetal circulation or persistent pulmonary hypertension of the newborn (PPHN) and delayed-onset and/or progressive sensorineural hearing loss (Sell et al, 1985; Naulty et al, 1986; Nield et al, 1986; Hendricks-Munoz and Walton, 1988; Walton and Hendricks-Munoz, 1991; Kawashiro et al, 1996). Investigators speculate that both clinical and treatment variables underlie the development of sensorineural hearing loss in infants with PPHN. Newer treatment protocols using inhaled nitric oxide rather than prolonged mechanical ventilation may reduce hearing loss associated with this condition (Fujikawa et al, 1997).

Ototoxic medications, including antibiotics, diuretics, and antineoplastic treatments, are

an important cause of progressive hearing loss in children and may account for significant childhood deafness in developing countries (Zeng, 1995). Both cisplatin and carboplatin chemotherapeutic agents are ototoxic and used to treat childhood malignancies (Walker et al, 1989; Pasic and Dobie, 1991; Weatherly et al, 1991; Parsons et al, 1998). The ototoxic effect of cisplatin is enhanced by prior or simultaneous cranial irradiation (Walker et al, 1989). For preterm infants who receive care in the neonatal intensive care unit, aminoglycoside antibiotics used in conjunction with loop diuretics are implicated in development of sensorineural hearing loss (Salamy et al, 1989; Borradori et al, 1997).

Parents typically seek the cause of their child's newly identified hearing loss. Because progressive hearing loss is a common finding in many conditions associated with childhood hearing loss, determining etiology of hearing loss in an infant or young child is especially important. All children with hearing loss should receive thorough medical/otologic and other evaluations appropriate to determine etiology of hearing loss and probability of progression (JCIH, 1994).

Signs of Progressive Hearing Loss in Children

Detecting fluctuating and/or progressive hearing loss in children is difficult, especially in very young infants or developmentally delayed children. Follow-up surveys of children who received newborn hearing screening reveal a small but important number of cases of hearing loss undetected by newborn screening (Nield et al, 1986; Lutman et al, 1997; Finitzo et al, 1998). Are these cases false-negative newborn hearing screenings or cases of late-onset progressive hearing loss? As universal newborn hearing screening becomes widespread, parents, primary care physicians, and other care givers should be alert to the signs of late-onset progressive hearing loss to ensure prompt care for infants and children with this condition.

Brookhouser et al (1994) document the importance of unilateral and asymmetric hearing loss as predictive of fluctuation and/or progression in hearing loss in the contralateral ear. In their study, the investigators found a 0.67 probability of progressive hearing loss in the ear contralateral to an ear with documented progression. Unilateral and/or asymmetric hearing loss has been reported in infants with asymptomatic congenital CMV (Williamson et al, 1992), an etiology strongly related to progressive hearing loss. Several investigators have reported identification of newborns with unilateral sensorineural hearing loss through universal newborn hearing screening programs (Barsky-Firsker and Sun, 1997; Finitzo et al, 1998; Mehl and Thomson, 1998). Because asymptomatic congenital CMV is often undetected at birth, diagnosis of unilateral hearing loss in a newborn infant should raise the suspicion of congenital CMV. Children of any age with unilateral and/or asymmetric hearing loss of unknown etiology should be considered at risk for late-onset progressive hearing loss in their contralateral ear.

Behaviors or complaints of related conditions must also be considered. Children who exhibit delay in development of walking, develop unexpected clumsiness, or complain of dizziness may be experiencing vestibular dysfunction. In a review of 16 patients with surgically confirmed perilymph fistula, Parnes and McCabe (1987) reported evidence of vestibular symptoms including complaint of intermittent dizziness or observed spells of imbalance in nine children. Limited reports on tinnitus in childhood hearing loss suggest that children experience tinnitus associated with both hearing loss and ear disease (Mills and Cherry, 1984; Mills et al, 1986; Hausler et al, 1987; Drukier, 1989; Viani, 1989).

Changes in auditory behavior—inattentiveness, inability to be consoled with sounds and voices, frequent requests for repetition, and increasing the volume on the television—are common-sense warning signs for parents and care givers to observe. For children with preexisting hearing loss, change in auditory behavior associated with hearing aid use is especially important. Refusal to wear a hearing aid or complaint that a hearing aid "hurts" or is broken must be promptly investigated. After ruling out hearing aid malfunction or ear canal irritation, parents should seek immediate audiologic and otologic assessment for potential change in hearing thresholds.

MONITORING CHILDREN FOR PROGRESSIVE HEARING LOSS

Despite the well-warranted concern about progression of sensitivity loss in children with preexisting sensorineural hearing loss, a universally accepted protocol of audiologic and medical/otologic surveillance and monitoring cannot be defined. Because the natural history and clinical course of auditory dysfunction varies depending on etiology and patient-specific individual differences, a single monitoring protocol

will not be appropriate for all conditions and all children. Investigators have suggested specific audiologic follow-up protocols for children with specific etiologic conditions based on their assessment of the natural course of the disorder. For example, Brookhouser et al (1988) suggest a protocol for longitudinal assessment of hearing in the postmeningitic child. Their protocol includes audiologic re-evaluation every 3 months until hearing thresholds stabilize and at regular intervals (as often as 3 months or no less than annually depending on the age of the child) thereafter. Walton and Hendricks-Munoz (1991) instituted a follow-up protocol for infants with PPHN that included audiologic evaluation before discharge and at 4, 12, 24, and 48 months of age. Based on the observation that progression of hearing loss in children with asymptomatic congenital CMV may occur as late as 4 years of age, Williamson et al (1992) suggest that

> hearing-impaired children without identified causes for their hearing losses should be managed as though the losses were caused by congenital CMV. Their management should include periodic audiologic assessments...to monitor hearing status for further deterioration. Such vigilance must continue uninterrupted until further longitudinal studies are completed and an endpoint for audiologic changes in hearing loss caused by CMV can be identified...

For children who receive ototoxic drugs, the American Speech-Language-Hearing Association (1993) recommends audiologic evaluation immediately after treatment and at 3 months, 6 months, and 1 year after cessation of treatment.

For children with documented normal hearing but indicators for delayed-onset sensorineural or conductive hearing loss, the JCIH (1994) recommends hearing evaluations at least every 6 months until age 3 years, and at appropriate intervals thereafter. The JCIH indicators associated with delayed-onset sensorineural hearing loss include (1) family history of hereditary childhood hearing loss; (2) in utero infection such as cytomegalovirus, rubella, syphilis, herpes, or toxoplasmosis; and (3) neurofibromatosis type II and neurodegenerative disorders. The JCIH indicators associated with conductive hearing loss include (1) recurrent or persistent otitis media with effusion, (2) anatomic deformities and other disorders that affect eustachian tube function, and (3) neurodegenerative disorders.

Regardless of the etiology of hearing loss or established monitoring protocol, any child who exhibits change in auditory behavior, complains of decreased sensitivity or diminished benefit from amplification, or demonstrates signs of related vestibular or auditory effects should receive immediate audiologic and medical/otologic assessment and intervention. Conservative treatment in these cases should be just that: treatment to conserve hearing and auditory function.

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