Causes of Pediatric Sensorineural Hearing Loss

Yesterday and Today

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Objective: To ascertain the present common causes of sensorineural hearing loss (SNHL) in children and compare them with those of previous reports.


Setting: A tertiary care children’s hospital.

Patients: Three hundred one children, aged 1 week through 18 years, who presented for evaluation of SNHL.

Results: Of the 301 children, 68.1% had a definite or probable cause of their SNHL identified; 18.9%, 1 or more possible causes; and 31.9%, no obvious cause. A family history of SNHL or prematurity and/or complicated perinatal course was found in 28.6% of patients. Named syndromes, multiple congenital anomalies, meningitis, or perinatal maternal factors, including maternal prenatal substance abuse, were present in another 38.5%. However, syndromes commonly reported to be associated with SNHL, such as Waardenburg syndrome, were seen in less than 1% of patients. The average age at diagnosis was 3.02 years for the bilateral moderate or worse SNHL, for unilateral SNHL, the average age was 3.97 years. The most useful diagnostic study was computed tomographic scanning.

Conclusions: Sensorineural hearing loss is fairly common in children. Extensive workups, often without clear direction, should be reconsidered based on the children with SNHL who otolaryngologists are now seeing. Infant screening programs, although identifying many children earlier, will also provide the opportunity to fine-tune the evaluation (ie, cytomegalovirus titers and/or cultures at birth), increasing the diagnostic yield.


T he incidence of severe to profound sensorineural hearing loss (SNHL) in children is approximately 1:2000 at birth and 6:1000 by 18 years of age.¹ Although these numbers indicate that SNHL is relatively common, it remains underappreciated and underdiagnosed in children. For example, the severe to profound unilateral losses are often not recognized until kindergarten, when the child undergoes the first audiometric evaluation. The high-risk register, which was designed to help decide who needs early audiometric screening, only captures 50% of the significant losses in infancy; the other 50% of children do not have obvious risk factors, or their risk factors are not appreciated. At present, most states do not have mandatory hearing screening at birth for all children, regardless of the risk factors, so many children are missed. In addition, 50% of the losses occur after the newborn period, so only ongoing surveillance will identify losses in these children.

Adding to the confusion about screening and who, when, and how are the uncertain results of the diagnostic process. Even when a loss is identified, most studies indicate a “hit rate” for an identified cause of 60% or less.²⁻⁴ This low number and the expense and nonuniform nature of the workup often discourage physicians from pursuing any further studies, leaving the patient and the physician unsatisfied. Finally, because of the belief that there is “nothing that the physician can do” to help these patients, the children are often seen only once, rather than having follow-up visits that may eventually yield a diagnosis, and they may not be referred for appropriate habilitative services.

One of the reasons that physicians are often reluctant, or uncertain, about pursuing an evaluation of the cause of SNHL is that many historical studies indicate that most of the causes are obscure and may actually have never been encountered by the evaluating otolaryngologist. Therefore, in an effort to provide more up-to-date information on the causes of SNHL, which could be used to guide future diagnostic evaluation, we performed a retrospective review of 301 children seen in the Department of Otolaryngology and Communication Disorders at the Children’s Hospital, Boston, Mass. Dr Billings is now with the Department of Otorhinolaryngology, University of Texas Southwestern Medical Center and Children’s Medical Center, Dallas.

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PATIENTS AND METHODS

The medical charts of 405 children aged 1 week through 18 years who presented for evaluation of SNHL from January 1, 1993, through September 30, 1996, were reviewed. Ninety patients were excluded based on age greater than stated criteria, normal hearing noted on results of auditory brainstem response testing, or tympanostomy tube placement. An additional 14 patients with unilateral mild (<35 dB) SNHL were excluded due to overall lack of clinical information available and of need for intervention. A total of 301 patients underwent evaluation for sex, age at diagnosis, unilateral vs bilateral hearing impairment, and degree of impairment. Other medical problems and/or congenital anomalies were noted. Particular attention was paid to identifying those children with known syndromes, birth factors, or maternal factors that may contribute to hearing loss and with acquired factors (ie, meningitis, cisplatin chemotherapy). Abnormalities noted on results of computed tomographic (CT) scanning (performed in 51.8% of patients) were documented. Finally, method of rehabilitation for the hearing loss, including hearing aids, cochlear implants, or no treatment, was recorded.

Hospital of Boston, Mass. Because the evaluations were performed by 9 different pediatric otolaryngologists during a period of years, the workups were nonuniform and based on the physicians’ best judgment at the time the patient was seen. If a probable diagnosis was obvious, the workup was frequently curtailed, whereas children with no obvious cause of SNHL underwent more extensive testing.

RESULTS

The male and female distribution of the 301 patients was approximately equal. The age at diagnosis of SNHL ranged from less than 1 month to 13 years (mean age, 3.52 years). More than half (70.1%) of the patients undergoing evaluation had bilateral severe to profound SNHL, whereas 19.9% had a unilateral severe to profound SNHL (Table 1). Overall, 80.1% of patients had a bilateral hearing impairment.

The most common associated medical problems encountered were a history of recurrent otitis media and neurologic abnormalities (ie, developmental or motor delays, cerebral palsy, and seizure disorders). A known syndrome was identified in 12.0% of patients, and 7.6% had assorted craniofacial anomalies. Twenty-four patients (8.0%) had a history of premature birth, often requiring a stay in a neonatal intensive care unit (NICU). Maternal factors, such as prenatal substance abuse, placental abruption, and toxemia, were present in 2.3%. The most common acquired factors potentially contributing to SNHL included meningitis (4.0%), cisplatin chemotherapy (2.3%), and extracorporeal membrane oxygenation (ECMO; 2.0%). Twenty patients with fluctuating hearing loss or widened vestibular aqueducts underwent middle ear exploration. Perilymph leakage from the round and/or oval window(s) was noted in 11 (55.0%). Computed tomographic scanning was performed in 51.8% of patients as part of their workup; 7.6% of patients had abnormal CT findings. These included dilated vestibular aqueducts (n = 8), vestibulocochlear dysplasia (n = 7), and Mondini malformations (n = 4). Overall, a definite or probable cause of SNHL was identified in 68.1% of patients; 31.9% had no obvious cause.

Comparative data for each group of patients undergoing evaluation, based on degree of hearing impairment, are shown in Table 1. Two hundred eleven patients with bilateral severe to profound SNHL (35 dB) were identified. The hearing loss was identified before 1 year of age in 66 (31.3%) of these patients, but the overall mean age at diagnosis was 3.02 years. Birth factors including prematurity (8.5%), prolonged NICU stay (3.8%), and elevated bilirubin levels (3.3%) were the most common etiologic factors for SNHL, followed by acquired factors, including meningitis, ECMO, and cisplatin chemotherapy. Known syndromes were identified in 12.3%. The most common syndrome was CHARGE association (associated symptoms include coloboma, hearing deficit, choanal atresia, retardation of growth, genital defects, and endocardial cushion defect) (n = 6), followed by Pierre Robin syndrome (n = 2), Usher syndrome (n = 1), Waardenburg syndrome (n = 1), achondroplasia (n = 1), and Down syndrome (n = 1). These patients had the highest rate of abnormalities identified on results of CT scanning, including dilated vestibular aqueducts (n = 8) and Mondini malforma-
Hearing Loss

No. of children 117 (100.0) 127 (100.0) 94 (100.0) 211 (100.0)
Known cause 85 (72.6) 81 (63.8) 69 (73.4) 159 (75.4)
Unknown cause 32 (27.4) 46 (36.2) 25 (26.6) 52 (24.6)
Genetic 39 (33.3) 28 (22.0) 31 (33.0) 52 (24.6)
Family history 32 (27.4) 10 (7.9) 25 (26.6) 62 (31.9)
Syndromal 7 (6.0) 18 (14.2) 7 (7.4) 26 (12.3)
Inner ear defects 0 12 (9.4) 0 25 (11.8)
Prenatal insult 16 (13.7) 16 (12.6) 14 (14.9) 40 (19.0)
TORCH infections 19 (16.2) 25 (19.7) 17 (18.1) 3 (1.4)
Meningitis 3 (2.6) 16 (12.6) 7 (7.4) 12 (5.7)
Chronic otitis media 2 (1.7) 0 1 (1.2) 51 (24.2)

* Data are given as number (percentage) of patients. SNHL indicates sensorineural hearing loss; TORCH, toxoplasmosis, other, rubella, cytomegalovirus, and herpes simplex infections.

Table 3. Comparison vs Previous Study of Unilateral SNHL

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<tr>
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<tr>
<td>No. of patients</td>
<td>324 (100.0)</td>
<td>60 (100.0)</td>
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<tr>
<td>Known cause</td>
<td>211 (65.1)</td>
<td>30 (50.0)</td>
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<tr>
<td>Unknown cause</td>
<td>113 (34.9)</td>
<td>30 (50.0)</td>
<td></td>
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<tr>
<td>Genetic</td>
<td>41 (12.6)</td>
<td>9 (15.0)</td>
<td></td>
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<tr>
<td>Prenatal insult</td>
<td>35 (10.8)</td>
<td>7 (11.7)</td>
<td></td>
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<tr>
<td>TORCH infection</td>
<td>4 (1.2)</td>
<td>1 (1.7)</td>
<td></td>
</tr>
<tr>
<td>Meningitis</td>
<td>21 (6.5)</td>
<td>0</td>
<td></td>
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<tr>
<td>Head trauma</td>
<td>35 (10.8)</td>
<td>4 (6.7)</td>
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<tr>
<td>Inner ear abnormality</td>
<td>10 (3.1)</td>
<td>4 (6.7)</td>
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* Data are given as number (percentage) of patients. SNHL indicates sensorineural hearing loss; TORCH, toxoplasmosis, other, rubella, cytomegalovirus, and herpes simplex infections.

The cause of SNHL is often not obvious. Of our 301 patients undergoing evaluation, 31.9% had no obvious cause of their hearing loss and, in some cases (4.7%), only a possible cause (ie, maternal prenatal drug abuse, congenital anomalies) could be entertained. The diagnostic search for an underlying cause can be expensive, time-consuming, and unrevealing. Determining a cause becomes important for those treatable causes of SNHL (eg, perilymphatic fistula) or can allow the physician to stop potentially cochleotoxic drug therapy, such as cisplatin chemotherapy or aminoglycoside therapy. Knowing a diagnosis is helpful in allowing the family to cope with the child’s SNHL and allows genetic counseling.

The most useful tools for establishing a potential cause of SNHL are thorough history taking and documentation. Often, a good prenatal or birth history is overlooked in favor of searching for medical problems or congenital anomalies known to be associated with SNHL. Pappas and Schaibly2 suggested that family and medical histories as well as a complete physical examination are essential steps in early pediatric surveillance for hearing loss. Similarly, another study found that reexamining children in a school for the deaf via audiological and non-audiological means in time reduced the incidence of unknown causes by half.3 Since many different physicians played a role in evaluation in our patient population, the diagnostic approach varied a great deal. Often, good prenatal, family, and birth histories were completely excluded. An approach for the evaluation and diagnostic workup have been outlined in a previous study (unpublished data, M.A.K. and M. W. Neault, PhD, February 1997) and are summarized in Table 4.

Since the search for a cause of SNHL can often be exhaustive and expensive, many debate the futility of...
ordering an expensive battery of tests that have a low yield in achieving a diagnosis. Kenna and Neault found that if results of a patient's examination and history are un-
revealing, additional testing should be considered. The test with the highest yield of a diagnosis (26%) in that re-
port was high-resolution CT scanning. In our study, 51.8%
of patients underwent CT scanning. Of these, an abnor-
mality was found in 7.6%. Additional diagnostic tests were
tailored to the individual patient, and many times de-
pended on physician preference. Of primary impor-
tance is observing the child in hopes that the diagnosis
can become apparent in time (ie, retinitis pigmentosa
in a patient with Usher syndrome, or the birth of a close
relative or sibling with hearing loss).

Comparing our data with those of several earlier stud-
ies (Table 2) suggests that the ability to determine a cause
of SNHL has improved for those children with bilateral
severe to profound SNHL (63.7%-73.4% vs 75.4% in our
study). \(^5\) The primary reason for the small increase ap-
ppears to reflect earlier detection of and screening for
SNHL in the NICU. Of 211 patients, 40 (18.9%) had a history
of a neonatal insult, including prematurity and/or low
birth weight, prolonged NICU stay (>3 days), and el-
evated bilirubin levels. This is a small but significant change from the incidence in previous reports of 12.6%
to 14.9%. Improved survival of patients in the NICU may
be a contributing factor to these figures. Another im-
pressive difference from the previous reports is the de-
creased incidence of TORCH infections (toxoplasmo-

dosis, other, rubella, cytomegalovirus [CMV], and herpes
simplex infections) in those with SNHL from 16.2% to
19.7%, down to 1.4%. Earlier studies contained a large
number of children with congenital rubella, the inci-
dence of which has decreased with more widespread
immunizations. Congenital CMV infection, however, is be-
ing increasingly diagnosed at earlier ages, and if CMV
infection is diagnosed in the first month of life, drug
therapy may be able to modify some of the neurologic
sequelae of the disease, including SNHL. \(^7\)

A multi-institutional phase 2 study evaluating the
efficacy of gancyclovir therapy at doses of 8 or 12 mg/kg
day showed hearing improvement or stabilization in
5 (16%) of 30 infants with congenital CMV infection. \(^7\)

Almost 50% of neonates have no high-risk factors that
would warrant early screening for SNHL. \(^2\) Therefore, a
diagnosis may be delayed until abnormalities in speech and
language are noted. Parving\(^8\) found that only 33% of the
children studied received a diagnosis of SNHL by 1 year
of age and that there was a delay in testing from when hear-
ing loss was first suspected to audiologic confirmation in
43%. Parents were the first to suspect the hearing loss in
up to 60% of the patients. Our data show an average age
at first diagnosis from 1.74 years for those with bilateral
moderate to profound SNHL to 4.3 years for those chil-

D | Tests to Consider in the Evaluation of SNHL in Children* |
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<tbody>
<tr>
<td><strong>Test</strong></td>
<td><strong>Reason for Test</strong></td>
<td><strong>Yield</strong></td>
<td><strong>Possible Consequences If Missed</strong></td>
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<tr>
<td>History</td>
<td>Illness, trauma, drugs</td>
<td>High</td>
<td>Depends on what missed</td>
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<tr>
<td>High-resolution CT scan</td>
<td>Anatomical abnormality</td>
<td>High</td>
<td>SNHL progression, PLF, other diagnosis</td>
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<tr>
<td>Magnetic resonance imaging</td>
<td>Anatomical abnormality</td>
<td>Medium</td>
<td>SNHL, progression, PLF, other diagnosis</td>
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<td>TORCH titers</td>
<td>Congenital infection</td>
<td>Depends on timing of test</td>
<td>Missed opportunity to treat</td>
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<td>Electrocardiogram</td>
<td>Long QT interval</td>
<td>Very low</td>
<td>Syncope, sudden death</td>
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<tr>
<td>Complete blood cell count</td>
<td>Anemia</td>
<td>Low</td>
<td>Depends on anemia type</td>
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<td>Urinalysis</td>
<td>Hematuria, proteinuria</td>
<td>Low</td>
<td>High (renal failure, Alport syndrome)</td>
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<tr>
<td>Sedimentation rate, antinuclear antibody†</td>
<td>Elevated levels</td>
<td>Low</td>
<td>Depends on diagnosis</td>
<td></td>
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<tr>
<td>BUN and creatinine levels</td>
<td></td>
<td>Low</td>
<td>High (renal failure, Alport syndrome)</td>
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<tr>
<td>RPR, fluorescent treponemal antibody</td>
<td>Syphilis</td>
<td>Low</td>
<td>Missed treatment opportunity</td>
<td></td>
</tr>
<tr>
<td>Glucose level</td>
<td>Diabetes</td>
<td>Very low</td>
<td>High</td>
<td></td>
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<tr>
<td>Thyroid function tests‡</td>
<td>Hypothyroid</td>
<td>Low</td>
<td>High if hypothyroid</td>
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<tr>
<td>Liver function tests</td>
<td>Liver abnormalities</td>
<td>Very low</td>
<td>Depends</td>
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<tr>
<td>Connexin 26</td>
<td>Recurrent SNHL</td>
<td>High</td>
<td>None (has educational value)</td>
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<td>Genetics consultation</td>
<td>Genetic SNHL</td>
<td>Variable</td>
<td>Long-term prognosis, other children</td>
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<td>Neurology consultation</td>
<td>Associated diseases</td>
<td>Low</td>
<td>Educational, medical</td>
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<tr>
<td>Ophthalmology consultation</td>
<td>Retinitis pigmentosa, others</td>
<td>Low in infants unless CMV, toxoplasmosis, rubella, syphilis</td>
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*SNHL indicates sensorineural hearing loss; CT, computed tomographic; PLF, perilymphatic fistula; TORCH, toxoplasmosis, other, rubella, cytomegalovirus, and herpes simplex infections; BUN, serum urea nitrogen; RPR, rapid plasma reagin; and CMV, cytomegalovirus.
†If autoimmune process suspected, may need Western blot and/or additional testing.
‡If Pendred syndrome suspected, may need perchlorate test.
bilirubin levels, and hypothermia. Bergman et al found that the most significant factors for predicting hearing loss in premature infants were prolonged respiratory support, hyperbilirubinemia, and hyponatremia. The postulated causes for SNHL in low-birth-weight infants include hypoxic-ischemic injury to the brainstem, hemorrhage into the inner ear, toxic effects of bilirubin or aminoglycoside. CMV infections, and acoustic trauma to cochlear hair cells due to noise levels in the NICU. In our study, 17.6% of the patients had a birth factor, including low birth weight and/or prematurity and elevated bilirubin levels, as a suspected cause of hearing loss. Of interest was the high incidence of neurologic impairment noted in the patients studied, including developmental delays, cerebral palsy, and seizure disorders. These may correspond to these earlier neonatal insults.

In another study on early detection of SNHL in infants in the NICU, 631 of 3767 high-risk infants were identified. Ninety-two percent of the patients in the NICU met the high-risk criteria for screening, as did 8% of the infants not treated in the NICU. Although the NICU group who underwent subsequent screening had the more severe hearing loss, there was a substantial incidence of abnormalities in the non-NICU group. The impairment was usually mild or associated with an obvious craniofacial defect. At present, Rhode Island, Colorado, Hawaii, Mississippi, and Connecticut have adopted screening programs for all newborns. This has allowed for earlier rehabilitation for those with SNHL.

Sixty of our patients undergoing evaluation had a unilateral severe to profound SNHL. The ability to determine the cause of the unilateral impairment was lower (50.0%) than in those with bilateral severe to profound SNHL (75.4%). This may have been influenced by less-involved histories and diagnostic workups given or the finding that many of the children had no functional impairment (61.7% required no rehabilitation) and were identified at a later age on average (when a detailed birth history may have been overlooked). The importance of early detection in this group should not be overshadowed. Bess et al found that one third of children with unilateral SNHL were required to repeat a grade and were easily distractible, prone to daydreaming, and unable to follow directions well. Brookhauser et al evaluated unilateral SNHL in 324 children. A known cause was found in 65.1%; cause was unknown in 34.8%. Overall, the causes of unilateral SNHL were similar in our study (Table 4). More than half of the patients undergoing evaluation at Boy's Town had academic or behavioral problems at school. This emphasizes the importance of close follow-up and early intervention in children who decide to assist in appropriate rehabilitation. Despite the unilateral SNHL, almost 30% of our patients required a hearing aid or FM system in the classroom.

Eighty-five percent of children with SNHL are thought to suffer from congenital or early acquired hearing loss. Our data show that 30.6% of children undergoing evaluation had a hereditary factor that may have contributed to their hearing loss. These include known syndromes (12.0%), a family history of hearing loss (11.0%), and an inner ear abnormality on results of CT scanning (7.6%). These data have not changed significantly compared with previous studies. Nowadays, many deaf children are not confined to schools for the deaf but are “mainstreamed” with hearing children. This may make it more difficult to study the population of hard-of-hearing children in search of causes and to provide genetic counseling. It is our hope that more states adopt policies for mandatory newborn hearing screening. In addition, recent genetic studies have identified connexin 26 mutations in a significant proportion (50%-80%) of patients with sporadic deafness. Such information will undoubtedly give us better insight into the potential causes of SNHL and allow more timely intervention and parental counseling.

Given the improved survival rates of premature infants, children with multiple medical and neurologic problems, and children with congenital anomalies or childhood tumors, we cannot overemphasize the importance of early screening and ongoing surveillance for SNHL. Earlier education and rehabilitation can then be instituted, allowing these children to reach their potential.

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REFERENCES