

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.

Design: A retrospective review of the medical records for all children with a diagnosis of SNHL seen from January 1, 1993, through September 30, 1996, at our institution.

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 pre-

natal 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.

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THE 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 uncer-

tain 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

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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 7.0% 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 ab-

Table 1. Comparative and Epidemiologic Data for the Different Degrees of Hearing Impairment*

Hearing Loss Criteria	Bilateral Severe-Profound	Bilateral Mild-Moderate	Unilateral Severe-Profound	Total
Female	98 (46.4)	13 (43.3)	28 (46.7)	139 (46.2)
Male	113 (53.6)	17 (56.7)	32 (53.3)	162 (53.8)
Total	211 (70.1)	30 (10.0)	60 (19.9)	301 (100.0)
Medical problems				
Neurologic	39 (18.5)	6 (20.0)	11 (18.3)	56 (18.6)
Childhood tumors	9 (4.3)	0	1 (1.7)	10 (3.3)
Congenital anomalies				
Craniofacial	13 (6.2)	4 (13.3)	4 (6.7)	21 (7.0)
Other	28 (13.3)	2 (6.7)	10 (16.7)	40 (13.3)
Known syndromes	26 (12.3)	4 (13.3)	6 (10.0)	36 (12.0)
Birth factors				
Prolonged NICU	8 (3.8)	2 (6.7)	2 (3.3)	12 (4.0)
Prematurity	18 (8.5)	3 (10.0)	3 (5.0)	24 (8.0)
Hypoxemia	7 (3.3)	0	2 (3.3)	9 (3.0)
Elevated bilirubin level	7 (3.3)	1 (3.3)	0	8 (2.6)
Maternal factors	4 (1.9)	0	3 (5.0)	7 (2.3)
Acquired factors				
Meningitis	12 (5.7)	0	0	12 (4.0)
Chemotherapy	7 (3.3)	0	0	7 (2.3)
ECMO	5 (2.4)	1 (3.3)	0	6 (2.0)
Aminoglycosides	2 (0.9)	0	0	2 (0.7)
Abnormal results of CT scans	18 (8.5)	1 (3.3)	4 (6.7)	23 (7.6)
Family history	26 (12.3)	3 (10.0)	4 (6.7)	33 (11.0)
Rehabilitation				
Hearing aids	170 (80.6)	19 (63.3)	17 (28.3)	206 (68.4)
Cochlear implant	18 (8.5)	0	0	18 (6.0)
None	11 (5.2)	11 (36.7)	37 (61.7)	59 (19.6)
Known or probable cause	159 (75.4)	16 (53.3)	30 (50.0)	205 (68.1)

*Data are given as number (percentage) of patients. NICU indicates neonatal intensive care unit; ECMO, extracorporeal membrane oxygenation; and CT, computed tomographic.

normal 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-

Table 2. Epidemiologic Features of Previous Studies for Children With Bilateral Moderate to Severe SNHL*

Hearing Loss Criteria	Reference (Year)			
	Parving ⁶ (1983)	Pappas and Schaibly ² (1984)	Parving ⁴ (1985)	Present Study (1997)
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)	26 (12.3)
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.

tions (n = 3). Most patients (80.6%) underwent rehabilitation with hearing aids, and 18 children (8.5%) underwent cochlear implantation. The definite or probable causes of SNHL were identified in 75.4%.

Those children with high-frequency or sloping SNHL constituted 17.1% of the children undergoing evaluation in the bilateral severe to profound SNHL group. Birth factors associated with SNHL were less prevalent among these patients. Unique to this subset was the high average age of diagnosis (4.3 years) and the high percentage of acquired factors (25.0%) resulting in the hearing impairment. Seven patients had documented hearing loss related to cisplatin chemotherapy used in the treatment of a variety of childhood tumors.

Those children with mild to moderate bilateral SNHL (≤ 35 dB) were identified later in life (average age at diagnosis, 3.57 years) than those with severe to profound SNHL, often not until school screening audiography was performed. Six (20.0%) of these patients had birth factors that may have contributed to their hearing impairment, and the acquired causes of SNHL were less frequently noted (3.3% vs 12.3%). A significant proportion (63.3%) of these children underwent rehabilitation with hearing aids.

Sixty patients underwent evaluation for severe to profound (> 35 dB) degrees of unilateral SNHL, constituting 19.9% of the population studied. The mean age at diagnosis for these patients was 3.97 years, and 70% (42/60) did not receive initial diagnoses until at least 3 years of age. Relatively few of these patients had congenital anomalies or identifiable syndromes (including Goldenhar syndrome [n = 2], Evans syndrome, Crouzon disease, and Klippel-Feil syndrome). Although many children had neonatal risk factors for SNHL, a history of head trauma was a common cause noted in 6.7% (4/60) of this group. Most patients received no rehabilitation for unilateral hearing loss (61.7%). The definite or probable causes of SNHL were identified in only 50.0% of these patients.

Our data were compared with those of previous reports to look for changes in the common etiologic risk

Table 3. Comparison vs Previous Study of Unilateral SNHL*

Hearing Loss Criteria	Reference (Year)	
	Brookhauser et al ¹⁵ (1991)	Present Study (1997)
No. of patients	324 (100.0)	60 (100.0)
Known cause	211 (65.1)	30 (50.0)
Unknown cause	113 (34.9)	30 (50.0)
Genetic	41 (12.6)	9 (15.0)
Prenatal insult	35 (10.8)	7 (11.7)
TORCH infection	4 (1.2)	1 (1.7)
Meningitis	21 (6.5)	0
Head trauma	35 (10.8)	4 (6.7)
Inner ear abnormality	10 (3.1)	4 (6.7)

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

factors associated with bilateral SNHL (**Table 2**). The 2 primary changes have been the increased number of children with neonatal risk factors as a cause of their SNHL and the decreased incidence of congenital rubella infections. For those with unilateral SNHL, findings are consistent with those of a previous report (**Table 3**).

COMMENT

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.⁵ 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

Table 4. Tests to Consider in the Evaluation of SNHL in Children*

Test	Reason for Test	Yield	Possible Consequences If Missed
History	Illness, trauma, drugs	High	Depends on what missed
High-resolution CT scan	Anatomical abnormality	High	SNHL progression, PLF, other diagnosis
Magnetic resonance imaging	Anatomical abnormality	Medium	SNHL, progression, PLF, other diagnosis
TORCH titers	Congenital infection	Depends on timing of test	Missed opportunity to treat
Electrocardiogram	Long QT interval	Very low	Syncope, sudden death
Complete blood cell count	Anemia	Low	Depends on anemia type
Urinalysis	Hematuria, proteinuria	Low	High (renal failure, Alport syndrome)
Sedimentation rate, antinuclear antibody†	Autoimmune	Low	Depends on diagnosis
BUN and creatinine levels	Elevated levels	Low	High (renal failure, Alport syndrome)
RPR, fluorescent treponemal antibody	Syphilis	Low	Missed treatment opportunity
Glucose level	Diabetes	Very low	High
Thyroid function tests‡	Hypothyroid	Low	High if hypothyroid
Liver function tests	Liver abnormalities	Very low	Depends
Connexin 26	Recessive SNHL	High	None (has educational value)
Genetics consultation	Genetic SNHL	Variable	Long-term prognosis, other children
Neurology consultation	Associated diseases	Low	Educational, medical
Ophthalmology consultation	Retinitis pigmentosa, others	Low in infants unless CMV, toxoplasmosis, rubella, syphilis	Double handicap

*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.

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 unrevealing, additional testing should be considered. The test with the highest yield of a diagnosis (26%) in that report was high-resolution CT scanning. In our study, 51.8% of patients underwent CT scanning. Of these, an abnormality was found in 7.6%. Additional diagnostic tests were tailored to the individual patient, and many times depended on physician preference. Of primary importance is observing the child in hopes that the diagnosis may 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 studies (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).^{2,5,6} The primary reason for the small increase appears 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 elevated 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 impressive difference from the previous reports is the decreased incidence of TORCH infections (toxoplasmosis, 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 incidence of which has decreased with more widespread immunizations. Congenital CMV infection, however, is being 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 per 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.^{2,8} Therefore, a diagnosis may be delayed until abnormalities in speech and language are noted. Parving⁹ 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 hearing 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 children with high-frequency or sloping moderate to profound SNHL. Early detection of SNHL cannot be overemphasized. Primary care physicians and school systems must be encouraged to avoid delays in patient referrals for children suspected of having a hearing loss.

In 1993, the National Institutes of Health recommended universal screening for hearing loss by 3 months of age and that all patients in the NICU undergo screening before their discharge.¹⁰ This was followed by similar recommendations by the Joint Commission on Infant Screening in 1994; they further recommended habilitation by 6 months.¹¹ With shorter postnatal hospital stays for healthy newborns, as well as frequent interhospital transfers for many infants in the NICU, it is often difficult to perform even a screening test, let alone more detailed diagnostic testing and timely intervention, for these patients.

Low birth weight (<1500 g) is the most common risk factor for SNHL in the NICU population. In 1 study of low-birth-weight infants, those with SNHL were found to have had more frequent periods of apnea, increased

bilirubin levels, and hypothermia.¹² Bergman et al¹³ 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.⁸ 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 al¹⁴ 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 al¹⁵ 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.¹⁵ This emphasizes the importance of close follow-up and early intervention in these children 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.^{7,16} 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.^{17,18} Such information will undoubtedly give us better insight into the potential causes of SNHL and allow more timely intervention and parental counseling.

CONCLUSIONS

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

- Bergstrom L, Hemenway WG, Downs MP. A high risk registry to find congenital deafness. *Otolaryngol Clin North Am.* 1977;4:369-399.
- Pappas DG, Schaibly M. A two-year diagnostic report on bilateral sensorineural hearing loss in infants and children. *Am J Otol.* 1984;5:339-343.
- Stein L, Clark S, Kraus N. The hearing-impaired infant: patterns of identification and habilitation. *Ear Hear.* 1983;4:232-236.
- Parving A. Aetiological diagnosis in hearing-impaired children: clinical value and application of a modern examination program. *Int J Pediatr Otorhinolaryngol.* 1984;7:29-38.
- Hollen A, Parving A. Aetiology of hearing disorders in children at the school for the deaf. *Int J Pediatr Otorhinolaryngol.* 1985;10:229-236.
- Parving A. Epidemiology of hearing loss and aetiological diagnosis of hearing impairment in childhood. *Int J Pediatr Otorhinolaryngol.* 1983;5:151-165.
- Whitley RJ, Cloud G, Gruber W, et al. Gancyclovir treatment of symptomatic congenital cytomegalovirus infection: results of a phase II study: National Institute of Allergy and Infectious Diseases Collaborative Antiviral Study Group. *J Infect Dis.* 1997;175:1080-1086.
- Alberti PW, Hyd ML, Riko K, Hollis C, Fitzhardinge PM. Issues of early identification of hearing loss. *Laryngoscope.* 1985;95:373-381.
- Parving A. Early detection and identification of congenital/early acquired hearing disability: who takes the initiative? *Int J Pediatr Otorhinolaryngol.* 1984;7:107-117.
- Early identification of hearing impairment in infants and young children. *NIH Consensus Statement.* 1993;11:1-24.
- American Academy of Pediatrics Joint Committee on Infant Hearing. Joint Committee on Infant Hearing 1994 Position Statement. *Pediatrics.* 1995;95:152-156.
- Anagnostakis D, Petmezakis J, Papazissis G, Messantakis J, Matsoniotis N. Hearing loss in low-birth-weight infants. *AJDC.* 1982;136:602-604.
- Bergman I, Hirsch RP, Frice TJ, et al. Cause of hearing loss in the high-risk premature infant. *J Pediatr.* 1985;106:95-101.
- Bess FH, Klee T, Culbertson J. Identification, assessment, and management of children with unilateral SNHL. *Ear Hear.* 1986;7:43-51.
- Brookhauser PE, Worthington DW, Kelly WJ. Unilateral hearing loss in children. *Laryngoscope.* 1991;101:1264-1271.
- Parving A. Hearing disorders in childhood: some procedures for detection, identification, and diagnostic evaluation. *Int J Pediatr Otorhinolaryngol.* 1985;9:31-57.
- Estivill X, Fortina P, Surrey S, et al. Connexin-26 mutations in sporadic and inherited sensorineural hearing loss. *Lancet.* 1998;351:394-398.
- Cremers FPM. Genetic causes of hearing loss. *Curr Opin Neurol.* 1998;11:11-16.