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Use of Laboratory Evaluation and Radiologic Imaging in the Diagnostic Evaluation of Children With Sensorineural Hearing Loss

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Objective: **Laboratory testing and radiologic imaging are commonly used to delineate syndromic from nonsyndromic sensorineural HL (SNHL). The aim of this study was to examine the yield of laboratory tests and radiologic imaging commonly used in the diagnostic evaluation of SNHL in children.** ***Study Design:*** **Retrospective analysis of 114 (54 female, 60 male) consecutively investigated children with SNHL between 1998 and 2000 at a tertiary-care university hospital.** ***Methods:*** **Results of routine laboratory testing to assess autoimmunity, blood dyscrasias, endocrine abnormalities, renal function, infection, and cardiac testing were reviewed. Results of radiologic evaluation were also reviewed. In general, computed tomography (CT) was obtained in patients with symmetric SNHL, whereas magnetic resonance imaging (MRI) with or without CT was obtained in asymmetric SNHL.** ***Results:*** **Laboratory evaluation of the blood did not yield the etiology of SNHL in any patient. Blood tests for autoimmune disease were often positive but did not correlate with clinical disease. Nonspecific elevation of erythrocyte sedimentation rate (ESR) and antinuclear antibody (ANA) was present in 22% of cases. An abnormal electrocardiogram with a prolonged QT interval resulted in the diagnosis of Jervall and Lange-Nielsen syndrome. In the 97 patients who underwent radiologic studies, abnormalities were present in 38 of 97 studies (39%). Isolated inner ear malformations were twice as common as multiple abnormalities with large vestibular aqueducts as the most common isolated finding.** ***Conclusion:*** **In the evaluation of children with unexplained SNHL, routine laboratory evaluation should be reconsidered given its low diagnostic yield. However, radiologic abnormalities of the inner ear are common. Identification of inner ear malformations has direct impact on management of these children, suggesting that all children should undergo radiologic imaging as an integral component of evaluation of SNHL.**
Key Words: **Sensorineural HL, laboratory testing, computed tomography, magnetic resonance imaging, electrocardiograms.**

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INTRODUCTION

The prevalence of permanent, moderate-to-severe sensorineural HL (SNHL) is estimated to be between 1 to 2 per 1000 live births.[1,2] This disorder has a variety of causes and can be generally classified according to genetic and nongenetic etiologies. Estimates drawn from studies of hearing-impaired populations indicate a genetic basis in approximately 50% of all HL.[3] Approximately 15% to 30% of hereditary hearing impairment (HHI) involves other organ systems and occurs as a syndrome.[4] Nonsyndromic HL occurs in the absence of association with any other systemic manifestations. Identifying syndromic causes of HL may allow for anticipation of associated complications and their appropriate management. This often involves an extensive evaluation requiring the coordination of a multidisciplinary team of professionals. Thorough evaluation includes a complete case history, physical examination, audiologic testing, radiographic studies, consultation with other specialists, and multiple laboratory testing.

Over 200 different syndromes include deafness or hearing impairment.[5] A number of these syndromes may be detected with diagnostic laboratory tests. Some otolaryngologists routinely order a battery of diagnostic laboratory studies to detect causes of HL. For example, a complete blood count (CBC) is useful to rule out leukemias that may rarely be associated with HL.[6] Platelet studies may exclude Fechner syndrome, a rare autosomal-dominant disorder consisting of macrothrombocytopenia

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and leukocyte inclusions. Autoimmune tests include ANA, ESR, and rheumatoid factor (RF) to exclude lupus and other autoimmune disorders. Thyroid function studies may lead to identification of Pendred syndrome and the need for thyroid supplementation. Blood urea nitrogen (BUN), creatinine, and urinalysis may help exclude Alport's syndrome. Random blood glucose can assess glucose intolerance as a finding in Alstrom syndrome and diabetes. Fluorescent treponemal antibody-absorbed (FTA-ABS) and rapid plasma reagent (RPR) can aid in the diagnosis of syphilis in which prompt intervention may arrest further HL. Medical management of prolonged QT interval associated with Jervall and Lange-Nielsen syndrome identified on electrocardiogram (EKG) can be life saving and avert unnecessary pain and suffering.

Radiographic imaging of the temporal bone can identify inner ear malformations that may be responsible for hearing impairment. Computed tomography (CT) of the temporal bone is the first-line recommended imaging modality for SNHL.[7] Even infants with nongenetic causes have a high incidence of radiologic findings. When asymmetric SNHL is present, magnetic resonance imaging (MRI) is superior to identify retrocochlear pathology. In the current managed-care environment, it is important to investigate the use of obtaining appropriate laboratory studies in evaluating a child with HL. We reviewed our experience in a tertiary-care university hospital in an effort to identify if laboratory studies and radiologic imaging were effective in identifying the etiology of a newly diagnosed HL.

MATERIALS AND METHODS

We conducted a retrospective analysis of 114 (54 female, 60 male) children with unidentified causes of SNHL. The patients were seen between 1998 and 2000 at the University of California, San Francisco Medical Center and were less than 18 years of age. Patients with SNHL attributed to recurrent otitis media, maternal cytomegalovirus, rubella, and toxoplasmosis were excluded. The following clinical information was retrieved from clinical notes, hospital charts, and outside records and recorded on a clinical data sheet: demographic information, pertinent prenatal, perinatal, and postnatal factors (e.g., gestational diabetes, low birth weight); family history of HL; CT and MRI findings; and laboratory tests. The following tests were reviewed: hematocrit (HCT), platelet count (PLT), ANA titer, RF, ESR, free thyroxine (FT4), thyroid-stimulating hormone (TSH), triiodothyronine (T3), BUN, creatinine, urinalysis, random blood glucose, FTA-ABS, RPR, and EKG. CT and MRI findings were also reviewed. An etiology for the HL was noted if the clinical staff was able to establish a diagnosis. The data were tabulated and analyzed with Microsoft Access 2000. Subgroups within the study group were compared using χ^2 analysis.

RESULTS

Of the 114 patients with HL, 54 were female and 60 were male. The age range was 1 year to 18 years of age with a mean and median of 9. Audiometric results were available in the medical records of 111 of 114 patients. Ninety-five patients (83%) had bilateral SNHL and 13 (11%) had unilateral SNHL. The severity of hearing impairment was moderate to profound HL in 81% of patients, whereas the remainder had mild HL. The vast majority had SNHL; however, mixed HL was found in six patients.

Physical examination revealed 6 (5%) children with craniofacial abnormalities, 6 (5%) children with ear deformities, and 6 (5%) children with musculoskeletal anomalies. One patient with bilateral microsomia, torticollis, ear tags, and strabismus was diagnosed with cervico-oculo-acoustic (Wildervanck) syndrome.

Laboratory Testing

The general panel of laboratory tests to evaluate syndromic causes of SNHL, obtained in most patients, consisted of CBC, platelet count, BUN, creatinine, glucose, ANA, RF, ESR, TSH, and FTA-ABS (Table I). EKG and urinalysis were obtained in some but not all patients and were based on the clinician's judgment. The following are the number of abnormal labs found as a percentage of the total number of labs obtained: ESR 14/62 (23%), ANA 14/66 (21%), CBC 10/80 (13%), urinalysis 4/42 (10%), creatinine 3/77 (4%), BUN 3/76 (4%), TSH 2/66 (3%), RF 1/59 (1.7%), RPR 0/13 (0%), FT4 0/44 (0%), and T3 0/7 (0%). CBC, platelet count, BUN, creatinine, blood glucose, and thyroid function test were rarely abnormal and never associated with clinical disease. Blood tests for autoimmune disease were often positive but did not correlate with clinical disease; ESR and ANA were elevated in 22% of

TABLE I.
Selected Laboratory Tests Ordered for Evaluation of Syndromic Hearing Loss.

Lab Test Ordered	No. of Tests Ordered	No. of Abnormal Tests	Associated Syndromes or Etiology
ANA	66	14	Autoimmune, lupus, etc
Rheumatoid Factor	59	1	Autoimmune, rheumatoid arthritis
ESR	62	14	Autoimmune, lupus, etc
CBC	80	10	Anemia, infections
Platelet count	80	4	Fechtner syndrome
BUN	76	3	Alport's syndrome
Creatinine	77	3	Alport's syndrome
Urinalysis	42	4	Alport's syndrome
Glucose (serum)	56	0	Diabetes, Alstrom syndrome
FT4	44	0	Pendred syndrome, cretinism
TSH	66	2	Pendred syndrome, cretinism
T3	7	0	Pendred syndrome, cretinism
FTA-ABS	46	0	Syphilis
RPR	13	0	Syphilis
EKG	15	1	Jervell and Lange-Nielsen syndrome

*This patient had family history of Jervell and Lange-Nielsen syndrome.
ANA = antinuclear antibody; ESR = erythrocyte sedimentation rate; CBC = complete blood count; BUN = blood urea nitrogen; FT4 = free thyroxine; TSH = thyroid-stimulating hormone; T3 = triiodothyronine; FTA-ABS = fluorescent treponemal antibody–absorbed; RPR = rapid plasma regent; EKG = electrocardiogram.

cases. Of the 14 patients with positive ANA, the titer was 1:320 in 4 patients, 1:80 in 4 patients, and 1:40 in 6 patients. The 4 patients with the highest titer (1:320) were referred to pediatric rheumatologists; however, none of those referred resulted in identification of the etiology of HL. Urinalysis was normal in nearly every case.

Electrocardiography

Only one abnormal finding (1 of 15) aided in determining the etiology of SNHL: an EKG with a prolonged QT interval resulted in the diagnosis of Jervall and Lange-Nielsen syndrome.

Radiologic Imaging

Ninety-seven of the 114 patients with SNHL underwent radiologic imaging: 76 underwent CT, 14 had CT and MRI, and 7 underwent MRI alone. Radiologic abnormalities were present in 38 of 97 cases (39%), with 33 (37%) CT scans and 7 (33%) MRIs identified as abnormal (Table II). Among the abnormal CT scans, 18 demonstrated isolated and 9 showed multiple inner ear malformations. Large vestibular aqueducts were the most common isolated finding (n = 7), followed by lateral semicircular canal dysplasia (n = 5), cochlear dysplasia (n = 3), otic capsular lucency (n = 1), small internal auditory canals (n = 1), and hypoplastic cochlea (n = 1). In the 9 scans with multiple abnormalities, cochlear dysplasia was the most common (n = 7; 78%), followed by large vestibular aqueduct (n = 6; 67%) and lateral semicircular canal dysplasia (n = 6; 67%). The remaining 6 scans showed abnormalities that were not related to the HL. Of the 7 abnormal MRI scans, 4 had findings distinct from the CT with the abnormality primarily involving the central nervous system. These findings included a lipoma within the quadrigeminal cistern, a possible fistulous connection between the internal auditory canal and temporal bone, a flattened floor in the posterior fossa, and butterfly vertebrae. In 2 cases, there were abnormalities of the inner ear; one was consistent with bilaterally large vestibular aqueducts and the other had prominent bilateral vestibules consistent with a Mondini malformation. Abnormal CT or MRI scans were not associated with gender, family history of hearing impairment, or presence of external ear abnormalities ($P > .05$).

DISCUSSION

Early identification and appropriate treatment of HL in infants is critical for normal development. Although universal newborn hearing screening programs are leading to the increased diagnosis of HL,[2] identifying the etiology in children can be challenging. A thorough investigation of the child may include laboratory testing, imaging studies, and referral to other specialists. These evaluations often result in extensive and expensive assessments in trying to identify the cause of hearing impairment. HL may exist as part of a syndrome and its identification may allow for anticipation of associated complications and appropriate management.

Initial evaluation in identifying children with HL should begin with a comprehensive clinical history. A thorough pregnancy and postnatal history should be elicited to identify risk factors associated with HL (Table III). Approximately 25% to 33% of childhood SNHL results from environmental or nongenetic causes, including intrauterine infections, maternal metabolic disorders, ototoxic therapeutic agents (aminoglycosides), maternal alcohol or illicit drug use, and exposure to teratogens (e.g., Accutane [Roche Pharmaceuticals, Nutley, NJ], thalidomide).[8] In comparison, our series had 22% of SNHL attributed to environmental or nongenetic causes. This number may be a conservative estimate because we excluded SNHL at-

TABLE II.
Radiologic Findings in Children With Sensorineural Hearing Loss.

Finding	No.	Percent
Normal	59	60.8
Isolated CT abnormalities		
Large vestibular aqueduct	7	7.2
Lateral semicircular canal dysplasia	5	5.1
Cochlear dysplasia	3	3.1
Otic capsular lucency	1	1.0
Small internal auditory canals	1	1.0
Hypoplastic cochlea	1	1.0
Multiple CT abnormalities		
Cochlear dysplasia	7	7.2
Large vestibular aqueduct	6	6.2
Lateral semicircular canal dysplasia	6	6.2
MRI findings distinct from CT		
Lipoma within quadrigeminal cistern	1	1.0
Fistulous connection	1	1.0
Flattened posterior fossa floor	1	1.0
Butterfly vertebrae	1	1.0

CT = computed tomography; MRI = magnetic resonance imaging.

TABLE III.
Syndromic and Non-syndromic Hearing Loss and Associated Clinical History.

Pregnancy history
Intrauterine infections: toxoplasma, rubella, CMV, herpes simplex virus, HIV, syphilis
Maternal metabolic disorders: diabetes, hypothyroidism
Toxic agents: alcohol, tobacco, methyl mercury, ototoxic medications (aminoglycosides, furosemide)
Birth history: hypoxia, hyperbilirubinemia, persistent pulmonary hypertension, extracorporeal membrane oxygenation, low birth weight, prolonged perinatal hospital stay
Postnatal history
Noise-induced hearing loss, head trauma, infections (measles, mumps, meningitis), systemic illness (hypothyroid, Kawasaki disease), fainting spells, altered visual acuity
Family history
Familial patterns of hearing loss through at least two generations
History of consanguinity
Familial pattern of other health problems (e.g. history of white forelock associated with Waardenburg's syndrome, history of blindness and ataxia may be Usher's syndrome)

CMV = cytomegalovirus; HIV = human immunodeficiency virus.

tributed to maternal infections such as rubella, toxoplasmosis, and cytomegalovirus. Our findings included 9 (8%) born premature, 6 (5%) with low birth weight (<1500 g), 3 (3%) with ototoxic medications, and 3 (3%) with abnormal Apgar scores. Of these patients, none had abnormal laboratory findings. However, 5 of these 21 patients (24%) had abnormal CT findings (4 with semicircular canal dysplasia and 1 with cochlear dysplasia). Maternal systemic disorders such as gestational diabetes or hypothyroidism should be documented. Craniofacial anomalies and HL can be associated in infants of diabetic mothers.[9] In our series, 3 (3%) were born to mothers with gestational diabetes. Potential toxic agents (e.g., alcohol, cocaine) or use of certain medications (e.g., aminoglycosides, antimalarials) may result in auditory system injury.

A careful family history helps screen for hereditary hearing impairment. We found 17% reporting a family history of HL. Of these, one case led to the diagnosis of Jervall and Lange-Nielsen syndrome. This patient had an older sibling who had died from this particular syndrome. Knowing that it is inherited in an autosomal-recessive pattern, obtaining a screening EKG was appropriate and revealed a prolonged QT interval.

Systematic physical examination can help uncover cases of syndromic hereditary hearing impairment.[10] Physical findings associated with syndromic HL should be referred for evaluation to a multispecialty clinic. Of the 6 patients in our study with craniofacial abnormalities, 4 were referred for consultation with a specialist; the 2 not referred had findings not associated with syndromic HL (e.g., cleft palate). Referral to specialists is useful when a child has patterns of physical features associated with syndromes that include HL.

Until routine gene identification tests are available to identify hereditary hearing impairment, no individual or set of laboratory tests is universally recommended for screening. We have previously recommended a comprehensive battery of tests, including CBC, thyroid function tests, glucose, syphilis serologies, autoimmune tests, urinalysis, EKG, ophthalmologic consultation, and radiographic examination, to rule out syndromic HL.[3] However, similar to the findings of Ohlms et al.,[11] our current study suggests that routine laboratory evaluation should be reconsidered, given its low diagnostic yield, in the evaluation of children with unexplained SNHL.

A total of 10 children out of 80 (13%) had abnormal CBCs in our study; however, all of the abnormal findings were borderline-low hematocrits ranging from 32 to 34, which did not correspond to the cause of HL. Few blood disorders in children have been associated with HL. Otologic manifestations of leukemias and lymphomas are rare and have only been reported in case reports.[6,12–17] The HL associated with leukemias is thought to be the result of hyperviscosity or infiltration of the temporal bone interfering with cochlear nerve conduction.[18] Even more rare is the presentation of HL as the initial manifestation of leukemia. We were only able to find one such case report in the literature.[15] The decision to order a CBC should be based on clinical and physical findings that might indicate acute leukemia. Most patients with acute lymphoblastic leukemia present with a history of being ill for several weeks and associated physical findings often include gingival bleeding and bone and joint pain. In the absence of signs or symptoms indicating acute leukemia, the CBC is likely to be low yield and uninformative.

Platelet studies are ordered to exclude hereditary macrothrombocytopenia; also known as Fechner syndrome. This disorder is a variant of Alport's syndrome and is associated with high-frequency sensorineural HL, proteinuria, macrothrombocytopenia, and ocular disease. It is inherited in an autosomal-dominant pattern and the clinical manifestations are usually present in varying combinations in several members of the same family.[19] The genetic defect has been localized to chromosome 22[20] and the responsible gene MYH9 has also been identified.[21] Although the exact mechanism of HL is unknown, it is thought to be the result of a cochlear defect and not a neural defect.[22] Our experience revealed only one abnormal platelet count (1.5%) that was not associated with the HL. Fechner's syndrome is extremely rare. In the absence of a family history, routine platelet studies may be of low value.

HL can be associated with lupus and juvenile rheumatoid arthritis (JRA).[23–25] Laboratory tests commonly ordered for these disorders and other autoimmune causes of HL include ANA, RF, and ESR. However, ESR has little diagnostic value in rheumatoid arthritis.[26] Body temperature has been correlated with elevated sedimentation rate,[27] and illnesses such as upper respiratory infections or urinary tract infections common in children may have contributed to the high false-positive findings in our series. In fact, we found elevated ANA and ESR in 22% of cases; however, none of these abnormal findings led us to a diagnosis of an autoimmune disease or a specific cause of SNHL. ANA is highly sensitive for lupus but the positive predictive value was only 11% in one retrospective series.[28] In fact, positive ANA are found in normal blood donors, with up to 32% of normal individuals being positive at 1:40 serum dilution 5% at 1:160 dilution and 3% of normal individuals at 1:320 serum dilution.[29] Our study had a total of 14 patients with elevated ANA titers. The 4 patients with the highest titers (1:320) were referred to rheumatology specialists, but the etiology of HL was not identified after consultation. Similarly, RF is an insensitive test in determining the cause of HL. The sensitivity and positive predictive value for RF in detecting juvenile rheumatoid arthritis has been reported to be 5% and 0.7%, respectively, indicating that RF is an insensitive test in diagnosing this disorder.[30]

Blood glucose can identify juvenile diabetes that may be associated with HL.[31] Several different genetic syndromes have the co-association of HL and diabetes. These include: Wolfram or DIDMOAD (diabetes insipidus, diabetes mellitus, optic atrophy, and SNHL) syndrome; syndrome of maternally inherited diabetes (representing 1%–3% of all cases of type 2 diabetes mellitus) and deafness; thiamine-responsive megaloblastic anemia, diabetes, and deafness syndrome, among others. It may also exclude Alstrom syndrome, a rare autosomal-recessive inherited disorder. Only 50 cases have been reported since the syndrome was first described in 1959. This syndrome is characterized by obesity, impaired glucose tolerance

with insulin resistance, retinal degeneration, neurosensory deafness, acanthosis nigricans, hepatic dysfunction, and other endocrine abnormalities.[32] Given the low prevalence of Alstrom and other deafness and diabetes syndromes, ordering blood glucose as a routine screening examination for these disorders may be of low use. In addition, it is unusual for HL to be the initial presentation of juvenile diabetes.

Thyroid studies can be obtained to exclude Pendred syndrome, an autosomal-recessive disorder associated with familial goiter and congenital deafness. However, thyroid function may also be normal. There is no specific biochemical marker of this disease, and the diagnosis depends on the demonstration of the triad of congenital sensorineural HL, goiter, and abnormal perchlorate discharge test. Pendred syndrome is caused by mutations within the putative Cl^-/I^- ion transporter gene (PDS gene), located on chromosome 7q.[33] Radiologically, Pendred patients may have Mondini deformity and a large vestibular aqueduct. Given the rare abnormalities on thyroid function tests in our study, we recommend that they are obtained only in the presence of clinical signs and symptoms of hypothyroidism, presence of goiter, or when there is radiologic evidence of large vestibular aqueduct or Mondini deformity.

In 1995, Grundfast and Lalwani recommend urinalysis in all children with HL to assess for proteinuria or hematuria associated with Alport's syndrome.[3] This disorder can be X-linked, autosomal dominant, or autosomal recessive, and is associated with a slow, progressive, bilateral SNHL and progressive renal failure. Mutations in collagen genes COL4A3, COL4A4 and COL4A5 have been associated with Alport's syndrome.[34] It is unclear whether this defect in collagen is the cause of HL. Deafness is frequently but not universally associated with the Alport's renal lesion, occurring in approximately 55% of males and 45% of females with the disease.[35] Persistent microscopic hematuria is a constant feature in male patients and is probably present from birth. Many patients also have episodic gross hematuria, precipitated by upper respiratory infections, during the first two decades of life. One of the most common abnormalities is pigmentary changes in the perimacular region, consisting of whitish or yellowish granulations surrounding the foveal area.[36] Given the clinical characteristics of this disorder, obtaining routine BUN and creatinine as an indicator for renal disease is not as useful as urinalysis, which would reveal proteinuria or microscopic hematuria long before elevations in BUN or creatinine. Despite the convenience and low cost in obtaining a urinalysis, it should not be routinely ordered without a clinical history that might indicate the presence of Alport's (e.g., family history of slow progressive HL in males, episodes of gross hematuria). Our experience revealed a total of 10 abnormal results (5%) in BUN, creatinine, and UA. None of these patients had Alport's or other identifiable cause of HL associated with renal insufficiency.

Jervall and Lange-Nielsen syndrome, also known as long QT syndrome (LQTS), is a disorder thought to result from mutations in genes that encode proteins forming the delayed rectifier potassium channel. The responsible genes have been identified as KVLQT1 and KCNE1, located on chromosome 11 and 21, respectively.[37,38] Mutations in these genes encoding cardiac ion channels result in delayed myocellular repolarization.[39] It is a rare disorder with an estimated incidence of 1.6 to 6 cases per million.[40] However, identification of patients with LQTS can be lifesaving. An EKG may be valuable when a history of syncope, arrhythmias, or a family history of sudden death in a young child is elicited.

In evaluating children with unexplained SNHL, radiologic studies such as high-resolution CT and MR imaging have made it possible to identify a specific cause of auditory impairment.[11,41–45] In general, CT is the preferred modality for delineating bony pathology of the temporal bone such as trauma, inner ear bony dysplasia, otosclerosis, and erosive and destructive lesions of the temporal bone. On the other hand, high-resolution MR imaging provides excellent resolution of the membranous labyrinth, and imaging of lesions of the central auditory pathway in the internal auditory canal, cerebellopontine angle, brainstem and cerebral cortex.

Our study suggests that CT imaging is useful in evaluation of children with SNHL; however, there is insufficient data to define a role for MRI based on our investigation. High-resolution CT scanning of the temporal bone demonstrated abnormal findings in 33 (37%) patients in our series. Temporal bone CT may be helpful in certain syndromes, such as Pendred syndrome in which Mondini cochlear abnormality and large vestibular aqueduct (LVA) has been described. LVA is the most common imaging abnormality detected in children with SNHL. Similarly, the most common abnormality we found in our imaging studies was large vestibular aqueduct (n = 13; Fig. 1). At least 40% of children with LVA will develop profound SNHL.[46] The identification of LVA should raise the suspicion of Pendred syndrome and thyroid abnormality. The presence of LVA may also indicate additional malformations and has been associated with Stapes

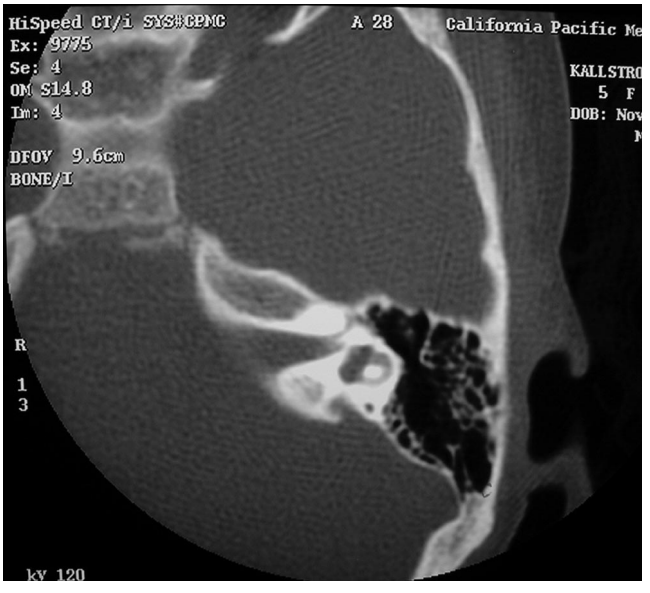

Fig. 1. Large vestibular aqueduct on axial CT scan.

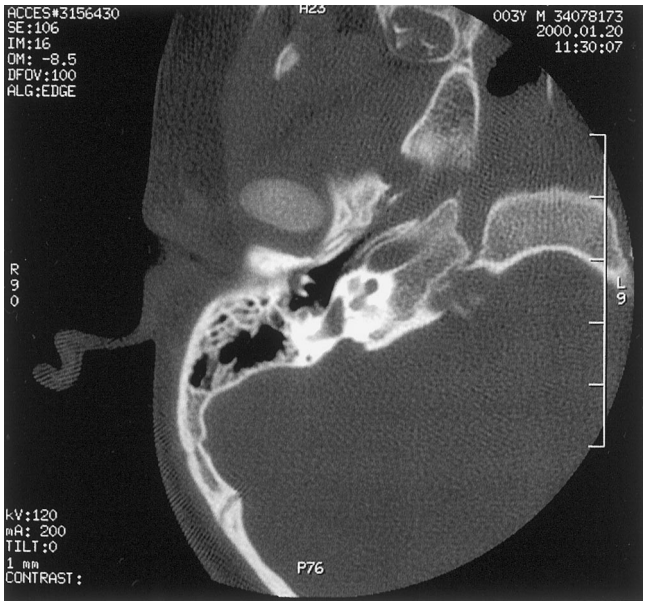

Fig. 2. Large vestibular aqueduct and lateral semicircular canal dysplasia on axial CT scan.

Gusher syndrome, lateral semicircular canal dysplasia, and Mondini deformity[47] (Fig. 2). Patients with LVA are at risk for progressive HL after minor head trauma. Identifying this anomaly influences parent counseling with respect to the dangers of incidental head trauma.[48–50] This suggests that all patients should undergo radiologic imaging as an integral component of evaluation of SNHL.

CONCLUSION

In the evaluation of children with unexplained SNHL, routine laboratory evaluation should be reconsidered given its low diagnostic yield. Clinical suspicion should lead to specific testing. Radiologic abnormalities of the inner ear are common and contribute significantly to the identification of the etiology of SNHL in childhood. Identification of inner ear malformations has direct impact on management of these children, suggesting that all children should undergo radiologic imaging as an integral component of evaluation of SNHL.

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