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Epidemiology of Unilateral Sensorineural Hearing Loss with Universal Newborn Hearing Screening

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Abstract

Objective—Compare the epidemiology of pediatric Unilateral Sensorineural Hearing Loss before and after implementation of Universal Newborn Hearing Screening in Missouri.

Study Design—Inception cohort

Methods—Charts of 134 children born between January 1 1990 and December 31 2007 diagnosed with Unilateral Sensorineural Hearing Loss at a single institution in Missouri were reviewed to determine the effects of Universal Newborn Hearing Screening on age of detection and etiology of hearing loss.

Results—Mean age of detection declined from 4.4 (SD 1.8) to 2.6 (SD 2.6) years of age while rate of detection by six months of age increased from 3% to 42%. The majority (58%) of cases had normal hearing at birth. The most common etiological category was unknown (41%) before screening and congenital (45%) after screening. The use of Magnetic Resonance Imaging has increased by 21% (2-fold) while use of Computed Tomography has declined by 8% since 2002. Yields of Connexin, Pendred, electrocardiogram, and syphilis testing were 0/48 and 2/31 before and after screening respectively.

Conclusion—Implementation of Universal Newborn Hearing Screening in Missouri is associated with a decrease in age of detection of Unilateral Sensorineural Hearing Loss. The majority of cases are either not present or not detectable at birth. The combination of hearing status at birth and imaging findings suggests that the majority of cases are congenital rather than of unknown etiology.

Keywords

Unilateral; Sensorineural; Hearing; Deaf; Children; Newborn; Screening

INTRODUCTION

Historically, unilateral hearing loss (UHL) was considered to have few, if any, adverse functional consequences on children. However, since the early 1980s, many studies have been done showing the adverse effects of UHL on speech and language development, behavior, and academic achievement.^{1–6} Prior to the implementation of Universal Newborn Hearing Screening (UNHS), Unilateral Sensorineural Hearing Loss (USNHL) often went

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undetected until school-age unless specific predisposing conditions such as meningitis prompted earlier evaluation.^{3,7-9}

All the published epidemiologic studies focusing on the age of detection and etiologies of UHL were performed in the pre-UNHS era.^{3,7-9} The implementation of UNHS allows for better delineation of the timing of onset and better differentiation between congenital and acquired etiologies. The goal of the current study was to compare children with USNHL diagnosed before and after the implementation of UNHS.

MATERIALS & METHODS

Design

Inception cohort

Patient population

The Human Research Protection Office at Washington University Medical Center provided a waiver of informed consent for this study. Saint Louis Children's Hospital (SLCH) Department of Audiology provides diagnostic testing for 23% of the children who follow-up after failing newborn hearing screening in Missouri (Personal communication: Kris Grbac M.S. Newborn Hearing Screening Project Director, Missouri State University, Department of Communication Sciences and Disorders, 901 S. National Ave., Springfield, MO 65897). The charts of children seen at SLCH Department of Audiology and the Washington University Division of Pediatric Otolaryngology with an audiogram documenting Unilateral Sensorineural Hearing Loss (USNHL) were reviewed. Children were included in the study if they were born in Illinois or Missouri between December 1 1993 and December 31 1999 or between January 1 2002 and December 31 2007. Children with unilateral mixed hearing loss were included due to the presence of a USNHL component.

Definition of pre- and post-UNHS eras

Universal newborn hearing screening was implemented on January 1 2002 in Missouri and January 1 2003 in Illinois. In the year 2000, 34% and 11% of hospitals in Illinois and Missouri respectively had implemented UNHS. By the year 2002, 98% and 96% of hospitals in the respective states had implemented UNHS.¹⁰ The majority (80%) of children in our cohort were born in Missouri. In order to account for the transition period of UNHS implementation, the pre-UNHS era was defined as prior to December 31 1999 while the post-UNHS era was defined as after January 1 2002.

Definition of Unilateral Hearing Loss

Bone conduction hearing thresholds >30 dB in any three consecutive frequencies in the worse ear. The better ear did not have any three consecutive frequencies with hearing thresholds > 20 dB.

Severity of hearing loss

The three consecutive pure tone thresholds which when averaged gave the largest value were used to define severity of hearing loss. Hearing loss was classified as mild (30–45 dB HL), moderate (46–70 dB HL), severe (71–90 dB HL), or profound (> 90 dB HL). The hearing loss was classified as high frequency if the lowest of the abnormal frequencies was > 2000 Hz, and as low frequency if the highest of the abnormal frequencies was < 2000 Hz.

Age at detection

This was the age at which a USNHL confirmed by auditory brainstem response (ABR) or pure tone audiometry was initially detected by otoacoustic emissions (OAE) testing, auditory brainstem response (ABR) audiometry, or pure tone audiometry.

Temporal bone imaging

Radiologist and Otolaryngologist notes were reviewed to determine whether abnormalities including enlarged vestibular aqueduct (EVA), small internal auditory canal (IAC), cochlear or labyrinthine dysplasia, enlarged cochlear aqueduct (ECA), temporal bone fractures, cochlear nerve aplasia or hypoplasia, or other significant temporal bone abnormalities were detected on Computed Tomography (CT) scan or Magnetic Resonance Imaging (MRI).

Lab work-up

Physician notes and lab reports were reviewed to determine results of the following diagnostic testing: Electrocardiogram (EKG) for long QT interval, Rapid Plasma Reagin (RPR) and Fluorescent Treponemal Antibody Absorbed (FTA-ABS) tests for syphilis, testing for genetic mutations in Connexin 26/30 and Pendred SLC26A4 genes.

Etiologies

Etiology was determined by reviewing physician notes, radiologist notes, and parent surveys. Trauma was assumed to be the etiology if significant head trauma with / without temporal bone fracture occurred prior to the diagnosis of hearing loss. Meningitis was assumed to be the etiology if the diagnosis was documented in the chart prior to diagnosis of USNHL. Congenital etiology included all patients with inner ear abnormalities on imaging or with otherwise unexplained USNHL present at birth. Hereditary was assigned if an immediate family member had a diagnosis of SNHL or if genetic testing was abnormal. 'Risk factors' was assigned as the etiology if there was no clear etiology in the presence of any of the following: Neonatal Intensive Care Unit (NICU) stay, prematurity (<37 weeks), ototoxic medications, jaundice requiring treatment, confirmed autoimmune disease, encephalopathy, brain infarcts, hydrocephalus, and syndromes associated with hearing loss. Cytomegalovirus (CMV) was assumed to be the etiology in cases where the mother or child tested positive for the virus prior to detection of hearing loss.

Statistical Analysis

Data was analyzed using IBM SPSS Statistics, Release 20.0.0, Chicago: IBM Corporation. Data distributions were reported as means with standard deviations for continuous variables and proportions for categorical variables. Statistical testing for difference between the means was done using the t-test with 95% confidence intervals (CI) reported. Chi-squared testing was used for statistical hypothesis testing with categorical variables. A two-sided *p* value of 0.05 was considered to be statistically significant.

RESULTS

Demographics and clinical characteristics

There were 134 children diagnosed with USNHL: 67 before, and 67 after the establishment of UNHS. Gender, race, side and severity of hearing loss, and birthplace were statistically identical in both groups. Half of patients were male, two-thirds white, two-thirds had severe-profound hearing loss, and 80% were born in Missouri. Seven percent of pre-UNHS cases underwent hearing screening at birth. (Table 1)

Age at detection

The mean age of detection was 2.6 years (SD 2.6) after UNHS. This is 1.8 years earlier (95% CI 1.0, 2.5) than age of detection prior to UNHS. Stratified by severity, mild-moderate and severe-profound hearing losses were detected 2.3 years (95% CI 0.8, 3.8) and 1.5 years (95% CI 0.5, 2.4) earlier respectively. Age at detection was statistically identical for mild-moderate and severe-profound hearing loss within each group. The percentage of USNHL detected before 6 months of age was 3% before and 42% after UNHS. In the post-UNHS group, hearing loss that was not detected at birth was detected by school screening (39%), parents (28%), and primary care physicians (8%). (Table 2 & 3, Figure 1)

Etiologies

There were significant changes in the distributions of etiologies for USNHL in patients born before and after UNHS. Prior to UNHS, the most common etiology was 'unknown' (41%), followed by 'congenital' (27%) and 'risk factors' (22%). After UNHS, the most common etiology was 'congenital' (45%), followed by 'unknown' (31%) and finally 'risk factors' (10%). Other etiologies including hereditary, meningitis, CMV, and trauma together accounted for 14% of etiologies in both groups. Subgroup analysis of patients in the post-UNHS group who passed newborn hearing screening suggests that 91% had delayed onset or progressive hearing loss that was not detectable at birth, while 9% had acquired losses due to meningitis and trauma. (Table 4)

Temporal bone imaging

Trends in choice of diagnostic imaging have changed since the implementation of UNHS. Compared to the pre-UNHS era, the frequency of MRI in the work-up of USNHL has increased from 14% to 36% while the use of CT has declined from 73% to 64% over the same period. Distribution of imaging findings was similar in both groups. On CT, EVA and cochlear / labyrinthine dysplasia remain the most common etiologies, with the majority of cases appearing normal. MRI was normal in the majority of cases. Among patients who underwent both CT and MRI, six patients had hearing-related ipsilateral abnormalities. Based on the reports, half were discordant, with MRI missing two cases of EVA and one case of cochlear dysplasia. Review of these three cases showed that the findings were obvious on MRI but were not commented upon in the report. (Table 5, 6)

Lab work-up

Of the total of 79 tests done throughout the study period to look for mutations in Connexin or Pendred SLC26A4 genes, long QT intervals, or presence of syphilis, there were two cases of Connexin 26/30 mutations identified.. (Table 7)

DISCUSSION

Age at detection

This is the first study to specifically examine the age of detection of USNHL in a population of patients born after UNHS. In our study, the mean age of detection was found to be 4.4 and 2.6 years before and after UNHS respectively. Prior to screening, only 3% of patients were identified by age 6 months. Since onset of UNHS, this percentage increased to 42%. The pre-UNHS results are similar to previous publications documenting a mean age of diagnosis of greater than four years, corresponding to school age.^{3,7-9}

Our study shows that while UNHS leads to early detection of many cases of USNHL, there was a significant (60%) proportion of patients who do not have detectable USNHL at birth. Given the high sensitivity of hearing screening, and the fact that the results of 97% of the

post-UNHS newborn hearing screens were known, it suggests that patients who passed their hearing screens and were later found to have hearing loss may have simply developed the hearing loss later. The exact time of onset of the majority (91%) of these patients is unknown due to the absence of obvious triggering events. In this group of patients, at least 39% were missed by caregivers and primary care physicians and were not detected until school age screening was performed. This finding underscores the continued importance of parents and physicians in detection of USNHL in the post-UNHS era. It also suggests a role for pre-school age (2–4 years) hearing screening to allow for intervention prior to school age.

Etiologies

In this study, prior to UNHS, 42% of cases were of ‘unknown’ etiology, followed by ‘congenital’ and ‘risk factors.’ After UNHS, there was a significant shift in the relative prevalence of etiologies; due to the fact that children with no known risk factors were screened, the relative prevalence of ‘risk factors’ dropped by 50%. In addition, the prevalence of ‘congenital’ etiologies replaced ‘unknown’ as the most common etiology in the post-UNHS era. This finding is logical since a child born with isolated significant USNHL would be classified as ‘unknown’ if it was discovered at school age and ‘congenital’ if it was discovered at birth. As expected, implementation of UNHS was not associated with significant changes in the relative frequency of genetic losses or those caused by trauma, meningitis, or CMV infections.

Studies prior to UNHS suggested that 35–50% of unilateral SNHL is of unknown etiology.^{3,9} Among known causes of hearing loss, it is difficult to compare across studies due to differences in categorization, multiple etiologies per patient, and variations in work-ups. The Brookhouser study (1991) was most similar in design to our study and had similar findings: unknown cause (35%), genetics (13%), head trauma (11%), prenatal insult (11%), meningitis (7%), inner ear abnormality (3%), and TORCH (1%).³ The similarity in outcomes suggest that the populations examined were similar pre-UNHS and thus the results of this study can perhaps be applied to their population.

Imaging

Trends in use of CT and MRI in the evaluation of UNHS have evolved at our institution over the last twenty years, with a gradual increase in the use of MRI and a corresponding decrease in the use of CT scans. Temporal bone CT had the greatest yield of all diagnostic tests, with EVA and cochlear / labyrinthine dysplasia being the first and second most common etiologies over both study periods. In cases where both CT and MRI were performed, the majority of cases (26/29) were concordant. The three discordant cases (EVA, cochlear dysplasia) were obvious on MRI, but these bony abnormalities which had already been commented upon in the CT were not again described in the MRI report which tended to focus on soft tissue findings.

In the Brookhouser et al (1991) study, CT was performed in 17% (57/324) and MRI in 2% (7/324) of patients with unilateral SNHL and 3% (10/324) had abnormal findings.³ Simons et al (2006) found 85% (83/98) had a CT scan, 41 (41%) had a MRI, and 36 (37%) had both modalities. Overall, in children with both CT & MRI, 6 (17%) had findings from the CT that would have been missed if MRI was done alone.¹¹ Preciado et al (2005) found 9 of 28 children (43%) with unilateral SNHL had abnormal CT findings.¹² Song et al (2009) found a 25% (82/322) rate of abnormal ipsilateral CT findings in children with USNHL.¹³

Comparison of our results to prior literature first shows that there is tremendous variation in practice patterns, with use of CT varying from 20 to 70% of children with hearing loss. This

variation in practice patterns is reflected in the changing trends seen in our department as a result of variability in provider preferences for imaging modalities in the absence of clear protocols for imaging of hearing loss. EVA tends to be the most common finding on abnormal CT scans. In the study by Song et al (2009), cochlear / labyrinthine dysplasia was the most common abnormality (12%), followed by EVA or EVAS (10%). Compared to our study, the Song et al study population was seen in South Korea, had significantly more cases of profound hearing loss (76% vs. 52%), and did not include children with significant medical conditions, syndromes, or a history of temporal bone trauma. These factors may contribute to the different outcomes in the two studies.

Labs

The overall yield of lab diagnostic testing for USNHL was low. Two of seven were positive for Connexin 26/30 mutations. There were no confirmed cases of Pendred mutations, long QT syndrome, or congenital syphilis. The low test frequency and yield of genetic testing is consistent with the fact that these mutations tend to be associated with bilateral rather than unilateral SNHL.

These findings are very similar to those found in the 2005 Preciado et al prospective study on 150 children with sensorineural hearing loss (SNHL) who underwent full lab work-ups including Connexin 26 (GJB2) screens, syphilitic blood tests, and EKGs.¹² Yields for USNHL patients were 0/21(0%) for Connexin tests, syphilitic screens, and thyroid function tests. They found two abnormal EKGs during screening, but none were diagnostic.

Limitations

The strength of the study was limited by the design and available data. The generalizability of these results may be limited by multiple factors: our institution manages many (23%), but not all the cases of hearing loss diagnosed in Missouri; the most likely etiology is a good but not perfect estimate of true etiology since many patients did not have their hearing tested immediately before events such as meningitis or head trauma led to the discovery of a hearing loss; and there were cases where a newborn received hearing testing after initiation of ototoxic medications, preventing absolute certainty in assigning etiology as congenital or due to risk factors. Assuming that most children are born with normal hearing, etiology tended to favor presence of risk factors rather than congenital when present. The frequency of 'congenital' etiologies is an underestimate since it is known that many patients with congenital hearing loss have progressive losses that may have been undetectable at birth. Finally, distribution of etiologies is imperfect in a study where the work-up was not identical in all patients.

IMPLICATIONS

USNHL has adverse consequences on speech and language development and is often not present or detectable at birth. Pre-school age (2–4 years old) hearing screening would allow for earlier detection and intervention for hearing loss that would otherwise not be detected until school age. At this time, parents and primary care physicians are still critical to the early identification of USNHL.

CONCLUSIONS

Implementation of UNHS in our region was associated with a significant decrease in the age of detection of USNHL. At birth, the majority of cases were either not present or undetectable. Combination of timing of detection and findings on imaging suggests that the majority of cases were congenital.

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Figure 1. Mean age at detection of Unilateral Sensorineural Hearing Loss (USNHL) before and after Universal Newborn Hearing Screening (UNHS)

White: pre-UNHS

Black: post-UNHS

Table 1

Demographics & Clinical Characteristics of Children with Unilateral Sensorineural Hearing Loss (USNHL) before and after Universal Newborn Hearing Screening (UNHS)

	N (%)		p-value
	Pre-UNHS	Post-UNHS	
Total	67	67	--
Gender Male	30 (45)	35 (52)	0.39
Race Black White Other	13 (19) 46 (69) 8 (12)	17 (25) 42 (63) 8 (12)	0.70
Side of hearing loss Right	35 (52)	28 (42)	0.23
Severity of loss Mild Moderate Severe Profound	7 (10) 14 (21) 11 (16) 35 (52)	3 (4) 20 (30) 11 (16) 33 (52)	0.44
State Illinois Missouri	13 (19) 54 (81)	14 (21) 53 (79)	0.83
Screened at birth	5 (7)	66 (99)	< 0.001

Table 2

Mean age at detection of Unilateral Sensorineural Hearing Loss (USNHL) before and after Universal Newborn Hearing Screening (UNHS)

	Age in years (SD)		Difference in years (95% CI)	p-value
	Pre-UNHS	Post-UNHS		
Overall	4.4 (1.8)	2.6 (2.6)	1.8 (1.0, 2.5)	< 0.001
Mild-Moderate	4.3 (2.2)	2.0 (2.5)	2.3 (0.8, 3.8)	0.004
Severe-Profound	4.4 (1.7)	3.0 (2.6)	1.5 (0.5, 2.4)	0.002

*
CI – Confidence Interval.

Table 3

Frequency of case detection by 6 months of age before and after Universal Newborn Hearing Screening (UNHS)

	N (%)		p-value
	Pre-UNHS	Post-UNHS	
Before 6 months	2 (3)	28 (42)	< 0.001
After 6 months	60 (97)	39 (58)	

Table 4

Etiologies of Unilateral Sensorineural Hearing Loss (USNHL) before and after Universal Newborn Hearing Screening (UNHS)

	N (%)		p-value
	Pre-UNHS	Post-UNHS	
Congenital	18 (27)	30 (45)	0.13
Unknown	27 (41)	21 (31)	
Risk factors	14 (22)	7 (10)	
Other	5 (9)	2 (3)	
Hereditary	2 (3)	3 (5)	
Meningitis	0	2 (3)	
CMV	1 (2)	2 (3)	
Trauma			

* Column total > 100% due to accumulated rounding error.

Table 5

Imaging performed before and after Universal Newborn Hearing Screening (UNHS)

	N (%)	
	Pre-UNHS	Post-UNHS
None	13 (20)	21 (31)
Computed Tomography (CT) only	44 (67)	22 (33)
Magnetic Resonance Imaging (MRI) only	1 (2)	3 (5)
CT & MRI	8 (12)	21 (31)

Table 6

Ipsilateral imaging findings before and after Universal Newborn Hearing Screening (UNHS)

CT scan		
	N (%)	
	Pre-UNHS	Post-UNHS
Normal	34 (63)	34 (77)
Enlarged Vestibular Aqueduct *	8 + 2 (19)	2 + 4 (14)
Cochlear / Labyrinthine dysplasia	2 + 2 (7)	0 + 4 (9)
Small Internal Auditory Canal	0 + 3 (6)	1+1 (4)
Enlarged Cochlear Aqueduct	2 + 1 (6)	0 + 1 (2)
Temporal bone fracture	1 (2)	1 (2)
Other	3 (6)	1 (2)
Multiple	4 (7)	5 (11)
Total	54	44
MRI		
	N (%)	
	Pre-UNHS	Post-UNHS
Normal	10 (63)	22 (81)
Enlarged Vestibular Aqueduct	0+1 (6)	1 (4)
Cochlear / Labyrinthine dysplasia	1+1 (12)	0
Absent / Hypoplastic cochlear nerve	0	1 (4)
Other	4 (25)	2 (7)
Unknown	0	1 (4)
Multiple	1 (6)	0
Total	16	27

* a + b represents patients with isolated (a) versus multiple (b) findings on MRI

Table 7

Yield of diagnostic tests before and after Universal Newborn Hearing Screening (UNHS)

	Pre-UNHS		Post-UNHS	
	N	Abnormal n (%)	N	Abnormal n (%)
Connexin 26/30	9	0	7	2 (29)
Pendred SLC26A4	4	0	4	0
EKG (QT interval)	25	0	12	0
Syphilis	10	0	8	0