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Evidence of Vestibular and Balance Dysfunction in Children With Profound Sensorineural Hearing Loss Using Cochlear Implants

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Objectives/Hypothesis: Similarities between the peripheral auditory and vestibular systems suggest that children with sensorineural hearing loss (SNHL) may demonstrate vestibular and balance impairments. This hypothesis was studied in 40 children with severe to profound SNHL and unilateral cochlear implants.

Study Design: Prospective cross-sectional study with repeated measures.

Methods: Vestibular function was assessed with caloric, rotational, and vestibular evoked myogenic potential testing; balance was assessed using the balance subset of the Bruininks-Oseretsky Test of Motor Proficiency-II, a standardized test of static and dynamic balance.

Results: Horizontal semicircular canal function was abnormal in response to a caloric stimulus in 50% (16/32), with a large proportion of those [6/16 (38%)] reflecting mild to moderate unilateral abnormalities. In comparison, horizontal semicircular canal function in response to rotation was abnormal in 38% (14/37). Saccular function was absent bilaterally in 5/26 (19%) and unilaterally in 5/26 (19%) with vestibular evoked myogenic potential. Age standardized balance abilities were significantly poorer in the study population [$\mu = 12.9 \pm 5$(SD)] compared with normal hearing controls [$\mu = 17 \pm 5$(SD); $P = .0006$] and correlated best with horizontal canal function in response to a rotational stimulus ($P = .004$; $R^2 = 0.24$). SNHL from meningitis was associated with worse balance function than other etiologies.

Conclusions: Vestibular and balance dysfunction occurred in >1/3 of children with SNHL and cochlear implants, and is highly dependent on etiology. Although compliance with all tests was high, rotational chair testing, which assesses higher frequency motion (0.25–5 Hz) and thus more "real world" vestibular function, correlated best with dynamic balance. For this reason, rotational chair testing may represent the test of choice in this population, particularly given that it is amenable to testing children of all ages.

Key Words: Cochlear implant, vestibular function, pediatric, sensorineural hearing loss, rotational chair testing, vestibular evoked myogenic potentials, caloric testing.

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INTRODUCTION

Given the embryological and anatomical connection between the cochlea and vestibular end-organs, children with profound sensorineural hearing loss (SNHL) may exhibit abnormalities of vestibular and balance function. Documentation of vestibular (dys) function in children with SNHL has a long and rich history, which indicates that 20% to 70% of children with SNHL demonstrate an element of vestibular end-organ dysfunction.[1] The prevalence of vestibular dysfunction seems also to correlate with severity of the cochlear loss.[2,3] More recently, evaluation of vestibular end-organ function in the setting of profound SNHL has garnered much attention given its implications for the safety of bilateral cochlear implantation and the potential risk of bilateral vestibular injury that this intervention carries. Although there are numerous reports of vestibular function in profoundly deaf adults receiving

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cochlear implants (CI), there is a paucity of data detailing vestibular function in children with profound SNHL receiving CI. The largest series of children to date was published by Buchman et al. in 2004 (n = 22) and nearly 70% of the pediatric cohort demonstrated absent or low intensity responses to caloric irrigation preoperatively.[4]

Clearly, the importance of vestibular testing is to provide an indication of the child's ability to achieve adequate dynamic balance to participate in daily activities. We have recently shown that children with SNHL and unilateral CI on average demonstrate significantly poorer performance on a test of dynamic balance [Bruininks–Oseretsky Test of Motor Proficiency II (BOT-2)] than their age-matched, normal hearing peers.[5,6] Similarly, impairments in postural control in children with SNHL and hypoactive vestibular rotary responses have been recently demonstrated using dynamic posturography.[7]

Dynamic balance results from a complex interplay between the vestibular end-organs, vision and proprioception. Although the available tests of vestibular end-organ function allow us to assess the function of the horizontal canals (rotational and caloric testing) and the saccule [vestibular evoked myogenic potentials (VEMP) reflects saccular function], however, these tests may fall short of predicting functional outcomes in disease and nondisease states. Given that children with SNHL and CI demonstrate abnormal dynamic balance measured using the BOT-2, we asked if abnormalities would also be found in formal assessments of the vestibular end-organs in the same population.[6] Such assessments, if they can be done in children, would provide important information regarding the etiology of cochleovestibular losses and potential for vestibular dysfunction after unilateral or bilateral CI. Moreover, the information might provide an awareness of potential challenges and initiate or direct habilitation.

Although this information is important to understand early development, it may be difficult to perform traditional tests of vestibular function in very young children. With this in mind, we aimed in the present study to: 1) Assess and correlate vestibular and balance function in children with profound SNHL and unilateral CI; and 2) Define an age appropriate test battery for use in this population. This work builds on our previous findings demonstrating balance dysfunction in a similar cohort,[6] by examining balance function in an additional group of children in whom tests of vestibular end-organ function were also performed. We hypothesized that given the similarities between the peripheral auditory and vestibular systems, children with SNHL would demonstrate vestibular and balance impairments. In addition, static and dynamic balance performance may directly correlate with particular deficits in vestibular end-organ function.

METHODS

Forty children (3–19.3 years) with severe to profound SNHL using unilateral CI underwent vestibular function testing including caloric, rotational, and VEMP testing. Dynamic balance was assessed using the balance subset of the BOT-2.[5] All children had unilateral Nucleus 22, 24M, 24RCS, 24CA, or 24RE devices (Cochlear Corporation, Melbourne Australia). This study was approved by the research ethics board at the Hospital for Sick Children, Toronto, Canada. Written consent and verbal assent were obtained from the guardians and children, respectively. Most children were undergoing assessment for sequential bilateral CI (30/40). There were no exclusions based on age, etiology of SNHL, or sidedness of implant. Children with coexisting uncompensated visual loss or motor deficits were excluded.

TABLE I.
BOT-2 Balance Subtest.

Balance Subtest Items	Raw Score (Max)	Point Score
Standing with feet apart on a line		
Eyes open	10 sec	0–4
Eyes closed	10 sec	0–4
Walking forward on a line	6 steps	0–4
Standing on one leg on a line		
Eyes open	10 sec	0–4
Eyes closed	10 sec	0–4
Walking forward heel to toe on a line	6 steps	0–4
Standing on one leg on a balance beam		
Eyes open	10 sec	0–4
Eyes closed	10 sec	0–4
Standing heel-to-toe on a balance beam	10 sec	0–5
Total Score		0–37

Total score then looked up in normative value table provided with test for given age and gender category and converted to age adjusted score (range 1–35).

BOT-2 = Bruininks-Oseretsky Test of Motor Proficiency II.

Balance Testing

Balance was assessed using the balance subset of the BOT-2,[5] which includes nine items of which four are performed alternately eyes open and closed (Table I). The raw score for each item is converted into a point score and point scores on each of nine items are summed producing a total point score (0–37 points). The total point score and the subject's age is used to obtain an age-matched scale score based on population norms provided by the test (1–35 units) (Table I). Scores used in this study were obtained with the unilateral CI in place and functioning. Although age-matched normative scores are provided a second control group of normal hearing children were tested to estimate the uniformity and replication of the testing environment. This normative group overlapped with a previously published study.[6] A single individual, blinded to vestibular test results, administered the BOT-2 on the same day as vestibular testing in 89% of children and within a month (n = 4) in the remainder.

Saccular Function—VEMP Testing

A 105 dB click was administered through Telephonics TDH-39 headphones (Telephonics Huntington, NY). Recordings were made on a Bio-logic Navigator SE system (Bio-logic Systems Corp., Illinois). Electrodes were placed at the mid-point of the muscle belly of the sternocleidomastoid (SCM) bilaterally and referenced to the ipsilateral mastoid tip. A contralateral head turn was used to activate SCM and if this did not yield a tracing, a head lift activating both SCM's was used. At least 100 sweeps within the acceptance criteria were averaged and averaged signals were replicated. VEMP responses were judged as either present or absent within a range of acceptable latencies of the P_I

N_{II} waveform (P_I: 8.5–14 ms; N_{II}: 15–23 ms) based on published age-appropriate ranges.[8]

Horizontal Canal Function—Caloric Testing

ENG-caloric testing was carried out by the Fitzgerald–Hallpike method of binaural alternating caloric stimulation after oculomotor and positional ENG.[9] Asymmetry was calculated based on peak slow-phase velocities using Jongkee's formula.[10] All ears were inspected and debrided of wax before testing. Abnormal ENG-caloric tests were defined as follows:

1. Unilateral:
   a. Normal: <15% excitability difference (ED)
   b. Mild loss: 15% to 29% ED
   c. Moderate loss: 30% to 50% ED
   d. Severe loss: 51% to 100% ED
2. Bilateral:
   a. Moderate: sum of bithermal caloric responses <20°/S AND responses >3°/S with ice water
   b. Severe: negative ice water caloric response on both sides

CIs were on and activated at the time of caloric testing although the ear-level microphone and was removed from the ear and protected against water.

Horizontal Canal Function—Rotational Testing

Horizontal canal function in response to rotation was assessed using a custom-built hydraulic chair with a 10-hp motor and torque capability of 138 m per kilogram (Zonic Technical Laboratories Inc., Cincinnati, Ohio). Subjects were seated alone, padded and secured coupling head and chair movement tightly with the body. Implants and hearing aids were removed throughout the test. Horizontal and vertical electro-oculography electrodes were placed at the lateral canthus, cheek and midbrow, with a ground on the forehead to measure the vestibuloocular reflex (VOR). Calibration was performed using 10° horizontal gaze deviations. Signals were amplified (band pass 0–30 Hz) and digitized. In 29/36 cases, sinusoidal harmonic acceleration was completed at a maximum rate of 100°/second over a frequency range of 0.25 to 5 Hz. Pseudorandom rotation (0.32–5.01 Hz) was used in four and both were used in three. Two trials were attempted, the first in light with subjects fixated on a distant visual target and the second in dark with fixation on a distant imaginary earth fixed target. Low frequency rotation was defined as ≤2 Hz and high frequency rotation >2 Hz. Calibrated eye and chair position signals were digitized online. During analysis, eye position records were edited for saccades followed by differentiation of chair and cumulative eye positions yielding velocity profiles which underwent power spectral analysis. Normative data were obtained from healthy adult volunteers in our center, all had normal corrected vision and abstained from alcohol and medications 48 hours before testing [n = 30, 8:22 men:women, mean age 27.2 years (range 20–45 years)].[11] Abnormally low and high scores fell beyond the 95% CI for at least two frequencies.

Data and Analysis

SAS v. 9.1 software (SAS, Cary, North Carolina) was used for statistical analyses. Mean age-standardized scale score on the balance subset of the BOT-2 (Table I) of our study population was compared with both normative test data and our separate controls using independent Student *t* test.

Based on the criteria outlined above, VEMP, caloric- and rotational results were categorically defined as normal or abnormal. Caloric data were further broken down into bilateral vs. unilateral loss and rotational testing into low and high frequency categories and defined continuously based on the mean VOR gain in light (VVOR) and dark (VORD). Age standardized scale score on the BOT-2 balance subset was the dependant variable. Univariate regression analysis was performed to determine the predictive nature of each of the vestibular tests on BOT-2 score and potential covariates were introduced into the model (i.e., etiology of deafness, duration of implant use, progressive vs. nonprogressive SNHL, and age at implantation). Significant and borderline significant variables (P = .05) on univariate analysis were then included in multivariate regression analysis. The regression equation, which maximized significance of the predictor variable and best fit of the model was found.

RESULTS

Forty children with profound SNHL and unilateral CI participated. The 22 boys and 18 girls ranged in age from 3 to 19.3 years [$\mu = 9.6 \pm 4.6$(SD)], all were experienced users with >1 year of implant use [$\mu = 5.4 \pm 2.6$(SD)]. Age at implantation was variable with a mean of 5.4 years [±2.6(SD)] as was the etiology and onset of SNHL (Table II). Most children (36/40) had stable SNHL although four experienced delayed onset with progression to profound SNHL (n = 4).

Compliance with Testing

Compliance with testing was high (>88%) and the youngest child on which all testing was attempted and successfully completed was 3 years old. Younger children have not yet been tested in our center at this time. In our study group, a single child (age = 4 year) refused all testing despite two attempts 6 months apart. BOT-2 could not be completed in one child (age = 5.6 year) because of poor attention. Only 2/36 children who completed rotational testing, required repeat visits (ages 4.6 and 8.2 year) and nine were unable to complete testing in dark [mean age 5.1 year ± 1.15(SD)]. Compliance for VEMP testing was high, however, concerns regarding the younger child's ability to generate adequate muscle contraction to obtain reliable results occurred in five [mean age 4.9 year ± 1(SD)]. These results were labeled indeterminate and not used in further analyses. Caloric testing, was accomplished in entirety in 28/31 (90%) children. Three of the 31 (10%) were unable to complete all temperature trials [mean age 4.3 years ± 1(SD)]. All children

TABLE II.
Etiology and Onset of Deafness.

Etiology of Deafness	N (%)
Meningitis	7 (18)
Connexin 26	12 (30)
Unknown	16 (40)
Abnormal cochlea	5 (13)
Mondini	4 (80)
CHARGE syndrome	1 (20)
Total	40
Nature of SNHL	
Nonprogressive	36 (90)
Progressive	4 (10)
Total	40

SNHL = sensorineural hearing loss.

requiring ice water evaluation (n = 9) completed testing. A single child (age = 17 years) experienced nausea and vomiting with caloric testing. Implants were left on and active during caloric testing and greatly facilitated adequate alerting. No artifact because of the implant was noted on eye tracings.

Dynamic Balance Testing

The published age-adjusted mean (n = 1520) for the BOT-2 Balance Subset is 15 ± 5 (scale range 1–35). In comparison, the mean of our normal hearing control group (n = 19) was 17 ± 5 units. There was no statistically significant difference between our control group and the published normative data ($t_{(18.5)} = 1.77$, $P = .09$). In comparison, mean BOT-2 scores for children with SNHL and CI were significantly poorer [$\mu = 12.9 \pm 5.2$(SD)]. The difference between the implant group and both the normative and control group was statistically significant ($t_{(37.3)} = 2.98$, $P = .005$; $t_{(29)} = 2.72$, $P = .01$, respectively).

Horizontal Canal Function—Caloric Testing

Abnormalities of low frequency horizontal canal function detected using caloric stimuli were observed in 50%(16/32), with a large proportion of these (6/16, 38%) reflecting mild (n = 5) to moderate (n = 1) unilateral abnormalities. Nine children (28%) demonstrated complete horizontal canal dysfunction (areflexia) bilaterally and one child had complete unilateral canal dysfunction (areflexia). The majority of the children with bilateral areflexia (5/9, 56%) and the single child with unilateral areflexia had acquired SNHL from meningitis. Of the remaining four children with bilateral areflexia, one had a cochleovestibular anomaly, one had *GJB2* related deafness, one had both *GJB2* mutation and a cochleovestibular anomaly and one had SNHL of unknown etiology with normal temporal bone imaging. Of those with unilateral losses (n = 7), the implanted ear was affected 71% (5/7) of the time, however, this difference did not reach statistical significance (paired t-test, $t_{(30)} = 1.44$, $P = .16$).

Horizontal Canal Function-Rotational Stimulus

Higher frequency horizontal canal function was assessed by measuring the VOR in response to rotation and dysfunction was shown in 14/37 (38%) with the majority (10/14, 71%) having a loss spanning all frequencies (0.25–5 Hz). Four children had bilateral loss of horizontal canal function detectable only at high frequencies of rotation [>1 Hz (2) and >3 Hz (2)]. The VOR gain in dark by frequency for these children is plotted in Figure 1.

Saccular Function

Abnormalities of saccular function (VEMP) was common with 10/25 (40%) demonstrating an abnormality, half occurring bilaterally [5/25 (20%)] and half unilaterally [5/25 (20%)]. In those with unilateral abnormalities, absence of VEMP occurred on the implanted side in 3/5 (60%). Differences in the incidence of abnormalities on the implanted side vs. nonimplanted side were not statistically significant [paired *t*-test: ($t_{(22)} = 1.37$, $P = .19$)].

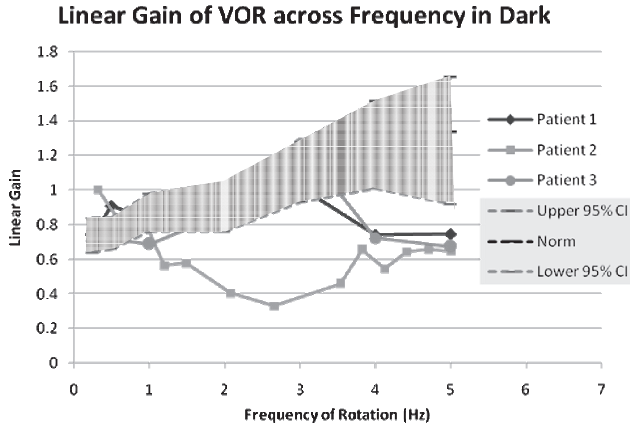

Fig. 1. Linear VOR gain across frequency of sinusoidal rotation in dark for three individual patients with loss of high frequency horizontal canal function (solid lines). Normative data and the 95% CI are also plotted (dashed line). Note the reduced gain that occurs exclusively above 3Hz in patient 1 (♦) and 3 (●) and above 1 Hz in patient 2 (■) signaling an isolated high frequency loss of vestibular function.

Correlation Between Tests of Vestibular End-organ Function

Many children with identifiable losses of horizontal canal function on caloric testing also demonstrated abnormalities on rotational testing and vice versa. This relationship is reflected in the moderate correlation between these tests (VOR-light $F_{(1,\ 23)} = 17.86$, $P = .0003$, r = 0.66; VOR-dark $F_{(1,\ 15)} = 27.46$, $P < .0001$, r = 0.80) and is consistent with the fact that both are testing horizontal canal function although over a different frequency spectrum. In comparison, a child with an abnormality of saccular function (VEMP) was not more likely to demonstrate an abnormality of horizontal canal function (caloric and/or rotation) as indicated by the lack of significant relationship between abnormalities of caloric or rotational and VEMP testing in our population ($F_{(1,\ 27)} = 8.43$, $P = .0074$, r = 0.49; $F_{(1,\ 27)} = 8.43$, $P = .0074$, r = 0.49, respectively).

Correlation Between Vestibular End-organ Function and Balance Ability

In Figure 2, balance ability, (BOT-2 score), is plotted according to horizontal canal and saccular function defined by the categorically [normal vs. abnormal VOR and VEMP; normal vs. abnormal (unilateral vs. bilateral) caloric results]. There is considerable spread in performance but the mean difference is largest when balance ability is grouped according to horizontal canal function in response to rotation. Specifically, horizontal canal function as measured by rotation, correlated significantly with balance ability (BOT-2 performance) ($P = .0036$; r = 0.49).

Given that horizontal canal function in response to rotation was measured both in light and dark, linear regression was repeated using mean VVOR and VORD as predictor variables. The correlation between balance function (BOT-2 score) and horizontal canal function based on rotation remained significant regardless of whether results were defined categorically (normal vs. abnormal) or continuously

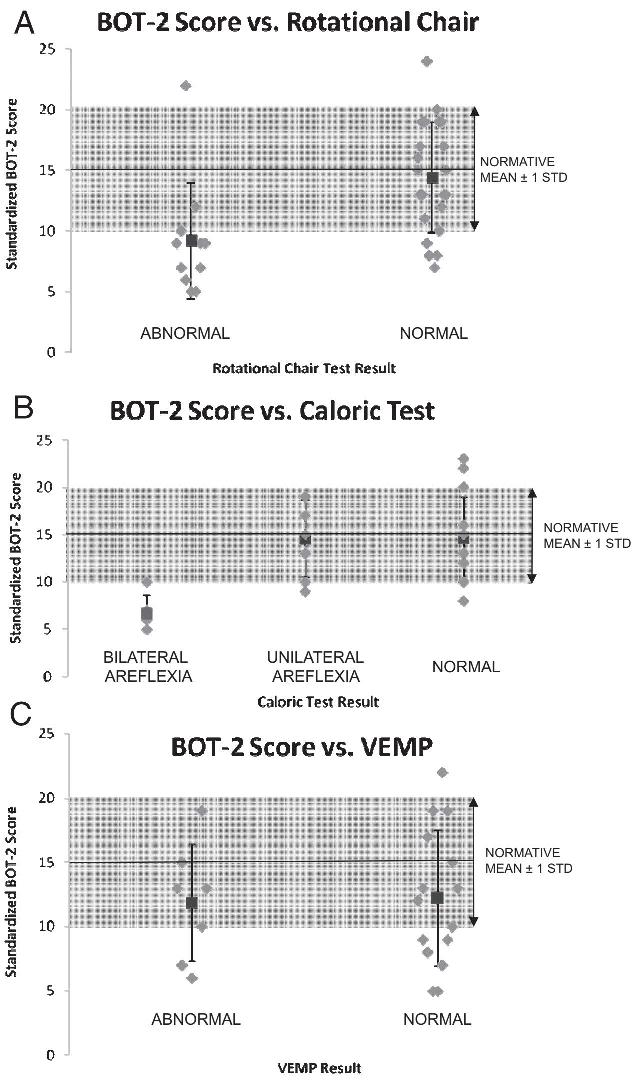

Fig. 2. Age standardized score on balance subset of BOT-2 against results of rotational chair testing (A), caloric-ENG testing (B), and VEMP testing (C). (■ Mean ± 1 SD).

(mean VOR gain in light/dark). Given that four children demonstrated normal gains at low frequencies of rotation, with abnormal gain at high frequencies (Fig. 1), a subanalysis was performed to assess for correlation between balance ability and horizontal canal function at low vs. high frequency. In this analysis, rotational results were defined both categorically (normal vs. abnormal) and continuously (mean gain VVOR and VORD). Linear regression analyses revealed that horizontal canal function during rotation, correlated well with balance function (BOT-2 scores) at high and low frequencies when test results were defined categorically (Table III). This relationship was preserved for mean VORD gain at both low ($\leq$2 Hz) and high ($>$2 Hz) frequency. However, mean VVOR gain was strongly correlated with balance ability (BOT-2 score) at high frequency ($>$2 Hz), it was no longer a good correlate at low frequency ($\leq$ 2 Hz) (Table III).

Abnormalities of low frequency horizontal canal function, as measured by caloric testing, correlated well with balance ability (BOT-2 score) (β = 7.90, SE = 2.07, P = .0008, r = 0.71). However, saccular dysfunction (absence of VEMP) was not correlated with balance ability (BOT-2 score) (P = .88; r = 0.03). Figure 2 plots the relationship between balance ability (BOT-2 score) and abnormalities of low frequency horizontal canal function and saccular function, respectively.

In an effort to control for potentially confounding factors related to our population demographics, additional variables were examined by univariate analysis (Table IV) including etiology of SNHL, age and duration of implantation, nature of SNHL (progressive vs. nonprogressive) and rotation paradigm used (pseudorandom vs. sinusoidal). Age at testing was not considered in the analysis given that BOT-2 scores are age standardized and our previous study demonstrated that this relationship holds across a spectrum of ages and performance.[6]

In Figure 3, balance ability (BOT-2 score), is plotted against age at implantation and duration of implant use. Considerable variability in balance ability across our study population is apparent. Although age at implantation does not significantly affect BOT-2 performance (P = .46, r = 0.13), longer duration of implantation seems to predict significantly poorer balance performance (BOT-2 score) (P = .015, r = 0.4) on univariate analysis.

Balance ability (BOT-2 score) is plotted according to etiology and onset of SNHL (progressive vs. nonprogressive) in Figure 4 which demonstrates a clustering of performance according to etiology of SNHL. Children with meningitis or cochleovestibular anomalies display poorer balance than those with *GJB2* deafness or SNHL of unknown etiology, although the latter two groups also contained a number of poor performers. Differences in balance abilities and scores across etiology are reflected in the significance of etiology as a predictor of balance function (BOT-2 score), on univariate analysis (P = .002, r = 0.61). Only four children in our study had progressive SNHL and Figure 4B demonstrates that 3/4 had very good balance as reflected by their high BOT-2 score, although one child with a progressive loss performed quite poorly. Given this variability and small sample size, nature of the SNHL (progressive vs. nonprogressive) approached but did not reach statistical significance on univariate analysis (P = .06, r = 0.32) (Table III).

In summary, univariate regression indicates that horizontal canal function in response to rotation or caloric stimulation, correlated well with balance ability (BOT-2 score). Balance ability was also significantly correlated with etiology and duration of implant use on univariate analysis. In addition, the correlation between balance ability and onset of SNHL (progressive vs. nonprogressive) was only marginally nonsignificant. Likely many variables found to be significant on univariate analysis are interrelated. To determine their independent influence, significant variables) were introduced into a multivariate regression model. Before proceeding with multivariate regression, however, we must remember that a significant, although not absolute, correlation exists between rotational and caloric testing as discussed above. Given this strong correlation, entering both

TABLE III.
Predictive Ability of the Mean VOR Gain in Light and Dark for BOT-2 Score at Low (≤2 Hz) and High Frequencies (>2 Hz).

		Univariate Analysis		Multivariate Analysis	
Independent Variable	n	*P*	Correlation Coefficient (*r*)	*P*	Multiple R
CHAIR–categorical, low frequencies	32	.0001*	0.63	.0002*	0.75
CHAIR–categorical, high frequencies	32	.006*	0.48	.002*	0.70
VVOR-≤2 Hz	31	.33	0.18	.20	0.57
VVOR->2 Hz	31	.01*	0.46	.005*	0.68
VORD-≤2 Hz	24	.01*	0.50	.009*	0.72
VORD->2 Hz	24	.02*	0.47	.01*	0.72

* Denotes significance at $P < 0.05$.
VOR = vestibuloocular reflex dark; BOT-2 = Bruininks–Oseretsky Test of Motor Proficiency II; VVOR = VOR light; VORD = VOR dark.

parameters, into the regression analysis leads to loss of significance of all variables in question. Therefore our multivariate regression analysis was conducted for three separate models, each of which included different but correlated measures of horizontal canal function, notably: 1) rotational testing in light; 2)rotational testing in dark; and 3) caloric testing.

Controlling for effects of additional variables, the *P*-values related to the slope of each parameter remained largely unchanged from those obtained on univariate analyses (Table V). In this analysis, horizontal canal function measured by mean VORD gain remained the most significant and robust correlate of balance ability (T-2 score) ($P = .005$, multiple R = 0.78) followed by rotational testing in light ($P = .02$, multiple R = 0.70) and finally caloric testing ($P = .03$, multiple R = 0.64). The nature of the SNHL (progressive vs. nonprogressive) remained a significant correlate of balance ability in models that contained mean VOR gain in light and dark (light: $P = .006$, multiple R = 0.70; dark: $P = .04$, multiple R = 0.78) but not in the analysis of caloric testing ($P = .20$, multiple R = 0.64). Neither etiology of SNHL nor duration of implant use were significantly correlated with balance function (BOT-2 score) in any multivariate regression model (Table V).

The best fit regression model (R = 0.77) contained a measure of horizontal canal function, mean VOR gain in dark, and etiology and onset of HL (progressive vs. nonprogressive). Specific statistics for each of the parameters in this analysis are provided in Table VI.

In summary, in children with severe to profound SNHL, a strong relationship exists between static and dynamic balance ability (BOT-2 score), and horizontal canal function particularly when it is measured by mean VOR gain in the dark in response to a high frequency rotational stimulus. Furthermore, etiology and onset of SNHL seems to significantly modulate this relationship.

Impact of Implantation

Although not an ideal model, vestibular assessment in children with unilateral implants does afford a means of testing the impact of implantation by examining side

TABLE IV.
Univariate Linear Regression Analysis of Factors Influencing Standardized Score on Balance Subset of BOT-2, a Test of Dynamic Balance.

Effect	*F*-Value	$df_{(num., denom.)}$	Slope	Standard Error	Correlation Coefficient (*r*)	*P*
VEMP	0.02	1,20	0.34	2.33	0.033	.88
Caloric	10.37	2,20	−7.91	2.01	0.72	.0008*
VVOR	7.46	1,29	14.80	5.42	0.45	.01*
VORD	10.13	1,23	10.45	3.28	0.56	.004*
Etiology	6.40	3,32	7.50	2.37	0.61	.002*
Duration of implant use	6.58	1,34	−0.77	0.30	0.40	.01*
Age at implantation	0.56	1,34	0.18	0.24	0.13	.46
Progressive vs. nonprogressive SNHL	3.83	1,34	5.22	2.67	0.32	.059
Test type	0.49	1,32	1.84	2.63	0.13	.49

* denotes significance at $P = .05$ level.
BOT-2 = Bruininks–Oseretsky Test of Motor Proficiency II; df = degrees of freedom; VEMP = vestibular evoked myogenic potentials; VOR = vestibuloocular reflex dark; VVOR = VOR light; VORD = VOR dark; SNHL = sensorineural hearing loss.

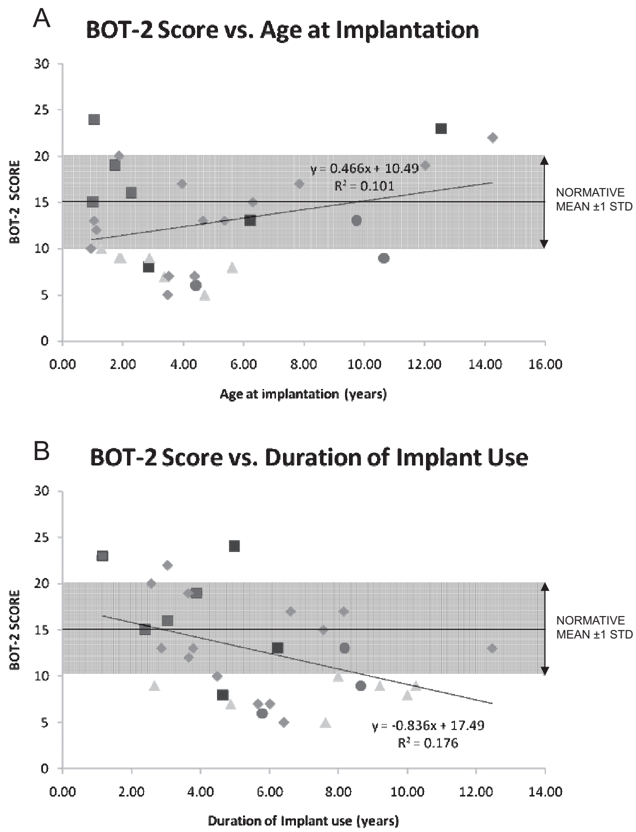

Fig. 3. Age standardized score on balance subset of BOT-2 plotted by age at implantation (A) and duration of implant use (B). Univariate linear regression equation and R^2 shown on graph. ◆ unknown ▲ connexin-26 ■ meningitis ● abnormal cochlea.

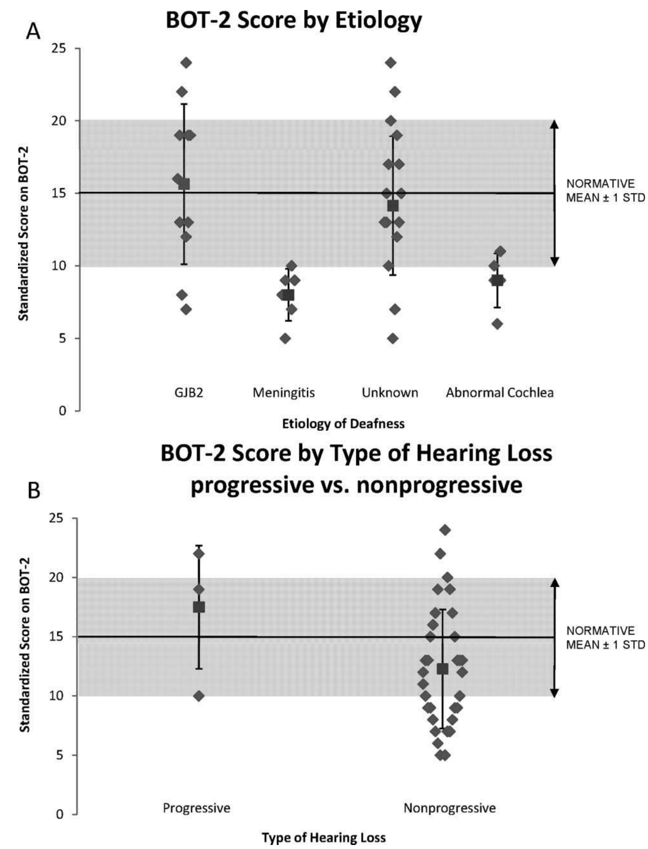

Fig. 4. Age standardized score on balance subset of BOT-2 plotted by etiology (A) and type of HL (progressive vs. nonprogressive) (B) (■ Mean ± 1 SD).

differences in vestibular function. For children undergoing sinusoidal rotation, a paired comparison of the VOR gain in light and dark on the implanted side [light $\mu = 0.90 \pm 0.28$ (SD); dark $\mu = 0.79 \pm 0.41$(SD)] vs. the nonimplanted side [light $\mu = 0.921 \pm 0.29$(SD); dark $\mu = 0.86 \pm 0.58$(SD)] was performed. No significant difference existed between horizontal canal function assessed by individual VOR gains on the implanted side vs. those on the nonimplanted side in either light or dark using a paired analysis across frequencies. Although the difference between horizontal canal function at 3Hz (VORD) appeared significant ($t_{(23)} = 2.26$, $P = .03$), this did not remain significant once levels were adjusted for multiple comparison (Bonferroni adjusted level of significance 0.004).

In many of the children with unilateral horizontal canal hypofunction on caloric testing ($n = 7$), the implanted ear was affected (71% (5/7)). This difference, however, did not reach statistical significance (paired t-test: $t_{(30)} = 1.44$, $P = .16$). Similarly, in those children with unilateral abnormalities in saccular function, the VEMP was absent on the implanted side in 3/5 (60%). The difference between saccular function on the implanted vs. the nonimplanted side was likewise not statistically significant (paired t-test: ($t_{(22)} = 1.37$, $P = .19$)).

DISCUSSION

Compliance with Testing in the Pediatric Setting

Overall compliance with testing was high although the youngest child in whom we attempted testing was 3 years of age. This high degree of compliance, however, underestimates the challenges and time commitment involved in testing and retesting this population in an effort to achieve reliable results. Many minor but important adjustments have been made in our testing center and protocols to accommodate pediatric patients. To our advantage, all participants had unilateral CIs which were left in place wherever possible and appropriate. Although there is redundancy in using both caloric and rotational testing, this was useful in that inconsistencies between tests identified when repeat evaluation would be useful. Proper assessment of the VEMP was questionable in a number of younger s participants. Specifically, absence of a myogenic response in this group may indicate an inability to produce sufficient and consistent tonic activation of the SCM to allow replication. Therefore, yield on VEMP testing was lower given exclusion of cases where muscle activation was deemed insufficient. The ability to obtain reliable VEMPs would be significantly improved by incorporating a commercially available video feedback system that promotes maintenance of ongoing tonic SCM contraction.

TABLE V.
Multivariate Linear Regression Analysis of Factors Influencing Standardized Score on Balance Subset of BOT-2, a Test of Dynamic Balance.

	Multivariate Analysis					
Variable	*F*-Value	$df_{(num., denom.)}$	Slope	Standard Error	*P*	Multiple R
Light						
VVOR	6.71	4,26	12.29	5.74	0.02*	0.70
Etiology	0.48	4,26	0.577	0.84	0.50	
Duration of implant use	0.78	4,26	−280	0.32	0.39	
Type of hearing loss (progressive vs. nonprogressive)	8.88	4,26	8.18	2.75	0.006*	
Dark						
VORD	9.97	4,19	8.81	2.79	0.005*	0.78
Etiology	1.64	4,19	1.26	0.98	0.22	
Duration of implant Use	0.76	4,19	−0.32	0.37	0.39	
Type of hearing loss (progressive vs. nonprogressive)	5.04	4,19	6.69	2.98	0.04*	
Caloric testing						
Caloric testing	5.54	4,22	2.94	1.25	0.03	0.64
Etiology	1.59	4,22	1.09	0.86	0.22	
Duration of implant use	0.33	4,22	−0.22	0.40	0.57	
Type of hearing loss (progressive vs. nonprogressive)	1.79	4,22	3.88	2.91	0.20	

* denotes significance at $P = .05$.
BOT-2 = Bruininks–Oseretsky Test of Motor Proficiency II; df = degrees of freedom; VOR = vestibuloocular reflex dark; VVOR = VOR light; VORD = VOR dark.

In summary, having a hierarchy of tests based on the yield of useful information is essential. From the perspective of yield and compliance, we would argue that a test of dynamic balance (BOT-2) coupled with rotational testing (0.25 to 5 Hz) would be the ancillary tests of choice following history and physical examination and in addition to radiological and genetic evaluation for children with profound SNHL.

Dynamic Balance

Identifying a peripheral vestibular loss based on measures of functional balance in a child can be difficult. Depending on the specific test chosen, children with peripheral dysfunction can appear quite normal as evidenced by the fact that measures of bilateral coordination, strength, or running speed are not affected by loss of peripheral vestibular functional whether complete or partial, symmetrical or asymmetrical.[12] The results of the present work indicate that when children with SNHL are challenged by sufficiently difficult balance tasks that emphasize the contribution of the peripheral vestibular system, deficiencies in function surface. The variety of tasks that comprise the balance subset of the BOT-2 provide this necessary level of difficulty and place emphasis on peripheral vestibular function by removing vision during several tasks. Our results in the current cohort are consistent with those previously published by our group.[6] Certainly as a group, balance ability (BOT-2 performance), of children with SNHL is poorer than the standardized norms, although the range of abilities within this group spans a large spectrum. More importantly on an individual level, the BOT-2 has allowed us to accurately identify which children with profound SNHL have concurrent abnormalities of balance. The balance subset of the BOT-2 is a simple and cost-effective tool requiring only 10 minutes to administer. Given that the score is age standardized it allows for repeated measures, making it ideal to assess the impact of sequential interventions. One con-

TABLE VI.
Best Fit Linear Regression Model.

Effect	*F*-Value	$df_{(num., denom.)}$	Slope	Standard Error	Multiple R	*P*
VORD	11.41	3,23	9.22	2.73	0.77	0.003*
Etiology	1.79	3,23	8.00	0.97		0.0053*
Progressive vs. nonprogressive SNHL	9.79	3,23	1.31	2.56		0.1955

* denotes significance at $P = 0.05$ level.
df = degrees of freedom; VORD = vestibuloocular reflex dark; SNHL = sensorineural hearing loss.

cern is that the minimum age of administration is 4 years and the majority of our pediatric implant candidates are much younger. It therefore remains difficult to obtain pre- and postoperative assessments in many pediatric implant candidates. Computerized dynamic posturography is certainly an alternative to the BOT-2 and likely provides additional information, however, it is more costly, and also does not allow for assessment of very young children.

Incidence of Abnormalities on Vestibular End-organ Testing

Our results indicate an overall incidence of horizontal canal dysfunction on caloric testing of 50%, which is lower than that of previous reports.[4] The incidence of dysfunction is lower still (31%) if those children with only mild caloric losses are excluded from the count. Abnormalities of horizontal canal function based on rotational testing occurred in 38% of subjects and four children had isolated high frequency losses. In many cases, isolated high frequency loss can be symptomatic and missed using caloric or low frequency rotational testing.[13] Our incidence of rotational abnormalities is slightly higher but in line with the 22% found by Suarez et al.[7] using passive low frequency ($<$0.05 Hz) rotation, given that we would expect to identify additional cases using a higher frequency spectrum. A significant proportion of our study population (38%) demonstrated saccular dysfunction indicated by absence of VEMP (bilaterally or unilaterally), which is consistent with a smaller study by Jin et al. who reported that 6/12 children with profound SNHL had abnormalities of saccular function based on VEMP measured preoperatively.[14]

Impact of Cochlear Implantation on Vestibular End-organ Function

No significant side differences were detected in horizontal canal function (caloric or rotational stimuli), or in saccular function (VEMP). There was, however, a trend towards an increased incidence of saccular dysfunction and mild unilateral horizontal canal dysfunction measured by caloric stimuli in the implanted ear. The current study paradigm did not allow us to assess the impact of implantation on balance function as all children had unilateral implants at the time of testing. Whether mild, unilateral caloric reductions, particularly those associated with the implanted ear, are clinically significant is questionable. It is plausible that these changes may simply be a function of an alteration in the thermal stimulability of the horizontal canal after mastoidectomy with facial recess approach. Given that these small unilateral EDs in the implanted ear do not correlate with dynamic balance function, and it is our perspective that such differences should not preclude sequential bilateral implantation. However, long-term vestibular effects in children receiving bilateral CI should continue to be studied.

It has been suggested that the saccule is particularly susceptible to damage during CI.[14] Although unilateral absence of VEMP occurred on the implanted side 60% of the time, this was not significant. At baseline in individuals with SNHL VEMP is often absent attributing the absence of VEMP on the implanted side to the act of CI without having confirmed their presence preoperatively would be inappropriate. Therefore no conclusion with regards to the impact of CI on the saccule can be made from the current study at this time.

Correlation Between Vestibular End-organ Testing and Dynamic Balance

Of the tests of vestibular end-organ function used, performance on high frequency rotational testing displayed the strongest and most robust correlation with static and dynamic balance ability (BOT-2 score). Horizontal canal dysfunction based on caloric areflexia also correlated with balance ability; however, horizontal canal function based on rotational testing identified an additional population of children with isolated high frequency loss, which was deemed clinically significant based on their poor BOT-2 performance. This population is easily missed by caloric testing alone. Other advantages to rotational testing is that it is better tolerated than caloric testing, particularly given that confirmation of bilateral areflexia requires the use of ice water irrigation. In summary, our results suggest that caloric testing may identify horizontal canal dysfunction that is not clinically significant likely as a result of physiological compensation and may also miss clinically significant high frequency losses of function identified by rotational testing. For these reasons, we feel that the true incidence of vestibular dysfunction is best predicted by abnormalities of the VOR in response to high frequency rotation, which in this study occurred in 38% of our cohort.

Recommendations for Testing

We are only beginning to understand the functional significance of vestibular end-organ deficits in children with profound SNHL and the implication to and of CI in this context. As such it is premature to outline specific recommendations for testing in this population. Similarly our understanding of the implications of abnormal vestibular end-organ function on the safety of bilateral CI is incomplete. With this in mind we continue to advocate the use of a battery of vestibular end-organ and balance tests in any CI candidate in whom testing can be obtained, although we do not currently use this information to guide our decisions for implantation. Although we recognize that this is not feasible in all centers and in all individuals, a targeted approach according to etiology may be more appropriate. Likewise, using the BOT-2, and performing additional vestibular end-organ testing on only those children identified as having balance dysfunction may improve yield. Finally, the correlation between BOT-2 and rotational testing was high and tended to identify the same individuals, it was not absolute and we would recommend that at a minimum, a test of vestibular end-organ function and a test of static and dynamic balance be used in assessing any individual.

CONCLUSION

A significant proportion of children with profound SNHL requiring CI demonstrate abnormalities in static and dynamic balance. Etiology plays an important role and the characteristic vestibular fingerprint of many causes of SNHL, such as meningitis, has been well defined. However, subtleties likely exist in the balance and vestibular phenotype of other etiologies of deafness which remain to be examined. The identification of even mild abnormalities of vestibular end-organ and balance function is important both to understand the potential for vestibular dysfunction after unilateral or bilateral CI and to provide education regarding a child's balance abilities, leading to improved compensation and appropriate safety.

Although compliance with all tests was high, abnormalities of horizontal canal function in response to rotation, which assesses higher frequency motion (0.25–5 Hz) and thus more "real world" vestibular function, correlated best with balance. In addition, rotational testing may be most amenable to testing very young children and infants. For these reasons, rotational testing and the BOT-2 balance subset are at the top of our hierarchy of age-appropriate adjuncts to history and clinical examination in the evaluation of children with profound SNHL.

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