



Cervical Vestibular-Evoked Myogenic Potentials and Balance Testing in Children with Down Syndrome

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Abstract

Introduction Vestibular otolith function plays a major role in balance control.

Objective To investigate the saccular and balance functions of children with Down syndrome (DS).

Methods In total, 15 children with DS aged between 9 and 11 years were included. An age- and gender-matched control group (CG) composed of 15 normal participants was also included. The subjects with DS had trisomy 21, without hearing or organic problems, and they had independence in stance. The saccular function among the children with DS and among the controls was tested using air-conduction cervical vestibular-evoked myogenic potentials (cVEMPs). In addition, the static and dynamic balance statuses were evaluated using the following assessments; the Pediatric Balance Scale (PBS), the modified Clinical Test of Sensory Interaction on Balance (mCTSIB), the Romberg test, and the Timed Up and Go (TUG) test.

Results In the present study, the results of the saccular function test showed that there was a significant difference between children with and without DS ($p < 0.05$). The DS subjects had significantly earlier N1 latency and lower amplitude of the cVEMPs ($< 70 \mu\text{V}$) compared with the control subjects. The static-dynamic balance ability was statistically and significantly different in children with DS compared with the controls ($p < 0.05$).

Conclusion These results revealed that saccular function seems to be affected in DS subjects. The dysfunction in static and dynamic balance abilities of the children with DS may be attributed to vestibular dysfunction as well as low gross motor skills. This knowledge should be taken into account when assessing motor performance in those subjects. Additional larger studies testing other dimensions of the vestibular system in children with DS are needed.

Keywords

- ▶ down syndrome
- ▶ cervical vestibular-evoked myogenic potentials
- ▶ postural balance

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Introduction

Down syndrome (DS) is one of the most common chromosomal birth defects in children,¹ and it is the phenotypic consequence of trisomy 21. As consequence of nonhereditary intellectual disability, several problems arise, such as developmental delay, respiratory dysfunctions, vision problems, hearing problems, and balance dysfunction.^{2,3} Control of balance requires the integration of vestibular sensory information with somatosensory and visual information,⁴ and its dysfunction may be evaluated via standardized scales and tests. Some of those are the Pediatric Balance Scale (PBS),⁴ the Romberg test,⁵ the Modified Clinical Test of Sensory Interaction on Balance (mCTSIB),⁶ and the Timed Up and Go (TUG) test.⁷ These tests and their uses are presented in ►Table 1.

The vestibular system is responsible for maintaining the sense of head orientation and acceleration, both at rest and in motion.^{8,9} Vestibular inputs through vestibulo-spinal connections play a major role in trunk stabilization.¹⁰ The activation of otolith organs evokes the vestibulocollic reflex (VCR), which provides stabilization of the position of the head in space.¹¹ The cervical vestibular evoked myogenic potential (cVEMP) is a test used for neurotologic examination both in adults and in children. It verifies the integrity of vestibular otolith function through a muscular response evoked by a high-intensity acoustic stimulation which activates the saccular macula and records the transient inhibition of the sternocleidomastoid (SCM) muscle via surface electrodes.

While several studies have reported balance and gait dysfunction in DS,¹²⁻¹⁴ the impact of vestibular otolith function in balance control in DS has not been fully addressed. The principal aim of the present study was to assess vestibular otolith function using cervical vestibular-evoked myogenic potentials (cVEMPs) in children with DS and matched control children. In addition, the static and dynamic balance performances were also measured through conventional balance tests and correlated with cVEMP responses.

Methods

The present study was conducted in compliance with the Declaration of Helsinki, and it was approved by the local Ethics in Research Committee. In total, 15 children with DS, 7 females and 8 males, aged between 9 and 11 years (mean age:

10.25 ± 0.70 years) were included. An age- and gender-matched control group (CG) composed of 15 participants (7 females and 8 males) without DS was also included. The inclusion criteria for the subjects in the DS group were: the presence of trisomy 21; the absence of any visual or organic disorders, and independence in stance and ambulation (having no hearing loss, no additional impairment, and no neurologic or vestibular system problem). The participants in the control group were healthy, with no orthopedic or neurological disorders, no impairment in somatosensory activity, hearing, vestibular or uncorrected visual functions, and free of medications for at least three months before the beginning of the study. Written informed consent was obtained from all parents and children.

All children (DS and control) underwent the cVEMP, the PBS, the Romberg test, the mCTSIB and the TUG test. The CVEMPs were recorded with the Neurosoft Neuro-Audio.Net (Neurosoft Ltd., Ivanovo, Russia) software. The electromyographic activity of the SCM muscle was recorded while the children were sitting on a chair and were asked to turn their head to both sides to activate their neck flexors, and the saccular receptors were acoustically stimulated. Electromyography (EMG) biofeedback was used to reject all traces outside the minimum/maximum root mean square (RMS) EMG limits. Prior to the cVEMP test, the maximum contraction level is determined for each individual. Then, the target level for all subsequent cVEMP tests is set at ~ 70% to 80% of the maximum contraction level. The RMS is calculated for the EMG before each stimulus (prestimulus area) and displayed on a dial on a computer screen for the patient to see. The subjects are instructed to keep the EMG level constant and at a predefined target level (50 µV ± 20 µV). The acoustic stimulus was a click of 500 Hz at an intensity of 105 dB nHL. The rate was of 4 stimuli per second, and it was presented monoaurally through earphones. Recordings were obtained with an average of 200 stimuli response, and 2 traces from each side were obtained to assess reproducibility. In each trace, we determined the latency of positive-negative waves (P1 and N1), in addition to the amplitude of the P1-N1.

Functional balance in everyday activities is measured with PBS on a 5-point scale to obtain overall scores for each subject. In the Romberg test, children were asked to stand upwards for 30 seconds with their eyes closed, and then we noted whether they could sustain their position or not. In the mCTSIB, children were asked to preserve their balance in 4 different conditions

Table 1 Scales and tests used to evaluate balance dysfunction

Pediatric Balance Scale (PBS)	Functional balance in everyday life activities (Franjoine et al., ⁴ 2003).
Romberg test	The integrity among different sensory organs and neuronal conduction pathways utilized in the constitution of balance (Black et al., ⁵ 1982).
Modified Clinical Test of Sensory Interaction on Balance (mCTSIB)	Sensory system dysfunction on a stable and unstable floor, with eyes open and closed. In the tests on unstable ground with the eyes closed, the effects of visual and somatosensory inputs are eliminated, and the effects of vestibular inputs on postural stability are evaluated more efficiently (Shumway-Cook et al., ⁶ 1986).
Timed Up and Go Test (TUG)	Functional mobility and balance through different variables such as walking speed, postural control, functional mobility, and balance are evaluated (Podsiadlo et al., ⁷ 1991).

Table 3 Results of the balance tests, latencies of P1 and N1, amplitude of P1-N1 in the Down-syndrome and control subjects and their comparison

Grups			Pediatric Balance Scale	Timed Up and Go Test	P1	N1	P1-N1
Down syndrome group	N	Valid	30	30	30	30	30
		Missing	0	0	0	0	0
	Median		30.00	20.00	12.40	18.30	62.30
	Interquartile range		7.00	8.00	1.00	1.70	19.75
Control Grup	N	Valid	30	30	30	30	30
		Missing	0	0	0	0	0
	Median		56.00	9.00	12.70	20.00	104.75
	Interquartile range		0.00	1.00	1.10	2.30	59.05
Mann Withney U test			$p = 0.000$	$p = 0.000$	$p = 0.08$	$p = 0.000$	$p = 0.000$

in which sensory inputs were systematically altered. Balance times were recorded. All subjects in both groups were able to fully maintain their balance for 30 seconds. The Romberg test was negative for the whole sample, which means that all subjects could stand with their eyes closed for 30 seconds. The median PBS score (IQR) for the DS subjects was of 30.00 s (7.00 s), which was significantly lower than that of the control subjects (56.00 s [0.00 s]; $p = 000$). Task in the TUG test for DS 20.00 (8.00) which significantly longer than control subjects 9.00 (1.00) ($p = 000$).

Discussion

In the present study, the results of the saccular function test showed that there was a significant difference between children with and without DS. The DS subjects had significantly earlier N1 latency and lower amplitude in the cVEMPs ($< 70m V$) compared with the control subjects. In addition, there was a statistically significant difference in the static-dynamic balance ability in DS children compared with the controls.

Under everyday conditions, the vestibular system works in a complementary way with the somatosensory and visual systems to provide balance/postural control. Balance functions of DS has been put forth before,¹²⁻¹⁵ and the cVEMPs have been extensively studied in subjects without DS,¹⁶⁻¹⁹ as well as some other pathological conditions.²⁰ On the contrary, balance function and cVEMP testing have not been correlated in DS before.

The present study was performed to compare balance and saccular functions between DS children and typically developing children. In this regard, the static balance and the dynamic balance were tested and correlated with the saccular function test (cVEMP). As expected, the DS children had significant differences in all areas of balance ability in comparison with typically developing children ($p < 0.05$). Typically developing children showed higher balance ability than the DS children. This is in line with the following previous study results: the static-dynamic balance of DS

children showed more differences compared with typically developing children and DS children lack static balance ability.^{12,14,15} The reason for this is that their muscle reaction velocity is significantly slower compared with their peers,²¹ and there is much difficulty with motor control.

We used the cVEMP with air-conduction stimuli that measures saccular function. The cVEMPs were recorded while the children were sitting on a chair and were asked to turn their head to both sides to activate their neck flexors bilaterally. As mentioned in the literature,¹⁶ activation of the the SCM muscle by turning the head while sitting was sufficient to generate the cVEMP responses without early fatigue. Reproducible, unrectified cVEMP tracings were easily obtained with clicks at 105 dB in all DS and control children. In other words, the response rate in the present study was of 100%. Despite the usual muscle weakness and motor control handicap among DS children, the cVEMPs could be easily recorded in those subjects.

Our results demonstrated that the cVEMP parameters for the control group are similar to the normative values, as reported in the literature.¹⁸ On the other hand, we detected that the fundamental parameters of the cVEMP test (latency and amplitude) among DS children are significantly different than those obtained among the controls. Differences in latencies and amplitudes should be considered as N1 latencies were shorter and P1-N1 amplitudes were lower than those obtained among the controls. These results suggest that cVEMPs recording, demonstrating that the saccule, the vestibular afferent fibers and the vestibular nuclei are not similar to typically developing children. No statistically significant difference was detected between the right and left sides regarding the P1 and N1 latencies and the P1-N1 amplitude. This agrees with the studies by Picciotti et al.¹⁷ and Gonzalez et al.,²² who demonstrated no differences between the right and left ears.

The recording rate was of 100%. This means that the cVEMP test could be applied successfully to diagnose saccular function among the pediatric DS population, as it can be

recorded easily in all children to make appropriate decisions about interventions and program placement and planning, and to track the progress of children with DS.

Regarding the use of cVEMPs in the examination of the vestibular system in the pediatric population, Tribukait et al.²³ showed that the function of the semicircular canal correlated best with the function of the saccule, and, if the hearing was better than 90 dB, the function of the vestibular otolith was often normal, while for hearing levels of 100 dB to 120 dB, otolith function declined significantly. De Kegel et al.²⁴ found that children between 3 and 12 years of age with moderate hearing impairment and absent cVEMP perform significantly weaker on static balance measurements in which visual and/or somatosensory information is unreliable. This confirms that the saccule has an important role in the development of static balance.

Conclusion

In the present study, the most remarkable result is that cVEMP responses are effected in children with DS means that saccular function is altered in those children. This finding may have implications to the understanding of the basis of the balance problem that affects children with DS in their everyday lives. When assessing static and dynamic balance dysfunction among DS children, vestibular otolith functions should be considered. Future studies on the functions of the utricle and semicircular canal are needed for a more complete understanding of vestibular dysfunction among DS children.

Availability of Data and Material

The datasets used during the current study may be made available by the corresponding author upon reasonable request.

Conflict of Interests

The authors have no conflict of interests to declare.

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