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Vestibular system and neurodevelopmental disorders

PRODUCT INSIGHTS

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This document provides an overview of the critical role of the vestibular system in neurodevelopment and the evaluation of its functionality in children. The SYNAPSYS vHIT system, equipped with a remote camera, offers a unique approach to assessing vestibular function without the need for goggles or calibration, ensuring accurate and efficient testing. By analyzing head and eye movements, this system provides valuable insights into the development of motor, cognitive, and spatial skills, highlighting its importance in pediatric vestibular assessments.

The newborn must quickly adapt to the effect of "gravity," managing head position, sitting position, standing posture, and walking. For this reason, it is not surprising that the vestibular system (VS) is the first to develop. The labyrinth is morphologically complete at birth, the connections with the oculomotor nuclei are already present between the 12th and 24th week of gestation, and the vestibular branch of the VIII nerve is the first to complete myelination.

This results in extremely active vestibular activity from the very first months of life. For example, as early as 2 months, head rotation stimulates the labyrinth, which generates a slow counter-rotational movement of the eyeballs (doll's eye reflex) without the rapid phase, which, as is known, originates from the brainstem and develops later. This finding is confirmed by thermal stimulation, which generates the same slow counter-rotational phase. Between 3 and 6 months, the optokinetic response appears, indicating proper integration between visual stimulus and vestibular nuclei. At 4 months, the vestibulo-collic reflex can be generated, confirming the normal development of the saccule macula, the lower component of the vestibular branch, and the vestibular nuclei.

In line with its importance in the acquisition of motor skills, the vestibular apparatus is very active between 6 and 12 months of life. Between 10 and 14 years, cerebellar inhibitory activity renders the VS functionally similar to that of an adult.

Control of saccadic and pursuit eye movements also reaches full functionality very early, becoming fully active by the age of 2.
Among the primary developmental anomalies is the absence of the vestibulo-ocular reflex at 10 months. It is now well-known that the incomplete or partial development of the vestibular apparatus has clear repercussions on the development of neuro-motor skills. Possible warning signs include the lack of head control at 3 months, hypotonia (ragdoll appearance), and the backward head drop when the infant is lifted from a lying position between 4 and 7 months, failure to maintain a sitting position, inability to crawl or to hold a seated position even when supported between 9 and 12 months.
At later stages, the child may be unable to maintain balance with feet together or with eyes closed, may exhibit asymmetry in sitting position or upright posture, resulting in delayed autonomous walking, frequent falls, reliance on support, and inability to gauge step distances correctly. Table 1 shows the results of different items of the balance subset of the Bruininks-Oseretsky Test (BOT-2) in a normal children, while table 2 compares the control and children with sensorineural hearing loss without vestibular loss (group II), children with sensorineural hearing loss and unilateral vestibular loss (group III) and children with sensorineural hearing loss and bilateral vestibular loss (EL-Badry MM et al, 2023).

Test item	Maximum row score
Standing with feet apart with eye opened	10 s
Standing with feet apart with eye closed	10 s
Standing on one leg with eye opened	10 s
Standing on one leg with eye closed	10 s
Walking for 6 steps on a line on the floor	6 steps
Walking forward 6 steps on a line with heel to-toe gait	6 steps
Standing on a balance beam with heel to-toe	10 s
Standing on one leg on a balance beam with eye opened	10 s
Standing on one leg on a balance beam with eye closed	10 s

Table 1

Test item	Mean ± SD				*F* value	*P* value
	Group I	Group II	Group III	Group IV		
1) Standing with feet apart on a line-eyes open	4 ± 0	4 ± 0	4 ± 0	3.9 ± 0.3	5.918	0.001
2) Walking forward on a line	4 ± 0	4 ± 0	4 ± 0	3.5 ± 1.1	10.594	< 0.001
3) Standing on one leg on a line-eyes open	3.6 ± 0.7	3.4 ± 0.9	2.9 ± 0.9	2.3 ± 1.6	19.748	< 0.001
4) Standing with feet apart on a line-eyes closed	4 ± 0.2	4 ± 0.2	4 ± 0.2	3.8 ± 0.6	4.811	0.003
5) Walking forward heal to toe on a line	3.7 ± 0.8	4 ± 0.2	3.7 ± 0.7	2.3 ± 1.5	27.089	< 0.001
6) Standing on one leg on a line-eyes closed	2.9 ± 0.9	2.6 ± 1	1.8 ± 0.6	0.9 ± 0.8	39.147	< .001
7) Standing on one leg on a balance beam-eyes open	3.7 ± 0.7	3.5 ± 0.8	2.7 ± 0.8	2.2 ± 1.8	24.215	< 0.001
8) Standing heal to toe on a balance beam	3.9 ± 0.3	4 ± 0.2	3.4 ± 0.8	2.9 ± 1.5	19.240	< 0.001
9) Standing on one leg on a balance beam-eyes closed	3.3 ± 1.3	2.7 ± 1.4	1.4 ± 0.5	1 ± 1	40.273	< 0.001
Total point score	33.1 ± 3.1	32 ± 3.4	27.8 ± 2.5	22.7 ± 2.7	47.361	< 0.001
Scale score	17.3 ± 3.7	13.8 ± 4.6	7.9 ± 2.1	7.2 ± 3.1	89.909	< 0.001

Table 2

While the aforementioned alterations seem fairly intuitive, it is essential to recall that the vestibular apparatus is actually involved in much more complex activities, referred to as "high-level" functions. These include the perception of accelerations, visuospatial skills, attention, spatial and body representation, as well as a modulatory effect on pain, mood, emotions, and mental activity.

Thanks to acceleration perception and visuospatial skills, we can construct and maintain a highly precise mental representation of the world. Specifically, we can understand and organize two-dimensional and three-dimensional space, perceive distance and depth, mentally rotate images, and, above all, navigate space and build spatial memory.

Notably, spatial navigation is supported by vestibular activity, which develops this skill through direct connections to the hippocampus, mediated by the thalamus. In child development, it has been shown that spatial localization is rather rudimentary at 2 years old, a true navigation ability develops by age 7, and by age 11, this ability is similar to that of an adult, with a direct correspondence between chronological age and the increase in volume and myelination of the involved areas. Another extremely important skill is spatial memory, which enables one to plan a route, remember where an object is located, and recall where an event occurred. It is fascinating that, thanks to spatial memory, we can process concepts such as large-small, distant-close, above-below.

The vestibular apparatus is also involved in spatial and body representation, as demonstrated in patients with hemi-neglect or phantom limb pain.

In these patients, vestibular stimulation temporarily alters this distorted representation and re-establishes correct spatial coordinates by reactivating multisensory networks. This produces specific effects that can be seen as mechanisms through which the vestibular apparatus manages sensory signaling. By modulating other sensory modalities, vestibular inputs can thus aid in adapting flexibly to changing information in the body-world relationship and in anticipating future motor and perceptual events.

Regarding its effect on mental activity, even simply imagining rotating to the left facilitates the production of small numbers, and vice versa, or lifting facilitates addition. Similarly, thinking about the past is accompanied by a backward body movement, while thinking about the future is accompanied by a forward movement. Therefore, the vestibular apparatus provides information about the spatial relationship between the self and the world, and space is used to structure, represent, and understand abstract concepts such as numerical magnitudes or temporal events, which cannot be directly experienced through our senses. Thus, it is unsurprising that vestibulopathy results in altered body self-awareness and cognitive disturbances.

It is therefore extremely clear that altered vestibular development will have repercussions not only on motor function but also on learning that relies on proper visuospatial, cognitive, and memory capabilities. Hence, it is essential to assess vestibular function in at-risk children in order to initiate rehabilitative measures as quickly as possible to facilitate motor and cognitive skill acquisition. Table 3 shows hearing loss (HL) origin and vestibular function status of candidates for cochlear implantation (Wiener-Vacher SR et al, 2024).

HL origin	Sample size	No. (%)		
		NVF	PVF	CBVL
Genetic nonsyndromic	60	45 (75.0)	15 (25.0)	0
Connexin 26	41	31 (75.6)	10 (24.4)	0
Nonsyndromic, no connexin 26	19	14 (73.7)	5 (26.3)	0
Otoferlin	1	1 (100)	NA	NA
Mitochondrial disease	1	NA	1 (100)	NA
Consanguinity and familial HL (17)	17	13 (76.5)	4 (23.5)	0
Genetic syndromic	60	13 (21.6)	34 (56.6)	13 (21.6)
Usher[a]	22	1 (4.5)	12 (54.5)	9 (40.9)
Waardenburg[a]	20	8 (40.0)	10 (50.0)	2 (10.0)
Rare syndromic	18	5 (27.8)	11 (61.1)	2 (11.1)
Jervell and Lange-Nielsen (SQTL)	2	1 (50.0)	NA	1 (50.0)
Alport (glomerulonephritis)[a]	1	NA	1 (100)	NA
Bartter type IV (tubulopathy)	1	1 (100)	NA	NA
BOR[a]	2	1 (50.0)	1 (50.0)	NA
Down (trisomy 21)	2	1 (50.0)	NA	1 (50.0)
CHARGE	1	NA	1 (100)	NA
Di Georges (deletion 22q11)	1	NA	1 (100)	NA
Duplication 22 q 11.2	1	NA	1 (100)	NA
DOOR	1	NA	1 (100)	NA
Duane with Mondini	1	NA	1 (100)	NA
Hurler	1	NA	1 (100)	NA
Klinefelter (XXY)	1	NA	1 (100)	NA
Polymalformation	2	1 (50.0)	1 (50.0)	NA
Brown-Vialetto-Van Laere	1	NA	1 (100)	NA
Inner ear malformation	67	28 (41.7)	36 (53.7)	3 (4.5)
Nonsyndromic	56	28 (50.0)	28 (50.0)	0
Syndromic[a]	11	0	8 (72.7)	3 (27.2)
Infectious	74	23 (31.0)	43 (58.1)	8 (10.8)
CMV	52	16 (30.7)	30 (57.6)	6 (11.5)
Meningitis	17	7 (41.1)	9 (52.9)	1 (5.8)
Various infections	5	0	4 (80.0)	1 (20.0)
Mumps	1	0	1 (100)	0
Materno-fetal infection	2	0	1 (50.0)	1 (50.0)
Kabuki	1	0	1 (100)	0
Sarcoidosis	1	0	1 (100)	0
Toxic	4	1 (25.0)	3 (75.0)	0
Chemotherapy	2	0	2 (100)	0
Aminosid	1	1 (100)	0	0
Neonatal jaundice	1	0	1 (100)	0
Perinatal distress	12	3 (25.0)	7 (58.3)	2 (16.7)
Undetermined	326	216 (66.3)	99 (30.3)	11 (3.4)

Table 3

Abbreviations: BOR, Branchiootorenal; CBVL, complete bilateral vestibular loss; CHARGE, involving coloboma, heart defects, atresia choanae, retardation of growth and development, genitourinary disorders, and ear anomalies; CMV, cytomegalovirus; DOOR, deafness, onychodystrophy, osteodystrophy, and mental retardation; NA, not applicable; NVF, normal vestibular function; PVF, partially impaired vestibular function; SQTL, splicing quantitative trait locus.
[a]Note that within the 67 malformations of the inner ear identified (Mondini, DAV, Gusher), 56 were isolated without any clinical associated syndrome and 11 were part of an identified syndrome: 8 Waardenburg, 1 Alport, 1 BOR, and 1 Usher type 1 syndromes.

In this regard, we note that vestibular function can even be assessed in very young children through specialized equipment, such as the Inventis video head impulse test. The advantages of this method are numerous, including:

- Thanks to the remote camera, there is no need to wear any mask.
- The system does not require calibration.
- During the test, the child sits in a caregiver's lap while watching, for example, cartoons or images that capture their attention.
- Although it is necessary to acquire numerous trials and discard those in which the child did not focus on the "target," the examination usually lasts no more than 4-5 minutes.
- The ability to study all semicircular canals, both horizontal and vertical, depends solely on the compliance of the young patient.

In Figure 1, we see two cases of children with congenital cytomegalovirus infection. At the top left is the evaluation of the lateral semicircular canals of AG, 11 months old, whose dynamic VOR gain is normal. At the bottom right is the evaluation of the lateral semicircular canals of AG, 14 months old, with a cochlear implant on the right side. The deficit in dynamic VOR gain on the left is clearly evident.

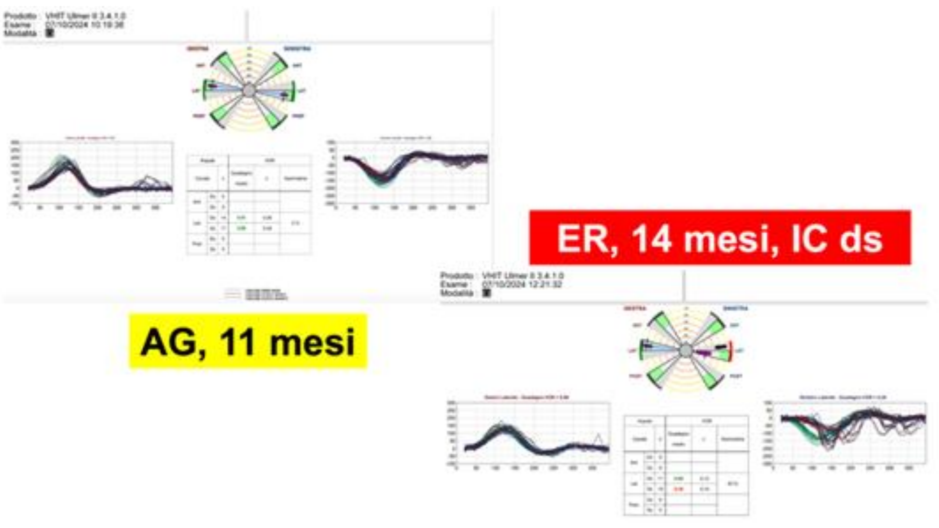

Figure 1: two cases of congenital cytomegalovirus infection

In this latter case, parents were advised to subject the young patient to vestibular stimulation, encouraging activities such as spinning around, swinging with both anterior-posterior and lateral accelerations, rolling on a mat, and engaging in as much physical activity as possible.

Finally, another significant aspect involves the possibility of promoting the acquisition of “high-level” skills by stimulating the child as previously mentioned, even in cases with a normal vestibular apparatus. In fact, as early as forty years ago, the possibility of improving conditions for children with delayed development of these skills was discussed through structured and personalized vestibular rehabilitation programs.

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