Vestibular Examination of Children With Alterations in Balance (II): Results by Pathologies

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Abstract
We review the findings of vestibular examinations in children according to the disease. Just as in adults, the dizziness can be classified following a physiopathological scheme. © 2010 Elsevier España, S.L. All rights reserved.

Introduction
This article reviews the results of vestibular examination in the various forms of the disease experienced by children. As in adults, vestibular disorders can be classified according to a physiopathological scheme.1

Otitis Media With Effusion

Children With Otitis Media With Effusion

According to the findings of the Peabody Developmental Motor Scale (PDMS), the most serious involvement occurs in children with bilateral otitis media with effusion (OME), whereas no significant differences are observed between normal controls and children aged 1–4 years old with unilateral involvement. Surprisingly, there is no correlation between the information given by parents about the balance
of children and the examination findings, as detailed below. Differences exist both in the overall score and in some of the test categories such as: (1) dynamic equilibrium in which the children’s centres of mass move continuously in various directions (standing on one foot, grasping a toy), and (2) locomotion tests in which children are asked to move in order to carry out an activity. Consequently, these are children with alterations in the acquisition of gross motor requiring dynamic balance. The involvement is possibly due to the fact that weight information for balance from the inner ear is erroneous, which makes the child dependent on visual and kinaesthetic information. The most interesting point is that receiving only some correct vestibular information from the healthy ear is sufficient to correct the deficit. This situation is reflected in the posturography results obtained when maintaining children in a colourful and stimulating visual environment: children with OME show more balancing and destabilisation than normal controls. However, given the age of the children, there are no data for a specific test that locates vestibular function (neither clinical tests, with cephalic impulse, nor instrumental ones, with caloric or rotary test) that can ascertain the pathophysiological basis of vestibular damage. In fact, studies carried out using electronystagmography (ENG) show no actual record of caloric nystagmus. Instead, this is obtained of only spontaneous and positional nystagmus (with little or scarce locator value), which shows highly pathological values in 58% of children with otitis media. The need for an accurate assessment of this type is greater if we consider that the only study carried out using rotational stimulation in children aged 4–9 years showed no significant differences in the findings of children with OME with respect to those in normal controls.

Children With Otitis Media With Effusion Before and After Placement of Eardrum Drainage Tubes

From the time of placement of the tubes, it is possible to observe that both spontaneous nystagmus and positional nystagmus disappear in 96% of children who showed this sign before surgery. The static posturography of children with bilateral OME shows a significant increase in the length and area of balance, which is increased by 20% when modifying the study conditions, both closing the eyes and testing on foam; the two alterations are not added together when the alterations are combined. When the otitis is resolved, the abnormal values of path followed and area generated by the centre of pressures become normalised.

Regarding the reliability of the information provided by parents, 29% mentioned that their children had symptoms of instability, dizziness and frequent falls and that, in general, they were more awkward than expected. However, 61% of children with OME showed a significantly worse score than the normal population in the efficacy test or Bruininks-Oseretsky test of motor proficiency (BOTMP), and this occurred both in children with unilateral involvement and with bilateral involvement, regardless of the degree of hearing loss. More specifically, the score was worse in the categories of balance, bilateral coordination, running speed and agility. After the placement of the tubes, 90% of parents mentioned a noticeable improvement in the stability of the child, and the BOT2 became normalised in 86% of children.

These results were worse than in the control group and, in the long term, this phenomenon became more evident in the vestibulo-oculomotor reflex (VOR), which is clearly hypometric in children who have suffered repeated OME. The ENG data did not provide information of interest in these children, as they were frequently normal or the findings had no locator value.

Benign Paroxysmal Vertigo of Childhood

This form of disease, listed within the "periodic syndromes of childhood", is the most frequently diagnosed, regardless of the age of the child. Benign paroxysmal vertigo of childhood (BPVC) is possibly a central syndrome, given the examination findings during the intercritical phase: (1) when confronted with an optokinetic (cortical) or visual-vestibular interaction (subcortical) visual preference paradigm, the children show an abnormal preponderance for the latter; (2) the dissociation in the response in vestibular evoked myogenic potentials (VEMP) depending on the location of registration and therefore on the stimulation of the inner ear and the neurological pathway followed, since the ocular are usually conserved and the cervical show a significant latency delay; (3) the caloric test is normal in 75% of diagnosed children; (4) both children with BPVC and children with migraine show a similar behavioural disorder, indistinguishable but mild (emotional difficulty, anxiety, depression, symptoms of hyperactivity), in the limit of normality and (5) 33% of children with migraine without vestibular symptoms present vestibular function abnormalities, which are therefore subclinical. Beyond the terminological confusion, the separation line between this form of vertigo and the idiopathic benign paroxysmal positional form (BPPV) in children is sometimes very tenuous.

Immune-mediated Inner Ear Disease

As in adults, immune-mediated inner ear disease (IMIED) is very rare in children. The differential diagnosis of localised forms is sudden and rapidly progressive hearing loss, and of systemic forms: Cogan’s disease, lupus erythematosus, antiphospholipid syndrome, antiphospholipid syndrome, Sjögren’s syndrome, Behçet’s disease, Wegener’s granulomatosis, idiopathic juvenile arthritis and Hashimoto’s thyroiditis. In the few cases of sudden hearing loss described, the vestibular function was normal according to the caloric test result but, as in adults, the association of vertigo to the episode of hearing loss adds a poor hearing prognosis. Fluctuations in uneven vestibular function have been described in Cogan’s disease: intense in the caloric and rotatory tests and hardly any in VEMP tests. In all these cases, the evolution of both hearing and balance should be monitored.

Congenital or Acquired Hearing Loss

Children With Congenital or Acquired Hearing Loss

The children were divided into several groups according to computerized dynamic posturography (CDP) results.
1) With normal vestibular function: Children who showed balance and motor development similar to that of children without hearing or vestibular deficit.

2) With vestibular deficits (most frequently bilateral hypofunction): Children who showed few problems in motor skills expect for balance and in whom balancing was significantly higher in conditions requiring vestibular support (Conditions 5 and 6 of the CDP).

3) With sensory organisation deficit and normal vestibular function: Children showing further deterioration in all measurements of motor skills including balance.¹⁹

Such evident alterations of balance in Conditions 5 and 6 of the CDP reflect, among other things, a change in the coordination of muscle response in the lower extremities, where it has been found that the response of the tibialis anterior muscle is slower and more prolonged than in normal children when they are subjected to a disturbance of the supporting surface. Children in group 2 would benefit from a vestibular rehabilitation plan that could work on several actions aimed at maintaining balance in demanding environments and surface conditions, while children in group 3 would require a global plan to tackle problems such as coordination and ability to solve conflicting sensory situations.²⁰ This plan should take into account the need to work on the visual acuity deficits suffered by children with vestibular hypofunction due to the profound alteration of the VOR that so intensely affects the ability to read, which is significantly hindered as a result.²¹

Children With Congenital or Acquired Severe-Profound Hearing Loss

Tests conducted prior to the placement of a cochlear implant: caloric test with ice water, rotatory test and VEMP. In general terms, 85% of children with severe-profound hearing loss present a certain degree of vestibular deficit. Patients were divided into 4 groups: (1) normal vestibular function (15%), in which the response was normal in all tests; (2) asymmetric caloric response (35%), in which the caloric response was different in both ears, with the value exceeding 30% but the test results of the rotary test and VEMP were normal; (3) bilateral vestibular hypofunction (25%), where the caloric response and the amplitude of the VEMP were less intense than normal, while the response to the rotatory stimulus was normal and (4) bilateral areflexia (25%), where the caloric response was zero, as in the VEMP and no nystagmic or postrotatory bursts were recorded.

In general, taking into account the limitations of the tests performed, it can be considered that in children with severe-profound hearing loss, the canalicular function is more vulnerable than the otolithic; canalicular function suffers a significant deterioration in a situation of severe hearing loss and the latter shows severe deterioration only occasionally in the case of profound hearing loss, while both are affected in a situation of cophosis.²²

Changes Resulting From Implantation in the Cochlea: Vestibular Function

Tests performed: clinical examination of VOR, bi-thermal caloric test, rotational and VEMP. In a study with a large number of children assessed systematically before surgery, the results of a caloric test allowed them to be grouped into 4 groups: (1) normal (50%); (2) unilateral hypofunction (22.5%); (3) bilateral hypofunction (7.5%); and (4) bilateral areflexia (20%). Unlike in other studies (with a smaller number of patients), there was no correlation between the degree of hearing loss and vestibular deficit. Placing an implant in the cochlea produced the following changes: (1) canalicular function deteriorated in 31% of children who had a normal previous response (most frequently generating hypoxectability, not areflexia); (2) otolithic function deteriorated in 55% of children with normal preoperative response; (3) 27% of children presented symptoms consistent with acute vestibulopathy and (4) changes could occur even months after the intervention. In general terms, 30% presented alterations in all tests after implantation and it is considered that the changes have already taken place definitively after 3 months.²³ It is in that period of time that a more complete and efficient vestibular compensation process can be expected.

The most abnormal result in the caloric test (vestibular areflexia) is the subject of another study in a similar group of children who underwent a complete videoystagmography (VNG); this revealed that 49% of children referred vestibular symptoms (without specifying their type or degree) in the immediate postoperative period, that those children who had a normal caloric response went from 63% before surgery to 39% after surgery, and that those with caloric areflexia went from 14% before surgery to 34% after surgery. This relevant change in the vestibular function is compensated in the vast majority of children without creating medium-term symptoms except in a few; so far, only an alteration in visual suppression of caloric nystagmus has been identified as a risk factor for poor prognosis for such an eventuality.²⁴

The abnormal postoperative response is also detected effectively with VEMP. This test is recommended for its simplicity and consistency of response. The percentage of children who presented alterations in the VEMP before implantation was high and correlated well with age of onset of hearing loss and motor development, in that the earlier hearing loss became established, the worse the result in VEMP and motor development tests. The VEMP revealed that, although there was a certain degree of postoperative impairment in 80% of children, this depended directly on the surgical inner ear approach, namely the performance of the cochleostomy and the implantation procedure. However, when studying a population of children implanted in a single ear, there were no differences in the various parameters of the VEMP obtained between the implanted ear and the non-implanted.²⁵

Changes Resulting From Implantation in the Cochlea, Vestibular Function and Motor Skills

Children with congenital or acquired severe-profound hearing loss users of a cochlear implant: Tests performed:
exploration of dynamic equilibrium (BOTMP, balance subtest) and rotatory test with an active implant. The average value of the BOT2 was 13 ± 5, which was significantly different from the value in the normal population; there was a trend towards improved results when the implant was active with respect to when it was turned off. There was also good correlation between the findings and developments in the BOT2 and the gain and phase values of the VOR at intermediate stimulation frequencies from 0.16 to 0.64 Hz. 26 It is interesting to observe that the children who used cochlear implants showed an increased visual dependence compared to those with normal hearing. In general terms, when analysing multiple variables (many of them related to each other), it is possible to find good correlation between degree of balance and aetiology (worse if the cause of deafness was meningitis or a cochlear-vestibular malformation), but not with age of implantation or time of implant use. 27

In these children, it is particularly necessary to implement a vestibular rehabilitation plan that aims to encourage substitution through exercises for visual-vestibular facilitation and proprioceptive function enhancement, similarly to those mentioned previously. In a study with implanted children with varying degrees of severe-profound hearing loss and bilateral vestibulopathy, it was possible to achieve better and more complete sensory organisation and coordination and, ultimately, better balance. 28 The improvement was objectified in the PDM5 score and postural analysis with Condition 3 of the CDP and the somatosensory and visual indexes, which showed improvement nearing the values of normal children. This study observed that the group of children with no rehabilitation evidenced a worsening of results between studies in the previously mentioned tests. Consequently, it is possible to state that it was also possible to stop the motor delays that these children suffered additionally. As with auditory rehabilitation, there seems to exist a period that should not be exceeded or else there will be a significant limitation on the recovery of balance and posture. Similarly, we must not forget that these are children with bilateral vestibulopathy and the results should thus be evaluated in the long term or a maintenance plan to retain the gain without reverting to motor impairment should even be implemented. It is possible that over time they could be engaged in more complex exercises interfering or distracting vision and proprioception. The compensation capability is documented and depends on the age of onset of damage; both in congenital and acquired cases, they will be able to obtain a normal Romberg, but only congenital cases will be able to stand on one foot with eyes open, perform a tandem walk, ride a bicycle or even swim, activities which the latter will be able to do with errors or difficulty. In contrast, it will be difficult for both (but especially for the latter) to stand on one foot with closed eyes or walk on a balance bar. 29

Alterations do not consist of only the deleterious effect of vestibular damage. Hearing loss in itself generates a series of changes in neuropsychological development of children, among which is a relevant delay in visual-spatial coordination and maturation (without reaching a pathological range), which disappears in the group of children implanted in a single ear. 30 A cascade of alterations takes place, which become difficult to restore after some time as motor development follows a predetermined sequence or program. In this sequence, auditory development takes place very early, already before birth and, among this cascade, vestibular function probably acts as a catalyst of deterioration.

Genetic Alterations

Given the high number of genetic alterations and the frequent association of hearing loss, we will review a small number of alterations that are worth clarifying because of their uniqueness or relevance. Hypoplasia of the semicircular canals is very frequent in the CHARGE complex. This leads to 52% of children presenting vestibular symptoms or a significant alteration in vestibular testing; surprisingly, it seems that only vestibular alteration is specific for the mutation CHD7. 30 In the case of mutations of the connexin gene associated with non-syndromic hearing loss, we can expect some degree of vestibular alteration since, in Cx30−/− mutant mice, there is degeneration evident only in saccule cells. Gap junctions are probably hybrid and require both Cx26 and Cx30; in contrast, in the case of isolated deletion of Cx30, only the saccular ones are lost, which can be retrieved experimentally by overexpressing Cx26. 31 This phenomenon may underlie the disorder found in children with the A1555G mutation in whom the frequency of vestibular symptoms is variable depending on the series, but who show a curious combination of complete VEMP involvement with normal caloric response in 60% of cases. 32 This finding is identical to that obtained in patients with mitochondrial pathology in whom the saccule endolymphatic space is collapsed (as described histologically), which in turn generates the fundamental vestibular damage. 33

Motion Sickness

Despite being classified as one of the forms of physiological dizziness, the impact of motion sickness is extremely high in the daily evolution of children and their families. It is the combination of 2 different mechanisms: (1) inconsistency of sensory information converging at a given time at the central level and (2) excessive release of acetylcholine in the vestibular nuclei. The above factors eventually lead to confusion or loss of spatial orientation. Children are influenced by several factors: (1) size, which makes them lose visual references; (2) incongruous physical activity, natural in childhood games, which facilitates overstimulation and (3) lack of habituation. The smaller the child, the less susceptibility to motion sickness there is, but, surprisingly, the only symptom may be a postural disorder and headache in the smallest children; as the vestibular system matures, the postural disorder decreases in severity but the vegetative symptoms increase. 34

Head Traumatism

Children with Minor Head Traumatism

The frequency of vertigo or instability in children who have suffered cerebral concussion is 51% and the percentage of children referring noise intolerance during the first days is 65%; in any case, these figures are not different from
those found in children who have suffered banal non-cranial trauma, which leads to doubts about the existence of post-concussion syndrome in children. In BOT2, the result is lower than expected at 1, 4 and 12 weeks after the traumatism, but a progressive improvement is documented. The situation is notably worse when children must perform exercises on the balance beam such as standing on one foot with eyes closed, walking in tandem and overcoming an obstacle. These are exercises that not only alter the sensory information, they also require a higher level of motor coordination. The level of demand lies in the anticipatory activity for balance, in other words, that which enables an individual to prepare and organise the motor activity necessary to overcome a new or complex situation. These are not mechanisms subject to feedback control, which would explain the difficulty or near impossibility to recover them and hence adapt. In children with mild head traumatism who do not improve at the beginning with conventional measures of resting and vigilance after the traumatism, vestibular rehabilitation has shown an improvement in symptoms (more intense in children than in adults) and in the results obtained in Conditions 1 and 2 of the CDP. We must not forget that BPPV in its various forms is also within the post-traumatic vestibular manifestations in children that require precise clinical assessment and adequate treatment.

Among the most unusual but potentially dangerous post-traumatic phenomena is dissection of the vertebral artery, which occurs occasionally after a forced cervical manoeuvre (for example, during sports practice). In these cases, ataxia and vertigo are severe enough to insist on the differential diagnosis even though the initial radiological studies do not include an examination of the neck and its vascular contents.

**Emergencies**

Despite being very uncommon, the desire for good management of children with vertigo in the emergency service seems to be one of the priorities in GP environments. It is possible to diagnose and treat systemic disorders that often begin with a vague and imprecise feeling of dizziness, as occurs in arterial hypertension and metabolic disorders or specifically otological emergencies, such as an exacerbation of OME in the form of acute otitis media leading to labyrinthitis. This is a true emergency, which requires an adequate medical and surgical approach. As for the former, one should consider that a bacterial cause of a “labyrinthisation” process of the middle ear is only detected in 65% of children, without there being a specific predisposition for a specific agent.

**Tumours of the Internal Auditory Canal and Posterior Fossa and Central Vestibular Syndrome**

The incidence of vestibular schwannoma (outside the context of neurofibromatosis) is very low and there are fewer than 50 cases described in the literature. The onset symptoms are otological in only 60% of cases and, given that there is a predominance of intracranial hypertension (headache and vomiting), cerebellar syndrome (ataxia and falls) and paralysis of several cranial nerves (predominantly of the facial), it is necessary to maintain a high degree of suspicion if these symptoms appear in the clinical evolution of the vestibular process of the child. In contrast, the most common symptoms of tumours of the posterior fossa and brainstem include vertigo and ataxia with headache (and less frequently tinnitus) and, almost as an oddity, hearing loss; in terms of exploration, ocular motility (tracking) is highly abnormal, as is the optokinetic nystagmus, and there is also nystagmus evoked by lateral gazing, looking vertically downwards or even opsoclonus, with normal caloric test results in which the visual suppression of caloric or per-rotatory nystagmus was normal.

In addition to tumours in the locations mentioned above, other causes of central vestibular syndrome are encephalitis and multiple sclerosis. In children with hydrocephalus treated by means of a shunt, irrespective of the aetiology, it is common to find signs of central location during vestibular exploration (82%). However, 46% also show signs of peripheral location. This high rate of mixed findings may well explain the high rate of motor development that they present.

Along with the alterations mentioned above, one cause of suspicion that should lead to a radiological study is cranial traumatism. In general, the presence of persistent headaches and/or neurological signs indicates the need for a radiological study; isolated vertigo does not seem to be relevant enough to provide information both in the MRI and in the CT.

**Temporal Bone Abnormalities**

Cochlear-vestibular nerve aplasia represents a diagnostic challenge that requires a complete functional study, not only auditory or vestibular but also radiological; when in doubt, the vestibular examination can be very useful, in particular the study of per-rotatory nystagmus and the precise levels of its gain and symmetry.

Dilatation of the vestibular duct affects the canal and endolymphatic sac. The prevailing symptoms are auditory but 50% of children (as in adults) present additional vestibular symptoms (episodic vertigo, motor retardation or instability), although this incidence increases in the case of children who carry the Pendred disease mutation. Vestibular disorders are usually found in the caloric exploration: unilateral or bilateral hypofunction. VEMP studies find a significant decrease in the response threshold, possibly due to a third-window phenomenon in the inner ear.

**Ménière’s Disease**

Ménière’s disease can be manifested in surprising manners, such as cyclical vomiting without other apparent symptoms, until after inquiry by the physician, perhaps due to how strange or peculiar it may be for a child to perceive a sound in the ear. It is common to find a high degree of canalicular paresis in children, despite the process being nascent and undeveloped, due to a phenomenon of fluctuation of vestibular function as intense as that of hearing function; this is a phenomenon that is less common in adults. The
glycerol test and the electrocochleography provide highly pathological data in children.\(^5\)

Delayed endolymphatic hydrops (DEH) is related to Ménière’s disease. It appears in adults who have suffered a severe-profound hearing loss in childhood; in general, in the case of children with severe unilateral hearing loss, the risk of suffering both ipsi- and contralateral DEH is 17% in the first 15 years and 30% for the rest of their lives.\(^5\)

### Somatoform Disorders

The incidence of somatoform disorders is almost 3% among children with vertigo. Risk factors that must be taken into consideration for an adequate approach include: frequent crises or "continuous vertigo" with associated headache, low class attendance and relationship problems with family or friends.\(^5\)

#### Conflict of Interests

The authors have no conflicts of interest to declare.

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