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Comprehensive Vestibular and Balance Testing in the Dizzy Pediatric Population

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Abstract

Objective. To describe the spectrum of balance disease in a large population of children presenting to a tertiary care vestibular and balance laboratory.

Study Design. Case series with chart review.

Setting. Tertiary care pediatric hospital.

Main Outcome Measures. Results of audiometric, vestibular, and balance tests and final diagnosis.

Subjects and Methods. Retrospective review of audiometric, vestibular, balance testing, and final diagnosis from a patient database.

Results. Between September 2003 and September 2007, 132 children were evaluated at the Alfred I. duPont Hospital for Children Vestibular Disorders Program. Sixty-nine of the patients were boys and 63 were girls. The average age was 9.7 ± 5.0 years (range, 1-17 years). Although not all were able to complete the entire test battery (99 children completed at least 50% of the tests in the protocol), a diagnosis was achieved in most cases. The most common diagnoses were peripheral vestibulopathy (29.5%), migraine/benign recurrent vertigo of childhood (24.2%), motor/developmental delay (10.6%), traumatic brain injury (9.8%), and central nervous system structural lesion (9.1%).

Conclusions. Peripheral vestibular deficits and migraine disease account for most of the pathology in the pediatric population. With a multidisciplinary approach, diagnosis of the source of vertigo and imbalance is possible in most children.

Keywords

balance, pediatric, vertigo, vestibular disorders

Common signs and symptoms of vestibular disturbances include vertigo, imbalance, vegetative symptoms such as nausea and vomiting, and frequent falls. In children, the spectrum of pathology is different than in adults, and therefore, the presentation may differ, such as delay in gross motor skills.

There are few published reports regarding vestibular and balance disorders in the pediatric age group. This may be because of a lack of recognition of this pathology. Because children infrequently complain of vertigo, they may be considered "clumsy," and signs and symptoms of a balance disorder may be ignored. They have also been noted to rapidly and readily accommodate for balance disorders.[1] In addition, there are intrinsic difficulties in obtaining complete clinical histories and objective vestibular and balance testing in this age group.

The goal of the current report was to examine the spectrum of disease in a large population of children presenting to a tertiary care vestibular and balance laboratory. These children

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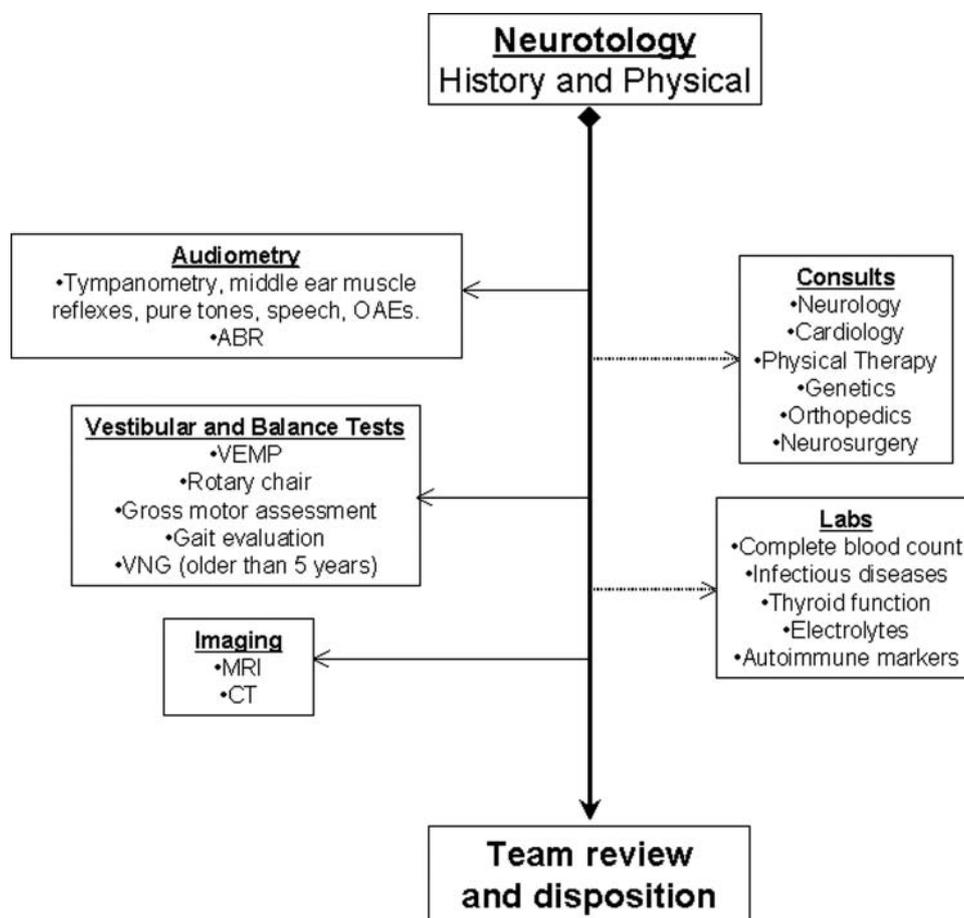
Figure 1. Clinical evaluation of children with balance disorders. All patients are evaluated by neurotology. Select patients receive labs and other specialist consultations (right dashed lines). All patients receive imaging, audiometric and vestibular, and balance testing (left solid lines). All data are then reviewed by a multidisciplinary team (black arrow), and a disposition is reached.

underwent comprehensive vestibular and balance testing and review by a multidisciplinary balance disorders team. This information may assist in more effective strategies to identify and treat children with balance disorders.

Methods

This study received Institutional Review Board approval from the Nemours Office of Human Subjects Protection, Wilmington, Delaware.

Chart Review

A retrospective database review, approved by the Nemours Office of Human Subjects Protection, was conducted on 132 children (age range, 1-17 years) evaluated consecutively from September 2003 to September 2007 at the Alfred I. duPont Hospital for Children Balance and Vestibular Disorders Program. The database included patient demographics, final diagnoses, and all audiometric, vestibular, and balance-testing data.

The general diagnostic algorithm followed is detailed in **Figure 1**. This represents a general framework by which children were evaluated because a balance between obtaining as much objective information to confirm or elucidate the diagnosis and individualization of the workup to accommodate the

particular circumstances of the patient was necessary (age, ability to cooperate, existence of comorbidities, etc). No analysis was made of the cost-benefit ratio of these tests or of their positive and negative predictive values because the small number of patients in each diagnostic subgroup precluded this. Briefly, each patient was first evaluated by a fellowship-trained neuro-otologist. Depending on each clinical scenario, following the history and physical examinations, selected patients were referred to other pediatric specialists from neurology, cardiology, physical therapy, orthopedics, genetics, and neurosurgery when necessary. Some patients also had blood testing for complete blood count, electrolytes, thyroid function, infectious diseases, and autoimmune markers if the clinical history indicated that their balance symptoms might result from abnormalities in these areas. All patients were referred for audiometry, vestibular, and balance testing and temporal bone and central nervous system (CNS) imaging either by computed tomography or magnetic resonance imaging, or both, if not already completed. Criteria used to select vestibular and balance tests were up to the discretion of the neuro-otologist. In general, acquisition of the maximum amount of objective data was attempted in each patient, with the exception of videonystagmography (VNG) testing in 42 children younger than 5 years.

Audiometric, Vestibular, and Balance Testing

The testing battery was modified according to the patient's age and ability to cooperate with and complete testing. Results were compared with published normative data (vestibular evoked myogenic potential [VEMP],[2,3] vestibulo-ocular reflex [VOR],[3,4] VNG[5-7]), norms provided by manufacturers, and our own norms (gait analysis). As noted above, VNG testing was not completed in children younger than 5 years. The complete test battery included the following audiometric, vestibular, and balance tests:

1. Tympanometry using a 226-Hz probe tone and middle ear muscle reflex testing
2. Age-appropriate pure tone and speech audiometry
3. Distortion product otoacoustic emissions (DPOAEs): The presence of DPOAEs was determined when the amplitude of the response was at least 3 dB above the noise floor.
4. When warranted by the results of the tests noted above, such as absent or elevated middle ear muscle reflex, an auditory brainstem response test was performed.
5. VEMPs: VEMP testing was completed using a 2-channel Intelligent Hearing Systems SMART EP evoked potential unit to obtain recording of the response of the sternocleidomastoid muscles to air- or bone-conducted auditory stimuli, or both.
6. Rotational chair-generated sinusoidal harmonic accelerations: The VOR gain, asymmetry, and phase angle were measured at 4 separate frequencies (0.01, 0.04, 0.16, and 0.64 Hz) at a maximum velocity of 50°.
7. VNG: The VNG battery of tests was performed using a goggle-mounted infrared video system to record eye movement. The oculomotor system was tested with random saccades, pendular smooth pursuit, and optokinetic nystagmus. Spontaneous, positional, and positioning nystagmus were recorded. Air-induced alternating binaural, bithermal, and calorics were performed with unilateral reduced responses (>25%) and directional preponderance (>30%) calculated via the Jongkees formula.
8. Gross motor development assessment: In patients younger than 4 years, the locomotion, stationary, and object manipulation subtests of the Peabody Developmental Motor Scales–second edition were administered. The balance, bilateral coordination, running speed and agility, and strength subtests of the Bruininks-Oseretsky Test of Motor Proficiency, second edition, were administered to children aged 4 years and older. A standardized z score ≤ -1.5 was indicative of impairment for both tests.
9. Modified Clinical Test for Sensory Interaction on Balance: The Balance Master (NeuroCom International, Clackamas, OR) was used to measure patient postural sway under varying conditions of sensory input.
10. Limits of Stability Test: The Balance Master (NeuroCom International, Clackamas, OR) was used to measure the maximum distance patients can intentionally displace their center of gravity without losing balance, stepping, or reaching for assistance.
11. Gait analysis: Computerized gait analysis was performed to measure the kinematics and kinetics of dynamic balance. Reflective markers were placed on the skin on standard landmarks (Cleveland Clinic Marker Set). These markers were traced by an 8-camera computer motion analysis system (Motion Analysis Corporation, Santa Rosa, CA) collecting at 60 Hz while the patient walked in a straight line at a self-selected speed for a distance of 9 m. The position and movement of the individual's center of mass were calculated. Dynamic balance was assessed via the analysis of deviations in joint angles between steps and from normal curves and with analysis of stability of the child's center of mass.

Data Review and Assignment of Diagnoses

A multidisciplinary panel assigned diagnoses after evaluation of the clinical history, physical examination, and results of audiometric, vestibular, and balance testing according to clinical standards. The criteria used to arrive at each diagnosis are not listed here but can be found in the literature.[8]

Results

Between September 2003 and September 2007, 132 children were evaluated at the Alfred I. duPont Hospital for Children Vestibular Disorders Program. Sixty-nine of the patients were boys and 63 were girls. The average age was 9.7 ± 5.0 years, with a range of 1 to 17 years. Because of our clinical algorithm as well as time constraints and age limitations, not all children completed the entire test battery; however, the medical diagnosis was reached in most cases. Ninety-nine children completed at least 50% of the tests in the protocol. Five patients underwent clinical evaluation but did not complete any objective testing (final diagnoses for this group: 1 malingering, 1 vestibular neuritis, 1 peripheral vestibulopathy after cochlear implantation, and 2 migraine).

The pathologic diagnoses are shown in **Table 1**. Approximately one-third of the patients had a peripheral vestibular disturbance, one-fourth had migraine-related vertigo, and the remainder exhibited a variety of pathologies.

The most common abnormality, peripheral vestibulopathy, was found in 29.5% of the children. These diagnoses are detailed in **Table 2**. Eight of 9 children with inner ear malformations had enlarged vestibular aqueducts (EVAs; unilateral or bilateral). These patients presented with sensorineural hearing loss (SNHL) of various degrees and either episodic or static vertigo and imbalance. Four children had idiopathic, nonfluctuating, stable SNHL associated with disequilibrium. Current or prior history of otitis media with effusion was thought to be the cause of the vestibulopathy in 3 children because their symptoms coincided with the presence of

Table 1. Distribution of Diagnoses in 132 Children Evaluated for Vestibular and Balance Disorders

Diagnosis	n	%
Peripheral vestibulopathy	39	29.5
Migraine/benign recurrent vertigo of childhood	32	24.2
Motor/developmental delay	14	10.6
Traumatic brain injury	13	9.8
Central nervous system structural lesion	12	9.1
Behavioral/psychogenic	8	6.1
Idiopathic/normal	4	3.0
Movement disorder/neurodegenerative	4	3.0
Encephalopathy	2	1.5
Vascular	2	1.5
Peripheral neuropathy	1	0.8
Oculomotor abnormality	1	0.8
Total	132	99.9

Table 2. Distribution of Individual Diagnosis in Children With Peripheral Vestibulopathy

Peripheral Vestibulopathy	n
Inner ear malformations (EVA and third window)	9
Sensorineural hearing loss (excluding EVA)	4
Benign paroxysmal peripheral vertigo	3
Ménière's disease	3
Otitis media	3
Progressive cochleovestibulopathy	2
Cytomegalovirus	2
Temporal bone fracture	2
Congenital vestibular hypofunction	2
Post–cochlear implantation	2
Vestibulopathy NOS	2
Vestibular neuritis	2
Autoimmune inner ear disease	2
Viral labyrinthitis	1
Total	39

Abbreviations: EVA, enlarged vestibular aqueduct; NOS, not otherwise specified.

effusion and resolved with the elimination of the effusion. Posttraumatic benign paroxysmal positional vertigo (BPPV) and probable Ménière's disease were diagnosed in 3 children. Vestibular neuritis was identified in 2 children, and congenital cytomegalovirus infection accounted for the vestibular deficit in 2 children. In the children with congenital cytomegalovirus, the peripheral vestibular deficit was accompanied by fluctuation or drop in their sensorineural hearing thresholds. Temporal bone fracture with traumatic brain injury was found in 2 children. Uncompensated vestibulopathy was found in 2 children who previously underwent unilateral cochlear implantation, and 2 other children displayed characteristics of congenital bilateral vestibular hypofunction. Two children had progressive, idiopathic cochleovestibulopathies, and 2 had confirmed autoimmune inner ear disease. Two children had recurrent vestibulopathies without hearing loss but could not be further

Table 3. Distribution of Individual Diagnoses in Children With Migraine Disease (24.2% of the Total Population)

Migraine/Benign Recurrent Vertigo of Childhood	n
Migraine equivalent	16
Migraine	7
Benign recurrent vertigo of childhood	6
Paroxysmal torticollis	3
Total	32

categorized. There was 1 case each of viral labyrinthitis and third window of the otic capsule.

Migraine with aura and benign recurrent vertigo of childhood (BRVC)/paroxysmal torticollis were frequently diagnosed (24.2%) in our study (**Table 3**). These diagnoses were assigned according to the criteria of the International Headache Society (IHS).[9] As expected, the mean age of the group with migraine with aura was older (11.7 years) than that of the children with BRVC/paroxysmal torticollis (3.5 years).

Motor and developmental disorders were found in 10.6% of patients. Included in this classification were mitochondrial myopathy, cerebral palsy, and pervasive developmental delay.

Traumatic brain injury independent of temporal bone fracture was found in 9.8% of patients. These patients had findings consistent with vestibular concussion, brainstem injury, or a mixed peripheral and central vestibular lesion.

Approximately 9% had CNS structural lesions including Arnold-Chiari malformation with or without syrinx, spinocerebellar atrophy, posterior fossa arachnoid cyst, and delayed myelination.

Behavioral disorders or psychogenic vertigo was found in 6.1%. These children were diagnosed with sensorimotor integration disorders, conversion disorders, anxiety, tic, autism, and attention-deficit hyperactivity disorder. Some children with normal testing and clinical examinations were suspected to complain of symptoms for secondary gain. Three percent of the patients were suspected of having hypoglycemic episodes and migraine with aura but did not meet standards for diagnosis and so were termed *idiopathic imbalance*. Another 3% were diagnosed with a movement disorder and neurodegenerative disease.

Disorders related to the vascular system were found in 1.5% of the children (orthostasis and cerebrovascular accident). Postviral encephalopathy was also found in 1.5%. Less than 1% of the children were found to have peripheral neuropathies or oculomotor abnormalities.

Discussion

The differential diagnosis of vertigo in childhood is large and includes a spectrum of disease that is not mirrored in the adult population.[10] Common diseases seen in adulthood, such as BPPV, viral infections of the inner ear, and Ménière's disease, are very uncommon in children in our experience. However, migraine-related vertigo is quite commonly seen in the pediatric dizzy clinic, although its manifestations can differ from those seen in adults.

In our series, most patients (29.5%) had a peripheral vestibular cause of their complaints. In this group, EVA accounted for all but 1 patient with vestibulopathy and inner ear malformation. Although EVA is the most commonly identified inner ear abnormality in children with SNHL, the predominance of this pathology in our study likely represents a bias toward testing these children because they are known to develop complaints of vertigo and to lose further hearing following minor head injury.[11] We have found that children with EVA tend to clinically present with balance instability at a younger age and endolyphatic hydrops in their early teens with fluctuating hearing loss associated with episodic vertigo. However, longitudinal studies are required to better define the vestibulopathy in this condition. The true incidence of peripheral vestibular deficits in children with EVA and other inner ear anomalies has not been rigorously studied and remains unknown.

Migraine-related vertigo and the migraine variant BRVC are a common cause of childhood vertigo.[1,12,13] In our study, 32 children (24%) were diagnosed with migraine-related vertigo or BRVC. Consistent with prior studies, the average age of the migraine group was older (11.7 years) than the BRVC group (3.5 years). There were no objective peripheral vestibular abnormalities in either group. However, subtle deficiencies in gross motor development were noted in the migraine and BRVC groups. This finding is of uncertain significance but may reflect an element of deconditioning in these groups or possibly an association with the migraine pathology.

Although grouped together, migraine-related vertigo and BRVC have separate diagnostic criteria. First described by Basser in 1964, BRVC has an estimated prevalence of 2.6%.[14] It is described as repeated, paroxysmal vertigo occurring in otherwise neurologically normal children. During these events, nystagmus, head tilt, and, occasionally, torticollis are observed, and associated symptoms can include pallor, perspiration, nausea, and vomiting.[14,15] To diagnose BRVC, other neurologic abnormalities such as seizures and CNS structural lesions must be ruled out. Although the etiology of BRVC remains unknown, a vascular disturbance of the posterior circulation with effects on the vestibular nuclei has been postulated.[16]

Migraine-related vertigo was diagnosed according to the revised IHS criteria.[17] The diagnosis in children allows for a longer duration and bilateral headaches. The pathogenesis of migraine and migraine with aura is felt to have a common mechanism and involve a spreading oligaemia and cortical depression.[18]

Benign recurrent vertigo of childhood may be a precursor to migraine and is often associated with a strong family history of migraine, although this was not found in our study.[17,19] In a study involving more than 1100 patients with migraine by Al-Twaijri and Shevell,[19] 68% to 100% of children with various migraine equivalents (BRVC, abdominal migraine/cyclical vomiting, acephalgic migraine, and acute confusional migraine) were found to have a positive family history of migraine. In a study by Drigo et al,[15] a family history of migraine was found in 53% of children. Lindskog et al[20] found that 21% of children with BRVC went on to develop migraine.

In contradistinction to the experience of Bower and Cotton,[13] we found very few patients with otitis media–related disequilibrium. In our series, 3 patients had symptoms of disequilibrium that coincided with the presence of effusions that resolved with elimination of the effusion. This difference in findings is most likely due to patient selection. The mechanism of disequilibrium in otitis media is not entirely clear but may be the result of inflammatory mediators crossing the round-window membrane or pressure changes within the middle ear.[21]

Although Erbeck et al[12] reported that 12% of their patients had BPPV, we found very few (2%) patients with this condition, and all were children with a history of traumatic brain injury. Certainly, the prevalence of BPPV in children is much lower than that found in adults.[22] This difference is thought to be the result of the aging labyrinth.[23]

Ménière's disease is another entity that is common in adults but not often found in children. Diagnosis is strictly clinical and based on the presence of intermittent vertigo, fluctuating low-frequency hearing loss, roaring tinnitus, and aural fullness.[24] In our study, 3 children (2.3%) were diagnosed with Ménière's disease according to the criteria described above. Choung et al[25] found that only 2.6% of their dizzy pediatric patients had Ménière's, whereas the incidence in the dizzy adult population has been reported to be up to 7%.[26]

Motor and developmental delays were determined to be the cause of imbalance in 10.6% of the children we studied. These conditions have been recognized to affect postural control and balance function.[27] Affected children included those with cerebral palsy, neuromuscular diseases such as mitochondrial myopathy, and pervasive developmental delay. The high incidence of these disorders in our study population is likely related to referral patterns to our clinic.

Traumatic brain injury was an important cause of imbalance in our study and manifested in 3 different forms: vestibular concussion (history of brain injury and objective documented peripheral vestibular deficits), brainstem concussion (history of brain injury, imbalance, and centrally mediated nystagmus), and mixed injury. These injuries can be independent of temporal bone fractures, and the balance dysfunction contributes a significant degree of morbidity in this patient population.[28] Rehabilitation of these children is challenging because of the associated behavioral and cognitive impairment and limited ability for central compensation for the peripheral vestibulopathy.[29]

Central nervous system lesions including Arnold-Chiari malformation accounted for disequilibrium and vertigo in 10% of our patients. Although the true incidence of imbalance and vestibular deficits in Arnold-Chiari malformation is not known, we found objective evidence of balance dysfunction in several of our patients with this condition.[30]

To our knowledge, this series is one of the first in the literature to include careful, multidisciplinary assessment of patients presenting with balance complaints including as thorough as possible assessment of objective vestibular and balance function. In our study, peripheral vestibulopathy and migraine/BRVC accounted for most of the pathology (54%).

These findings are similar to those reported in other, smaller series in which consistently more than half the pediatric patients were diagnosed with a peripheral vestibular disorder or migraine/BRVC (from 49.9% to 83.1%).[1,12,13,21,25,31] However, these studies show great variability in the percentage of patients in each diagnostic subgroup (peripheral vestibular, migraine, or other disease). These differences are probably due to the completeness of the retrospective review, the specialists assigning diagnoses, the number of patients evaluated, the completeness or variety of the workup, and the patient population that was studied (type of clinic from which the patients were accrued and country of origin of the study). All of the studies cited were retrospective reviews that relied on billing records or diagnostic codes to identify patients and so are liable to underreport the total number of patients with vestibular and balance problems evaluated. Although our study involved a retrospective review of the clinical and laboratory information, all patients seen through the balance program were enrolled in our database at the time of testing.

The size of the study population may also have an influence on the distribution of pathology. In the only other study with more than 100 patients, the distribution of patients in each diagnostic category was roughly equivalent to ours.[31] This may represent more accurately the type of pathology seen in children who present for evaluation of vestibular and balance complaints; however, the incidence of balance disorders in children remains unknown.

Our report is also one of the few in the literature in which multiple pediatric specialists were involved in the patient evaluation. The above-mentioned studies[1,12,13,21,25,31] were limited to specialists either from otorhinolaryngology or neurology. We believe that a multidisciplinary approach to the dizzy child allows for the diagnosis of more obscure pathologies. Evidence of this can be found in the lower percentage of patients in our review that had an idiopathic diagnosis (3%). With the exception of the series reported by Ravid et al,[31] the percentage of patients without a diagnosis in other series was greater than 5%. Indeed, Eviatar and Eviatar[32] assigned the diagnosis of vestibular seizure/idiopathic to a full 50% of the patients they evaluated.

The standard workup of the patients varied markedly in prior reviews. All of the studies cited included audiometrics and electronystagmography as part of the evaluation; however, the remainder of the evaluation protocol was inconsistent. In our current study, all patients were offered a standard battery of vestibular and balance tests, and all patients had some form of CNS imaging. Other tests (blood work, electroencephalography, etc) were tailored to the patient's history and physical examination. The small number of patients in each diagnostic subgroup precluded statistical analysis of the diagnostic utility and cost-effectiveness of each diagnostic test. Our group is currently in the process of acquiring enough study subjects to allow this type of analysis.

The distribution of diagnoses in these series may also result from differences in study populations. In several of the published series, the patients were seen in an otolaryngology balance clinic, and, in other reviews, the patients were culled from referrals to a neurology clinic.[1,12,13,21,25,31] Differences in referral patterns to these 2 specialties may influence the type of pathology seen, but the type of clinic in which the patients were evaluated did not appear to correlate with the preponderance of disease. It can be seen, however, that with the exception of the series from Eviatar and Eviatar[32] in which a high percentage of patients were assigned the diagnosis of "vertiginous seizures/idiopathic," those series originating from neurology clinics showed a higher percentage of migraine patients than patients with peripheral vestibulopathy.[1,30]

Conclusion

Peripheral vestibular deficits and migraine disease account for most pathology in the dizzy pediatric population that we studied. Motor and developmental disorders, traumatic brain injury, and CNS structural lesions appear also to be important in the differential diagnosis of balance and vestibular disorders in children. When evaluating children with dizziness, it is important to complete a comprehensive subjective and objective evaluation. Although this study has significant limitations, including retrospective design and a select group of patients referred for evaluation, the ability to diagnose the source of vertigo and imbalance is possible in the vast majority of children. However, larger, multi-institutional, epidemiological studies are necessary to better define this population.

Author Contributions

Robert C. O'Reilly, design, data analysis, manuscript preparation; **Jewell Greywoode**, data analysis and acquisition, manuscript preparation; **Thierry Morlet**, design, data analysis, manuscript preparation; **Freeman Miller**, design, data analysis, manuscript approval; **John Henley**, design, data analysis, manuscript approval; **Chris Church**, design, data analysis, manuscript approval; **Jeffrey Campbell**, design, data analysis, manuscript approval; **Jason Beaman**, design, data analysis, manuscript approval; **Anne Marie Cox**, design, data analysis, manuscript approval; **Emily Zwicky**, data acquisition, data analysis; **Charles Bean**, design, data analysis, manuscript approval; **Stephen Falchek**, design, data analysis, manuscript approval.

Disclosures

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