Routine vestibular function assessment in children with congenital CMV: are we ready?

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Routine Vestibular Function Assessment in Children With Congenital CMV: Are We Ready?

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Congenital Cytomegalovirus (cCMV) is a leading non-genetic cause of sensorineural hearing loss (SNHL) and is an important cause of neurodevelopmental delay.1,2 Vestibular hypofunction can result in gross motor delay, hypotonia, postural instability, and lack of spatial awareness. Vestibular physiotherapy and safety advice may be indicated. Studies have shown vestibular impairment to be common in children with cCMV, in both those with, and without, sensorineural hearing loss.3 Routine screening for, and appropriate management of, vestibular hypofunction in all children with hearing loss has been recommended in NICE accredited guidelines issued by the British Association of Audiovestibular Physicians.4 One author5 has recommended that all children with cCMV with and without SNHL are tested for vestibular dysfunction. However, how and when, to test vestibular function in children with cCMV does not form part of standard guidance for the management of congenital CMV in the UK.

The majority of referrals to vestibular clinics are made by audiovestibular physicians and pediatricians. We, therefore, invited audiovestibular physicians and pediatricians in audiology, to take part in a national survey, to record current practice in the UK.

MATERIALS & METHODS

An electronic survey requesting details of audiovestibular services and assessments in children with cCMV was designed and trialed by four audiovestibular physicians. It was subsequently revised with their feedback and sent to all 100 members of the British Association of Paediatricians in Audiology (BAPA) and all 25 members of the British Association of Audiovestibular Physicians (BAAP) in pediatric practice, via email for completion over three weeks in June 2019. No patient identifiable data was requested. Survey questions can be found in appendix 1.

RESULTS

Twenty-one clinicians responded. This is approximately half of those with relevant clinics, as 25/50 members of BAAP are trained in pediatrics and a minority of pediatricians in audiology have had vestibular training.

19/21 clinicians reported assessing the vestibular system in children with cCMV: 16/19 carried out clinical vestibular assessments in children with cCMV and SNHL and 11/19 also carried out clinical vestibular assessments in children with cCMV and normal hearing.

The most common time to carry out vestibular assessments was at diagnosis of hearing impairment (42.1 %) or if balance symptoms were reported (36.8%), however, clinicians reported assessing children at all ages. The clinical assessments most frequently used to assess general balance function are demonstrated in Fig 1 and included the Romberg test, gait assessment, hopping, tandem gait, and one leg stance. Farmer’s test and head thrust test, which specifically test vestibular function, were also frequently used (see Fig 1.). One clinician commented that he would avoid testing jumping in a severely disabled child with cCMV as it might not be safe.

Sixteen clinicians had access to quantitative vestibular function tests. Tests most commonly used were videonystagmography (VNG), (4/16), video head impulse test (vHIT) (3/16), and cervical vestibular evoked myogenic potentials (cVEMP) (3/16). 2/16 clinicians used ocular vestibular evoked myogenic potentials (oVEMP), rotation testing, bi thermal caloric tests, and mastoid vibration. Rates of clinician referral for quantitative vestibular testing varied between 0-100 percent of children with cCMV. Of the 13 clinicians who had referred patients for quantitative vestibular testing...
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10 (77%) had seen cases with confirmed vestibular dysfunction. One clinician reported diagnosing congenital CMV in a child presenting primarily with imbalance, rather than hearing loss.

When asked to comment on the management of children with CMV and vestibular dysfunction, nearly eighty percent of respondents said they would refer children with vestibular dysfunction to physiotherapy and 63% said they would give safety advice or advice regarding activities to promote vestibular balance/function. One respondent commented that they would make a referral for situational counseling, psychological assessment, and a full medical screen for detecting factors for decompensation. Another respondent mentioned “customized vestibular/sensory rehabilitation, and, where appropriate, psychologist input for stress management” as a management strategy for children with CMV and vestibular dysfunction.

Several barriers to vestibular function assessment were identified; children not being well enough, hearing being a priority, and lack of training, awareness, and resources. It is also acknowledged that children with congenital CMV with severe neurodevelopmental impairments may not be able to take part in or tolerate some of the assessments indicated to evaluate vestibular function.

WHilst pediatric vestibular medicine is included in Audi-vestibular Medicine higher specialist training, it remains a specialized and lesser-known field. The UK has an estimated 50 Consultant Audiovestibular Physicians, with approximately half working in a pediatric setting, whereas there is little formal vestibular training for pediatricians working in audiology. There are only a handful of dedicated pediatric vestibular clinics in the UK.

It is therefore important that all clinicians working in pediatric audiology have access to training and equipment to diagnose vestibular impairment in children with hearing loss. Some tests, such as VEMP, employ equipment that audiology departments already have, and some test equipment is relatively inexpensive, such as mastoid vibrators, video Frenzel goggles, and vHIT. These tests are comparatively well tolerated and could be used more widely to increase access to vestibular testing. However, the Farmer’s test (used up to age 6 weeks) and head impulse test (used for any child who can fixate on a target), are bedside tests of the vestibular function which require no additional equipment and are easy to learn. They could be used as a screening tool in children with cCMV with balance difficulties and/or delay in gross motor milestones to guide referral for vestibular testing

This study was limited by its small sample size. Conclusions therefore cannot be drawn about the prevalence of vestibular dysfunction in cCMV cases. It is possible that physicians working in the field and/or with relevant experience were more likely to respond which may mean that an even smaller proportion of children with congenital CMV are referred for vestibular testing than reported in this survey. cCMV itself, despite being the leading non-genetic cause of deafness, is under-diagnosed, and not all children diagnosed with vestibular impairment will have had tests for congenital CMV. The ability to retrospectively diagnose children with cCMV by retrieval of stored newborn dried blood spots is essential in being able to give a diagnosis to individuals presenting outside the newborn period with vestibular dysfunction and

**DISCUSSION**

Vestibular dysfunction in children with cCMV is common (3), and may even be more common than SNHL(6). However, whilst awareness of cCMV-related hearing loss has grown, there is a lack of awareness of its impact on vestibular function and a lack of dedicated pediatric vestibular clinics in the UK to refer children to.

In this study, 75 percent of responding clinicians regularly assessed children with SNHL and cCMV for vestibular dysfunction, and yet the timing and investigations used varied from clinician to clinician. Clinicians reported a number of barriers to vestibular testing including; lack of time, training, awareness, and resources. It is also acknowledged that children with congenital CMV with severe neurodevelopmental impairments may not be able to take part in or tolerate some of the assessments indicated to evaluate vestibular function.

**Table 1. Clinician perceived barriers to vestibular function assessment in children with cCMV**

<table>
<thead>
<tr>
<th>Barriers to vestibular function assessment</th>
<th>Number of comments</th>
</tr>
</thead>
<tbody>
<tr>
<td>Lack of time in the clinic</td>
<td>3</td>
</tr>
<tr>
<td>Child not well enough</td>
<td>2</td>
</tr>
<tr>
<td>Hearing is the priority</td>
<td>2</td>
</tr>
<tr>
<td>Not enough training</td>
<td>2</td>
</tr>
<tr>
<td>Lack of awareness amongst key professionals</td>
<td>2</td>
</tr>
<tr>
<td>Lack of pediatric test facilities</td>
<td>2</td>
</tr>
<tr>
<td>Decommissioned services</td>
<td>1</td>
</tr>
<tr>
<td>Lack of access to specialized therapies</td>
<td>1</td>
</tr>
</tbody>
</table>
investigations for cCMV are worthy of consideration in future guidelines on the investigation of vestibular disorders.

The survey is the first to capture current views and practices in pediatric vestibular medicine in the UK. There is a shortage of trained clinicians and dedicated clinics in this field. Given the recent evidence that vestibular dysfunction is one of the commonest sequelae of cCMV, the case for more clinicians with expertise in this area, who can diagnose vestibular dysfunction and advise on appropriate management is growing. Including simple clinical tests of vestibular function assessment in management guidelines for cCMV, should be considered, particularly when there is a delay in early gross motor milestones, as diagnosis informs therapy provision and safety advice. Moreover, consideration should be given to the inclusion of vestibular function as an outcome measure in future randomized controlled treatment trials.

This study demonstrates the variation in vestibular clinical practice and access to resources. This coupled with NICE approved guidelines (4) recommending that all children with SNHL should have vestibular assessments and Teissier (5) suggesting all children with cCMV should have vestibular assessments, reinforces the argument for greater awareness, training, and access to pediatric vestibular assessment nationally. 

References for this article can be found at http://bit.ly/HJcurrent.