Prevalence of Vestibular Dysfunction in Children with a Congenital CMV Infection or Connexin 26 Variance: A Systematic Review

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BACKGROUND

Vestibular dysfunction is well documented and studied in the general adult population, yet there is less research examining the prevalence in the pediatric population. Vestibular dysfunction can be caused by multiple factors including genetic causes and viral/bacterial infections. Two of the most common causes of hearing loss in the pediatric population are due to the viral infection, Cytomegalovirus (CMV), and variation in the gene that codes for Connexin 26 proteins, or GJB2 (Maes et al., 2017).

Through the use of a systematic review, we were interested in determining if there is a prevalence of vestibular dysfunction present in children with CMV and Connexin 26. We focused on studies using vestibular-evoked myogenic potential (VEMP) testing, which has been found to be an effective tool for measuring vestibular function in children (Inoue et al., 2013).

PURPOSE

The purpose of this systematic review was to determine the estimated prevalence of vestibular dysfunction in children with congenital hearing loss related to a CMV infection or Connexin 26 variance, using vestibular-evoked myogenic potential (VEMP) testing.

METHODS

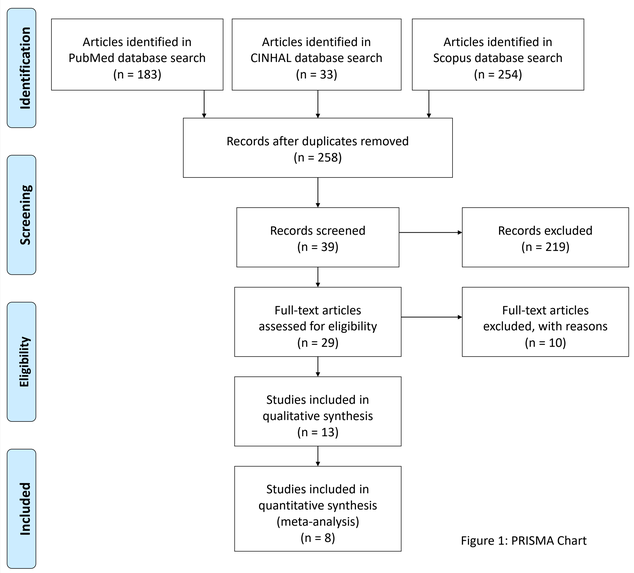

Figure 1: PRISMA Chart

Search Strategy: [(Vestibular) AND (CMV OR Cytomegalovirus OR GJB2 or Connexin 26)]

TABLE 1: FINAL REVIEW ARTICLES

	Subjects	Evaluation	GJB2 Variation	CMV Infection
Bernard et al. (2015)	52 CMV infection 5 m.o. - 11 y.o.	Caloric test Rotation chair vHIT cVEMP test Gross motor development	No subjects	74% Abnormal 87% Abnormal 57% Abnormal 78% Abnormal Significantly delayed head control, unaided sitting & walking
Inoue et al. (2013)	13 GJB2 variance (20 - 33 m.o.) 8 CMV infection (24 - 63 m.o.) Cochlear implant candidates	Dampened rotation test Caloric test VEMP test Gross motor development	100% Normal 100% Normal 17% Bilateral dysfunction 85% Normal head control; 100% Independent walking	20% Unilateral dysfunction; 20% Bilateral dysfunction 17% Unilateral dysfunction; 17% Bilateral dysfunction 17% Unilateral dysfunction; 50% Bilateral dysfunction 50% Delayed head control; 50% Delayed independent walking
Karltorp et al. (2014)	11 CMV infection (7 - 16 y.o.) Cochlear implant candidates	vHIT Caloric test VEMP test Gross motor development	No subjects	27% Unilateral dysfunction; 36% Bilateral dysfunction 50% Unilateral dysfunction; 40% Bilateral dysfunction 27% Unilateral dysfunction; 18% Bilateral dysfunction 70% Obvious balance disturbance
Kasai et al. (2010)	9 GJB2 variance (25 - 37 y.o.) Cochlear implant candidates	Caloric test VEMP test	18% Unilateral dysfunction; 9% Bilateral dysfunction 27% Unilateral dysfunction	No subjects
Maes et al. (2017)	40 Children (4.8 - 8.9 m.o.) Asympt cCMV Sympt cCMV_NH Sympt cCMV_HL Connexin 26	cVEMP test Gross motor development	100% normal 100% normal	Asympt cCMV = 100% normal; Sympt cCMV_NH = 14% abnormal; Sympt cCMV_HL = 57% abnormal Sympt cCMV_HL = significantly delayed motor development
Nassar (2014)	Case study (3 y.o. female) Vestibular calcification due to CMV infection	cVEMP test	No subjects	Asymmetric otolithic responses Abnormally small amplitudes on right side Normal amplitudes on left side
Tsukada et al. (2015)	24 GJB2 variance Mean age: 16 y.o.	Caloric test cVEMP test	9% Unilateral dysfunction 80% Abnormal	No subjects
Zagolski (2008)	26 CMV infection (3 m.o.)	Caloric test VEMP test	No subjects	31% Absent 33% Absent

RESULTS

VEMP testing between the two populations showed a general trend of more vestibular dysfunction present in the CMV group compared to the Connexin 26 variance group. This dysfunction includes both unilateral and bilateral dysfunction. Additionally, the degree of vestibular dysfunction for children with symptomatic CMV was more severe compared to those with non-symptomatic CMV. Studies that evaluated gross motor performance also showed children with CMV were more likely to have developmental delays compared to their peers with Connexin 26 variation.

DISCUSSION

The increased prevalence of vestibular dysfunction found in children with CMV has implications for overall gross motor function and performance, which can impact developmental milestones. This supports the need for vestibular testing in children diagnosed with CMV. However, the prevalence of vestibular dysfunction in these studies varied greatly. There is a continued need for further research investigating the prevalence of vestibular dysfunction in the pediatric population.

DISCLOSURES, ACKNOWLEDGEMENTS & REFERENCES

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