**ABSTRACT**

**Objective:** To determine if saccular dysfunction plays a probable role in infants with congenital torticollis

**Study Design:** Saccular function was evaluated in an 18-month-old infant with torticollis who was referred for evaluation of postural imbalance and observed rotary nystagmus. The infant demonstrated full passive range of motion but a left lateral head tilt was apparent. Lack of postural control when reaching down was also noted upon observation. The infant had undergone treatment of left torticollis for nearly 15 months. Pediatric ophthalmology examination was unremarkable and neck ultrasound to rule out muscle fibrosis was normal.

**Methods:** Informed consent was obtained. cVEMPs were recorded to assess saccular function. Caloric stimulation, positional and rotational testing were performed to evaluate other vestibular receptors.

**Results:** Abnormal cVEMP findings were observed with a low amplitude response on the left, indicating left sided saccular dysfunction. Caloric testing was symmetrical and rotational chair study revealed a normal pattern. Rotary-torsional nystagmus upon right positional testing suggested a presence of positional vertigo presenting secondary to abnormal saccular function.

**Conclusions:** We propose that saccular dysfunction plays a possible role in infants with torticollis and is an important consideration when timely resolution of congenital torticollis is not obtained with physical therapy. Early awareness of abnormal saccular function in this population is necessary in order to propose suitable explorations and ensure appropriate intervention. Larger studies will be necessary to validate these findings but to the best of our knowledge, this is the first documented report of saccular dysfunction identified in an infant with CPT.

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**INTRODUCTION**

Torticollis is a term used to describe a shortened or fibrotic sternocleidomastoid muscle (SCM) which can lead to tilting of the head towards the tightened side. Usually presenting at birth or soon after, congenital torticollis has an estimated incidence of 1 in 250 live births.\(^1\)

The etiology of torticollis can be due to congenital muscular or abnormal postural position, osseous, neurological, or ocular abnormalities. The evaluation of a child with torticollis should be comprehensive including a full physical examination, neurologic assessment, plain radiographs, and ophthalmologic exam.

Congenital muscle torticollis (CMT) is defined by unilateral fibrosis or shortening of the SCM, lateral flexion of the head and deviation of the chin to the contralateral side. With CMT there is loss of active and passive range of motion. There is a palpable mass in the SCM during the first 3 months of life. There are also compensatory changes in posture, flattened parietal-occipital area, and frontal flattening.\(^2,3\)

In contrast, congenital postural torticollis (CPT) has similar signs as CMT but no palpable mass in the SCM. CPT patients tend to have a decreased ability to actively rotate or laterally flex their head but have normal passive cervical range of motion. They tend to intermittently tilt their head rather than fixed as in CMT.

Neurological abnormalities can lead to torticollis. Neurological causes are differentiated due to presence of other symptoms such as headaches, nausea, vomiting, and positive neurological signs.\(^1,4\) Ocular abnormalities may also cause torticollis. There may be associated nystagmus with restrictive ocular movement but no cervical range of motion restrictions.\(^5\)

We present a unique case, the first report to our knowledge, of a child with CPT due to abnormal saccular function.

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**CASE REPORT**

An 18-month-old male infant was referred for evaluation of possible imbalance issues. There was a history of congenital postural torticollis. He demonstrated left lateral tilt and rotation of the head to the right side (figure 1), but was noted to have full range of motion and intermittent left lateral tilt. Pediatric ophthalmology and neurologic examinations were unremarkable, and neck ultrasound to rule out muscle fibrosis was normal.

At 2 months of age, the infant presented to physical therapy (PT) with a classic torticollis posture. PT treatment consisted of positioning, strengthening, stretching, and neuromuscular facilitation techniques. A home stretching and exercise program was also incorporated.

After 15 months of PT and home stretching exercises, the infant demonstrated some gain of lateral flex of the head but continued to exhibit imbalance concerns, primarily with postural stability. Difficulty maintaining the head in midline also persisted. Our physical examination revealed obvious postural instability, tightness of the left SCM, and a less prominent right SCM. The infant otherwise had normal development and age appropriate milestones. Audiologic evaluation was unremarkable.

Cervical vestibular evoked potential (cVEMP) testing was performed to evaluate saccular function. Abnormal asymmetric cVEMP amplitudes of the p13-n23 complex were recorded on the left (affected side) compared to the right (normal side). A 69% amplitude reduction on the left side (figure 2) indicates saccular dysfunction or abnormal saccule pathway. There was no pattern of hypersensitivity to sound reflected by cVEMP findings. Caloric testing was symmetrical and rotational chair sinusoidal harmonic acceleration (SHA) study revealed a normal pattern. Rotary-torsional nystagmus upon right positional testing suggested a presence of positional vertigo secondary to abnormal saccular function.

**DISCUSSION**

The role of the otothlic organs, the maculae of the utricle and saccule, is to sense linear accelerations and static orientation of the head by responding to dynamic changes in linear velocity. Disruption to the otothlic organs, or more specifically the sacculocollic pathways, can influence the development of postural control and put children at risk for impairment of sensory information critical to normal development of locomotion and motor coordination.\(^6,7\)

To our knowledge, an underlying saccular dysfunction has not been previously considered part of the differential diagnosis for CPT. Early diagnosis and determination of the cause of torticollis are imperative to recommend further workup and develop the most efficacious treatment plan.

Saccular function can be noninvasively assessed by use of cVEMPs. Although not commonly used in infants, cVEMPs offer an objective test to explore the vestibular system and the sacculocollic pathways in infants and very young children.\(^6,8\) The utilization of cVEMP testing can refine the differential diagnosis of CPT to avoid delayed motor development and postural impairment due to saccular deficits.

**CONCLUSIONS**

The findings of the present case suggest that saccular dysfunction plays a possible role in infants with postural torticollis and is an important consideration when timely resolution of congenital torticollis is not obtained with PT.

Early detection of abnormal saccular function in this population is necessary in order to propose suitable explorations and ensure appropriate intervention.

Undiagnosed saccular deficit(s) in patients with CPT could result in impaired motor coordination, reduced locomotion, and postural instability.

Larger studies may be undertaken to validate these findings but awareness of the presented case is important in order to optimize clinical decision and focus of treatment for young patients with unresolved CPT, especially when demonstrating imbalance or lack of postural control.

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**REFERENCES**