Article ▸ Why Does this Kid Keep Winking at Me?

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ABSTRACT

Background: One of the more common types of congenital synkinetic abnormalities is Marcus Gunn Jaw Winking Syndrome. It is typically a unilateral condition in which the affected superior eyelid moves with mouth movement, and the identified ptosis improves upon opening the mouth.

Case Report: A ten-week-old infant presented to The Eye Center at Southern College of Optometry due to the right superior lid moving simultaneously with mouth movement. The child had been seen at eight weeks of age for the same presentation at Le Bonheur Children's Hospital, where neurological testing was found to be normal. At TEC, she was diagnosed with Marcus Gunn Jaw Winking Syndrome. Possible sequelae of this condition include, but are not limited to, binocular vision and accommodative dysfunctions and potential amblyopia of the affected eye.

Conclusion: This case presentation will serve to help other eye care professionals to recognize and to understand Marcus Gunn Jaw Winking Syndrome in the pediatric patient.

Keywords: Marcus Gunn Jaw Winking Syndrome (MGJWS), oculomotor nerve (CN 3), synkinetic abnormality, trigeminal nerve (CN 5)

Case Presentation

AR, a ten-week-old Caucasian female, presented to The Eye Center at Southern College of Optometry with her mother in July 2013. The chief complaint given was that the child's right upper eyelid "jerked uncontrollably." This movement occurred more frequently when feeding, did not resolve when not feeding or using a pacifier, and had occurred since birth. It was also noted that the child slept with the left eye closed and the right eye partially open.

AR was born prematurely by four weeks, weighing five pounds, thirteen ounces. No supplemental oxygen was given. She had been diagnosed with gastroesophageal reflux disease (GERD) and had no known drug allergies. Medications reported included lansoprazole (Prevacid) and poly-vitamin drops with iron. There was a history of Haemophilus influenzae infection at four weeks old, which caused conjunctivitis in the right eye. This quickly resolved with erythromycin ointment. At eight weeks old, AR received a round of immunizations. Within hours of receiving said immunizations, the specific types of which were unknown to the mother when questioned, the child's body began twitching. She was taken to Le Bonheur Children's Hospital, where an electroencephalogram (EEG), sedated magnetic resonance imaging (MRI), and a computed tomography scan (CT) were performed. All tests were found to be normal; however, AR's parents stated that neurology verbally gave them an abnormal neurological report without a conclusive diagnosis.

Visual acuities were taken via preferential looking using the Lea Grating Paddles. AR achieved 8 cycles per centimeter (CPCM) at 38 cm OU, which is equivalent to approximately 20/80 to 20/100 Snellen at 38 cm. Both pupils were equal, round, and reactive to light with no relative afferent pupillary defect. Ocular motility testing showed a full range of motion OU as the child fixed and followed a moving target. Cover test at near revealed a combination of esophoric and exophoric postures, which varied throughout testing. No strabismus was noted via cover test or Hirschberg. A Bruckner test was performed, showing equally bright red reflexes OU. Intraocular pressure was assessed via digital palpation and was found to be soft and equal to touch OU. Gross observation throughout the exam noted that the right superior eyelid moved downward 100% of the time with mouth movement but did not obscure the pupillary axis at any point. Just Look retinoscopy, a form of near retinoscopy, was found to be +0.75 DS OD and +1.00 DS OS. The anterior segment health was unremarkable OU, as assessed with a 20 diopter lens and a transilluminator. Anterior chamber angles were open via the shadow test OU, and no central lens opacities were noted using the direct ophthalmoscope OU. AR was dilated with one drop of 0.5% tropicamide OD, OS. The dilated fundus exam was unremarkable, with a cup-to-disc ratio of approximately 0.25 round OD, OS. The macula was flat and intact, with a positive foveal light reflex OD, OS. Only fleeting views of the retina could be obtained in each eye.

AR was diagnosed with suspected Marcus Gunn Jaw Winking Syndrome OD and hyperopia OU. The mother was educated about the condition and was asked to watch the right upper eyelid when it moved from a superior position to a more inferior position. This was recommended to see whether any change of the most inferior position of the upper lid occurred. It was also suggested that the mother engage the child with eye contact whenever possible, and the importance of switching sides when feeding to maintain body and eye

symmetry was discussed. The child was to return to the clinic in three months.

AR returned in December 2013. The right upper lid was still "jerking" with mouth movement according to the mother, and there were no changes to the frequency or amplitude of the lid movement. No changes to medications and no known drug allergies were reported. It was recommended by AR's pediatrician that she start seeing an occupational therapist (OT) due to difficulty lifting her head during tummy time and overall low muscle tone. In November 2013, an episode occurred where AR clenched her left fist abnormally. The OT was concerned about possible seizure activity, and the patient was sent for another EEG. The results were found to be normal, but the parents were still given a verbal abnormal neurological report regarding their daughter. Neurology gave no conclusive diagnosis and could not give a reason for the right upper eyelid moving with mouth movement when asked. Marcus Gunn Jaw Winking Syndrome was not considered by neurology when probed by AR's parents. Chair skills were found to be unremarkable OD, OS. The Keystone Basic Binocular (KBB) test was performed to assess the presence or absence of stereopsis. AR attempted to touch the duck with her hand (above the page) and then pulled her hand back towards herself, indicating gross stereopsis. A reach and grasp test was also done, where a pen was placed on the child's midline. AR grabbed the pen with both hands and pulled it back towards herself with what would be subjectively considered normal strength for her age. Near point of convergence, performed with a transilluminator, showed gross ability to converge. It was noted on gross observation that the right superior lid did not seem to be "winking" 100% of the time, but possibly 80-90% of the time, a possible reduction in frequency from the initial assessment.

A diagnosis of congenital MGJWS OD was made, and the parents were educated on the importance of changing sides when feeding in order for the child to develop the ability to look both to the right and left equally, leading to better symmetry of the body and ocular alignment. There was no change to the magnitude of the ptotic state during jaw winking (still not reaching the superior pupil). Eye Stretches were recommended (moving a target in all positions of gaze so that the child looks in all positions of gaze), and the patient was to return to the clinic in four months.

Discussion

Marcus Gunn Jaw Winking Syndrome occurs either when the jaw is thrust forward or upon opening the mouth.[1] Ptosis of the affected eye in primary gaze will improve upon opening the mouth.[2] It was first reported in 1883 in a 15-year-old female who presented with what was considered to be a unilateral congenital ptosis in primary gaze.[2] The child demonstrated jaw movement, with an associated winking of the affected eyelid.[2] Marcus Gunn Jaw Winking Syndrome is considered to be a synkinetic abnormality. Synkinesis occurs with simultaneous movements or a coordinated sequence of muscle movements innervated by different branches of the same nerve or innervated by different nerves.[3]

There are several thoughts as to the cause of Marcus Gunn Jaw Winking Syndrome. The more popular theory maintains that there is an aberrant connection between the superior branch of cranial nerve three (the oculomotor nerve) and the motor branch of cranial nerve five (V3 of the trigeminal nerve).[3] The superior branch of cranial nerve three innervates the levator muscle of the upper eyelid, and the motor branch of cranial nerve five innervates the pterygoid muscle.[2] Electromyographic studies looking specifically at simultaneous contraction of the levator muscle and the pterygoid muscle have shown that a synkinetic innervation is present.[4]

Another thought as to the cause of Marcus Gunn Jaw Winking Syndrome is that there is an inability to inhibit "pre-existing phylogenetically more primitive mechanisms."[5] This is potentially why some people who have not been diagnosed with Jaw Winking Syndrome open their mouths while opening their eyes wide (e.g., instilling eye drops).[5] This cause has not been explored to the same extent as has the previously mentioned synkinetic abnormality theory.

Other synkinetic abnormalities have been discovered; however, only two of the more relevant conditions will be discussed here. The first of these conditions is Inverted Marcus Gunn Jaw Winking Syndrome. In this condition, the ptosis in primary gaze becomes worse with opening the mouth,[6] as opposed to Marcus Gunn Jaw Winking Syndrome, when the ptosis improves with opening the mouth. In the case of Inverted MGJWS, there is an aberrant connection between the superior branch of cranial nerve three innervating the levator muscle of the upper eyelid and the motor branch of cranial nerve five innervating the internal pterygoid muscle.[6] In MGJWS, the motor branch of cranial nerve five innervates the external pterygoid muscle.[6]

In a second condition, called Marin-Amat Syndrome, there exists an aberrant connection between the motor branch of cranial nerve five and the frontal and zygomatic branches of cranial nerve seven (facial nerve). Both eyes close when the mouth is open due to blepharospasm.[6]

Ptosis of the affected eye is seen in MGJWS in primary gaze; therefore, it is logical that a review of ptosis be presented to the reader. The normal position of the eyelid in primary gaze is one to two millimeters below the superior corneal limbus; thus, any measurement greater than two millimeters is considered ptosis.[7] Causes of ptosis include aponeurotic, mechanical, myogenic, and neurogenic. Aponeurotic causes include senile ptosis, which tends to occur in the elderly and is bilateral. This is due to a disinsertion of the levator aponeurosis.[7] In a mechanical cause of ptosis, the eyelid is too heavy for the levator muscle to lift. This occurs with scarring or in the presence of tumors that could increase the weight of the eyelid.[7] The most common myogenic cause of ptosis is Myasthenia Gravis. In this case, the muscles responsible

Table 1. Characteristics of MGJWS[2]

Eyelid ptosis that is usually unilateral (rarely bilateral)
Worse on down gaze
Occurs in 2-13% of congenital ptoses
Mostly sporadic but can be irregular autosomal dominant inheritance
No abnormal neurological findings on MRI, CT scan, or EEG
Ptosis may improve, remain stable, or become worse

Table 2. Associated Conditions of MGJWS[2,9]

Condition	Prevalence in MGJWS	Characteristics
Strabismus	50-60%	Vertical: hypertropia of uninvolved side, more common Horizontal: esotropia
Amblyopia	30-60%	Mostly strabismic or refractive amblyopia
Congenital nystagmus		
Congenital fibrosis of extraocular muscles		Autosomal dominant or autosomal recessive
Anisometropia	5-25%	
Superior rectus palsy	25%	Weakness of superior rectus muscles in both eyes
Double elevator palsy	25%	Possibly linked to a supranuclear lesion

for elevation of the upper eyelid become too weak to lift the eyelid.[7] Neurogenic causes of ptosis include Horner Syndrome, MGJWS, and Inverted MGJWS. In these cases, any damage to cranial nerve three or to the sympathetic nervous system can cause a ptosis. Recall that the superior branch of cranial nerve three innervates the levator muscle, and the sympathetic nervous system innervates Muller's muscle, which also assists in the elevation of the superior lid.[7]

There are several characteristics (Table 1) and conditions (Table 2) associated with MGJWS. Many motor actions are also recognized as causes of the elevation or retraction of the superior eyelid (Table 3).

The ptotic state in primary gaze in MGJWS is categorized based on the degree of ptosis. The condition is considered to be mild when the superior eyelid is positioned three to four millimeters below the superior corneal limbus in primary gaze.[7] Moderate MGJWS is characterized by the superior eyelid being positioned four to five millimeters below the superior corneal limbus, and severe is characterized as ptosis measured to be greater than or equal to five to six millimeters.[7]

Treatment options for patients diagnosed with MGJWS include home-based activities and vision therapy.[2,8] These options are ideal in the situation where the patient presents with associated conditions such as amblyopia, strabismus, etc. Another treatment option is surgery. If surgery is going to be performed, "there is a better outcome when the degree of ptosis is assessed accurately."[7] Table 4 provides the type of measurements, average measurements for patients not diagnosed with MGJWS, and how to obtain the specified measurements.[7]

Table 3. Causes of Elevation or Retraction of Superior Eyelid[2]

Moving the mandible from side to side
Chewing
Sucking
Contraction of the sternocleidomastoid muscle
Smiling
Breathing
Valsalva maneuver (e.g., coughing, sneezing, straining)
Sticking out the tongue

Table 4. Measurements Used for Surgery

Measurement	Average Measurement	How to Measure
Eyelid crease	7 to 8 mm in males 9 to 10 mm females	Distance from the eyelid margin to the eyelid crease while the patient is looking down at a 45 degree angle
Marginal reflex distance (MRD)	4 mm or greater	Distance from the inferior margin of the upper eyelid to the corneal reflex; important if lower eyelid position is not symmetric
Dominant eye	Levator muscle tone is influenced by ocular dominance when ptosis is present	Multiple methods to determine this
Palpebral fissure	8 to 10 mm	The distance between the bottom of the upper eyelid margin to the top of the bottom eyelid margin at the center
Levator function	Greater than 11 mm	Measure the movement of the upper eyelid from down gaze to up gaze; must stabilize the frontalis muscle when measuring

Table 5. Common Surgical Procedures in MGJWS

Condition(s) Present	Surgical Procedure
Amblyopia and vertical strabismus	Eyelid repair
Severe ptosis and levator muscle dysfunction	Frontalis muscle elevation and myectomy of the levator
Severe ptosis and normal levator function	Shorten eyelid levator muscle

Indications for surgery include, but are not limited to, the presence of amblyopia and vertical strabismus, severe ptosis and levator muscle dysfunction, and severe ptosis with normal levator function.[7] Table 5 shows the common surgical procedures performed when certain conditions are present.[7,8]

Thoughts on surgery for MGJWS are currently controversial.[7] Some surgeons believe that the child should not have surgery until the age of one to two years old because general anesthesia is safer at those ages, as compared to in infancy.[3] It is believed by some surgeons that a child less than the age of one with severe MGJWS, with or without associated conditions (amblyopia, etc.), should have corrective surgery.[7] It has been noted in the literature that some children may learn

to alter the position of their jaw, which reduces the degree of ptosis in primary gaze; thus, surgery may not be necessary.[9]

Conclusion

MGJWS is a benign condition that is easily recognized and diagnosed accurately. Electrodiagnostic testing and possible surgical procedures can be minimized if this condition is understood and diagnosed appropriately. This can lead to a timely diagnosis and a decrease in health care costs, as well as a decrease in time spent by the eye care professional and the patient. Knowing the associated conditions, such as amblyopia and strabismus, can help the optometrist to develop a successful plan for vision therapy and/or home-based therapy. This can, therefore, serve to help develop the best and most efficient visual system for the patient.

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