Bilateral Marcus Gunn Jaw-Winking in a Child with Developmental Delay

Abstract

Background: Marcus Gunn jaw-winking phenomenon (MGP) is a rare condition that results from synkinesia between the external pterygoid muscle and the levator. MGP is usually unilateral and associated with unilateral ptosis. During chewing, sucking, or movement of the jaw to the side the ptotic eyelid elevates. MGP is often associated with amblyopia, anisometropia, and strabismus. MGP rarely occurs in a bilateral form.

Case Report: A 5-year-old boy had a history of unusual eyelid movements since birth, while sucking on a bottle or chewing. He also had general developmental delay and severe speech delay. He presented with 1-2 mm left ptosis, but no strabismus, amblyopia, or limitation of ocular motility. Retinoscopy yielded anisometropic hyperopia and astigmatism. Ocular health was unremarkable. Observation of the patient while chewing showed that as his jaw opened the left eyelid elevated while the right eyelid fell. Upon mouth closure the left eyelid fell while the right eyelid elevated.

Conclusion: This rare case of bilateral MGP showed findings consistent with a possible misdirection of a branch of CN V to both levator muscles. There appears to be a reciprocal excitation of one levator and simultaneous inhibition of the other when the external pterygoid muscles are activated during chewing. To our knowledge, MGP has not been previously reported to occur commonly in children with developmental delay, as it does here. We discussed the possibility of eyelid surgery with our patient’s mother but did not recommend it at this time due to the slight ptosis and the equal acuities.

Key Words
Marcus Gunn jaw-winking, eyelid, ptosis, developmental delay

Background

Marcus Gunn jaw-winking phenomenon (MGP), first described by Marcus Gunn in 1883, is a rare condition associated with ptosis in 90% of all cases. When the external pterygoid is activated during chewing, sucking, or movement of the jaw to the side the levator is also stimulated and the ptotic eyelid elevates, producing the wink. These movements are the result of a synkinesia, or coordinated sequence of movements of muscles supplied by different nerves, between the external pterygoid and the levator. Inverse MGP has been reported to occur when the eyelid falls or closes as the mouth opens. Bilateral MGP is quite rare. A study by Beyer-Machule et al. reported that of 1500 cases of congenital ptosis, 80 were due to the jaw-winking phenomenon but only three of the 80 cases were bilateral. Doucet and Crawford reported on 55 patients with MGP, only two of them having the bilateral condition. MGP can be either congenital or acquired secondary to aberrant regeneration of cranial nerve (CN) III after nerve palsy. Congenital forms may be due to trauma or other insult in utero. MGP is often associated with other ocular anomalies. Beyer-Machule and colleagues found amblyopia in 54%, anisometropia in 26%, and strabismus in 56% of patients with MGP. Doucet and Crawford found amblyopia in 35% and strabismus in 36%.

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Case Report

A 5-year-old boy presented for his first eye examination with a history of severe speech delay (inability to form sentences, and his spoken words were difficult to understand). His mother reported that he was experiencing general developmental delay, with a mental age of 16 months. The child had suffered a series of anoxic events at age 7 months, reportedly having stopped breathing and turned purple for several minutes on two or three occasions. The cause of these episodes was never determined. He was the product of a full-term pregnancy, weighing 7 pounds, 8 ounces (3.4 kg) at birth. His mother denied use of tobacco, alcohol or other drugs during the pregnancy. There was a positive family history of learning disability in two older siblings, and strabismus in the child’s paternal grandmother, father, and one brother. There was no family history of glaucoma.

At the time of his eye examination, the patient was healthy and was taking no medications. His mother noted no signs of reduced vision, but remarked that his eyes did “funny things.” Since birth, unusual eyelid movements had been observed while he was sucking from a bottle (prior to the anoxic events at age 7 months). It could not be determined if the anoxic events had aggravated the condition. The condition was not present in any other family members.

Entering visual acuities were 20/25 (6/7.5) with each eye at distance, using Allen symbols, and 20/20 (6/6) OU at 40 cm, using Lea symbols (the patient resisting occlusion of either eye). Unaided cover test revealed orthophoria at distance and near. The patient appreciated random dot stereopsis on the Lang I Stereotest. Color vision appeared to be normal, but the patient had difficulty communicating his responses. Extraocular muscle motilities were full, and pupils were equal in size and reacted briskly to light, with no afferent defect. A left eyelid ptosis of 1-2 mm was evident (Figure 1) when the patient’s jaw was closed. Observation of the patient while chewing showed that as his jaw opened the left eyelid elevated while the right eyelid fell (Figure 2). Upon mouth closure the left eyelid fell while the right eyelid elevated (Figure 3). These peculiar reciprocal eyelid movements occurred continuously during chewing, and were quite obvious to all observers.

Both dry and cycloplegic retinoscopy revealed OD +0.50 sph., OS +1.00-0.50 x 180, although accuracy was questionable due to the child’s poor cooperation. Tonometry was unobtainable. Biomicroscopy revealed healthy, normal anterior segments OU, and direct and binocular indirect ophthalmoscopy demonstrated clear media, cup/disk ratios of 0.5, healthy optic nerve heads with clear margins, clear maculas, and positive foveal reflexes OU.

The patient was diagnosed with bilateral Marcus Gunn jaw-winking phenomenon (MGP). No spectacles were prescribed at that time, because the refractive error was minimal and acuities were normal. We discussed eyelid surgery with the patient’s mother but did not recommend it at that time due to the slight ptosis and equal visual acuities. We did not believe it was likely that he would develop amblyopia secondary to this degree of ptosis, but suggested that surgery might be a future option for cosmetic reasons. We advised further evaluation by a pediatric neurologist due to his developmental delay. Follow-up in our clinic was planned in 6 months to monitor the eyelid condition, unless any changes were observed by the mother sooner.

The patient came for follow-up 10 months later, still demonstrating 20/25 (6/7.5) distance acuity (Allen symbols) with each eye at far, and 20/40 (6/12) acuity at 40 cm (Lea chart, refusing occlusion of either eye for near). Dry retinoscopy revealed OD +1.00-0.50 x 180, OS +2.00-1.00 x 180, with better reliability compared to the first visit. A trial frame containing OD +0.75-0.50 x 180, OS +1.50-0.50 x 180 was tried on the patient, but he was inattentive for further acuity testing. Monocular estimate method (MEM) retinoscopy through the trial frame showed an approximately equal, slight lag of accommodation in both eyes, without appreciable residual astigmatism, as compared to unaided MEM.

Further evaluation of the MGP showed the same appearance of the eyelids habitually and while chewing.
as at the previous visit. Observations of the eyelids with the jaw moved to the right and left revealed the habitual mild left ptosis with the jaw to the left (Figure 4), and right ptosis with the jaw to the right (Figure 5). While sucking through a straw, the eyelids remained in their habitual position (mild left ptosis, Figure 6). No eyelid movements were elicited upon lateral gaze to either side.

We reassured the patient’s mother that the unusual eyelid movements were not interfering with his vision, and were not likely to limit his abilities in school (according to his mother, he had been determined to be learning disabled and was to receive appropriate school services). He was not cooperative enough to allow us to evaluate his visual information processing skills at this time. However, due to the magnitude of anisometric hyperopia with astigmatism found at this visit, and the resultant risk of developing anisometric amblyopia, we prescribed OD +0.75-0.50 x 180, OS +1.50-0.50 x 180 for school and near work. We felt that the lens prescription would aid focusing and possibly increase his interest in performing school work. A follow-up phone call 2 months later confirmed that the patient had obtained his glasses and was performing better using them for near tasks. He was subsequently lost to follow-up, despite numerous attempts to contact the family.

Discussion

This case was assessed as bilateral Marcus Gunn jaw-winking phenomenon with standard presentation on the left side and inverse presentation on the right. The superior division of CN III supplies the levator. CN V innervates the external pterygoid while CN VII innervates the orbicularis oculi. It is well accepted that true Marcus Gunn jaw-winking phenomenon is caused by a trigemino-oculomotor synkinesis. The affected eyelid is ptotic but appears normal or slightly retracted when the jaw muscles are activated during chewing, movement of the mandible to the opposite side, forward projection of the mandible, or wide opening of the mouth. The above actions require either ipsilateral or bilateral contraction of the external pterygoid. It was initially believed that a peripheral branch of the CN V is misdirected to the levator but several investigators have challenged this theory. Lyness et al. histologically analyzed the levator muscles in patients with MGP and concluded that the underlying process is a neurogenic atrophy with aberrant regeneration. They found that both levators were affected, with the clinically normal side being affected to a lesser degree. They noted that this indicates the initiating pathologic process is within the brainstem and theorized that the initial lesion occurs in utero. Their data also offer an explanation for bilateral MGP. Lepore and Glaser also suggest a central process rather than a peripheral nerve misdirection for acquired oculomotor nerve synkinesis.

There is controversy concerning the mechanism of inverse MGP. In 1948 Wartenberg reported that the appearance of inverse MGP is actually an intrafacial associated movement, which occurs after CN VII (facial nerve) paralysis. CN V was assumed to be uninvolved because the closure of the eye appeared to be associated with movement of the lower facial muscles and not of the mandible. In 1978 Lubkin used electromyography to analyze one case of inverse MGP. She found that the closure of the eye was not mediated by CN VII stimulation of the orbicularis oculi but by inhibition of the levator, suggesting a trigemino-oculomotor synkinesis. In contrast, a 2003 report by Oh et al. also used electromyography to study one patient with inverse MGP. They found consistent and reproducible co-contraction of the ipsilateral orbicularis oculi muscle and the lateral pterygoid muscle suggesting a synkinesis of the trigeminal and facial nerves. Thus, the exact mechanism of inverse Marcus Gunn phenomenon remains poorly understood. The rarity of inverse MGP makes it impossible to gather enough data to draw definite conclusions. It is certainly possible that either a pterygoid-levator synkinesis or a trigemino-facial synkinesis could cause this condition. Unfortunately, electromyographic studies were not possible in the present case. It was our observation that when our patient...
opens his mouth the right upper eyelid droops but is not actively closing. It therefore appears as if CN VII is not involved. Our observations instead suggest a reciprocal excitation of one levator and simultaneous inhibition of the other when the external pterygoid muscles are activated during chewing.

To our knowledge, MGP has not previously been reported to occur in children with developmental delay, as it does here. With the information we could obtain, it is impossible to know whether a prenatal insult led to both the MGP and the developmental delay, or whether the delay resulted from the anoxic events at age 7 months (after the MGP had already manifested). Although our patient did not have strabismus or amblyopia, commonly associated with MGP, he did show anisometropia. Strabismus in MGP is usually caused by a superior rectus muscle palsy. In the study by Doucet and Crawford 36% of MGP patients had an associated strabismus with 24% of these due to superior rectus palsy. Amblyopia occurs secondary to strabismus or significant ptosis in MGP. It is not surprising that our patient did not have amblyopia due to the absence of strabismus and presence of only a mild ptosis. It is difficult to draw any firm conclusion about the etiology of our patient’s refractive condition, because his habitual ptosis was insufficient to interfere with normal development of the eye. Additionally, anisometropia of 1 diopter or more has been shown to be present in 2%11 to 3.1%12 of children this age, without any associated MGP.

Surgical options exist for patients with MGP. Before surgery it must be decided if the ptosis or the wink is the greater cosmetic issue. If it is decided to improve the appearance of the ptosis, shortening the levator aponeurosis can elevate the eyelid. The patient and parents must be aware that shortening the levator, although improving the appearance of the ptosis, increases the movement of the winking phenomenon. Doucet and Crawford4 found that many patients who perceived the ptosis as the more severe problem before the surgery became concerned about the jaw-winking after the ptosis was reduced or eliminated. The jaw-winking is most often considered the major cosmetic problem. The most acceptable method for correcting the winking involves severing the levator tendon to destroy its action and then performing a fascia lata sling. The presence of significant jaw-winking necessitates disinsertion of the levator with subsequent suspension of the upper eyelid.3,13,14 Doucet and Crawford4 and Wong et al. 14 found favorable results using bilateral frontalis suspension surgery with levator disinsertion. It has been suggested that MGP improves in adulthood, but the Doucet and Crawford study 4 found no objective evidence among patients who reported improvement.

Conclusion

We have presented a rare case of bilateral MGP in a young boy with developmental delay. It is important to evaluate and follow such a patient for development of strabismus and/or amblyopia, as well as to counsel the patient/parents on the advisability of eyelid surgery for the ptosis or the winking phenomenon, depending on the severity of the presentation and its potential to affect vision.

References