



## Jaw Winking Syndrome -A Review

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### Abstract

JWS is a syndrome which is characterized by synkinesis: that is when two or more than two muscles which have independent nerve supply, having simultaneous or coordinated movements. In 1883, Marcus Gunn had described a 15-year-old girl with a rare condition of a type of congenital ptosis that showed an associated motion of winking of the affected eyelid on the movement of the jaw. Hence this syndrome also termed as Marcus Gunn phenomenon or Marcus Gunn jaw winking syndrome. JWS is usually seen unilaterally, but can present bilaterally in rare cases [1-3]. The winking of eyelid may be elicited by opening the mouth, thrusting the jaw to the contralateral side, jaw protrusion, chewing, smiling, or sucking. This wink phenomenon is often discovered early, as the infant is bottle-feeding or breastfeeding. Both the sexes are equally affected with no racial predilection. Incidence of this syndrome is approximately 5% amongst the population [1].

**Keywords:** Jaw Winking; Autosomal Dominant; Marcus Gunn; Synkinesis

### Introduction

Jaw-Winking Syndrome (JWS) or trigemino-oculomotor synkineses or pterygoid levator synkinesis is an autosomal-dominant condition with incomplete penetrance, in which the patients manifest with rhythmic upward jerking of their upper eyelid.

### Pathophysiology

Marcus Gunn jaw-winking syndrome is condition usually seen at birth. The winking of the infant is first noted by the parents while feeding the infant. However it has been suggested, and the older patients have often noticed, that the jaw-winking improves over time; but, it has not proven to be true on objective evaluation. In most cases patients stop taking care as they get older, or they learned to compensate and mask the winking response [6]. JWS is thought to be a form of synkinetic ptosis. There is an aberrant connection thought to exist between the motor branches of the trigeminal nerve innervating the external pterygoid muscle and the fibers of the superior division of the oculomotor nerve which innervate the levator superioris muscle of the upper eyelid [6,7].

Several electromyographic studies have demonstrated the synkinetic innervation by showing simultaneous contraction of the external pterygoid and the levator muscle. In rare cases, synkinesis

may be present between the internal pterygoid and levator muscles. In these cases, the eyelid elevates on closing the mouth and clenching the teeth [8].

A few authors have speculated that the jaw-winking is not due to a new aberrant pathway, but rather the disinhibition of preexisting phylogenetically more primitive mechanisms. This is thought to explain why individuals who are not affected will often open their mouths while attempting to widely open their eyes to place eye drops or to apply makeup. External pterygoid-levator synkinesis is the more common group.

There are two groups of trigemino-oculomotor synkinesis [9].

1. External pterygoid-levator synkinesis: in this synkinesis the eyelid raises upon:
  - Jaw thrust to contralateral side (unilateral external pterygoid)
  - Jaw is projected forward (bilateral external pterygoid).
  - Mouth is opened wide.
2. Internal pterygoid-levator synkinesis: in this synkinesis the eyelid raises upon clenching of teeth.

### Mortality/morbidity

JWS is associated with strabismus in 50 - 60% of cases. Superior rectus palsy is found in 25% of cases, and double elevator palsy is found in another 25% of cases [10]. In double elevator palsy, a deficiency in elevation of the globe occurs in all positions of gaze, secondary to an apparent weakness of the superior rectus and inferior oblique muscles. In rare cases, there is horizontal strabismus seen in the absence of a vertical motility disturbance seen. Incidence of anisometropia among patients with JWS is reported to be 5 - 25%. Amblyopia occurs in 30 - 60% of patients with JWS and is almost always secondary to strabismus or anisometropia, and only rarely, is due to occlusion by a ptotic eyelid [10].

### Clinical presentation

It has been estimated that the synkinetic movement of the mandible and eyelid is found between 2% and 13% of all cases of congenital ptosis. Previous authors have reported that the Marcus Gunn phenomenon is more commonly seen in males as opposed to females and that the left eyelid is more frequently involved than the right. However, this predilection has been refuted by Doucet, Crawford and Pratt et al. Bilateral and familial instances of the phenomenon are rare although Doucet and Crawford (1981) reported two patients with bilateral involvement in their series but none with a family history of the disorder. Ophthalmological conditions may accompany the Marcus Gunn phenomenon and an incidence of 36% for strabismus and 34% for amblyopia was found in Doucet and Crawford's series. Similarly, weakness of the superior rectus muscle on the affected side is associated in approximately three out of four cases of the condition. 11 Signs and symptoms of JWS include mild to moderate blepharoptosis, usually unilateral. Synkinetic movement of the upper eyelid with jaw-winking after one of the movements either mouth opening or jaw movement towards the opposite side, chewing, sucking, jaw protrusion, clenching of teeth together, swallowing, jaw-winking worsening in downgaze, decreased vision secondary to amblyopia, Strabismus (Vertical deviation and Horizontal deviation (rare cases)) [10,11].

### Clinical examination

Complete ophthalmic examination has to be carried out with the help of an Ophthalmologist. That includes pupillary examination, visual acuity, (amblyopia in infants and children needs to be ruled out), cycloplegic refraction (anisometropia has to be ruled out). Externally, one has to examine for extraocular motility, perform the cover test (performed to rule out a superior rectus or double elevator palsy), Bell phenomenon (can be decreased with a superior rectus or double elevator palsy), checking the head position, if a child does not elevate the chin in the presence of moderate-to-severe ptosis, then amblyopia has to be considered. The level or severity of ptosis should be assessed with the jaw immobilized in

a central position and after fusion is disrupted with brief ocular occlusion. Attempt to elicit synkinesis of eyelid movement by asking the patient to open the mouth and move the jaw from side to side, or protrude the jaw forward. Jaw-wink can be quantified as follows- Mild (less than or equal to 2 mm), moderate (3-6 mm), or severe (greater than or equal to 7 mm) [12].

### Histologic findings

Light microscopy of surgical specimens usually reveals normal striated muscle. Fibrosis within the muscle has been reported [13].

### Surgery

Any patient requiring an eyelid surgery, associated strabismus must be assessed.

### Superior rectus palsy

Correction of superior rectus palsy can be done by resecting the superior rectus muscle but only if there is no inferior rectus restriction. As the superior rectus is loosely attached to the overlying levator, the upper eyelid will be pulled inferiorly during resection, exaggerating the ptosis which is already present. It can be managed by subsequent ptosis repair.

### Double elevator palsy

Double elevator palsy presents as a deficit in the elevation of the eyeball in all fields of vision. It can be due to restriction of superior rectus and inferior oblique palsy and/or inferior rectus muscle. Inferior rectus restriction may be elicited by positive forced ductions in elevation, normal force generations in upward gaze which indicates an absence of superior rectus or inferior oblique muscle palsy, Bell phenomenon may be absent on the affected side. Inferior rectus restriction is treated by recession of the inferior rectus muscle. The double levator palsy requires a transposition procedure to displace the medial and lateral recti muscles superiorly (Knapp procedure). Many techniques have been suggested for the correction of jaw-winking ptosis, which shows the treatment controversy regarding the surgical management of this condition. If the jaw-winking is cosmetically acceptable, it can be ignored in the treatment of the ptosis. If the ptosis is mild, the patient may not want to go ahead with surgery. If at all the correction is desired by the patient, Müller muscle and conjunctival resection (MMCR), a Fasanella-Servat procedure [14], or a standard external levator resection [6,15], is performed. If the ptosis is moderate to severe, a levator resection may be advised. Beard advocated performing more resection than normal to avoid undercorrection. In severe ptosis, a supermaximum (> 30 mm) levator resection or frontalis suspension is necessary. 6 Although the amount of ptosis and synkinetic eyelid movement is variable, those patients with more severe ptosis tend to have the worse aberrant upper eyelid movement. The jaw-wink is considered cosmetically significant if it is 2 mm or more. Any treatment to repair the ptosis

without treating the jaw-winking would lead to an exaggeration of the aberrant eyelid movement to a level well above the superior corneal limbus, which would be unacceptable to the patient.

If the jaw-wink is significant, ablation of the levator and resuspension of the eyelid to the brow are necessary. Several techniques have been suggested to obliterate levator function, which effectively dampens the aberrant eyelid movement. Bullock advocated complete excision of the levator aponeurosis and muscle all the way to the orbital apex [16]. Dillman and Anderson argued that removal of a portion of the levator muscle above the Whitnall ligament (ie, myectomy) is adequate to obliterate its function without extensive dissection and damage to eyelid structures [17]. Bowyer and Sullivan describe the removal of a portion of levator muscle above the Whitnall ligament through a posterior conjunctival approach [18]. Dryden et al proposed suturing the transected levator aponeurosis to the arcus marginalis of the superior orbital rim [19]. This technique not only effectively deactivates the muscle but also allows the procedure to be reversed, if necessary. Beard and others have advocated bilateral excision of the levator muscle and bilateral frontalis suspension [6]. While this approach almost completely eliminates the wink and arguably results in better symmetry, it is often difficult to persuade the parents and the patient to perform surgery on and effectively damage the normal contralateral levator muscle.

Satisfactory and predictable results also can be obtained after only unilateral levator excision on the affected side, combined with bilateral frontalis suspension. This leaves the normal functioning levator muscle to elevate the nonptotic eyelid in primary position but produces a lag in downgaze for improved symmetry. Kersten et al advocate unilateral levator muscle excision and frontalis sling only on the affected side [20]. If the postoperative result is judged to be unsatisfactory, the parents or the patient can opt for further surgery to the contralateral side. Any amblyopia and strabismus should first be addressed, as there may be insufficient drive to lift the disinserted eyelid. Islam et al described a technique of dissecting a frontalis flap hinged superiorly through a suprabrow incision that is then brought down into an eyelid crease incision. The frontalis flap is used to suspend the ptotic eyelid after extirpation of the levator muscle [21]. Lemagne and Neuhaus described techniques that involve transection of the involved levator followed by transposition of the distal segment to the brow, which effectively suspends the eyelid to the frontalis muscle. Their techniques maintain normal eyelid contour, as the levator aponeurotic attachments are left undisturbed [22].

### Complications of surgery

Complications of surgery include the following:

- Height or contour of the eyelid may be unacceptable.
- Postoperative wink may get exaggerated.

- Asymmetry of eyes.
- Disturbed wound healing.
- Wound infection.
- Bleeding or hematoma formation at the site of surgery.

### Prognosis of the surgery

Results achieved through medical and surgical approach for the treatment of Marcus Gunn jaw-winking ptosis are usually satisfactory.

### Conclusion

Surgery is only indicated if the droopiness of the eyelid or of the jaw winking is objectionable or only if the patient complains that symptoms are more cosmetically unacceptable. Surgeries are available for many different intensities depending on the droop of the eyelids.

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