Jaw-winking marcus gunn management in Ophthalmology Department
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ABSTRACT

Introduction: Jaw-Winking Marcus Gunn was first introduced in 1883 as unilateral ptosis with eyelid retraction due to activation of pterygoid muscle (jaw movement). Marcus Gunn jaw-winking usually appear from birth. Some of the techniques proposed to eliminate thelevator function, which levator function can effectively reduce the movement of the eyelid. This report aims to describe the case of Jaw-Winking Marcus Gunn treated with levator resection procedures and frontal suspension.

Case Description: A woman 23 years old came to Sanglah Hospital complaining the left eyelid moves up and down with the movement of the mouth, especially when eating and talking. Right eye visual acuity 6/6, left eye 6/30 pinhole 6/20 C-1.00 Ax 180 6/12, orthophoria in primary position. Vertical fissure 5 mm, margin reflex distance -1 mm when the mouth is closed and +5 mm when the mouth is opened, levator action 4 mm, margin limbal distance 3 mm. Funduscopic examination of the posterior segment of both eyes in this patient was normal. The Patient was diagnosed as Marcus Gunn jaw-winking ptosis and planned for management with myectomy and fascia lata frontal sling. Ten months after surgery, eyelid movement when chewing is still visible. The results of the examination showed right eye visual acuity is 6/6 and left eye 6/30 pinhole 6/12 with a correction of the C-1.00 Ax 180 is 6/12. Vertical fissure 9 mm, margin reflex distance +2 mm when the mouth is closed and +3 mm when the mouth is opened, levator action 0.

Discussion: Levator resection technique and frontal suspension procedures can result in resolution of Jaw-Winking Marcus Gunn.

Keyword: Jaw-winking Marcus Gunn, Ptosis, Eyelid movement, Myectomy


INTRODUCTION

Jaw-winking Marcus Gunn was first described in 1883 as unilateral ptosis with eyelid retraction associated with activation of the pterygoid muscle (jaw movement). Approximately 50% of blepharoptosis cases are congenital.1,2 Marcus Gunn incidence is an example of congenital synkinesis ptosis that accounts 5% of all cases of congenital ptosis with various degrees. Acquired forms have been described after eye surgery, trauma, post-Bell’s palsy and pontine tumors.

Jaw-winking ptosis is almost always sporadic, but familial cases have been reported with autosomal dominant. No known racial predilection exists. Early reports showed jaw-winking ptosis to be more prevalent in females than in males; however, larger case series have shown an equal prevalence among males and females. Marcus Gunn jaw-winking syndrome is usually evident at birth. The winking phenomenon is often first noted by the parents when the infants are feeding.

The etiology of Marcus Gunn jaw-winking is obscure. It was originally assumed that the syndrome was is caused by aberrant innervation of the levator palpebrae superioris from the motor branch of the trigeminal nerve because of the close approximation of the nuclei of the oculomotor and trigeminal nerves. Jaw-winking is a congenital condition that is thought to occur as a result of the pterygoid-levator synkinesis. Abnormal appropriate neural pathways can not be explained, but the basic idea is that of a branch the 5th cranial nerve is responsible directly or indirectly supplies the levator palpebra muscle. Levator activation derived from proprioceptive receptors pterygoid muscles, thereby connecting the pterygoid movement that causes the eyelid elevation. Movement of the eyelid ptosis is most often due to the movement of the contralateral side of the mandible.

The characteristic feature of the Marcus Gunn jaw-winking phenomenon is that the raising and not winking of the affected eyelid is synchronous with and proportionate to the opening of the mouth. When the baby or child is opening his or her mouth, sucking or chewing, the ptotic upper eyelid elevates to an equal or even higher level than the contralateral normal eyelid. Another

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Nerves of Marcus Gunn movement with activation of ipsilateral pterygoid muscle could also lead to eyelid elevation including jaw movement toward the contralateral side or jaw protrusion. The amplitude of the wink tends to be worse in downgaze. The rapid, abnormal motion of the eyelid can be the most disturbing aspect of the jaw-winking syndrome. It is usually unilateral but may be bilateral in rare cases. The amount of jaw-winking is the excursion of the upper eyelid with synkinetic mouth movement as mild < 2mm; moderate 2-5 mm; severe > 6 mm.

Marcus Gunn jaw-winking associated with other ocular diseases such as strabismus (50-60%) as a result of superior rectus palsy (25%) and double elevator palsy (25%). Double elevator palsy manifests as a deficit in the elevation of the globe in all fields of gaze. The incidence of anisometropia among patients with Marcus Gunn jaw-winking syndrome is reported to be 5-25%. Amblyopia occurs almost always secondary to strabismus or anisometropia, and only rarely, is due to occlusion by a ptotic eyelid.

Medical management of Marcus Gunn jaw-winking is if amblyopia is encountered, treat aggressively with occlusion therapy and/or correction of anisometropia prior to any consideration of ptosis surgery. Patients who require eyelid surgery, first address associated strabismus. Superior rectus palsy can be corrected by resecting the superior rectus muscle but only in the absence of inferior rectus restriction. Double elevator palsy may be the result of superior rectus and inferior oblique palsy and/or inferior rectus restriction. Inferior rectus restriction is treated by recession of the inferior rectus muscle. A combined superior rectus and inferior oblique palsy requires a transposition procedure to displace the medial and lateral recti muscle superiorly.

Considering eyelid surgery only when the parents or the patient and the surgeon agree about whether the most cosmetically objectionable condition is the ptosis or the jaw-winking or whether it is a combination of both. It can be ignored in the treatment of the ptosis if the jaw-winking is cosmetically insignificant. The patient may elect not to proceed with surgery if the ptosis is mild, if correction is desired, perform a Muller muscle and conjunctival resection (MMCR), a Fasanella-Servat procedure or a standard external levator resection. Levator resection may be indicated if the ptosis is moderate to severe. Beard advocated performing more resection than normal to avoid undercorrection. Maximum levator resection (> 30 mm) or frontal suspension is necessary for severe in severe ptosis.

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. Dillman and Anderson argued that removal of a portion of the levator muscle above the Whitnall's ligament (myectomy) is adequate to obliterate its function without extensive dissection and damage to eyelid structures. Beard and others have advocated bilateral excision of levator muscle and bilateral frontalis suspension. Kersten et al advocated unilateral levator muscle excision and frontalis sling only on the affected side. The parents or the patient can opt for further surgery to the contralateral side if the postoperative result is judged to be unsatisfactory.

Fascia lata has been primarily used for the permanent surgical correction of congenital ptosis with poor levator function (0-4 mm). Fascia lata can be harvested from autogenous source or donor or irradiation materials can be used. Other materials are also used for frontalis sling which includes palmaris tendon graft, deep temporalis fascia graft, mersilene mesh, gortex, silicon rods, supramid or various other sutures. Fascia lata has the advantage of having the best biocompatibility with least chances of extrusion or granuloma formation. Silicon rod has an advantage of being elastic and is the material of choice in chronic external ophthalmoplegia to overcome residual lagophthalmos. Suture sling is usually used in children less than five years of age because of inadequate length of fascia lata. Mersilene mesh is the preferred material of some surgeons but it does have a tendency to extrude and produce granulomas.
CASE REPORT

A woman 23 years old came to Sanglah Hospital complaining the left eyelid moves up and down with movement of the mouth, especially when eating and talking. Patient was also disturbed by the movement of her eye. The winking of the left eye has been recognized since birth by her parent when breastfeeding. Patient also said that the right eye sees more clearly than the left eye. Another eye diseases and systemic complaints were denied. History of labor in shaman with unknown gestational age.

Examination results on 3rd January 2013 obtained the visual acuity of the right eye 6/6, the left eye 6/30 pinhole 6/20 C-1.00 Ax180 6/12, no signs of strabismus with orthophoria in primary position. Further examination of ptosis is as follows:

<table>
<thead>
<tr>
<th>RE (mm)</th>
<th>LE (mm)</th>
</tr>
</thead>
<tbody>
<tr>
<td>Vertical Fissure (VF)</td>
<td>5 (closed mouth)</td>
</tr>
<tr>
<td>Horizontal Fissure (HF)</td>
<td>11 (open mouth)</td>
</tr>
<tr>
<td>Lid crease</td>
<td>30</td>
</tr>
<tr>
<td>Margin Reflex Distance (MRD)</td>
<td>4</td>
</tr>
<tr>
<td>Margin Limbal Distance (MLD)</td>
<td>-1 (closed mouth) +5 (open mouth)</td>
</tr>
<tr>
<td>Levator Action (LA)</td>
<td>4</td>
</tr>
<tr>
<td>Bell’s phenomenon</td>
<td>+</td>
</tr>
<tr>
<td>Eyelid lag</td>
<td>+</td>
</tr>
</tbody>
</table>

Both eyes movements were normal in all gaze. Examination of the anterior segment of the right and left eyes with slit lamp were within normal limits. Funduscopic examination of the posterior segment of both eyes in this patient were normal. Patient was diagnosed as Marcus Gunn jaw-winking ptosis and planned for management with myectomy and fascia lata frontal sling.

The surgery was performed on 29 January 2013. After anesthesia with general anesthesia and disinfection of the surgical field created markers in the region of the left femur 5 cm from condylus to obtain fascia lata, perform incision until there was tensor fascia lata muscle. Fascia lata was harvested with size 1 cm x 3 cm. Created markers on the left eyelid by measuring the contralateral normal eyelid crease of 6.5 mm from the border of the lid, then made the points parallel to the pupil, medial and lateral limbal upon the skin crease, connect the dots as if forming a skin crease approximately 25 mm. The dots parallel to the medial and lateral limbal were made straight lines toward the frontalis up over the brow approximately 3 cm. Performed a horizontal incision at the lid crease line, separated layer by layer, so that it looked the levator muscle. Separated the levator muscle by using desires hook, performed myectomy levator muscle above the Whitnall’s ligament as much as 2 mm. Performed incision above the eyebrow to look frontalis muscle, made tunnel to penetrate further towards the palpebral margin. Fascia lata placed vertically in the tunnel by connecting aponeurosis levator and frontalis muscle. Sutured the bottom of fascia lata to the levator muscle with vicryl 6.0, then performed suturing the skin-fascia-skin as much as 3 stitches in the middle to form a lid crease, followed by suturing skin to skin around the edges with prolene 7.0. Fascia lata over the brow stitched to frontalis muscle with vicryl 6.0 followed by suturing skin to skin with prolene 7.0.

Post operatively, the patient was given oral antibiotic, oral anti inflammatory, oral analgetic, added with antibiotic-steroid eye drop, antibiotic eye ointment, lubricant eye drop and eye ointment. First day post operatively, patient complained of pain and difficult to open eyelid. Examination results on 30 January 2013 were obtained the visual acuity on the right eye 6/6 and the left eye 6/20, palpebral swelling, good stitch, MRD +1, lid crease is formed, the levator function is still difficult to be evaluated.

Ten days after surgery, patient had no complains. Examination results showed visual acuity on the right eye 6/6 and left eye 6/30 pinhole 6/20, MRD +1 when the mouth is closed and +2 when the mouth is opened, lid crease 4 mm, then the sutures were released.

One month post operatively, visual acuity on the right eye 6/6, left eye 6/12 with C-1.00 Ax 180 6/12, MRD +2 when the mouth is closed and +5 when the mouth is opened, lid crease 4 mm. Lid lag evident in the left eye.

Ten months after surgery, patient complained about watery on the left eye without discharge,
DISCUSSION

Congenital neurogenic ptosis is caused by innervational defects during embryonic development. Congenital neurogenic ptosis may also be synkinetic. Marcus Gunn jaw-winking syndrome is the most common form of congenital synkinetic neurogenic ptosis. In this synkinetic syndrome, the unilaterally ptotic eyelid elevates with jaw movements. The lateral mandibular movement to the contralateral side is the most commonly causes elevation of the ptotic eyelid. The wink phenomena may be elicited by opening the mouth, thrusting the jaw to the contralateral side, jaw protrusion, chewing, smiling or sucking. This phenomenon is usually first noticed by the mother when she is feeding or nursing the baby. In this case, patient complained flickering movements in the left eye became apparent during chewing. The movements have been realized by her parents since birth.

Marcus Gunn jaw-winking is usually unilateral, it can present bilaterally in rare cases. In this case, ptosis occured only in the left upper eyelid since birth.

The incidence of anisometropia among patients with Marcus Gunn jaw-winking syndrome is reported to be 5-25%. Amblyopia occurs 30-60% and almost always secondary to strabismus or anisometropia, and only rarely, is due to occlusion by a ptotic eyelid. Examination results showed, visual acuity of the right eye 6/6 and left eye 6/30 with C-1.00 Ax 180 6/12, where it is proved that there is amblyopia in the left eye.

Ptosis can be measured with three measurements such as margin reflex distance (MRD), levator function and vertical interpupillary distance. Margin Reflex Distance 1 (MRD1) is the single measurement of skin crease.

**Figure 3** Ptosis Examination. A. Measurement of skin crease. B. Margin Reflex Distance. C. Levator Action. D. Bell’s phenomenon. E Eyelid lag appears when opening the mouth.

**Figure 4** RLE fundus

**Figure 5.** Surgery Procedures
The normal interpupillary fissure height averages 10 mm with the range from 8-12 mm in adults. The levator function directly determines the type and amount of surgery that needs to be performed. Levator function may be classified as excellent (13-15 mm), good (8-12 mm), fair (5-7 mm), poor (<4 mm).\(^2\)\(^3\)\(^1\)\(^3\) Ptosis measurements results that have been performed in this patient showed that vertical interpupillary fissure is 5 mm, levator function is 4 mm.

The position of the ptotic eyelid in downgaze can help differentiate between congenital and acquired causes. The congenitally ptotic eyelid is typically higher in downgaze than the contralateral, as a result of eyelid lag.\(^4\)\(^4\) The results of examination in this case has positive-looking eyelid lag when opening the mouth.

Bell's phenomenon which normally would indicate eyeball rolling upward. Eyeball that can not move up or when rolling to another position like downward will increase the incidence of exposure keratopathy due to lagophthalmos as complication after surgery.\(^4\)\(^4\) The patient in this case had a positive Bell's phenomenon so that the incidence of exposure keratopathy can be avoided.

The amount of jaw-winking is the excursion of the upper eyelid with synkinetic mouth movement. It classified as: mild (< 2 mm); moderate (2-5 mm); severe (> 6 mm).\(^1\)\(^7\) Patient in this case can be categorized into severe jaw-winking ptosis because of the distance of vertical eyelid interpupillary when the mouth is closed and opened is 6 mm.

Correction of anisometropia and/or occlusion therapy prior to any consideration of ptosis surgery to encounter amblyopia. The jaw-wink is considered cosmetically significant if it is 2 mm or more, so the parents and the patient consider the surgical management. The results of the examination showed that the eye position was orthophoria. Right eye visual acuity 6/6 and left eye 6/30 with C-1.00 Axs180 6/12. Amblyopia has occurred in the ptotic eye. The patient was planned for eyeglasses after surgery because she feels more disturbed by the movements of her jaw-wink. Indication of surgical management is for cosmetic purpose.

Several techniques have been suggested to obliterate levator function, which effectively dampens the aberrant eyelid movement. Frontalis suspension using fascia lata is frequently used after obliterate the levator function. The frontal suspension technique is reserved for patients with poor levator function (<4 mm) and intact frontalis muscle. Kersten et al (2005) advocate unilateral levator muscle excision and frontalis sling only on the affected side. Fascia lata has the advantage of having the best biocompatibility with least

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Figure 6. One day after surgery

Figure 7. Ten days after surgery

Figure 8. One month after surgery. A. MRD +1 when the mouth is closed. B. Lid lag apparent.

Figure 9. Ten months after surgery. A. MRD +2 when the mouth is closed. B. MRD +4 when the mouth is open.

most important measurement in describing the amount of ptosis, which is in severe ptosis the light reflex may be obstructed by the eyelid and therefore have a zero or negative value.\(^4\) The normal MRD 1 is between 4-4.5 mm.\(^1\)\(^3\) MRD 1 test result in this patient showed a value of -1 when the mouth is closed.
chance of extrusion or granuloma formation or rejection.\textsuperscript{1,2,10,13,15} The patient has a 4 mm levator function which can be categorized into severe ptosis and planned for frontalis suspension using fascia lata. Fascia lata was harvested from the tensor fascia lata muscle of the left leg. The surgical procedures were myectomy levator muscle about 2 mm followed by attaching the fascia lata to levator muscle upon eyelid and frontalis muscle, thus obtained tarsal lynching in frontalis muscle.

Zelinsky (2006) suggest antibiotic eye ointment, lubricant ointment, and eye drop four times a day to protect the eye and comfort the patient.\textsuperscript{16} Edema usually improves after 2-4 weeks, but in severe cases can be given antiinflammatory corticosteroid (methylprednisolone) if needed.\textsuperscript{17} After surgery, patient in this case was given oral antibiotic to prevent infection, oral analgetic to relieve pain and anti inflammatory, antibiotic-steroid topical, lubricant eye ointment and eye drop.

The desired results after surgery is resolution (\(\leq 1\) mm movement upper lid along synkinetic jaw movement), improved (\(\leq 2\) mm and > 1 mm residual motion), did not improved (\(< 2\) mm, but > 1 mm residual motion).\textsuperscript{9} The results after ten months post operatively showed vertical fissure 8 mm, MRD when the mouth is closed +2 and when the mouth is opened is +3, it is difference is 1 mm. The difference of 1 mm can be categorized as resolution in this case.

CONCLUSION

A 23 years old female patient was diagnosed with left eye Marcus Gunn jaw-winking ptosis and myectomy levator muscle and frontalis suspension with fascia lata has been performed. Examination results of ten months post operatively showed good result in eye cosmetic.

Treatment of Marcus Gunn jaw-winking patient requires a proper diagnosis, good planning, through understanding the anatomy of the eyelid and good surgical techniques. Severity of the ptosis, the type, and the degree of levator function are the most desicive factor in order to obtain good results in function and cosmetics.

REFERENCES