

Treatment of Marcus Gunn Jaw Winking Ptosis

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ABSTRACT

Objective To share the experience of treating Marcus Gunn jaw winking ptosis by levator disinsertion followed by frontalis sling procedure.

Study design Case series.

Place & Duration of study Oculoplastic Department, Khalid eye clinic Karachi, from May 2019 to October 2019.

Methodology Patients of unilateral congenital Marcus Gunn jaw winking ptosis of both genders with an age range of 9 to 15 years were enrolled. Levator disinsertion followed by frontalis sling procedure was performed in all cases. The inclusion criteria for undergoing levator muscle disinsertion followed by frontalis sling comprised of drooping of the eyelid (ptosis) of greater than 2mm with moderate to severe jaw winking of the affected eye. The rectification of ptosis, eyelid equilibrium, improvement of winking and postoperative complications were assessed. A successful surgical outcome was classified as jaw winking of less than 1mm of affected eyelid with synkinetic jaw movement. Postoperative follow up period was of three months. Approval of ethical review committee and informed consent were obtained from the parents.

Results Marcus Gunn jaw winking ptosis was more common in females. Ten out of eleven patients had excellent cosmetic outcome comparable to the contralateral eye at the end of follow up period of three months. Residual jaw winking was only seen in one patient despite good ptosis correction.

Conclusion Levator disinsertion followed by frontalis sling is an excellent technique for the rectification of ptosis and Marcus Gunn jaw winking phenomenon.

Key words Marcus Gunn jaw winking, Ptosis, Frontalis sling, Levator disinsertion.

INTRODUCTION:

First described in 1883 by Gunn, Marcus Gunn jaw winking phenomenon (MGJWP) is a synkinetic reflex retraction of the upper eyelid occurring simultaneously. When mouth opens, the jaw moves to the contralateral side (pterygoid-levator synkinesis). It may also occur with chewing, sucking, unilaterally or bilaterally.¹ It is a synkinesis that is associated with congenital eyelid ptosis (2% to 13% of cases) in an unusual manner; accompanied by

ophthalmologic conditions such as a high incidence of strabismus, amblyopia, anisometropia, superior rectus muscle weakness and double elevator palsy.^{2,3} The concurrent movements or a synchronized sequence of actions of muscles supplied by different nerves or by different branches of the similar nerve is known as synkinesis.⁴

MGJWP comprises of anomalous congenital relation among motor branches of trigeminal nerve controlling mastication muscles and superior division of oculomotor nerve controlling the levator palpebrae superioris, ensuing in concurrent contraction. Although most cases are congenital, acquired sporadic forms also exist.³ MGJWP may develop after ocular surgery, infection, trauma and tumors with spontaneous remission but the congenital form persists and does not improve as the person ages.

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It is usually first noticed by the mother while feeding when there is atypical elevation of the ptotic lid.⁵

Ophthalmologists usually recommend MGJWP to be treated with observation, without surgical intervention if jaw winking phenomenon is mild with less than 2mm ptosis and no cosmetic concern, whereas, surgical treatment is imminent for moderate to severe jaw winking and ptosis. The goal is to get rid of the synkinetic movement of the eyelid and improve the cosmetically unappealing drooping of the eyelid. There are various surgical procedures such as levator sling, levator resection/ plication with a tailored technique and excising the levator muscle with subsequent frontalis suspension available for the correction of MGJWP.⁶⁻⁸ A gold standard surgical procedure is still a controversy since complete elimination of synkinesis is difficult with present surgical techniques. This study aimed to share the experience of performing levator disinsertion followed by frontalis sling among patients having one-sided congenital Marcus Gunn jaw winking and ptosis.

METHODOLOGY:

This case series was conducted at the Oculoplastic Department of Khalid Eye Clinic Karachi, from May 2019 to October 2019. This study included eleven patients of unilateral congenital Marcus Gunn jaw winking ptosis of both genders with an age range of 9 years to 15 years. Approval was obtained from the institutional ethical review committee and carried out in agreement with the Declaration of Helsinki. Informed assent was taken from each of the eleven patients undergoing surgical intervention before the surgery and consent signed by their parents. Patients' demographics and history including personal and family history, past surgical record and additional eye related diseases were documented. Ocular examination and careful assessment was done before surgical intervention. The grading of ptosis was done as less than 2mm (mild), between 2-4mm (moderate) and more than 4mm (severe). Levator function was assessed as being good if it was more than 10mm, fair if between 5-10mm, or poor if less than 5mm. The winking phenomenon was labeled as mild when less than 2mm, moderate when between 2-5mm, or severe if more than 5mm.⁵

The inclusion criteria for undergoing levator muscle disinsertion followed by frontalis sling comprised of drooping of the eyelid (ptosis) of greater than 2mm with moderate to severe jaw winking of the affected eye. The rectification of ptosis, eyelid equilibrium, improvement of winking and postoperative

complications were assessed after the surgery. A successful surgical outcome was classified as jaw winking of less than 1mm of affected eyelid with synkinetic jaw movement. Patients were followed up for a period of three months postoperatively.

All the surgeries were carried out using general anesthesia. Unilateral levator muscle disinsertion with subsequent frontalis suspension of the affected eye was performed in all the patients. Silicone tube was used as the suspension material. Incision was made at the level of the eyelid skin crease, followed by revealing the Müller muscle and levator aponeurosis and horizontally transecting the levator aponeurosis from tarsal plate and Müller muscle with cautious incision and dissection of the medial and lateral edges of the levator aponeurosis as much as possible up to 1-2mm under the Whitnall's ligament, held with a hemostat and excised. Eyelid height was set according to the height of the contralateral normal eyelid. Frontalis suspension was done using two silicone tubes, the lateral, middle part and medial parts of which were sutured on the tarsal plate with 5-0 non absorbable suture. The two free edges of silicone tubes were coursed through the lateral and medial orbital septum tunnels and conical frontalis suspensions was formed on both medial and lateral side which were then taken out of the forehead cut (double rhomboid). The skin incisions were sutured with 6-0 sutures after cutting the remaining silicone tubes. Froste suture was applied to prevent exposure of the cornea and prescription of lubricating eyedrops and ointment was given for corneal safeguard followed by removal of the skin sutures after 2 weeks of the operation. The patients were called on 1st postoperative day for the removal of Froste suture and advised eyelid closure exercise. The follow up period was upto 3 months postoperatively.

RESULTS:

This study included eleven patients with ages ranging from 9 years to 15 years with the mean of 11 ± 3.08 year. There were 4 (36.3%) boys and 7 (63.6%) girls in the study. Three (27.7%) patients had involvement of right eye whereas eight (72.7%) patients had involvement of the left eye. Six (54.5%) patients had moderate ptosis with moderate winking while the five (45.5%) had severe ptosis with severe winking. At the end of the three month follow up period, ten (90.09%) patients had excellent cosmetic outcome, with negligible winking and vertical fissure height was similar to the contralateral eye. One (9.09%) patient was unsatisfied due to residual winking despite good ptosis correction. No other

complications were observed in this study.

DISCUSSION:

Marcus Gunn jaw winking phenomena and ptosis is a rare condition with variable severity and resultant cosmetic disability. There is a tendency for improvement with the age but no scientific evidence is provided.^{9,10} The patients included in this study did not report any such improvement prior to surgery. Patients suffering from MGJWP usually present early in life. Bower and Sullivan described 'habitual ptosis' in which the sufferers acclimatize and recompensate the jaw in a precise position to curtail the drooping of the eyelid which is usually less than the patient's actual ptosis.¹¹

MGJWP commonly occurs unilaterally but bilateral cases have been reported. It is more frequent on left side, which was in accordance with this study where MGJWP involved the left eye in 72.7% of the patients.¹² Studies have shown that the age of presentation is between 5 years to 11 years.^{13,14} In this study, the mean age was 11.5 year. It is important that the patients suspected of suffering from MGJWP should be referred to neurologists and ophthalmologists for the evaluation of associated conditions and secondary complications such as imbalance while focusing, squint, elevator palsy, superior rectus palsy, inborn nystagmus and hereditary fibrosis syndrome.¹⁵⁻¹⁷ A retrospective study observed the outcome of surgical rectification of eyelid drooping related to Marcus Gunn phenomenon and established that the level of ptosis was unsatisfactorily examined during ocular examination, thereby concluding that the surgical result is enhanced if extent of ptosis is thoroughly assessed before operating by immobilizing the jaw and temporarily occluding the ipsilateral eye.¹⁸

The main treatment for MGJWP is surgical especially for moderate-to-severe cases. The surgical outcome for the correction of MGJWP has been variable among different studies. A study was done on 9 patients, all undergoing unilateral frontalis suspension after levator excision and showed that 67% of the patients resulted in good surgical outcome with resolution of jaw winking and ptosis.¹⁹ Another study included 19 patients with 84.2% of the patients achieving resolution of MGJWP.²⁰ In this study, 90.09% of the patients who underwent unilateral levator muscle disinsertion with frontalis suspension showed excellent surgical outcome in terms of resolution of jaw winking and ptosis. It was also noticed that 63.6% of the recruited patients were females whereas no such gender disparity was found in other studies.^{11,21} One study noticed a

higher incidence of MGJWP among males with frequency of 78%.⁸

Lemagne described the transposition of levator muscle to the frontalis muscle to reinstate a little function of the levator muscle and resolve the jaw winking but this procedure did not correct the ptosis and required a further process of suspension of the brow for further tightening of the transposed levator.²² A modified technique was reported by Bajaj et al involving levator plication but this resulted in 70% of patients more than 1mm residual ptosis with synkinesis.⁷ In the aforementioned procedures, the levator muscle was not disinserted completely but held in reserve as a suspension for the frontalis muscle which lead to an increase chance of retaining a left over synkinesis owing to the lingering anomalous function of levator muscle and connection between the levator muscle and the upper eyelid.

The surgical technique used in this study included disinserting the levator muscle entirely followed by frontalis suspension using silicone tube in relieving both the jaw winking and associated ptosis. Demirci and Khwarg performed similar surgical technique but used autologous fascia lata for the frontalis suspension and reported a resolution of 97% and 85.2% jaw winking synkinesis in their patients.^{5,18} In this study silicone tube was used instead of fascia lata for frontalis suspension and resulted in an equally successful cosmetic and surgical outcome. Cates and Tyers performed a similar technique and reported minimal recurrent jaw winking after surgery in two of their patients comparable to this study with one patient having a residual jaw winking.¹³

CONCLUSIONS:

Unilateral levator muscle disinsertion followed by frontalis suspension with silicone tube is an effective surgical technique for the treatment of unilateral MGJWP, resulting in satisfactory symmetrical outcome and resolution of jaw winking and ptosis.

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