

Systematic Review

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# Surgical management of jaw-winking synkinesis and ptosis in Marcus Gunn syndrome: a systematic outcomes analysis

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## Abstract

**Aim:** Marcus Gunn jaw-winking synkinesis (MGJWS) is characterized by congenital ptosis in conjunction with rapid and involuntary elevation of the affected upper eyelid upon contraction of the ipsilateral external pterygoid muscle. Selecting an approach to the surgical management of eyelid malposition in this syndrome is challenging and requires careful discussion with each patient's family. In this systematic review, we describe reported surgical approaches, assess outcomes data, and attempt to identify areas of consensus in the management of MGJWS.

**Methods:** Twenty-seven peer-reviewed studies were identified, describing a variety of interventions.

**Results:** The most commonly-used surgical techniques included: bilateral levator excision with bilateral frontalis sling, unilateral levator excision with bilateral or unilateral frontalis sling, the Neuhaus/Lemagne method, and levator plication surgery. However, no clear outcomes-based consensus regarding choice of surgical approach was identified, highlighting the ongoing role of surgeon and family preference in the selection of management strategy. Further, there was considerable variability in the literature for reporting outcome measures, including grading schemes for ptosis and jaw-wink.

**Conclusion:** The existing literature on management of MGJWS does not enable the development of an evidence-based consensus algorithm regarding the selection of an appropriate surgical technique. The disorder is



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treated according to a case-by-case approach governed by surgeon and family preference. Standardization of nomenclature and outcome measures is crucial for obtaining higher-quality, generalizable data in futures studies.

**Keywords:** Marcus Gunn jaw-winking synkinesis, pterygoid-levator synkinesis, maxillopalpebral synkinesis

## INTRODUCTION

Marcus Gunn jaw-winking synkinesis (MGJWS), first described by Robert Marcus Gunn in 1883, is a disorder characterized by congenital ptosis accompanied by synkinetic elevation of the affected eyelid upon movement of the jaw<sup>[1]</sup>. Electromyographic studies demonstrate that contraction of the levator palpebrae superioris muscle of the ptotic eye occurs with stimulation of the ipsilateral external pterygoid muscle (by mouth opening, suction, or lateral excursion of the mandible). It is therefore also known as “maxillopalpebral synkinesis” or “pterygoid-levator synkinesis”. Among individuals with congenital ptosis, MGJWS is relatively common, with a reported incidence of 12.1%, and should be ruled out in all patients with this presenting complaint<sup>[2]</sup>.

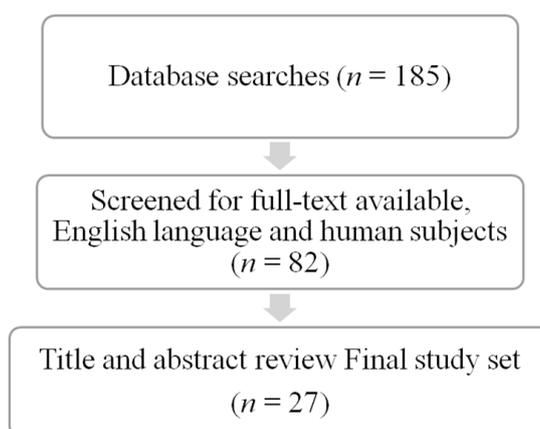
Although the exact etiology is unknown, a combination of genetic, myogenic, and neurological factors has been implicated. Some evidence suggests underlying KIF21A mutation, while histological analyses demonstrate a degree of levator muscle atrophy on both the affected and unaffected side, thus suggesting a critical role for neural input<sup>[3-5]</sup>. But while the existence of congenital aberrant connections is widely accepted, their exact location within the brain or peripheral nervous system remains unclear. One possible location is in the midbrain between the trigeminal mesencephalic nucleus and oculomotor nucleus while the most likely alternative is between the mandibular division of the trigeminal nerve (CN V3) and the oculomotor nerve (CN III)<sup>[6,7]</sup>. Patients may present with other congenital aberrant innervation disorders or neurological disorders such as morning glory disc anomaly, inverse Marcus Gunn syndrome, or Duane's retraction syndrome type 1<sup>[8-10]</sup>.

Treatment for MGJWS typically takes into account the extent of both functional and cosmetic impairments. Strabismus and amblyopia, when present, should be addressed. Repair of the baseline ptosis can be considered independent of jaw-winking correction, especially when the amplitude is low. However, in cases of moderate to severe jaw-winking, it is typically desirable to address the wink surgically. In general terms, surgery to eliminate the wink involves the disabling of levator function. Four types of surgeries are most commonly performed [Table 1]. These include: (1) bilateral levator excision with bilateral frontalis sling; (2) unilateral levator excision with bilateral or unilateral frontalis sling; (3) transection of the levator muscle followed by frontalis eyelid suspension using the distal segment of the muscle and aponeurosis, independently described by Neuhaus<sup>[11]</sup> and Lemagne *et al.*<sup>[12,17]</sup>; and (4) levator plication/resection without subsequent eyelid suspension (for patients with lower-amplitude jaw-winking). The primary difference between the Lemagne and Neuhaus procedure is that while the Lemagne procedure involves connecting the levator muscle to the frontalis muscle, with the intent of neurotizing the former with the latter, the Neuhaus procedure involves dividing the levator muscle into three strips as a mechanical sling. Apart from these four groups of procedures, several less commonly deployed options exist, including the Motais-Parinaud procedure (suspension of upper eyelid tarsus from the superior rectus muscle), the Friedenwald-Guyton technique (anchoring of the upper lid to the frontalis muscle with a single buried suture), and the Reese method (attachment of strips of orbicularis to the frontalis muscle)<sup>[18,19]</sup>.

The variety of surgical approaches for MGJWS, coupled with the absence of systematic and comprehensive outcomes analysis, present clinicians with significant challenges in terms of evidence-based technique selection. We herein evaluate the peer-reviewed outcomes literature pertaining to various surgical approaches for the management of MGJWS and attempt to identify common themes or areas of consensus.

**Table 1. Description of commonly performed surgical techniques for MGJWS**

Surgical technique	Description
Bilateral levator excision with bilateral frontalis sling	This procedure is well suited for severe blepharoptosis. Both levator muscles are dissected. The bilateral frontalis suspension is then most commonly performed using autogenous fascia lata. This procedure helps in achieving a symmetrical result in primary gaze and also during a downgaze <sup>[13]</sup>
Unilateral levator excision with bilateral (or unilateral) frontalis sling	In this approach, only the levator muscle to the ptotic eyelid is resected, while frontalis suspension is performed either only for the ptotic lid or in the normal lid as well <sup>[14]</sup> . Bilateral frontalis suspension after unilateral levator excision is designed to provide symmetrical lid height in downgaze while allowing the intact levator muscle to function in primary gaze <sup>[16]</sup>
Neuhaus/Lemagne method	In 1985, Russell Neuhaus and Jean-Michel Lemagne independently described a method to treat MGJWS by transection of the levator muscle followed by suspension of the eyelid to the frontalis using the distal segment of the muscle and aponeurosis, as opposed an autograft of fascia lata or a silicone rod <sup>[11,12]</sup> . While the Lemagne procedure involves connecting the levator muscle to the frontalis muscle with the intent of neurotizing the former with the latter, the Neuhaus procedure involves dividing the levator muscle into three strips as a mechanical sling
Levator plication or excision	In this technique, the levator muscle is resected or levator aponeurosis is plicated to effectively shorten the levator muscle and correct the ptosis. The amount of levator plication/resection to be performed depends on the desired eyelid height to be obtained. This a simpler technique but primarily addresses only the ptosis component and may not resolve jaw-winking <sup>[15]</sup>

**Figure 1.** Flowchart of the review process to identify the final study sample

## METHODS

A systematic review of the peer-reviewed literature as of March 31, 2020 was conducted using the search terms “Marcus Gunn jaw”, “Marcus Gunn jaw synkinesis”, “Marcus Gunn jaw ptosis”, “Marcus Gunn syndrome” and “maxillopalpebral synkinesis”. Terms related to acquired forms of eyelid ptosis and synkinesis, such “Marin-Amat syndrome”, were omitted. The databases searched were PubMed, Medline, and Cochrane. Search term and database selection, as well as inclusion/exclusion criteria, were based upon guidelines from the Cochrane Handbook of Systematic Reviews of Interventions<sup>[20]</sup>.

The initial search retrieved 185 unique articles. This pool was further limited to human studies in the English language, available in full-text either online or through interlibrary loan. These parameters narrowed the sample size to 82 articles. Upon title and abstract review, we identified 27 articles related to surgical management of the disorder, which were included in the final study sample [Figure 1].

Full text of the articles was reviewed for each article and the following data extracted: (1) publication details (authors, year, references); (2) sample size; (3) ptosis and jaw-wink severity, (4) management approach (type of surgery performed, sling material, and follow-up); (5) documented efficacy outcomes; and (6) short- and long-term complications.

## RESULTS

### Study characteristics

The 27 identified articles included seven case reports, one case series, and 19 retrospective analyses [Table 2]. Studies that describe jaw-winking synkinesis in broader contexts of non-ophthalmic conditions were not included in our analysis. The remaining 23 studies reported a variety of surgical interventions, some of which were experimental in nature or modifications of established procedures. Larger case series often included the results of multiple types of surgical intervention. No randomized controlled surgical trials were identified.

Reported surgical interventions included bilateral levator excision with bilateral frontal sling<sup>[13,19,21]</sup> (3 articles), unilateral levator excision with bilateral frontal sling<sup>[16,19,23,33]</sup> (4 articles), unilateral levator excision with unilateral frontal sling<sup>[16,28,29,32,33]</sup> (5 articles), the Neuhaus/Lemagne method<sup>[11,13,17,29,35,37]</sup> (6 articles), levator plication/resection without eyelid suspension<sup>[13,15,30,33]</sup> (4 articles), the Motais-Parinaud procedure<sup>[26,33]</sup> (2 articles), and the use of an orbicularis oculi muscle flap<sup>[27,30]</sup> (2 articles). Although the Neuhaus and Lemagne methods differ slightly, the procedures are not significantly differentiated with regard to practice and analyses of outcomes in the majority of studies we found. For details of studies that discuss experimental procedures, see Table 2.

### Efficacy outcomes

There was considerable variability between studies in terms of how severity of ptosis and jaw-winking was documented and reported. Eyelid position was described using a variety of parameters, including quantitative measures such as eyelid height (margin to reflex distance, or MRD-1), palpebral aperture, and upper eyelid excursion; and qualitative measures such as eyelid contour and eyelid symmetry. Even in instances when studies employed similar outcome measures, definitions of these parameters were noted to vary. For instance, Doucet and Crawford<sup>[35]</sup> categorized jaw-wink amplitude as Grade I (< 2 mm of change in MRD-1), Grade II (2-6 mm), and Grade III (> 6 mm), while Dave *et al.*<sup>[22]</sup> considered < 2 mm mild, 2-4 moderate, and > 5 mm severe. Raw data for individual patients were not provided. Doucet and Crawford acknowledged that the jaw-winking severity scale they utilized was arbitrary, based on personal observation of the magnitude of eyelid movement deemed “noticeable”. The degree of eyelid symmetry was also reported using varying scales; Morris *et al.*<sup>[34]</sup> defined the outcome of “satisfactory” symmetry as a difference of ≤ 1 mm between the MRD-1 of the operated and unoperated eyes, while Cates and Tyers<sup>[21]</sup> and Ibrahim<sup>[35]</sup> reported symmetry as “good” or “acceptable” without quantitatively defining these terms. Eyelid contour was reported descriptively in all studies.

Given the variability of grading schemes used, we evaluated the studies that reported surgical outcomes of a given procedure for a discrete number of MGJWS patients. Three out of 3 studies that analyzed bilateral levator excision followed by bilateral frontalis suspension resulted in satisfactory improvement of both ptosis and jaw-winking<sup>[13,19,21]</sup>. Two out of 4 studies that reported on unilateral levator excision followed by bilateral frontalis suspension resulted in improvement of both ptosis and jaw-winking<sup>[16,33]</sup>; the other two studies<sup>[19,23]</sup> revealed unsatisfactory outcomes for ptosis but improvement in jaw-winking. Four out of 5 studies that described unilateral levator excision with unilateral frontalis suspension resulted in satisfactory improvement of both ptosis and jaw-winking<sup>[16,28,29,32]</sup>; however, one study<sup>[33]</sup> did not report any improvement in either ptosis or jaw-winking. Six out of 6 studies that assessed the Neuhaus/Lemagne method resulted in satisfactory improvement of both ptosis and jaw-winking (with significantly more improvement in jaw-winking). Of 4 studies that analyzed levator plication/resection without eyelid suspension, one resulted in satisfactory improvement of both ptosis and jaw-winking<sup>[15]</sup>; two of these four studies<sup>[13,30]</sup> revealed resolution of ptosis but undercorrection of jaw-winking; and one of these four studies<sup>[33]</sup> revealed only moderate correction of ptosis (in 63% of patients) and no improvement in jaw-winking.

**Table 2. Included peer-reviewed articles on surgical management of Marcus-Gunn jaw-winking synkinesis**

Author(s)	Year	Sample size	Ptosis severity	Management	Outcome
Beard <sup>[25]</sup>	1965	1	Severe	Levator excision of the unaffected eyelid followed by bilateral brow suspension	This approach is recommended for a failed prior surgery. Resulted in satisfactory outcomes
Nagpaul and Charan <sup>[26]</sup>	1968	1	5 mm	Motais-Parinaud procedure	Moderate improvement shown after the operation. Paretic superior rectus did not improve
Tsai <i>et al.</i> <sup>[27]</sup>	2002	1	Severe	Orbicularis oculi muscle flap	Used the orbicularis oculi muscle flap to elevate dynamically the ptotic eyelid and to eliminate the synkinetic reflex without levator excision. This approach had successful outcomes
Yoshikata and Yanai <sup>[28]</sup>	1999	1	Severe	Unilateral excision of levator muscle followed by unilateral frontalis suspension	33-yr-old patient had satisfactory surgical outcomes
Carbajal <sup>[18]</sup>	1959	5	N/A	A case-by-case approach: levator tucking, Blaskovics, tenectomy and Friedenwald-Guyton, tenectomy and Reese	Except for one case, all patients experienced recurrence between 6 and 23 months
Bajaj <i>et al.</i> <sup>[15]</sup>	2015	10	4.25 ± 0.79 mm	Levator plication	10 patients underwent modified levator plication surgery. 9 patients showed correction of ptosis and 3 had resolution of MGJWS. Resolution of MGJWP was defined as less than 1 mm of excursion of upper eyelid with synkinetic mouth movement. Ptosis correction (2.40 ± 0.50 mm) was statistically significant
Betharia and Kumar <sup>[14]</sup>	1987	15	Severe ( <i>n</i> = 9); mild-moderate ( <i>n</i> = 6)	Unilateral levator transection with levator aponeurosis for frontalis suspension (Neuhaus/Lemagne method)	Good correction in 10 cases. Under-correction in 5 cases
Bartkowski <i>et al.</i> <sup>[29]</sup>	1999	19	Marked ( <i>n</i> = 15)	Unilateral levator transection with levator aponeurosis for frontalis suspension (Neuhaus/Lemagne method; <i>n</i> = 16); unilateral levator transection followed by unilateral frontalis suspension ( <i>n</i> = 3)	84% patients showed no symptoms after the surgery. 1 patient had lagophthalmos
Park <i>et al.</i> <sup>[30]</sup>	2008	20	Mild-moderate ptosis	Unilateral levator resection only ( <i>n</i> = 10); frontalis muscle flap or orbicularis oculi muscle flap ( <i>n</i> = 10)	After ~30 months, blepharoptosis was corrected; however, there was only mild to moderate resolution of jaw-winking reflex
Shah <i>et al.</i> <sup>[31]</sup>	2019	23	Moderate to severe	Unilateral tarsofrontal silicone sling without levator excision	Unilateral tarsofrontal silicone sling without disinsertion or extirpation of the levator reduces the severity of symptoms in MGJWS. "good" = upper eyelid height was <1 mm, "fair" = 1-2 mm and "poor" ≥ 2 mm
Khwarz <i>et al.</i> <sup>[19]</sup>	1999	24	Minimal ( <i>n</i> = 5); moderate ( <i>n</i> = 11); severe ( <i>n</i> = 9)	Bilateral ( <i>n</i> = 19) or unilateral ( <i>n</i> = 5) levator excision, all followed by bilateral frontalis suspension	The procedure provides satisfactory correction (62% cases). But 5 patients reported recurrence
Bowyer and Sullivan <sup>[13]</sup>	2004	31	Severe ( <i>n</i> = 10); mild-moderate ( <i>n</i> = 21)	Unilateral levator advancement surgery ( <i>n</i> = 4, mild cases); bilateral levator weakening followed by bilateral frontalis suspension ( <i>n</i> = 13, moderate-severe cases)	The surgical approach will differ according to the condition. Patients with bilateral surgery had wink elimination while unilateral surgery had detectable wink
Ning <i>et al.</i> <sup>[32]</sup>	2019	42	Mild ( <i>n</i> = 7); moderate ( <i>n</i> = 24); severe ( <i>n</i> = 11)	Unilateral levator excision followed by unilateral frontalis suspension	34 patients with moderate to severe MGJWS underwent surgery and had satisfactory outcomes at 6-month follow-up
Demirci <i>et al.</i> <sup>[16]</sup>	2010	48	Mild ( <i>n</i> = 8); moderate ( <i>n</i> = 36); severe ( <i>n</i> = 4)	Unilateral levator excision followed by bilateral/unilateral frontalis suspension	The management was effective. Symptoms resolved in 97% patients and improved in 3%

Doucet and Crawford <sup>[33]</sup>	1981	55	Severe ( $n = 34$ ); mild-moderate ( $n = 21$ )	Unilateral levator disinsertion with bilateral ( $n = 2$ ) or unilateral ( $n = 12$ ) fascial suspension; Fasanella-Servat procedure ( $n = 1$ ); Motais-Parinaud procedure ( $n = 11$ ); levator excision only ( $n = 26$ ); no treatment ( $n = 17$ )	Bilateral fascial suspension had superior outcomes as compared to other treatments or no treatments
Ho <i>et al.</i> <sup>[24]</sup>	2017	8 out of 319 patients with ptosis	N/A	Levator muscle excision; frontalis suspension; frontalis muscle flap advancement (procedures not stratified by MGJWS status)	Presence of MGJWS had poorer outcomes after surgical correction for congenital ptosis
Cates and Tyers <sup>[21]</sup>	2008	7 out of 13 patients with ptosis	N/A	Bilateral frontalis brow suspension after bilateral levator excision	The researchers report satisfactory cosmetic results with good symmetry of lid movement and position
Morris <i>et al.</i> <sup>[34]</sup>	2008	7 out of 89 patients with ptosis	MRD1 > 2 mm	Silicone rod frontalis suspension	The surgery was found to be modestly effective (57% cases improved). Postoperative eyelid symmetry (< 1 mm = satisfactory)
Ibrahim <sup>[35]</sup>	2007	3 out of 8 patients with ptosis	N/A	Use of the distal portion of levator aponeurosis as a flap for frontalis suspension (similar to the Neuhaus/Lemagne method)	Synkinetic muscle movements disappeared, and hence, it is an effective treatment
Dave <i>et al.</i> <sup>[22]</sup>	2019	43 out of 95 patients	Severe ( $n = 91$ )	Frontalis sling with silicone; unilateral levator excision	Outcomes were not stratified by association with MGJWS. For all ptosis repairs, as compared to LR, FS gives a better eyelid elevation but also has greater regression requiring more surgeries
Kemp and MacAndie <sup>[23]</sup>	2001	3 out of 29	NA	Unilateral levator excision followed by bilateral Mersilene mesh brow suspension	MGJWS was associated with poorer outcomes
Neuhaus <sup>[11]</sup>	1985	1	Severe	Unilateral levator transection with distal levator muscle and aponeurosis for frontalis suspension (Neuhaus/Lemagne method)	No residual aberrant eyelid movement
Lemagne <sup>[17]</sup>	1988	1 out of 2	Severe	Unilateral levator transection with distal levator muscle and aponeurosis for frontalis suspension (Neuhaus/Lemagne method)	Synkinetic muscle movements disappeared. Moderate ptosis recurred 6 months postoperatively but was later corrected with an additional levator excision
Xiang <i>et al.</i> <sup>[36]</sup>	2010	13	Minimal ( $n = 1$ ); moderate ( $n = 7$ ); severe ( $n = 5$ )	Unilateral anastomosis of levator and frontal muscles	For moderate-to-severe MGJWS, this procedure provided generally satisfactory outcomes of both ptosis and jaw-winking
Manners <i>et al.</i> <sup>[37]</sup>	1996	28 out of 35	2-4 mm ( $n = 20$ ); 5-7 mm ( $n = 8$ )	Unilateral levator transection with distal levator muscle and aponeurosis for frontalis suspension (Neuhaus/Lemagne method)	This method was effective in eliminating jaw-winking. Ptosis often required additional levator excision to resolve

NA: not available

In studies with internal comparison of techniques, there was an apparent advantage of bilateral levator excision with bilateral frontalis suspension over other procedures. Khwarg *et al.*<sup>[19]</sup> reported that 100% of patients (19/19, including 3 who had bilateral MGJWS) who received bilateral levator excision with bilateral frontalis suspension saw significant improvement of ptosis, *vs.* only 40% of patients (2/5) who saw improvement after unilateral levator excision with bilateral frontalis suspension; across all operated eyelids, 37% (10/27) saw complete resolution of jaw-winking, while 48% (13/27) had mild residual jaw-winking. Bowyer and Sullivan<sup>[17]</sup> reported that all patients (13/13) who received bilateral levator excision with bilateral frontalis suspension had complete resolution of jaw-winking, *vs.* all 4 patients who received unilateral levator advancement who had persistent jaw-winking, despite the former group having more severe baseline MGJWS.

There was also an apparent slight advantage of unilateral levator excision with bilateral frontalis suspension over unilateral levator excision with unilateral frontalis suspension. Doucet and Crawford<sup>[33]</sup> reported

**Table 3. Types of sling materials**

Types of sling materials	Number of studies
Autogenic fascia lata	9
Muscle flaps	5
Silicone or other materials	5
None	7

**Table 4. Reports of post-surgical complications**

Post-surgical complications	Number of studies
Present	11
Absent	14

that all patients (2/2) who received bilateral frontalis suspension had complete resolution of both ptosis and jaw-winking, whereas patients who received unilateral frontalis suspension had residual moderate/severe ptosis (8%; 1/12), jaw-winking (33%; 4/12), and lid lag (100%; 12/12). They concluded that bilateral frontalis suspension is the more desirable option when possible. Demirici *et al.*<sup>[16]</sup> reported that patients who received bilateral (88%; 23/26) frontalis suspension had better upper eyelid symmetry than those who received unilateral (75%; 3/4) frontalis suspension, though this was not statistically significant.

For broader studies of ptosis that included a subset of MGJWS patients, ascertaining the efficacy of surgical intervention was hampered in some instances due to lack of reported postoperative outcomes for jaw-winking as well as for ptosis. For example, neither Dave *et al.*<sup>[22]</sup>, who assessed 95 ptosis patients, nor Ho *et al.*<sup>[24]</sup>, who assessed 319 ptosis patients, reported postoperative presence or absence of jaw-winking for MGJWS patients. Thus, for these studies, the efficacy of surgical interventions for MGJWS could not be definitively ascertained. Nevertheless, Ho *et al.*<sup>[24]</sup> found that patients with MGJWS achieved a less ideal lid height (postoperative MRD-1 < 3 mm) than in those without the condition (25% *vs.* 75.7%,  $P = 0.004$ ), while Dave *et al.*<sup>[22]</sup> did not find the presence of MGJWS to significantly affect outcome.

### Frontalis sling materials

Among articles describing the use of frontalis slings ( $n = 19$ ), the greatest number ( $n = 9$ ) used autologous materials such as tensor fascia lata, temporalis fascia, or frontalis fascia. Studies using autologous fascia lata did not report any post-surgical complications directly related to the choice of sling material. Synthetic materials such as silicone slings and tantalum wires were used in a smaller number of studies ( $n = 5$ ). Two studies reported post-surgical complications such as a sling-associated abscess or sling migration ( $n = 1$  for each). Five studies proposed the use of a muscle flap instead of a sling, using either the levator or orbicularis oculi muscle. These case studies did not report any subsequent complications [Table 3].

### Postoperative complications and recurrences

In total, there were 383 patients with MGJWS across all studies. Many of the series reported post-surgical complications [Table 4], including suture granuloma (3 patients), eyelash ptosis (12 patients), entropion (3 patients), undercorrection of ptosis (46 patients), overcorrection of ptosis (5 patients), lagophthalmos (26 patients), exposure keratopathy (22 patients), silicone sling complications (2 patients), and lid contour abnormalities (8 patients). Nonetheless, the noted number of patients with the above complications is complicated by the studies that did not specify whether the reported complications occurred in the subset of patients with MGJWS among all ptosis patients analyzed<sup>[22-24,34]</sup>.

Six studies reported recurrence of MGJWS symptoms. Carbajal<sup>[18]</sup> reported recurrence of both ptosis and jaw-winking in 2/2 patients who underwent levator plication and recurrence of ptosis in 1/1 patient who underwent the Reese procedure and 1/1 patient who underwent the Friedenwald-Guyton procedure.

**Table 5. Follow-up period**

Follow-up period	Number of studies
Less than 1 year or not reported	14
Between 1 and 5 years	9
More than 5 years	4

Khwarz *et al.*<sup>[19]</sup> reported recurrence of ptosis in 1 of 5 patients (20%) who underwent bilateral levator excision and bilateral frontalis suspension and in 3/19 patients (16%; of whom 3 had bilateral jaw-winking) who underwent bilateral levator excision and bilateral frontalis suspension. Cates and Tyers<sup>[21]</sup> reported recurrence of ptosis in 1/7 patients (14%) who received bilateral levator excision and bilateral frontalis suspension. Ho *et al.*<sup>[24]</sup> reported recurrence of ptosis in 2/8 patients (25%), although the procedures involved were not specified. Ning *et al.*<sup>[32]</sup> reported recurrence of jaw-winking in 1/34 patients (2.9%) who received unilateral levator excision and unilateral frontalis suspension. Demirci *et al.*<sup>[16]</sup> reported recurrences of ptosis in 3/30 of patients (10%) who received unilateral excision and bilateral frontalis suspension. These pooled results are similar to prior reports suggesting that the presence of MGJWS was associated with poorer surgical outcomes when compared to isolated congenital ptosis<sup>[23]</sup>. Recurrence of ptosis was more common than recurrence of jaw-winking. It is difficult to determine whether particular surgeries yielded fewer recurrences due to the limited sample size and the fact that the severity of preoperative ptosis and jaw-winking was often used as criteria for determining the type of surgery performed.

In these studies, the follow-up interval varied from two months to 16 years [Table 5]. The length of reported follow-up did not correlate with the type of surgery performed or postoperative complications recorded. In one study, although the patients were systematically followed for six months, late recurrences (e.g., 8 years) were also noted<sup>[21]</sup>.

## DISCUSSION

Surgical approaches for the management of MGJWS have historically been nuanced, with varying considerations employed for management of the ptosis, jaw-wink, and/or both. Procedures that correct only the ptosis component, such as levator plication, can potentially exaggerate the presentation of the jaw-winking. Therefore, management of MGJWS with clinically significant ptosis and jaw-winking typically involves disabling levator muscle function and suspension of the eyelid to the frontalis muscle. In this systematic review assessing reported outcomes of surgery for MGJWS, we found marked heterogeneity in management, even among cases with similar baseline clinical characteristics. Additionally, meta-analysis was challenging due to considerable differences in grading schemes for ptosis and jaw-wink as well as in reported outcome measures and follow-up intervals. Accordingly, even after thorough evaluation of the published literature, it was not possible to articulate a consensus algorithm regarding the selection of appropriate surgical technique.

Several articles<sup>[16,25,28,30,33]</sup> suggest that bilateral levator excision followed by bilateral frontalis suspension is the theoretically ideal surgical intervention for MGJWS from the perspective of achieving improvement of eyelid symmetry and jaw-wink. Nonetheless, they also acknowledge the difficulty this can present in practice, as excising a normally functioning levator on the unaffected side requires significant confidence on the part of the surgeon, and trust on the part of the patient and his or her family. Understandably, the potential ethical implications of operating on a normal eyelid and eyebrow must be carefully considered by the surgeon and weighed against potential functional and cosmetic benefit.

Along with the lack of consensus regarding choice of surgical approach, we also found considerable variability in the methodologies for reporting outcome measures. In particular, reports characterizing

ptosis and MGJWS typically describe surgical outcomes using qualitative terms such as “satisfactory”, “cosmetically acceptable”, “good symmetry”, or “improved”. In some studies<sup>[22-25]</sup>, the postoperative jaw-winking status was not mentioned. In cohort studies, we observed a greater preponderance of four objective measurements: magnitude of jaw-wink, MRD-1, levator excursion, and eyelid height. However, there remained no consistent manner by which these parameters were graded and reported [Table 6]. For future studies and case reports, we recommend including millimeter measurements of jaw-wink amplitude, MRD-1, levator excursion, and lagophthalmos in evaluations of MGJWS, with qualitative characterizations included as ancillary outcome descriptors. This will enable future post-hoc statistical analyses of outcome measures to better quantify the effectiveness of particular surgical approaches.

It is important to note that the efficacy of individual procedures may be affected by the severity of MGJWS, as this is in turn often a factor that determined the type of surgery that authors chose to perform. For example, Bowyer and Sullivan<sup>[17]</sup> performed unilateral levator advancement on patients with mild jaw-winking but bilateral levator weakening and brow suspension on patients with severe jaw-winking. In spite of this documented preference on the part of some surgeons, there was insufficient evidence on systematic review to support this as a consensus practice.

We observed that postoperative recurrence of both ptosis and jaw-wink was relatively common even in studies with limited follow-up duration, suggesting that a more structured approach to outcomes research would be beneficial for optimizing clinical results. Autologous fascia was the preferred sling material in most series, and the available evidence suggests that autogenous materials may be associated with fewer complications. The majority of case series reported only minor post-surgical complications with a limited impact on cosmetic or functional results, but heterogeneity of cohort size, reporting and analysis make it difficult to ascertain whether there are meaningful underlying lessons regarding surgical technique and sling material selection.

This review highlights findings that can be applied to clinical practice. First, we recommend that clinicians report preoperative and postoperative clinical findings, as described herein, in a quantitative, consistent manner, to enable more reliable systematic analyses. In addition, physicians should thoroughly counsel patients and families that the literature on management of MGJWS does not provide clear consensus guidelines, and that there is no clearly defined optimal approach to all cases. Furthermore, thorough discussions regarding potential complications - including the possibility for over-/undercorrection and recurrence - are critical to properly manage expectations preoperatively.

There were a number of limitations associated with this review. The primary limitation was the small sample size of 26 peer-reviewed articles (comprising 383 patients) that met inclusion criteria and the heterogenous reporting of pre-surgical metrics and outcome measures. Furthermore, in some studies with more statistical power<sup>[21,23,34]</sup> patients with MGJWS were a subset of a larger ptosis population. This analysis was a systematic review of the existing literature and therefore does not provide new prospective data. In addition, this review is limited to the peer-reviewed literature and does not describe surgical techniques that may be employed by surgeons anecdotally.

In conclusion, evidence-based lessons on the surgical management of MGJWS are limited, even when post-hoc analysis is applied to the existing literature in a systematic fashion. No clear consensus was noted, and at present, the disorder is treated according to a case-by-case approach governed by surgeon and family preference. Patients and physicians alike should be aware that recurrences in either ptosis or synkinetic jaw-winking movements are not uncommon after initial surgeries, but that in some cases, postoperative decreases in lid elevation can be corrected with subsequent interventions (such as additional levator excisions for patients who received the Neuhaus/Lemagne procedure). Future analyses may identify

**Table 6. Outcome measures reported in case series and retrospective reviews**

Author(s)	Postoperative jaw-winking	MRD-1	Upper eyelid excursion	Eyelid height
Bajaj <i>et al.</i> <sup>[15]</sup>	Resolution (< 1 mm of excursion of upper eyelid with synkinetic mouth movement) in three patients. Improvement in MGJWP (> 2 mm decrease but > 1 mm of excursion of upper eyelid with synkinetic mouth movement) in seven patients	N/A	Lid lag 1.20 ± 0.48	Postoperative lagophthalmos was 0.80 ± 0.88 mm. Amount of ptosis correction 2.40 ± 0.50
Betharia and Kumar <sup>[14]</sup>	Resolution in all patients	N/A	N/A	“Good correction” in 66.6% of patients; lagophthalmos average 2 mm (“minimal”)
Bartkowski <i>et al.</i> <sup>[29]</sup>	Resolution in 84% of patients; improvement in 31.6% of patients	N/A	N/A	68% of patients had proper width and symmetry of palpebral fissures. Remaining had marked improvement
Park <i>et al.</i> <sup>[30]</sup>	Moderate degree of residual jaw-winking	N/A	N/A	“Ideal”
Shah <i>et al.</i> <sup>[31]</sup>	Resolution in 39.1% of patients, “improvement” in 47.8% of patients, and no improvement in 13.04% of patients	From -1.13 ± 0.916 to 3.17 ± 0.865 mm	N/A	Ptosis correction was “good” in 65.21% of patients and “fair” in 26.08% of patients
Khwarg <i>et al.</i> <sup>[19]</sup>	Resolution in 37.0% of patients, mild winking (1 mm or less) on the lateral jaw movement in 48.2% of patients	For bilateral frontalis suspension with unilateral levator excision: 40% of patients had “good” and “60%” had poor results. For bilateral frontalis suspension with bilateral levator excision: 68.4% of patients had good and 31.6% had fair results	Only initial excursion was recorded: poor (≤ 4 mm) in 22.2% of eyelids, fair (5-7 mm) in 19%, and good (8 mm or more) in 59%	N/A
Bowyer and Sullivan <sup>[13]</sup>	Unilateral levator advancement: persistent. Bilateral levator weakening with bilateral frontalis suspension: resolution	N/A	Only initial excursion was recorded: poor (≤ 4 mm) in 7% of eyelids, fair (5-7 mm) in 25%, and good (8 mm or more) in 68%	Improved
Ning <i>et al.</i> <sup>[32]</sup>	Resolution in all patients	From -0.57 mm to 2.96 ± 0.48 mm.	Preoperative mean excursion was 3.69 ± 1.09 mm	Postoperative palpebral fissure height of the operated eye in primary gaze was 7.93 ± 0.58 mm, with no significant difference to the unaffected side
Demirici <i>et al.</i> <sup>[16]</sup>	Resolved in 97% of patients, improved (from 6 mm to 2 mm) in 3%	N/A	From 7.5 ± 2.3 mm for the ptotic eyelid and 13 ± 2.2 mm for the normal eyelid, to 3.2 ± 0.9 mm for the ptotic eyelid and 10.3 ± 3.3 mm for the opposite normal eyelid	Upper eyelid margin distance from 0.1 ± 1.8 to 1.1 ± 0.9 mm
Doucet and Crawford <sup>[33]</sup>	For bilateral suspension: resolution. For unilateral suspension: no improvement	N/A	N/A	Moderate/severe ptosis in 37.5% of patients with levator muscle excision and 8.3% of patients with unilateral fascial suspension
Ho <i>et al.</i> <sup>[24]</sup>	N/A	Final MRD-1 > 3 mm in 25% of patients	N/A	Lid height determined by MRD-1

Cates and Tyers <sup>[21]</sup>	Resolution in 85.7% of patients	N/A	N/A	Successful lid height (within 1 mm of desired height) in 85.7% of patients
Morris <i>et al.</i> <sup>[34]</sup>	NA	Mean MRD from 0 to 2 mm; satisfactory MRD-1 elevation in 85.7% pts	N/A	57% of patients had satisfactory symmetry (< 1 mm difference between eyelid height)
Ibrahim <sup>[35]</sup>	Resolution in all patients	Mean MRD from 3.6 mm to 5.3 mm	N/A	N/A
Dave <i>et al.</i> <sup>[22]</sup>	N/A (results not stratified by MGJWS status)	N/A	NA	N/A
Xiang <i>et al.</i> <sup>[36]</sup>	Resolution in all patients	N/A	N/A	77% of patients had equal palpebral apertures, 23% had mild residual ptosis
Manners <i>et al.</i> <sup>[37]</sup>	Resolution in 93% of patients	N/A	N/A	62% of patients had improved ptosis, 21% had no change in ptosis, and 14% had worsened ptosis

whether particular surgical approaches are more appropriate, effective, and safe for particular clinical scenarios.

## DECLARATIONS

### Authors' contributions

Made substantial contributions to conception and design of the study, performed data analysis and interpretation, and wrote the manuscript: Bair H, Garcia GA, Erickson BP

### Availability of data and materials

Not applicable.

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### Conflicts of interest

All authors declared that there are no conflicts of interest.

### Ethical approval and consent to participate

Not applicable.

### Consent for publication

Not applicable.

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