

# Brown syndrome associated with Marcus-Gunn jaw winking ptosis

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## Abstract

Brown syndrome is a rare mechanical disorder characterized by restriction of the superior oblique trochlea-tendon complex. Marcus-Gunn jaw winking ptosis is a more common congenital oculofacial synkinesis in which blepharoptosis is associated with upper eyelid contraction that accompanies jaw movement. In this report, we present the case of a 50-year-old woman with unilateral Brown syndrome and Marcus-Gunn jaw winking ptosis in the fellow eye.

## Introduction

Brown Syndrome, first reported in 1950, is a rare mechanical disorder characterized by restriction of the superior oblique trochlea-tendon complex; it may be congenital or acquired, secondary to trauma or inflammation. Clinical signs, which may be constant or intermittent, include deficiency of elevation in adduction, and in moderate to severe disease, downshoot of the eye occurs in adduction. The vast majority of cases, approximately 90%, are unilateral.<sup>1</sup>

Marcus-Gunn jaw winking ptosis, also known as the jaw-winking phenomenon or Marcus-Gunn Syndrome, first reported in 1883, is a more common congenital oculofacial synkinesis in which blepharoptosis is associated with upper eyelid contraction that accompanies jaw movement. As with Brown Syndrome, the vast majority of cases of Marcus-

Gunn jaw winking ptosis are unilateral.

Approximately one-half of all cases of Marcus-Gunn are associated with strabismus: superior rectus palsy in one-quarter of cases and a double elevator palsy on another one-quarter of cases.<sup>2-4</sup>

## Case Report

A 50-year-old woman with a history of Brown Syndrome, without any current visual complaints, presented for routine strabismus evaluation. The patient was diagnosed with Brown Syndrome in her right eye at age 7, when she was found to have active and passive restriction of elevation in adduction in the right eye, with divergence in upgaze, downshoot in adduction, and hypotropia in primary position with a compensatory chin elevation. Her family history is unknown. At age 20, she underwent surgical recession of superior oblique tendon of the right eye, twice.

Now, at age 50, on examination, her visual acuity was 20/30 and 20/25 in the right and left eyes, respectively, with -2.50 Diopter spherical correction in each eye. External examination demonstrated a head tilt to the left; she had mild blepharoptosis of the left upper eyelid. Hirschberg corneal reflex testing demonstrated 18 prism diopters of extotropia and 12 prism diopters of right hypotropia with left eye fixation, at distance, and 20 prism diopters of extotropia and 6 prism diopters of right hypotropia with left eye fixation, at near. The unilateral cover test

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demonstrated the capacity of the patient to maintain fixation with the right eye, confirming her ability to alternate fixation with both eyes. Prism cover testing confirmed 18 prism diopters of extotropia and 12 prism diopters of right hypotropia with left eye fixation, at distance, though 30 prism diopters of extotropia and 10 prism diopters of right hypotropia with left eye fixation, at near. Worth four dot testing revealed alternating suppression, although fixation was mostly observed with the left eye. During the red filter test, the patient complained of transitory diplopia only at the moment of the switch of the fixing eye. Extraocular motility testing demonstrated a mild downshoot of the right eye in adduction, with bilateral medial rectus hypofunction. Ipsilateral superior oblique hyperfunction was absent.

During the orthoptic examination, left upper eyelid contraction was incidentally observed while the patient was speaking. A directed evaluation was then performed: the patient to open her mouth, and upon doing so, the left upper eyelid retracted concurrently by approximately 3 mm (Figure 1). When her mouth was closed, the left eyelid returned to its normal position (Figure 2). The patient made note that she had been experiencing this peculiar phenomenon for many years, but she had never complained about it as she was not bothered by the eyelid retraction.



**Figure 1**  
Approximately 3 mm of left upper eyelid retraction is present when the patient's mouth is open.

## Discussion

Brown Syndrome and Marcus-Gunn jaw winking ptosis are rare ocular conditions. Brown Syndrome is the result of restriction of passage of the superior oblique muscle tendon and sheath through the pulley-like trochlea.<sup>1</sup> While it may be secondary to trauma or inflammation, it is often congenital, as in this case. The Marcus-Gunn jaw winking ptosis is the result of aberrant nerve connections, specifically a co-innervation between the elevator palpebrae superior muscle from the oculomotor nucleus and external pterygoid portion of the trigeminal nucleus.<sup>2,5</sup> The Marcus-Gunn jaw winking ptosis may also be acquired in rare cases, but is most often congenital. In this case, it is uncertain whether the patient's condition was acquired or congenital.

A high incidence of strabismus and amblyopia have been associated with the Marcus-Gunn jaw winking ptosis.<sup>5</sup> Amblyopia has been reported in just over one-half (59%) of cases, and strabismus in approximately one-half of cases: superior rectus palsy in one-quarter of cases and a double elevator palsy on another one-quarter of cases.<sup>2-6</sup> Other ocular associations have been reported in rare cases, such as synergistic divergence<sup>7</sup> and Duane's retraction syndrome.<sup>8</sup> This case presented is rare, as we are aware



**Figure 2**  
Upon closing her mouth, the left upper eyelid retraction resolves and the eyelid returns to its normal position.

of only two other cases of Brown Syndrome associated with Marcus-Gunn jaw winking ptosis reported in the referenced medical literature: Sztetter, et al. in 1968<sup>9</sup> and Artifoni, et al. in 1965.<sup>10</sup>

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