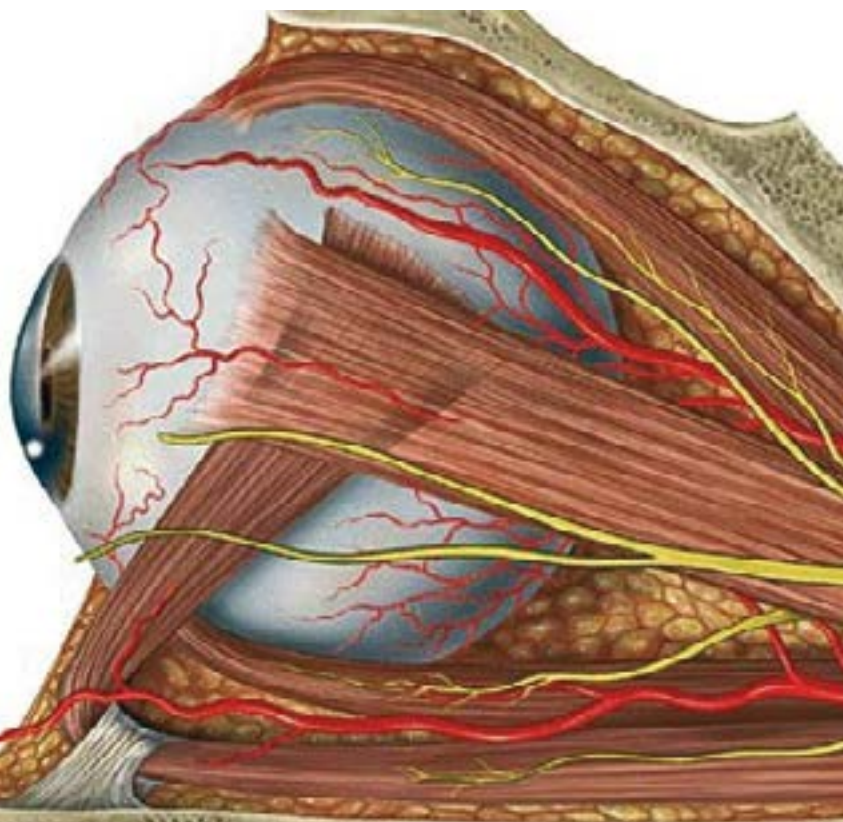


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Marcus Gunn's jaw
winking phenomenon

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Following an uncomplicated pregnancy and delivery, this female infant was seen for ptosis of the right eyelid. Apart from her ptosis, physical examination was unremarkable and she was dismissed with the diagnosis peripheral facial nerve paresis. At the age of 3 months, her physician referred her again for investigation of an unclear neurological disorder affecting her face with unilateral ptosis. Family history was unrevealing.

The girl was in good general condition, her weight was 4.83 kg (P25), her length was 58 cm (P25), and her head circumference was 40 cm (P50-75). There was a plagiocephalus on the left side without restrictions in the movement of the cervical spine. Her psychomotor development was normal. When the girl was calm on her mother's lap, there was ptosis of her right eye. At the moment the girl started to breastfeed or suck on her pacifier, the ptotic lid jerked upward with each sucking movement (see Fig. A and B). In contrast, the movements of the left eyelid were coordinated with the movements of the left eye independent of the sucking movements.

The girl was able to fixate and follow with normal eye movements in all directions. Pupillary reactions were normal. There was no nystagmus or strabismus and no evidence of amblyopia. Based on the typical findings a diagnosis of Gunn's jaw winking phenomenon (trigemino-oculomotor synkinesis) was made. The patient

was seen again at the age of 6 months; at this time, the synkinetic movements had already diminished.

Rhythmic movement of the affected right upper eyelid is easily recognizable during bottle feeding (age 3 months).

Fig. A



*Note: movies can be accessed at
www.neonet.ch/case-2007-12*



Fig. B

DISCUSSION

This phenomenon was first described in 1883 (1) by the British ophthalmologist Robert Marcus Gunn (1850–1909). Synkinesis means the simultaneous movement of muscles normally innervated through different nerves or different branches of the same nerve. The term pathologic synkinesis is used when muscles are innervated through nerves other than their own. This can occur congenitally or after an injury. In Marcus Gunn's syndrome the eyelid is elevated with mouth opening and jaw movement towards the contralateral side (ipsilateral external pterygoid firing) or jaw protrusion (bilateral external pterygoid firing), chewing, sucking or swallowing or rarely by clenching of the teeth (internal pterygoid-levator synkinesis). It is due to aberrant innervation of the levator palpebrae muscle by a branch of the motor division of the trigeminal nerve that supplies the muscles of mastication. Interestingly, there is also an inverse Marcus Gunn's phenomenon: with opening of the mouth or different jaw movements the affected eyelid closes (inhibition of the oculomotor branch).

Marcus Gunn's syndrome is associated with strabismus (50-60%), ansiometropia (25%) and amblyopia (30-50%). The latter is due to strabismus or anisometropia, and only rarely due to ocular occlusion by the ptotic eyelid. Other associated malformations are very rare (spina bifida occulta, ectrodactyly, pes cavus, cryptorchidism, dental enamel defects).

Marcus Gunn's phenomenon is almost always sporadic

with an irregular autosomal dominant inheritance pattern. There is no racial or gender preference. Different forms of aberrant innervations have been reported with thalidomide embryopathy. It is the most common congenital synkinesis with a prevalence of 2- 5 % in patient with congenital ptosis.

Patients with this diagnosis must be referred to an ophthalmologist for assessment of amblyopia and/or anisometropia. They may need surgical interventions to correct severe ptosis. Most children learn to suppress the synkinetic movements, and the condition is often hard to identify in adults.

1. Gunn RM. Congenital ptosis with peculiar associated movements of the affected lid. Transactions of the Ophthalmological Societies of the United Kingdom, London 1883;3:283-287

REFERENCES

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