et al. proposed that in acute zonal occult outer retinopathy viral introduction in the retina may occur at the optic nerve margin and ora ser- rata because of the absence of sur- rounding neuroepithelium at these sites. The initial manifestation of transient discrete peripapillary outer retina or retinal pigment epithelium involvement centered around the optic nerve in our patient would be consistent with such a hypothe- sis. The funduscopic appearance subsequently evolved into more typical MEWDS findings, including deep retinal white dots surrounding the paramacular area with foveal sparing, blurring of the disc margin, and vascular involvement at angiography. The patient’s subjective complaints of photopsia and temporal scotoma and the blind spot enlargement at visual field testing are all consistent with the diagnosis of MEWDS. Because of their transient nature, these peri- papillary findings may be more common in MEWDS than previously appreciated.

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Bilaterial Congenital Trigemin-abducens Synkinesis

To our knowledge, congenital bilateral trigemino-abducens synkinesis has not previously been re- ported. Herein we report a case in which ipsilateral jaw movement oc- curred.

Report of a Case. A 21-year-old man had ipsilateral jaw movement asso- ciated with abduction of either eye. The patient had been aware of this phenomenon since early childhood. He was a term infant with a normal vaginal delivery and had no dysmorphic features or developmental delay. There was neither history of orbital or ocular trauma, strabismus, or amblyopia nor family history of a similar phenomenon. On examina- tion, his visual acuity was 6/6 OU and his pupils were 3 mm equal and re- active to light and accommodation. Extraocular movements were full with no nystagmus or widening or narrowing of the palpebral fissures in any position of gaze. On abduction to the left or right, subtle ipsilateral jaw movement was noted(Figure). Findings from neurological examination were normal and electomyographic study demonstrated that abduction of the eyes produced ipsilateral mas- seter and pterygoid activity.

Comments. Freedman and Kush- ner documented that congenital ocular aberrant innervations are more common than previously de- scribed and exist in many varieties. Ocular “miswirings” typically in- volve the sixth nerve and the most recognized clinical entities are Du- ande syndrome and Marcus Gunn jaw-winking ptosis.

We believe we have now pre- sented the first case of congenital bi- lateral trigemino-abducens synkine- sis. Furthermore, ocular abduction producing ipsilateral masseter and pterygoid activity is contrary to pre-
viously reported cases in which jaw movement produced abduction of the eye. Kodsi described a 4-year-old girl who exhibited a right intermittent exotropia with mouth opening and chewing movement from birth and McGovern et al documented a case of acquired right trigemino-abducens synkinesis following a major head trauma from a motor vehicle crash.

Freedman and Kushner hypothesized that trigemino-abducens synkinesis may be due to abnormal synkinesis between the abducens nerve controlling the lateral rectus muscle and the mandibular division of the trigeminal nerve controlling muscles of mastication, including the masseter and pterygoid muscles. The etiology of congenital aberrant innervations involving the nucleus of the abducens nerve may be due to abnormal development and degeneration of the abducens nerve fibers during embryologic development that leads to central or peripheral miswiring. The exact reasons for the common involvement of the abducens nerve and nucleus with aberrant innervation are unknown. The abducens nerve fibers and nucleus develop earlier than the fibers and nuclei of the oculomotor and trochlear nerves, and subsequent degeneration of the caudal branches may lead to the development of abnormal abducens nerve connections, either in the brainstem or peripherally. An abnormal wiring pattern of abducens nerve fibers to the adjacent trigeminal nerves and nuclei in the brainstem could account for the clinical findings of trigemino-abducens synkinesis. On the other hand, miswiring may also result from a peripheral cause. Embryonic nerves are attracted to developing mesenchyme and muscle masses during development and inappropriate attraction of a cranial nerve branch to an extraocular muscle may lead to aberrant ocular innervation.

Our case further supports the findings of Freedman and Kushner that congenital ocular aberrant innervation may be more common than previously reported and highlights the importance of diagnosing bilateral trigemino-abducens synkinesis and its many varieties.

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