

## Congenital third nerve palsy with synergistic depression on attempted adduction and trigemino-oculomotor synkinesis: Underpinnings of a spectral dysinnervation disorder

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The authors describe a case of congenital partial pupil-sparing third cranial nerve palsy with absent adduction, synergistic depression of globe and widening of palpebral fissure on attempted adduction and synergistic elevation and adduction on mouth opening and sideways thrusting of jaw. The case illustrates trigemino-oculomotor synkinesis associated with congenital third nerve palsy. The possible mechanism of miswiring involving the medial longitudinal fasciculus and trigeminal nuclei is discussed. At least some cases of congenital third cranial nerve palsy may fall in the realm of congenital cranial dysinnervation disorders (CCDDs) sharing a much wider spectrum of presentation.

**Key words:** Congenital cranial dysinnervation disorder, congenital third nerve palsy, synergistic depression, trigemino-oculomotor synkinesis

We report a case of absent adduction with unilateral partial pupil sparing congenital third cranial nerve palsy without ptosis manifesting as synergistic depression of globe and widening of the palpebral aperture on attempted adduction. Jaw movements led to synergistic elevation and adduction of globe suggesting trigemino-oculomotor synkinesis, riding piggyback on congenital third nerve palsy.

### Case Report

A 7-year-old boy presented with widened lid fissure, constant exotropia (XT) and inability to adduct, elevate, or depress his

right eye since birth. Family history was unremarkable. There were no dysmorphic features. Unaided visual acuity was 20/20 OU with a cycloplegic retinoscopy of +2.5 dioptres. He displayed a large constant XT of >100 prism dioptres OD, fixing OS. Fixing OD, he adopted a face turn to the left and elevated his chin [Fig. 1a]. There was -3 limitation of depression, -4 of elevation and adduction OD on versions and ductions [Fig. 1b]. Convergence was absent OD. On levo-version/attempted adduction OD, there was synergistic depression with widening of palpebral aperture [Video 1]. Ocular motility OS was unremarkable.

On the opening of mouth, eye elevated and adducted, no globe retraction was noted. Sideways thrusting of the jaw produced similar findings [Video 1]. Magnetic resonance imaging brain and orbits was unremarkable except for mildly hypoplastic medial rectus OD. Linkage analysis and electromyography recordings were not done. A 12 mm recession of tight lateral rectus OD did not significantly alter alignment or improve adduction or synergistic depression.

### Discussion

Synergistic divergence with deficient/absent adduction has been recognized as a distinct ocular motility dysinnervation pattern, often related to Duane's retraction syndrome and congenital fibrosis of extraocular muscles.<sup>[1-3]</sup> Anecdotal reports have described unique dysinnervational patterns such as supraduction on attempted adduction and infraduction on attempted abduction as part of the spectrum of congenital cranial dysinnervation disorders (CCDDs).<sup>[4]</sup> Synergistic depression on attempted adduction has not been described with congenital third nerve palsy. Trigemino-oculomotor synkinesis manifesting globe movements associated with jaw movements have been described anecdotally with or without Marcus Gunn phenomenon.<sup>[5,6]</sup> Synkinetic globe and jaw movements have not been described with congenital third nerve palsy.

As there was no recovery of function for adduction, no aberrant innervation of oculomotor nerve can be factored in to account for widening of the palpebral fissure or synergistic depression on attempted adduction. Widening of palpebral fissure could be due to weaker superior rectus with overflow of innervations to levator palpebrae superioris. Synergistic depression on attempted adduction has not been described with aberrant innervations of third cranial nerve palsy.

Oculomotor miswiring was gaze induced, likely at the level of medial and inferior rectus subnuclei in the third cranial nerve nucleus. Miswiring was likely routed through medial

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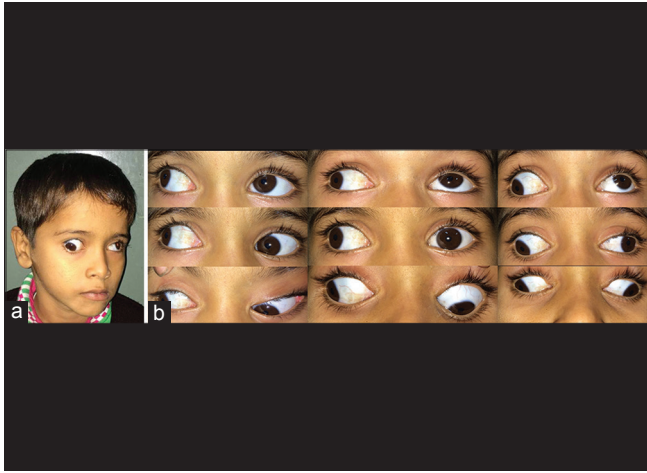
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**Figure 1:** (a) Anomalous head posture with face turn to the left and chin elevation, fixing OD. (b) Versions demonstrating large exotropia OD (fixing OS), with absent adduction OD, limited elevation and depression OD, synergistic depression and widening of palpebral aperture on attempted adduction OD

longitudinal fasciculus (MLF) as attempted convergence failed to elicit it. Large recession of tight lateral rectus hardly ameliorated the clinical picture stressing that it was not the culprit.

Trigemino-oculomotor miswiring in the setting of congenital third cranial nerve palsy remains speculative. Proprioceptive afferents from extraocular muscles synapse with branches of trigeminal nerve and may be relayed to its mesencephalic nucleus. The nucleus integrates monosynaptic reflexes for proprioception.<sup>[7]</sup> Motor nucleus of the fifth nerve has connections to mesencephalic nucleus and integrates chewing and salivation. Neural pathways known to exist between the masticatory mesencephalic trigeminal nucleus neurons and the oculomotor system may explain the occurrence of trigemino-oculomotor synkinesis.<sup>[8]</sup>

The present case showcases that at least some of the congenital third cranial nerve palsies may fall into the realm of CCDDs sharing much wider spectrum of presentation with involvement of the MLF as well as nuclei of trigeminal nerve.

### Declaration of patient consent

The authors certify that they have obtained all appropriate patient consent forms. In the form the patient(s) has/have given his/her/their consent for his/her/their images and other clinical information to be reported in the journal. The patients understand that their names and initials will not be published and due efforts will be made to conceal their identity, but anonymity cannot be guaranteed.

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### Conflicts of interest

There are no conflicts of interest.

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