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Teaching Video NeuroImages: Bilateral complete horizontal gaze palsy with preserved convergence

The 1 + 1 syndrome

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Bilateral complete horizontal gaze palsy, the 1 + 1 syndrome, is extremely rare. An otherwise healthy 25-year-old woman shows a recently acquired complete horizontal saccadic palsy. Its association with the absence of improvement with oculocephalic maneuvers and the preservation of convergence clinically suggests a bilateral cranial nerve VI nucleus dysfunction[1,2] (video at Neurology.org; figures 1 and 2). Main causes include CNS inflammation, metabolic/toxic disorders, and regional expansive lesions. Mimickers such as myasthenia gravis or Fisher syndrome and anti-GQ1b-mediated disorders have to be meticulously excluded.

Despite exhaustive investigations, including repeated MRI, the cause remains unknown in this patient and no changes were observed over years.

AUTHOR CONTRIBUTIONS

D. Biotti: first author, corresponding author, principal investigator, neuro-ophthalmologic management. C. Tilikete: data collection, neuro-ophthalmologic management.

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DISCLOSURE

The authors report no disclosures relevant to the manuscript. Go to Neurology.org for full disclosures.

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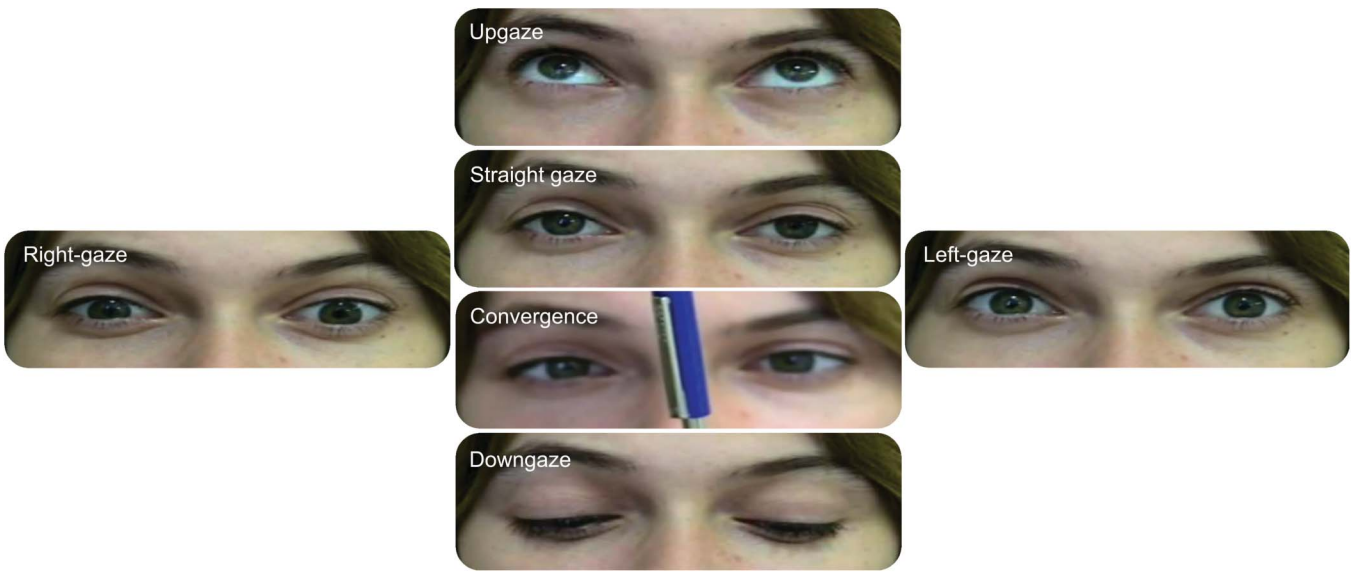
2. Leigh R, Zee D. Disorders of ocular motility due to disease of the brainstem, cerebellum, and diencephalon. In: The Neurology of Eye Movements, 5th ed. Oxford: Oxford University Press; 2015.

Figure 1 Ocular motor examination



Supplemental data at Neurology.org

Download teaching slides: Neurology.org

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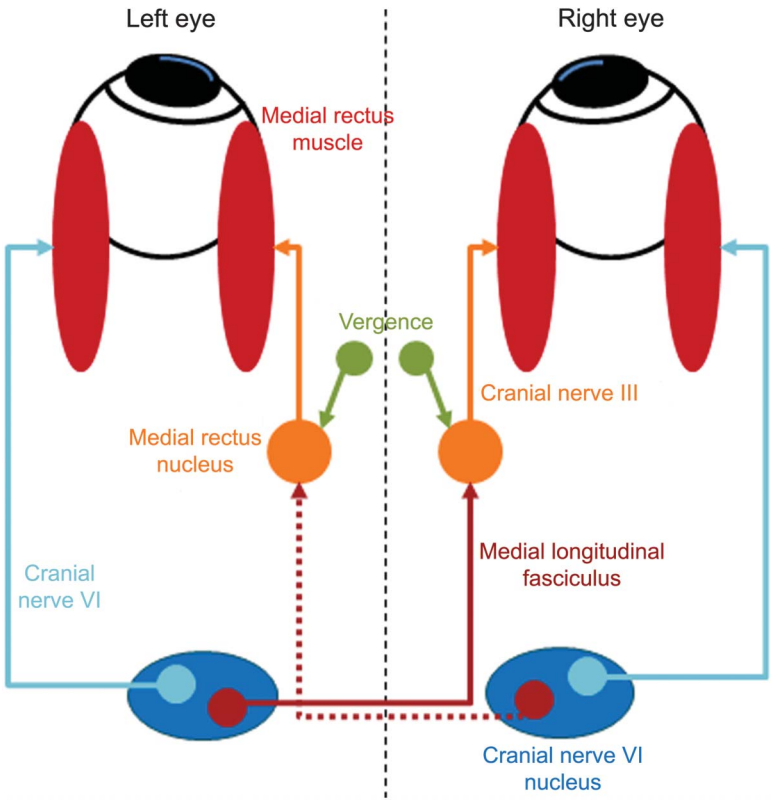
Figure 2 Anatomical scheme of nuclear and infranuclear horizontal gaze pathways



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