VIDEO Ptosis in myasthenia gravis: Extended fatigue and recovery bedside test

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Fluctuating double vision and ptosis are the hallmarks of extraocular muscle weakness in myasthenia gravis (MG). On sustained upward gaze, ptosis usually increases temporarily. The first description of using this fatigue sign has been attributed to the Scottish neurologist John Simpson. Cogan added his lid-twitch sign to the diagnostic bedside armamentarium.¹ For many years, we have applied a fatigue-recovery test at the bedside, whereby the strength of the previously fatigued levator palpebrae muscle is improved shortly after voluntary maximal contraction of the antagonistic orbicularis oculi muscles, leading to a temporary improvement of lid opening. This extended fatigue maneuver provides an additional hint to the typical myasthenic weakness, in particular in the absence of double vision. Furthermore, we often observed that Cogan's lid-twitch sign may only be elicited when provoking recovery. This short-lived lid twitch is supposed to reflect the temporary recovery of phasic more than tonic motor units in the levator palpebrae muscles. Recovery of ptosis after 3 minutes of lid closure has been mentioned earlier by the late

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Myasthenia – Fatigue and Recovery Test 'Simpson plus'

Figure. (A) A 61-year-old woman with marked generalized myasthenia gravis, 2 days after plasma exchange treatment, with only minimal spontaneous ptosis at baseline. (B) After 10 seconds of upward gaze. (C) After 30 seconds of upward gaze. (D) Maximal voluntary lid closure for 10 seconds. (E) Almost complete recovery upon lid opening. (F) Reoccurrence of ptosis after another 10 seconds of upward gaze. Note that the patient has no heterotropia and did not report double vision at time of testing.

Dutch neurologist Hans Oosterhuis.² However, the recovery test presented here requires only 10 to 30 seconds of maximal voluntary eye closure. In the differential diagnosis of pure ocular MG, the test proves particularly helpful in differentiating MG from other common disorders causing ptosis, e.g., Horner syndrome without miosis, mild forms of progressive external ophthalmoplegia, or habitual ptosis of the elderly where fatigue on upward gaze and re-

covery after lid closure is less pronounced or absent (figure).

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See also pages 1333, 1521–1523, 1525

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