

Sir,
Congenital ptosis associated with combined superior rectus, lateral rectus, and levator palpebrae synkinesis: the first reported case

Superior rectus (SR) to levator palpebrae superioris (LPS) synkinesis has recently been described in patients with congenital or longstanding ptosis.^{1,2} However, aberrant innervation between lateral rectus (LR) and LPS has not been previously reported. We report a combined SR and LR to LPS synkinesis in a patient with congenital ptosis.

Case report

A 22-year-old man presented for ptosis assessment in his left eye. The ophthalmic history referred to a previous unsuccessful operation for correction of his unilateral, congenital ptosis. No history of squint or trauma was reported. His medical history was unremarkable.

The patient displayed a marked ptosis, more noticeable in the temporal half of upper lid. Ptosis assessment measurements were palpebral aperture (PA) = 7 mm, levator function (LF) (two-phase procedure as suggested by Jones and colleagues³): on primary position: phase 1 = 4 mm, phase 2 = 8 mm (total = 12 mm), abduction: phase 1 = 4 mm, phase 2 = 8 mm, while on adduction: phase 1 = 4 mm and phase 2 = 4 mm (total 8 mm). Skin crease height (SC) = 8 mm, symmetrical to contralateral eyelid. Bell's phenomenon was moderately weak, while 1–2 mm lagophthalmos was apparent. Cover test and eye movements were normal.

A complete resolve of ptosis was noted on up-gaze (PA = 11 mm) as well as on abduction (PA = 11.5 mm; Figure 1). Slit lamp examination was normal, while patient's visual acuity was 6/6 OU.

The patient underwent a large levator resection, as indicated in cases with simple SR to LPS synkinesis, with satisfactory results. The surgery resulted in 4 mm lagophthalmos, which progressively improved to 2–3 mm. At 4 weeks post-op, PA was 10.5 mm on primary, 14 mm on up-gaze, and 11.5 mm on abduction, while symmetry between fellow eyes was achieved. (Figure 2).

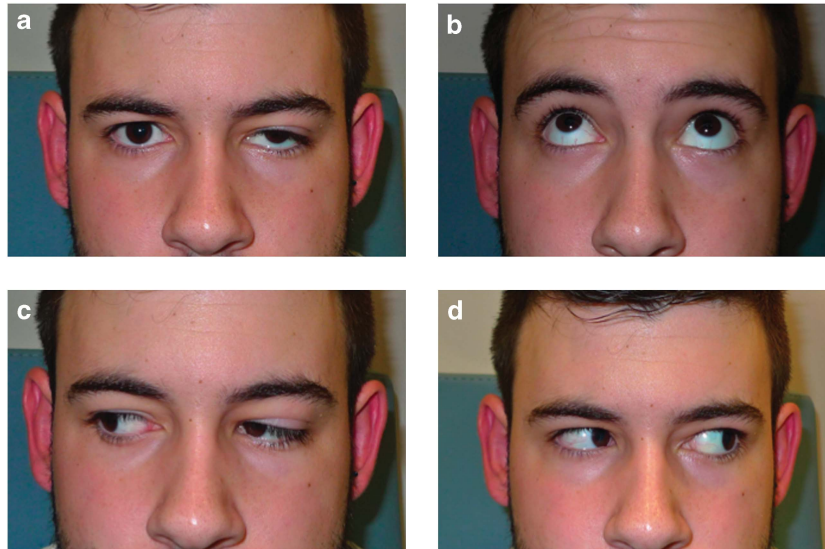


Figure 1 Pre-operative photographs depicting the eyelids on four positions of gaze: (a) primary: note the marked ptosis on left side, (b) up-gaze: complete resolve of ptosis is noted, (c) adduction: ptosis becomes apparent, and (d) abduction: complete resolve of ptosis.

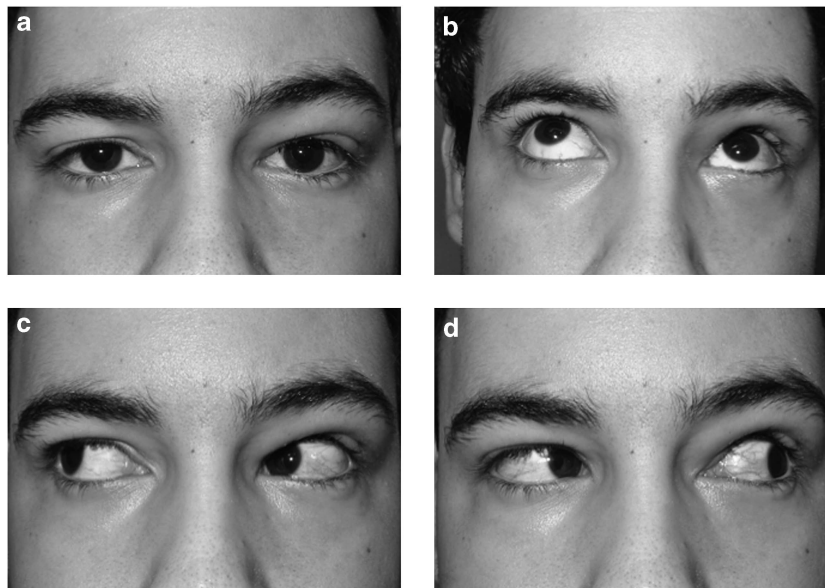


Figure 2 Early post-operative photographs on same four positions of gaze: (a) primary: successful ptosis correction is shown despite the post-op tissue oedema, (b) up-gaze: slight overcorrection of ptosis compared to fellow eye, (c) adduction: symmetrical elevation is noted, and (d) abduction: mild overcorrection is apparent.

Comment

Synkinetic innervation between muscles nerved by 3rd cranial nerve has been described as a result of acquired or congenital palsies. SR to LSP synkinesis is considered a poor prognostic factor affecting ptosis surgery and therefore a new method of ptosis assessment has been proposed.³ As the neurogenic dysfunction along the course of 3rd nerve seems to play a major role in LPS weakness, the phenomenon should always be sought in this group of patients and if apparent, a larger than usual LPS resection is recommended.¹

However, and to our knowledge, synkinesis between SR, LSP, and LR has never been reported. This pattern of aberrant innervation involves 3rd and 6th nerve simultaneously and represents an addition to the range of congenital cranial dysinnervation disorders.⁴

Conflict of interest

The authors declare no conflict of interest.

References

- 1 Harrad RA, Shuttleworth GN. Superior rectus-levator synkinesis: a previously unrecognized cause of failure of ptosis surgery. *Ophthalmology* 2000; **107**(11): 1975–1981.
- 2 McMullan TFW, Robinson DO, Tyers AG. Towards an understanding of congenital ptosis. *Orbit* 2006; **25**(3): 179–184.
- 3 Jones CA, Lee EJ, Sparrow JM, Harrad RA. Levator function revisited: a two-phase assessment of lid movement to better identify levator-superior rectus synkinesis. *Br J Ophthalmol* 2010; **94**(2): 229–232.
- 4 Oystrek DT, Engle E, Bosley TM. Recent progress in understanding congenital cranial dysinnervation disorders. *J Neuroophthalmol* 2011; **31**(1): 69–77.

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