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Rectus Pulley Displacements without Abnormal Oblique Contractility Explain Strabismus in Superior Oblique Palsy

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Purpose: Using high-resolution magnetic resonance imaging (MRI), we investigated whether rectus pulleys are significantly displaced in superior oblique (SO) palsy and whether displacements account for strabismus patterns.

Design: Prospective case-control study.

Participants: Twenty-four patients diagnosed with SO palsy based on atrophy of the SO muscle on MRI and 19 age-matched orthotropic control subjects.

Methods: High-resolution, surface coil MRI scans were obtained in multiple, contiguous, quasicoronal planes during monocular central gaze fixation. Pulley locations in oculocentric coordinates in the following subgroups of patients with SO palsy were compared with normal results in subgroups of patients with SO palsy: unilateral versus bilateral, congenital versus acquired, and isotropic (round) versus anisotropic (elongated) SO atrophy. Expected effects of pulley displacements were modeled using Orbit 1.8 (Eidactics, San Francisco, CA) computational simulation.

Main Outcome Measures: Rectus pulley positions and ocular torsion.

Results: Rectus pulleys typically were displaced in SO palsy. In unilateral SO palsy, on average the medial rectus (MR) pulley was displaced 1.1 mm superiorly, the superior rectus (SR) pulley was displaced 0.8 mm temporally, and the inferior rectus (IR) pulley was displaced 0.6 mm superiorly and 0.9 mm nasally from normal. Displacements were similar in bilateral SO palsy, with the SR pulley additionally displaced 0.9 mm superiorly. However, the lateral rectus pulley was not displaced in either unilateral or bilateral SO palsy. The SR and MR pulleys were displaced in congenital SO palsy, whereas the IR and MR pulleys were displaced in acquired palsy. Pulley positions did not differ between isotropic and anisotropic palsy or between patients with cyclotropia of less than 7° versus cyclotropia of 7° or more. Simulations predicted that the observed pulley displacements alone could cause patterns of incomitant strabismus typical of SO palsy, without requiring any abnormality of SO or inferior oblique strength.

Conclusions: Rectus pulley displacements alone, without abnormal oblique muscle contractility, can create the clinical patterns of incomitant strabismus in SO palsy. This finding supports accumulating evidence that clinical binocular misalignment patterns are not reliable indicators of contractile function of the SO muscle. Ocular torsion does not correlate with and thus cannot account for pulley displacements in SO palsy. *Ophthalmology 2016;■:1–10 © 2016 by the American Academy of Ophthalmology.*

The directions of forces exerted by the extraocular muscles (EOMs) are determined by the connective tissue pulleys through which the EOMs transit; thus, EOM force vectors are determined by pulley positions.[1–4] Although magnetic resonance imaging (MRI) has shown that pulley positions are quite uniform among normal people,[2,5] abnormalities of rectus pulley positions are associated consistently with incomitant strabismus that may simulate oblique EOM dysfunction.[6,7] For example, inferolateral displacement of the lateral rectus (LR) pulley caused by connective tissue involution in sagging eye syndrome was recognized recently as a cause of age-related distance esotropia and hypertropia in older people.[8]

Rectus pulley positions in the coronal plane normally are subject to the actions of the orbital layers of the oblique EOMs and consequently shift by modest amounts under physiologic conditions.[9–11] For example, the array of the 4 rectus pulleys rotates in torsional fashion around the long axis of the orbit during ocular counter-rolling[12] and during convergence,[13] paralleling the direction and amount of the corresponding physiologic ocular torsion. It seems plausible that cyclovertical strabismus, commonly associated with oblique EOM dysfunction, may alter rectus pulley positions. An MRI study demonstrated abnormal pulley shifts during head tilt in subjects with head tilt–dependent hypertropia both with and without superior oblique (SO) muscle atrophy.[14] Unlike physiologic ocular counter-rolling, the LR and inferior rectus (IR) pulleys paradoxically shifted into intorted positions in orbits having SO atrophy. In head tilt–dependent hypertropia without SO atrophy, the LR, medial rectus (MR), and superior rectus (SR) pulleys in the hypertropic orbit

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exhibited reduced or reversed extorsional shift during head tilt. These differences in pathologic pulley shifts suggest that pulleys may contribute to the development of head tilt–dependent hypertropia and that this contribution may be influenced by the presence of SO atrophy.

By both histologic and MRI criteria, a monkey model of SO palsy produced by intracranial trochlear neurectomy confirmed that SO denervation reduces the maximum SO cross-section to approximately half of normal and does so within 5 weeks.[15–18] Because comparable SO atrophy and loss of contractile enlargement on infraduction are unequivocal evidence of human SO palsy, the MRI finding of SO atrophy can be regarded as a sufficient (albeit perhaps not a necessary) objective confirmation of the diagnosis of SO palsy.[14,18–20] This confirmatory criterion makes sense because EOM size correlates well with contractile function,[15,17,20,21] so a markedly atrophic SO cannot generate normal force. Using the criterion of SO atrophy for unequivocal diagnosis of SO palsy, clinical series have demonstrated that the 3-step test is only 70% sensitive and 50% specific.[18] Poor reliability of the 3-step and other time-honored clinical tests for diagnosis of SO weakness implies that strabismus practice may be confounded by inclusion of other pathologic features masquerading as SO weakness.[6,7,22,23] Therefore, it has become important to evaluate the features and associations of actual SO weakness, rather than risking the perils of circular reasoning resulting from defining SO palsy as equivalent to a set of clinical motility findings that we now know are only occasional and inconsistent associations of palsy. Thus, an important validating role for orbital MRI has emerged.

In our earlier, small study, only the MR pulley was displaced significantly in patients with SO palsy.[24] However, biomechanical modeling suggested that displacement of the MR pulley alone could not cause the clinical patterns of strabismus observed in SO palsy. Moreover, the earlier MRI study suggested that isolated MR pulley displacement was unlikely to be a secondary effect of ocular excyclotorsion because any ocular torsion should have displaced all the rectus pulleys to the same degree; none of the other 3 rectus pulleys were displaced.

This study reinvestigated pulley positions in SO palsy in a substantially larger number of patients and in several common subtypes of SO palsy. In light of the recent finding that the trochlear nerve bifurcates to innervate separately medial versus lateral compartments of the SO muscle that are specialized for vertical versus torsional actions,[25] a novel classification has been proposed based on the shape of the palsied SO muscle: isotropic atrophy with a rounded SO muscle cross section versus anisotropic atrophy with elongated SO muscle cross section.[26] It also has been suggested that the anatomic features of congenital versus acquired SO palsy may differ.[27–30] This study analyzed pulley positions in unilateral versus bilateral, congenital versus acquired, and isotropic versus anisotropic SO palsy and investigated using computational simulation whether any associated rectus pulley displacements may influence the clinical patterns of strabismus observed in SO palsy.

Methods

Subjects

We studied 24 cases (17 males and 7 females) of SO palsy and 19 age-matched, healthy, orthotropic control subjects (10 males and 9 females). The median age of patients with SO palsy was 34 years (range, 1–83 years), closely matching that of normal subjects at 27 years (range, 18–74 years; $P = 0.89$). The median ages of patients with unilateral and bilateral SO palsy were 35 years (range, 1–80 years) and 31 years (range, 5–83 years), respectively ($P = 0.93$). Hypertropic subjects were diagnosed as having SO palsy based on evidence of atrophy of one or both SO muscles on coronal plane MRI. Patients were excluded when there was a history of previous strabismus surgery or if they had additional causes of strabismus such as dissociated vertical deviation, orbital trauma, or thyroid ophthalmopathy. The diagnosis of congenital or acquired SO palsy was based on the history of the age at which strabismus was first noted and early photographs when available. Probable causes of acquired SO palsy included cranial trauma, brain stem hemorrhage, and surgical manipulation causing trochlear nerve palsy. All other acquired cases were regarded as idiopathic. Three patients with SO palsy included in the previous study[24] were reanalyzed in this study using methods differing from the original ones. All subjects gave written, informed consent according to a protocol approved by the University of California, Los Angeles, Institutional Review Board that conformed to the tenets of the Declaration of Helsinki.

Control subjects underwent comprehensive eye examinations to verify normal corrected vision, binocular alignment, and stereoacuity. Patients with SO palsy underwent complete ophthalmic evaluation, including binocular alignment measured by alternate prism cover testing by turning or tilting the patient's head approximately 30° in cardinal gazes and head tilt positions, as well as ocular versions, and stereoacuity was measured by the Titmus test. Deviations were neutralized by placing a base-down prism in front of the hypertropic eye. Diagnostic occlusion was not performed. Subjective ocular torsion was measured by double Maddox rods when age appropriate. Confirmatory Hess screen testing was performed in patients who were consistently able to appreciate diplopia with normal retinal correspondence.

Magnetic Resonance Imaging

High-resolution T1- or T2-weighted fast spin echo MRI was performed for each subject using a 1.5-Tesla scanner (Signa; GE Health Care, Milwaukee, WI) and a face mask–mounted, dual-phased surface coil array (Medical Advances, Milwaukee, WI), as described in detail elsewhere.[17,20] Each orbit was imaged during monocular fixation by that eye in central gaze. Contiguous 2- or 3-mm-thick quasicoronal images were obtained perpendicular to the long axis of the orbit using a matrix of 256 × 256 pixels over a 6-, 8-, or 10-cm field of view, giving 234- to 390-μm in-plane resolution.

Analysis

Digital MRI images were processed using Adobe Photoshop (Adobe Systems, San Jose, CA) and were quantified using the program ImageJ (National Institutes of Health, Bethesda, MD). Images of left orbits were reflected to the orientation of right orbits to analyze EOM positions uniformly.

Cross sections of the rectus and SO EOMs were outlined manually in contiguous images, and the area centroid was taken as the EOM position. The globe was assumed to be spherical and its 3-dimensional center was determined as previously described.[5]