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Editorial: Muscle Paths Matter in Strabismus Associated with Axial High Myopia

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The relationship between high axial myopia and severe strabismus has long been recognized. Our understanding of the pathogenesis of this form of strabismus has, however, evolved tremendously, as illustrated by the paper in this issue by Yamaguchi and colleagues. Classical concepts of pathogenesis of severe myopic strabismus included supposition that the enlarged, highly myopic globe was gravitationally heavy and would physically sink to orbital floor¹, or compress the lateral rectus muscle², causing ischemic paresis resulting in esotropia. These pathophysiologic suppositions provided ready explanations for the typical failure of standard recess/resect strabismus surgery to correct severe strabismus associated with high myopia. In retrospect, these formerly-reasonable concepts now seem outdated.

Yamaguchi and colleagues here present a persuasive series of highly myopic patients in whom magnetic resonance imaging (MRI) demonstrated superotemporal globe shift out of normal relationship to the array of the rectus extraocular muscles³. Thus, both the superior rectus becomes abnormally nasal and the lateral rectus muscle inferior relative to the globe. Since the oculorotatory effect of a muscle is determined by pulling direction relative to the globe, superotemporal globe dislocation adds adducting and infraducting effect, while reducing supraducting and abducting effect. The muscles in the myopic patients described by Yamaguchi et al. had no intrinsic abnormality of force generally, but muscle paths were abnormal. Yamaguchi and colleagues demonstrated that the severity of this anatomic abnormality is associated with the severity of strabismus. Moreover, they demonstrated that surgical union between the corresponding margins of the superior and lateral rectus muscles can restore the normal anatomic relationship of muscles, and markedly improve the esotropia and hypotropia. This rectus union operation sometimes required supplemental medial rectus recession, but in several cases was fully effective even without medial rectus surgery. Although no lateral rectus resection was performed, the surgical result was highly successful and durable over as long as 7 years. The technique described by Yamaguchi and colleagues represents a powerful addition to our surgical armamentarium.

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It is interesting consider the broad implications of what has been demonstrated so clearly by Yamaguchi and colleagues. Severe esotropia and hypotropia can be corrected by suturing together the corresponding borders of the superior and lateral rectus muscles. Until recently, such an operation would have been unimaginable in this setting. How did ophthalmology reach the point where this operation could have been conceived? A likely answer lies in recent advances in the knowledge of the anatomy and physiology of extraocular muscles and their associated connective tissues. Joel M. Miller first demonstrated MRI evidence that the rectus muscles have paths that are constrained anteriorly in the orbit by connective tissues that he termed the "pulleys."⁴ Miller proposed that pulleys were responsible for maintaining the stability of the deep paths of the rectus muscles, preventing them from side-slipping over the globe in secondary and tertiary gazes. Miller's seminal concept motivated systematic reexamination of orbital anatomy, with surprising results. It has since emerged that each muscles consists of two substantial layers: the global layer continuous with the tendinous insertion on the sclera that rotates the eye, and the orbital layer that inserts posteriorly to the globe on a connective tissue of the pulley⁵. Rectus pulleys consist of collagenous rings stiffened by elastin that coincide with muscle penetrations through posterior Tenon's fascia. Although demonstrable histologically, pulleys also can be demonstrated by high quality MRI in living people. The orbital layers of muscle normally shift pulley locations to control muscle pulling directions. Recent neurophysiological findings strongly suggest that pulleys are responsible for fundamental aspects of ocular kinematics⁶⁻⁸, including Listing's Law⁹. Especially relevant here is that the superior and lateral rectus pulleys are normally joined by a ligament, the "LR-SR band," that suspends the LR pulley and prevents the globe from dislocating superotemporally¹⁰.

Another fundamental development that enabled Yamaguchi and collaborators to develop and study their powerful new surgical approach was widespread application of orbital imaging to the study of strabismus. Imaging, usually including coronal or sagittal planes, can be performed by computed x-ray tomography or better yet by MRI¹¹. Application of imaging has demonstrated that three pulley pathologies can cause strabismus: (1) heterotopy, representing mislocation, as in the present case of strabismus associated with high axial myopia; (2) instability that intermittently shifts pulleys, causing a complex incomitant strabismus; and (3) hindrance, representing impediment to physiologic shift of rectus pulleys during muscle contraction, and is associated with restrictive strabismus⁸. Recognition of these pathologies has motivated increasing application of clinical imaging in strabismus, so that cases of strabismus due to muscle path abnormalities are more frequently recognized.

Severe strabismus associated with high axial myopia is rare, except perhaps in certain genetic "hot spots" where it may be locally more common. Nevertheless, strabismus due to relative dislocation of rectus muscles is widespread, deserving routine consideration in clinical strabismus evaluation. Clark et al. employed MRI to demonstrate inferior displacement of both the medial and lateral rectus muscles as a general phenomenon associated with the aging of otherwise asymptomatic patients¹², and an explanation for prevalent age-related deficiency of supraduction¹³. This is likely because the LR-SR band degenerates during normal aging^{10, 14}. Rutar and Demer demonstrated that inferior shift of the lateral rectus relative to the globe may occur excessively or asymmetrically in non-myopic patients who develop strabismus similar to that in high axial myopes¹⁴. Inferior shift of the lateral rectus was associated with age-related degeneration of the LR-SR band¹⁴. Rutar and Demer proposed that where the lateral rectus sags symmetrically in two orbits, abduction is limited, but symmetric limitation to supraduction prevents development of vertical strabismus¹⁴. Such patients present with findings of divergence paralysis esotropia, esotropia present mainly or exclusively during distance fixation and absent at near¹⁵. Divergence paralysis esotropia is associated with normal saccadic velocities, for which it may be inferred that the lateral rectus remains normally innervated¹⁵. It makes sense, however, that if the lateral rectus is not pulling effectively into

an abducting direction, even normal lateral rectus force cannot prevent esotropia during distance fixation. Rutar and Demer also proposed that asymmetric lateral rectus sagging muscle would produce hypotropia as well as possible esotropia¹⁴. This strabismus responds to the operation described in this issue by Yamaguchi and colleagues³.

It can be no accident that there is strong association between high axial myopia and severe globe dislocation. While Yamaguchi and colleagues suggested that the bulky globe might displace the extraocular muscles, an alternative mechanism deserves consideration. It might be that a connective tissue abnormality expressed in both the sclera and the LR-SR band bridging the superior and lateral rectus muscles¹⁴ may be the common factor allowing both development of high axial myopia, and LR-SR band degeneration permitting superotemporal globe dislocation. There is likely to exist a genetic association between these axial high myopia and LR-SR band degeneration.

Clinicians should be mindful that while Yamaguchi and colleagues have demonstrated effectiveness of their operation uniting the lateral and superior rectus margins for the patients described, severe strabismus may arise in other axial myopic patients for whom this operation would be inappropriate. For instance, highly myopic patients can still experience neurogenic abducens paralysis, thyroid ophthalmopathy, or myasthenia gravis. Finally, patients with severe myopic staphylomata may develop such bizarre globe shapes that the eye cannot rotate in the orbit without collision with the orbital walls¹⁶. In such cases, no operation on the extraocular muscles could correct the resulting strabismus, and it would probably be harmful to attempt such surgery. For these reasons, clinicians may find it prudent to conduct preoperative imaging in highly myopic patients with severe strabismus before selecting a surgical procedure. Imaging would allow the selection of an operation such as described by Yamaguchi and colleagues where appropriate, while alternatively suggesting a different operation where indicated.

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Biography

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