HETEROTOPY OF EXTRAOCULAR MUSCLE PULLEYS CAUSES INCOMITANT STRABISMUS

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I. INTRODUCTION

Re-examination of orbital anatomy has shown that the rectus and inferior oblique muscles pass through connective tissue sleeves in posterior Tenon's fascia that constrain muscle paths and thus act as functional origins of the muscles, i.e. soft pulleys. The pulleys, which are composed of collagen and elastin stiffened by richly innervated smooth muscle, are anchored to the orbital bones and to one another via connective tissue bands[1]. Quantitative analysis of magnetic resonance images (MRI) of normal orbits shows the locations of each of the pulleys to be highly uniform, with the confidence limits of less than a millimeter for their horizontal and vertical coordinates[2]. This suggests that abnormal location of pulleys could produce strabismus. We therefore employed high resolution orbital imaging to determine if rectus pulleys are mislocated in strabismic patients.

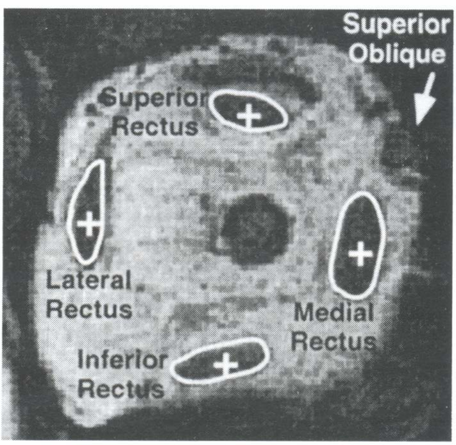

Fig. 1. Three mm thick, 390 µm resolution coronal MRI scan of the right orbit of a 26 year old woman with right hypertropia that increased in right gaze. Note 1.8 mm inferior displacement of MR and 1.7 mm nasal displacement of SR muscles. There was also a 2.3 mm superior displacement of the left LR muscle (not shown). The rectus cross sections have been digitally outlined and their area centroids marked with crosses.

II. METHODS

We obtained coronal orbital images by surface coil MRI [3] or computed radiographic tomography (CT) [4] in 18 strabismic patients. Geometric corrections were made for variations in head position during scanning[2]. Using the computer program *NIH Image* for analysis of a coronal plane close to the junction of the globe with the optic nerve, the rectus muscles were outlined to determine coordinates of their area centroids (Fig. 1). Since the foregoing image plane approximates the anteroposterior location of the rectus muscle pulleys[2, 5], area centroids of the muscles correspond to the coordinates of the pulleys themselves. Imaging findings were compared with a group of 18 normal subjects and correlated with Hess screen test results. Computer simulation of binocular alignment was performed using the *Orbit 1.8* program (Eidactics, San Francisco, CA). Initial simulations were performed using only pulley coordinates determined from analysis of the orbital images. Observed patterns of binocular alignment were them compared with the initial simulations to determine of the pulley abnormalities could explain the qualitative incomitance present. More refined computer simulations were later performed in selected cases using reasonable secondary changes in muscle lengths to account for comitant components of deviations.

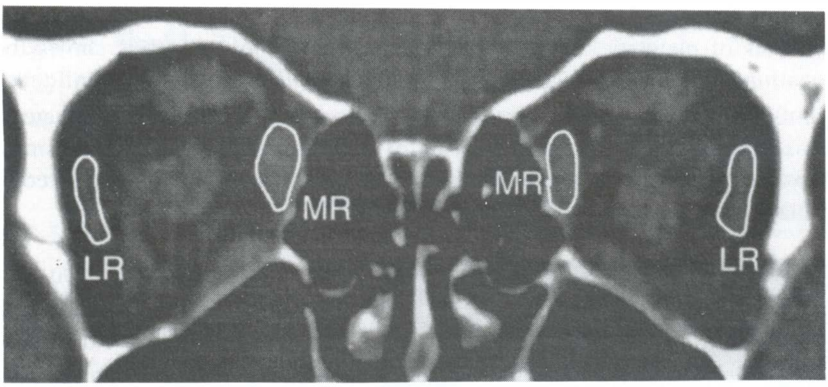

Fig. 2. 211 μm resolution, 1 mm thick coronal CT scan of the orbits of a 5 year old girl with large "V" esotropia and marked overelevation and underdepression of the right eye in adduction. Note inferior displacement of right LR more than left LR.

III. RESULTS

In each patient the size and contractility of the superior oblique (SO) muscle was normal, ruling out SO palsy[3]. Every patient had one or more rectus muscles significantly (two or more standard deviations) displaced from the normal position in the coronal plane.

Imaging

Six patients had "V" pattern incomitance. Most commonly this was associated with a relative superior placement of the medial rectus (MR) pulley relative to the lateral rectus (LR) pulley, usually in both orbits but unilaterally in one case. Two patients had lateral displacement of the superior rectus (SR) pulley relative to the inferior rectus (IR) pulley, usually bilaterally. Fig. 2 shows the orbits of a 5 year old girl with an esotropia that increased by 50Δ from elevation to depression; she also had in adduction of the right eye marked overelevation and underdepression. Note inferior displacement of the right LR more than the left LR, and nasal displacement of the SR in both eyes.

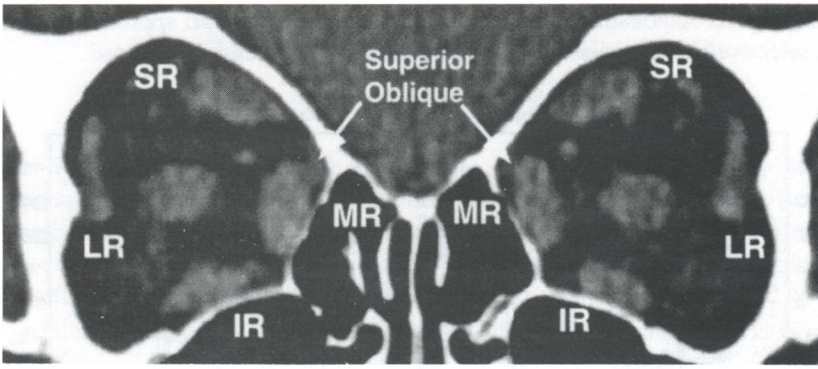

Fig. 3. CT scan of 6 year old girl with "A" pattern esotropia 60Δ greater in elevation than depression. Note LR displaced superiorly and SR displaced nasally in both eyes.

Eight patients had "A" pattern incomitance. This was always associated with superior displacement of the LR pulley relative to the MR pulley, and frequently also with nasal displacement of the SR pulley relative to the IR pulley. This pattern is seen in Fig. 3, a CT scan of the orbits of a child.

Four patients had hypertropias with incomitance not suggestive of isolated muscle weakness. Each had one or more heterotopic extraocular muscles. An example is seen in Fig. 1 for a patient who had right hypertropia greatest in right gaze. MRI shows inferior displacement of LMR pulley and nasal displacement of RSR pulley, with inferior displacement of the LLR pulley.

Computer modeling using observed pulley positions accounted for the clinically observed patterns of strabismus in all patients. Agreement between observed patterns of strabismus and those predicted on the basis of pulley positions is shown for the typical patient of Fig. 4. Patients responded to strabismus surgeries, usually including rectus muscle transpositions, designed to correct the imbalances produced by pulley heterotopies.

IV. DISCUSION

Orbital imaging now indicates that malpositioning of rectus muscle pulleys is present in many cases of incomitant strabismus that have been conventionally attributed to "oblique muscle dysfunction." Orbital imaging in patients with confirmed SO palsy suggests that rectus pulley heterotopy is not the result of ocular torsion[6] Computer modeling as performed here indicates that malpositioning of the pulleys in these cases is alone sufficient to cause the observed patterns of incomitance. The foregoing evidence thus makes it reasonable to suppose that pulley heterotopy is the cause of many cases of incomitant strabismus. It is not necessary to suppose unproved mechanisms of oblique muscle over- or underaction. In cases where orbital imaging has demonstrated rectus muscle heterotopy, surgical approaches such as selective transposition of rectus muscles can be effectively selected.

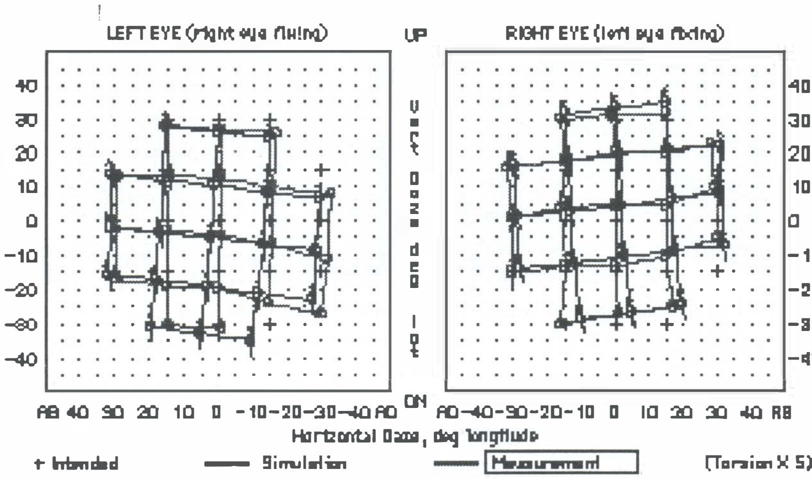

Fig. 4. Hess screen charts for patient shown in Fig. 1. Note excellent agreement between observed alignment and that predicted based on observed location of muscle pulleys, with 1.5 mm shortening of right SR, and 1.5 mm lengthening of right IR.

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