Orbital Disorders

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Topics

Common Non-neurological Causes of Strabismus:
- CHILDREN: Congenital malpositioning of structurally sound rectus pulley system.
- OLDER ADULTS: Acquired rectus pulley malpositioning due to connective tissue degeneration.

Novel Neurological Causes of Strabismus:
- Compartmental lateral rectus palsy.
- Compartmental superior oblique palsy.

Rectus Pulley System: Inner Gimbal of the Eye

Functional Pulleys

Childhood Onset Pulley Heterotopy

34 cases (from prospective imaging data set of 577 total strabismus cases and 157 normal controls)

MRI ALWAYS shows robust LR-SR band.

Congenital A Pattern Pulley Heterotopy
Congenital A Pattern Pulley Heterotopy

Lid fissures usually correlate.

Congenital V Pattern Pulley Heterotopy

Robust Band

+4

-4
**Congenital V Pattern Pulley Heterotopy**

Lid fissures sometimes correlate.

**Late Onset Pulley Heterotopy**

- 28+ cases
- Diplopia onset: 68 ± 12 (std. dev.) years

MRI NEVER shows robust LR-SR band. It is usually ruptured.

**Age-Related Degeneration of LR-SR Band**

Sagging Eye Syndrome
MRI Shows Age-Related Degeneration of LR-SR Band

Bilaterally Symmetrical LR Sag: Divergence Paralysis Esotropia (DPE)

Asymmetric LR Sag Causes Cyclovertical Strabismus

Cases of Divergence Paralysis Esotropia (DPE) = Age-related Distance Esotropia

Criteria
1. ET at distance, vertically comitant
2. Fusion at Near
3. Normal Abduction Saccades

Patients- 11
1. Age: 72 ± 11 years
2. Gender: 7 women and 4 men
3. Distance ET: 11.5 ± 10.6 Δ (SD)
4. Near E’: 1.3 ± 3.1 Δ

Exclusions:
1. Orbital Trauma
2. Restrictive Strabismus (i.e. thyroid)
3. Prior strabismus surgery
4. High myopia
Cases of Cyclovertical Strabismus (CVS)

Patients - 17
1. Age: 68 ± 2 years
2. Gender: 10 women and 7 men
3. Distance HT: 9.9 ± 3.4 D (SD)

Exclusions:
1. Superior Oblique Palsy
2. Orbital Trauma
3. Restrictive Strabismus (i.e. thyroid)
4. Prior strabismus surgery
5. High myopia
6. Skew deviation

Magnetic Resonance Imaging
Surface coils
Central target fixation
Image Planes:
Quasi-coronal planes for pulley positions
Axial planes for horizontal rectus lengths

Controls
Younger Normal: 52 orbits, 28 subjects
Normal eye exam
Age 23 ± 5 years

Age-Matched Older Normal: 25 orbits, 14 subjects
No ocular disease (pseudophakia allowed)
Age 65 ± 5 years

Superior Sulcus Defect in SES
64% of subjects

Blepharoptosis in SES
29% had ptosis and high lid crease.
29% had prior blepharoplasty, brow lift, or face lift surgery.
Rectus Pulleys Heterotopic in SES

Table 1. Rectus Pulley Positions Relative To Globe Center, mm.

<table>
<thead>
<tr>
<th>Group</th>
<th>Medial Rectus</th>
<th>Superior Rectus</th>
<th>Inferior Rectus</th>
</tr>
</thead>
<tbody>
<tr>
<td></td>
<td>Lateral Mean</td>
<td>Lateral Mean</td>
<td>SD</td>
</tr>
<tr>
<td>DPE</td>
<td>12.19 ± 0.84</td>
<td>2.35 ± 1.52</td>
<td>0.75</td>
</tr>
<tr>
<td>CVS</td>
<td>12.19 ± 0.84</td>
<td>2.35 ± 1.52</td>
<td>0.75</td>
</tr>
</tbody>
</table>

* Significant differences between DPE and control groups, P < 0.005.
† Significant differences for the hypotropia eye of CVS vs. controls.
‡ Significant differences for the hypotropia eye of CVS vs. controls.

Lateral Rectus Pulley

P = 0.02
OD - OS
Symmetrical < 0.5 mm
P = 0.8
Excyclo 12 ± 6°
P = 0.01
Excyclo 7 ± 5°
P < 0.005

Always Asymmetrical
> 1 mm

14/22 Ruptured
31/34 Ruptured

Lateral Rectus Angle - deg

P < 10°
Horizontal Rectus Muscle Elongation in SES

Avoids surgery near the fragile LR-SR band.

Medial Rectus Recession for DPE in SES

MR recession dose:
1. Doubling the measured distance ET.
2. Recess MR based on surgical dose table of Parks et al. for doubled ET.

Graded Vertical Rectus Tenotomy

For Cyclovertical Strabismus in SES

Partial tenotomy at margin of scleral insertion
Reduces EOM tension
20 for 40% to 6D for 80%.
SES is **NOT** “Heavy Eye Syndrome”

- Inferonasal shift of lateral rectus, hugging the globe.
- Nasal shift of inferior rectus.
- Nasal shift of superior rectus.

“Heavy Eye Syndrome”

- Inferotemporal shift of lateral rectus.
- Temporal shift of inferior rectus.
- No shift of superior rectus.
- Lateral rectus shifted away from globe.

“Sagging Eye Syndrome”

Age-related degeneration of LR-SR band allows lateral rectus pulley to shift and tilt inferolaterally.
Rectus EOMs elongate.
When symmetrical: divergence paralysis esotropia.
When asymmetrical: ipsilateral hypotropia and excyclotropia
(Note: SO palsy causes ipsilateral hypertropia and excyclotropia).

Heavy Eye Syndrome

Adnexal “sag” strongly correlates with pulley sag.
Diagnosis does NOT require imaging in most cases.
Divergence paralysis ET treatable by large MR recessions.
Cyclovertical strabismus treatable by partial vertical rectus tenotomy.
SES is probably a major cause of adult acquired strabismus.
Compartmental Pathology of Horizontal Rectus Muscles

Frequent congenital cranial dysinnervation disorders: Duane syndrome
  Congenital fibrosis
  Occasionally in normal subjects.

Longitudinal Fissure In Human Lateral Rectus Muscle

Human Abducens Nerve Has Superior & Inferior Divisions
Abducens Nerve Has Superior & Inferior Divisions

Human Medial Rectus Motor Nerve Has Superior & Inferior Divisions
Patient WS
Left Superior Compartment Lateral Rectus Palsy

- Limited Abduction
- Incomitant Esotropia
- Superior LR Atrophy

1. Plicate superior LR tendon 10 mm
2. Transpose superior LR tendon 1/2 tendon width
3. Recess left MR 5 mm

Pre-operative

Patient WS
Left Superior Compartment Lateral Rectus Palsy

Post-operative

Improved

Incomitant Esotropia
Ipsilesional Hypotropia in 15/39 Cases

Trochlear Nerve Branches in Adult Superior Oblique Muscle
(Confirmed in 4 specimens)

Adult Human Superior Oblique

Superior Oblique Compartments

- Torsional
- Vertical

Adult Human Superior Oblique

Medial   Lateral


Superior Oblique Compartments

- Torsional
- Vertical

Anisotropic: More hyper in infraversion
More extorsion

| Binocular Alignment Unilateral Superior Oblique Atrophy |
|-----------------|-----------------|-----------------|-----------------|
|                  | Anisotropic Atrophy (n=36) | Isotropic Atrophy (n=26) | P value          |
| Age (years)      | 39 ± 17.4        | 26 ± 13.3        | 0.733           |
| Hypertropia (Δ)  |                  |                  |                 |
| Central Gaze     | 14 ± 7.4         | 13 ± 7.1         | 0.899           |
| Sursumversion    | 9 ± 7.5          | 9 ± 8.7          | 0.914           |
| Infaversion      | 16 ± 13.7        | 6 ± 4.8          | 0.048           |
| Incompliance of Hypertropia (Δ) |                  |                  |                 |
| Central Minus Sursumversion | 3 ± 2.9         | 3 ± 6.3          | 0.956           |
| Central Minus Infaversion | -4 ± 7.7        | 6 ± 5.7          | 0.014           |
| Lateral          | 10 ± 7.5         | 10 ± 5.7         | 0.916           |
| Right Minus Left Head Tilt | 10 ± 6.5        | 14 ± 6.1         | 0.596           |
| Exycyclotorsion (degrees) | 8 ± 3.5         | 4 ± 2.6          | 0.032           |

Conclusions

Lateral rectus (LR) muscle has two neuromuscular compartments that differentially supplement abduction:
Superior adds supraduction
Inferior adds infraduction

Each compartment is innervated by a separate abducens nerve branch.
Superior compartment LR palsy is common, and features hypotropia of the affected eye.

Compartmental atrophy correlates with clinical presentation.
Some patients probably have selective abducens nerve lesions affecting only one compartment.

Conclusions

Superior oblique (SO) muscle has two neuromuscular compartments that function differentially:
Medial mainly for incycloduction
Lateral mainly for infraduction

Each compartment is innervated by a separate trochlear nerve branch.
Some cases of SO weakness exhibit patterns of atrophy suggesting predominant paresis of the lateral, vertical compartment.

Compartmental atrophy correlates with clinical presentation.
Some patients probably have partial trochlear nerve lesions affecting only one compartment.

Take-Home Message

Disorders of rectus muscle paths cause some cases of strabismus.

CHILDREN: Congenital malpositioning of structurally sound rectus pulley system.

OLDER ADULTS: Acquired rectus pulley malpositioning due to connective tissue degeneration.
Implications

Differential surgeries have long been employed for SO.
Harada-Ito advances the anterior SO to create intorsion.
Posterior tenectomy reduces SO infraduction without torsion effect.
Anterior tenectomy reduces intorsion without much vertical effect.

We could consider differential surgeries on the respective compartments of the horizontal rectus muscles.

We can selectively tighten a paretic LR superior compartment, for example.

Consider other clinical possibilities.

Peripheral Ocular Motor Apparatus is More Complex Than You Learned In School!

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