Pediatric Ophthalmology 2014
A Magnificent Mile of Innovations

Program Directors
Jane C Edmond MD and Daniel E Neely MD

In conjunction with the American Association for Pediatric Ophthalmology and Strabismus and the American Academy of Pediatrics

McCormick Place
Chicago, Illinois
Saturday, Oct. 18, 2014

Presented by:
The American Academy of Ophthalmology
2014 Pediatric Ophthalmology
Subspecialty Day Planning Group

On behalf of the American Academy of Ophthalmology, the American Association for Pediatric Ophthalmology and Strabismus (AAPOS) and the American Academy of Pediatrics (AAP), it is our pleasure to welcome you to Chicago and Pediatric Ophthalmology 2014: A Magnificent Mile of Innovations.

Jane C Edmond MD
Program Director
Alcon Laboratories, Inc.: L

Daniel E Neely MD
Program Director
None

Stephen P Christiansen MD
National Eye Institute: S

Daniel J Karr MD
None

R Michael Siatkowski MD
National Eye Institute: S

Laura B Enyedi MD
Pediatric Eye Disease Investigator Group: S

David A Plager MD
Alcon Laboratories, Inc.: S
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Pediatric Ophthalmology 2014 Contents

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CME Credit

Academy’s CME Mission Statement
The purpose of the American Academy of Ophthalmology’s Continuing Medical Education (CME) program is to present ophthalmologists with the highest quality lifelong learning opportunities that promote improvement and change in physician practices, performance or competence, thus enabling such physicians to maintain or improve the competence and professional performance needed to provide the best possible eye care for their patients.

2014 Pediatric Ophthalmology Subspecialty Day Meeting Learning Objectives
Upon completion of this activity, participants should be able to:
■ Evaluate new disease entities, practices, technologies and treatment that may change current practice
■ Plan the surgical treatment of complex strabismus
■ Expand the strabismus surgeon’s surgical armamentarium
■ Discuss various common pediatric anterior segment and corneal diseases and explore new developments in their diagnosis and treatment
■ Provide patients with new information about amblyopia, including its pathophysiology and novel new treatments
■ Assess challenging pediatric neuro-ophthalmology cases and discuss the latest diagnostic and treatment options

2014 Pediatric Ophthalmology Subspecialty Day Meeting Target Audience
The intended target audience for this program is pediatric ophthalmologists, comprehensive ophthalmologists, medical professionals, visual physiologists and orthoptists who are involved in maintaining high-quality health care for the pediatric and strabismus populations.

2014 Pediatric Ophthalmology Subspecialty Day Meeting CME Credit
The American Academy of Ophthalmology is accredited by the Accreditation Council for Continuing Medical Education to provide CME for physicians.

The American Academy of Ophthalmology designates this live activity for a maximum of 7 AMA PRA Category 1 Credits™. Physicians should claim only the credit commensurate with the extent of their participation in the activity.

Self-Assessment Credit
This activity meets the Self-Assessment CME requirements defined by the American Board of Ophthalmology (ABO). Please be advised that the ABO is not an accrediting body for purposes of any CME program. The ABO does not sponsor this or any outside activity and does not endorse any particular CME activity. Complete information regarding the ABO Self-Assessment CME Maintenance of Certification requirements are available at

NOTE: Credit designated as “self-assessment” is AMA PRA Category 1 Credit™ and is also preapproved by the ABO for the Maintenance of Certification (MOC) Part II CME requirements.

Teaching at a Live Activity
Teaching instruction courses or delivering a scientific paper or poster is not an AMA PRA Category 1 Credit™ activity and should not be included when calculating your total AMA PRA Category 1 Credits™. Presenters may claim AMA PRA Category 1 Credits™ through the American Medical Association. Please contact the AMA to obtain an application form at www.ama-assn.org.

Scientific Integrity and Disclosure of Financial Interest
The American Academy of Ophthalmology is committed to ensuring that all CME information is based on the application of research findings and the implementation of evidence-based medicine. It seeks to promote balance, objectivity and absence of commercial bias in its content. All persons in a position to control the content of this activity must disclose any and all financial interests. The Academy has mechanisms in place to resolve all conflicts of interest prior to an educational activity being delivered to the learners.

Attendance Verification for CME Reporting
Before processing your requests for CME credit, the Academy must verify your attendance at Subspecialty Day and/or AAO 2014. In order to be verified for CME or auditing purposes, you must either:
■ Register in advance, receive materials in the mail and turn in the Final Program and/or Subspecialty Day Syllabus exchange voucher(s) onsite;
■ Register in advance and pick up your badge onsite if materials did not arrive before you traveled to the meeting; or
■ Register onsite.

CME Credit Reporting
South, Level 2.5; Academy Resource Center, Booth 508
Attendees whose attendance has been verified (see above) at AAO 2014 can claim their CME credit online during the meeting. Registrants will receive an email during the meeting with the link and instructions on how to claim credit.

Onsite, you may report credits earned during Subspecialty Day and/or AAO 2014 at the CME Credit Reporting booth.

Academy Members: The CME credit reporting receipt is not a CME transcript. CME transcripts that include AAO 2014 credits entered onsite will be available to Academy members on the Academy’s website beginning Nov. 13, 2014.
NOTE: CME credits must be reported by Jan. 15, 2015. After AAO 2014, credits can be claimed at www.aao.org.

The Academy transcript cannot list individual course attendance. It will list only the overall credits spent in educational activities at Subspecialty Day and/or AAO 2014.

Nonmembers: The Academy will provide nonmembers with verification of credits earned and reported for a single Academy-sponsored CME activity, but it does not provide CME credit transcripts. To obtain a printed record of your credits, you must report your CME credits onsite at the CME Credit Reporting booths.

Proof of Attendance
The following types of attendance verification will be available during AAO 2014 and Subspecialty Day for those who need it for reimbursement or hospital privileges, or for nonmembers who need it to report CME credit:
- CME credit reporting/proof-of-attendance letters
- Onsite Registration Form
- Instruction Course Verification Form

Visit the Academy’s website for detailed CME reporting information.
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Miles Professor of Ophthalmology and
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Storm Eye Institute
Medical University of South Carolina
Pediatric Ophthalmology 2014: A Magnificent Mile of Innovations

In conjunction with the American Association for Pediatric Ophthalmology and Strabismus and the American Academy of Pediatrics

SATURDAY, OCT. 18, 2014

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<tr>
<th>Time</th>
<th>Event</th>
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<tr>
<td>7:00 AM</td>
<td>CONTINENTAL BREAKFAST</td>
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<tr>
<td>8:00 AM</td>
<td>Welcome and Introductions</td>
<td>Jane C Edmond MD*</td>
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<td>Daniel E Neely MD</td>
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Section I: Reoperation Strategies in Strabismus
Moderator: David A Plager MD*

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<thead>
<tr>
<th>Time</th>
<th>Topic</th>
<th>Presenter(s)</th>
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<tr>
<td>8:02 AM</td>
<td>Introduction and Self-assessment</td>
<td>David A Plager MD*</td>
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<tr>
<td>8:04 AM</td>
<td>Tort Reform</td>
<td>Burton J Kushner MD</td>
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<tr>
<td>8:13 AM</td>
<td>A Twist on Brown Syndrome—Turning It Upside Down</td>
<td>David L Guyton MD*</td>
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<tr>
<td>8:22 AM</td>
<td>Entering the In Crowd: Esotropia Following Lateral Rectus Recession</td>
<td>M Edward Wilson Jr MD</td>
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<tr>
<td>8:31 AM</td>
<td>The Outcast: Exotropia Following Medial Rectus Recession</td>
<td>Jane C Edmond MD*</td>
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<tr>
<td>8:40 AM</td>
<td>If at First You Don’t Succeed . . .</td>
<td>Edward G Buckley MD</td>
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<tr>
<td>8:49 AM</td>
<td>I. O. You Won</td>
<td>David A Plager MD*</td>
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<tr>
<td>8:58 AM</td>
<td>Conclusion and Self-assessment</td>
<td>David A Plager MD*</td>
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Section II: Tough Pediatric Cornea and Anterior Segment Cases—Advice From the Experts
Moderator: Daniel J Karr MD

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<tr>
<td>9:02 AM</td>
<td>Introduction and Self-assessment</td>
<td>Daniel J Karr MD</td>
</tr>
<tr>
<td>9:04 AM</td>
<td>White Eye, White Corneal Lesion</td>
<td>M Bowes Hamill MD*</td>
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<tr>
<td>9:13 AM</td>
<td>Red Eye, White Corneal Lesion</td>
<td>Sonal S Tuli MD</td>
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<tr>
<td>9:22 AM</td>
<td>Red Eyes, Red Lid Margins</td>
<td>Ivan R Schwab MD FACS</td>
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<tr>
<td>9:31 AM</td>
<td>Recurrent Red Eye</td>
<td>Kathryn A Colby MD PhD*</td>
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<tr>
<td>9:40 AM</td>
<td>Red, Itchy Eyes</td>
<td>Stephen C Pflugfelder MD*</td>
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<tr>
<td>9:49 AM</td>
<td>Congenital White Eyes, Turned into Bright Eyes</td>
<td>Kanwal K Nischal MBBS*</td>
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<tr>
<td>9:58 AM</td>
<td>Conclusion and Self-assessment</td>
<td>Daniel J Karr MD</td>
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<tr>
<td>10:00 AM</td>
<td>REFRESHMENT BREAK and AAO 2014 EXHIBITS</td>
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Section III: Cutting-edge Innovations in Amblyopia
Moderator: Laura B Enyedi MD

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<th>Topic</th>
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<tr>
<td>10:35 AM</td>
<td>Introduction and Self-assessment</td>
<td>Laura B Enyedi MD</td>
</tr>
<tr>
<td>10:37 AM</td>
<td>Where Are We Now? Screening for Amblyopia in 2014 and Beyond</td>
<td>Sean P Donahue MD PhD*</td>
</tr>
<tr>
<td>10:46 AM</td>
<td>Amblyopia: It’s a Binocular Disorder</td>
<td>Eileen E Birch PhD*</td>
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<tr>
<td>10:55 AM</td>
<td>New Noninvasive Treatment Modalities for Amblyopia</td>
<td>Robert Hess PhD*</td>
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<tr>
<td>11:04 AM</td>
<td>New Medical Treatment Modalities for Amblyopia</td>
<td>Michael X Repka MD MBA*</td>
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* Indicates that the presenter has financial interest.
No asterisk indicates that the presenter has no financial interest.
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<tr>
<th>Time</th>
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<tbody>
<tr>
<td>11:13 AM</td>
<td>New Intraocular and Corneal Refractive Surgeries in the Management of Amblyopia</td>
</tr>
<tr>
<td>11:22 AM</td>
<td>The Pediatric Eye Disease Investigator Group and Amblyopia: Where We Have Been and Where We are Going</td>
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<tr>
<td>11:31 AM</td>
<td>Conclusion and Self-assessment</td>
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**Section IV: Enhanced Techniques for Strabismus Surgery**
Moderator: Daniel E Neely MD

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<th>Time</th>
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<tbody>
<tr>
<td>11:35 AM</td>
<td>Introduction and Self-assessment</td>
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<tr>
<td>11:37 AM</td>
<td>Minimally Invasive Strabismus Surgery</td>
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<td>Daniel S Mojon MD*</td>
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<tr>
<td>11:52 AM</td>
<td>A New Twist on the Inferior Oblique</td>
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<tr>
<td>12:00 PM</td>
<td>Plications for Strabismus Surgery</td>
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<td>12:09 PM</td>
<td>Strabismus Surgery After Scleral Buckle Procedure</td>
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<tr>
<td>12:18 PM</td>
<td>Short Tag Noose Adjustable Suture</td>
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<tr>
<td>12:27 PM</td>
<td>Superior Oblique Z-Tenotomy</td>
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<td>12:35 PM</td>
<td>Conclusion and Self-assessment</td>
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**Late Breaking News**

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<tr>
<td>12:37 PM</td>
<td>Telemedicine and ROP—Is It Ready for Prime Time?</td>
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<tr>
<td>12:44 PM</td>
<td>Advocating for Patients</td>
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<tr>
<td>12:49 PM</td>
<td>LUNCH and AAO 2014 EXHIBITS</td>
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**Section V: Video Bonanza**
Moderator: Stephen P Christiansen MD*

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<thead>
<tr>
<th>Time</th>
<th>Session</th>
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<tbody>
<tr>
<td>1:58 PM</td>
<td>Introduction and Self-assessment</td>
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<tr>
<td>2:00 PM</td>
<td>Persistent Fetal Vasculature Syndrome: Divide and Conquer</td>
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<tr>
<td>2:07 PM</td>
<td>Persistent Fetal Vasculature Syndrome: A Posterior Approach</td>
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<tr>
<td>2:13 PM</td>
<td>Masquerade Uveitis</td>
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<tr>
<td>2:19 PM</td>
<td>Superior Oblique Exaggerated Traction Test</td>
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<td>2:25 PM</td>
<td>Show Me My Cataract</td>
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<td>2:31 PM</td>
<td>Sinskey Procedure for Nystagmus</td>
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<td>2:37 PM</td>
<td>Pilomatrixoma of the Eyelid</td>
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<td>2:43 PM</td>
<td>The Induced Tropia Test</td>
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<tr>
<td>2:49 PM</td>
<td>Conclusion and Self-assessment</td>
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<tr>
<td>2:51 PM</td>
<td>REFRESHMENT BREAK and AAO 2014 EXHIBITS</td>
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**Section VI: OMIC Closed Cases**
Moderator: R Michael Siatkowski MD*

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<thead>
<tr>
<th>Time</th>
<th>Session</th>
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<tbody>
<tr>
<td>3:25 PM</td>
<td>Introduction and Self-assessment</td>
</tr>
<tr>
<td>3:27 PM</td>
<td>25-Year Review of Closed Claims in Pediatric Ophthalmology and Strabismus</td>
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* Indicates that the presenter has financial interest.
No asterisk indicates that the presenter has no financial interest.
### Lessons to Learn: Pearls from Individual Cases

**3:42 PM**
- **Lessons to Learn: Pearls from Individual Cases**
  - Robert E Wiggins MD MHA*
  - Robert S Gold MD*
  - Anne M Menke RN PhD

### Conclusions

**4:22 PM**
- **Conclusions**
  - Anne M Menke RN PhD

### Conclusion and Self-assessment

**4:25 PM**
- **Conclusion and Self-assessment**
  - R Michael Siatkowski MD*

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### Section VII: Perplexing Problems in Pediatric Neuro-Ophthalmology

**Moderator:** Jane C Edmond MD*

**Virtual Moderator:** Gena Heidary MD

**4:29 PM**
- **Introduction and Self-assessment**
  - Jane C Edmond MD*

**4:31 PM**
- **Case 1: A Star is Born – Acute Visual Loss**
  - Paul H Phillips MD

**4:43 PM**
- **Case 2: An Eyelid Has Fallen and Can’t Get Up – Acute Ptosis**
  - Stacy L Pineles MD

**4:55 PM**
- **Case 3: Seeing Two Mommies – Acute Onset Esotropia**
  - Gena Heidary MD*

**5:07 PM**
- **Case 4: Not Seeing Mommy – Infant With Poor Vision**
  - Mark S Borchert MD

**5:19 PM**
- **The Hardest Conversation: Functional Vision Loss and What to Say to the Parents and the Patient**
  - R Michael Siatkowski MD*

**5:29 PM**
- **Conclusion and Self-assessment**
  - Jane C Edmond MD*

**5:31 PM**
- **Closing Remarks**
  - Jane C Edmond MD*

**5:35 PM**
- **ADJOURN**

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No asterisk indicates that the presenter has no financial interest.
Tort Reform

Burton J Kushner MD

Case Summary
A 12-year-old girl has a recurrent head tilt to the right to damp nystagmus after prior successful torsional surgery.

History
The girl was born with unilateral optic nerve hypoplasia in her right eye with NLP. Her left eye is normal and corrects to 20/40. I first saw her at 8 years of age. She had nystagmus that damped in right gaze and head tilt right. For a 40-degree right tilt, 30-degree left turn, and a right esotropia that varied from 0 to 30 PD she underwent left medial rectus recession of 7 mm with supraplacement, a left lateral rectus resection of 9 mm with infraplacement, a left Harada-Ito, and anterior 7/8’s disinsertion of the left inferior oblique. Also, for the anticipated consecutive exotropia, the right lateral rectus was recessed 6 mm. This improved her head posture and substantially intorted the left eye. She returned at 12 years of age with a recurrence of the head tilt to the right.

Exam at Age 12
Alignment was approximately straight, but she fixated with a variable head tilt right that was about 30 degrees with visual effort. In that position her nystagmus damped.

Surgery
She underwent further surgery with the intention of further intorting her already intorted left eye.

Outcome
The surgery completely eliminated her head tilt, and the results have remained stable for 2 years. However, despite the surgical success, the procedure did not further intort her left eye.

What I Learned
Although surgery to rotate the fixing eye in the direction of the head tilt can improve an abnormal head posture, there must be another mechanism by which surgery can work. By shifting force vectors so that muscles that normally are torsional antagonists act synergistically after surgery, the nystagmus may be damped.
A Twist on Brown Syndrome: Turning It Upside Down
Inverted Brown Pattern: More Common Than You Think

David L Guyton MD

Introduction

A particular pattern has emerged from routine use of the Lancaster red-green test in the analysis of cyclovertical strabismus. We have dubbed this pattern the “inverted Brown pattern.” It is caused by a tight or inelastic inferior oblique muscle and is best treated by weakening this muscle, usually a large amount.

The Lancaster red-green test is invaluable in analyzing patterns of cyclovertical strabismus. A and V patterns are immediately obvious, the subjective torsion is noted in the 9 diagnostic positions of gaze, and torsion of the eye movement patterns with respect to one another can be analyzed in detail.

Our paper in 2006 that described the inverted Brown pattern (see reference) concentrated more on the less common causes, such as orbital trauma and overcorrection following superior oblique tenotomy for Brown syndrome. By far the more common presentation of the inverted Brown pattern, however, is as a recurrent superior oblique paresis after initial inferior oblique weakening surgery. It can also appear simply as a variant of unilateral superior oblique paresis without previous surgery.

When associated with superior oblique paresis, the inverted Brown pattern, which again is caused by a tight or inelastic inferior oblique muscle, masquerades simply as an underacting superior oblique muscle, without significant inferior oblique overaction (without significant elevation in adduction of the involved eye). The diagnosis is made by at least several of the following: (1) awareness of this possibility, (2) typical history of previous inferior oblique weakening surgery, (3) fundus extorsion of the involved eye, usually accompanied by subjective extorsion of the involved eye on Lancaster red-green testing, (4) the greatest hyperdeviation in the field of action of the paretic superior oblique muscle, (5) no or minimal overaction of the inferior oblique muscle, and (6) a tighter than normal inferior oblique muscle on exaggerated forced duction testing at the time of surgery.

Treatment is surgical—usually involving only a weakening procedure of the inferior oblique muscle of the involved eye. A large weakening procedure such as a denervation / extirpation is needed if the inferior oblique muscle has been weakened before. A barrier to this needed treatment is that most surgeons are very hesitant to do inferior oblique weakening when there is no significant inferior oblique muscle overaction, especially when the inferior oblique muscle has already been weakened. And yet the inverted Brown pattern is best treated precisely with further weakening of the inferior oblique muscle, often with excellent results.

Lancaster red-green plots are shown in Figure 1 (red or dark streaks = RE, green or light streaks = LE), from a patient with a relatively mild inverted Brown pattern on the left, before and after a large (~18-20 mm) myectomy of the left inferior oblique muscle.

Figure 1.

This 55-year-old male had a head tilt to the right for decades, and began to see double intermittently at approximately age 53. He had an intermittent left hypertropia, worse in gaze down and to the right in the field of action of the left superior oblique muscle. He had good stereoacuity. Clinically he showed only trace underaction of the left superior oblique muscle and had only trace extorsion of each fundus. The left hypertropia increased from 7 PD on forced right head tilt to 16 PD on forced left head tilt. The Lancaster red-green plot showed a small V pattern, extorsion of one eye with respect to the other, increasing in down gaze, and a left hyperdeviation most typical of underaction of the left superior oblique muscle.
Figure 2.

At the time of surgery, forced ductions of the oblique muscles were normal on the right, but on the left the inferior oblique muscle was definitely tighter than the superior oblique muscle. A large, 18-20 mm left inferior oblique myectomy was performed, with an excellent postoperative result, with only a small remaining left hyperdeviation in down gaze across the board.

Another patient, with a large inverted Brown pattern on the left, had the Lancaster red-green plots (red or dark streaks = RE, green or light streaks = LE) seen in Figure 3, before and after a large left inferior oblique myectomy.

Figure 3.

This 28-year-old male had a long-standing right head tilt and intermittent left hyperdeviation, with good stereoacuity with his anomalous head posture. Clinically he showed only +1 overaction of the left inferior oblique muscle, but −2 underaction of the left superior oblique muscle. The hyperdeviation increased from 5 PD on forced right head tilt to 28 PD on forced left head tilt. The left fundus was 2+ extorted. The Lancaster red-green plot showed a left hyperdeviation of close to 40 PD on straight right gaze, decreasing significantly on gaze up and to the right.

Figure 4.

At the time of surgery, forced ductions showed the left inferior oblique muscle to be minimally tighter than the right inferior oblique muscle. A large left inferior oblique myectomy was performed, with an excellent postoperative result, specifically without creating significant underaction of the left inferior oblique muscle.
A third patient, 55 years old, had had a mild left superior oblique paresis 5 years before, for which a right inferior rectus muscle recession was performed, with an initial excellent postoperative result. The hypertropia recurred, however, several years later (as has been the case in all 4 such patients that could be located in our database). A left inferior oblique muscle recession had essentially no effect, and the Lancaster red-green plots seen in Figure 5 (red or dark streaks = RE, green or light streaks = LE), show the deviation before and after a left inferior oblique denervation/extirpation.

Figure 5.

After a left inferior oblique muscle recession, with essentially no effect, there is still an inverted Brown pattern on the left, with the greatest deviation in down and right gaze.

Figure 6.

The previously operated left inferior oblique muscle was found to be 3+ tight, and a large denervation/extirpation procedure (including a 20-mm myectomy) of the left inferior oblique muscle was performed, yielding an excellent result.

Recently, over a 6-year period, 13 patients with superior oblique muscle paresis presented with an inverted Brown pattern. Eight of these 13 patients had had a prior inferior oblique muscle recession, now with recurrence of the hypertropia and fundus extorsion in the higher eye. Twelve of the 13 patients had a tight inferior oblique muscle on exaggerated forced ductions at the time of surgery. Three of the previously unoperated patients were operated with an inferior oblique muscle recession, although one of these had to be reoperated with a denervation/extirpation. The other 10 of the 13 patients had an inferior oblique muscle denervation/extirpation, all with a good result.

In conclusion, just as the first procedure to use for a significant Brown syndrome is a weakening procedure of the superior oblique muscle, the first procedure to use for an inverted Brown pattern (with fundus extorsion and a tight inferior oblique muscle on forced ductions) is an inferior oblique weakening procedure, usually a large one—even though the inferior oblique muscle is not significantly overacting in the inverted Brown pattern.

References


Entering the In Crowd: Esotropia After Lateral Rectus Recession

M Edward Wilson MD

I. History
A. A 6-year-old presented with an intermittent exotropia of 30 PD at distance and 15 PD at near.
B. The parents had noted a progression of the frequency of out-turning from only with fatigue to almost half the waking hours.
C. Part-time patching was poorly tolerated but had been tried, with no change in the frequency or magnitude of the exotropia.

II. Examination
A. The deviation measured 30 PD at distance and 15 at near. The intermittent exotropia was comitant with no A- or V-pattern.
B. Ductions and versions were normal.
C. After 40 minutes of unilateral occlusion in the office, the near deviation increased to 25 PD. Accommodative convergence-to-accommodation (AC/A) measured using minus 2.00 lenses at distance was normal.

III. Surgery
A. Lateral rectus recessions (7 mm) were performed bilaterally.
B. Parents had been told that there might be temporary diplopia after surgery.
C. At 2 week postop, parents indicated (via scheduled postop phone call) that the child still saw double and that the eyes appeared crossed. Photos were sent via email.

IV. One Month Postoperative Examination
A. The patient was found to be 20 PD of esotropia for both distance and near. Versions were normal.
B. Cycloplegic refraction revealed +0.5 D in both eyes.

V. Two-Month Postoperative Examination
A. The patient remained unchanged and measured 20 PD of comitant esotropia. Ductions and versions were normal.
B. Plano glasses with a total of 16 PD base-out Fresnel prism were prescribed (fusion verified in prism in office).

VI. Five-Month Postoperative Examination
A. The patient remained unchanged and measured 20 PD of comitant esotropia. Ductions and versions were normal.
B. Surgery was scheduled.
C. Medial rectus muscle recession (3.5 mm) bilateral performed 4 weeks later (at 6 months after the first surgery).

VII. One Month After Second Surgery
Exophoria measured 12 PD distance and near; parents and patient were happy.

VIII. Six Months After Second Surgery
Intermittent exotropia measured 20 PD distance and near with poor control. Parents reported that the eyes drift nearly 100% of the time. A third surgery was now needed.

IX. Points for Discussion
A. Persistent overcorrection (beyond 6 weeks) after bilateral lateral rectus muscle recession surgery for intermittent exotropia is rare (5% of surgery for intermittent exotropia).1,2
B. After some period of waiting (and using hyperopic spectacles and prisms as indicated) for spontaneous improvement (6 months), surgery may be needed. When there is an abduction limitation (and normal forced duction test), lateral rectus exploration and advancement is the procedure of choice. If ductions are normal, bilateral medial rectus recession is often chosen.
C. Surgical dosage for consecutive esotropia is rarely written about or discussed. Textbooks often advise applying Cooper’s dictum (if versions are normal, treat it as a fresh case).3
D. Should surgical dosage be reduced since recidivism is so high in intermittent exotropia?

X. Take Home Point
Surgery for consecutive esotropia after lateral rectus recession for intermittent exotropia often leads to a return of esotropia – at times with less fusional control than the initial exotropia. Reduced dosage bilateral surgery or, alternatively, unilateral surgery should be considered in these rare cases of persistent overcorrection with normal abduction after bilateral lateral rectus recession.

References
The Outcast: Exotropia Following Medial Rectus
Recession

My learning curve . . . is long and steep 🙃

Jane C Edmond MD

I. Three-month-old boy with Down syndrome and infantile esotropia (ET) of 45 PD.

A. 8 months: 45 ET
   1. Low hyperopia
   2. Bilateral medial rectus (BMR) recess 5.5 mm
   3. Immediate postop: slight undercorrection

B. 17 months: 35 ET
   1. Low hyperopia
   2. Bilateral lateral rectus (BLR) resections 6.5 mm
   3. Immediate postop: slight undercorrection

C. 2 years: 40 ET
   1. +2.00 spec prescribed
   2. Refused to wear
   3. BLR re-resection 5.5 mm
   4. Immediate postop: slight undercorrection

D. 3 years: 45 ET
   1. Still won’t wear low hyperopic specs
   2. BMR re-recess, 12 mm from limbus
   3. Immediate postop: looks straight!!

E. 4 years: 45XT, with BMR underaction/adduction deficits
   1. Right medial rectus advancement with pseudo-tendon resection
   2. Found 8 mm from old insertion (not 12 mm)
   3. Tendons looked thin. Maybe I didn’t resect the floppy part completely . . .
   4. Immediate postop: looks straight!!

F. 4.5 years: 50 XT, with BMR underaction/adduction deficits
   1. Mother informs me that her whole family questions why she keeps me as her son’ physician. 🤷‍♂️🤷‍♂️🤷‍♂️
   2. Both MR found recessed, ~7 mm of pseudotendon (that was really resected), both advanced to 6 mm from limbus
   3. Immediate postop: straight

G. 6 years: 20 E(T)
   1. Increased hyperopia, +3.00
   2. Specs prescribed, worn

H. 9 years: 🤷‍♂️🤷‍♂️🤷‍♂️
   1. Ortho w +250 specs

II. The surgical approach to large (> 50 PD) angle infantile esotropia is controversial.

Surgical options:

A. Large BMR muscle recessions of 6-8 mm, regardless of the age of the child. In the United States, this is probably more popular than 3-4 muscle surgery.

B. Three or 4 horizontal rectus muscle surgery, with MR recession no greater than 5.5-6 mm

C. Less utilized in the United States
   1. Monocular recession-resection
   2. BOTOX

III. For large-angle infantile ET, large BMR recessions and 3-4 muscle surgery have been shown in the literature to have good initial success, with late overcorrections (and undercorrections).

IV. Overcorrections following surgery for large-angle congenital ET may be attributed to the following factors:

A. Younger age at time of surgery (< 15 months, more effect from recession if surgery performed on a smaller eye)

B. Amount of the MR recession (over 6 mm is considered “large”)

C. Amount of LR resection (if preforming 3-4 muscle surgery)

D. Duration of follow-up (The longer the follow-up, the more frequently overcorrections occur.)

E. Development of a stretched scar / pseudotendon at insertion of recessed MR


V. Reported Rates of Overcorrection Long Term in Infantile ET Status Post Surgery

A. Routine BMR recess

B. Large BMR recess: 27%, developing at average of 3.4 years postop BMR recess 7 mm (Stager, et al), although overcorrections reported to occur 6-45 years after MR recession)

C. 3-muscle surgery: 23%, developing at average 2 years postop, overcorrections 3x more common than late undercorrections, average follow-up 8 years (Chatzistefanou, et al.)
VI. Treatment Options for Late Overcorrections After Surgery for Infantile ET

A. Bilateral LR recess
   1. Easiest procedure to do
   2. Will be unsuccessful if MR is over-recessed or a stretched scar/pseudotendon is present

B. MR advancement alone
   1. Very good procedure (Marcon)
   2. Will be unsuccessful if MR has a stretched scar/pseudotendon


D. Unilateral LR recess + MR advancement or MR resection without advancement (no stretched scar/pseudotendon). Reported good outcomes (Mohan, et al.)

E. MR stretched scar/pseudotendon excision alone (MR not advanced). Unlikely to be unsuccessful, will need more surgery

F. MR stretched scar/pseudotendon excision and MR advancement
   1. Best surgery for this situation (Tinely, et al.)
   2. Be more aggressive than I was.
   3. For larger XT, can add LR recess

VII. Clues That You Need to Operate the MR

A. Early overcorrection after > 6 mm BMR recession (MR are too recessed)
B. Late onset overcorrection when previously well aligned (stretched scar)
C. Adduction deficit (over-recessed MR and stretched scar)
D. XT at near > distance (convergence insufficiency type XT) (over-recessed MR and stretched scar)
E. Significant undercorrection after lateral rectus resections performed
F. Tight LR on forced ductions in OR

VIII. Surgical Tips for Operating a Previously Recessed MR

A. Limbal approach gives great exposure
B. Suture material? Debate over whether to use absorbable or nonabsorbable
C. Strongly suspect stretched scar if:
   1. MR tendon seems thin.
   2. Muscle hook is visible under the tendon’s insertion.
   3. Pulling up on the muscle hook creates a linear ridge or bump in the “tendon.”
   4. Pseudotendon length: 4-12 mm

D. If stretched scar is found and tendon attachment is not over recessed, I proceed with:
   1. Complete scar excision
   2. MR advancement
   3. If unilateral, will typically correct up to 25 XT (in my hands)
   4. If XT > 25
      a. Add LR recess (I usually use less than suggested LR recess dosing suggested in the surgical tables for recession/resection for XT)
      b. Or do other MR
E. If no stretched scar is found, I proceed with:
   Advance MR, one or both, ± LR recess, depending on the size of the deviation
   1. 1 mm of MR advancement = 1 mm MR resection (Donaldson, et al.). I think advancement is more slightly powerful than a resection.
   2. 1 mm advancement = ~4 PD esoshift (Marcon, et al.). For example: 3 mm MR advancement = ~12 PD esoshift
   3. LR recess amounts are derived from R+R XT tables as above, with modifications.
F. If using adjustable sutures, slight ET is desirable (as with all surgery for XT).

References

If at First You Don’t Succeed

Edward G Buckley MD

Transpositions

Transpositions provide a dynamic force vector in the field of the transposed muscles that is not an active force, but rather a passive force, much like a spring. This passive force can be modulated by the antagonist muscle to allow movement to occur. The saccadic velocity in the direction of the transposed muscles is significantly decreased from normal, and patients still experience difficulties, especially during fast movement. There are a variety of transposition procedures to choose from, with the most popular being the full tendon transposition. Other procedures can be used in special circumstances when compromised anterior segment blood supply or previous surgeries make it necessary (Jensen, Hummelsheim, Knapp procedure).

In the full tendon transposition, the entire rectus muscle is relocated adjacent to the paralytic muscle.

Figure 1.

In the case of a sixth nerve palsy, the superior and inferior recti are transposed adjacent to the paralytic lateral rectus. The advantages of this technique include minimal rectus muscle manipulation, commonly used surgical techniques, and ease of reoperation. This procedure also can be combined with a resection and/or recession of the transposed muscles for a complicated horizontal and vertical deviation. (See Figure 1.)

Figure 2.

Several modifications to the full tendon transposition procedure have been shown to increase its effectiveness. Buckley described using a nonabsorbable suture to attach the edges of the transposed muscle to the paralytic muscle. In the case of a sixth nerve palsy, the lateral border of the superior rectus and the superior border of the lateral rectus would be sutured together. The suture is placed approximately 8 mm posterior to the insertion and incorporates approximately one quarter of each muscle. The inferior rectus and the inferior border of the lateral rectus are attached in a similar manner. This provides a slightly increased force vector by directing more of the transposed muscles laterally. Foster described a similar technique whereby the portion of the transposed muscle that is closest to the paralytic muscle is sutured to the globe, much in the same manner as in the fadenoperation at a similar location. Results using this technique also showed a greater improvement in abduction. (See Figure 2.)
The Knapp procedure is very similar to the full tendon transposition except that the muscle is reattached parallel to the lateral border of the paralytic muscle as opposed to parallel to the insertion. (See Figure 3.) The advantages and disadvantages are the same as for a full tendon transposition.

The Jensen procedure is more complicated in its surgical approach. One half of the adjacent rectus muscle is isolated and sutured to one half of the paralytic muscle. (See Figure 4.) The major advantage is the preservation of anterior ciliary vessels and anterior segment blood flow. The disadvantages include significant manipulation of the rectus muscles and complicated reoperations. Jensen procedures do not seem to generate as much abduction as full tendon transpositions.

The Hummelsheim procedure consists of taking one-half of the adjacent rectus muscle and relocating it to the insertional border of the paralytic muscle (see Figure 5). The major advantage is preservation of anterior ciliary arteries. The disadvantages include splitting the rectus muscle that causes hemorrhage and possible fibrosis, and complicated reoperations.

References

I. O. You Won
Reoperations on the Inferior Oblique

David A Plager MD

Traction testing of the oblique muscles—both superior and inferior—is an indispensable tool for helping to manage patients with complicated strabismus due to oblique dysfunction. Although traction testing of the superior oblique is informative in many cases whether the tendon has been operated previously or not, inferior oblique (IO) traction testing is most useful in cases involving reoperation of the muscle.

For instance, following inferior oblique myectomy, even if the original surgery was performed flawlessly the residual muscle can come back to cause a recurrent or residual strabismus. In almost all cases, the muscle will either reattach to the globe somewhere or in cases where the myectomy involved only excising a small section in the middle of the muscle, the 2 muscle ends can reattach to each other through a fibrous band. Either of these eventualities can cause an obvious recurrent overaction of the IO or a more subtle “inverted Brown syndrome,” as described by D Guyton.

When such a condition is suspected based on clinical exam, careful evaluation with traction testing of the IO will guide the surgeon’s operative strategy. In general, if no “band” of the IO can be felt on traction testing, surgical exploration looking for the IO will not be fruitful. On the other hand, if a definite “band” is felt on traction testing, the surgeon will find a structure to explain it. In most cases of recurrent overaction or “inverse Brown,” lysis of the band including excising all of the sub-Tenon portion of the IO is indicated. Repeat traction testing should confirm that the attachment has been nullified.
Technique for IO Traction Test

The globe is grasped with strong-toothed forceps obliquely (eg, at 4:00 and 10:00) on the left eye. The eye is pushed down and in, as if the eye is looking toward the right foot. The globe is then swept across temporally, as if to make the eye look toward the left foot. The broad band of the IO will be felt as a bump as the eye is swept back and forth from left to right and back again.

In reoperation cases where the IO has reattached to the globe or to itself, the “band” may be felt to be more prominent (tighter) than in the fellow eye or the band may be felt to be displaced from its normal position. The band is normally felt most prominently just past the midline, eg, around 5:00-5:30 on the left eye or 6:30-7:00 on the right eye (surgeon’s view).

Selected Readings


Section II: Tough Pediatric Cornea and Anterior Segment Cases—Advice From the Experts

Case Presentations

White Eye, White Corneal Lesion

M Bowes Hamill MD

Case Presentation

Called by a pediatrician from South Texas who had just examined a newborn with “a white eye.”

On Exam

The patient was a healthy 2-day-old LAM who was the product of a normal pregnancy and delivery. There was no family history of any ocular conditions or diseases.

*Vision was blink to light O.U.* The right cornea and anterior segment appeared normal to hand-held slitlamp examination. The left cornea showed an opacity that involved the central 2/3 of the cornea. There appeared to be some iridocorneal adhesions at the margin of the opacity. The pupil and lens were not well visualized. B-scan of the left eye was normal.

Red Eye, White Corneal Lesion

Sonal S Tuli MD

Case

A 10-year-old boy presented with a history of redness, pain, and blurry vision in the right eye. He was playing outdoors and thought he got something in the eye. When his mother flushed the eye, she noticed it was red and the front of the eye seemed “cloudy.” When she checked his vision, it was extremely blurry and he could barely see fingers in front of his face. They used OTC eye drops, but the next day he started complaining of a headache and increasing pain and was brought to the eye clinic.

Past medical and surgical history was unremarkable. Ocular history was significant for previous episodes of red eye (not sure which one) on a number of occasions that resolved spontaneously. Parents were not sure when the child’s last eye examination was but think it was unremarkable.

On examination, the visual acuity was counting fingers at 2 feet in the right eye and 20/20 in the left eye. The IOP was 46 mmHg. Pupillary examination was difficult in the right eye but there was no afferent pupillary defect by reverse in the right eye. Extraocular movements were full. External examination was within normal limits other than mild edema and ptosis of the right upper eyelid. Slitlamp and dilated fundus examination were completely within normal limits in the left eye. The right eye revealed diffuse erythema of the conjunctiva and a mild follicular reaction. The cornea was diffusely cloudy, especially temporally, with stromal as well as microcystic edema. There were a number of large granulomatous keratic precipitates on the endothelium.

Red Eyes, Red Lid Margins

Recurrent Red Eye

Ivan R Schwab MD FACS

History

A 9-year-old female presents with decreased vision, photophobia, and a red right eye. No history of eczema in the young girl. Family history is positive for asthma and eczema in her mother.

Exam

20/80 O.D., 20/20 O.S. with the right eye showing 2+ conjunctival injection inferiorly and 2+ PEEs on the inferior half of the ocular surface with neovascularization inferiorly. Bulbar follicles are noted. The upper tarsus has 1-2+ papillary response with scattered follicles. O.S. has similar findings but much less prominently. The iris, lens, and dilated funduscopic examination are completely normal O.U. IOP is normal.

Recurrent Red Eye

Kathryn Colby MD PhD

History and Exam

A healthy 4-year-old girl from Bermuda presented for a second opinion on intermittent recurrent redness, tearing, and photophobia in the left eye since the age of 8 months. Over the years, she had been diagnosed with staphylococcal hypersensitivity, phlyctenular keratoconjunctivitis, and limbal vernal keratoconjunctivitis. Previous therapies included topical antibiotics and topical steroids. Despite these treatments, her symptoms continued to recur. There was no history of systemic allergic disease. The right eye was asymptomatic.

On examination, she had mild blepharitis in both eyes, with unilateral conjunctivitis, extensive peripheral corneal neovascularization, and a small focus of anterior stromal haze in the left eye. Corneal sensation was reduced O.S. and was normal O.D.
Red, Itchy Eyes
Stephen C Pflugfelder MD

History and Exam
An 11-year-old boy presented with a several-year history of redness, itching, and tearing in both eyes. In the past month, he had complained of difficulty seeing the board at school, and his mother had observed swelling of his eyelids. He had a history of asthma and was otherwise healthy. He had experienced minimal improvement with olopatadine drops; however, his symptoms improved with topical prednisolone acetate.

Visual acuity was 20/50 in both eyes. He had mild bilateral upper eyelid swelling. There was edema of the inferior puncta, and the height of the inferior tear meniscus was increased. There was velvety papillary reaction, edema of the inferior palpebral conjunctiva, and marked papillary reaction of the superior palpebral epithelium. Both corneas had a vortex pattern of epithelial erosions migrating from the superior limbus to the central cornea. IOP was 12 in each eye. Lenses were clear, and the remainder of exam was normal. Dry scaly dermatitis was noted in the antecubital fossa of both arms. How would you manage this patient?

Congenital White Eyes, Turned Into Bright Eyes
Kanwal K Nischal MBBS

History and Exam
A 5 year old adopted female of Asian descent presented for a fifth opinion. She had counting fingers vision in either eye and on examination had bilateral corneal opacities with corneal vascularisation. She had focal opacities scattered randomly through the superficial cornea. There was no view of the fundus or anterior chamber. Exam under anesthesia showed a shallow anterior chamber but it was formed in each eye. Impression cytology proved to be equivocal. Despite presenting at the age of 4 months to an ophthalmologist in the USA soon after adoption, corneal transplantation had been discouraged on the grounds that there was likely limbal stem cell deficiency and that since she had no know living relatives that any limbal stem cell transplant would likely fail despite systemic immunosuppression. Evaluation of limbal stem cell presence by a novel technique together with better phenotyping allowed consideration of corneal transplantation.
Section II: Tough Pediatric Cornea and Anterior Segment Cases—Answers and Teaching Points

White Eye, White Corneal Lesion

M Bowes Hamill MD

Now what? Final Diagnosis

Unilateral Peter’s anomaly

After extensive discussion with the parents, the patient underwent a penetrating keratoplasty 2 days later.

Teaching Points

1. Peter’s is a rare condition with a variety of genetic causes.
2. The management of this condition depends on a variety of factors, including severity of the disease, parental involvement, and access to specialized care (ie, pediatric glaucoma, etc.). It’s better to do nothing and have a poorly seeing, comfortable eye than a failed graft with painful edema.
3. I believe earlier surgery (within days to weeks of birth) is preferable to later surgery (weeks to months after birth).
4. Even with aggressive surgical management and good parental involvement, the prognosis for a clear graft and “good” vision is guarded. It’s far better to have a frank discussion with the parents about reasonable visual expectations and risks of graft failure before surgery than a long explanation of a poor visual outcome later.

Red Eye, White Corneal Lesion

Sonal S Tuli MD

Diagnosis and Clinical Outcome

Based on the presence of granulomatous keratic precipitates (KPs) and overlying corneal edema along with extremely high IOP, a presumptive diagnosis of herpes simplex endotheliitis (disciform keratitis) with trabeculitis was made. Additional clues to the diagnosis were the follicular reaction and a history of unilateral red eye in the past on multiple occasions. Further direct questioning revealed that the child and both parents were prone to getting fever blisters around the mouth.

The child was started on prednisolone acetate eye drops every 2 hours along with apraclonidine and timolol eye drops twice a day each. He was also started on oral acyclovir at the appropriate dose for his weight. He returned the next day with a significant improvement in his pain. His vision was now 20/400 with an IOP of 20. He rapidly improved over the next 2 weeks and was 20/20 two weeks following the initial presentation. His IOP was in the low teens at that time, and both the pressure-lowering drugs were stopped. The prednisolone drops were tapered very slowly over the following 4 months. He was asked to continue the oral acyclovir at the prophylactic dose long term. His vision continued to be stable at 20/20 over the year of follow-up.

Teaching Points

1. Granulomatous KPs indicate a lymphocytic reaction, which should suggest a very specific list of differential diagnoses.
2. Significant corneal edema overlying KP should indicate an active involvement of endothelial cells (endotheliitis) rather than a passive deposition of lymphocytes on the endothelium.
3. Acute uveitic syndromes usually cause a low IOP due to cyclitis. An extremely high IOP should suggest trabeculitis.
4. Uveitis or endotheliitis with trabeculitis is usually herpes simplex virus (HSV), herpes zoster virus, or cytomegalovirus.
5. A very rapid response of IOP and endotheliitis to topical steroids suggests HSV.
6. Steroids, once started for stromal HSV, should be tapered extremely slowly, as rapid taper will result in recurrence.
7. Prophylactic acyclovir should be considered long term in vision-threatening HSV infections to decrease the incidence of recurrences.

Selected Readings

Red Eyes, Red Lid Margins

Recurrent Red Eye

Ivan R Schwab MD FACS

Diagnosis

Staph hypersensitivity or ocular rosacea

Selected Readings


Recurrent Red Eye

Kathryn Colby MD PhD

Clinical Course and Outcome

The history and exam was consistent with recurrent herpes simplex keratitis. She was started on oral acyclovir 300 mg 3 times a day. Her mother reported rapid improvement in her symptoms. The acyclovir was stopped 1 week later during a visit with another ophthalmologist, with recurrence of her symptoms. Acyclovir was restarted, together with 0.12% prednisolone acetate and topical azithromycin to her eyelashes. Her eye was quiet at a visit 2 months later, with regressed neovascularization. The steroid was tapered and eventually stopped. She was sent back to her local doctor on long-term acyclovir prophylaxis, which was continued for a total of 1 year. Her corneal disease remained quiescent off acyclovir.

Final Diagnosis

Atopic keratoconjunctivitis (AKC)

Teaching Points

1. Atopic dermatitis (AD) is a significant cause of ocular morbidity, with eye or eyelid involvement occurring in 20%-43% of patients. AKC is a chronic and potentially blinding condition. Diagnosis may be missed if eczema is mild and affects non-eyelid skin.
2. Corneal involvement is common, ranging from punctate corneal erosions to epithelial erosion, epithelial filaments, and ulceration. Corneal scarring and neovascularization from chronic inflammation can lead to vision loss. In a study of 20 patients with 3 or more years of follow-up, Power et al found that 70% of patients with AKC developed epithelial disease and corneal neovascularization developed in 60%.
3. Treatment of AKC is aimed at controlling symptoms, decreasing recurrence and exacerbations, and reducing vision loss. A stepwise treatment approach is advised utilizing mast cell stabilizers, corticosteroids, calcineurin inhibitors, and corneal protection. The calcineurin inhibitors cyclosporine A and FK506 (tacrolimus) inhibit many immune mediators responsible for AKC and permit reduction in frequency of topical steroids. Two randomized controlled trials using 0.05% cyclosporine A vs. placebo (either vehicle or artificial tears) reported significant improvement in AKC. In a study by Attas-Fox, topical tacrolimus 0.03% ointment applied to the conjunctival sac was similarly found to be safe and effective in patients with seasonal allergic conjunctivitis. In our experience, application of tacrolimus 0.03% ointment to the eyelid margins or conjunctival sac twice a day in 7 patients with AKC has been effective in reducing eyelid margin and conjunctival inflammation. There is currently no FDA-approved ophthalmic formulation of tacrolimus, and its use on the ocular surface is currently off-label.

4. Cornea involvement requires an aggressive approach with immunomodulatory therapy to prevent cornea opacity and vision loss.

References and Selected Readings


Congenital White Eyes, Turned Into Bright Eyes

Kanwal K Nischal MBBS

Teaching Points

1. Accurate phenotypic determination is very important in considering treatment for children with congenital or neonatal corneal opacification.
2. Newer imaging modalities can be utilized to image potential limbal stem cell presence.
3. Strict postoperative surveillance is necessary for children undergoing corneal transplants.

Selected Readings

Where Are We Now? Screening for Amblyopia in 2014 and Beyond

Sean P Donahue MD PhD

I. The Past
   A. Direct testing of acuity: Optotype-based screening
   B. Detection of risk factors
      1. Analog / Polaroid photoscreening
      2. Autorefractors (SureSight and Retinomax)

II. The Present
   A. Detection of decreased acuity
   B. Detection of amblyopia risk factors

III. The Future
   A. Detection of decreased acuity
   B. Detection of amblyopia risk factors
   C. Detection of amblyopia

IV. What We Don’t Know
   A. Prevalence of risk factors
   B. Likelihood ratios for risk factors
   C. Factors that modify risk: genetic or environmental

V. Things You Should Know
   A. American Academy of Pediatrics Photoscreening statement
   B. U.S. Preventive Services Task Force Position Statement
   C. CPT Codes
      1. 99173: acuity screening
      2. 99174 automated screening
   D. How sensitivity and specificity affect predictive value and false positive rate

VI. Which instrument should I purchase?
   Should ask: Which referral criteria should I use?

VII. Main Take-Home Message
   Choice of referral criteria is the main variable that will impact your pediatrician’s opinion about automated screening.
Amblyopia: It’s a Binocular Disorder

Eileen E Birch PhD

Amblyopia occurs when discordant images are present during visual development, usually due to strabismus and/or anisometropia. If one eye is habitually suppressed, the monocular visual acuity deficits by which amblyopia is defined soon develop. Along with amblyopia, a constellation of other visual functions may be compromised, resulting in spatial distortion, abnormal hyperacuity, fixation instability, and fine motor skill deficits.

Patching and atropine are the mainstays of amblyopia treatment. Both have been shown to be effective treatments for the monocular visual acuity loss. Over 80% of children treated with patching and/or atropine show significant improvement in visual acuity in the amblyopic eye. However, patching and atropine do not treat the binocular abnormality. That may be why only 40% of children treated with patching or atropine achieve 20/20 visual acuity and more than 35% experience a recurrence of amblyopia.

The visual cortex devotes enormous resources toward providing a binocularly integrated view of the world, combining separate inputs from the two eyes and using image disparities to calculate depth. Binocular combination and stereoacuity require balanced visual sensitivity and visual acuity, and symmetric mutual suppression. In amblyopia the balance is disrupted; the amblyopic eye provides a weak, noisy signal and little suppression of the fellow eye. The dominant eye strongly suppresses the amblyopic eye.

The dominant theory is that suppression is a necessary adaptation to avoid visual confusion and diplopia due to strabismus or anisometropia. However, converging evidence suggests that imbalanced interocular suppression plays a primary role in the genesis of amblyopia and the constellation of visual and motor deficits that accompany the monocular visual acuity deficit. Armed with this new understanding of suppression, we can appreciate that suppression may be a roadblock to successful amblyopia treatment.

Until recently, the dominant theory of visual development posited an immature primary visual cortex that failed to develop strong connections with the suppressed, amblyopic eye during a critical period of maturation. This resulted in a major shift in ocular dominance and, once the critical period ended, the amblyopic visual system was thought to be structurally monocular. We now know that this hypothesis was incorrect. Instead, the primary visual cortex (V1) matures early and has a normal distribution of ocular dominance even when infantile strabismus or anisometropia disrupts visual experience. The prevalence of binocular suppression in both V1 and V2 neurons is correlated with the depth of amblyopia. In other words, imbalanced interocular suppression renders a structurally intact binocular intact visual system functionally monocular.

The new evidence that the binocular visual system of amblyopic children is structurally intact but functionally monocular provides a foundation for a broadened scope of amblyopia research and for crafting more effective evidence-based treatments. If imbalanced interocular suppression plays a primary role in the genesis of amblyopia, can we rebalance binocular vision in amblyopia as an amblyopia treatment? Yes, by reducing the contrast of the fellow eye image we can allow the amblyopic eye to “break through” and the amblyopic child can experience binocular vision. Our recent studies have shown that we can incorporate this approach into dichoptic iPad games that the amblyopic child can play while at home wearing anaglyptic red-green glasses. Repeated binocular experience, provided by dichoptic iPad game play, was effective in improving visual acuity in amblyopic children.

Selected Readings

New Noninvasive Treatment Modalities for Amblyopia

Robert F Hess PhD

Two pieces of new information offer the possibility of recovering vision in adults who have suffered amblyopia in childhood. The first is that the adult brain is more plastic than previously thought, and the second is that loss of binocular vision is the primary problem in amblyopia.1

Noninvasive brain stimulation can be used to recover function from the amblyopic cortex in adulthood. A number of different procedures can be used, including repetitive transcranial magnetic stimulation,2 theta burst stimulation,3 and DC current stimulation.4 Improvements can occur in as little as 40 seconds and, if repeated for 3 consecutive days, can be made permanent.3

If the loss of binocular function is treated as a first step, an effective and novel behavioral therapy for adults can be demonstrated.5 This treatment involves the dichoptic presentation of stimuli designed to strengthen fusion / reduce suppression. It has been implemented on an iPod / iPad platform as a videogame and involves 1 hr/day for 4-6 weeks.6 During this time, binocular single vision is restored in over 90% of cases, stereopsis in over 60%, and improved monocular acuity in over 60%.7

We have also shown that the combination of the behavioral video game treatment with DC current stimulation results in a better outcome for stereopsis.4

What is clear is that function in amblyopic adults is not lost but suppressed, leaving open the possibility of significant recovery of vision using a variety of different approaches. At present there is no treatment for adult amblyopes in the clinic, yet a variety of different ways to recover vision have been demonstrated in laboratory studies.

References
New Medical Treatment Modalities for Amblyopia

Michael X Repka MD MBA

I. Pharmacologic Treatment of Amblyopia

A. Atropine

B. Levodopa

C. Citicoline (cytidine-5’-diphosphocoline)

D. Many pilot studies show varied visual acuity improvement with each drug. However, many of the studies are small, uncontrolled, and/or nonrandomized.

II. Atropine

One drop of atropine 1% ophthalmological solution is applied to the fellow eye at varied frequencies.

A. Daily dosing has been traditional. Such dosing was equivalent to 6 or more hours of daily patching in children 3 to 6 years of age. The improvement was maintained through age 15 years.

B. Twice weekly treatment is as effective as daily patching for visual acuity 20/40 to 20/80.

C. Addition of a plano lens increases the risk of “reverse” amblyopia. A Pediatric Eye Disease Investigator Group (PEDIG) study of the use of a plano lens for residual amblyopia has been completed, and the results are pending.

D. Twice weekly atropine will improve the amblyopia in some older children and those with more severe amblyopia. Thus twice weekly atropine would be suitable as an option, especially if compliance with occlusion in these children is problematic.

III. Levodopa

A. Levodopa is a medication used to treat adults with Parkinson disease and children with dopamine-responsive dystonia. Dopamine is a neurotransmitter that does not cross the blood-brain barrier. Levodopa, which is an intermediate in the biosynthesis of dopamine, is used as pharmacological replacement therapy as it will cross the blood-brain barrier, where it is converted to dopamine. Levodopa is combined with carbidopa. Carbidopa is a peripheral decarboxylase inhibitor that prevents the peripheral breakdown of levodopa. Concomitant administration of carbidopa reduces the dose of levodopa required by about 75%, while allowing sufficient levodopa to enter the brain for the desired central effect. The reduced dose of levodopa reduces the peripherally mediated side effects, including nausea and emesis.

B. Some improvement of visual acuity in the amblyopic eye occurs within 1 hour of medication administration and then begins to decline 5 hours after administration. Prolonged dosing has been associated with an improvement in visual acuity in younger and older children.

C. Multiple uncontrolled or small studies have reported on the outcome of levodopa.

1. In one prospective randomized controlled trial, 72 subjects with amblyopia were distributed into 3 groups of 24. Group A subjects received levodopa alone, Group B received levodopa (0.50 mg/kg/t.i.d.) and part-time occlusion (3 hours/day), and Group C received levodopa and full-time occlusion (during all waking hours) of the dominant eye. Fifty-three of 72 subjects (74%) had an improvement in visual acuity (mean: 1.6 Snellen lines, ≤ 10 years; mean: 1.1 Snellen lines, > 10 years) after treatment, 52% of those who improved had regression in visual acuity when measured after 1 year.

2. A follow-up report of 3 longitudinal studies (9 to 27 months) using levodopa (0.55 mg/kg, 3 times per day) plus occlusion for treatment of amblyopia included 30/33 subjects (91%). Those who received levodopa plus occlusion demonstrated significant regression of visual acuity after stopping the levodopa. The regression over approximately 6 months of follow-up averaged 1.4 lines. This recurrence was similar in magnitude to that experienced by those receiving occlusion only.

3. A PEDIG pilot study randomized 33 subjects 8 to < 18 years old between 2 dosages of levodopa (0.51 mg/kg and 0.76 mg/kg, both with carbidopa 0.17 mg/kg, 3 times a day) and followed them for 8 weeks. At the 8-week primary outcome, visual acuity in the amblyopic eye improved an average of 4 letters in the 15 subjects randomized to the 0.51 mg/kg dosage, and an average of 6 letters in the 17 subjects randomized to the 0.76 mg/kg dosage.

D. PEDIG has conducted (2010-2013) a randomized clinical trial of residual amblyopia in children 7 to 12 years of age comparing oral levodopa 0.76 mg/kg 3 times per day (t.i.d.) with carbidopa 0.17 mg/kg t.i.d. with oral placebo with patching 2 hours per day in both groups. Medication was continued for at least 16 weeks. Follow-up was conducted for an additional 10 weeks. Adverse events including headache and nausea were elicited. Results are expected in Fall 2014.
IV. Citicoline

A. Used for movement disorders such as Parkinson disease.

B. One amblyopia therapy protocol used intramuscular injections of citicoline (1 gm intramuscular daily) for 15 days in an open (nonrandomized, unmasked) clinical trial. Ten additional subjects were studied in a randomized double-masked design. A significant improvement in visual acuity in both the amblyopic (mean: 1.7 lines) and the fellow (mean: 1.0 lines) eyes, which remained stable for at least 4 months.12

C. Oral treatment: 61 participants (5 to 10 years of age) suffering from anisometropic or strabismic amblyopia were randomized: Group A, 800 or 1200 mg of oral CDP-choline and 2 hours patching a day; Group B, 2 hours patching a day. Both groups were treated for 30 consecutive days. A follow-up visit was set 60 days after the treatment was discontinued. The addition of CDP-choline to patching therapy was not found to be more effective than patching alone after a 30-day treatment.13

References


The time for refractive surgery in some children is now, not years from now. Why? Because for a few it is the only way out of blur-induced blindness, and for others, it is an effective way to augment treatment of amblyopia. I outline herein the compassionate-use, “off-label” application of refractive surgery to treat special subpopulations of ametropic children. A general strategy is explained, along with the benefits and risks associated with adapting adult techniques for use in pediatric eyes.

CHILDREN WITH BILATERAL HIGH AMETROPIA WHO ARE SPECTACLE NONCOMPLIANT BECAUSE OF A NEUROBEHAVIORAL DISORDER

These are the hard cases and the children with greatest need. Their spherical-equivalent myopia or hyperopia exceeds approximately 4 to 5 diopters (D). The uncorrected myopia translates to a distant acuity of 20/200 or worse (legal blindness). The uncorrected acuity for the child with hyperopia can be better during epochs when the child accommodates partially, but accommodation is often impaired in children with neurobehavioral disorders.1 The prevalence of isometropic amblyopia in children with uncorrected high hyperopia approaches 50%.2,4 Add to this the substantial risk (about 80%) of refractive esophoria or esotropia.

A variety of neurobehavioral disorders impede spectacle wear. The most common are moderate to severe cerebral palsy (ie, Gross Motor Function Classification Scale levels 3-5), autism, Angelman syndrome, more severe Down syndrome, and suboptimally controlled seizure disorders.6-10 Other causes are idiopathic developmental delay/mental retardation and progressive childhood encephalopathies. The uncorrected ametropia exacerbates the neurobehavioral disorder. The child lives in a cocoon of blur, promoting fearfulness, reduced interest in the outside world, and blunted social interactions (a constellation labeled “visual autism”).3 The extreme refractive error is seldom the only visual deficit. Common comorbidities include strabismus, amblyopia, optic neuropathies, foveopathies, and nystagmus.6-10 Comorbidities do not mean that the refractive error is unimportant or that correcting the error would not improve visual function. Too many ophthalmologists, after examining a child with an intimidating array of neurobehavioral and visuomotor deficits, succumb to the bigotry of low expectations. The uncorrected vision is dismissed as “good enough for a child with multiple disabilities.”

Why is prescribing glasses ineffective in a substantial fraction of these children? Many are aversive to wearing any head or facial appliance. They dislodge glasses repeatedly, despite heroic efforts to enforce compliance on the part of parents, teachers, therapists, and caregivers. If the battle is won to keep the glasses in place, the benefit may be negated by poor motor control of the head, neck, and trunk or gaze palsy and apraxias.5 The motor-control disorders cause chronic viewing over, around, or at the edges of spectacle lenses. Contact lenses are seldom a suitable option. Insertion and removal may require bodily restraint by multiple family members and vigorous eyelid manipulation to overcome intense blepharospasm. Repeated rounds of this exhaust—physically and emotionally—even the most committed parents.

CHILDREN WITH ANISOMETROPIA WHO ARE SPECTACLE NONCOMPLIANT AND UNSUITED TO CONTACT LENS WEAR

These children should be considered for refractive surgery if spectacle compliance is poor and the anisometropia is amblyopiogenic. Poor compliance for purposes herein can be defined as wearing glasses 25% or less (only 1 of every 4) of waking hours (a permissive boundary). Amblyopiogenic magnitudes of anisometropia can be defined as approximately 2 D for children with hyperopia and approximately 3 to 4 D for children with myopia.11,12 The prevalence of any amblyopia (defined as ≥2 optotype lines’ difference between the eyes) in preschool children who have more than 1 D of anisometropia is 66%. The prevalence of moderately severe (≥4 lines’ difference) amblyopia is 38%. Children with higher magnitudes of anisometropia have higher prevalence and greater depth of amblyopia. The prevalence and depth of amblyopia also increases with duration (ie, age). At each level of anisometropia, older preschoolers have more amblyopia than younger preschoolers.11

The majority of anisometropic children do well in glasses, and a select subset can be managed with a contact lens. But a significant mi-
majority will not wear glasses and are unsuitable for contact lens wear. This minority includes anisometropic children who have the neurobehavioral disorders discussed and neurologically normal children who do not tolerate standard correction. Intolerance can be caused by nonadaptation to spectacle anisokoria and anisosvergence, with chronic asthenopia or diplopia. For special-needs children and their parents, the glasses may represent yet another social stigma subjecting the child to ridicule, and contact lenses may be problematic. Contacts can be difficult to insert and remove, frequently lost, irritating to the eye, or too costly. In a recalcitrant subset of anisometropic amblyopic children, refractive surgery is the best treatment option.

CHILDREN WITH OTHER DISORDERS AND SPECIAL NEEDS THAT IMPEDE SPECTACLE WEAR

Refractive surgery should also be considered in certain children with issues that lie outside the categories covered earlier. Examples include children with high ametropia and significant craniofacial or ear deformities, hearing aids, a cochlear implant, or a prominent subcutaneous hydrocephalus shunt tube. The majority of these children and their families manage with specially fitted glasses and head straps, but some do not. Refractive surgery may also be considered for the rare child with high bilateral hyperopia and refractive/accommodative esotropia who exhausts all efforts (eg, repeated trials of bilateral atropine drop therapy) to enforce spectacle wear. The refractive surgery can achieve a doubly beneficial effect: reversing isoametropic amblyopia and restoring binocular fusion.

PEDIATRIC REFRACTIVE SURGERY STRATEGY AND TECHNIQUES

The surgical techniques, strategies, goals, and perioperative care used for refractive surgery in adults require key modifications for children. Beginning with the most obvious, children are more difficult to examine than adults. Obtaining the preoperative quantitative information needed often requires multiple sets of measurements by examiners skilled in examining uncooperative children, supplemented by a confirmatory set obtained during a brief examination under anesthesia. The surgical devices and techniques used, progressing from least to most invasive, are advanced surface ablation (ASA) using the excimer laser (ie, photorefractive keratectomy); implantation of a phakic intraocular lens (IOL); and clear lens extraction (CLE) with or without insertion of a posterior chamber IOL (refractive lens exchange). The following is a general strategy. Children with refractive errors +6 to −10 D are treated by ASA (laser-assisted in situ keratomileusis [LASIK]) if less advantageous, see later). Children with refractive errors beyond this range, who have anterior chamber depths 3.2 mm or more, receive a phakic IOL. The remainder usually require CLE/RLE. In the overwhelming majority of children, ASA requires general anesthesia (volatile anesthetic induction with conversion to intravenous propofol). General anesthesia is obligatory for pediatric phakic IOL or CLE/RLE surgery. All of these are performed as outpatient surgeries. The preoperative and postoperative care entail a team with pediatric experience. Detailed descriptions are available.

GAINS IN VISUAL ACUITY AND VISUAL FUNCTION

Is refractive surgery in children effective? Yes, with a qualification. The relevant measure of effectiveness in children noncompliant with spectacles is uncorrected visual acuity (UCVA). Work from our group, and that of other investigators, shows substantial gains in UCVA using either ASA, phakic IOLs, or CLE/RLE. Impressive gains are achieved in children with bilateral ametropia. Modest but consistent gains are achieved in the amblyopic eyes of children with anisometropia. The qualification is that tolerances are wider in pediatric refractive surgery; seldom does one achieve the precision commonplace in adult surgery.

In children with bilateral ametropia averaging 7.1 D (and visuomotor comorbidities) treated using ASA, the average UCVA gain was 13-fold, from a mean 20/810 to a mean 20/60. In the subset of these children who would wear glasses during testing, the gain in best-corrected visual acuity (BCVA) was an average 2-fold. In children with ametropia averaging 15 D (and visuomotor comorbidities) treated by phakic IOL implantation, the average UCVA gain was 60-fold, from a mean 20/3400 to a mean 20/57. The average BCVA gain was 2-fold. Similar UCVA (100-fold) and BCVA gains have been reported in children with bilateral ametropia, averaging 19 D, treated by CLE/RLE. The majority of reports on pediatric refractive surgery have described using ASA to treat anisometropic amblyopia. These case series show a reliable response, ie, initial correction of large refractive error to ±1.5 D of emmetropia in approximately 90% of treated eyes. The gains in UCVA or BCVA range from mild to excellent (2-7 lines’ improvement), with no reported losses of acuity. One-half or more of the children treated have improved binocular fusion and stereopsis.

Beyond the gains measured in office testing of acuity or binocularity, refractive surgery has also been shown to have positive effects on children’s day-to-day visual function. Enhanced visual awareness, attentiveness, and social interactions have been reported in approximately 80% or more of children treated for high bilateral ametropia. When measured using validated visual function questionnaires, scores for eye contact, tracking, finding and avoiding, judging depth and distance, and reading improved by an average 73% in children with bilateral ametropia and 58% in children with anisometropia.

COMPLICATIONS AND CAVEATS

Advanced Surface Ablation

Advanced surface ablation is safe in children, if safety is gauged as a negligible rate of sight-threatening com-
Phakic IOL

Phakic IOL implantation is less subject to significant regression and may be considered the preferred method currently for surgical correction of pediatric myopia and hyperopia beyond the range of ASA.5 The anterior chamber depth required for an iris-enclaved IOL precludes the use of this lens in many children younger than 3 years. Children who have high lenticular myopia after retinopathy of prematurity may also be unsuitable because of their shallow chambers (arrested anterior segment growth).59 The major concern with use of any phakic IOL in a child is the long-term effect on the corneal endothelium. Experience to date indicates that endothelial cell loss is low,8,26,27,31 no greater than that reported in adult implantation. However, the data here are thin.8 Few children have been implanted with phakic IOLs, and accurate endothelial cell counts are difficult to obtain in the children who may benefit most from implantation.8 Any refractive surgery procedure, including ASA or CLE/RLE, can be expected to cause some reduction of endothelial cell density. What we do not know and need to know is the comparative loss. Posterior chamber phakic IOLs have also been implanted in children.38-30 Because these implants lie immediately adjacent to the iris pigment layer and lens, they pose greater risk for pigment dispersion and cataract formation over the long-term in a pediatric eye.

CONCLUSIONS

The majority of children with high ametropia—unilateral or bilateral—do well with glasses or contact lenses, but a minority do not. Pediatric refractive surgery fills an important need for this minority. The surgery deserves debate, careful scrutiny, full disclosure of potential benefits and risks, and candid reporting of successes and failures. What we know from experience to date is that select children in need benefit substantially, and often dramatically, from refractive surgery.

Submitted for Publication: March 10, 2008; final revision received October 1, 2008; accepted October 10, 2008.

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Financial Disclosure: None reported.

Funding/Support: This work was supported by a Walt and Lilly Disney Award for Amblyopia Research from Research to Prevent Blindness and a Gustavus and Louise Pfeiffer Foundation Award (Dr Tychsen).

REFERENCES

The Pediatric Eye Disease Investigator Group and Amblyopia: Where We Have Been and Where We are Going

David K Wallace MD MPH

I. Occlusion vs. Atropine
   A. Randomized clinical trial (RCT) enrolled 419 children 3 to < 7 years.
   B. After 6 months, mean improvement was 3.16 lines in occlusion group and 2.84 lines in atropine group.
   C. Mean difference was 0.34 lines (0.05 to 0.6).

II. Occlusion Dosage Studies
   A. Moderate amblyopia
      1. RCT of 2 vs. 6 hours prescribed patching
      2. After 4 months, both groups had improved 2.4 lines.
   B. Severe amblyopia
      1. RCT of 6 hours vs. full-time prescribed patching
      2. After 4 months, 6-hour group improved 4.8 lines and full-time group improved 4.7 lines.

III. Amblyopia Treatment in Older Children and Teenagers
   A. 507 subjects 7 to < 17 years
   B. RCT of optical correction with vs. without occlusion (and atropine for < 13 years)
      C. For age 7 to < 13, 20/25 or better VA was achieved after 6 months by 36% with augmented treatment vs. 14% with optical correction alone (P < .001).
      D. For age 13 to < 17, there was no significant difference between groups.
      E. If 13 to < 17 and no prior treatment, augmented treatment was superior to optical correction alone.

IV. Spectacles for Amblyopia
   A. 84 children age 3 to < 7 wore spectacles alone until no improvement between 2 visits, 5 weeks apart.
   B. 77% had improvement of BCVA of at least 2 lines.

V. Occlusion vs. Spectacles Alone
   A. RCT of 2 hours prescribed daily patching vs. spectacles alone in children 3 to < 7 years
      B. After 5 weeks, mean improvement was 1.1 lines in the occlusion group vs. 0.5 lines in the spectacles group (P = .006).

VI. Near Activities for Amblyopia
   A. 425 children age 3 to < 7 years
   B. RCT of near vs. distance activities while patching 2 hours per day
      C. After 8 weeks, mean improvement was 2.6 lines in the distance group and 2.5 lines in the near group; mean difference was 0.1 line (-0.3 to 0.3).

VII. Increasing Patching for Amblyopia
   A. 169 children age 3 to < 8 who had stopped improving with 2 hours of daily occlusion
   B. RCT of increasing occlusion dosage to 6 daily hours vs. continuing with 2 hours
      C. After 10 weeks, mean improvement was 1.2 lines in the 6-hour group and 0.5 line in the 2-hour group (P = .002).

VIII. Levodopa for Amblyopia
   A. 139 children age 7 to < 12 with residual amblyopia following occlusion therapy
   B. RCT of oral levodopa vs. placebo while patching 2 hours per day
      C. After 18 weeks, mean improvement was 1.0 line (5.2 letters) in the levodopa group vs. 0.8 line (3.8 letters) in the placebo group (P = .06).

IX. A New Approach: Binocular Therapy for Amblyopia
   A. Ongoing RCT comparing occlusion to binocular treatment
   B. 512 subjects age 5 to < 17 years will be enrolled.
   C. Children wear red-green glasses and play a Tetris-like game (Hess falling blocks).
   D. Contrast levels in the fellow eye are adjusted to optimize combination of visual information from both eyes and overcome amblyopic eye suppression.
Minimally Invasive Strabismus Surgery

Video Presentations of Basic and Advanced Principles

Jon Peiter Saunte MD and Daniel S Mojon MD

Introduction
Minimally invasive surgical techniques allow reduced trauma to the tissues and enhanced postoperative quality of life. In ophthalmology, many minimally invasive procedures have been developed—for example, for cataract, glaucoma, vitreoretinal diseases, the lacrimal system, and lids. In strabismus surgery, several muscle access techniques have been described to reduce the conjunctival incision size, since it is one of the major components influencing the postoperative quality of life, cosmesis, and later the function of the operated muscle. For rectus muscles, many surgeons use the limbal approach first described by Harms in 1949. Other access techniques have been described by Swan and Talbot, Parks, Velez, and Santiago et al. In previous studies, the term “minimally invasive strabismus surgery technique” (MISS) has been proposed for all operations that significantly reduce tissue trauma.

Studies have showed that primary rectus muscle recessions and plications performed through keyhole openings are feasible and induce less trauma than those with the usual limbal opening. The operation is performed by applying 2 small radial cuts along the muscle insertion. After muscle separation from surrounding tissue, a recession or a plication is performed through the resulting tunnel. MISS patients have been shown to have better visual acuity and less lid swelling the day after surgery, indicating that the technique is superior in the immediate postoperative period. A second study found that repeat rectus muscle surgeries—recessions, advancements, and plications—also are feasible through 2 small perimuscular cuts. MISS repeat rectus muscle surgery also reduces postoperative swelling and avoids limbal openings, which might further increase perilimbal scarring and lead to a permanent visible redness. Minimal dissection techniques can also be used to perform graded inferior oblique recessions and retroequatorial rectus muscle fixation sutures. Rectus muscle transpositions might be indicated for strabismus associated with complete paralysis. They generate new force vectors that can replace the weak muscle. So far, partial or full rectus muscle transpositions are usually performed through large limbal openings. This often produces considerable conjunctival and lid swelling during the immediate postoperative period and may even lead to corneal surface problems. This can now be performed by MISS openings.

The Surgical Technique for MISS

The whole surgical procedure is performed under the operating microscope under general anesthesia. All surgical steps can be performed by oneself, so there is no need for an assistant. First, a limbal traction suture (Silkam 6-0 or Safil 6-0, B Braun Medical; Seesatz, Switzerland) is applied to rotate the eyeball away from the field of surgery. During surgery, direct contact of the traction suture with the cornea has to be avoided. Then, 2 small radial cuts are performed, one along the superior and the other along the inferior muscle margin. The anterior margin of the cut is at the level of the tendon insertion.

The size of the cuts will depend on the amount of muscle displacement that has to be achieved. As a rule of thumb, the opening size will be 1 mm less than the amount of muscle displacement that has to be achieved. For example, a recession or plication of 4 mm can be performed through two 3-mm openings. Usually, for sizes > 5 mm, an opening of 2 mm less than the amount is sufficient. In patients with reduced elasticity of the conjunctival tissue, slightly larger openings will be necessary.

With blunt Wescott scissors using the 2 cuts for access, the episcleral tissue is separated from the muscle sheath and the sclera. When the borders of the muscles have been identified, the muscle is hooked. Then, a meticulous dissection of the check ligaments and intramuscular membrane is performed. This dissection is performed 6-7 mm backward to the insertion. The resulting tunnel allows a recession or plication to be easily performed. We performed plications; however, the tunnel also allows resections.

To perform a recession, 2 sutures (Vicryl 7-0, Ethicon; Spreitenbach, Switzerland) are applied to the superior and inferior borders of the muscle tendon as close as possible to the insertion. Then, the tendon is detached using a Wescott scissor. If necessary, hemostasis is performed. After measurement of the amount of recession, the tendon is reattached with the 2 sutures to the sclera. The tendon has to be stretched to avoid the middle part of the muscle bowing backward.

To perform a plication, 2 sutures (Vicryl 7-0, Ethicon; Switzerland) are applied to the upper and lower borders of the muscle at the distance from the tendon insertion site corresponding to the plication amount. Then the sutures are passed at the superior and inferior tendon insertions. An iris spatula is inserted between the tendon and the sutures and the muscle is plicated. The surgical procedure is finished by applying 2 sutures (Vicryl Rapid 8-0, Ethicon) to each of the 2 small cuts.

At the end of surgery, TobraDex ointment (1 mg dexamethasone and 3 mg tobramycin per gram of 0.5% chlorobutanol) is applied. No eye patch is used. For the first 2 weeks after surgery, the following treatment is prescribed: TobraDex suspension (1 mg dexamethasone and 3 mg tobramycin per ml of 0.01% benzalkonium chloride) 3 times daily and TobraDex ointment in the evening.

Conclusion
MISS is a gentler way to perform strabismus surgery and can be used for most types of strabismus surgery. A conjunctival opening situated at a reasonable distance from the cornea should decrease the incidence of corneal dellen formation, avoid a prolapse of the Tenon capsule, and minimize postoperative discomfort. There is also increasing evidence that nonlimbal strabismus surgery affects less perilimbal blood supply and may safeguard anterior segment ischemia in high-risk patients. However, because of the low incidence of such complications, only larger studies with more statistical power will be able to show whether such complications will be less frequent with the new technique.
MISS seems to be superior in the immediate postoperative period as, compared with controls, the visual acuity decrease on Day 1 was much less pronounced and no abnormal lid swellings could be seen. In the immediate postoperative period, patient discomfort has been shown to be reduced. After 6 months, no significant difference has been found between MISS and the control group for final alignment, binocular single vision, other visual acuities, refractive changes, number of allergic reactions, and dellen formation. No increased conjunctival redness could be seen in patients operated by MISS. Only a minimal cicatriziation was found along the incision lines, which did not hinder free movement of the conjunctiva over the sclera and muscle. Sometimes even biomicroscopically it was not possible to see the scars after MISS. It could be hypothesized that this minimal scarring might facilitate reoperations.

Why MISS? Is the surgery worth the effort?

Standard strabismus surgery by perilimbar conjunctival opening causes postoperative ocular irritation and scarring. The MISS approach is a useful advanced technique when performing strabismus surgery, but it has a steep learning curve for the surgeon. The MISS technique is more time consuming than traditional strabismus surgery. MISS produces less congestion and foreign body sensation at 2-3 weeks, and less redness at 6 weeks, compared to limbal openings for strabismus surgery. The method can be used by experienced surgeons for both simple and complicated cases. In patients with dry eyes requiring strabismus surgery, MISS may be a better choice than the limbal approach. The postoperative results after MISS are markedly better, with decreased patient discomfort and better cosmetic outcomes, as the conjunctiva heals with almost invisible scarring—and yes, MISS is worth the extra effort for the surgeon.

In this talk, surgery videos of basic and advanced MISS surgery techniques will be presented.
A New Twist on the Inferior Oblique

Sonal R Farzavandi FRCS

CASE

An 11-year-old girl presented with a left eye Marcus Gunn jaw winking ptosis. The parents were keen on surgery. The child had 20/20 vision in each eye, no strabismus, and excellent binocular function.

Surgical Plan

Left eye extirpation of the levator palpebrae superioris and fascia lata brow suspension

Intraoperative

- Skin incision
- Dissection to reflect the tarsus
- Levator muscle isolated above the Whitnall ligament and cut
- Inadvertently the superior oblique was transected. Cut ends identified and sutured with 5-0 Mersilene.
- Surgery completed with fascia lata brow suspension.

Postoperative

One week later she presented with symptomatic vertical and torsional diplopia, which she attempted to fuse with a right head tilt.

Examination

Left hypertropia 20 PD, increasing in right gaze to 25 PD and reducing in left gaze to 3 PD.

Head tilt test

- Head tilt to right: Orthotropia
- Head tilt to left: Left hypertropia, 25 PD

Double Maddox rod test

- Excyclotorsion in primary position, 8 degrees
- Excyclotorsion in downgaze, 13 degrees

Surgical options

1. Left eye inferior oblique anteriorization
2. Left eye Harada Ito
3. Combined left eye inferior oblique anteriorization and Harada Ito
4. Recess inferior rectus of the right eye (parents refused surgery on the right eye)

Main concern was would any of the above options completely correct the excyclotorsion?

What did I finally do?
Plications for Strabismus Surgery

Joseph L Demer MD PhD

I. Extraocular Muscle Shortening Techniques
   A. Resection
      1. Traditional method taught and practiced in Western Hemisphere
      2. Excises tendon and muscle anterior to new scleral insertion of posterior muscle
   B. Plication
      1. Long practiced in Europe
      2. Creates new scleral insertion of posterior part of muscle (not muscle-to-muscle)
      3. Leaves folded anterior tendon and muscle in situ

II. Surgical Planning of Plication Dose
   A. Equivalent mm for mm to dose effect of resection
   B. Adjustable plication can be performed similarly to resection.

III. Disadvantages of Resection
   A. Surgical trauma and inflammation due to tenectomy and myectomy
   B. Pain from application of clamp or cautery
   C. Ciliary arterectomy
      1. Interrupts ciliary circulation, probably permanently
      2. Prone to bleeding from cut muscle
   D. Potential for muscle loss if sutures break

IV. Advantages of Resection
   A. Tendon and muscle specimen are obtained. (But how often is this microscopically examined?)
   B. No lump of redundant anterior tendon and muscle remain.

V. Advantages of Plication
   A. Does not involve vessels
      1. Does not divide ciliary vessels running in muscle
      2. Iris angiography suggests immediate maintenance of ciliary circulation.
      3. Avoids intraoperative bleeding
   B. Less tissue trauma
   C. Reversible in immediate postoperative period
   D. No potential for lost muscle; breakage of all sutures returns anatomy to preoperative state.
   E. Not ugly: Clinically significant tissue elevation results. Even an 8-mm inferior rectus plication in a child heals to look like an unoperated muscle after 6 months.
   F. Not painful: No more complaints than after resection, possibly fewer

VI. Technique for Rectus Plication
   A. Fixed plication
      1. Conjunctival incision of your choice: limbal, Swan, fornix, etc.
      2. Isolate the tendon on 2 muscle hooks.
      3. At the desired mm distance from the insertion, lock 1 single-armed, absorbable suture at each muscle margin.
      4. Pass each suture partial thickness through the sclera at the corresponding pole of the scleral insertion.
      5. Using a thin instrument (eg, iris sweep), fold the anterior tendon posteriorly against the globe.
      6. Tie each suture so that the anterior fold lies against the original insertion.
   B. Adjustable plication
      1. Conjunctival incision at the insertion: Swan works well.
      2. Isolate the tendon on 2 muscle hooks.
      3. At the desired mm distance from the insertion, tie 1 double-armed, absorbable suture in the muscle center, and lock each end at the muscle margins.
      4. Pass each suture partial thickness through the sclera at the corresponding pole of the scleral insertion.
      5. Using a thin instrument (eg, iris sweep), fold the anterior tendon posteriorly against the globe.
      6. Tie the sutures temporarily so that the anterior fold lies against the original insertion.
      7. If there is an overcorrection, suture loosening permits partial or complete postoperative release of the plication.
      8. Of course, you cannot increase the effect of the plication by adjustment, so consider this in dose determination.
   C. Mini-plication (Kenneth Wright MD)
      1. Absorbable suture secured to central 3 to 4 mm of the muscle belly 5 mm posterior to the insertion
2. Suture passed through the sclera just anterior to the muscle insertion to plicate the central portion of the muscle.

3. Fixed procedure achieves 5-6Δ effect.

D. Minimally invasive plication (Daniel Mojon MD)

1. Similar to fixed plication

2. But performed under the conjunctiva through small slit incisions paralleling insertion ends, and muscle margins at the point of suture placement

3. Requires use of operating microscope and takes additional time

4. Reduces postoperative inflammation in expert hands

VII. Advantageous Situations for Plication

A. Minimizes risk of anterior segment ischemia for operation of several rectus muscles

B. Topical anesthesia

C. When the option of reversibility is important

VIII. Reoperations

A. Early: Tendon and muscle simply unfold in the first few days postoperatively.

B. Late: Plicated tissues become irreversibly fused together.

C. After suture dissolution, the plication does not spontaneously unfold.

D. For further shortening at reoperation, you can plicate again, or resect as desired.

Selected Readings


Grant Support: USPHS National Eye Institute EY-08313, and Research to Prevent Blindness. No financial conflict of interest exists.
Strabismus Surgery After Scleral Buckle Procedure

Scott R Lambert MD

I. Scleral buckling vs. Pars Plana Vitrectomy
A. In the 1950s, Schepens/Lincoff introduced scleral buckling as a treatment for rhegmatogenous retinal detachments (RD).
B. In the 1970s, Machemer developed the pars plana vitrectomy procedure.
C. Vitrectomies have increased 80%, while scleral buckling procedures have decreased 70% between 1997 and 2010 in the United States (Medicare).
D. Scleral buckling is still the “gold standard” for uncomplicated rhegmatogenous RD.1
1. European Vitreo-Retinal Society recently compared the treatment of uncomplicated rhegmatogenous RDs with scleral buckling vs. vitrectomy in 4179 patients from 48 countries.
2. Final failure rate was lower with scleral buckling than vitrectomy (SB: 0.5% vs. Vtx: 1.2%; \( P = .04 \)).2

II. Complications
A. Vitrectomy
1. High rate of late failures due to new breaks, missed breaks and proliferative vitreoretinopathy (PVR)
2. Cataracts develop in 80% of phakic eyes.
3. PVR: 3%-11%
4. More expensive than scleral buckling
B. Scleral buckle
1. Strabismus reported in 3.8% of patients after scleral buckling procedure; most treated with prismatic correction or occluder lens.3
2. Induced myopia
3. Band/explant exposure
4. Ptosis
5. Anterior segment ischemia

III. Emory Experience
A. Medical records of 23 patients who underwent strabismus surgery following a scleral buckling procedure were reviewed.4 Successful motor outcome was defined as a horizontal deviation ≤ 10 PD and a vertical deviation ≤ 4 PD.
B. Results
1. Mean patient age: 48 years (range: 14 to 67 years)

IV. UCLA Experience
A. Medical records were reviewed for 25 patients who underwent strabismus surgery following a scleral buckling procedure.5
1. Successful motor outcome was defined as a horizontal deviation ≤ 10 PD and a vertical deviation ≤ 4 PD.
2. Successful sensory outcome was defined as no diplopia in primary position.
B. 25 patients were evaluated. Successful motor outcome was achieved in 36% of patients after 1 surgery.
C. After multiple surgeries (1.8 operations/patient), motor success was achieved in 72% of patients; sensory success was achieved in 62% of patients with preoperative fusion potential ($n = 21$).

D. Scleral buckle was removed in 15/25 patients; there was no correlation between motor or sensory outcome and buckle removal ($P = .60$).

E. No RD occurred after scleral buckle removal.

V. Case Study 1

A. 38-year-old pharmacist with vertical diplopia x 18 years. Scleral buckling procedures at age 20 years O.S. and 21 years O.D. Constant diplopia since surgery on right eye.

B. Preoperative exam

1. VA: 20/30 O.U.
2. M: O.D. -11.50 +2.50 x 75; O.S. -10.50 +1.75 x 65
3. Motility: 25Δ RHT, 10Δ XT, -2 limitation of infraduction O.D.
4. SLE: Scleral buckle exposed inferiorly O.D.
5. Fundus: Low encircling scleral buckle, retina attached O.U.

C. Strabismus surgery / scleral buckle removal

1. Joint surgery with retinal surgeon
2. Exposed scleral buckle removed, inferior rectus muscle found to be inserted behind a large silicone explant, advanced to original insertion
3. Right superior rectus muscle recessed 5 mm using adjustable suture, recessed an additional 10 mm postoperatively

D. Postoperative exam (5 months)

Orthotropic, no diplopia in primary position, residual -2 infraduction O.D.

VI. Contraindications for Scleral Buckle Removal

A. Presence of vitreous traction
B. Presence of retinal tear
C. Failure to identify retinal breaks at initial RD surgery
D. Increased number of quadrants of RD prior to retinal reattachment
E. Short interval between primary RD repair and buckle removal

VII. Conclusions

A. Consider scleral buckle removal if strabismus appears to be restrictive.
B. Refer to retinal surgeon to determine if it is safe to remove scleral buckle.
C. I prefer to have the retinal surgeon remove the scleral buckle prior to strabismus surgery—in some cases strabismus has resolved with buckle removal alone.

References

The Short Tag Noose Adjustable Suture
Late, In-Office Suture Adjustment

David G Hunter MD PhD

Overview
The short tag noose adjustable suture is a small sliding noose that tucks under conjunctiva and need not be tied down, allowing for repeated or delayed suture adjustment days after the initial strabismus surgery. In this presentation I will use brief videos to describe the surgical procedure and an in-office adjustment 3 days postop.

Operative Procedure
The surgeon places a standard sliding noose adjustable suture. The noose and pole sutures are trimmed to about 5 mm in length. The short “tags” are then tucked under the conjunctiva, which is left partially open (5-10 mm) to preserve access.

Timing
There are many options for adjusting (or not adjusting) patients: In the OR (topical cases); in the recovery room; 3-7 days later. I do not schedule a visit for these patients; instead, we stay in touch by phone or text message and I bring them in only if they have concerns. Delayed adjustments will take 15-45 minutes to complete.

Preparation
Anxious patients may benefit from Ativan (1-2 mg, PO or sublingual)—first dose prior to leaving home, second dose on arrival. Consider oxycodone and ibuprofen on arrival as well; patients often develop a postadjustment headache.

Topical Meds
I use topical proparacaine and phenylephrine, followed either by lidocaine 2% or 3.5% gel. I also keep 2% lidocaine in a syringe on cannula available.

Observations/Pearls
Conjunctival incision will tease open, no incision required. The muscle must move freely when the pole sutures are pulled. If not, it is still adherent to globe, and it cannot be adjusted until freed up with a muscle hook. The sutures do not slide as smoothly after 1-2 days, it is more of a “quantum” adjustment of 1-2 mm at a time.

Table 1. Adjustable Suture Tray Contents

<table>
<thead>
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<th>Item</th>
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<tr>
<td>Westcott tenotomy scissors, blunt</td>
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<tr>
<td>Straight tying forceps</td>
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<tr>
<td>0.3-mm nonlocking toothed forceps</td>
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<tr>
<td>Straight needle holders, 1 standard and 1 delicate</td>
</tr>
<tr>
<td>Stevens tenotomy hook</td>
</tr>
<tr>
<td>Randolph cyclodialysis cannula</td>
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<tr>
<td>Calipers</td>
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<td>Conway lid retractor</td>
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Oculocardiac Reflex
Many patients are prone to this, especially at the beginning (suture retrieval) or in the middle (freeing up muscle). Be constantly vigilant and stop the procedure at the earliest sign (feeling “hot,” mild diaphoresis, feeling “funny,” change in demeanor). With sufficient attention to these signs and prompt interruption of the procedure, syncopal spells can be avoided.

Conclusion
Delayed suture adjustments for any one patient can be easy or difficult to perform. Factors influencing the ease of adjustment include the patient’s propensity to scar, access to sutures (eg, medial vs. lateral rectus muscle), the number of days that have elapsed since surgery, and the patient’s overall level of anxiety. The option adds another dimension of complexity to the logistics of using adjustable sutures. The procedure, while not pleasant, is tolerable, and it makes it possible to rescue the occasional patient from what would have almost certainly been a poor outcome.

References
Superior Oblique Z-Tenotomy

David G Morrison MD

Superior oblique Z-tenotomy can be used to treat A-pattern strabismus and over-depression in adduction. In this video presentation, the technique for superior oblique Z-tenotomy is demonstrated. A fornix-based conjunctival incision is created, and the superior oblique tendon is isolated. The tendon is spread between 2 muscle hooks. A needle tip Bovie cautery is then used to make 50% partial thickness cuts on opposite sides of the superior oblique tendon, about 8 mm apart from each other. The tenotomies are then enlarged to a 60%-70% total thickness to titrate the desired effect.

In our cohort of 20 patients, 90% had resolution of over-depression in adduction, with an average pattern correction of 16 PD. Three patients developed small, vertical deviations postoperatively that were not considered clinically significant. One patient developed strabismus consistent with superior oblique palsy.
Telemedicine and ROP: Ready for Prime Time?

Graham E Quinn MD on behalf of the e-ROP Cooperative Group

Purpose

To describe a telemedicine system for the detection of serious acute-phase ROP and the implications of the e-ROP study.

The present strategy for identifying infants needing treatment for ROP relies on repeated eye examinations of at-risk infants, few of whom require treatment. Retinal imaging by nonphysicians with remote image interpretation by nonphysicians may provide a more effective, accessible, and efficient strategy than that currently in place.

The results of the e-ROP study provide important information about the value of an ROP telemedicine system. Using a systematic approach with trained imagers and graders with quality management can decrease the burden of examinations on infants, with sufficient identification of those infants that require more intensive eye care. There is an urgent need to develop programs for detection of serious ROP that efficiently and effectively identify at-risk infants worldwide and that reduce the reliance on examinations by ophthalmologists.

Selected Readings


Grant identification: NIH/NEI U10 EY017014
2014 Advocating for Patients

Bradley C Black MD

Ophthalmology’s goal in protecting quality patient eye care remains a key priority for the American Academy of Ophthalmology (the Academy). All Eye M.D.s should consider their contributions to the following three funds as (a) part of their costs of doing business and (b) their individual responsibility in advocating for patients:

- Surgical Scope Fund (SSF)
- OPHTHPAC® Fund
- State Eye PAC

Your Eye M.D. colleagues serving on the Academy’s Secretariat for State Affairs commit many hours on your behalf while strategizing and collaborating with state ophthalmology society leaders to ensure the success of Surgery by Surgeons. Their ultimate goal—protecting quality patient eye care in the states—requires a robust Surgical Scope Fund, and we need every single Eye M.D. to step up to the plate and deliver with their checkbooks.

The Academy’s federal advocacy arm works to protect ophthalmology practices from payment cuts, burdensome regulations, and scope of practice threats, as well as to advance the profession by promoting funding for vision research and expanded inclusion of ophthalmology in public and private programs. It is critical for our OPHTHPAC Fund to also be strong.

Surgical Scope Fund

The Surgical Scope Fund (SSF) provides grants to state ophthalmology societies to support their legislative, regulatory and public education efforts. Since its inception, the Surgery by Surgeons campaign, in partnership with state ophthalmology societies and with support from the SSF, has helped 31 state/territorial ophthalmology societies reject optometric surgery proposals.

2014 has proved to be a challenging year, with several battleground states facing major optometric surgery initiatives. A number of state ophthalmic societies benefited from SSF disbursements and were able to successfully implement patient safety advocacy campaigns to defeat attempts by optometry to expand its scope of practice to include surgery. The Nebraska Academy of Eye Physicians and Surgeons was successful in its patient advocacy and public education efforts to derail legislation that would have granted optometrists the authority to perform eyelid surgery and injections. Additionally, the Arizona Ophthalmological Society succeeded in protecting patients by stopping legislation that would have allowed optometrists to gain authority to perform injections. The SSF is also at work assisting ophthalmic societies with their efforts to protect patients in California, Delaware and Massachusetts.

Proactively, the Georgia Society of Ophthalmology introduced a bill that would establish a formal definition of “surgery” into state law. While the legislative session expired before the bill could advance, Georgia ophthalmologists will be back in 2015 in an effort to pass this important safeguard for their patients.

2014 was certainly not without its challenges. Despite a vigorous battle for patient safety on the part of the Tennessee Academy of Ophthalmology, the Tennessee Medical Association and the Academy, the legislature passed a bill allowing optometrists to inject anesthesia into the eyelids. Previously, optometrists were authorized to perform only therapeutic injections and any surgical procedure that required no more than a topical anesthetic. And in Louisiana, the Academy, the Louisiana Ophthalmology Association and the Louisiana State Medical Society vigorously opposed legislation that would authorize optometrists to perform certain scalpel and laser surgeries and injections. On June 1, 2014, Louisiana Governor Bobby Jindal signed into law a laser surgery bill that will allow optometrists to perform scanning laser trabeculoplasty and argon laser trabeculoplasty glaucoma surgery procedures, as well as YAG capsulotomy surgery procedures, with the completion of as little as 32 hours coursework. The Academy’s Secretariat for State Affairs knows from past experience that with this success in Louisiana, organized optometry will push hard in 2015 to see if they can gain additional surgery states. This is why everyone must “advocate for patients,” engage in the state political process and aggressively support the SSF.

California, Delaware and Massachusetts remain “in play” and are still faced with active O.D. surgery legislation. When it comes to state legislation of any kind, California and Massachusetts are often considered bellwether states for the rest of the nation. Now more than ever, your contribution to the SSF is needed as a critical tool of the Surgery by Surgeons campaign to protect quality surgical care for our patients. The Academy relies not only on the financial contributions to the SSF from individual Eye M.D.s and their business practices, but also on the contributions made by ophthalmic state, subspecialty and specialized interest societies. The American Association for Pediatric Ophthalmology and Strabismus (AAPOS) contributed to the Surgical Scope Fund in 2013, and the Academy counts on its contributions in 2014.

OPHTHPAC® Fund

OPHTHPAC is a crucial part of the Academy’s strategy to protect and advance ophthalmology’s interests in key areas, including physician payments from Medicare as well as protecting ophthalmology from federal scope-of-practice threats. Established in 1985, today OPHTHPAC is one of the largest and most successful political action committees in the physician community. In the past, Politico highlighted OPHTHPAC as one of the most successful PACs in strategic giving. By making strategic election campaign contributions and independent expenditures, OPHTHPAC helps us elect friends of ophthalmology to federal leadership positions, ultimately resulting in beneficial outcomes for all Eye M.D.s. For example, in the 2012 election cycle, OPHTHPAC was able to help retain 20 physicians in Congress. Among the significant impacts made by OPHTHPAC are the following:

- Prevented onerous national patient prescription requirements for compounded drugs and preserved access to most ophthalmic compounded drugs for office use
Advocating for Patients

2014 Subspecialty Day | Pediatric Ophthalmology

Averted significant cuts to Medicare payments due to the Sustainable Growth Rate (SGR) formula

Protected practice expense increases for ophthalmology when other specialties sought legislative carve-outs

Protected ophthalmologists’ ability to provide in-office diagnostic testing without triggering self-referral violation

Prompted congressional action that helped reduce ophthalmology’s multiple procedure payment reduction

Secured appointment of full-time ophthalmology national program director in the U.S. Department of Veterans Affairs

Provided further exemptions from both the Electronic Prescribing and Meaningful Use EHR penalties

Leaders of AAPOS are part of the Academy’s Ophthalamic Advocacy Leadership Group (OALG), which has met for the past seven years in January in the Washington, D.C., area to provide critical input and to discuss and collaborate on the Academy’s advocacy agenda. The topics discussed at the 2014 OALG meeting included a focus on the collaboration needed among the Academy and its OALG partners on the issue of compounding. As a 2014 Congressional Advocacy Day (CAD) partner, the AAPOS and the AAP-Section on Ophthalmology ensured a strong presence of pediatric specialists to support ophthalmology’s priorities as nearly 400 Eye M.D.s had scheduled CAD visits to members of Congress in conjunction with the Academy’s 2014 Mid-Year Forum in Washington, D.C. The AAPOS and AAP-Section on Ophthalmology remain crucial partners with the Academy in its ongoing federal and state advocacy initiatives.

State Eye PAC

We all must also support our respective State Eye PACs, because state ophthalmology societies cannot count on the Academy’s SSF alone. The presence of a strong State Eye PAC providing financial support for campaign contributions and legislative education to elect ophthalmology-friendly candidates to the state legislature is also critical. The Secretariat for State Affairs strategizes with state ophthalmology societies on target goals for state eye PAC levels.

ACTION REQUESTED: Advocate for your patients!!

Academy Surgical Scope Fund contributions are used to support the infrastructure necessary in state legislative/regulatory battles and for public education. PAC contributions are necessary at the state and federal level to help elect officials who will support the interests of our patients. Contributions to each of these three funds are necessary and should be considered the costs of doing business. Surgical Scope Fund contributions are completely confidential and may be made with corporate checks or credit cards, unlike PAC contributions, which must be made by individuals and are subject to reporting requirements.

Please respond to your Academy colleagues who are volunteering their time on your behalf to serve on the OPHTHPAC® and Surgical Scope Fund Committees, as well as your state ophthalmology society leaders, when they call on you and your subspecialty society to contribute. Advocate for your patients now!

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Michael X Repka MD (MD)
Gregory L Skuta MD (OK)
George A Williams MD (MI)

**State EyePAC**

To derail optometric surgical scope-of-practice initiatives that threaten patient eye safety and quality of surgical care

Political grassroots activities, lobbyists and media; no funds may be used for candidates or PACs

Contributions: Unlimited.

Individual, practice and organization.

Contributions are 100% confidential.

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<td>Individual, practice and organization.</td>
<td>Contributions above $200 are on the public record.</td>
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Persistent Fetal Vascular Syndrome: Divide and Conquer

Elias I Traboulsi MD

The surgical management of pediatric cataracts associated with persistent hyperplastic primary vitreous (or persistent fetal vascular syndrome) can be challenging. In addition to the microphthalmia in some cases, the thick posterior capsular complex can be quite difficult to remove. This complex is formed from a combination of capsule and fibrovascular tissue that remains after partial regression of the secondary vitreal structures. As the fetal eye is developing, this abnormal fibrovascular tissue and the incorporated posterior capsule are presumed to contract, shrinking and pulling the zonule and ciliary processes toward it centripetally. Because of its thickness and leathery consistency, this complex is difficult to remove in order to clear the visual axis.

This video presentation illustrates the final stages of the surgical intervention. After the anterior capsule is removed with the vitrector through a clear corneal incision and using a separate infusion of saline from a blunt handheld cannula introduced through another corneal incision, the posterior capsular complex is incised using an MVR blade. The complex is then divided into slivers using curved intraocular scissors. Each sliver is cut and aspirated using the vitrector. Additional radial cuts are then performed using the scissors all the way to the edge of the capsule and in between the ciliary processes in multiple locations, dividing the capsular complex ring and allowing the ciliary processes to relax close to their normal anatomic location. If patent hyaloid blood vessels are encountered in the process of surgery, they are cauterized. A limited anterior vitrectomy is then performed. The 2 corneal incisions are closed using 10-0 polyglactin sutures.
Persistent Fetal Vasculature Syndrome:
A Posterior Approach
The Case of Progressive Lens Changes in PFV and an Illustration of What to Do and What Not to Do at Surgery

M Edward Wilson MD

The surgical management of cataracts and other lens disorders in children presents challenges at every turn. The surgeon must develop a set of principles and preferred techniques but must also be willing and able to customize or individualize the approach to each case as it evolves.

Persistent fetal vasculature (PFV) encompasses a very wide spectrum of findings, each of which may vary in their severity. One-size-fits-all approaches to these findings are not optimal. In general, I am cautious about approaching a severe PFV from a pars plana or pars plicata incision. This is true especially when ciliary processes are displaced toward the center of the pupil. Ciliary process displacement can be a marker for an abnormally anterior retinal insertion into the abnormal PFV material. For those cases, and in all cases where an IOL is not being inserted, I approach the lens, posterior capsule, and vitreous with an anterior bimanual approach. For these cases, I follow the same principles just illustrated by Dr. Traboulsi in the preceding video bonanza presentation.

In the absence of severe PFV, my preferred technique is to place an IOL within the capsular bag prior to opening the posterior capsule. All of the ophthalmic viscosurgical device is removed, and then the posterior capsulectomy and vitrectomy are performed via a pars plana incision, with the irrigation port remaining in the anterior chamber. This has worked well for me even when a stalk or mild to moderate posterior capsule plaques are found.

The video illustrated in this presentation is a unique case where a posterior approach was used for a portion of the surgery in spite of the presence of PFV. In this 3-year-old child, progressive lens changes (weakening and bulging of the posterior capsule) were documented in association with a large PFV stalk. In infancy, the refraction was +1.00 and the lens was clear other than the stalk. No surgery was recommended initially. By age 3 years, the lens was bulging posterior and the refraction was -10.00+6.00X125. During surgery, the posterior lens capsule ruptured prematurely when the irrigation cannula entered the eye. As in lentiglobus, a low-flow surgery would have been safer in this case and might have prevented an uncontrolled posterior rupture.
A 2-year-old Hispanic male was referred to the Pediatric Ophthalmology Department at the University of Illinois at Chicago for evaluation of esotropia. His parents denied other ocular or systemic issues. The patient was able to fixate and follow bilaterally with a 40 PD esotropia and full extraocular movements. IOPs were elevated by globe palpation. Anterior segment examination revealed peripheral band keratopathy bilaterally. Pupils were 2 mm, minimally reactive, and failed to dilate despite multiple rounds of mydriatics.

An examination under general anesthesia (EUA) was performed. Elevated early IOPs of 40 and 31 mmHg were found in the right and left eye, respectively. Axial length was approximately 26 mm O.U. B-scan ultrasound did not reveal vitritis or intraocular mass. Chronic anterior uveitis with secondary glaucoma was suspected; systemic and rheumatologic workup were within normal limits.

An ultrasound biomicroscopy (UBM) with a 50-mHz probe (Sonomed; Lake Success, NY) was performed. Very shallow anterior chambers in both eyes with closed angles, and multiple iridocorneal adhesions were visualized. The crystalline lenses were small and spherical in shape and appeared to be pushing the irises forward.

**Diagnosis: Microspherophakia**

The diagnosis of microspherophakia was suspected on UBM and confirmed during surgical exploration as shown in the video. Lysis of synechiae, lensectomy, and anterior vitrectomy were performed in each eye. Aphakic spectacle correction was given. At 6-month follow-up, the patient’s vision was central, steady, and maintained in both eyes. The IOP was controlled with a combination of timolol and dorzolamide drop in his right eye only. Visualization of the posterior pole revealed moderate cupping in both eyes.
Superior Oblique Exaggerated Traction Test

Jonathan M Holmes MD

I. Index Case
Referral of an unexpected hypotropia following “planned complete tenotomy of both superior oblique (SO) tendons” elsewhere
A. Suspected “incomplete SO tenotomy” on one side
B. Confirmed at the time of reoperation
C. How could this be avoided?

II. Judging the Height of the Speed Bump
A. Normal SO exaggerated traction test – normal speed bump
B. Abnormal SO exaggerated traction test – Brown syndrome – “the wall”
C. Retesting SO exaggerated traction test after “planned SO disinsertion” (or planned complete tenotomy): Is the speed bump still there?
D. If so, explore for residual fibers.
E. Repeat SO exaggerated traction test; need to feel “the flat line” to confirm there are no residual SO fibers

III. Conclusion
Get proficient at assessing the height of the SO “speed bump” to avoid nasty surprises following SO surgery.

Selected Reading
Although advanced or anterior cataracts may be visible to a patient in a mirror, that is typically not the case for focal and posterior lens opacities. This video will present a simple examination technique allowing in vivo patient visualization (entoptic perception) of anterior segment opacities.

The examination technique is simple to accomplish for cooperative patients of all ages. With the patient positioned in a dark room, a light source is pointed at the patient’s face from a distance. A high plus lens is introduced approximately 1 inch in front of the tested eye while the fellow eye is covered. The patient is asked to describe what they perceive.

It is my experience that patients with focal cataracts and good overall acuity greatly appreciate visualizing their optical problem. The experience and related discussion bring rewards to the patient-physician relationship.
Sinskey Procedure for Nystagmus
Early Observations on the Sinskey Subtotal Exirpation Procedure for Nystagmus

Robert W Lingua MD

In 2002, Drs. Sinskey and Eshete1 published on 4 cases of adult nystagmus managed with anterior subtotal extirpation of the 4 horizontal recti. The anterior 20-22 mm of extraocular muscle is removed with the aide of an enucleation snare, where the muscle exits posterior Tenon fascia. Cotton tip compression is applied for 5 minutes.

It was reported that all patients experienced a reduction in normal and abnormal eye movements, and all experienced subjective improvement in vision and appearance. In this series, 2 of the 4 experienced exotropia, reported as the most common complication. Despite the removal of a large portion of the muscle, lateroverision is reduced, but intact, as evidenced in the videos presented. The muscles are still able to execute voluntary pursuit and saccade. This procedure does not result in paralysis.

In 2012, I performed 2 traditional procedures on a 17-year-old patient with infantile nystagmus syndrome. A tenotomy and reattachment of the 4 horizontal recti initially2 was followed by a 4 muscle large (> 10 mm) recession of the horizontal recti.3,4 Both procedures failed to control the nystagmus and improve acuity. We performed the anterior extirpation as previously described in this patient, documented nystagmographic evidence for quieting the nystagmus, and recorded visual acuity improvement from 20/50 to 20/25, binocular best corrected (BBCVA), distance and near. Videos will be presented demonstrating preservation of lateroverision and quieting the nystagmus.

Infrared videonystagmography depicting spontaneous nystagmus, gaze-associated nystagmus, pursuit and saccade was performed pre- and postoperatively with a commercially available nystagmograph from Interacoustics. All numeric data derive from the proprietary software in the unit.

Figure 1. Nystagmograph of preoperative fixation in primary position, at 70 cms.

Figure 2. Nystagmograph of 8-month postoperative fixation in primary position, at 70 cms.

Figure 3. Nystagmograph of preoperative smooth pursuit-horizontal pendular target, 70 cms.
We report on a subsequent pediatric case treated primarily with the Sinskey procedure, experiencing the reported large angle exotropia, and their response to secondary augmentation. In 1 patient, the BBCVA improved from 20/150 to 20/60 distance and from 20/150 to 20/30 at near, and the eyes are aligned with intact stereopsis (400 seconds of arc). In a second case treated with the Sinskey alone, a large exotropia resulted that has not responded to secondary augmentation procedures.

We now augment the Sinskey procedure primarily, to prevent the risk of a large, difficult, postoperative strabismus. Augmentations have included either nasal transposition of the nasal \( \frac{3}{2} \) superior and inferior rectus to the superior or inferior border of the original medial rectus insertion site or anteriorization of the accessory abductors, the superior and inferior obliques. The inferior oblique is anteriorly transposed to the temporal border of the inferior rectus insertion and the superior oblique to the nasal border of the superior rectus. Both augmentations are effective to avert a clinically significant strabismus. However, the patients undergoing the oblique transposition retained better lateral gaze, while those undergoing the nasal transposition were more tethered from abduction.

As of this writing we have performed this procedure on 12 patients with a follow-up of 3 to 18 months. A more comprehensive report, when all are at least 6 months postop, is in preparation.\(^1,^2\)

Clinical and nystagmographic data will be presented to evidence the effectiveness of this procedure in controlling nystagmus and improving acuity, in our initial cases.

References


5. Lingua RW, Sinskey RM, Gerling A. Clinical and nystagmographic data of 12 patients treated with the augmented Sinskey anterior extirpation for nystagmus. In prep.

A 20-month-old female with a history of right eyelid swelling for 1 month presented to the pediatric ophthalmology clinic for evaluation. She had previously been treated with topical antibiotic drops and topical antihistamine drops as well as oral antibiotics without resolution of the swelling. On exam, she was found to have a whitish-gray lesion of the palpebral conjunctiva of the right upper eyelid on eyelid eversion. In clinic, some of the lesion was able to be removed with a forceps and a portable slitlamp. The material appeared to be calcific in nature and was thought to be a conjunctival foreign body. The corneal had a small area of epithelial defect superiorly, but no infiltrate. Given the potential for deprivational amblyopia and the difficulty of removing the lesion in the clinic, the decision was made to proceed to the operating room the next day for removal.

The edited video of her surgery is presented today. The lesion was viewed with the surgical microscope, and areas of the lesion appeared calcific. Forceps were used to remove the lesion in multiple pieces, as it was brittle and not cohesive. At one point, thick white material was noted to exude from the area of the lesion. When the lesion had been completely excised, the remaining tarsal defect measured 4mm x 2.5mm. The pieces of the lesion were sent to pathology, and the final pathology report indicated a pilomatrixoma. After the procedure, the patient’s eyelid healed well and the eyelid swelling resolved.

A pilomatrixoma (also called a benign calcifying epithelioma of Malherbe) is a tumor of the hair cortical cells. It presents most often in children and young adults, with a female predominance. A pilomatrixoma can appear almost anywhere on the body, but the most common sites of involvement are the head and upper extremities. It is often misdiagnosed as an epidermoid or dermoid cyst when it presents on the eyelids. Histology can show shadow cells (anucleated), basophilic cells, inflammation, and calcification.
The Induced Tropia Test

Stephen P Christiansen MD

Fixation testing to assess visual acuity in infants, preverbal children, children with developmental delay, or those with limited attention can be challenging. Monocular fixation testing is usually documented by noting whether the child’s fixation is central, steady, and maintained in each eye, but such testing is easily confounded or misinterpreted. The induced tropia test refines fixation testing and can better identify children with amblyopia or organic visual deficit. In performing the test, a 16 PD base-down prism is presented before one eye, creating a vertical retinal disparity that allows simultaneous observation of the quality and maintenance of fixation in both eyes. In this video presentation, the correct use of the induced tropia test is demonstrated. Interpretation and limitations of the test will be discussed with the panel.
Twenty-Five-Year Review of Closed Case Claims in Pediatric Ophthalmology and Strabismus
The OMIC Experience

Robert Wiggins MD MHA, Robert Gold MD, Anne M Menke RN PhD

I. Introduction

A. Although reports have drawn attention to medical liability and ROP, there are no comprehensive claims studies in the field of pediatric ophthalmology and strabismus.

B. The Ophthalmic Mutual Insurance Company (OMIC) was founded in 1987 and provides professional liability insurance for the majority of ophthalmologists in private practice. This database was used to analyze the claims in the field of pediatric ophthalmology and strabismus between 1988 and 2013.

C. Those clinical conditions and risk management issues that pose the highest risk for all ophthalmologists who treat this patient group will be discussed. Ophthalmologists can use these findings to improve the quality of care they provide their patients and reduce the personal and societal costs associated with professional liability litigation.

II. Methods

A. Retrospective analysis of 140 claims: 119 individual claims, 21 entity claims

B. Data collected
   1. Age, gender, professional liability coverage level, specialty, and locale of the insured ophthalmologist
   2. Age of the plaintiff
   3. Allegation and clinical category of the claim
   4. Location of the incident
   5. Date of the incident and when the claim was opened and closed
   6. Expert determination of whether or not the standard of care was breached
   7. Cause of the claim
   8. Outcome of the case (dismissal, settlement, mediation, or trial)
   9. Claim expense and indemnity payments
   10. Lessons learned from each case

III. Results

A. Cases/claims: 117 cases (unique plaintiffs) resulted in 140 claims.

B. Insured ophthalmologist characteristics:
   1. 100 male, 19 female ophthalmologists
   2. Mean age at time of incident = 45 years

C. Plaintiff characteristic: Most commonly < 1 year of age

D. Location of incident: Most commonly occurred in the office, followed by a surgical setting

E. Allegations: Treatment related most common

F. Standard of care: In 62 of 119 individual claims, expert review determined standard of care was met.

G. Claims course
   1. Time
      a. Incident to opening of claim: 2.24 years (mean)
      b. Opening to closing of claim: 2.58 years (mean)
   2. Resolution
      a. 44/119 (37%) individual claims had indemnity payments
      b. 3/21 (14%) entity claims had indemnity payments
   3. Indemnity payments and expenses
      a. Mean indemnity payment (individuals): $444,559
      b. Median indemnity payment (individuals): $238,722
      c. 105/119 (88%) of individual claims had associated expenses (mean: $60,933)

H. Claims characteristics by clinical condition
   1. Frequency (number of cases): Most common category was strabismus.
   2. Severity (percentage of claims closed with an indemnity payment): Greatest severity associated with ROP and highest category, mean indemnity payment ($932,928)

I. Risk management issues
   1. The most common risk management issues among individual claims included the categories of informed consent, follow-up, and the litigation process. The highest total indemnity payments were made in the category of “follow-up.”
2. Among entity claims the litigation process was most common.

IV. Conclusions

A. Claims in the field of pediatric ophthalmology and strabismus are low in frequency but high in severity among all ophthalmology claims at OMIC.

B. Claims were most common among subspecialists in the field of pediatric ophthalmology and strabismus, followed by comprehensive ophthalmologists, and occurred among a broad range of other subspecialists.

C. The most common claims were in the area of strabismus. Claims related to ROP were those associated with the highest severity and highest average indemnity payments.

D. A number of risk management factors were identified. Those related to follow-up accounted for the highest total indemnity payments paid by OMIC over the course of the study. These factors and risk management recommendations will be further explored in the following case studies.

V. Case Studies

A. Case 1: Failure to Diagnose Orbital Floor Fracture: Diagnostic Process

A 5-year-old male was fighting with his brother and supposedly was pushed to the floor and his face was rubbed against the carpet. The trauma was not witnessed by a parent. The patient was seen by the insured on Day 2 and a corneal abrasion was diagnosed. On Day 3 the patient had increased pain and the insured performed an EUA to rule out a foreign body. Four months later the pediatrician noted that the eye did not move properly. Another ophthalmologist was consulted and a CT revealed an orbit fracture requiring surgical intervention.

B. Case 2: The Unhappy Patient after Strabismus Surgery—Informed Consent

A 51-year-old female underwent strabismus surgery by the insured ophthalmologist because she did not like the appearance of the misaligned eye. She remained unhappy with the alignment, as well as the appearance of scar tissue after the first surgery. One year later the insured performed further surgery. After the second surgery, patient felt she was even worse off and scheduled additional strabismus surgery with another ophthalmologist. Plaintiff claimed no informed consent (IC) had been given. The insured ophthalmologist did not use a procedure-specific consent form but does mention that IC was obtained and documented the discussion was performed in the chart.

C. Case 3: Failure to Treat Corneal Ulcer—Patient Hand-Off

A 2-year-old reportedly fell into “oily dry matter” in her driveway. An ER doctor called the insured on a Sunday to report a diagnosis of a “corneal abrasion with acute inflammatory response.” The insured was leaving for vacation and did not follow his usual practice of alerting his office about an ER consult, so they did not know the mother had been instructed to come to the office. The patient/family showed up at the insured ophthalmologist’s office the next day, did not identify themselves as a referral from the ER, and mistakenly reported the condition as conjunctivitis; they were not seen, as the office had a policy of not seeing “public aid” patients. The patient was ultimately seen at the local Children’s Hospital 2 days later and examined by the pediatric ophthalmologist and cornea specialist, who diagnosed a significant corneal ulcer that led to a permanent decrease in vision.

D. Case 4: Glaucoma and Poor Vision After Infantile Cataract Surgery—Noncompliance / Follow-up

A 4-month-old male underwent monocular cataract surgery by the insured ophthalmologist. The surgery and postoperative course were uneventful. Nine months later the child was being followed by a comprehensive ophthalmologist from out of town who noted the eye was “rock hard.” The insured ophthalmologist made medication recommendations and on follow-up the IOP was 26 mmHg. Arrangements were made for follow-up with the comprehensive ophthalmologist and with the insured ophthalmologist. The patient did not return for further follow-up. Several phone calls were made, and a letter was sent to the family. A claim was later filed alleging poor management of the child’s case resulting in poor vision in the left eye.

E. Case 5: Failure to Diagnose Intraocular Foreign Body—Telephone Care

A 10-year-old boy’s parents reported that he was hammering a penny in his garage and a fragment flew up and hit him in the eye. The parents flushed the eye. The next day the child was seen by his pediatrician, who noted decreased vision (which was difficult to assess) and pain. The pediatrician found no evidence of a penetrating injury. The pediatrician telephoned the insured, who was on call for the local children’s hospital ER. The insured advised the pediatrician to refer the child to his office or the ER if the condition worsened. The pediatrician spoke to the insured several days later to inform him that the patient did not improve and did not go to an ER, but was seen by another ophthalmologist who diagnosed an intraocular foreign body requiring surgical intervention.

F. Case 6: ROP and Bilateral Retinal Detachments (Systems Issues / Follow-up)

A premature infant born at 28 weeks gestation with a birth weight of 949 grams was followed by the insured ophthalmologist for ROP. The insured ophthalmologist evaluated the child on one occasion 7 weeks after birth and diagnosed stage 1, zone 3 ROP and recommended a follow-up exam 1 week later. The NICU did not contact the ophthalmologist for a 1-week visit as was customary. The insured ophthalmologist sent the family a letter after...
the missed appointment but is not sure why his staff did not contact the hospital, as the patient was still in the hospital at the time. The infant was transferred to another hospital 2.5 weeks after the first eye exam, but the neonatologist did not mention ROP in the discharge summary. The infant was later diagnosed with bilateral retinal detachments.

Selected Readings

Lessons to Learn: Pearls from Individual Cases

Robert E Wiggins MD MHA, Robert S Gold MD, Anne M Menke RN PhD
Section VII: Perplexing Problems in Pediatric Neuro-Ophthalmology—Case Presentations

Case 1: A Star Is Born—Acute Visual Loss
Paul H Phillips MD

CASE

A 16-year-old girl was in her usual state of good health until 3 weeks prior to presentation, when she had a flu-like illness with fever and myalgia. These symptoms resolved during the following 2 weeks. However, 1 week prior to presentation she had floaters and decreased vision O.D. She denied eye pain. Past medical history was unremarkable and she was on no medications.

On the day of presentation she had a visual acuity of 20/200 O.D; 20/20 O.S. External, slitlamp, and ocular motility examination were normal. Pupils were equal and reactive with a relative afferent pupillary defect O.D. Dilated fundus examination revealed vitreous cells, disc edema, and a macular exudate in a star formation O.D.

Case 2: An Eyelid Has Fallen and Can’t Get Up—Acute Ptosis
Stacey L Pineles MD

CASE

History and Exam

A 3-year-old child presents with ptosis of the left eye for as long as her parents can remember. At 9 months of age, she presented to an ophthalmologist for ptosis and intermittent drifting out of either eye. Her left eye drifted more than the right eye, but the right eye also occasionally drifted out. She underwent treatment with patching and glasses, but the eyes continued to drift out so she underwent strabismus surgery at 2 years of age. Her ptosis never really improved.

On examination, her visual acuity was 20/130 in each eye by preferential looking testing (PLT). Cycloplegic refraction was +2.50 sphere in each eye. Versions were full with no abduction limitation. No nystagmus was noted. She had a comitant esotropia measuring 30 PD at distance, at near, and in right and left gaze. External examination was normal without evidence of ptosis. Anterior segment examination was normal. Dilated fundus examination was normal, with no evidence of optic nerve edema.

Case 3: Seeing Two Mommies—Acute Onset Esotropia
Acute Acquired Comitant Esotropia
Gena Heidary MD

CASE

History and Exam

An otherwise healthy 2-year-old girl presented to the pediatric ophthalmology clinic with a sudden onset esotropia of her right eye. Her parents had noted an intermittent right esodeviation for 1 week that then became constant. In that setting, the child was noted to rub and cover her right eye. There was no antecedent viral illness, trauma, recent immunization, or family history of strabismus. Her review of systems was negative for headache, vomiting, ataxia, or prior episode of strabismus or ptosis.

On examination, visual acuity was 20/130 in each eye by preferential looking testing (PLT). Cycloplegic refraction was +2.50 sphere in each eye. Versions were full with no abduction limitation. No nystagmus was noted. She had a comitant esotropia measuring 30 PD at distance, at near, and in right and left gaze. External examination was normal without evidence of ptosis. Anterior segment examination was normal. Dilated fundus examination was normal, with no evidence of optic nerve edema.

Case 4: Not Seeing Mommy—The Infant With Poor Vision
Mark S Borchert MD

CASE

History and Exam

A 2-year-old child presents with poor vision. At 9 months of age, she presented to an ophthalmologist for poor vision and intermittent drifting out of either eye. Her left eye drifted more than the right eye, but the right eye also occasionally drifted out. She underwent treatment with patching and glasses, but the eyes continued to drift out so she underwent strabismus surgery at 2 years of age. Her ptosis never really improved.

On examination, her visual acuity was 20/20 O.D. and 20/40 O.S. She had no measurable stereoaucuity. Pupils were round and symmetric and reacted normally to light and near. External examination revealed ptosis in the left eye. Her palpebral fissures measured 12 mm O.D. and 6 mm O.S. She had a large exotropia and hypotropia in primary position. She had severe limitation to elevation, adduction, and infraduction in the left eye. The remainder of her extracocular movements were normal. Slitlamp and fundus examinations were normal.
Case 1: A Star is Born—
Acute Visual Loss

Paul H Phillips MD

I. Definition
   A. Syndrome-disc edema and retinal star exudate
   B. Leber (1916)¹
      1. Focused on retinal star exudate
      2. Leber stellate maculopathy
   C. Gass (1977)² — disc edema before / concurrent with retinal star
      1. Disc vessel leakage source of retinal fluid
      2. Subsequent fluid resorption results in star

II. Demographics
   A. All ages
   B. No gender predominance
   C. Unilateral or bilateral

III. Clinical Manifestations – Systemic Prodrome
   A. Malaise
   B. Headache
   C. Nausea
   D. Vomiting
   E. Upper respiratory symptoms
   F. Arthralgias
   G. Diarrhea
   H. Fever
      I. Lymphadenopathy
      J. Some asymptomatic³

IV. Clinical Symptoms, Ocular
   Occur days to weeks after systemic symptoms
   A. Decreased vision
   B. Painless or dull periocular pain
   C. Floaters
   D. Flashes³

V. Clinical Signs, Ocular
   A. Disc edema
   B. Retinal fluid
   C. Retinal exudate in star formation with resolution
   D. Hemorrhage
   E. Cotton wool spots
   F. Anterior uveitis
   G. Vitritis

VI. Etiology
   A. Bartonella henselae (catscratch disease)
   B. Idiopathic
   C. Viral
   D. Syphilis
   E. Lyme disease
   F. Leptospirosis
   G. Toxoplasmosis
   H. Toxocariasis
      I. Tuberculosis
      J. Sarcoid
   K. Chronic/autoimmune
   L. Acute retinal necrosis
   M. ?? Progressive outer retinal necrosis
   N. Inflammatory etiology implied
   O. Retinal star may occur from noninflammatory disc edema.
      1. Papilledema
      2. Hypertensive retinopathy
      3. Anterior ischemic optic neuropathy

VII. Neuroretinitis and Multiple Sclerosis
   Parmley et al⁴: No multiple sclerosis among 50 patients with neuroretinitis
   A. Neuroretinitis: Inflammation involves prelaminar disc vessels, resulting in retinal edema.
   B. Multiple sclerosis: Inflammation involves retrolaminar myelin with less retinal edema.

VIII. Catscratch Disease
   A. Bartonella henselae: Gram-negative bacillus, responsible organism
   B. Diagnosed by lymph node biopsy, culture, or skin test prior to 1993
   C. Indirect fluorescent-antibody testing available in 1993; convenient and sensitive
D. Most common etiology of neuroretinitis
   1. Suhler et al\(^6\): 64% of 14 cases
   2. Accounts for idiopathic cases
E. Transmitted by cat scratch/bites or fleas
F. Flu-like symptoms
G. Adenopathy
H. Parinaud ocular glandular conjunctivitis
I. Neuroretinitis\(^5\)

IX. Catscratch Disease – Neuroretinitis
   A. Disc edema and retinal scar
   B. Discrete white retinal/choroidal lesions
   C. Branch retinal artery/vein occlusion / central retinal artery/vein occlusion – often at location of foci
D. Uveitis
E. Vitreous cysts
F. Macular hole
G. Peripapillary angiomatosis\(^3,7\)

X. Catscratch Disease: Systemic Complications
   A. Encephalitis
   B. Cranial nerve palsy
   C. Hepatomegaly/splenomegaly\(^8,9\)

XI. Neuroretinitis Workup
   A. History/physical examination
   B. Bartonella serology: Repeat if initially negative
   C. Consider FTABS/PPD/CXR – other serology
   D. Neuroimaging not necessary in typical case.
   E. Presume and treat for catscratch if findings are typical\(^3,9\)

XII. Neuroretinitis Natural History
   A. Natural history: resolution with good vision in most cases
   B. Recurrence is rare.

XIII. Neuroretinitis Treatment
   Antibiotics: azithromycin, doxycycline, rifampin, doxycycline, Bactrim, ciprofloxacin
   A. Reed\(^10\): Doxy/rifampin hastened recovery in 7 patients.
   B. Solley\(^3\): Treated patients (11) no different than untreated patients (13).

XIV. Neuroretinitis: What about the cat?
   A. Catscratch disease in familial members demonstrated
   B. Cat forms immunity by the time disease is detected in humans.

References

Case 2: An Eyelid Has Fallen and Can’t Get Up—Acute Ptosis

*Stacey L Pineles MD*

**CASE**

Clinical Course and Outcome
On further inspection, it was determined that the ptosis in her left eye was associated with an eyelid twitch on attempted upgaze. She had curtaining of the right eyelid when her left eyelid was elevated by the examiner. She also had improvement of her ptosis with 10 minutes of rest in a dark room. Acetylcholine receptor antibodies were sent and were elevated. The patient was started on Mestinon and there was mild improvement in her ptosis, but she continued to have a large exotropia and residual ptosis, so oral corticosteroids were started and systemic immunosuppression with azathioprine was eventually initiated. An MRI of the chest did not reveal any thymus abnormalities. Her amblyopia was adequately treated with the addition of patching treatment.

**Final Diagnosis**
Myasthenia gravis
**Teaching Points**

1. The underlying etiology of ptosis in a child can be determined by evaluating associated history and examination findings.
2. Isolated congenital ptosis can usually be diagnosed based on historical features and old photographs. Congenital ptosis can also be seen with congenital oculomotor nerve palsy, congenital Horner syndrome, and congenital / neonatal myasthenia gravis.
3. Acquired myogenic causes of ptosis such as chronic progressive external ophthalmoplegia and myotonic dystrophy are typically seen in association with diminished ocular ductions in all directions and typically cause symmetric bilateral ptosis.
4. Acquired ptosis due to neuromuscular transmission defects are almost always attributable to myasthenia gravis in children. Ptosis from myasthenia gravis is usually bilateral but asymmetric and may be associated with motility defects. Variability, fatigability, and eyelid twitch are all characteristically seen. Diagnosis can be made with acetylcholine receptor antibody testing, single fiber electromyogram (although this is difficult in young children, especially those under the age of 8 years), Tensilon testing, or rest/ice test.
5. Neurologic causes of ptosis in children are typically associated with pupillary abnormalities and are most often caused by third nerve palsy or Horner syndrome. If the pupil on the ptotic side is larger, then suspect third nerve palsy. If the pupil on the ptotic side is smaller, then consider Horner syndrome.

**Selected Readings**


**Case 3: Seeing Two Mommies—Acute Onset Esotropia**

**Acute Acquired Comitant Esotropia**

**Gena Heidary MD**

**CASE**

**Clinical Course and Outcome**

Patient was prescribed her full cycloplegic refraction for a presumed diagnosis of accommodative esotropia with acute onset. Follow-up examination 8 weeks later revealed incomplete resolution of the esodeviation to an intermittent esotropia of 14 PD at distance, near, right and left gaze. Visual acuity in the right eye showed a slight decrement compared with the left eye. The remainder of the examination was unchanged. Patching of the left eye was initiated.

One month later, the parents called describing an unusual gait with an inwardly turned left foot, new imbalance, and difficulty with walking. Neurologic examination was notable for ataxia but was otherwise nonfocal. Urgent neuroimaging was performed. MRI brain with and without contrast was normal. There was improvement in the gait without intervention.

Follow-up eye examinations revealed worsening control of the esodeviation, and the patient underwent bilateral medial rectus recession with good restoration of ocular alignment.

**Final Diagnosis and Teaching Points**

**Final Diagnosis**

Acute acquired comitant esotropia without evidence of an intracranial process

**Teaching Points**

1. Acute acquired comitant esotropia is generally considered to be a benign condition, commonly described in the context of an interruption of oculomotor fusion through occlusion, in association with mild/moderate myopia, or as an idiopathic entity.
2. Acute acquired comitant esotropia may be the presenting sign in patients who harbor neurologic disease, including tumors of the corpus callosum, thalamus, brainstem, and cerebellum or in association with a Chiari I malformation or hydrocephalus.
3. A detailed ophthalmological evaluation with a focus on signs of a neurologic process, including the presence of nystagmus or optic nerve edema, may guide the determination of when to obtain neurologic evaluation and neuroimaging. Failure to re-establish oculomotor fusion following surgical intervention may also reflect underlying neurologic disease and should warrant further evaluation.

**Selected Readings**

Case 4: Not Seeing Mommy—
The Infant With Poor Vision
Mark S Borchert MD

Answer and Teaching Points

Ophthalmoplegia, gaze palsies (eg, Moebius syndrome), and ocular motor apraxia may present as poor visual behavior before the child develops good head and neck control. If eye movements are normal with vestibular stimulation, the critical diagnostic point is in distinguishing cortical vision impairment from ocular disease. Congenital optic nerve or retina diseases generally have diminished pupillary responses and nystagmus by age 3-4 months. Subsequent workup then depends on the fundus findings. If fundus exam is normal, an ERG should be performed at 6 months of age.

On the other hand, a child with poor vision, normal pupillary responses, and no nystagmus must be considered to have cortical vision impairment (CVI), regardless of ophthalmoscopic findings.

At this point, our patient has brisk pupillary response to light and no nystagmus.

Children with cortical vision impairment frequently have superimposed optic atrophy, the presence of which helps direct further workup. If there is no optic atrophy and the child is otherwise developing normally, she can be presumed to have visual maturational delay, and further evaluation can be delayed until age 6 months if vision does not spontaneously improve.

In our child, moderate temporal disc pallor is noted O.U.

Electrophysiology has no role in the diagnosis or management of cortical vision impairment. A flat visual evoked potential (VEP) in a child with absolutely no visual behavior is prognostic of poor visual outcome. Otherwise, the VEP has no prognostic value. In the face of optic atrophy, an MRI is necessary to rule out treatable causes of CVI, such as hydrocephalus, or to anticipate neurologic problems, such as seizures associated with gyral malformations.

MRI shows encephalomalacia with hydrocephalus ex vacuo.

Diagnosis

Cortical vision impairment and optic atrophy due to congenital ischemic encephalopathy
The Hardest Conversation: Functional Vision Loss and What to Say to the Parents and the Patient

R Michael Siatkowski MD

I. Classification of Functional Vision Loss (FVL)
   A. Conversion (primary gain) vs. malingering (secondary gain)
   B. Pure FVL vs. functional overlay: Relative contribution of organic vs. non-organic visual loss
   C. Adult vs. pediatric FVL
   D. Unequivocal vs. possible FVL
      1. Unequivocal: definitely 20/20 and full field O.U.
      2. Possible
         a. Known organic disease with FVL but uncertain of contribution of each
         b. Are you are missing true organic disease? (2%-3%) Stargardt, cone dystrophy, amblyopia, bilateral homonymous hemianopsia most common missed diagnosis

II. Demographics of FVL
   A. Adults
      1. Trigger factors
         a. Antecedent trauma/injury: 58%
         b. Recent surgery: 20%
         c. Fear of eye disease / blindness
      2. Psychiatric disease
         a. 30% with stress, anxiety, depression
         b. 15% on psych meds
      3. Associated conditions
         a. Female: 75%
         b. Migraine: 15%-20%
         c. Trigeminal neuralgia: 6%
         d. Involved in litigation: 10% overall, but 50% of those with prior trauma
      4. Outcomes: 45%-78% recovery
   B. Children
      1. Trigger factors
         a. Social (school, family relationships, need for attention): 45%
         b. Friends/peers with glasses, eye drops, etc.: 5%
         c. Family member with organic eye problem
         d. Sexual abuse: 2%
      2. Psychiatric disease: Prior diagnosis rare but often acknowledge stress factors
      3. Associated conditions
         a. Female: 67%
         b. Migraine: 15%-20%
         c. Trigeminal neuralgia: 0%
      4. Outcomes: 75%-95% recovery

III. New Research
   A. Data not consistent, often contradictory
   B. Conversion disorders associated with dysregulation of modulating pathways
   C. Limbic system, orbitofrontal cortex, anterior cingulate gyrus involved
   D. Functional MRI abnormal in some cases

IV. Management
   A. No comparative data exist.
   B. Historical methods
      1. IV amobarbital
      2. Sensory isolation tent with bilateral patching
      3. ECT
      4. Psychoanalysis
      5. Placebo drops or glasses
   C. Standard recommendations
      1. Stress good prognosis, encourage recovery, supportive manner.
      2. Engineer a “way out.”
      3. Do not employ confrontation.
      4. Placebo Rx only 50% successful
      5. Make a follow-up appointment unless sure 20/20 and a full-field O.U.
   D. Personal recommendations
      1. Explain eye exam normal, no reason to explain visual loss.
      2. Gentle confrontation may be helpful by explaining that inconsistencies in exam (stereo, fields, etc.) are mathematically impossible—if presentation is for secondary gain only, discussion may end here.
      3. Reassure that there is no evidence of brain tumor or blinding disease.
      4. Introduce concept of SADness (stress, anxiety, depression) and ask them to identify possible areas in their life.
5. Empathize with patient by explaining how you experience SADness.

6. Be aware of possibility of talking to patient or some family members separately if abuse is a possibility or confidentiality is an issue.

7. Have a technician or resident with you in the room for private conversations with patients or selected family members.

8. DMEI psych referral / no improvement rate 11% (cf up to 50% in literature).

References


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