Pediatric Ophthalmology 2017
Sightseeing in New Orleans—For the Pediatric Ophthalmologist

Program Directors
Yasmin Bradfield MD and Jonathan M Holmes MD

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

Ernest N Morial Convention Center
New Orleans, Louisiana
Saturday, Nov. 11, 2017

Presented by:
The American Academy of Ophthalmology

Pediatric Ophthalmology 2017
Planning Group
Jonathan M Holmes MD
Program Director
Yasmin Bradfield MD
Program Director
Erick D Bothun MD
Sean P Donahue MD PhD
Nils K Mungan MD
R Michael Siatkowski MD
Serena X Wang MD
Tammy L Yanovitch MD

Subspecialty Day Advisory Committee
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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 New Orleans and Pediatric Ophthalmology 2017: Sightseeing in New Orleans—For the Pediatric Ophthalmologist.
2017 Subspecialty Day
Advisory Committee

Daniel S Durrie MD, Chair
(Refractive Surgery)
Abbott Medical Optics: L,C
AcuFocus, Inc.: C,L,O
Alcon Laboratories, Inc.: C
Alphaeon: C,L,O | Avedro: L,O,C
Hoopes Durrie Rivera Research Center: C
Strathspey Crown LLC: C,L,O

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(Pediatric Ophthalmology)
National Eye Institute: S

Kuldev Singh MD
(Glaucoma)
Abbott Medical Optics Inc.: C
Aerie: C
Alcon Laboratories, Inc.: C
Allergan: C
Belkin Laser Ltd: C
Glaukos Corporation: C
InjectSense: C
Ivantis: C
Mynosys: C
National Eye Institute: S
Novartis Institute for Biomedical Research: C
Santen, Inc.: C
Shire: C
Thieme Medical Publishers: C
U.S. Food and Drug Administration: S,C

Nicholas J Volpe MD
(Neuro-Ophthalmology)
Ophthotech: C
Opticent Inc.: O

AAO Staff
Ann L'Estrange
None
Melanie Rafaty
None
Lisa Romero
None
Debra Rosencrance
None
Beth Wilson
None
Pediatric Ophthalmology 2017 Contents

Program Planning Group ii
CME vi
Faculty Listing viii
Mobile Meeting Guide xii
Program Schedule xiii

Section I: The Heart and Soul—Bourbon Street and Strabismus 1

Section II: Lagniappe—“Bonus Gifts” to Enhance Your Strabismus Surgery Outcomes 13
Advocating for Patients 19

Section III: Architectural Details—Corneal Structure and Disease 21

Section IV: Ghost Tours—Frightening Postoperative Surprises 28

Section V: Satchmo's Got Nothing on Me—“Inventive” Management of Intermittent Exotropia 35

Section VI: When the Complaints Go Marching In—Glasses Problems and How to Fix Them 42

Section VII: Jambalaya—A “Mishmash” of Extreme Benign Disease 49

Faculty Financial Disclosure 55
Presenter Index 58
CME Credit

The 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.

2017 Pediatric Ophthalmology Subspecialty Day Meeting Learning Objectives

This meeting will enable attendees to:

■ Improve their surgical management of complex secondary strabismus in adults
■ Expand their armamentarium to improve strabismus surgery outcomes
■ Evaluate new pediatric corneal disease surgery and devices that may change current practice
■ Understand how to prevent and manage unexpected postoperative strabismus outcomes
■ Incorporate new clinical trial data into their current management of intermittent exotropia
■ Recognize glasses prescribing and lens optical errors that can significantly impact patient satisfaction and vision quality
■ Improve their management of atypical presentations of common eye disease

2017 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.

2017 Pediatric Ophthalmology Subspecialty Day CME Credit

The American Academy of Ophthalmology is accredited by the Accreditation Council for Continuing Medical Education (ACCME) to provide continuing medical education 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.

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. To obtain an application form, please contact the AMA 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 relevant financial interest. The Academy has mechanisms in place to resolve all conflicts of interest prior to an educational activity being delivered to the learners.

The Academy requires all presenters to disclose on their first slide whether they have any financial interests from the past 12 months. Presenters are required to verbally disclose any financial interests that specifically pertain to their presentation.

Control of Content

The American Academy of Ophthalmology considers presenting authors, not coauthors, to be in control of the educational content. It is Academy policy and traditional scientific publishing and professional courtesy to acknowledge all people contributing to the research, regardless of CME control of the live presentation of that content. This acknowledgement is made in a similar way in other Academy CME activities. Though they are acknowledged, coauthors do not have control of the CME content and their disclosures are not published or resolved.

Attendance Verification for CME Reporting

Before processing your requests for CME credit, the Academy must verify your attendance at Subspecialty Day and/or AAO 2017. 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 Subspecialty Day Syllabi exchange voucher(s) onsite;
■ Register in advance and pick up your badge onsite if materials did not arrive before you traveled to the meeting;
■ Register onsite; or
■ Scan the barcode on your badge as you enter an AAO 2017 course or session room.
CME Credit Reporting

Lobby B, Lobby G, and Academy Resource Center, Hall G – Booth 3140

Attendees whose attendance has been verified (see above) at AAO 2017 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 2017 at the CME Credit Reporting booth.

Academy Members: The CME credit reporting receipt is not a CME transcript. CME transcripts that include AAO 2017 credits entered onsite will be available to Academy members on the Academy’s website beginning Dec. 7, 2017.

After AAO 2017, 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 2017.

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 2017 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 receipt
- Instruction course and session verification

Visit www.aao.org/cme for detailed CME reporting information.
Faculty

Gillian G W Adams MD
London, United Kingdom

Michelle T Cabrera MD
Seattle, WA

Laura B Enyedi MD
Cary, NC

Kyle A Arnoldi CO
Buffalo, NY

Kathryn A Colby MD PhD
Chicago, IL

K David Epley MD
Kirkland, WA

Erick D Bothun MD
Rochester, MN

Janine E Collinge MD
Oklahoma City, OK

Sergul A Erzurum MD
Poland, OH

Yasmin Bradfield MD
Madison, WI

Sean P Donahue MD PhD
Nashville, TN

Katherine J Fray BS CO
Little Rock, AR
Richard S Freeman MD
Minnetonka, MN

Kristin M Hammersmith MD
Philadelphia, PA

Malcolm R Ing MD
Honolulu, HI

Beatrice E Frueh MD
Bern, Switzerland

Edward J Holland MD
Union, KY

Burton J Kushner MD
Madison, WI

Nandini G Gandhi MD
Sacramento, CA

Jonathan M Holmes MD
Rochester, MN

Gregg T Lueder MD
Saint Louis, MO

David L Guyton MD
Baltimore, MD

David G Hunter MD PhD
Boston, MA

Sarah Mackinnon CO COMT
Boston, MA
Nils K Mungan MD
Ridgeland, MS

S Grace Prakalapakorn MD MPH
Durham, NC

Shira L Robbins MD
La Jolla, CA

Sarah M Nehls MD
Madison, WI

Cindy Pritchard BA COT
New Orleans, LA

Erin O Schotthoefer MD
Charlotte, NC

Faruk H Orge MD
Cleveland, OH

Radha Ram MD
Austin, TX

R Michael Siatkowski MD
Oklahoma City, OK

Stacy L Pineles MD
Los Angeles, CA

Michael X Repka MD MBA
Baltimore, MD

Yi Ning Strube MS MD FRCSC
Kingston, ON, Canada
Donny Won Suh MD
Omaha, NE

Serena X Wang MD
Dallas, TX

Pamela E Williams MD
Baton Rouge, LA

Lawrence Tychsen MD
St Louis, MO

Constance E West MD
Belmont, MA

Tammy L Yanovitch MD
Owasso, OK

Deborah K VanderVeen MD
Boston, MA
Ask a Question Live During the Meeting Using the Mobile Meeting Guide

To ask a question during the meeting, follow the directions below.

- Access at www.aao.org/mobile
- Select “Program Handouts & Evaluations”
- Filter by Meeting—Pediatric Ophthalmology Meeting
- Select Current Session
- Select “Ask the presenter a question (live)” Link
- Click Submit Question
Pediatric Ophthalmology 2017: Sightseeing in New Orleans—For the Pediatric Ophthalmologist

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

SATURDAY, NOV. 11

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<td>CONTINENTAL BREAKFAST</td>
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<td>8:00 AM</td>
<td>Welcome and Introductions</td>
<td>Yasmin Bradfield MD</td>
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<td>Jonathan M Holmes MD*</td>
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Section I: The Heart and Soul—Bourbon Street and Strabismus

Moderator: Jonathan M Holmes MD*

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<td>Introduction</td>
<td>Jonathan M Holmes MD*</td>
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<td>Case 1: Stuck Outside—Paralytic Exotropia following Sinus Surgery</td>
<td>Stacy L Pineles MD</td>
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<td>8:07 AM</td>
<td>Medial Rectus Retrieval</td>
<td>Gillian G W Adams MD</td>
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<td>8:12 AM</td>
<td>Periosteal Fixation</td>
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<td>8:17 AM</td>
<td>Panel Discussion of Alternatives</td>
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<td>8:22 AM</td>
<td>Case 2: Struck Inside—Esotropia with Conjunctival Scarring</td>
<td>Malcolm R Ing MD</td>
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<td>8:27 AM</td>
<td>Conjunctival Dissection and Graft</td>
<td>Yi Ning Strube MS MD</td>
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<td>FRCSC*</td>
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<td>Amniotic Membrane</td>
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<td>Panel Discussion of Alternatives</td>
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<td>Case 3: Down, Not Out—Hypotropia due to Brown Syndrome</td>
<td>Malcolm R Ing MD</td>
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<td>Superior Oblique Suture Spacer</td>
<td>Gillian G W Adams MD</td>
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<tr>
<td>8:52 AM</td>
<td>Superior Oblique Silicone Band Spacer</td>
<td>Yi Ning Strube MS MD</td>
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<td>FRCSC*</td>
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<td>Case 4: Hanging Out—Exotropia after Scleral Buckle</td>
<td>Jonathan M Holmes MD*</td>
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<tr>
<td>9:07 AM</td>
<td>Horizontal Rectus Muscle Surgery</td>
<td>Malcolm R Ing MD</td>
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<td>9:12 AM</td>
<td>OnabobotulinumtoxinA</td>
<td>Gillian G W Adams MD</td>
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<td>Panel Discussion of Alternatives</td>
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<tr>
<td>9:22 AM</td>
<td>Wrap-up</td>
<td>Jonathan M Holmes MD*</td>
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Section II: Lagniappe—“Bonus Gifts” to Enhance Your Strabismus Surgery Outcomes

Moderator: Sean P Donahue MD PhD*

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<td>Introduction</td>
<td>Yasmin Bradfield MD</td>
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<td>9:31 AM</td>
<td>Prism Adaptation</td>
<td>Kyle A Arnoldi CO</td>
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<tr>
<td>9:40 AM</td>
<td>Adhesion Barriers</td>
<td>Yasmin Bradfield MD</td>
</tr>
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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>
<td>9:49 AM</td>
<td>Pullover Traction Suture</td>
<td>Burton J Kushner MD</td>
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<td>9:58 AM</td>
<td>Delayed Adjustable Suture</td>
<td>Richard S Freeman MD</td>
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<td>10:07 AM</td>
<td>Phospholine Iodide for Residual Esotropia</td>
<td>Laura B Enyedi MD</td>
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<td>10:16 AM</td>
<td>Postoperative Eye Exercises</td>
<td>Cindy Pritchard CO COT</td>
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<td>10:25 AM</td>
<td>Wrap-up</td>
<td>Yasmin Bradfield MD</td>
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<td><strong>REFRESHMENT BREAK and AAO 2017 EXHIBITS</strong></td>
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**Section III: Architectural Details—Corneal Structure and Disease**

Moderator: Serena X Wang MD

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<td>Advocating for Patients</td>
<td>Pamela E Williams MD</td>
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<td>Pediatric Keratoprosthesis: Con</td>
<td>Kathryn A Colby MD PhD*</td>
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<td>11:16 AM</td>
<td>Pediatric Keratoprosthesis: Pro</td>
<td>Sarah M Nehls MD</td>
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<td>11:26 AM</td>
<td>Ocular Surface Stem Cell Transplantation for Aniridia</td>
<td>Edward J Holland MD*</td>
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<td>Pediatric Keratoconus: Crosslinking vs. Penetrating Keratoplasty vs.</td>
<td>Beatrice E Frueh MD*</td>
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<td>11:46 AM</td>
<td>Red Eye in Children: A Cornea Specialist’s View</td>
<td>Kristin M Hammersmith MD*</td>
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<td>11:56 AM</td>
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<td>Serena X Wang MD</td>
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**Section IV: Ghost Tours—Frightening Postoperative Surprises**

Moderator: Erick D Bothun MD

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<th>Presenter</th>
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<td>Introduction</td>
<td>Erick D Bothun MD</td>
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<tr>
<td>1:37 PM</td>
<td>Case 2: Discomfort and Dyscoria</td>
<td>Lawrence Tychsen MD</td>
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<td>1:47 PM</td>
<td>Case 3: When the Third Eye Is Not So Wise</td>
<td>Shira L Robbins MD*</td>
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<td>1:57 PM</td>
<td>Case 4: That Bump Shouldn’t Be There!</td>
<td>Gregg T Lueder MD</td>
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<td>2:07 PM</td>
<td>Case 5: “Weird Vision” after Strabismus Surgery</td>
<td>Deborah K VanderVeen MD*</td>
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<td>2:17 PM</td>
<td>Case 6: Forgiving SINS</td>
<td>Radha Ram MD</td>
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<td>2:27 PM</td>
<td>Wrap-up</td>
<td>Erick D Bothun MD</td>
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**Section V: Satchmo’s Got Nothing on Me—“Inventive” Management of Intermittent Exotropia**

Moderator: Nils K Mungan MD

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<td>Classification of Intermittent Exotropia</td>
<td>Katherine J Fray BS CO</td>
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<td>2:33 PM</td>
<td>The Natural History of Intermittent Exotropia</td>
<td>Donny Won Suh MD*</td>
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<td>2:40 PM</td>
<td>Occlusion for Intermittent Exotropia: IXT2 Results</td>
<td>Faruk H Orge MD</td>
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<td>Overminus Spectacles for Intermittent Exotropia: Initial Results from a Pilot Study</td>
<td>Sergul A Erzurum MD</td>
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<td>Surgery for Intermittent Exotropia: Initial Results from the IXT1 Trial</td>
<td>Sean P Donahue MD PhD*</td>
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<td>Wrap-up</td>
<td>Nils K Mungan MD</td>
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<td><strong>REFRESHMENT BREAK and AAO 2017 EXHIBITS</strong></td>
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* Indicates that the presenter has financial interest. No asterisk indicates that the presenter has no financial interest.
**Section VI:** When the Complaints Go Marching In—Glasses Problems and How to Fix Them  
**Moderator:** R Michael Siatkowski MD*  

<table>
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<td>Introduction</td>
<td>R Michael Siatkowski MD*</td>
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<tr>
<td>3:33 PM</td>
<td>Case 1 and Case 2: I Want My Money Back</td>
<td>David L Guyton MD*</td>
<td>42</td>
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<tr>
<td>3:44 PM</td>
<td>Case 3 and Case 4: These Glasses Make Me Sick</td>
<td>Michael X Repka MD MBA*</td>
<td>44</td>
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<td>3:55 PM</td>
<td>Case 5 and Case 6: My Child Will Not Wear His Glasses</td>
<td>Constance E West MD*</td>
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<tr>
<td>4:06 PM</td>
<td>Case 7 and Case 8: I Have Never Had a Pair of Glasses I Could See With</td>
<td>David G Hunter MD PhD*</td>
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<td>4:17 PM</td>
<td>Case 9 and Case 10: These Glasses Make Me See Double</td>
<td>Sarah E Mackinnon CO COMT*</td>
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<td>4:28 PM</td>
<td>Wrap-up</td>
<td>R Michael Siatkowski MD*</td>
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**Section VII:** Jambalaya—A “Mishmash” of Extreme Benign Disease  
**Moderator:** Tammy L Yanovitch MD

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<th>Event</th>
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<td>Introduction</td>
<td>Tammy L Yanovitch MD</td>
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<td>Case 1: Corneal Conundrum</td>
<td>Nandini G Gandhi MD</td>
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<td>4:44 PM</td>
<td>Case 2: Un-nerving—Optic Disc Drusen with Visual Field Loss and Nerve Fiber Layer Thinning on OCT</td>
<td>Erin O Schotthoefer MD</td>
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<td>Case 3: Just Another Failed Vision Screen</td>
<td>Janine E Collinge MD*</td>
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<td>5:06 PM</td>
<td>Case 4: What Is This White Spot?</td>
<td>S Grace Prakalapakorn MD MPH*</td>
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<td>5:17 PM</td>
<td>Case 5: An Extreme Case of Blepharitis</td>
<td>Michelle T Cabrera MD</td>
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<td>5:28 PM</td>
<td>Wrap-up</td>
<td>Tammy L Yanovitch MD</td>
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<td>Yasmin Bradfield MD</td>
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<td>ADJOURN</td>
<td>Jonathan M Holmes MD*</td>
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* Indicates that the presenter has financial interest. No asterisk indicates that the presenter has no financial interest.
Case 1: Stuck Outside—Paralytic Exotropia following Sinus Surgery

Jonathan M Holmes MD

CASE

History

- 82-year-old female
- Unable to move left eye on waking from sinus surgery
- Diplopia in all positions of gaze

Examination

- Visual acuity: 20/20 right eye, 20/30 left eye
- Unable to adduct left eye to midline
- Distance: 35 PD exotropia in straight ahead gaze, 50 PD exotropia in right gaze, 10 PD exotropia in left gaze
- Downgaze 35 PD exotropia and 4 PD left hypertropia
- Near: 40 PD exotropia, 6 PD left hypertropia
- Double Maddox rod: 11 degrees of excyclotropia

Additional Testing

CT scan of orbits showed severed left medial rectus muscle mid-belly.

Intraoperative Testing

Forced ductions: No restrictions

Surgical Options (audience votes)

1. Retrieve medial rectus and reattach
2. Periosteal fixation to medial orbital wall
3. Transpose superior rectus and inferior rectus medially
4. Other

Panel members present their recommended approaches, discussion, and audience re-votes.
Medial Rectus Retrieval

Stacy L Pineles MD

Approach

The approach to a transected muscle after endoscopic sinus surgery is dependent upon the status of the muscle and its innervation. Typically, an MRI or computed tomography scan can be performed to determine whether the proximal stump of muscle is contractile when gaze is directed toward the injured muscle. If there is evidence of contractility, or if the injury was sustained less than 3 months ago (thus allowing for potential neural recovery), then muscle retrieval should be attempted.

Details of Performing the Procedure

There are several published techniques for muscle retrieval, and most of them require the expertise of an orbital surgeon or a head and neck surgeon. Our technique is performed with an orbital surgeon. A transconjunctival approach to the subperiosteal space is utilized. Once the periosteal space is reached, the dissection is carried back toward the orbital apex and the rectus muscle is identified in the posterior orbit. Once the muscle is identified, a double armed nonabsorbable suture is placed through the muscle. The suture is then passed through a standard strabismus conjunctival incision, and the muscle is advanced as far as possible so that the eye is in an orthotropic position, or slightly overcorrected.

References

Periosteal Fixation

Gillian GW Adams MD

Diplopia due to muscle damage following functional endoscopic sinus surgery is a recognized but rare complication of sinus surgery. In this case the medial rectus has been damaged with very little evidence of function, and there should be concern about damage to the left superior oblique muscle. In most of these cases the muscle has been chewed up rather than having suffered a simple transection. Our experience has been that despite an anterior orbitotomy and retrieval of medial rectus remnants combined with botulinum toxin to the lateral rectus, there remains a significant adduction deficit because of the extensive medial rectus trauma.

In these cases of complete adduction failure including medial rectus trauma after ENT surgery, periosteal fixation has provided good alignment in primary position but the ocular movements are restricted. The technique that we have evolved is to fixate both lateral and medial recti to their respective orbital walls combined with skin traction sutures for 4 weeks. We use a retrocaruncular approach to the medial orbital wall to reduce scarring. A small overcorrection is a desirable postoperative alignment, as this will settle as the traction stitches are released. Before we fixed both rectus muscles we found that the alignment drifted, but we have found good long-term alignment with periosteal fixation of both rectus muscles.

References

Case 2: Struck Inside—Esotropia with Conjunctival Scarring

Jonathan M Holmes MD

C A S E

History

- 90-year-old male
- 2-year history of diplopia looking to the right
- Previous history of medial conjunctival intraepithelial neoplasia right eye
- Status post several episodes of excision and cryotherapy

Examination

- Visual acuity: right eye 20/30, left eye 20/25
- “−2” limitation of abduction of right eye
- Distance: 1 PD esophoria straight ahead, 18 PD esotropia in right gaze, orthophoria in left gaze
- Near: 6 PD exophoria
- Diplopia in right gaze, starting 10 degrees into right gaze

Additional Testing

- Divergence amplitudes at distance 4/1
- Convergence amplitudes at near 25/20

Intraoperative Testing

Forced ductions: Limitation of abduction of right eye

Surgical Options (audience votes)

1. Recess medial rectus with conjunctival dissection and graft
2. Recess medial rectus with amniotic membrane
3. Recess medial rectus with routine management of conjunctiva
4. Other

Panel members present their recommended approaches, discussion, and audience re-votes.
Conjunctival Dissection and Graft

*Malcolm R Ing MD*

**My Approach to This Type of Case**

1. Obtain previous operative note, if possible.
2. Measure strabismus in all fields of gaze.
3. Use forced duction test to determine the severity of the restriction of the horizontal (medial) rectus.
4. Use operating microscope to assist in dissection of scar tissue, starting above and below the medial rectus to free the eye and establish normal ductions.
5. Use muscle hook under the medial rectus during the dissection to carefully distinguish the muscle from the scar tissue (therefore, topical anesthesia would be inadequate). Must use general or subtenon local anesthesia.
6. The dissection of the scar tissue must be complete, and the forced duction test must be without any restriction at the end of the dissection.
7. Add a recession of the medial rectus if any esotropia in the primary position is found on original measurement.
8. Cover the defect with an autogenous conjunctival graft.

**Summary**

Restrictive strabismus may be relieved by the appropriate anesthesia and approach, the use of the operating microscope, utilizing a muscle hook under the medial rectus during the dissection, and covering the defect with an autogenous graft.

**Selected Reading**

Amniotic Membrane

Yi Ning Strube MS MD FRCSC

- Restrictive strabismus after periocular surgery is caused by at least 3 mechanisms:
  1. conjunctival scarring
  2. fat adherence syndrome, and/or
  3. rectus muscle contracture.
- Amniotic membrane transplant (AMT) may help prevent recurrence of adhesions in patients with restrictive strabismus.
- Amniotic membrane is immunologically inert; provides a substrate for epithelial growth and attachment; and reduces inflammation, fibrosis, and angiogenesis.
- Cryopreserved amniotic membrane provides results superior to those of lyophilized amniotic membrane, with less scarring and fibrosis reported.
- Secure the amniotic membrane with fibrin glue ± dissolvable sutures as needed to keep the AMT in place.
- Amniotic membrane is relatively easy to manipulate and quick to use, and provides a good cosmetic and functional result.

Selected Readings

Case 3: Down, Not Out—Hypotropia due to Brown Syndrome

Jonathan M Holmes MD

CASE

History
- Nine-year-old male
- Left eye drifts up since birth.
- Right eye doesn’t seem to elevate.

Examination
- Visual acuity: 20/20 right eye, 20/40 left eye
- Limitation of elevation right eye, particularly in adduction
- Distance: 16 PD right hypotropia, worse in left gaze and upgaze
- Near: 10 PD right hypotropia

Intraoperative Testing
- Forced ductions: Limited right eye elevation, particularly in adduction
- Qualitatively tight right superior oblique, assessed by Guyton exaggerated traction test
- Excyclorotation of right eye limited to 10 degrees, assessed by quantitative intraoperative torsional forced duction test

Surgical Options (audience votes)
1. Superior oblique suture spacer
2. Superior oblique silicone band spacer
3. Superior oblique tenectomy
4. Superior oblique recession
5. Other

Panel members present their recommended approaches, discussion, and audience re-votes.

References
Superior Oblique Suture Spacer

Stacy L Pineles

Approach

In a patient with Brown syndrome and a significant hypertropia, a tendon-lengthening procedure is often indicated.

In the past, superior oblique tenotomy was utilized for Brown syndrome, but this procedure has been replaced by tendon-lengthening procedures due to the risk of inducing a large and potentially irreversible overcorrection. Both suture spacers and silicone band spacers are reasonable options that preserve the anterior insertion location of the superior oblique tendon. However, suture spacers are preferred by many surgeons for the following reasons:\textsuperscript{1,2}

1. Ease of procedure
2. Ability to perform adjustable procedures
3. Decreased risk of extrusion or foreign body reaction

Details of Performing the Procedure

Various techniques have been described.\textsuperscript{3-4} Thuangtong and Isenberg\textsuperscript{4} described a useful technique that utilizes a single suture instead of two sutures tied together. For this technique, the superior oblique is isolated and explored. A double-armed 6-0 nonabsorbable suture is placed in the superior oblique tendon in a horizontal mattress fashion. The superior oblique is then transected between the two sides of the horizontal mattress suture. A caliper is used to measure the desired length of suture between the two ends of the tendon as the two ends are pulled apart. A typical starting point is 5 mm of separation. The suture is tied in a slipknot fashion, and forced duction testing is performed. Continued repetition of forced duction testing and subsequent suture length adjustment is performed until there is a restriction to elevation in adduction of approximately -1 to -2 on a 4-point scale. The suture knot is then converted to a permanent knot, and the intermuscular septum and conjunctiva are carefully closed.

References

Superior Oblique Silicone Band Spacer

Yi Ning J Strube MS MD FRCSC

- Silicone tendon expander uses a segment of a silicone 240 or 40 retinal band.
- Provides controlled lengthening of the superior oblique tendon.
- The effect of the spacer can be titrated based on the length of the spacer used, usually between 4 and 6 mm; 7 mm being the maximum length recommended.
- Silicone band is attached to the two cut ends of the superior oblique tendon with nonabsorbable double-armed sutures (ie, 5-0 Mersilene) in a horizontal mattress fashion.
- This creates a fixed separation between the two cut ends of the superior oblique tendon, preventing the two tendon ends from reuniting or scarring at an unpredictable location.
- Surgical technique requires careful manipulation of tissue—not violating the floor of the superior oblique tendon capsule and keeping the nasal intermuscular septum intact, to prevent adhesions and scarring.

Selected Readings

Case 4: Hanging Out—Exotropia after Scleral Buckle

Jonathan M Holmes MD

CASE

History
■ 70-year-old male
■ Nine-year history of right eye wandering out, with intermittent diplopia
■ Onset immediately after scleral buckle for right retinal detachment

Examination
■ Visual acuity: 20/20 right eye, 20/20 left eye
■ Slight limitation of adduction of right eye
■ Distance: 30 PD intermittent exotropia. Right and left gaze 25 PD exotropia
■ Near: 40 PD intermittent exotropia
■ Distance control: Tropic < 50% of time (30-second observation)\(^1,2\)
■ Near control: Recovers in 1-5 seconds after 10-second dissociation\(^1,2\)

Intraoperative Testing
Forced ductions: Mild limitation of adduction of right eye

Surgical Options (audience votes)
1. Recess right lateral rectus and resect right medial rectus (R R&R), leaving scleral buckle
2. R R&R, removing scleral buckle
3. Recess left lateral rectus and resect left medial rectus (L R&R)
4. Bilateral lateral rectus recess
5. Right lateral rectus onabotulinumtoxinA
6. Other

Panel members present their recommended approaches, discussion, and audience re-votes.

References
Horizontal Rectus Muscle Surgery

Malcolm R Ing MD

My Approach to This Type of Case

1. Obtain previous operative notes if possible.
2. Obtain a retinal consult before strabismus surgery. Try to analyze the risk of redetachment.
3. Measure deviation in all fields.
4. Note that the exotropia in this case is greater at the near point of fixation, suggesting restriction of the lateral rectus to inhibit the medial rectus or the medial rectus has slipped posteriorly.
5. Use general anesthesia.
6. Use operating microscope.
7. If the lateral rectus is bound down by scar tissue, dissect all the scar off of the muscle utilizing a muscle hook under the lateral rectus to help complete the dissection.
8. Recess the lateral rectus.
9. Explore the medial rectus; if it has slipped posteriorly, resect and advance the right medial rectus.
10. Since the deviation is so large at near, warn patient that surgery on the other eye may be necessary after the surgery on the right eye.
11. Removal of the scleral buckle may be necessary if it is under the lateral rectus and causing restriction.

Summary

What the surgeon finds at surgery will determine the steps taken to relieve the problem. Removal of the scleral buckle favorably influences the outcome.

Selected Reading

Diplopia immediately following retinal detachment surgery will often improve over a 4-week period. We have found the use of botulinum toxin therapy very helpful after this time to realign the eyes and importantly to check the ability to fuse images. This is particularly important in macular-off detachments where bifoveal fixation may not be possible, leading to intractable double vision. It is important to assess this accurately before considering adjustable suture squint surgery.

Our practice has been to symptomatically support patients (eg, with Fresnel prisms in the immediate postoperative period) and then treat with botulinum toxin therapy. In a small number of patients this produces a cure as the eyes lock together. If a good result is obtained squint surgery can be undertaken, but this can be a technically challenging intervention. In addition to conjunctival scarring there may be muscle damage as well as mechanical difficulties from the presence of a buckle. We have not found scleral buckle removal to benefit motility. Any surgery, therefore, has to be undertaken around the buckle, and the muscle may be stretched or cheese-wired and difficult to handle. Recurrence of retinal detachment is a possibility.

In this case I would use abobotulinumtoxinA injection (Dysport 2.5 units) to the right lateral rectus muscle. Botulinum toxin injection is an outpatient procedure under local anesthetic; it avoids a difficult surgical intervention that may not be successful and that carries with it a risk of retinal detachment. There are small risks from the toxin injection, and for this reason I would advise it be performed under electromyographic control.

Selected Readings

Prism Adaptation
Direction and Misdirection: The Disparity-Driven Vergence System

Kyle Arnoldi CO

I. Fast vs. Slow Vergence

II. Response to Retinal Image Disparity in the Normal System
   A. Fixation disparity
   B. Phoria adaptation
   C. How subtypes of vergence interact

III. Response to Disparity in the Abnormal System
   A. In maldevelopment of vergence systems
   B. In vergence insufficiency
   C. In loss of vergence
      1. Supranuclear lesion
      2. Infranuclear lesion

IV. Uses of Prism Adaptation
   A. Horizontal strabismus with distance–near disparity
   B. Incomitant strabismus
   C. Other

V. Misuses of Prism Adaptation
   A. Dissociation vs. adaptation: The goal of prism adaptation
   B. Anomalous binocular correspondence
   C. Orthotropia with diplopia
      1. Symptomatic phoria
      2. Dragged fovea diplopia syndrome (“retinal” diplopia)

VI. Alternatives to Prism Adaptation
   A. To determine target angle for surgery
   B. To determine fusion potential

Selected Readings
Adhesion Barriers

Yasmin Bradfield MD

I. Introduction and Concepts

II. Types of Adhesion Barriers

III. Animal Studies: Rabbits
A. Mitomycin C (MMC)
1. Strabismus surgery both eyes, then reoperation
2. MMC exposed 5 min to rectus muscles in one eye; other eye control
3. Reduced scarring and adhesions found in MMC eyes.
4. Tensile strength of muscle-scleral junction reduced in MMC eyes.
B. MMC in fat adherence syndrome
1. Adhesion created between inferior rectus and inferior orbital rim
2. One eye treated with MMC; control eye with sterile water
3. Measured passive forced ductions pre- and postop
4. Restriction similar in both groups
5. Histopathologic evaluation demonstrated fibrosis in MMC eyes.
C. Interceed vs. Surgicel vs. Control
1. Evaluated wound healing after trabeculectomy
2. Suppression of vascularization seen in Interceed and Surgicel groups.
3. No reduction in fibroblasts vs. control group
D. Seprafilm
1. Decreased adhesions after strabismus surgery
2. No reduction of inflammation
E. Guardix-SG (poloxamer-alginate mixture)
1. Changes from solution to gel from room to body temperature
2. Reduced adhesions and late fibrosis compared to control
F. Anti-allergy Tranilast
1. Used for symblepharon, pterygium, corneal haze following PRK
2. Tranilast 0.5% drops with fluoroquinolone drops vs. fluoroquinolone drops alone
3. Tranilast reduced adhesion and fibrosis.

IV. Surgical Uses, Other Medical Specialties: Gynecological Surgery
A. Interceed reduced pelvic adhesions in laparoscopic and laparotomy procedures.
B. Gore-Tex reduced pelvic adhesions but requires suture and later removal.
C. Seprafilm: Limited evidence for adhesion reduction in uterine surgery

V. Surgical Uses: Strabismus Surgery
A. MMC
B. MMC with amniotic membrane
C. Amniotic membrane
D. Interceed

Selected Readings
Pullover Traction Suture

Burton Kushner MD

I. Overview
A. Limited indications
B. Very useful in select cases
C. Technically easy
D. Definite downsides

II. Indications
A. Severe or recurrent restrictions that cannot be fully relieved
B. Inelastic muscles when suspension (aka hang-back) or adjustable is desired
   1. Thyroid eye disease
   2. Severely contractured muscles

III. Technique
A. Nonabsorbable suture
   1. Dacron
   2. Mersilene
B. Firm suture placement in sclera or muscle insertion stump
C. Place on side of limbus closest to area of restriction.
D. Two sutures to avoid dragging on cornea
E. Bring through opposite lid.
F. Can be done adjustably
   1. Upper lid if restriction is inferior
   2. Lower lid if restriction is superior
G. Tie over or through cotton bolsters.
H. Leave for 1 week if possible.

IV. Disadvantages
A. Inconvenient
B. Uncomfortable
C. Not necessary if semiadjustable suture used

V. Advantages
A. Keeps stiff muscle back where desired
B. Allows for adjustable sutures on inelastic muscles
C. Allows for sarcomere remodeling in muscle opposite restriction

Selected Readings
Delayed Adjustable Suture
The Great Adjustable Suture Cover-up (Unexposed)!

Richard S Freeman MD

I. History
A. C Burns, 1993
B. R Freeman, 1994
D. D Granet, AAPOS poster, Delayed Adjustable, 2001
E. BJO publication, 2010¹

II. Technique
A. Surgical
B. Postop

III. Results
A. % adjusted
B. Complications

IV. Advantages

V. Disadvantages

VI. Conclusion

Reference
Phospholine Iodide for Residual Esotropia

Laura Enyedi MD

I. Mechanism of Action
   Anti-cholinesterase increases parasympathetic activity.

II. History of Miotics for Esotropia

III. Uses of Phospholine Iodide (aka Ecothiophate Iodide)
   A. Accommodative esotropia
      1. Refractive (normal accommodative convergence-to-accommodation [AC/A] ratio)
      2. Nonrefractive (high AC/A ratio)
   B. Postoperative residual esotropia
   C. Diagnostic use
   D. Spasm of the near reflex

IV. Adverse Effects
   A. Local effects
      1. Common: brow ache, iris cyst, conjunctival hyperemia, allergy
      2. Rare in children: lens opacities, angle closure, retinal detachment
   B. Visual effects
   C. Systemic effects
      1. Usually mild: lacrimation, salivation gastrointestinal, bradycardia, urinary urgency, fatigue
      2. Considerations for general anesthesia using succinylcholine

V. Practical Tips for Obtaining and Using the Drug

Selected Readings
Postoperative Eye Exercises

Cindy Pritchard CO COT

I. Duction Exercises
   A. To minimize contracture and stimulate weakened muscle
      1. Physiology
   B. Methods
      1. Duction sessions
      2. Occlusion of uninvolved eye
   C. Efficacy

II. Fusion Exercises
   A. Indications
      1. To control residual deviation
      2. Control overcorrection
      3. To stabilize surgical results
   B. Methods
      1. Prisms
      2. Convergence training
         a. “Pencil pushups”
         b. Computer programs
         c. Base-out prisms
         d. Stereograms
      3. Divergence training
         a. Computer programs
         b. Base-in prisms
         c. Stereograms
      4. Vertical vergence training: Stereograms
   C. Efficacy of fusion training
      1. Not well supported in literature
      2. Can be cost-effective (stereograms)
      3. Low/no risk

III. Practice Patterns of AAPOS Members
2017 Advocating for Patients
Pamela E Williams MD

Ophthalmology’s goal to protect sight and empower lives requires active participation in and commitment to advocacy from every ophthalmologist. Contributions to the following three critical funds are a part of that commitment:

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

Please join the dedicated community of ophthalmologists who are contributing to protect quality patient eye care for everybody. The OPHTHPAC Committee is identifying Congressional Advocates in each state to maintain close relationships with federal legislators in order to advance ophthalmology and patient causes. At Mid-Year Forum 2017, we honored nine of those legislators with the Academy’s Visionary Award. This served to recognize them for addressing issues important to us and to our patients. The Secretariat for State Affairs is collaborating closely with state ophthalmology society leaders to protect Surgery by Surgeons at the state level. This year has seen an unprecedented effort by optometry to advance its scope of practice via education rather than legislation. Our mission of protecting sight and empowering lives requires robust funding of both the Surgical Scope Fund and the OPHTHPAC Fund. Each of us has a responsibility to ensure that these funds are strong.

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 and protecting ophthalmology from federal scope of practice threats. Established in 1985, OPHTHPAC is one of the oldest, largest, and most successful political action committees in the physician community. We are very successful in representing your profession to the U.S. Congress.

As one election cycle ends, a new one starts, yet the pressure to remain vocal on our issues remains. Advocating for our congressional issues is a continuous battle, and OPHTHPAC is always under financial pressure to support our incumbent friends as well as to make new friends with candidates. These relationships allow us to have a seat at the table with legislators willing to work on issues important to us and our patients.

The relationships OPHTHPAC builds with members of Congress is contingent on the financial support we receive from Academy members. Academy member support of OPHTHPAC allows us to advance ophthalmology’s federal issues. We need to increase the number of our colleagues who contribute to OPHTHPAC and the other funds. Right now, major transformations are taking place in health care. To ensure that our federal efforts and our PAC remain strong, we need the support of every ophthalmologist to better our profession and ensure quality eye care for our patients.

The significant impacts that OPHTHPAC has made include the following:
- Derailed the onerous global surgery data collection proposal
- Preserved global surgical payments
- Halted the Part B Drug Demonstration
- Continued efforts in collaboration with subspecialty societies to preserve access to compounded and repackaged drugs such as Avastin

Contributions to OPHTHPAC can be made here at AAO 2017 or online at www.aao.org/ophtpac by clicking “Join.”

Leaders of the American Association of Pediatric Ophthalmology & Strabismus (AAPOS) and the American Academy of Pediatrics (AAP)–Section on Ophthalmology are part of the Academy’s Ophthalmic Advocacy Leadership Group (OALG), which meets every 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 2017 OALG agenda included panel discussions on the Merit Based Incentive Payment System (MIPS) and APM implementation, as well as Academy analysis initiatives related to the IRIS® registry. In addition, meeting participants discussed the changing paradigm for optometric scope battles, held a roundtable to discuss challenges for surgical subspecialties, and considered opportunities to ensure physician and patient choice regarding access to pharmaceuticals.

At Mid-Year Forum 2017, the Academy, AAPOS, and AAP–Section on Ophthalmology ensured a strong presence of pediatric ophthalmologists to support ophthalmology’s priorities, and a record number of ophthalmologists visited members of Congress and their key health staff to discuss ophthalmology priorities as part of Congressional Advocacy Day. The AAPOS and AAP–Section on Ophthalmology remain crucial partners with the Academy in its ongoing federal and state advocacy initiatives.

Surgical Scope Fund

The Surgical Scope Fund (SSF) provides grants to state ophthalmology societies to support their efforts to derail optometric surgery proposals that pose a threat to patient safety. Since its inception, the Surgery by Surgeons campaign and the SSF, in partnership with state ophthalmology societies, has helped 32 state / territorial ophthalmology societies reject optometric scope of practice expansion into surgery.

In 2017, your colleagues serving on the Academy’s Secretariat for State Affairs, along with State Governmental Affairs staff and the leaders of state ophthalmology societies, have been put to the task while dealing with an unprecedented number of simultaneous legislative battles. Eleven states have been affected so far this year:
- Alaska
- California
- Florida
- Georgia
- Illinois
- Iowa
- Maryland
- Massachusetts
- Nebraska
- North Carolina
- Pennsylvania

Continued efforts in collaboration with subspecialty societies to preserve access to compounded and repackaged drugs such as Avastin

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Patient safety setbacks as well as victories will be reviewed during the presentation, but do know that in each of these legislative battles, the benefits from SSF distributions are abundantly clear. The best lobbyists and public relations consultants are contracted as necessary, and media campaigns (including TV, radio, and social media) to educate the voting public are launched when needed to secure success and stop optometry from expanding its scope of practice to include surgery. Each of these endeavors is very expensive, and no one state has the resources to wage one of these battles on its own. Ophthalmologists must join together and donate to the SSF to fight for patient safety when a state faces a scope battle over optometric surgery.

The Academy relies not only on the financial contributions to the SSF from individual ophthalmologists and their practices, but also on the contributions made by ophthalmic state, subspecialty, and specialized interest societies. The AAPOS contributed to the SSF in 2017, and we thank them. We look forward to AAP–Section on Ophthalmology’s contribution in 2017 as well. Contributions to the SSF can be made here at AAO 2017 or online at www.aao.org/ssf.

State Eye PAC

It is also extremely important for all ophthalmologists to support their respective State Eye PACs because campaign contributions to legislators at the state level must come from individual ophthalmologists and cannot come from the Academy, OPHTHPAC, or the SSF. 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 critical, as scope of practice battles and many regulatory issues are all fought on the state level.

Action Requested: ADVOCATE FOR YOUR PATIENTS

Academy SSF 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 help us protect sight and empower lives. SSF 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.

<table>
<thead>
<tr>
<th>Surgical Scope Fund</th>
<th>OPHTHPAC* Fund</th>
<th>State EyePAC</th>
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<td>To derail optometric surgical scope of practice initiatives that threaten patient safety and quality surgical care</td>
<td>Ophthalmology’s interests at the federal level</td>
<td>Support for candidates for state House, Senate, and governor</td>
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<td>Political grassroots activities, lobbyists, PR and media campaigns</td>
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<td>No funds may be used for campaign contributions or PACs</td>
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<td>Contributions: Unlimited</td>
<td>Contributions: Limited to $5,000</td>
<td>Contribution limits vary based on state regulations.</td>
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<td>Individual, Practice, and Organization</td>
<td>Contributions above $200 are on the public record.</td>
<td>Contributions are on the public record depending upon state statutes.</td>
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<td>Contributions are 100% confidential.</td>
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Please respond to your Academy colleagues and be part of the community that contributes to OPHTHPAC, the SSF, and your State Eye PAC. Please be part of the community advocating for your patients now.

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The Boston keratoprosthesis (KPro) is the most commonly implanted artificial cornea in the world, with approximately 13,000 implanted in total and approximately 900-1000 per year placed currently. Developed by Claes Dohlman, the Boston KPro has a collar-button design whose components are sandwiched between a donor cornea and sewn into place in a fashion similar to penetrating keratoplasty.

The most common indication for KPro is failed graft, although in recent years, primary KPro has been used for limbal stem cell dysfunction and in the setting of corneal vascularization or anesthesia, diagnoses that do poorly with traditional penetrating keratoplasty (PK). The device comes in aphakic and pseudophakic models and consists of a front optical piece, a backplate with holes to allow access of aqueous to the donor cornea, and a locking ring. A recently introduced 2-piece KPro with a “click-on” titanium backplate eliminates the need for the locking ring, thus simplifying device assembly.

Esen Akpek and colleagues described the first pediatric KPro cases more than a decade ago. Despite this, limited literature exists regarding use of the device in children. A recent AAO Technology Assessment on KPro\(^1\) concluded that there was not enough evidence in the literature to make a recommendation regarding Boston KPro use in children.

The most common complication after KPro is retroprosthetic membrane, which occurs in up to two-thirds of cases in adults. These can be especially aggressive in children, eventually resulting in device extrusion. Glaucoma is currently the biggest threat to long-term preservation of vision after KPro in adults and affects virtually all children after KPro implantation. This is an area that needs much more work. Other complications after KPro include persistent epithelial defects, infectious keratitis, sterile corneal necrosis, endophthalmitis, retinal or choroidal detachment, and idiopathic vitritis. Further work is needed to reduce complications and improve KPro outcomes in all patients, both young and old.
Pediatric Keratoprosthesis: Pro

Sarah Nehls MD

I. Boston Keratoprosthesis (KPro) Background
   A. Prosthetic corneal transplant suspended in donor corneal tissue
   B. 7-mm (pediatric) vs. 8.5-mm (adult) back plate
   C. Aphakic KPro is most often used in pediatric patients; optic power is based on axial length band can be selected anticipating growth of eye.

II. The Boston keratoprosthesis in children provides:
   A. Immediate visual rehabilitation
   B. More predictable visual rehabilitation
   C. Use of a soft contact lens to correct refractive error
   D. Immediate amblyopia management
   E. Immediate view of optic nerve and retina
   F. No rejection
   G. No astigmatism

III. Case
   A. The following case shows a 14-month-old with bilateral corneal opacities and glaucoma from Axenfeld-Reiger syndrome that underwent sequential, bilateral Boston keratoprosthesis surgery.

Figure 1. Corneal opacities and buphthalmos, right eye (A), left eye (B), age 14 months.
B. The patient showed immediate improvement in meeting developmental milestones. Retention and clarity of graft maintained to date, 20 months after surgery. Visual evoked potential (VEP) measurement predicts 20/70 vision in the right eye and 20/120 vision in the left eye.

References


Figure 2. Six months after KPro, lensectomy, endocyclophotocoagulation: right eye (A), left eye (B).
Conjunctival Stem Cell Transplantation for Aniridia

Edward J Holland MD

I. Incidence and Background
   A. Congenital aniridia is a disorder that affects 1/64,000 to 1/96,000 live births.
   B. It is inherited in an autosomal dominant pattern but can also have an autosomal recessive or sporadic pattern.
   C. Aniridia affects all parts of the eye and results in iris deformities, foveal hypoplasia, optic nerve hypoplasia, nystagmus, glaucoma, cataracts, and aniridic keratopathy.
   D. Over 90% of patients with aniridia develop aniridic keratopathy.

II. Aniridic Keratopathy Grading Scale Based on Severity
   A. Stage 1 includes a thickened, irregular peripheral epithelium and late staining with fluorescein.
   B. Stage 2 occurs when the peripheral epitheliopathy moves centrally but still spares the absolute central cornea.
   C. At Stage 3, the epitheliopathy involves the central cornea.
   D. At Stage 4, the cornea displays complete epitheliopathy and subepithelial fibrosis.
   E. At Stage 5, the cornea exhibits total epitheliopathy and deep scarring involving the stroma.
   F. Corneal neovascularization can occur during any stage and is not specific to any stage of keratopathy.

III. Management of Aniridic Keratopathy
   A. Supportive therapy (ie, lubrication, bandage contact lens, tarsorrhaphy)
   B. Primary keratoplasty: Inevitable failure
   C. Ocular surface stem cell transplantation (OSST)
   D. Keratoprosthesis

IV. Limbal Stem Cell Transplant Indications

V. Surgical Options
   A. Living-related conjunctival limbal allograft (LR-CLAL) vs. keratolimbal allograft (KLAL)
   B. Description of procedures

VI. Preoperative Management
   A. Addressing glaucoma, iris, and cataract comorbidities
   B. Laboratory testing
   C. Medical evaluation for systemic immunosuppression candidacy
   D. Identification of living related donors
   E. Trial of systemic immunosuppression

VII. Postoperative Management
   A. Follow-up
   B. Laboratory monitoring
   C. Systemic immunosuppression regimen
   D. Subsequent keratoplasty

VIII. Outcomes: Aniridic and Pediatric OSST
   A. Visual outcomes
   B. Ocular surface stability

IX. Complications
   A. OSST failure
   B. Keratoplasty failure
   C. Glaucoma
   D. Infectious keratitis
   E. Keratoprosthesis for aniridic keratopathy
Pediatric Keratoconus: Crosslinking vs. Penetrating Keratoplasty vs. Anterior Lamellar Keratoplasty

Beatrice E Frueh MD

Keratoconus (KC) diagnosed in childhood has a faster progression and a worse outcome than in adults. Lamellar and/or penetrating keratoplasties can dramatically improve best spectacle-corrected visual acuity (BSCVA), but they have all the drawbacks of corneal transplantation in the younger age group, even if the keratoconus group has the best prognosis among children. Keratoplasties can be performed at later stages of the disease, when contact lenses cannot correct the ametropia anymore, but they cannot prevent disease progression.

Corneal crosslinking (CXL) is a proven and safe method to stiffen the cornea and to halt keratoconus progression; it has recently been approved by the FDA for treatment of progressive keratoconus in patients older than 14 years.

There are some published articles on CXL in children, few with 4-5 year results, and most of them have lost a large percentage of subjects to follow-up. Deviations from the epi-off, standard Dresden protocol, riboflavin, 3 mW/cm², like accelerated CXL or iontophoresis, are not discussed due to lack of long-term results in children.

Our Results

- Standard CXL in children
- 65 eyes of children with KC with a minimum follow-up of 1 year
- Inclusion criteria: Follow-up of 6 years or progression at earlier time point
- 17 eyes of 15 patients aged 4 to 17 years, and 2 cases of progression at 3 and 2 years were included.
- Topographic and tomographic values remained unchanged or improved.
- No complications occurred; no cases of loss of ≥ 2 lines of BSCVA.
- The corneas thinned constantly during follow-up.
- The cases of progression have been retreated.

CXL in children is as safe as in adults and is very effective in halting KC progression. Children need to be checked postoperatively, because progression can occur 2 to 3 years after CXL, eventually necessitating a retreatment.

References

Red Eye in Children: A Cornea Specialist’s View

Kristin M Hammersmith MD

I. Common Causes of Red Eye in Children
   A. Viral conjunctivitis
      1. RPS Adenoplus: In-office device used to detect adenoviral conjunctivitis
      2. Traditional therapy and possible use of ganciclovir gel
   B. Bacterial conjunctivitis
   C. Allergic conjunctivitis
   D. Trauma
   E. Toxicity

II. Common “Red Eye” Conditions for Cornea Specialist
   A. Blepharokeratoconjunctivitis
   B. Herpes simplex keratitis (HSK)
   C. Atopic / vernal keratoconjunctivitis

III. Blepharokeratoconjunctivitis
   A. Epidemiology
      1. Most common condition for children at Wills Eye Hospital Cornea, accounting for 15% of referrals. Racial demographics affect the prevalence of this disorder. More common in Asian and Middle Eastern children.
      2. Girls more commonly affected than boys.
      3. May present from 6 months of age to teenage years, most common at 4-5 years.
   B. Clinical manifestation
      1. Anterior and posterior lid inflammation, often recurrent chalazia at initial presentation
      2. Conjunctival hyperemia
      3. Corneal inflammation: superficial punctate keratitis, peripheral ulcerations, and/or classic phlyctenule
      4. Corneal scarring
      5. Bilateral, but often asymmetric
      6. Reduced visual acuity and amblyopia
      7. Facial rosacea present in 20%-50%
   C. Treatments
      1. Lid therapy, dilute baby shampoo vs. commercially available products
      2. Warm compresses, consider moist heat packs (Bruder)
      3. Consider flax seed oil
      4. Topical antibiotics with broad gram-positive coverage (erythromycin, bacitracin, azithromycin)
      5. Oral erythromycin, azithromycin, or doxycycline (mature dentition)
      6. Preservative free lubricants
      7. Steroids, lower potency often effective
      8. Cyclosporine (0.0.5% to 2%)
      9. Rarely used: oral steroids, triamcinolone injection, oral immunosuppression

IV. Herpes Viral Keratitis
   A. Herpes simplex
   B. Varicella zoster
   C. Epstein-Barr
   D. Cytomegalovirus

V. Herpes Simplex Anterior Segment
   A. Clinical manifestation
      1. Herpes blepharoconjunctivitis (HBC)
         a. Suspect HSV in recurrent unilateral conjunctivitis or in conjunctivitis in eye with a history of HSV.
         b. Bilateral HBC more common in children than adults.
         c. Patients with bilateral HBC, increased rates of asthma, atopy, and systemic disease
      2. Dendritic keratitis
      3. Stromal keratitis
      4. Reduced vision from corneal scarring and refractive amblyopia
   B. Common misdiagnosis: blepharokeratoconjunctivitis, bacterial keratitis
   C. Recurrence common (40%-80%); median recurrence time, 12-15 months

D. Treatments
   1. Topical therapy with trifluridine 9 times a day or topical ganciclovir gel 5 times for dendritic keratitis. Oral therapy may be used as treatment in place of topical options.
   2. Dosing of oral therapy (acyclovir suspension 200 mg/5mL)³
      a. Infants (up to 18 months): 100 mg t.i.d. for treatment; 100 mg b.i.d. for prophylaxis
b. Toddlers (18 months to 3 years): 200 mg t.i.d. for treatment; 200 mg b.i.d. for prophylaxis

c. Young children (3-5 years): 300 mg t.i.d. for treatment; 300 mg b.i.d. for prophylaxis

d. Older children (6 and up): 400 mg t.i.d. for treatment; 400 mg b.i.d. for prophylaxis

3. Steroid therapy useful for stromal keratitis. Often very slow taper.

VI. Atopic / Vernal Keratoconjunctivitis

A. Atopic keratoconjunctivitis

1. Atopy: hereditary condition with ocular, respiratory, and dermatologic manifestations

2. Increased type I hypersensitivity and impaired cell mediated immune response

3. Increased HSV/staph infections

B. Vernal keratoconjunctivitis

1. Seasonal, ocular inflammatory disease involving tarsal and/or bulbar conjunctiva

2. Greater prevalence in boys living in warm climates

3. Personal or family history of atopic disease

4. Immunoglobulin E (IgE)-dependent (type 1) and IgE-independent (type 2) forms exist.

5. Medical therapy, localized

a. Mast cell stabilizers / antihistamines

b. Topical steroids

c. Topical cyclosporine A (0.05%-2%)

d. Topical tacrolimus (0.05%-0.1% drops, 0.03% ointment): Combination therapy with topical cyclosporine drops and tacrolimus ointment.

e. Interferon (IFN) alpha-2b: Randomized double-masked study comparing 0.005% topical tacrolimus to IFN alpha-2b (1,000, 000 units/cc) in 40 patients found both effective and safe in recalcitrant vernal keratoconjunctivitis. Cornea. 2017; 36(6): 675-678.

6. Medical therapy, systemic

a. Cyclosporine A, tacrolimus, oral steroids, or other immunosuppression

b. Omalizumab (Xolair): Recombinant humanized monoclonal antibody binding free IgE, approved for asthma. Small, recent case series demonstrating potential benefit for this subcutaneous therapy.

7. Surgical therapy

a. Surgical resection of papillae

b. Supratarsal triamcinolone injection

c. Shield ulcer: Debridement and amniotic membrane placement

References and Selected Readings

Blepharokeratoconjunctivitis


Herpes simplex anterior segment


Vernal keratoconjunctivitis


Case 1: What Did You Do to My Baby? A Ptosis Surgery Saga

K David Epley MD

I. Congenital Ptosis Frontalis Sling Surgery
   Surgical options include:
   A. Harvesting facia lata
   B. Banked fascia lata
   C. Suture
   D. Ptose-Up (ePTFE: expanded polytetrafluoroethylene)
   E. Silicone rods
   F. Others

II. Robert W Gore
   A. One of the “scientists you must know” along with Nicola Tesla, but that’s another talk. …
   B. Inventor, along with his father, Wilbert L. Gore, of ePTFE
      1. Gore-Tex
      2. Repels liquid while allowing vapor to pass through
   C. ePTFE is “stretched” PTFE, better known as Teflon.

III. Ptose-Up
   A. Designed by JM Ruban MD and sold by FCI Ophthalmics
   B. “Ptosis strips from FCI Ophthalmics have been developed to treat frontalis suspension in patients with significant ptosis and poor levator functioning. FCI Ptose-Up strips are easy to place and adjust, and they provide an excellent eyelid contour. The strips are made of a biocompatible, porous, inert, biointegrable, non-toxic, ready to use, non-allergenic material. FCI ptosis strips can be removed in the case of overcorrection, or if the patient experiences dry eye problems as a result of treatment.” http://www.fci-ophthalmics.com/ lid-repair#strips

IV. Case #1
   A. Two-month-old with nearly complete ptosis of the left eye
      1. Underwent frontalis sling procedure using 2-mm Ptose-Up
      2. Excellent lid crease and eyelid position until 20 days postop
   B. 20 days postop
      1. Preseptal cellulitis appearance with skin erythema and edema of the upper eyelid and brow
      2. Amoxicillin started
   C. 25 days postop
      1. Skin overlying the nasal infrabrow incision is erythematous with pustule at the site of this incision.
      2. Sutures removed and pus drained in office
      3. Neomycin / polymyxin and amoxicillin continued
      4. Slowly improves with multiple spontaneous episodes of drainage and 1 additional lancing with 30-gauge needle over the next 10 days
   D. 31 days postop
      1. It’s baaaack!
      2. Resumed higher dose amoxicillin (60 mg/kg/day vs. 40 previously)
      3. Proceeded to battle this repeatedly over the next 2 months, with the “infection” recurring each time the antibiotics were stopped.
      4. Oral steroids had no effect (thinking recurrent inflammatory reaction).
   E. Two months postop
      1. Returned to OR, drained abscess, removed cystic lining
      2. Problem solved!
   F. Five days status post abscess drainage and cystectomy
      1. Are you kidding me? Red, inflamed pustule is back.
      2. Culture at the time of surgery was positive for MRSA
      3. Trimethoprim / sulfamethoxazole was started.
      4. Ten days later, she was looking good, except…
   G. One month following incision and drainage of abscess
      1. Infection resolved
      2. Now the suprabrow incision has some of the Gore-Tex material protruding.
3. Cut flush with the skin using Wescott scissors in the office.

4. Continued use of topical antibiotic (mupirocin)

H. Two months following incision and drainage of abscess
   1. Material extruded further; parents pulled on it and pulled out a segment of the material that was, at this point, loose.
   2. Infection resolves.
   3. Skin heals.
   4. Unfortunately, her eyelid height is not as good as it was before the infections.

V. Case #2: What Did You Do to My Kid’s Eye?!?!?
   A. Even though you prepare them for the eye looking really open postoperatively …
   B. One month postop
      1. Suprabrow incision inflamed without pustule
      2. Low grade fever, 100.5
      3. Here we go again! Increased Tobradex, started cephalexin
      4. Family going to Hawaii for vacation …
   C. Eight weeks postop
      1. Finished cephalexin, pustule and inflammation returned 3-4 days later
      2. Placed on oral cephalexin and Tobradex topically
      3. Recurred off medication, to back to the OR to drain
   D. Three months postop, frontalis sling
      1. Wound debrided
      2. Gor-Tex “arms” cut flush with the knot
      3. Irrigated with antibiotics, 2-layer closure (frontalis and skin)
   E. One month postop debridement
      1. Mom calls to report another pustule at the suprabrow incision.
      2. Cephalexin started
      3. Mupirocin topically
   F. Two months status post debridement
      1. Back to the OR to remove the sling material
      2. Postop course this time was uneventful, thank goodness!
   G. Nine months status post frontalis sling
      1. Fully healed
      2. Some frontalis muscle atrophy from the recurrent infections
   H. Two years out: Muscle has filled in and appearance is good

VI. Summary of Complications
   A. Approximately 10% of my patients react to the Ptose-Up material (2 of 22 eyelids in my series).
   B. Chronic, recurrent infection with inflammation
   C. Can lead to local destruction of muscle
   D. Responds temporarily to antibiotics, but infection and inflammation recur repeatedly if the sling material is not removed.

VII. Lessons Learned
   A. The best solution is to remove the sling material if it has been long enough postop not to jeopardize the eyelid height (3 months at least in my experience).
   B. Antibiotics can be used to temporize until enough time has passed.
   C. Ptose-Up works to create a nice postop appearance with minimal thickening of the eyelid and eyebrow areas and it holds the eyelid well, but …
   D. 10% risk of reaction to the material is significant and needs to be discussed with families preoperatively during the consent process.
Case 2: Discomfort and Dyscoria

Lawrence Tychsen MD

C A S E

An intraocular collamer phakic IOL (Visian ICL) of −14 D was implanted in the right eye of a 5-year-old boy for treatment of anisometropic amblyopia. A YAG-laser peripheral iridotomy was performed under general anesthesia with the child in the lateral decubitus position before being repositioned supine for ICL surgery. The 2 small corneal incisions were closed with 9-0 Vicryl suture. The child was discharged from the same-day surgery unit 1 hour after the procedure.

At postop Day 1 exam the mother reported that the child had slept fitfully and attempted repeatedly to rub the operated eye. Examination revealed significant dyscoria with the iris peaked toward the 12 o’clock position.

Differential Diagnosis of Postoperative Day 1 Pediatric Phakic IOL Problem

Discomfort
- Photophobia of mild uveitis
- Ciliary muscle spasm
- Exposed suture end / knot
- Anesthesia-related nausea
- Diamox-related nausea
- Ocular hypertension (OHT) of retained anterior chamber viscoelastic
- OHT of pupillary block
- Eye rubbing wound rupture
- Postop ondansetron (Zofran)-induced migraine
- Endophthalmitis

Dyscoria / peaked pupil
- Impatient iridotomy OHT / pupillary block / wound gape
- OHT of retained viscoelastic causing wound leak
- Eye rubbing suture breakage wound gape
- Suboptimal spacing haptic enclavation (Ophtec pIOL)
- Poor suture placement / trimming wound leak
- Suture incarceration of nubbin peripheral iris at closure
- Displaced IOL
- Pupillary sphincter damage during surgery

Selected Readings
Case 3: When the Third Eye Is Not So Wise

Shira L Robbins MD

CASE

A 60-year-old man was found to have giant orbital mass as incidental finding during workup for peripheral neuropathy-type complaints. History of nystagmus and strabismus since infancy; underwent surgery for exotropia 30 and 55 years prior to presentation. No complaints of diplopia.

Examination revealed a small-angle strabismus and nystagmus with moderate limitation of O.S. to adduction. Review of orbital MRI showed large, well-circumscribed, nonenhancing mass measuring almost 2 cm within the distal left medial rectus muscle-tendon complex, with mass effect causing mild lateral displacement of the left globe.

Differential Diagnosis

Orbital mass resultant to strabismus surgery

- Orbital hemorrhage
- Orbital epithelial cyst
- Foreign body
- Fat prolapse

Orbital mass coincident to strabismus surgery

- Vascular-lymphatic malformations
- Optic nerve or nerve sheath based
- Infection / inflammation (mass forming)
- Benign
- Malignant

Treatment / Postoperative Course

Surgical approach by orbital surgeon: “Not taking that out without you in the room.” Joint surgical approach with strabismologist and oculoplastics specialist. Forced ductions showed restriction to adduction and abduction. Upon dissection, mass found between muscle and Tenon capsule just posterior to recessed insertion. The mass was found to have a few muscle fibers running above it, with a majority of the medial rectus muscle fibers compressed below the mass. Pathology will be discussed.

Postoperatively, the patient’s adduction deficit was greatly improved. Small-angle strabismus and nystagmus persisted consistent with lifelong conditions; however, no diplopia was appreciated.

Selected Readings


Case 4: That Bump Shouldn’t Be There!

Gregg T Lueder MD

CASE

A 9-month-old infant underwent bilateral medial rectus recessions for treatment of infantile esotropia. The surgery was performed through a fornix incision made 8 mm from the limbus. The wound was closed with cautery. One month after surgery the patient’s mother reported a lesion on the conjunctiva. Examination revealed a fleshy, elevated red mass at the site of the conjunctival incision.

Discussion

The patient’s lesion appeared to be a pyogenic granuloma (PG). Treatment of such lesions will be discussed.

Pyogenic Granuloma

- PGs are reactive lesions that occur in the setting of ocular inflammation or irritation.
- Etiology not clear
- May occur in patients with conjunctivitis, chalazia
- May develop following surgery involving transconjunctival incisions
- Develop in approximately 1 in 50 patients following strabismus surgery
- Presence of suture material not required for formation

Pathology

- Actually, neither pyogenic nor granulomas
- Mixed chronic and acute inflammation with capillary proliferation

Treatment

- Topical steroids or topical steroid / antibiotic combination, 2-4 times/day for 2-4 weeks
- Approximately 90% of lesions resolve with topical treatment.
- Surgical excision may be performed for those that persist.
- Newly described treatment: Topical timolol

Selected Readings

Case 5: “Weird Vision” after Strabismus Surgery

Deborah VanderVeen MD

CASE

A 6½-year-old boy with poorly controlled V-pattern exotropia (30 PD, distance and near, 3+ inferior oblique overaction) in setting of Schwartz-Jampel syndrome underwent bilateral lateral rectus recession and inferior oblique myectomy. The surgery was performed via inferotemporal fornix incisions, with care taken to avoid excessive manipulation of the globe, and lateral rectus recession was performed via hang-back technique. The patient was doing well at the first postoperative visit, but presented 5 weeks after surgery complaining of “weird vision” since surgery; and for the previous 2-3 days, “red floaters.”

Postoperative examination showed a residual small-angle intermittent exotropia brought out only after cover testing, minimal residual inferior oblique overaction, and full motility. Visual acuity in the right eye was stable at 20/70 (amblyopia), but decreased in the left eye to 20/200, with no improvement by pinhole or refraction.

Discussion

Systemic diagnoses and associated ocular findings may require special attention prior to, during, and after strabismus surgery.

Differential diagnosis of visual disturbance (not diplopia) after strabismus surgery:

- Ocular surface abnormality, corneal disease, change in refractive error, accommodative disorder, inflammatory reaction, infection, cataract / lens abnormality, hemorrhage (intraocular, orbital), retinal edema or detachment, optic neuropathy, central / intracranial process, iatrogenic injury, functional

- Timing and clinical findings help narrow differential diagnosis.

Findings for this case and management reviewed.

Selected Readings


Case 6: Forgiving SINS

Radha Ram MD

CASE

A 61-year-old otherwise healthy gentleman was referred for evaluation and management of exotropia. He had long-standing strabismus with no diplopia and was interested in surgical correction. He had no other complaints. His past medical history was negative for known autoimmune disorders. His past ocular history was significant for advanced glaucoma and prior strabismus surgeries in adulthood. He was monocular due to his advanced glaucoma. Examination revealed hand motion vision in the right eye and 20/25 vision in the left eye. He had a comitant, large-angle sensory exotropia. The patient underwent a re-resection and re-recession of the horizontal muscles in the right eye. One week after surgery, the patient denied pain, had a scant amount of discharge, and had excellent alignment.

Ten days after surgery, car debris fell into the patient’s eyes, and he subsequently developed pain in the operated eye. Exam revealed a subconjunctival abscess with mucopurulent material that was drained and cultured in the clinic. Cultures grew methicillin-resistant *Staphylococcus aureus* that was sensitive to clindamycin and vancomycin. The patient improved on oral clindamycin and topical fortified vancomycin.

After finishing his 14-day course of oral clindamycin, the patient developed severe pain. Since the inflammation continued to progress despite antimicrobial therapy, the patient was taken to the operating room for surgical drainage. Intraoperatively, there was no mucopurulent discharge. There was a scleral perforation with exposed uveal tissue anterior to the medial rectus muscle insertion. A corneal patch graft was placed over the area of scleral perforation. The area was debulked and cultured. Necrotizing scleritis was suspected and oral steroids were initiated. Cultures were negative. Laboratory evaluation revealed elevated uric acid, consistent with a diagnosis of gout. Over the ensuing weeks, complete remission of the scleral inflammation was observed on immunosuppressives.

Discussion

This is a rare case of surgically induced necrotizing scleritis (SINS) after strabismus surgery in a patient with gout that was treated with a corneal patch graft. This case illustrates the difficulty in identifying a rare complication that masquerades as and may be triggered by an infection.

Several aspects of this case set it apart from previously described cases. Although SINS has been associated with strabismus, the majority of SINS cases occur after pterygium and/or cataract surgery. The relationship between SINS and infection is not well characterized. Some believe SINS may be caused by infection, while others suggest it may predispose to secondary infection. The most commonly identified systemic diseases associated with SINS are rheumatoid arthritis and Wegener granulomatosis. SINS has been associated with gout in three cases in the literature, all of which were diagnosed after pterygium surgery. There has been only one other reported case describing the use of a corneal graft for SINS-related scleral melting which was done after pterygium surgery.

The diagnosis and management of SINS is challenging. This case offers further insights into a rare but vision-threatening and potentially life-threatening diagnosis.

Selected Readings

Classification of Intermittent Exotropia

Katherine J Fray BS CO

I. Distance–Near Relationship
A. Basic exodeviation (distance = near)
B. Convergence insufficiency (distance < near)
C. Divergence excess (distance > near)
1. True divergence excess
2. Pseudo divergence excess: Near deviation appears to be less than distance due to vergence after-effects or a high accommodative convergence-to-accommodation (AC/A) ratio

II. Testing Strategies That May Impact the Size, Classification, and/or Treatment of the Deviation
A. Stereopsis
B. Target size
C. Meticulous cover test technique
D. Measure at optical infinity
E. Patch test (prolonged occlusion)
F. Binocular visual acuity
G. AC/A ratio using the gradient method
1. Assess using plus lenses at near; best if done after prolonged occlusion
2. Can be assessed using minus lenses at distance without prolonged occlusion
H. Convergence accommodation to convergence (CA/C) = A measure of how much accommodation is stimulated by convergence
I. Convergence fusional amplitudes
J. Depth of suppression measured using a red filter bar
K. Prism adaptation test

III. Control of Near Deviation
A. Fusional mechanisms
1. Tenacious proximal fusion
2. Remeasure after prolonged occlusion
3. Respond better to surgical correction
B. Accommodative convergence
1. Assess the AC/A ratio
2. Respond better to optical correction

IV. Assessment of Control: Requires Multiple Measures or Longer Periods of Observation
A. Parental reporting
B. Office assessment using control scales
C. Time of day, alertness of patient, and level of fatigue

Selected Readings
The Natural History of Intermittent Exotropia

Donny Won Suh MD

Purpose
To determine the rate of development of constant exotropia (XT) over a 3-year period in children 12 to 35 months of age with untreated intermittent exotropia (IXT). The most recent results from the Pediatric Eye Disease Investigator Group (PEDIG) study will be discussed.

Methods
Ninety-seven children aged 12 to 35 months with previously untreated IXT were randomly assigned to observation as part of a multicenter trial. No IXT treatment other than refractive error correction was to be prescribed unless motor or stereoaucity deterioration criteria were met, with protocol-allowed exceptions for starting treatment being overwhelming social concern, debilitating diplopia, or failure to maintain stereoaucity age norms. Follow-up visits occurred at 3 and 6 months and at subsequent 6-month intervals until 3 years postrandomization. Ocular alignment was measured at each visit, with motor deterioration defined as constant XT of ≥ 10Δ at distance and near. Stereoaucity was measured at near starting at 36 months of age, with stereoaucity deterioration defined as a decrease of ≥ 2 octaves from best previous measure. Both assessments were performed by a masked examiner, with deterioration confirmed by a retest. The cumulative probability of meeting motor deterioration by 1, 2, and 3 years and 95% confidence intervals (CI) were calculated using Kaplan-Meier survival analysis.

Results
The cumulative probability of developing a constant XT of ≥ 10Δ at distance and near (motor deterioration) was 4% (95% CI, 1%-11%) by 1 year, 8% (95% CI, 4%-17%) by 2 years, and 10% (95% CI, 5%-19%) by 3 years. In addition to the 7 subjects who met motor deterioration, 2 had nonsurgical IXT treatment after meeting stereoaucity deterioration, and 15 had treatment without meeting either criterion (8 had surgery and 7 had nonsurgical treatment; 12 were protocol-allowed exceptions).

Conclusion
Because the development of a constant XT over 3 years was uncommon among untreated 12- to 35-month-old children with IXT, a close observation appears to be a reasonable approach for this population as long as patients do not manifest any signs of deterioration.
Intermittent exotropia (IXT) is the most common form of childhood exotropia and the most prevalent form of strabismus in some populations. IXT is characterized by periods of normal binocular alignment and sensory fusion some of the time and a manifest exotropia present at other times. Both surgical and nonsurgical management options are commonly prescribed, but there is controversy regarding both the optimal timing and method of treatment. Patching treatment—either patching the preferred eye or alternate patching—is one of several prescribed nonsurgical treatments for children affected by IXT.

Published studies have varied in terms of patching dosage, duration, and outcome measures and have reported varying success rates. Furthermore, studies have been primarily retrospective, with small sample sizes and conducted without comparison groups. As a result, there is no convincing evidence supporting the effectiveness of patching treatment for IXT.

The Pediatric Eye Disease Investigator Group has conducted the Intermittent Exotropia Study 2 (IXT2), a multicenter, randomized clinical trial to determine the effectiveness of prescribed part-time patching as treatment for reducing the risk of deterioration of IXT among 3 to < 11-year-old children over a 6-month period.

The results of the study from 358 children aged 3 to <11 years old with previously untreated (except for refractive correction) IXT and near stereoacuity of 400 arcsec or better will be discussed.

Selected Readings


Overminus Spectacles for Intermittent Exotropia: Initial Results from a Pilot Study

S Ayse Erzurum MD

Introduction

Intermittent exotropia (IXT), the most common form of childhood exotropia, is characterized by normal ocular alignment some of the time and a manifest exotropia at other times. Although surgery is often considered for treatment, many cases of IXT are treated using nonsurgical interventions such as overminus lenses or occlusion. One proposed mechanism for improvement with overminus lens therapy is that stimulation of accommodative convergence reduces the angle of exodeviation or triggers reflex convergence. An alternative hypothesis is that fusional convergence is exerted to control the exodeviation, inducing convergence accommodation and distance blur, and that overminus lenses may allow clear distance vision, facilitating fusion.

In some patients, overminus lenses alone appear to be successful in treating IXT, with eventual weaning of the overminus lenses to a point at which the IXT is well controlled in the regular refractive correction. Nevertheless, previous studies of overminus lens therapy have been limited to retrospective case series without comparison groups and have varied in terms of methods used to determine the amount of overminus power, treatment duration, and outcome measures.

The Pediatric Eye Disease Investigator Group did evaluate this question with a pilot study, which we will discuss. The objective of this study was to evaluate the initial, short-term effectiveness of prescribing overminus lens therapy to improve control of IXT among children aged 3 to 6 years.

Methods

We conducted a randomized clinical pilot trial enrolling 58 children, 3 to <7 years old, with IXT. Eligibility criteria included a distance control score of 2 or worse (mean of 3 measures during a single examination) on a scale of 0 (phoria) to 5 (constant) and spherical equivalent refractive error between −6.0 D and +1.0 D. Children were randomly assigned to either overminus spectacles (−2.50 D over cycloplegic refraction) or observation (non-overminus spectacles if needed, or no spectacles). The primary outcome measure was distance control score for each child assessed by masked examiner at 8 weeks. Outcome testing was conducted with children wearing their study spectacles, or plano spectacles for observation group children who did not need spectacles. The primary analysis compared mean 8-week distance control score between treatment groups using an analysis of covariance model that adjusted for baseline distance control. Treatment side effects were evaluated using questionnaires completed by parents.

Results

At 8 weeks, mean distance control was better in the 27 children treated with overminus spectacles than in the 31 children who were observed without treatment (2.0 vs. 2.8 points, difference = −0.80 points (95% CI = −1.49 to −0.11 points; P = .01 for one-sided test). Side effect profiles regarding headaches, eyestrain, avoidance of near activities, and blur appeared similar between treatment groups.

Figure 1. Mean distance control at 8 weeks.

Figure 2. Distribution of mean distance control at 8 weeks.
Figure 3a. Mean improvement in distance exotropia control over 8 weeks was 1.2 points in the overminus group vs. 0.4 points in the observation group. Bars above 0 indicate worsening of control, and bars below zero indicate improvement in control.

Figure 3b. Fifty-nine percent of children in the overminus group showed at least 1 point improvement in distance exotropia control at 8 weeks, compared to 39% in the observation group.

Figure 4. Baseline exotropia control at distance (left) and near (right). Baseline exotropia control scores appeared similar between the overminus and observation groups at both distance and near. Control score was assigned on a scale of 0 to 5, with 0 representing best control and 5 representing worst control (mean of 3 assessments during exam).

Table 1. Office Control Score

<table>
<thead>
<tr>
<th>Control Score</th>
<th>Control Score Description</th>
</tr>
</thead>
<tbody>
<tr>
<td>5</td>
<td>Constant XT during a 30-sec observation period (before dissociation)</td>
</tr>
<tr>
<td>4</td>
<td>XT &gt; 50% of time during a 30-sec observation period (before dissociation)</td>
</tr>
<tr>
<td>3</td>
<td>XT &lt; 50% of time during a 30-sec observation period (before dissociation)</td>
</tr>
<tr>
<td>2</td>
<td>No XT unless dissociated (10 sec); recovery in &gt; 5 sec</td>
</tr>
<tr>
<td>1</td>
<td>No XT unless dissociated (10 sec); recovery in 1-5 sec</td>
</tr>
<tr>
<td>0</td>
<td>Pure phoria; &lt; 1 sec recovery after 10-sec dissociation</td>
</tr>
</tbody>
</table>

Abbreviation: XT indicates exotropia.

Adapted from Mohney BG, Holmes JM. An office-based scale for assessing in intermittent exotropia. Strabismus 2006; 14:147-150.
Discussion
In a pilot randomized clinical trial, overminus spectacles improved distance control over 8 weeks in children 3 to <7 years old with IXT.

Conclusions
A larger and longer randomized controlled trial is warranted to assess the long-term effectiveness of overminus spectacles in treating IXT, particularly the effect on control after overminus treatment has been discontinued. Current RCT will be described and discussed.

References
Surgery for Intermittent Exotropia: Results from the IXT1 Trial

Sean P Donahue MD PhD

I. RCT: Recess–Resect (R/R) vs. Bilateral Lateral Rectus (BLRc) Recession

A. Basic type intermittent exotropia (XT)
B. Age 3 to < 11 years

II. Inclusion Criteria

A. Intermittent deviation
B. 15-40 PD at distance, at rest 10 at near
C. Detectable stereopsis
D. No previous surgery
E. No amblyopia

III. Follow-up

A. Postop 6 weeks to 8 weeks
B. Marked exam every 6 months
C. Follow-up at 3 years for primary outcome

IV. Primary Outcome

A. Substantial success at any marked examination up to and including 3 years
   1. XT (simultaneous prism and cover test [SPCT]) at distance or near ≥ 10 PD (reoccurrence)
   2. Esotropia (SPCT) at distance or near ≥ 6 PD
   3. Decrease in new stereo (2 octaves)
   4. Once failed, always failed
B. Secondary outcomes
   1. Above outcomes at 5-year visit
   2. Previous exams didn’t cover
   3. Reoperation at any time
   4. Potential for investigator bias

V. Results

A. Baseline demographics / visit completion
B. Suboptimal outcome rate by 3 years
   1. R/R: 37%
   2. BLRc: 46%
   3. 95% CI for difference: 6% to 23%
C. Secondary outcome at 3-year exam visit
   1. BLRc: 28%
   2. R/R: 15%
   3. CI: 3%-27%
D. Reoperation rate by 3 years
   1. BLRc: 10%
   2. R/R: 5%
   3. CI: 2%-13%

VI. Conclusion

Given there does not appear to be a clear advantage to either R/R or BLRc with the first postoperative years, both are reasonable approaches for the initial surgical treatment of basic IXT using surgical doses followed in our study.
Case 1 and Case 2: I Want My Money Back

David L Guyton MD

**CASE 1**

Patient is 63-year-old female. Chief complaint: Blurred vision when looking away from the center through expensive new lenses. “I want my money back!”

**Present Illness**

Moderate myopia and astigmatism since childhood. Recently obtained $600 new glasses with thinner, high-index, plastic lenses, made by a world-famous German optical company.

**Exam**

Power of present glasses in phoropter yields better vision in each eye.

- Refraction RE: −4.50 +0.50 x 58 = 20/20−
- Refraction LE: −5.75 +3.75 x 96 = 20/20+

**Diagnosis**

Off-axis chromatic blur and decreased vision caused by high dispersion of the high-index lenses

**Solution**

New glasses prescribed with normal refractive index lenses (CR39) provided resolution of complaints.

**Discussion**

“Dispersion” is a measure of how much the different colors of the visible spectrum are spread out upon refraction. High-index lens materials generally have high dispersion, with the resulting chromatic aberration, away from the optical axis, decreasing vision. The “Abbe number” is directly proportional to the reciprocal of the dispersion, such that the lower the Abbe number, the worse the dispersion.

<table>
<thead>
<tr>
<th>Lens Material</th>
<th>Refractive Index</th>
<th>Abbe Number</th>
</tr>
</thead>
<tbody>
<tr>
<td>Crown glass</td>
<td>1.52</td>
<td>59: low dispersion</td>
</tr>
<tr>
<td>1.7 glass</td>
<td>1.70</td>
<td>35</td>
</tr>
<tr>
<td>1.8 glass</td>
<td>1.80</td>
<td>35</td>
</tr>
<tr>
<td>1.9 glass</td>
<td>1.89</td>
<td>31</td>
</tr>
<tr>
<td>CR39 plastic</td>
<td>1.50</td>
<td>58: low dispersion</td>
</tr>
<tr>
<td>Polycarbonate</td>
<td>1.59</td>
<td>30: highest dispersion</td>
</tr>
<tr>
<td>Very high index</td>
<td>1.74</td>
<td>33</td>
</tr>
</tbody>
</table>

Crown glass and CR39 plastic lens materials have high Abbe numbers (59 and 58 respectively) and thus the least dispersion (see Table 1). Note that while polycarbonate lens material is practically indestructible, it scratches easily and has an unusually low Abbe number (30), causing the worst dispersion of all the common lens materials. I no longer prescribe polycarbonate lenses for my young patients, but specify CR39 lenses instead.

**CASE 2**

Patients is 86-year-old female. Chief complaint: Double vision after prism was increased in her left spectacle lens. “I want my money back!”

**Present Illness**

Bilateral pseudophakia with divergence insufficiency, corrected 1 year previously with a 2 PD base-out (BO) prism in her left lens only. Double vision worsened, especially on side gazes. In the office, 2 PD of additional BO prism eliminated the double vision, and the prism in the left lens was increased to 4 PD BO to avoid having to change both lenses. She returned 3 weeks later with worsened double vision.

**Exam**

The double vision persisted unchanged when she closed her right eye, but disappeared when she closed her left eye. There was good visual acuity in each eye, with minimal oblique astigmatism in the left eye only.

- Refraction RE: plano = 20/25
- Refraction LE: −0.50 +0.50 x 120 with 4 PD BO = 20/20+

No shift on cover test at distance or near, and moderately good stereoacuity. On questioning, she described the monocular double image in her left eye as a faint ghost image.

On inspection, the new left lens had been dispensed without an antireflection (AR) coating, whereas the fellow right lens did have such a coating.
**Diagnosis**

Monocular ghost image diplopia from multiple internal reflections within a new prism spectacle lens, exacerbated by lack of an AR coating.

**Solution**

New glasses were prescribed, with BO prism split between the two lenses and with AR coatings, giving complete resolution of symptoms.

**Discussion**

With only 3%-6% of the light reflected at each of the 2 non-coated lens surfaces, ghost images from multiple internal reflections are usually so faint (0.09% to 0.36% as bright as the primary image) and so far out of focus that they are not noticed. They are more easily seen, however, with very low-power corrections, and if there is also low-power prism in the spectacle lens, they are more displaced from the primary image and more bothersome, even in straight-ahead viewing.

In most cases, AR coatings are expensive and do not significantly improve vision. (They primarily help cosmetically by reducing external reflections that can hide the wearer’s eyes.) In addition, the lenses can appear dirty as the coating begins to wear off. For these reasons, I rarely prescribe AR coatings.

However, AR coatings can be effective in reducing or eliminating symptoms from ghost images from multiple internal reflections, especially with low-power spectacle lenses containing low-power prism.

**Selected Readings**

**Case 1**


**Case 2**


Case 3 and Case 4: These Glasses Make Me Sick

Michael X Repka MD MBA

CASE 3

I. Case

A. Patient is a 69-year-old man who presented with a several-month history of blurred vision O.D. with double vision.

B. Past ophthalmological history

1. Pseudophakia O.U.
2. Macula-off retinal detachment
   a. Gas tamponade and vitrectomy
   b. Periocular anesthesia; bupivacaine was used

C. Exam

1. Visual acuity, corrected
   a. O.D.: −4.75 +2.50 x 010 = 20/100
   b. O.S.: −4.50 +1.50 180 = 20/20
2. Pupils: mild relative afferent pupillary defect O.D.
3. Extraocular movement (EOM)
   a. Full ductions; comitant versions
   b. XT 8°, RHT 4°; XT' 12°
4. Confrontation visual fields
   a. O.D.: full
   b. O.S.: full

D. Clinical course

1. Prism helped in the office. Seemed to work.
2. 10° BD and BI at 320
3. Took a Fresnel prism home with that prescription. Stopped working in a day or so; ghost image on right returned. Prism increased. Felt good. Lenses constructed. He could not wear the glasses. Converted to Bangerter filter, which he tolerated.

II. Dragged Fovea Diplopia Syndrome (Retinal Misregistration)¹

A. Presentation

1. Unilateral or bilateral; binocular diplopia
2. Small angle, can be in any direction; often diagonal
3. Foveal / macular pathology: Idiopathic epiretinal membrane, postsurgical changes in internal limiting membrane, macular degeneration, other maculopathy
   About half of cases reported by De Pool; the macular disease was unknown prior to the evaluation for diplopia.

B. Diagnosis

1. Suspicion
2. Biomicroscopic evaluation of the fovea of both regions
3. Lights-on and lights-off test¹
4. OCT is the current gold standard for diagnosis.

C. Etiology

1. Epiretinal membrane (ERM)²
   a. In the Ververka et al study 50% of patients with ERM had symptomatic diplopia; 44% of those were felt to be due solely to the ERM. Other causes were strabismus and refractive error induced diplopia.
   b. More than a third of cases were associated with macular hole.
2. Nonexudative macular degeneration
3. Peripheral retinal tear
4. Other

D. Treatment

1. Difficult at best
2. Consider true strabismus and refractive issues
3. Prism generally does not work for foveal displacement; patients adapt.
   Conflict between central fusion, which is trying to use the shifted position, and peripheral fusion using the photoreceptors which are in register. Typically peripheral fusion wins out.
4. “Satin” packing tape or Bangerter filters are often effective; sometimes socially acceptable.

E. Recovery: Impact of ERM surgery is variable.

F. Quality of life: Not formally studied but it has been difficult
CASE 4

I. Case
A. Patient is a 59-year-old man with a several-month history of blurred vision; reading has become impossible.
B. Past ophthalmological history: Parkinson disease for 8 years
C. Exam
1. Visual acuity, corrected
   a. O.D.: −1.50; add +3.00 = 20/30
   b. O.S.: −2.25; add +3.00 = 20/25
2. Pupils: normally reactive
3. EOM
   a. Full ductions; versions comitant
   b. E 2\(^\circ\); X(T)’ 14\(^{D}\)
4. Confrontation visual fields
   a. O.D.: full
   b. O.S.: full
D. Clinical course
1. 5\(^{A}\) base-in O.U. readers prescribed
2. These never worked well; maybe some improvement some of the time

II. Parkinson Disease (PD) and Visual System
A. Progressive neurodegenerative disorder characterized by disruption of dopaminergic pathways involving the basal ganglia
1. Dopamine replacement is the mainstay of treatment.
2. Fluctuation of signs and symptoms is common. This phenomenon is referred to as “wearing off” or “on-off” fluctuations.
B. Ocular symptoms
1. Blepharospasm, apraxia of eyelid opening, dry eye, reduced blink rate, visual hallucinations, decreased visual acuity, color vision and contrast sensitivity, abnormal saccades and pursuit, diplopia, up-gaze limitation, and convergence insufficiency.\(^{3,5}\)
2. Convergence insufficiency is a frequent finding in PD, with no deviation at distance.\(^{5}\)
C. Convergence ability in one study showed substantial improvement after dopamine therapy, but did not normalize.\(^{6}\)
1. Convergence ability is likely to be in nearly constant flux during waking hours as dopamine levels in the central nervous system rise and fall. This would make treatment of near vision very difficult.
2. Severity of convergence insufficiency is unrelated to duration of PD.
D. Treatment
1. Counseling of patient and family
2. Evaluate in on and off states to try to find a prism prescription that might work; often has to be single vision near.

References
Case 5 and Case 6:  
My Child Will Not Wear His Glasses  

Constance E West MD

When pediatric patients won’t wear the glasses the ophthalmologist has prescribed, it can be frustrating for both physician and parents. The problem is often caused by one of the following:

- **Spectacle fit problems**
  - Frames too large overall
  - Temples too long
  - Poor bridge fit
  - Frames too heavy

- **Lens issues**
  - Lenses thicker than needed at dispensing
  - Incorrect prescription
    - Copied forward incorrectly in EHR
    - Incomplete cycloplegia
    - Poor retinoscopic technique or cooperation
    - Error at optician / lab
  - Lens(es) inadvertently swapped or rotated

- **Spectacle maintenance / upkeep problems**
  - Nosepads missing / damaged
  - Frames bent / loose
  - Bite marks on temples
  - Allergic reaction to frame component

- **Parental concerns**
  - Parent(s) doesn’t (don’t) want child to wear glasses.
  - Parent doesn’t reinforce need to wear glasses.
  - Other caregivers not consistent

Sometimes the solution is easy, as when the lenses are made incorrectly. Other times it is an iterative process involving the opticians and the caregivers. Parent, child, and optician may require positive reinforcement from the ophthalmologist.
Case 7 and Case 8: I Have Never Had a Pair of Glasses I Could See With

David G Hunter MD PhD

When adult patients are unhappy with their glasses, the differential diagnosis can generally be broken down into one or more of the following causes, not all of which may at first seem related to having blurry vision:

- Error in refraction
- Optical imperfections in the eye
  - Monocular diplopia
  - Irregular astigmatism
  - Lens or corneal opacities
- Binocular diplopia / heterophorias / incorrect prism correction
- High refractive errors
  - High astigmatism (especially oblique)
  - High hyperopia (field loss from prismatic effects)
  - High myopia (image minification)
- Incorrect bifocal fitting

Working through the differential diagnosis can be time consuming and frustrating for patient and doctor alike. Some key underlying principles and maneuvers can be helpful:

- Look at the glasses. Does the prescription match? Were they made well? Is there prism (intentional or accidental) in either lens? Are the optical centers in the correct place?
- Cover one eye. If vision improves with either eye covered, there may be a binocular vision issue. Perform a sensorimotor evaluation. Again, look for prism in the glasses.
- Look at the retinoscopic reflex. Is it “clean?” Are there opacities, irregularities, or surface anomalies? Do they improve with a blink?
- Check pinhole visual acuity after your refraction. If it improves, likely there is irregular astigmatism.
- Perform a cycloplegic refraction (even in adults) to look for latent hyperopia or too much minus power.
- Look at the bifocal design. Is the patient a poor candidate for progressive lenses? Is the bifocal power consistent with the accommodative amplitude? Are the segment heights okay?

If there are no problems with irregular astigmatism or strabismus, perform a careful refraction with subjective refinement. Place the new refraction in trial frames. Refine the refraction (with ±0.25 D) in a long hallway or with the patient looking out the window. Now sit the patient at the computer and see which add is most comfortable for intermediate work. Finally, have the patient read (paper or smartphone) and see which add is most comfortable for close-up work.

Using this information, it is usually possible to come up with either a satisfactory prescription with bifocal add, or, more commonly in these difficult situations, a separate pair of “driving” glasses (with or without an add) and “computer/reading” glasses (with progressive add).
Case 9 and Case 10:
These Glasses Make Me See Double

Sarah MacKinnon MSc OC(C) COMT

CASE 9

Patient is a 5-year-old girl with residual partially accommodative esotropia. Dad reports that her eyes are crossed, and she complains of occasional diplopia with her “purple glasses” but reports that she has straight eyes more often with her “pink glasses.” After both pairs of glasses are measured and Dad is further questioned, it becomes clear that the lenses of the pink glasses had popped out. Dad placed them back in, but the lenses were accidently flipped. The optical centers were temporally displaced, giving the patient enough base-out prism to help her control the partially accommodative esotropia.

CASE 10

A 67-year-old woman presented with complaints of diplopia following cataract surgery. The presentation will include differential diagnosis of diplopia following cataract surgery, but in this case, diplopia was resolved after careful refraction and correction of astigmatism.
Case 1: Corneal Conundrum

Nandini Gandhi MD

CASE

A 9-month-old female patient with a history of perinatal seizures and G-tube dependence presented to the emergency room with a 7-mm x 7-mm corneal ulcer in the right eye. The appearance of the lesion and lack of corneal sensation were suggestive of herpetic keratitis, and the patient was started on fortified antibiotic drops and oral acyclovir. Corneal cultures were negative for a pathogenic organism, and the epithelial defect improved to near resolution over the course of 2 weeks.

The patient returned the following month with bilateral neurotrophic keratitis in spite of good compliance with oral acyclovir. She was admitted for a 7-day course of IV acyclovir, and her corneal epithelial defects remained unchanged. Corneal cultures, serologies, CSF cultures, and polymerase chain reaction were all negative for herpes simplex virus.

Two weeks later, with persistent, nonhealing epithelial defects, the patient underwent bilateral amniotic membrane transplantation and lateral tarsorrhaphy. She was left with dense residual corneal scarring in both eyes, and in an attempt to clear her visual axis, a penetrating keratoplasty was performed in her right eye. This graft subsequently failed, and the child continues to have dense corneal scarring bilaterally and a variable response to light in each eye.

Congenital corneal anesthesia may be considered in the differential diagnosis for infants and children with unilateral or bilateral nonhealing epithelial defects that do not respond to standard antimicrobial therapy.

Selected Readings

Case 2: Un-nerving
Optic Disc Drusen with Visual Field Loss and Retinal Nerve Fiber Layer Thinning on OCT

Erin O Schotthoefer MD

Optic disc drusen are proteinaceous deposits anterior to the lamina cribosa in the optic nerve that can become calcified over time. When they present in young children, prior to calcification, they can give the appearance of optic disc edema and present a diagnostic dilemma—pseudopapilledema vs. true optic disc edema. They are present in < 4% of the general population and are found equally in males and females. Most patients will have optic disc drusen bilaterally; however, they can present asymmetrically or, less commonly, be unilateral.

Optic disc drusen are mostly an incidental finding on examination in patients who have no symptoms. Sometimes patients can complain of transient visual obscurations. Optic disc drusen can cause visual field changes and have been associated with choroidal neovascular membranes and hemorrhage, as well as anterior ischemic optic neuropathy and vascular occlusions. It can be challenging to discern visual field loss from concomitant glaucoma in patients with baseline visual field abnormalities from optic disc drusen.

On fundus examination, the optic nerve appears “lumpy bumpy.” The drusen can appear as yellow, round deposits visible on the surface of the optic nerve or they can be “buried” drusen. “Buried” drusen are often not calcified and can be harder to distinguish from optic disc edema. Sometimes no further diagnostic testing is necessary; however, several modalities are used for further evaluation. B-scan ultrasonography is very reliable given the highly reflexive nature of optic disc drusen. Optic disc drusen are also highly autofluorescent on fluorescein angiography, and this modality has been compared favorably to B-scan ultrasonography in evaluating children. OCT has also been used to evaluate optic disc drusen, both to try to image the drusen and also to detect thinning of the retinal nerve fiber layer related to the optic disc drusen. Intracranial imaging to rule out an intracranial cause for the appearance of optic disc edema will often miss drusen due to their small size, though CT scan can incidentally pick up the calcifications and sometimes patients will present with drusen seen on CT scan performed for another reason.

The case presented today was followed initially for accommodative esotropia, with the pseudopapilledema first noted on examination 14 years ago. She had buried drusen that gradually became more visible over the years, and when she was able to have OCT and complete a reliable Humphrey visual field, she was followed with both of these modalities. Of note, her visual field changes and OCT retinal nerve fiber thinning is quite dramatic; fortunately, she maintains good vision and these changes have not yet progressed over time.

Selected Readings


Case 3: Extreme Benign Disease
Just Another Failed Vision Screen

Janine Collinge MD

CASE

Case Presentation
- AW 10-year-old otherwise healthy male referred after failed vision screen
- Patient denies any vision problems.
- Teacher states patient can’t see the board.
- VA 20/100 O.U.; depressed color vision right eye
- Anterior segment normal
- Posterior segment: For your evaluation

Discussion

I. Stargardt Disease
   A. Most prevalent macular degeneration in children
      Prevalence: 10-12.5 per 100,000
   B. Clinical yellow-white flecks with macular atrophy
      Retinal pigment epithelial and photoreceptor atrophy causing secondary vision loss
   C. Worse prognosis with younger age of onset
      1. Average VA 20/100 at diagnosis
      2. Progressing to 20/200 to 20/400 over 10 years or less
   D. Generalized retinal dysfunction on electroretinography
   E. Autosomal recessive inheritance: mutation in the ABCA4 gene

II. Vision Screening
   A. Current screening recommendation and education tools available on the AAPOS website.
      1. www.aapos.org/terms/conditions/131
   B. Most vision screen failures are attributed to uncorrected or undercorrected refractive error.
   C. Amblyopia affects approximately 1%-4% of the pediatric population. Early childhood vision screening is aimed at treatment and prevention of amblyopia.
   D. Strabismus can be seen in 1%-5% of children.
   E. Significant refractive error has been reported in up to 30% of children, depending on population.
   F. Visual impairment from ocular disease can be identified in 0.1%-0.9% of vision screening patients.

Selected Readings

Case 4: What Is This White Spot?  
Vernal Keratoconjunctivitis  
S Grace Prakalapakorn MD MPH

I. Background  
A. Age distribution: Predominantly affects the pediatric population  
B. Sex distribution: Prior to puberty, affects more males than females  
C. Duration of disease: weeks to years  
D. Seasonal variation vs. year-round  
E. Severity of disease: Ranges from mild to severe

II. Symptoms  
A. Ocular itching and irritation  
B. Red eye  
C. Tearing  
D. Thick ocular discharge  
E. Photophobia

III. Clinical Findings  
A. Conjunctival hyperemia  
B. Palpebral papillae (especially under the upper eyelid) and limbal follicles (Horner-Trantas dots)  
C. Stringy / ropy secretions  
D. Corneal findings  
   1. Superficial punctate keratitis  
   2. Erosion  
   3. Epithelial defect  
   4. Ulcer  
      a. Grade 1: Transparent ulcer base  
      b. Grade 2: Translucent ulcer base ± opaque white or yellow deposits  
      c. Grade 3: Elevated plaque

IV. Treatments  
A. Topical therapy  
   1. Antihistamines  
   2. Mast cell stabilizers  
   3. Combination / dual-acting agents  
   4. Immunomodulators: cyclosporine, tacrolimus  
   5. Corticosteroids: prednisolone, loteprednol, flurometholone, dexamethasone  
B. Surgical management  
   1. Scraping  
   2. Superficial keratectomy  
   3. Excimer laser

V. Complications  
A. Corneal scarring and vascularization  
B. Microbial keratitis  
C. Amblyopia  
D. Strabismus

CASE  
- Eight-year-old AAM  
- Known history of VKC  
- Presents with active VKC and grade 2 corneal ulcer  
- Disease course  
- Treatment  
- Discussion

Selected Readings  
Case 5: An Extreme Case of Blepharitis

Michelle T Cabrera MD and Sailaja Bondalapati MD

CASE

We report the case of a 4-year-old girl with blepharitis and recurrent chalazia who presented with severe corneal opacifications, multiple hordeola, photophobia, and pain. Because she was very uncooperative with topical therapies and her family was Christian Scientist, she was nonadherent to recommended topical steroid-antibiotic combination treatments. She was lost to follow-up and 1 year later returned with worsening corneal scarring and decreased visual acuity to 20/100 binocularly. She underwent an examination under anesthesia with subtenon triamcinolone acetonide injection. She exhibited almost complete resolution of symptoms and improvement in corneal opacification. Final visual acuity achieved was 20/50 in the right eye and 20/70 in the left eye. Subtenon triamcinolone acetonide injections may be considered in cases of vision-threatening blepharo-keratoconjunctivitis and nonadherence to topical therapies.
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<td>Consultant fee, paid advisory boards, or fees for attending a meeting</td>
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</tr>
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</table>

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Presenter Index

Adams, Gillian 3, 12
Arnoldi, Kyle 13
Bradfield, Yasmin 14
Cabrera, Michelle 53
Colby*, Kathryn 21
Collinge*, Janine 51
Donahue*, Sean 41
Enyedi, Laura 17
Epley, K David 28
Erzurum, Sergul 38
Fray, Katherine 35
Freeman, Richard 16
Frueh*, Beatrice 25
Gandhi, Nandini 49
Guyton*, David 42
Hammersmith*, Kristin 26
Holland*, Edward
Holmes*, Jonathan 1, 4, 7, 10
Hunter*, David 47
Ing, Malcolm 5, 11
Kushner, Burton 15
Lueder, Gregg 32
Mackinnon*, Sarah 48
Nehls, Sarah 22
Orge, Faruk 37
Pineles, Stacy 2, 8
Prakalapakorn*, S 52
Pritchard, Cindy 18
Ram, Radha 34
Repka*, Michael 44
Robbins*, Shira 31
Schotthoefer, Erin 50
Strube*, Yi Ning 6, 9
Suh*, Donny Won 36
Tychsen, Lawrence 30
VanderVeen*, Deborah 33
West*, Constance 46
Williams, Pamela 19

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## Saturday, Nov 11

<table>
<thead>
<tr>
<th>Course Code</th>
<th>Course Title</th>
<th>Time</th>
<th>Room</th>
</tr>
</thead>
<tbody>
<tr>
<td>SYM61</td>
<td>MOC Exam Review: Core Ophthalmic Knowledge</td>
<td>10:00 AM to 12:00 PM</td>
<td>275-277</td>
</tr>
<tr>
<td>SYM60</td>
<td>Diagnose This Live Saturday</td>
<td>2:00 PM to 4:00 PM</td>
<td>275-277</td>
</tr>
</tbody>
</table>

## Sunday, Nov 12

<table>
<thead>
<tr>
<th>Course Code</th>
<th>Course Title</th>
<th>Time</th>
<th>Room</th>
</tr>
</thead>
<tbody>
<tr>
<td>Course 203</td>
<td>Evidence-Based Guidelines in the Management of Glaucoma</td>
<td>10:15 AM to 12:30 PM</td>
<td>388</td>
</tr>
<tr>
<td>Course 205</td>
<td>Decoding the Uveitis Workup: Why, When, and What to Order</td>
<td>10:15 AM to 12:30 PM</td>
<td>393</td>
</tr>
<tr>
<td>Course 206</td>
<td>Update on Diagnosis and Management of ROP: Pearls for ROP Screening, Introduction of Telemedicine, and Use of Anti-VEGF Medications in Practice</td>
<td>10:15 AM to 12:30 PM</td>
<td>389</td>
</tr>
<tr>
<td>Course 236</td>
<td>Examining the Optic Nerve and Evaluating the Visual Field: The 5 Rs</td>
<td>2:00 PM to 4:15 PM</td>
<td>388</td>
</tr>
<tr>
<td>Course 252</td>
<td>The Art and Science of Glaucoma Drainage Devices: How to Optimize Your Surgical Results</td>
<td>2:00 PM to 4:15 PM</td>
<td>389</td>
</tr>
<tr>
<td>Course 268</td>
<td>Optical Coherence Tomography: Basics and Beyond</td>
<td>3:15 PM to 5:30 PM</td>
<td>393</td>
</tr>
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</table>

## Monday, Nov 13

<table>
<thead>
<tr>
<th>Course Code</th>
<th>Course Title</th>
<th>Time</th>
<th>Location</th>
</tr>
</thead>
<tbody>
<tr>
<td>SPO2</td>
<td>Spotlight on Cataract Complications</td>
<td>8:15 AM to 12:15 PM</td>
<td>The Great Hall</td>
</tr>
<tr>
<td>Course 413</td>
<td>Herpes Simplex Keratitis: When Herpes Isn’t a Dendrite, and Vice Versa</td>
<td>9:00 AM to 11:15 AM</td>
<td>393</td>
</tr>
<tr>
<td>Course 414</td>
<td>Case Studies on the Use of OCT for Diagnosis of Unknown Causes of Visual Loss: Is it the Retina, Anterior Visual Pathway, or Misinterpretation of Normal?</td>
<td>9:00 AM to 11:15 AM</td>
<td>388</td>
</tr>
<tr>
<td>Course 445</td>
<td>Current Topics in Cornea/External Disease: Highlights of the Basic and Clinical Science Course 8</td>
<td>10:15 AM to 12:30 PM</td>
<td>389</td>
</tr>
<tr>
<td>Course 494</td>
<td>Principles of Pediatric Ocular Trauma Management</td>
<td>2:00 PM to 4:15 PM</td>
<td>388</td>
</tr>
<tr>
<td>Course 507</td>
<td>A Step-by-Step Primer to Starting LASIK in 2017</td>
<td>3:15 PM to 5:30 PM</td>
<td>389</td>
</tr>
<tr>
<td>Course 518</td>
<td>State of the Art of Intracameral Antibiotics, NSAIDs, Corticosteroids, and Drop-Free Cataract Surgery</td>
<td>3:15 PM to 5:30 PM</td>
<td>393</td>
</tr>
</tbody>
</table>

## Tuesday, Nov 14

<table>
<thead>
<tr>
<th>Course Code</th>
<th>Course Title</th>
<th>Time</th>
<th>Room</th>
</tr>
</thead>
<tbody>
<tr>
<td>Course 618</td>
<td>Surgical Management of Pediatric Glaucoma</td>
<td>9:00 AM to 11:15 AM</td>
<td>389</td>
</tr>
<tr>
<td>Course 673</td>
<td>Minimally Invasive Glaucoma Surgery: Surgical Techniques and Clinical Pearls</td>
<td>12:45 PM to 3:00 PM</td>
<td>393</td>
</tr>
<tr>
<td>Course 676</td>
<td>Update on Treatments for Diabetic Retinopathy: Clinically Relevant Results From the Diabetic Retinopathy Clinical Research Network</td>
<td>12:45 PM to 3:00 PM</td>
<td>388</td>
</tr>
</tbody>
</table>

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