Pediatric Ophthalmology 2011:
A Child’s View from Here...and There
Pediatric Ophthalmology 2011
A Child’s View From Here . . . and There

Program Directors
George S Ellis Jr MD, David B Granet MD, Ken K Nischal MBBS

In conjunction with the Section on Ophthalmology of the American Academy of Pediatrics

Orange County Convention Center
Orlando, Florida
Saturday, October 22, 2011

Presented by:
The American Academy of Ophthalmology

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Dear Colleague:

On behalf of the American Academy of Ophthalmology and the Section on Ophthalmology of the American Academy of Pediatrics, it is our pleasure to welcome you to Orlando and to Pediatric Ophthalmology 2011: A Child’s View From Here . . . and There.

Expertise does not reside in just one region of the world. Consequently, we have drawn not only on colleagues in disciplines throughout ophthalmology but also on specialists from around the globe. We are certain this group will be creative in their presentations and that the discussion will be lively! In fact, the format is interactive, with time for significant audience participation.

The outstanding faculty of internationally regarded experts will provide up-to-date and pertinent information on a variety of pediatric ophthalmology and strabismus topics. We are excited and honored to have brought in Helen Mintz-Hittner MD to discuss innovative new treatments for ROP—a timely, even controversial, topic.

In addition, the Section on Ophthalmology of the American Academy of Pediatrics (AAP) is proud to present experts on the effects of anesthesia and radiation on children. When is enough enough? Further, the discussion on health care change by the head of the AAP Washington office will be important to us all.

As cochairs of the Pediatric Ophthalmology Subspecialty Day Program Planning Group, we know this Pediatric Subspecialty Day will provide practical information that clinicians and subspecialists can use in their practices.

Again, we welcome you Pediatric Ophthalmology 2011: A Child’s View From Here . . . and There; we hope you find it educational and enjoyable.

Sincerely,

George S Ellis Jr MD  
Program Director

David B Granet MD  
Program Director

Kanwal K Nischal MBBS  
Program Director

PS: In an effort to put together innovative and interesting Subspecialty Day meetings in the future, we request that you assist us by completing the evaluation form. We carefully review all comments to better understand your needs, so please indicate the strengths and shortcomings of today’s program.
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CME Credit

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

2011 Pediatric Ophthalmology Subspecialty Day Meeting Learning Objectives
Upon completion of this activity, participants should be able to:
• Evaluate new disease entities, practices and treatment that may change practice
• Plan the surgical treatment of complex strabismus problems
• Assess the relative merits and disadvantages of some therapies in a balanced way
• Discuss ocular manifestations of various systemic diseases and explore new developments in the treatment
• Present various pediatric neuro-ophthalmology cases and discuss the latest diagnostic and treatment options

2011 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.

2011 Pediatric Ophthalmology Subspecialty Day CME Credit
The American Academy of Ophthalmology is accredited by the Accreditation Council for Continuing Medical Education 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.

Scientific Integrity and Disclosure of Relevant Financial Interest
The American Academy of Ophthalmology is committed to ensuring that all continuing medical education (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.

Attendance Verification for CME Reporting
Before processing your requests for CME credit, the Academy must verify your attendance at Subspecialty Day and/or the 2011 Annual Meeting. In order to be verified for CME or auditing purposes, you must either:
• Register in advance, receive materials in the mail and turn in the Final Program and/or Subspecialty Day Syllabus exchange voucher(s) onsite;
• Register in advance and pick up your badge onsite if materials did not arrive before you traveled to the meeting;
• Register onsite; or
• Use your ExpoCard at the meeting.

CME Credit Reporting
Level 2, Lobby B; Academy Resource Center, Hall A4, Booth 1359
Attendees whose attendance has been verified (see above) at the 2011 Annual Meeting can claim their CME credit online during the meeting. Registrants will receive an e-mail during the meeting with the link and instructions on how to claim credit.

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

Academy Members: The CME credit reporting receipt is not a CME transcript. CME transcripts that include 2011 Annual Meeting credits entered onsite will be available to Academy members on the Academy’s website beginning Nov. 16, 2011.

NOTE: CME credits must be reported by Jan. 18, 2012. After the 2011 Annual Meeting, credits can be claimed at www.aao.org.

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 the 2011 Annual Meeting and Subspecialty Day for those who need it for reimbursement or hospital privileges, or for nonmembers who need it to report CME credit:
• CME credit reporting/proof-of-attendance letters
• Onsite Registration Form
• Instruction Course Verification Form

Visit the Academy’s website for detailed CME reporting information.
Faculty

**Steven M Archer MD**  
Ann Arbor, MI  
Associate Professor of Ophthalmology  
University of Michigan

**Mark Del Monte JD**  
Washington, DC  
Director, Department of Federal Affairs  
American Academy of Pediatrics

**George S Ellis Jr MD**  
New Orleans, LA

**Michael J Callahan MD**  
Boston, MA

**Monte A Del Monte MD**  
Ann Arbor, MI  
Skillman Professor of Pediatric Ophthalmology  
University of Michigan  
Director of Pediatric Ophthalmology and Adult Strabismus  
W K Kellogg Eye Center

**David K Coats MD**  
Houston, TX  
Professor of Ophthalmology and Pediatrics  
Baylor College of Medicine  
Department Chief  
Pediatric Ophthalmology  
Texas Children’s Hospital

**Sean P Donahue MD PhD**  
Nashville, TN  
Professor of Ophthalmology, Neurology, and Pediatrics  
Vanderbilt University Medical Center  
Chief of Pediatric Ophthalmology  
Vanderbilt Children’s Hospital

**Brian J Forbes MD PhD**  
Wallingford, PA  
Assistant Professor of Ophthalmology  
Children’s Hospital of Philadelphia  
University of Pennsylvania School of Medicine

**Sharon F Freedman MD**  
Durham, NC  
Professor Of Ophthalmology and Pediatrics  
Duke Eye Center
David B Granet MD
La Jolla, CA
Anne Ratner Professor of Ophthalmology and Pediatrics
University of California, San Diego
Director of Pediatric Ophthalmology and Strabismus
Ratner & Shiley Eye Centers
University of California, San Diego

Mark E Jacobson MD
Santa Rosa, CA

Sylvia R Kodsi MD
New York, NY
Co-chief, Pediatric Ophthalmology and Strabismus
North Shore - Long Island Jewish Health Systems
Associate Professor
Hofstra North Shore-LIJ School of Medicine

Richard W Hertle MD
Akron, OH
Chief of Ophthalmology
Children’s Vision Center
Akron Children’s Hospital
Professor of Surgery (Ophthalmology)
Northeastern Ohio Universities College of Medicine

Constance S Houck MD
Boston, MA

Mark E Jacobson MD
La Jolla, CA
Director of Pediatric Ophthalmology and Strabismus
Ratner & Shiley Eye Centers
University of California, San Diego

Ramesh Kekunnaya MD FRCS
Hyderabad, AP, India
Associate Professor, Pediatric Ophthalmology
L V Prasad Eye Institute

Burton J Kushner MD
Madison, WI
John W and Helen Doolittle Professor of Ophthalmology
Department of Ophthalmology and Visual Sciences
University of Wisconsin, Madison

Don O Kikkawa MD
La Jolla, CA
Professor Of Ophthalmology
Chief, Division of Ophthalmic Plastic and Reconstructive Surgery
Department of Ophthalmology
University of California, San Diego

G Robert LaRoche MD
Halifax, NS, Canada
Professor of Ophthalmology
Dalhousie University
Thomas C Lee MD  
Los Angeles, CA  
Associate Professor of Ophthalmology  
University of Southern California  
Director, Retina Institute  
Childrens Hospital Los Angeles

Kanwal K Nischal MBBS  
Pittsburgh, PA  
Director, Children’s Hospital of Pittsburgh UPMC, Pittsburgh

Helen A Mintz-Hittner MD FACS  
Houston, TX  
Professor of Ophthalmology and Visual Sciences  
University of Texas Health Science Center at Houston Medical School

David A Plager MD  
Indianapolis, IN  
Professor of Ophthalmology  
Indiana University Medical Center  
Director of Pediatric Ophthalmology and Strabismus  
Indiana University Medical Center

Daniel S Mojon MD  
St Gallen, Switzerland

Kanwal K Nischal MBBS  
Pittsburgh, PA  
Director, Children’s Hospital of Pittsburgh UPMC, Pittsburgh

Paolo Nucci MD  
Milano, Italy  
Professor of Ophthalmology  
University of Milan  
Director  
San Giuseppe Eye Clinic

Jean E Ramsey MD MPH  
Boston, MA  
Associate Professor of Ophthalmology and Pediatrics  
Boston University School of Medicine Council, Vice-chair  
American Academy of Ophthalmology

Scott E Olitsky MD  
Kansas City, MO  
Chief of Ophthalmology  
Children’s Mercy Hospitals and Clinics  
Professor of Ophthalmology  
University of Missouri, Kansas City, School of Medicine

Shira L Robbins MD  
La Jolla, CA
Daniel J Salchow MD
New Haven, CT
Assistant Professor
Department of Ophthalmology & Visual Science and Department of Pediatrics
Yale University School of Medicine

Nicoline Schalij-Delfos MD
Voorschoten, Netherlands

Eduardo D Silva MD
Figueira Da Foz, Portugal
Assistant Professor of Ophthalmology
University of Coimbra, Portugal
MD, PhD
IBILI, School of Medicine, Coimbra, Portugal

Jane C Sowden PhD
London, United Kingdom

Elias I Traboulsi MD
Cleveland, OH
Professor of Ophthalmology
Cleveland Clinic Lerner College of Medicine

Abhay Raghukant Vasavada
MBBS FRCS
Ahmedabad, Gujarat, India
Professor of Ophthalmology
Raghu Deep Eye Clinic
Director
Iladevi Cataract & IOL Research Centre

M Edward Wilson Jr MD
Charleston, SC
Professor and Chairman of Ophthalmology
Storm Eye Institute
Medical University of South Carolina

Terri L Young MD
Durham, NC
Professor of Ophthalmology, Pediatrics, and Medicine
Duke University
Professor of Neuroscience
Duke - National University of Singapore

Gerald W Zaidman MD FACS
Valhalla, NY
Director of Ophthalmology
Westchester Medical Center
Professor of Ophthalmology
New York Medical College
Pediatric Ophthalmology 2011: A Child’s View From Here . . . and There

SATURDAY, OCTOBER 22, 2011

6:30 AM  REGISTRATION/MATERIAL PICKUP/CONTINENTAL BREAKFAST

8:00 AM  Welcome and Opening Remarks  George S Ellis Jr MD*

Section I:  Will This Change My Practice?
Moderator: David B Granet MD*

8:05 AM  Video: Minimally Invasive Strabismus Surgery  Daniel S Mojon MD  1

8:09 AM  Propranolol for Capillary Hemangiomas: Can I Use This All the Time?  David A Plager MD*  3

8:17 AM  Dynamic Retinoscopy  Burton J Kushner MD  5

8:25 AM  Anesthetic Neurotoxicity: Are We Poisoning Children’s Brains?  Constance S Houck MD  6

8:37 AM  Panel Discussion

Section II:  Oh, No!
Moderator: Kanwal K Nischal MBBS*

8:48 AM  Video: Rupture of Muscle During Strabismus Surgery  David B Granet MD*  8

8:51 AM  When to Order What!  Sylvia R Kodsi MD  9

8:59 AM  The Good, the Bad, and the Ugly: Nonaccidental Injury  Brian J Forbes MD PhD  11

9:07 AM  “Skew You”  Sean P Donahue MD PhD*  13

9:15 AM  Panel Discussion

Section III:  Oh, No! Part 2: What Do We Do Now?
Moderator: George S Ellis Jr MD*

9:30 AM  AAP Federal Affairs Update  Mark Del Monte JD  15

9:42 AM  Questions and Answers

9:47 AM  REFRESHMENT BREAK and ANNUAL MEETING EXHIBITS

Section IV:  New Techniques for Children
Moderator: David B Granet MD*

10:17 AM  Video: Goniotomy in an Aniridic Patient  Kanwal K Nischal MBBS*  16


10:28 AM  Endoscopic Vitreoretinal Surgery  Thomas C Lee MD  18


* Indicates that the presenter has financial interest.
No asterisk indicates that the presenter has no financial interest.
10:44 AM Fundus Autofluorescence in Pediatric Ophthalmology  
Elias I Traboulsi MD*  

10:52 AM Panel Discussion

11:07 AM Surgery by Surgeons Update  
Jean E Ramsey MD MPH  

Section V: Pediatric Ophthalmology Conundrums

Moderator: George S Ellis Jr MD*

11:12 AM COMETs CLAMP ATOM: The Myopia Studies—Can We Affect Refractive Outcome?  
Terri L Young MD*  

11:20 AM Iris Clip Lenses for Aphakia  
Nicoline Schalij-Delfos MD  

11:28 AM Should We Be Using Mitomycin So Readily in Pediatric Glaucoma Surgery?  
Sharon F Freedman MD*  

11:36 AM Do Adjustable Sutures Enhance Outcomes?  
Paolo Nucci MD*  

11:44 AM Congenital Corneal Opacifications: Time for a Re-think?  
Kanwal K Nischal MBBS*  

11:52 AM Panel Discussion

Keynote Lecture

Moderator: Kanwal K Nischal MBBS*

12:07 PM Avastin for ROP  
Helen A Mintz-Hittner MD FACS  

12:27 PM Questions and Answers

12:32 PM LUNCH and ANNUAL MEETING EXHIBITS

Section VI: Challenging Dogma (and Other Good Questions)

Moderator: David B Granet MD*

1:47 PM Video: Cataract Surgery for Marfan Syndrome  
Daniel J Salchow MD  

1:50 PM Why Don’t We Operate to Eliminate Lower-Power Hyperopic Spectacles in Accommodative Esotropia?  
Scott E Olitsky MD  

1:58 PM Is the Pediatric Eye Disease Investigator Group Wrong About . . . ?  
Steven M Archer MD  

2:06 PM Eye Drops for Nystagmus? Really?  
Richard W Hertle MD  

2:14 PM Do Study Design and Methodology Affect Pediatric Cataract Outcomes?  
Ramesh Kekunnaya MD FRCS  

2:22 PM Panel Discussion

Section VII: Challenging Dogma (and Other Good Questions)—Part II

Moderator: Kanwal K Nischal MBBS*

2:37 PM The Ciliopathies: What Are They?  
Eduardo D Silva MD*  

2:45 PM Retinal Repair by Transplantation of Photoreceptor Precursors  
Jane C Sowden PhD  

2:57 PM Radiation Exposure to Children From Medical Imaging: Is There a Problem?  
Michael J Callahan MD  

3:09 PM Questions and Answers

3:21 PM REFRESHMENT BREAK and ANNUAL MEETING EXHIBITS

* Indicates that the presenter has financial interest.
No asterisk indicates that the presenter has no financial interest.
Section VIII: Have You Thought About . . .
Moderator: David B Granet MD*

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<tr>
<td>3:51 PM</td>
<td>Video: Fishtail Sign in Posterior Lenticonus</td>
<td>Abhay Raghukant Vasavada MBBS FRCS*</td>
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<td>3:54 PM</td>
<td>Current Management Strategies for Blepharokeratoconjunctivitis</td>
<td>Mark E Jacobson MD</td>
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<td>4:02 PM</td>
<td>Evaluation of the Non-seeing Infant</td>
<td>Shira L Robbins MD*</td>
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<td>4:10 PM</td>
<td>Update on Oculoplastics</td>
<td>Don O Kikkawa MD</td>
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<td>4:18 PM</td>
<td>Managing Psychosocial Effects of Strabismus</td>
<td>Daniel S Mojon MD</td>
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<td>4:26 PM</td>
<td>Panel Discussion</td>
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Section IX: How Do You Handle Strabismus When . . .
Moderator: George S Ellis Jr MD*

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<td>4:41 PM</td>
<td>The Patient Has Thyroid Ophthalmopathy</td>
<td>David B Granet MD*</td>
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<td>4:49 PM</td>
<td>There Is a CN VI Palsy</td>
<td>G Robert LaRoche MD</td>
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<td>4:57 PM</td>
<td>The Patient Has Adult-Onset CN III Palsy</td>
<td>David K Coats MD</td>
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<td>5:05 PM</td>
<td>You Are Faced With Partially Accommodative Esotropia ± High Accommodative Convergence-to-Accommodation Ratio</td>
<td>Monte A Del Monte MD</td>
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<td>5:13 PM</td>
<td>Panel Discussion</td>
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<td>5:28 PM</td>
<td>Closing Remarks</td>
<td>George S Ellis Jr MD*</td>
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Video: Minimally Invasive Strabismus Surgery

Daniel S Mojon MD

Disclosure: Daniel Mojon has no relevant financial relationships.

I. Frequently Used Conjunctival Access Techniques for Strabismus Surgery

A. Limbal approach
1. Permits full visualization of the operated muscle
2. Patients have considerable postoperative discomfort.
3. Interpalpebral conjunctiva is red for weeks.
4. Prone for corneal dellen formation and Tenon prolapse
5. Large anatomical disruption between the muscle and perimuscular tissue

B. Fornix approach
1. Opening remains covered by the lids after surgery.
2. Minimal postoperative discomfort
3. Difficult in young patients (prominent Tenon tissue), old patients (inelastic conjunctiva), and for repeat surgery (scarring)
4. Moderate anatomical disruption between the muscle and perimuscular tissue
5. Need for an assistant surgeon to displace the opening over operated muscle

C. Minimally invasive strabismus surgery (MISS)
1. Permits performance of most types of strabismus surgeries (rectus muscle recessions, resections, plications, reoperations, retroequatorial myopexias, transpositions, and oblique muscle recessions and plications)
2. Minimal postoperative discomfort
3. Minimal anatomical disruption between muscle and perimuscular tissue
4. No need for assistant surgeon for most techniques
5. However, adjustable sutures are difficult with MISS.
6. Increased risk for scleral perforation for inexperienced surgeons
7. Rather long learning curve

II. MISS in More Detail

A. Best performed with operating microscope
B. Instead of one large opening, there are several keyhole cuts where main surgical steps (usually suturing) are performed.
C. For some surgical procedures, you need to create tunnels between the cuts.
D. Openings placed far away from limbus
E. Junction of cuts allows one to convert to usual, large limbal opening.
F. Perimuscular tissue dissection is reduced to absolute minimum, which still allows one to displace or anchor muscles.
G. Openings are all covered postoperatively by eyelids, minimizing visibility of surgery and patient discomfort.
H. Reduction of frequency of corneal complications, for example dellen formation
I. Minimal tissue disruption facilitates reoperations.
J. Transconjunctival suturing (TRASU) and marginal dissection (MADI) allow further reduction of tissue disruption and operating time.

III. Tips for Starting With MISS

A. Surgeons switching from magnifying glasses to operating microscope should perform several procedures with own technique with microscope before starting with MISS.
B. Start with primary horizontal rectus muscle displacements of 4 mm or less.
C. Use a corneal traction suture.
D. Use plications instead of resections (more demanding through small cuts); dose-response relationship will not change.
E. Ideal patient age between 14 and 40 years; abundant Tenon tissue makes surgery difficult in very young patients, reduced elasticity of the conjunctiva increases risk of a conjunctival tear in very old patients.
F. Before starting with MISS visit an already skilled surgeon; the author always welcomes colleagues in his operating theater.

IV. Instruments for MISS

A. Use small instruments in order to minimize risk of conjunctival tearing.
B. Clamp a serrefine to the eyelid speculum to avoid corneal rubbing of traction suture.
C. Use spatulas of different sizes to visualize tissue through the cuts.
D. Use a bipolar, coaxial diathermy tip.
E. Use only blunt scissors to minimize risks.
F. Use a blunt cannula to safely displace needles through tunnels.

References

Propranolol for Capillary Hemangiomas: Can I Use This All the Time?

David A Plager MD

Introduction

The serendipitous finding that propranolol has a salutary effect on the growth of infantile hemangiomas has led to an explosion of interest in this drug for treatment of this common tumor in infants. Several articles in the refereed medical literature describing its use have appeared in the brief time since that initial publication. Although it appears to be revolutionizing the treatment paradigm for such vision-threatening adnexal tumors, the indications, dosage, optimal treatment duration, and relative risks of treatment remain controversial or at least unknown.

Background

Prior to the discovery of the usefulness of propranolol for treating infantile hemangioma, the most common treatments available included systemic or intralesional injection of steroids, excisional surgery, and in especially difficult cases, more toxic drugs such as vincristine, cyclophosphamide, or alpha interferon. The side effects of those medications are common, well known, and significant. In contrast, the long-standing safety profile and well-tolerated nature of propranolol when used for other (cardiac) indications has led to its enthusiastic embrace as a potential cornerstone of treatment for hemangioma.

Pharmacology

Propranolol is a nonselective beta-blocker with effects on many target tissues, including the heart, lungs, and in gluconeogenesis. Its effects on hemangioma growth are thought to result from a combination of 3 mechanisms: Blockade of beta-2 receptors in capillary endothelial cells leads to vasoconstriction. Blockage of beta-1 receptors is thought to enhance apoptosis. The third mechanism is thought to be the reduction in vascular endothelial growth factor (VEGF) levels caused by beta-blockers leading to decreased angiogenesis.

Its current principle uses in children include treatment of cardiac arrhythmias, some congenital heart defects, and for migraine prophylaxis.

Side Effects of Propranolol

Propranolol has a long and well-documented safety profile as noted by Love and Sikka:4 “With 40 years of extensive clinical experience, not one documented case of death or serious cardiovascular morbidity as a direct result of a beta-blocker exposure is to be found in an English language review for children under 6 years of age.” However, there are many potential side effects.

- Most significant side effects: hypotension, bradycardia, hypoglycemia
- Miscellaneous side effects: bronchospasm, sleep disturbance, GI disturbance (diarrhea), rash

Hypoglycemia is probably the clinically most important side effect. The vast majority of case reports of symptomatic hypoglycemia have involved infants whose oral intake has been reduced as a result of illness or intentional or prolonged fast. However, there have been a small handful of reported symptomatic events requiring emergency room evaluation in otherwise healthy infants with reportedly normal oral intake. Symptoms of hypoglycemia with which the child’s caretakers must be made aware include early signs of shakiness, sweating, and anxiety, which can lead to lethargy, lack of responsiveness, and seizure. These symptoms respond rapidly to normalization of blood sugar level.

Summary

For many physicians, including this author, propranolol has become the treatment of choice for vision-threatening infantile hemangiomas that require systemic treatment. However, other treatment options including intralesional injection of steroid and surgical resection may be preferred for an individual patient. Clinical examples are shown of small astigmatism-inducing eyelid hemangiomas that will respond to 1 or 2 injections of steroid. This treatment may be preferable to subjecting the same infant to multiple months of a 3 times/day systemic medication.

Similarly, some well-circumscribed eyelid tumors with normal overlying skin will have an immediate, dramatic, and permanent response to surgical resection. For carefully selected cases, this outpatient surgical procedure may be preferable to a protracted course of systemic therapy.

Prospective trials of propranolol treatment are under way, but in the meantime, clinicians can be comfortable that propranolol has been shown to be very safe and effective in many infants with vision-threatening hemangiomas. Careful attention to potential side effects, especially hypoglycemia, and comanagement with physicians experienced in the care of young infants will optimize the safety for these young patients.

Indiana University Propranolol Treatment Algorithm

I. Premedication

A. Physical exam with baseline vital signs

B. Is the hemangioma segmental and over 5 cm?

1. If yes, MRI/MRA of head and neck and echo must be done to rule out PHACE syndrome and any contraindication for propranolol.

2. PHACE screening done

3. No contraindications for propranolol on MRI/MRA or echo

C. Baseline EKG

Figure 1.
II. Medication Administration
   A. Propranolol preparation: 20 mg/5 cc or 40 mg/5 cc
   B. Dosing schedule
      1. 0.5 mg/kg/d divided into t.i.d. x 2-3 days
      2. 1.0 mg/kg/d divided into t.i.d. x 2-3 days
      3. 1.5 mg/kg/d divided into t.i.d. x 2-3 days
      4. 2.0 mg/kg/d divided into t.i.d. x 2-3 days (final dose)

III. Monitoring
   Blood pressure/heart rate taken 24 hours after any dosage change including the initial administration (see example below). If abnormal then decrease dose and see cardiology.
   A. Day 1: Start medication.
   B. Day 2: Check blood pressure and heart rate.
   C. Day 4: Check blood pressure and heart rate.

Reference and Selected Readings
Dynamic Retinoscopy

Burton J Kushner MD

I. Technique

A. Dry retinoscopy as patient shifts fixation to target just below retinoscope light

B. End point
   1. Normal: Brisk change to neutrality
   2. Abnormal
      a. Sluggish change to neutrality
      b. Persistent with motion

II. Utility

A. Accommodative insufficiency
   1. Neurologic impairment
   2. Down syndrome

B. Determining whether to correct moderate hyperopia if orthophoric in preverbal children
   1. Orthophoric but normal and full accommodation: No glasses
   2. Orthophoric but not fully accommodating: Prescribe glasses
      a. Will become ET without glasses
      b. Or will be bilateral refractive amblyope

References

Anesthetic Neurotoxicity: Are We Poisoning Children’s Brains?

Constance S Houck MD

Introduction

Every year millions of preterm and newborn infants undergo general anesthesia for a variety of surgical procedures and imaging studies. The landmark studies of Anand and Hickey demonstrated more than 20 years ago that general anesthesia in infants ameliorates the surgical stress response and can lead to decreased morbidity and mortality perioperatively. However, recent research in immature animal models has revealed general anesthetic-induced neurotoxicity and thus has raised questions about the long-term safety of general anesthesia in human babies. All of this research has led to a concern that general anesthetics given to young, vulnerable children may be deleterious to their neurologic growth and development.

Background

Anesthetic agents that function as N-methyl-D-aspartate (NMDA) antagonists (eg, ketamine and nitrous oxide) and anesthetic agents that function as gamma amino butyric acid (GABA) agonists (eg, volatile anesthetics, midazolam, thiopental, and propofol) have been shown to cause neuroapoptosis in immature mice, rats, guinea pigs, and rhesus monkeys. It is important to recognize that neuroapoptosis is part of the normal pruning of redundant neurons during mammalian development and is a natural process as the brain differentiates into specific functions. However, the neuroapoptosis seen in the animal experiments was excessive and led to developmental impairments when the animals were allowed to mature. In all of these animal species, there is a specific period of vulnerability to neuroapoptosis that corresponds to the animal’s period of rapid brain growth. It is unclear when this vulnerable time would be for human infants, but neurodevelopmental studies would suggest that the period of rapid human brain growth occurs from 28 weeks postconception to 24 months of age.

Most of the animal experiments to date have involved relatively large doses of anesthetic agents per body weight (although the minimum doses needed to keep the animals anesthetized) and have exposed the animals for at least 4 hours. It is unknown whether 4 hours of anesthesia in an animal with a natural life span of 2 years would be comparable to 4 hours of anesthesia in a human with a life span of greater than 80 years. It is also impossible to replicate in animal experiments the careful monitoring that human infants receive during anesthesia, leading some researchers to speculate that at least some of the neurotoxicity seen in the animals may be due to poor anesthetic conditions.

A recent series of epidemiologic studies has linked exposure to anesthesia at a young age in humans with learning disabilities. Robert Wilder and his colleagues in Minnesota examined a cohort of over 5000 children and found an association between 2 or more anesthetics before the age of 4 and the cumulative development of learning deficits by age 18 (see Figure 1). Similar findings were seen in a study examining Medicare records in New York state.

However, a study of monozygotic twins from the Netherlands revealed that the intellectual attainments were similar between the anesthesia-exposed and nonexposed twin, leading these researchers to conclude that exposure to anesthesia was not a cause of learning deficits. In most association studies it is impossible to separate the confounders, ie, the effects of surgery and the morbidity that is associated with the need for surgery, from the effects of the exposure to anesthesia. A very recent study compared 2689 Danish adolescents (15-16 year olds) who had undergone inguinal hernia repair in infancy with a matched cohort of more than 15,000 adolescents who had not had surgery in infancy and found no difference in academic performance when adjusting for known confounders, including gender, birth weight, congenital anomalies, and academic achievement of both parents.

In response to these emerging concerns, in 2010 the FDA and the International Anesthesia Research Society (IARS) entered into a public-private partnership initially called SAFEKIDS (Safety of Key Inhaled and Intravenous Drugs in Pediatrics) to provide research support to investigators in this area. This partnership has subsequently expanded and evolved and is now called SmartTots (Strategies for Mitigating Anesthesia-Related Neuro-Toxicity in Tots; see www.smarttots.org). With funding from the FDA and other government agencies, several ongoing international and national studies are currently enrolling patients, including the PANDA and GAS studies. The PANDA study is assembling a cohort of U.S. children who underwent inguinal herniorrhaphy before age 3 and matching them with an unexposed sibling for extensive neurologic and developmental testing. The GAS trial is a multinational, randomized controlled trial of preterm and term infants undergoing inguinal herniorrhaphy utilizing either a spinal or general anesthetic and comparing the neurodevelopmental outcomes of each cohort at ages 2 and 5.

Conclusions

Although there are several recent epidemiologic studies that suggest an association between anesthesia exposure and developmental issues in humans, no study to date has been able to demonstrate a causal link. Because of the many concerning animal studies, however, the neurobehavioral effects of sedative and anesthetic agents are being extensively studied at the moment and this is a high priority for the FDA and the anesthesia community. Until further data are available, it may be prudent to consider delaying purely elective surgeries until children are older than 2.

References and Selected Readings

Reviews


Animal Studies


Human Studies


Video: Rupture of Muscle During Strabismus Surgery

David B Granet MD
When to Order What!
The Child With Nystagmus

Sylvia R Kodsi MD

I. Nystagmus in Childhood: Differentiate by Age of Onset
   A. Congenital: Before 3 months of age
   B. Acquired: After 3 months of age

II. Congenital Nystagmus Subtypes: Differentiate by Eye Exam and Electoretinogram (ERG)
   A. Motor nystagmus: Good vision
   B. Sensory nystagmus: Poor vision

III. Characteristics of Congenital Motor Nystagmus
   A. Onset within 3 months of age
   B. Usually horizontal
   C. Good vision
   D. No ocular or CNS abnormalities
   E. Bilateral
   F. Dampened by convergence and may be associated with esotropia
   G. May have a null point
   H. May have a family history
      I. Paradoxical inversion of optokinetic nystagmus response
      J. Normal ERG

IV. Characteristics of Congenital Sensory Nystagmus
   A. Poor vision
   B. Bilateral
   C. Usually horizontal
   D. Onset by 3 months of age
   E. Cause of bilateral loss of vision may or may not be readily apparent by eye examination.

V. Etiology of Sensory Nystagmus Visible on Eye Examination
   A. Cataracts: Family history, chromosomal studies
   B. Corneal opacities: Peters anomaly or herpes simplex virus
   C. Aniridia: PAX6, WT1 genetic testing
   D. Albinism: Hermansky-Pudlak syndrome
   E. Optic nerve hypoplasia: MRI of brain and endocrine evaluation
   F. Optic nerve colobomas: MRI of brain
   G. Chorioretinal scars in the macula: toxoplasmosis, cytomegalic virus and lymphocytic choriomeningitis virus titers
   H. Optic atrophy: MRI

VI. Etiology of Sensory Nystagmus With Normal Eye Exam: Need ERG
   A. Leber congenital amaurosis: Absent rod and cone response
   B. Congenital stationary night blindness: Absent rod response
   C. Achromatopsia (complete and incomplete): Absent cone response
   D. Blue cone monochromats: Partial cone response at short wavelengths

VII. Importance of Identifying Etiology of Congenital Sensory Nystagmus in Children With a Normal Eye Exam
   A. Gene therapy: Identification of patients with Leber congenital amaurosis RPE 65 gene mutation, which is the only subtype of patients that is eligible for gene therapy at the present time
   B. In vitro fertilization and preimplantation testing for future pregnancies

VIII. Acquired Nystagmus Characteristics
   A. Occurs after 3 months of age
   B. May or may not have oscillopsia
   C. Always requires MRI to rule out a structural lesion

IX. Main Etiologies of Acquired Nystagmus in Children
   A. Spasm nutans-head nodding and torticollis, diagnosis of exclusion
   B. Chiasmal or suprachiasmal glioma or mass
   C. Pinealoma: Convergence retraction nystagmus
   D. Craniopharyngioma: Seesaw nystagmus
   E. Opsoclonus: Postinfectious or paraneoplastic sign of neuroblastoma
   F. Arnold-Chiari malformation: Esotropia and downbeat nystagmus

X. Latent Nystagmus
   A. Although thought to be congenital, it is often not seen until early childhood.
   B. Occurs in children with poor fusion from strabismus or poor vision
   C. When one eye is occluded, a jerk nystagmus occurs in both eyes with a fast phase toward the uncovered eye.
   D. Binocular vision is better than monocular vision.
E. “Fogging with +6.00 lens” should be used to check vision monocularly.

F. This type of nystagmus does not require any further workup.

XI. Conclusion
A. Importance of identifying congenital vs. acquired nystagmus in children
B. If congenital nystagmus is present, identify the cause by either eye exam or ERG studies.
C. Acquired nystagmus in children always requires imaging studies.

Selected Readings

The Good, The Bad, and The Ugly: Nonaccidental Injury
Abusive Head Trauma (The Shaken Baby Syndrome)

Brian J Forbes MD PhD

Homicide is the leading cause of injury and death in infancy; 80% of infant homicides are thought to represent infant child abuse, and each year some 2000 children in the United States die as a result of child abuse. The majority of these deaths are caused by inflicted neurotrauma, which results from violent, nonaccidental shaking, blunt impact to the head, or both. Historically, the injuries resulting from repetitive unrestrained head and neck movements from shaking were termed the “whiplash shaken infant syndrome,” “shaken baby syndrome,” and now the more comprehensive term “abusive head trauma.”

Clinical findings in affected infants include subdural hemorrhage, hypoxic-ischemic brain injury, retinal hemorrhages, skeletal injuries, and cutaneous or other injuries. Unlike most other forms of ocular trauma, there are usually minimal external ocular signs of injury and no evidence of direct blows to the eye. Skeletal fractures are found in 30%-70% of injured children, and retinal hemorrhages are seen in approximately 80%. Victims of abusive head trauma are generally less than 3 years of age, and the majority are infants.

Acute Ophthalmic Findings in Abusive Head Trauma

Autopsy and in vivo studies of the acute ocular findings in infants and toddlers less than 3 years with nonaccidental head injury from abusive head trauma have described a consistent clinical picture. These characteristic ophthalmic findings include intraretinal hemorrhage with a reported frequency of 50%-100%, with most papers reporting approximately 80%. Retinal hemorrhage occurs at all levels of the retina, including blot, flame-shaped, and preretinal hemorrhage as well as vitreous hemorrhage. Retinal hemorrhages can be few in number, exclusively intraretinal, and confined to the posterior pole, although often they are too numerous to count, present at all layers, and extend to the ora serrata. Dense preretinal or vitreous hemorrhages may obscure underlying retinal hemorrhage. Retinoschisis may occur, most often in the macular area but also peripherally. Ophthalmoscopically, a dense central hemorrhage surrounded by a pale, elevated retinal fold in a circular shape. These lesions, seen both histopathologically and clinically, have also been called “hemorrhagic macula cysts” and “perimacular circular folds” and have a unique and characteristic appearance seen only rarely in other types of head trauma.

Late Ophthalmic Findings in Abusive Head Trauma

In contrast to the dramatic and relatively specific acute findings, late changes associated with abusive head trauma are neither consistent nor specific to abusive head trauma. Permanent visual impairment is frequent, and central visual impairment related to the hypoxic ischemic brain injury from abusive head trauma and optic atrophy is the most common cause of long-term reduced vision.

Abusive Head Trauma Prevention

The Shaken Baby Syndrome Prevention & Awareness Program was developed in 1998 in Upstate New York by Dr. Mark Dias MD, pediatric neurosurgeon. Since the inception of the SBS program, upstate New York has reduced the incidence of infant abusive head injuries by nearly 50%. The results of this research project were published in the Journal of Pediatrics in April 2005. In 2005, the upstate New York program expanded into pediatric offices and has shown an additional 10% decrease in infant abusive head injuries. Over the last several years, the body of robust, scientific research on infant crying, shaken baby syndrome/abusive head trauma (SBS/AHT) and the Period of PURPLE Crying program has grown significantly. Period of PURPLE Crying programs have grown extensively throughout North America over the past few years, with 49 of 50 U.S. States and 8 of 10 Canadian provinces having implemented the program at some level, as shown in Figure 1.

The Role of the Ophthalmologist in the Diagnosis and Management of Abusive Head Trauma

The primary role of the ophthalmologist in the care of these young children is to provide complete evaluation of the intraocular hemorrhages. Ophthalmic consultation allows complete assessment and documentation of the eye findings, frequently with retinal photography, an essential component of the diagnosis of abusive head trauma. In addition to establishing the diagnosis, examination provides prognostic information related to the eye findings. Physicians who treat infants and children are mandated to report suspected child abuse to child welfare agencies for investigation, and ophthalmologists who encounter children with ophthalmic manifestations of abuse need to ensure that the proper steps are taken to protect their patients from further harm.
References


“Skew You”
Skew Deviation

Sean P Donahue MD PhD

I. Definition
A. Vertical ocular deviation
B. Can be comitant or incomitant
C. Typically associated with ocular torsion
D. Often associated with abnormal head tilt
E. Pathology in brain stem
F. Can mimic oblique muscle palsy
G. Etiology typically infarction, demyelination, trauma

II. Background
A. Ocular counter-rolling reflex
1. Mediated by otoliths and semicircular canals
2. Semicircular canals project to vestibular nuclei and then to extraocular subnuclei (respond to acceleration, phasic)
3. Otolithic projections less well known but probably similar (respond to position, tonic)
B. Semicircular canal projections
1. Horizontal to horizontal recti
   a. Excitatory to ipsilateral superior rectus (SR) and contralateral inferior oblique (IO)
   b. Inhibitory to ipsilateral inferior rectus (IR) and contralateral superior oblique (SO)
2. Anterior canal
   a. Excitatory to ipsilateral superior rectus (SR) and contralateral inferior oblique (IO)
   b. Inhibitory to ipsilateral inferior rectus (IR) and contralateral superior oblique (SO)
3. Posterior canal
   a. Excitatory to ipsilateral SO and contralateral IR
   b. Inhibitory to ipsilateral IO and contralateral SR
4. Head pitch forward
   a. Excites both anterior canals
   b. Inhibits both posterior canals
   c. Elevators stimulated, depressors inhibited
   d. Eyes move up
5. Head pitch backward
   a. Excites both posterior canals
   b. Inhibits both posterior canals
   c. Depressors stimulated, elevators inhibited
   d. Eyes move down
6. Head tilt right
   a. Stimulates right anterior and posterior canals
   b. Inhibits left anterior and posterior canals
   c. Right eye incyclotorted activated
   d. Left eye excyclotorted activated
   e. Compensatory counter-rolling occurs
   f. Gain is less than 1
7. Head tilt left
   a. Stimulates left anterior and posterior canals
   b. Inhibits right anterior/posterior canals
   c. Left eye incyclotorted
   d. Right eye excyclotorted
C. Skew deviation is a perturbation of these projections.
1. Caused by lesions of the prenuclear vestibular ocular reflex pathways
2. Ocular tilt reaction
   a. Type of skew deviation
   b. Vertical strabismus head tilt
   c. Paradoxical ocular torsion
   d. Torsion and tilt in same direction
3. Can be comitant or incomitant and can mimic oblique muscle palsy depending on relative involvement of each pathway
4. Distinguishing feature is torsion as it is in the opposite direction for oblique palsy.

III. Localizing Skew Deviation
A. Caudal brainstem lesions
1. Typically caudal pons and medulla
2. Hypertropic eye contralateral to lesion
3. Lesion on side of lower eye for lower lesions
B. Rostral brainstem lesions
1. Typically upper pons and midbrain
2. Hypertropic eye ipsilateral to lesion
3. Lesion on side of higher eye for higher lesions
C. All cases are associated with torsion and head tilt toward lower-most eye.
IV. Naming Skew Deviation
   A. Neurologists typically name based upon hypotropic eye.
   B. Strabismus specialists in neuro-ophthalmology name based upon which eye is higher: “left over right skew”

V. Distinguishing Skew Deviation From Oblique Muscle Palsy: Background
   A. Asymmetric lesions to the otolithic pathways corresponding to those of a particular semicircular canal pathway produce an incomitant skew deviation that can mimic an oblique muscle palsy.
   B. Lesions most affecting contralateral posterior canal pathways can mimic a superior oblique palsy (ie, lesion on side of hypotropic eye).6
   C. Lesions most affecting anterior canal pathways on ipsilateral side can mimic an inferior oblique palsy (ie, lesion on side of hypotropic eye).2

D. Key feature is ocular torsion.
   1. Skew deviation has hypertropic eye incyclotorted or hypotropic eye excyclotored.
   2. Superior oblique palsy has hypertropic eye excyclotorted.
   3. Inferior oblique palsy has hypotropic eye incyclotorted.

E. Difference in vertical deviation from upright to supine may also be helpful.7,8

VI. Treatment of Skew Deviation10
   A. Await spontaneous resolution.
   B. Prism for small angle deviation
   C. Surgery must correct vertical and torsion.
   D. Consider using synoptophore in surgical evaluation.
      1. Assess if torsion significant.
      2. Determine potential stereopsis.
   E. Surgical options
      1. If torsion not significant (ie, fuse vertical in free space)
         a. Vertical rectus recession if large
         b. Horizontal rectus recession/resection
      2. If no fusion in free space
         a. Consider horizontal transposition of vertical rectus
         b. Consider oblique surgery

VII. Skew as a Mechanism in Childhood Strabismus
   A. Primarily theoretical
   B. Several papers postulate an association.3-5

References
AAP Federal Affairs Update

Mark Del Monte JD
Goniotomy for aniridic patients is challenging, as there is no protection for the lens due to an absent iris. This video demonstrates the angle anomaly seen in aniridia that may benefit from angle surgery. The use of prophylactic goniotomy in aniridia has not gained favor, but in those cases with early onset glaucoma where the pathophysiology of glaucoma is likely due to an anomalously developed angle (similar to that seen in primary congenital glaucoma) this procedure may still be considered as an option.
Descemet-Stripping Automated Endothelial Keratoplasty / Deep Lamellar Endothelial Keratoplasty: What Are They? Are They Good for Kids?

Gerald W Zaidman MD FACS

I. History of Corneal Transplant Surgery: PKP vs. Selective Corneal Transplant

II. Historical Overview of Corneal Transplant Surgery in Children

III. Indications for Pediatric Corneal Transplant Surgery
   A. Congenital: 65%-70%
      1. Peters anomaly, sclerocornea, dermoids: 65%-70%
      2. Dystrophies (congenital hereditary endothelial dystrophy [CHED], congenital hereditary stromal dystrophy, posterior polymorphous corneal dystrophy): 15%
      3. Congenital glaucoma: 15%
   B. Acquired nontraumatic: 20%
      1. Keratoconus: 50%
      2. Bacterial infections: 20% (contact lens wearers, rosacea)
      3. Herpes simplex virus: 14%
      4. Failed grafts: 7%
   C. Traumatic: 10%

IV. Development of Endothelial Keratoplasty
   A. Melles, 2000; Terry, 2003; Gorovoy, 2005-6
   B. Terminology and technique
      1. Deep lamellar endothelial keratoplasty (DLEK)
      2. Descemet-stripping automated endothelial keratoplasty (DSAEK), Descemet-stripping endothelial keratoplasty (DSEK)
      3. Descemet membrane endothelial keratoplasty (DMEK)

V. Pediatric Corneal Diseases That Might Require Endothelial Keratoplasty
   A. Endothelial decompensation
      1. CHED: 17 cases in literature
      2. Congenital glaucoma: 0 cases
      3. Failed grafts: 0 cases

VI. Results
   A. Adults
      1. Excellent, with high success rate in routine cases; 50% endothelial cell loss after 5 years
      2. Results more problematic in patients with glaucoma/shunts/filters
   B. Children: Limited data

VII. Conclusions
   A. An operation with good potential in a very select group of children
   B. Steep learning curve
   C. Very little short- or long-term data in children

References and Selected Readings
Endoscopic Vitreoretinal Surgery

Thomas C Lee MD
Intracameral Medications for Every Intraocular Surgery? Is This Safe for Kids?

M Edward Wilson MD, Rupal H Trivedi MD MSCR

Every pediatric eye surgeon has dreamed of the day when topical medications after surgery can be eliminated. Many parents struggle to comply with the surgeon’s instructions for preoperative and postoperative topical drops. We have all seen children with persistent inflammation, cell deposits on the surface of the IOL, and posterior synechia of the pupil. These complications are more often seen in situations where the parents have not been consistent in the application of postoperative medications.

As part of his Binkhorst Lecture in 2000, Robert Osher stated: “One of the major changes in ocular surgery I expect to see in my lifetime is the obsolescence of topical drops.” Since then, intracameral medications have been studied, in adults, for pupil dilation, infection prophylaxis, and control of postoperative inflammation. Questions remain, however. Are these medications safe for children? Do they add value to the topical regime we all use for children? Can intracameral medications replace topical applications, or can they at least reduce the consequences of parental noncompliance?

After surgery we still instruct the parents of our pediatric patients in the proper method and frequency of antibiotic and steroid drop application. However, at surgery, we are exploring every possible way to promote healing and prevent infection without having to rely on the compliance of the parents postoperatively. Can we achieve “no drops” pediatric cataract surgery? Will we see effective healing and the same low incidence of infection in noncompliant families as we do in those who comply with 4 weeks of multidose daily drops?1-7 Perhaps we can. Herein we will focus discussion on the safety and effectiveness of intracameral mydriatics, antibiotics, and triamcinolone for pediatric intraocular surgery.

References


Fundus Autofluorescence in Pediatric Ophthalmology

Elias I Traboulsi MD

Introduction
Fundus autofluorescence (FAF) imaging utilizes the fluorescent properties of lipofuscin to study the health and viability of the retinal pigment epithelium / photoreceptor complex. Lipofuscin is a heterogeneous fluorescent waste material that accumulates with age in some active postmitotic cells such as cardiac myocytes, select neurons, and the retinal pigment epithelium (RPE).1 RPE lipofuscin can be visualized in vivo using FAF imaging, and its patterns of distribution, accumulation, or absence can be characteristic of a variety of inherited or age-related retinal disorders.2 This presentation will review examples of FAF in selected inherited childhood retinal disorders and its usefulness in the diagnosis and follow-up of patients.

Physiology
Lipofuscin is derived from phagocytosed photoreceptor outer segments and normally accumulates in the RPE.3 RPE lipofuscin differs from that of other cells in that it is mainly derived from chemically modified residues of incompletely digested photoreceptor outer segments. It is composed of a mixture of lipids, proteins, and different fluorescent compounds, the main fluorophore of which is a derivative of vitamin A (retinoids). Formation of RPE lipofuscin fluorophores is almost completely dependent on a normal visual cycle, and the absence of retinal (both all-trans and 11-cis) for example in RPE65- knockout mice drastically reduces its formation. Hence normal FAF reflects the anatomic integrity of RPE and photoreceptors, normal outer segment turnover, and normal vitamin A metabolism.3

FAF Imaging Technology
Fundus autofluorescence (FAF) is recorded with a confocal scanning laser ophthalmoscope. The distribution of lipofuscin in fundus RPE using FAF was described by von Ruckmann et al.5 In the normal fundus in subjects over the age of 15 years, they found diffuse autofluorescence with the retinal blood vessels and optic disc appearing as negative shadows. In patients with long-standing retinal atrophy, they observed absent autofluorescence that corresponded spatially to the atrophy but present fluorescence in adjacent regions of surviving retina. FAF can be visualized with other cameras such as the Topcon TRC 50IX fundus camera. The highest degrees of fundus AF are detected in normal individuals at 7 degrees from the fovea and the lowest degrees are at the fovea. Physiologically reduced FAF is observed in the absence of RPE cells (eg, at the optic disc) or may be due to absorption of the incident short wavelength light by melanin, macular pigment, and the retinal vessels. Reduced FAF in retinal diseases may be due to a number of factors, including photoreceptor and/or RPE cell loss, disrupted phagocytosis, or disruption of the retinoid cycle.4

FAF in Children
Because lipofuscin accumulates with aging, levels of autofluorescence may be low in very young children.

Stargardt Disease
In Stargardt disease there are high levels of lipofuscin in the RPE. This results in high levels of autofluorescence on FAF imaging. As the disease progresses, patchy areas of loss of FAF are visualized and correspond to loss of retinal sensitivity reflecting photoreceptor cell death.6

Bestrophinopathies
In the bestrophinopathies, including Best disease, there is increased autofluorescence in the fovea and in extrafoveal lesions as a result of the accumulation of larger-than-normal levels of lipofuscin in the RPE.2, 7 A diffuse increase in FAF is detected due to the generalized accumulation of lipofuscin in RPE cells.

Leber Congenital Amaurosis
FAF may be preserved in the presence of severe photoreceptor dysfunction, as shown by undetectable full-field ERGs,4 and indicates structurally intact photoreceptors and preservation of the photoreceptor/RPE complex. In patients with CEP290 (NPHP5) and NPHP6 mutations, there is diffuse loss of FAF except in the foveal region, in which there is a preserved disc of FAF that corresponds to overlying remaining functioning cones; all rods and underlying RPE have degenerated.8 Patients with RPE65 mutations have reduced or severely reduced levels of FAF as a result of the severely reduced levels of retinoids.

Retinitis Pigmentosa
More than half of retinitis pigmentosa (RP) patients have an abnormally high-density parafoveal FAF ring (AF ring).9 This AF ring represents the border between functional and dysfunctional retina.10 Aizawa et al showed that that the size of AF ring decreased with the progression of RP. This was accompanied by a shortening of the length of the inner segment/outer segment line, a decrease in retinal sensitivity, and a worsening of BCVA.11

Fundus Albipunctatus
In fundus albipunctatus there are areas of increased AF of the RPE that correspond to the ophthalmoscopically visible lesions and RPE lesions on OCT images; in retinitis punctata albescens, in addition to the white lesions there is an enhanced AF ring in a parafoveal location.12 Mutations in RDH5 lead to a defect in oxidation of 11-cis-retinol into 11-cis-retinal. In the absence of this conversion, there is presumed storage of a retinoid, likely in an esterified form, within RPE cells. RLBP1 encodes the protein CRALBP, located within RPE and Müller cells, which binds...
to the vitamin A derivatives 11-cis-retinol and 11-cis-retinal. Impaired function of this protein could lead to the accumulation of a retinoid compound(s) within RPE cells, hence increased FAF.

**Conclusions**

Because of its ability to detect lipofuscin mainly at the RPE level, FAF is a useful method to assist in the diagnosis and progression of a wide variety of inherited and acquired retinal diseases even at stages in which fundus changes are not clearly evident on routine ophthalmoscopy. Normal or near-normal FAF may reflect the presence of structurally intact photoreceptors and preserved photoreceptor/RPE complex. Hence FAF imaging findings may have implications for gene and other therapies of inherited retinal disorders.

**References**

With this year’s passage of legislation in Kentucky that allows optometrists to perform laser surgery, the American Academy of Ophthalmology’s partnership with ophthalmic subspecialty and state societies on the Surgery by Surgeons campaign becomes even more important in protecting quality patient eye care across the country.

In 2009-2010, the Eye M.D.s serving on the Academy’s Secretariat for State Affairs collaborated with the leadership of many state ophthalmology societies on legislative battles in which optometry continued to push for expanded scope of practice. Leadership of subspecialty societies provided essential support in some of these battles. Success was reached with surgery provisions removed and/or bills defeated in Idaho, Maine, Mississippi, Nebraska, South Carolina, Texas, Washington and West Virginia.

In 2011, the stakes were raised with the disappointing outcome in Kentucky. The Kentucky legislation also includes the creation of an independent optometric board; no other board or state agency has the authority to question what constitutes the practice of optometry. The Secretariat for State Affairs continues to work diligently with state society leaders in South Carolina, Nebraska, Tennessee and Texas to ensure that a Kentucky outcome is not repeated. For example, following the passage of legislation in Kentucky, fundraising material by organized optometry in Tennessee made it clear that they would like to replicate optometry’s outcome in Kentucky and have begun discussions with state legislators.

The Surgical Scope Fund (SSF) is a critical tool of the Surgery by Surgeons campaign to protect patient quality of care. The Academy relies not only on the financial contributions via the SSF by individual Eye M.D.s but also on the contributions made by ophthalmic subspecialty and specialized interest societies. The American Association for Pediatric Ophthalmology & Strabismus (AAPOS) contributed to the SSF in 2010 and 2011, and the Academy counts on its continued support.

The results in Kentucky should be viewed as a failure neither of the SSF nor of the Academy’s Secretariat for State Affairs, which geared up immediately to strategize with Kentucky Academy physician leadership. In a period of fifteen days, with no advanced warning, optometry was able to introduce and pass a bill in the Kentucky state legislature and secure its passage into law. But a SSF disbursement actually assisted with critical media buys and powerful public messaging favoring ophthalmology and quality patient eye care for the citizens of Kentucky. This should be a lesson to each Eye M.D. in the country about the importance of contributions to your state eye PAC and to the SSF.

Leaders of AAPOS are part of the Academy’s Ophthalmology Advocacy Leadership Group (OALG), which has met for the past four years in the Washington DC area to provide critical input and to discuss and collaborate on the Academy’s advocacy agenda. The AAPOS and AAP-Section on Ophthalmology remain crucial partners to the Academy in its ongoing federal and state advocacy initiatives. As 2011 Congressional Advocacy Day (CAD) partners, the two pediatric societies ensured a strong presence of pediatric specialists to support ophthalmology’s priorities as over 350 Eye M.D.s had scheduled CAD visits to members of Congress in conjunction with the Academy’s 2011 Mid-Year Forum in Washington DC.

At the state level, the Academy’s Surgery by Surgeons campaign has demonstrated a proven track record. Kentucky was an outlier; the Academy’s SSF has helped 31 state ophthalmology societies reject optometric surgery language.

Help us help you protect our patients and quality eye care. The Academy’s SSF remains a critical tool in the Surgery by Surgeons campaign. The Academy’s SSF Committee works hard on your behalf to ensure the ongoing strength and viability of the SSF.

Thomas Graul MD (Nebraska): Chair
Arezio Amirikia MD (Michigan)
Kenneth P Cheng MD (Pennsylvania)
Bryan S Lee MD PhD (Maryland): Consultant
Richard G Shugarman MD (Florida)
Stephanie J Marioneaux MD (Virginia)
Bryan S Sires MD PhD (Washington)
Andrew Tharp MD (Indiana)
Ex-officio members:
Cynthia A Bradford MD
Daniel J Briceland MD

The SSF is our collective fund to ensure that optometry does not legislate the right to perform surgery. Do not forget about Congress, where ophthalmology’s influence is expressed through OPHTHPAC. Just as a strong state presence is needed, so do we need to remain strong in the federal arena. While OPHTHPAC is the third largest medical PAC, a mere 15% of the Academy’s membership contribute.

The Kentucky legislation is not in the best interests of patient safety and quality patient care. Ophthalmology needs the active support of every member—and this includes contributions to the Surgical Scope Fund, state eye PACs and OPHTHPAC.

Please respond to your SSF Committee and OPHTHPAC colleagues when they call on you and your subspecialty society to contribute. There are some decisions that require thought, but donating $500 to the SSF and OPHTHPAC is the easy answer for you and your patients. Do it today. Do it now.
COMETs CLAMP ATOM: The Myopia Studies—Can We Affect Refractive Outcomes?

Terri L Young MD

Myopia (nearsightedness) is a potentially blinding eye condition and the most common human eye disorder. With its increasing prevalence and earlier age of onset in recent birth cohorts, myopia now affects almost 33% of adults in the United States, and epidemic proportions of 85% to 90% of adults in Asian cities. The prevalence of myopia in Western population 7-year-old children is less than 5%, compared to 29% in Asian children. In addition to the direct economic and social burdens of myopia, associated ocular complications may lead to substantial visual loss. This presentation summarizes the current literature regarding myopia epidemiology, genetics, animal model studies, risk factors, and clinical treatments.
Iris Clip Lenses for Aphakia

Nicoline Schalij-Delfos MD

Lens dislocation can be found in children with hereditary ectopia lentis, Marfan syndrome, or homocystinuria, or after trauma. The absence of sufficient capsular support after removal of the lens interferes with in-the-bag fixation of an IOL, making IOL implantation in these cases a challenge, especially in children. The choice of lens design is open to debate, and no large, prospective, randomized clinical trials are available. In 2003 Wagoner et al performed an ophthalmic technology assessment (OTA) to evaluate options for IOL implantation in the absence of capsular support. They reviewed scientific literature published up to March 2002 and concluded that open-loop anterior chamber, scleral sutured posterior chamber (PC), and iris-sutured posterior chamber IOLs could all be used safely and effectively, without one being superior to the other. Reported complications were corneal edema, secondary glaucoma, IOL dislocation, cystoid macular edema (CME), suture erosion, and retinal detachment.

More recently other techniques have been introduced such as a capsular tension ring with PC-IOL in the capsular bag, sutured to the sclera, and endoscopic guided PC-IOL implantation in the sulcus during pars plana vitrectomy.

In 1978, the Artisan aphakia IOL (Ophtec, Groningen), a one-piece PMMA anterior chamber iris-fixated lens, designed by JG Worst, was introduced in the Netherlands. Since then, this lens has been used to correct aphakia, originally in adults but later also in children. Sminia et al published about the use of this lens in children with bilateral cataract, traumatic cataract, and lens dislocation. Nowadays, the Artisan IOL is only used in children with insufficient capsular support. The lens has a body of 5.0 mm and an overall diameter of 8.5 mm. For small eyes a pediatric design is available with a body of 4.4 mm and an overall diameter of 6.5 or 7.5 mm. The anterior chamber depth should be at least 3.3 mm, which is normally no problem in these children.

Surgical Technique

After removal of the crystalline lens and the capsular bag, constriction of the pupil is obtained with carbacholine 0.1 mg/ml or acetylcholine 10 mg/ml. A viscoelastic is used to fill the anterior chamber and protect the corneal endothelium. The Artisan is inserted through a 5-mm incision, and the claws are attached to the iris by enclavation of peripheral iris tissue. To prevent pupillary block glaucoma, a peripheral iridectomy is obligatory.

Concerns and Advantages

One of the concerns is the possible long-term negative effect on the corneal endothelial cell density. Small case studies with follow-up varying from 8 months to 10 years have shown no significant difference compared to control eyes. Prospective studies have been undertaken but follow-up is still short. Other possible causes of concern are IOL dislocation by erosion of the claws through the iris, prolonged postoperative inflammation, chronic anterior uveitis, cystoid macular edema, and iris atrophy. Dislocation of the lens is rare, since the design of the claws was adapted in the mid-1980s, but it can be seen after blunt trauma to the eye. However, repositioning is easy to perform. Other complications do not occur more frequently compared to PC-IOLs.

Advantages of implantation of an iris clip lens are that the implantation technique is straightforward and easy to acquire and there is no need for angle support, pupil fixation, or trans-scleral sutures. For post-traumatic aniridia an Artisan iris reconstruction lens is available. Furthermore, the IOL can be exchanged easily in case of substantial refractive errors when the eye is full grown. So, in conclusion, iris clip lenses are a good alternative for implantation in eyes of children without support of the capsular bag.

References

Should We Be Using Mitomycin So Readily in Pediatric Glaucoma Surgery?

Sharon F Freedman MD

I. The Problem: Poor Success of Filtration Surgery in Children
   A. Background
      1. The candidates: Refractory pediatric glaucoma
         a. Primary congenital/infantile glaucoma after failed angle surgery
         b. Aphakic/pseudophakic glaucoma (angle surgery failed first?)
         c. Juvenile open-angle glaucoma (selected cases)
         d. Other secondary glaucomas (selected cases)
      2.Mitomycin-less trabeculectomy
         a. Poor success in “plain” trabeculectomy in children?
         b. 5-fluorouracil and irradiation for pediatric filtration surgery
         c. Other filtration surgery: “Combined” trabeculectomy-otomy
         d. Alternatives to filtration surgery: Glaucoma drainage devices, cycloablation
      3. Mitomycin: Why it “works”
         a. Chemotherapeutic agent
         b. Inhibitor of cell (fibroblast) proliferation
   B. Mitomycin and adult glaucoma surgery
      1. The early days – high hopes
         a. Improved trabeculectomy success in adults
         b. Better than the competition (5-fluorouracil)
      2. Sobering truths
         a. Infections: Blebitis, endophthalmitis
         b. Leaks, early and late
      3. “Optimal” dosing: best kept secret
         a. Concentration
         b. Time of exposure
         c. Exposure location and methods
   III. Conclusions
      A. Mitomycin provides a useful tool in pediatric glaucoma surgery.
      B. There is no “free lunch.”
      C. Long-term follow-up needed.
      D. Bevacizumab – the “next” mitomycin?

Selected Readings

Do Adjustable Sutures Enhance Outcomes?

*Paolo Nucci MD, Massimiliano Serafino MD, Matteo Sacchi MD*

Not all strabismus surgeons use adjustable sutures (AS). To tell the truth, the attitude toward AS seems to be fideistic: some surgeons use them in almost all situations, while some are reluctant even when there is a strong evidence that AS could obtain better results.

Our attempt, apparently reactionary and “against the current thinking,” is to give voice to the group of surgeons not keen to do this surgery. We interviewed 33 surgeons reluctant to perform the AS technique, and we collected the reasons they asserted to justify their aversion to AS.

I. “Sincerely, I am not comfortable in operating awake patients.”

Corollary: Strabismus surgeons are often people not as used to finely move forceps and scissors as the majority of ophthalmic surgeons, and they can treat muscles and tissues quite aggressively. Topical or local anesthesia creates anxiety for these surgeons and does not warrant complete comfort for the patient.

II. “Dealing with a sixth cranial nerve palsy there is not much to adjust; I have no risk for overcorrecting the condition.”

Some surgeons prefer to maximally recess the antagonist, medial rectus, and consider resection of a paretic muscle unpredictable. Lastly, they are convinced that transposition surgery takes effect only few days after the procedure... so...!

III. “Inferior rectus recession, the most needed surgery for Graves ophthalmopathy, tends to increase in effect with time.”

Strabismus surgeons believe there is no reason to strive to obtain the alignment intraoperatively (or the day after) if muscle function will change weeks or months after the operation.

IV. “How reliable is cover test evaluation in the OR?”

Do surgeons' and patients' expectation to quickly complete the maneuvers concur with a truly accurate measurement? Is quality of vision after ocular manipulation always enough to disclose fine misalignment? These are questions related to the reliability of finely adjusting the eye position.

V. “AS have good effect only if you can restore the normal binocular vision (NBV); there is no reason to opt for this surgery in conditions in which NBV cannot be restored.”

In other words, we trust in stereopsis restoration to be sure our surgery obtained the best possible result.

VI. “AS are effective only with topical anesthesia.”

The idea behind this exception is that even local subtenon anesthesia could affect muscle movement and induce unpredictable results when the drug effect fades.

VII. “No prospective, randomized, double-blind studies show that the long-term results of strabismus surgery are better using AS.”

As a matter of fact, a recent Cochrane study did not find any randomized controlled trials comparing adjustable to non-AS for strabismus surgery.
Congenital Corneal Opacification: Time for a Re-think?

Kanwal K Nischal MBBS

Traditionally, congenital corneal opacification has been considered in terms of various etiologies and these have often been remembered by the following mnemonic:

S: Sclerocornea
T: Trauma
U: Ulcer
M: Metabolic
P: Peters anomaly
E: Endothelial dystrophy
D: Dermoid

Although this is helpful, it doesn’t help us formulate a plan or speculate on prognosis for a child with congenital corneal opacities. Over 12 years this author has seen over 200 children with congenital corneal opacification, and it has become apparent that there is a pattern that lends itself to a clinically more practical classification.1

Corneal opacities may be considered as being primary or secondary:

- Primary corneal problem
  - Corneal dystrophy (e.g., congenital hereditary endothelial dystrophy, posterior polymorphous dystrophy, congenital X-linked endothelial dystrophy)
  - Corneal dermoid
  - Isolated sclerocornea: Termed CNA1 or CNA2. This is not associated with total corneal opacification.2

- Secondary corneal problem
  - Congenital
    - Primary intraocular problem (e.g., affecting the lens, trabecular meshwork, or iris)
    - Acquired (e.g., infection, trauma, inflammation)

It is no coincidence that primary corneal problems do better in the published literature with corneal transplantation than do secondary ones.3

Now let us look at the secondary corneal problems in greater detail.

Secondary Corneal Problems

Lens

These can be termed kerato-irido-lenticular dysgenesis (KILD):

- Lens fails to separate from cornea (aka Peters anomaly type II)
  - Developmental (e.g., as seen in Dyl mouse4)
  - Mechanical. Clues to this include:
    - Lens epithelium discernible on ultrasound biomicroscopy
    - Evidence of persistent pupillary type membrane(s)
    - Intact iris stromal architecture

- Lens separates but fails to form thereafter.

- Lens fails to form (primary aphakia; e.g., as seen in aph mouse5)
- Due to persistent hyperplastic primary vitreous, lens is literally pushed forward.

Trabecular meshwork

Infantile glaucoma causes corneal opacity that reverses if IOP is controlled quickly enough.

Iris

- Iridocorneal adhesions (aka Peters type I)
- Iris anomalies (e.g., Axenfeld Rieger anomaly/syndrome; aniridia)

It is well known that if at the time of surgery the lens is removed due to keratolenticular adhesions (i.e., due to KILD above), then the chances of graft survival are reduced. Similarly if there is no lens (i.e., primary aphakia), the chances of successful corneal transplant are reduced significantly. Under these conditions it becomes important to consider alternatives such as optical iridectomy if possible.

This approach should not be surprising; we would never consider a corneal graft for a child presenting with hazy corneas due to infantile glaucoma since the primary problem is not the cornea but the pressure in the eye.

Similarly, we need to change our approach to corneal opacification and determine the primary disease before acting on a treatment algorithm.

The one exception to this is where there are only iridocorneal adhesions causing opacities in the cornea. Here there is published evidence that good results can be attained but only if the sole abnormality is iridocorneal adhesion and not abnormal iris with iridotrabecular anomalies.6

References

Avastin for ROP

Helen A Mintz-Hittner MD FACS

1. Off-Label Use of Bevacizumab (Avastin) for ROP: Efficacy = Benefit
   A. Timing is critical for antivascular endothelial growth factor (VEGF) for ROP.
      1. Pre-emptive strikes using bevacizumab in Stages 1 and 2 are not appropriate:
         A severe retinal dystrophy will likely occur if anti-VEGF therapy is given before the appearance of plus disease, usually before 31 weeks postmenstrual age.
      2. Rescue attempts using bevacizumab in Stages 4b and 5 are not appropriate:
         An accelerated tractional retinal detachment may well occur by contraction of dense vasoproliferative membranes, usually after 44 weeks postmenstrual age.
   3. Rescue therapy using bevacizumab following laser in advanced Stage 3+ and 4a can be very effective, although the ideal time for treatment (ETROP) has been missed and the infant already has been subjected to the natural consequences of and random complications of laser therapy, unnecessarily.
   4. Combination therapy using bevacizumab with laser for plus disease and extraretinal fibrovascular proliferation as conventional Stage 3 ROP or as aggressive posterior ROP does not increase efficacy.

Decreasing VEGF does not require this 2-pronged offensive: ablative laser therapy is not additive to bevacizumab (as anti-EPO is additive to anti-VEGF). Laser destroys the natural retinal barrier and allows bevacizumab to escape more easily.

Figure 1. Pathogenesis and therapy for ROP: Postmenstrual ages of Phase I (cessation of normal vessel growth) and Phase II (initiation of neovascularization) in relation to the development of retinopathy of prematurity stages. Reprinted with permission from the New England Journal of Medicine. Mintz-Hittner HA, Kennedy KA, Chuang AZ. Efficacy of intravitreal bevacizumab for Stage 3+ retinopathy of prematurity. NEJM 2011; 364(7):603-615.
freely. This may make it necessary to repeat bev-
acizumab injections before inner retinal vascu-
larization has been completed more frequently than
use of bevacizumab alone, and thereby exposes
the infant to higher systemic levels of this anti-
VEGF therapy.

5. Monotherapy using bevacizumab (without laser)
for plus disease and extraretinal fibrovascular
proliferation as conventional Stage 3 ROP or as
aggressive posterior ROP is recommended.

The regression of extraretinal fibrovascular pro-
liferation and the continued vascularization of
the peripheral retina subsequent to bevacizumab
monotherapy have been documented by several
case reports, case series, a prospective, random-
ized, multicenter clinical trial,1 human histopa-
thology,3 and rat histopathology.4,5 The BEAT-
ROP clinical trial 6-month results have been
published; however, 1- through 5-year results
will be forthcoming:

a. Long-term ocular structural data (fundus
photographs, fluorescein angiograms, and
OCTs) will be evaluated by masked examin-
ers with potentially lasered areas cropped.

b. Long-term ocular functional data will be
obtained by independent masked examin-
ers. These data will report ocular outcomes
including visual acuity (isolated and linear),
contrast sensitivity, color vision, depth per-
ception, visual field, refraction, and presence
or absence of strabismus.

B. Appearance of the retina following bevacizumab
therapy

1. With regression following bevacizumab, retinal
vessels advance to the location that the endothe-
lial precursors had reached at the time of preterm
birth, not necessarily to the ora serrata. At this
location, a ridge of variable thickness is usually
seen.

2. Often, in the most immature infants (gesta-
tional age 22 to 26 weeks), the peripheral retina
remains avascular and never differentiates with
thickening of the retina. Thus, the peripheral
avascular retina is not hypoxic, VEGF is not
increased, and ablative laser is not usually
required. (High myopia and tractional elements
may generate peripheral patholgy later if present.)

3. With recurrence following bevacizumab, plus
disease returns and extraretinal fibrovascular
proliferation is seen again (1) from the original
location of extraretinal fibrovascular prolif-
eration and (2) from the advancing edge as an
increasingly thick ridge—“two lines of recur-
rence.”

4. When recurrence is recognized early, re-treat-
ment with bevacizumab is possible—retinal ves-
sels advance to the location that the endothelial
precursors had reached at the time of preterm
birth, not necessarily to the ora serrata. (If thick
fibrovascular membranes have formed, some
peripheral laser may be required to prevent late
tractional retinal detachment.)

II. The Worrisome Unknown: Toxicity = Risk

A. A developmental evaluation will also be performed
by masked examiners including motor, language,
and other neurologic functions to identify any evi-
dence of central nervous system toxicity6-8 in survi-
ors of the BEAT-ROP clinical trial.

B. Additional organ systems will also be evaluated in
survivors of the BEAT-ROP clinical trial to identify
other organ system damage, especially effects of
abnormal pulmonary vascularization,9 that may
occur as a result of bevacizumab therapy.

C. The importance of limiting the dose of bevacizumab
(perhaps identifying a larger molecular weight drug
that will exit the eye less readily) and of being vigi-
1
lant for any signs of toxicity cannot be over empha-

III. Future Trials

A. Any prospective, randomized, multicenter clinical
trials must randomize infants, rather than eyes,
in order to clearly separate efficacy and toxicity.
Bevacizumab definitely gets out of the eye to some
extent, especially in association with leakage from
fragile neovascular vessels. Thus, bevacizumab
injection in one eye will affect the ROP in the fellow
eye. Also, VEGF circulates in the blood, so recur-
rence in one eye may stimulate recurrence in the fel-
low eye.

B. When randomizing eyes, rather than infants, reac-
tion to extensive laser, especially following treat-
ment for zone I disease, may cause deprivation
amblyopia by creating vitreous reaction (which can
be observed by slitlamp examination) or may cause
structural amblyopia by creating macular edema (as
shown by OCT) compared to the fellow eye.

C. Additionally, when randomizing eyes, rather than
infants, (1) all infants are being exposed to any
potential ocular and systemic toxicity of bevacci-
sumab, (2) all infants are at risk for the possible
consequences of laser in one eye (decreased field,
anisometropic amblyopia due to myopia, etc.), and
(3) all infants are at risk for the potential infrequent
complications of laser in one eye: corneal and len-
ticular opacities, ie, leukomas and cataracts, angle-
closure glaucoma, hemorrhage in the anterior or
posterior segments of the eye, decreased pressure
(phthisis), etc.

D. Determination of a minimally required dose is
essential. Thus, pharmacokinetics of all utilized
bevacizumab (and/or other anti-VEGF drugs) doses
should be planned. Comparison of VEGF levels
(and levels of other angiogenic factors) following
laser and different doses of bevacizumab should
be included in the study design. Documentation of
the retinal appearance photographically and with OCTs prior to and at predetermined times following treatment should be included in the trial design. The relationship of each dose to efficacy and the time of recurrence—number of recurrences associated with each dose (%) and the postmenstrual age at which recurrence is noted (weeks)—are both important to develop clinical guidelines for postinjection follow-up examinations. (Large numbers of infants will be required for these trials.)

E. Determination of efficacy of bevacizumab (compared to laser) for zone II is warranted. The BEAT-ROP clinical trial did not enroll enough infants to answer the question of bevacizumab efficacy for zone II ROP because recurrences are so infrequent in zone II. (Large numbers of infants will be required for these trials.)

F. Keep vigilant for very long-term toxicity. (Large number of infants—in national or international registry—will be required to identify any related toxicity definitively.)

IV. Conclusion

Because of severe zone I cases and imperfect results following laser surgery, further investigation of this less destructive, less labor intensive, readily available therapy is warranted.

References


Video: Cataract Surgery for Marfan Syndrome

Daniel J Salchow MD

The surgical treatment of a child with Marfan syndrome and lens subluxation is discussed using the edited surgical video of the case. The surgeon may be challenged in several ways, including (1) removal of the unstable lens, (2) treatment of vitreous prolapse, and (3) fixation of an IOL without capsular support. These and other aspects of the surgery are discussed, and different options are presented.
Why Don’t We Operate to Eliminate Lower-Power Hyperopic Spectacles in Accommodative Esotropia?

Scott E Olitsky MD

I. What causes accommodative esotropia?
   A. Accommodation
   B. Accommodative convergence
   C. Accommodative convergence-to-accommodation ratio (AC/A)
   D. Fusional divergence
   E. Esotropia occurs when the stimulus to cross outweighs the control to prevent it.

II. What is the natural history of accommodative esotropia?
   A. Level of hyperopia
   B. AC/A ratio
   C. Previous strabismus (fusional divergence)
   D. Changes in hyperopia
   E. Changes in AC/A ratio

III. The Child With Accommodative Esotropia and Normal or Low Levels of Hyperopia
   A. Normal AC/A ratio
   B. High AC/A ratio

IV. Why would we consider surgery to eliminate glasses in these children?
   A. Limited desire to wear glasses
   B. Minimal benefit in visual acuity
   C. Potential increase in amount of time eyes are aligned
   D. Eliminate diplopia in some children when not wearing glasses
   E. Eliminate the need for glasses in an active population

V. Surgery to Eliminate the Need for Spectacles
   A. The history of acceptance (or lack thereof)
      1. Stages of acceptance
      2. Current state of the art
   B. Types of refractive surgery
      1. Corneal
      2. Lens
      3. Strabismus?

VI. Potential Options for the Child With Normal or Low Levels of Hyperopia and Accommodative Esotropia
   A. High AC/A ratio
      1. Strabismus surgery for excess esotropia at near
      2. Medication
      3. Observation
   B. Normal AC/A ratio
      1. Strabismus surgery
      2. Refractive surgery
         a. Full correction of hyperopia
         b. Partial correction of hyperopia to maintain alignment
Is the Pediatric Eye Disease Investigator Group Wrong About . . . ? Amblyopia Treatments Are All Equal

Steven M Archer MD

I. Comparative PEDIG Amblyopia Treatment Studies
   A. ATS1: Daily atropine and patch 6 hours/day are equal.
   B. ATS2A: Patch 6 hours/day and full-time are equal.
   C. ATS2B: Patch 2 hours/day and 6 hours/day are equal.
   D. ATS4: Weekend atropine and daily atropine are equal.
   E. ATS6: Patch with near and distance activities are equal.
   F. ATS8: Atropine with and without optical penalization are equal.
   G. ATS9: Weekend atropine and patch 2 hours/day are equal.
   H. ATS10: Bangerter foil and patch 2 hours/day are equal.

II. Why is the result always “equal?”
   A. These are studies of “prescribed treatment.”
      1. If compliance is poor, all groups may actually get similar treatment.
      2. Consistent with “intention to treat” analysis, open to question
   B. Study design factors
      1. Not possible to prove the null hypothesis—“absence of proof is not proof of absence.”
         a. Statistical power calculations give some limits.
         b. Noninferiority design; design errors favor equivalence.
         c. Equivalence design (two one-sided tests—TOST)
      2. PEDIG study design is always meticulous, but could it be biased to favor an “equal” result?

III. Anatomy of an “Equal” Result: ATS1
   A. Motivation behind the study
      1. Atropine evangelists propose the study.
      2. Those who propose the study help design the study.
   B. Design the study to achieve the desired result
      1. Minimize disadvantage of atropine being slow
         a. Make this a relatively long (6-month) study
b. Give atropine a chance to catch up by allowing decreased/discontinued treatment if early “success” (for about one-fourth of patients; this may have actually been a trial of 5 weeks of patching compared to 13 weeks or 6 months of atropine.)
   2. Less effective treatment looks equal if you set the bar low enough.
      a. “Success” is final acuity ≥ 20/30 or ≥ 3 lines improvement
      b. Mean starting acuity is 20/68 (range: 20/40-20/100)
         i. Is 20/40 → 20/30 a success? (One patient in the atropine group actually started at 20/30.)
         ii. Does 20/70 → 20/30 meet expectations for 6 months of treatment?
   C. Results reported honestly but analysis reported selectively
      1. Difference in final acuity
         a. 0.034 logMAR
         b. 95% CI, 0.005-0.064
         c. P = .03 (not actually reported)
      2. “Success” (final acuity ≥ 20/30; improvement ≥ 3 lines)
         a. Patching: 164/208 (79%)
         b. Atropine: 144/194 (74%)
         c. P = .29 (not actually reported)
      3. Improvement ≥ 3 lines
         a. Patching: 146/208 (70%)
         b. Atropine: 116/194 (60%)
         c. P = .04 (not actually reported)
      4. Final acuity ≥ 20/30
         a. Patching: 132/208 (63%)
         b. Atropine: 103/194 (53%)
         c. P = .04 (not actually reported)
      5. Final acuity ≥ 20/25
         a. Patching: 85/208 (41%)
         b. Atropine: 56/194 (29%)
         c. P = .01 (not actually reported)
6. Parental questionnaire scores
   a. Worse on all 3 subscales for patching vs. atropine
   b. \( P = .002, P < .001, P < .001 \) (the only comparative amblyopic eye results with reported \( P \)-values)

D. Spin the conclusion
1. Primary outcome: 6-month visual acuity in amblyopic eye
   a. Result: Visual acuity at the final 6-month exam is significantly better in the patching group than the atropine group.
   b. PEDIG description: “Atropine and patching produce improvement of similar magnitude. . . . [T]he difference in mean visual acuity between groups was small . . . and clinically inconsequential.”
2. Secondary outcome: 6-month “success”
   a. Result: The difference in “success” rates is not statistically significant; however, patching is significantly better with regard to each component of “success” (acuity ≥ 20/30 and ≥ 3 lines improvement).
   b. PEDIG description: “The difference in the percentage of patients meeting our criteria for successful treatment . . . was also small.”
3. Other factors: Speed of improvement
   a. Result: Visual acuity in the atropine group is worse at 5 weeks and never catches up to the patching group, even after 6 months. This, in spite of the fact that treatment may have been reduced or discontinued in more of the patched patients well before the 6-month conclusion of the study.
   b. PEDIG description: “Improvement in the atropine group lagged behind that in the patching group. It is possible that if our primary outcome had occurred at a time point longer than 6 months, the atropine group might have shown further improvement, perhaps achieving the same proportion of patients with 20/30 or better amblyopic eye acuity as found in the patching group.”

IV. Is PEDIG wrong about atropine vs. patching?
A. 5 weeks of patching \( \equiv \) 15 weeks of atropine.
B. Atropine is effective, but for any treatment duration up to at least 6 months, it is significantly less effective than patching. Is the difference really “clinically inconsequential”?
   1. If each patient’s 6-month vision is only one-third line better with patching, as the authors imply, that may be of little importance.
   2. Alternatively, if one-third of patients are a full line better at 6 months with patching, that sounds more substantive.
C. In the long run, when investigators can prescribe subsequent treatment in whatever manner they choose, the initial 6 months of treatment has little bearing on the outcome at 2 or 10 years.

Selected Readings
Eye Drops for Nystagmus? Really?

Richard W Hertle MD

Introduction

Nystagmus comes from the Greek word *nystagmos*, “to nod,” “drowsiness,” and from *nystazein*, “to doze”; probably akin to Lithuanian *snusti*, also “to doze.” It is a rhythmic, involuntary oscillation of one or both eyes. Using the information obtained from a complete history, physical examination, and radiographic and eye movement recordings, over 40 types of nystagmus can be distinguished. Some forms of nystagmus are physiologic, whereas others are pathologic. Although the nystagmus is typically described by its more easily observable fast (jerk) phase, the salient clinical and pathologic feature is the presence of a slow phase in one or both directions. Thus, clinical descriptions of nystagmus are usually based on the direction of the fast phase and are termed horizontal, vertical, or torsional, or any combination of these. The nystagmus may be conjugate or dysconjugate. The nystagmus may be predominantly pendular or jerky, the former referring to equal velocity to-and-fro movement of the eyes, and the latter referring to the eyes moving faster in one direction and slower in the other. Involuntary ocular oscillations containing only fast phases are “saccadic oscillations and intrusions” and not nystagmus. It is well documented that these differences may be difficult, if not impossible, to differentiate clinically and can only be accomplished with eye movement recordings. Recent advances in eye movement recording technology have increased its application in infants and children who have clinical disturbances of the ocular motor system.

We define the ocular motor condition infantile nystagmus syndrome according to the National Eye Institute collaborative Classification of Eye Movement Abnormalities and Strabismus (CEMAS; see Table 1). Estimations of incidence of the most common form of nystagmus (infantile nystagmus syndrome, INS) vary from 1 in 500 to 1 in 6000. Other clinical characteristics include increased intensity with fixation and decreased with sleep or inattention, variable intensity in some position of gaze (a null position), changing direction in different positions of gaze (about a neutral position), decreased intensity (damping) with convergence, anomalous head posturing, strabismus, and the increased incidence of significant refractive errors. INS can occur in association with congenital or acquired defects in the visual sensory system (eg, albinism, achromatopsia, and congenital cataracts).

The visual symptoms are inversely proportional to the frequency (and speed) of the oscillation in patients with nystagmus. Visual sensitivity for both pattern and movement detection is reduced because of these eye movements. The object of regard spends little time in the foveal area, and image movement, often in excess of 80 degrees/second, causes blur, oscillopsia, diplopia, and vertigo. These symptoms begin at retinal slip velocities of greater than 4 degrees/second. Abolishing or reducing the nystagmus frequency would ameliorate these symptoms. Ideally, the treatment of nystagmus should be directed against the pathophysiological brain mechanism responsible for the ocular oscillation. In the absence of directly affecting neurological function, secondary ameliorative therapies treat the eyes directly (ie, prism glasses, contact lenses, occlusion, botulinum toxin and anesthetic injections, and eye muscle surgery).

Treatments

Infantile nystagmus may respond to surgical, medical, and optical treatments.

Table 1. A Classification of Eye Movement Abnormalities and Strabismus (CEMAS): Report of a National Eye Institute–Sponsored Workshop

<table>
<thead>
<tr>
<th>Classification</th>
<th>Description</th>
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<tbody>
<tr>
<td>1. Peripheral vestibular imbalance</td>
<td>Meniere, drug toxicity</td>
</tr>
<tr>
<td>2. Central vestibular imbalance</td>
<td>Downbeat, upbeat, drug toxicity</td>
</tr>
<tr>
<td>3. Instability of vestibular mechanisms</td>
<td>Periodic alternating nystagmus</td>
</tr>
<tr>
<td>4. Disorders of visual fixation</td>
<td>Vision loss, see-saw nystagmus, drug toxicity</td>
</tr>
<tr>
<td>5. Disorders of gaze holding</td>
<td>Gaze evoked, acquired pendular, drug toxicity</td>
</tr>
<tr>
<td>6. Acquired pendular nystagmus</td>
<td>Central myelin, oculopalatal, Whipple, drug toxicity</td>
</tr>
<tr>
<td>7. Saccadic intrusions and oscillations</td>
<td>Square wave jerks, macrosaccadic oscillations, opsoclonus</td>
</tr>
<tr>
<td>8. Miscellaneous eye movements</td>
<td>Superior oblique myokymia, oculor motor neuromyotonia</td>
</tr>
<tr>
<td>10. Fusion maldevelopment nystagmus syndrome</td>
<td>Latent, manifest latent, nystagmus blockage</td>
</tr>
<tr>
<td>11. Spasmus nutans syndrome</td>
<td>Without optic pathway glioma, with optic pathway glioma</td>
</tr>
</tbody>
</table>
### Table 2. Incidence of Operation Types

<table>
<thead>
<tr>
<th>Operation Type (100 Patients)</th>
<th>Percent</th>
</tr>
</thead>
<tbody>
<tr>
<td>Operation 1: Horizontal head posture alone</td>
<td></td>
</tr>
<tr>
<td><em>Horizontal rectus recess and resect or recess and tenotomy + reattach</em></td>
<td>22</td>
</tr>
<tr>
<td>Operation 2: Chin down head posture (± strabismus)</td>
<td></td>
</tr>
<tr>
<td><em>Superior rectus recess 5.0 mm + inferior oblique myectomy</em></td>
<td>16</td>
</tr>
<tr>
<td>Operation 3: Strabismus alone</td>
<td></td>
</tr>
<tr>
<td><em>Primary position deviation using at least 2 recti each eye</em></td>
<td>15</td>
</tr>
<tr>
<td>Operation 4: Horizontal head posture + strabismus</td>
<td></td>
</tr>
<tr>
<td><em>Fixing eye straightens head + nonfixing eye straightens eyes</em></td>
<td>10</td>
</tr>
<tr>
<td>Operation 5: Chin up head posture (± strabismus)</td>
<td></td>
</tr>
<tr>
<td><em>Inferior rectus recess 5.0 mm + superior oblique tenectomy 5.0 mm</em></td>
<td>10</td>
</tr>
<tr>
<td>Operation 6: No head posture, strabismus, or vergence damping</td>
<td></td>
</tr>
<tr>
<td><em>Horizontal rectus tenotomy + reattach</em></td>
<td>9</td>
</tr>
<tr>
<td>Operation 7: Multiplanar head posture (± strabismus)</td>
<td></td>
</tr>
<tr>
<td><em>Transposition of recti + combinations of oblique or recti recess</em></td>
<td>7</td>
</tr>
<tr>
<td>Operation 8: Vergence damping alone (artificial divergence)</td>
<td></td>
</tr>
<tr>
<td><em>Medical rectus recess 3.0 mm + lateral rectus tenotomy + reattach</em></td>
<td>6</td>
</tr>
<tr>
<td>Operation 9: Torsional head posture alone</td>
<td></td>
</tr>
<tr>
<td><em>Horizontal transposition of vertical recti 1 tendon width</em></td>
<td>5</td>
</tr>
</tbody>
</table>
Topical Medications

In 1979, Dell’Osso and Flynn recorded eye movements of 3 patients before and after surgery for INS. In addition to shifting the nystagmus null, they observed broadening of the null region and an overall reduction of nystagmus intensity at all gaze angles. This led them to speculate that the surgery caused “non-linear changes in ocular motor plant dynamics (i.e., changes in the characteristics of the muscles, tendons, Tenon’s capsule, fatty and scar tissue interactions) as a result of the surgical changing of the points of insertion and methods of attachment of the muscles to the globe.” Bosone et al found similar results. Subsequently, Dell’Osso et al and Hertle et al showed that eye muscle tenotomy and reattachment (T&R) alone had salutary effects on nystagmus amplitude and velocity in dogs with nystagmus and in 2 human trials in patients with INS. A hypothesis evolved that T&R damaged proprioceptive structures in the eye muscle tendon at its insertion on the globe (enthesis) that favorably affected the nystagmus oscillation. Enthesial neurons recently identified and studied by Hertle et al and Buttner-Ennever et al have been shown to have proprioceptive anatomy and physiology. They probably provide feedback that assists with alignment and stabilization of the eyes. It has been also shown over the last 10 years that surgical disruption of the enthesis (and associated enthesial neuroanatomy) in patients with INS results in long-standing beneficial effects on nystagmus and visual function. The neurological hypothesis for the “improvement” in the nystagmus is that there is a reduction of small-signal gain of the ocular motor plant by interfering with enthesial, neural proprioceptive tension control. Enthesial nerves are probably palisade type nonswitch motoneurons and are likely involved in modulating the gain of sensory feedback from the eye muscles analogous to the gamma motoneurons, which control the gain of proprioceptive feedback in skeletal muscles.

In general, the membrane potential of these neurons that are not transmitting signals is called their resting potential and is typically between -60 and -80 mV. In all neurons, the resting potential depends on the ionic gradients that exist across the plasma membrane. In general, mammalian neurons have an extracellular Na⁺ concentration of 150 millimolar (mM) and a K⁺ concentration of 5 mM. In the cytosol, the Na⁺ concentration is 15 mM and the K⁺ concentration is 150 mM. The gradients are maintained by sodium-potassium pumps in the plasma membrane. These ion pumps use the energy of ATP hydrolysis to actively transport Na⁺ out of the cell and K⁺ into the cell. Gradients of K⁺ and Na⁺ across the plasma membrane represent potential energy. Converting this chemical potential to electrical potential involves ion channels, pores formed by clusters of specialized proteins that span the membrane. Acid-sensing ion channels (ASIC) form a subset of voltage-independent cation channels that predominantly conduct Na⁺ ions, and were identified at the molecular level a little more than a decade ago. ASICs form effective proton sensors in both central and peripheral sensory neurons.

Carbonic anhydrase (CA) may play an important role in the neurochemical functioning of these enthesial ending’s membrane potential as it does in other sensory systems. Hansson’s enzyme histochemical method for the demonstration of carbonic anhydrase has found numerous carbonic anhydrase positive neurons in the trigeminal and geniculate ganglia as well as in the mesencephalic trigeminal nucleus. There is evidence that CA participates in the response of sensory stretch receptors of the trigeminal nerve and its nerve endings. CA inhibition has been shown to attenuate the steady-state inhibitory response of laryngeal receptors to airway CO₂ and to completely block the inhibition of pulmonary stretch receptor activity caused by airway CO₂. A functioning CA system may be involved in facilitating enthesial neuronal feedback to central ocular motor areas continuing to enhance the developmentally disturbed circuit, resulting in the ocular oscillation of INS. A CA inhibitor (CAI) may interfere with the sodium-potassium ATPase membrane bound system, thus interrupting enthesial neurophysiology (analogous to surgery), creating a damped circuit resulting in improvement in the ocular oscillation and subsequent enhanced visual function.

Case Report

Sixty-seven-year-old white male with INS known to PI and no medical or surgical treatment for INS other than prism spectacles was prescribed topical CAI as part of associated eye condition. Eye movement recordings and subsequent calculation of his NAFO as a function of gaze was accomplished prior to and after beginning his topical CAI and compared to no treatment, contact lenses, and convergence (all known to affect the nystagmus waveform and associated NAFO). The data from that single anecdotal case is presented below. The data show an improvement of NAFO and “predicted” acuities across all gaze angles after topical administration of a topical CAI. The curves represent NAFO as a function of gaze, the best NAFO is seen across gaze with convergence (Figure 1, top dashed line), followed by topical CAI only, contacts only and least with no convergence, contacts or CAI from top to bottom.

Topical Brinzolamide Ophthalmic Suspension vs. Placebo in the Treatment of Infantile Nystagmus Syndrome

This study is currently recruiting participants. Verified on March 2011 by Akron Children’s Hospital. Study NCT01312402. Information provided by Akron Children’s Hospital, First Received on January 21, 2011. Last Updated on March 9, 2011.

Brief summary

This study is a prospective, single crossover, double-masked, controlled clinical trial that will use topical brinzolamide (Azopt) ophthalmic medication to try to improve the nystagmus and visual consequences of nystagmus in patients with infantile nystagmus syndrome (INS). Subjects will undergo a clinical exam, questionnaire, and eye movement recordings on Day 1 and then receive either topical Azopt or placebo 3 times a day in both eyes for Days 2, 3, and 4, followed on the morning of Day 5 by a repeat clinical exam, questionnaire, and eye movement recordings. After at least 1 week, this protocol is repeated with the crossover regimen being taken by the subject. One week after all medications are discontinued, another clinical exam is done before study discharge. The hypothesis is that nystagmus and associated visual symptoms will be improved while on the Azopt compared to the placebo. There will be a total of 5 visits over a 1-2 month period. For more information on the current study, visit http://clinicaltrials.gov/ct2/show/NCT00967226.
Figure 1. Abbreviations: Poly indicates polynomial fit curve; positive gaze angle, gaze right; negative gaze angle, gaze left; LFD, patient PFD; Conv, convergence; PD, prism diopters.
Do Study Design and Methodology Affect Pediatric Cataract Outcomes?

Ramesh Kekunnaya FRCS, Sumit Monga FRCS

The advances in technology and refinements in surgical techniques over the past two decades have catapulted pediatric cataract surgery into a new era. This has led to vastly improved surgical and functional outcomes. Various innovative studies have contributed to the current understanding and evolution of surgical techniques that have helped to partly overcome the challenges of pediatric cataract management. However, pediatric cataract surgery has been associated with many complex issues that have been debated in literature. We believe that possibly the information on key aspects of pediatric cataract surgery may have been influenced by study methodologies and surgical techniques that have been reported in the literature.

Surgical Technique

Pediatric cataract surgery has evolved from the procedure of discission and aspiration in 1930s, lensectomy in the 1970s, and extracapsular cataract extraction in 1980s to the present surgical technique of anterior continuous capsulorrhexis with phaco-aspiration of lens matter with primary posterior capsulorrhexis with limited anterior vitrectomy.

The use of mechanized vitrectomy instrumentation to selectively perform a primary posterior capsulotomy and vitrectomy combined with IOL implantation resulted in decreasing the scourge of visual axis opacification and has led to fewer recoveries in younger children. In the literature, the rate of posterior capsular or visual axis opacification (PCO or VAO) is up to 100% when the posterior capsule remains intact. There is a strong relationship between age and incidence of PCO or VAO. The rate of membrane formation is high in young children, reflecting greater tissue reactivity of lens epithelial cells (LECs). When primary posterior capsulectomy (PPC) is not combined with anterior vitrectomy, the incidence of posterior capsule closure is up to 60%. The main reason for the occurrence of VAO after performing a posterior continuous curvilinear capsulorrhexis could be the increased LEC activity and also the presence of an intact anterior vitreous face, which acts as a scaffold for LEC migration. Reported rates of VAO after vitrectomy are less than 6%. The importance of anterior vitrectomy in pediatric surgery is emphasized in cataract surgery in patients younger than 7 years by several authors. In a study, the technique of optic capture through the posterior capsulorrhexis has been shown to prevent PCO. However, Vasavada et al found that optic capture without anterior vitrectomy did not always ensure a clear visual axis. Eyes with an obscured visual axis had reticular fibrosis of the anterior vitreous face in the first 2 months after surgery. Hence, vitreous opacification could be a primary response of the anterior vitreous face when it occurs with the IOL optic rather than a secondary scaffold response caused by proliferating LECs, inflammatory cells, and exudate deposits.

As per the current understanding, one can make the following recommendations. In children younger than 5 years, PPC with anterior vitrectomy is advisable. PPC without anterior vitrectomy may be considered in children between 5 years and 7 years. If the child is cooperative, Nd:YAG capsulotomy may be the other option. In older children, maintaining intact posterior capsule is debatable, as we see more and more PCO formation. There are no clear-cut data available on this issue. Personally we favor PPC until 10 years of age.

Primary IOL Implantation in Children

IOLs are being used increasingly for the optical correction of aphakia in infants following cataract surgery. In children older than 2 years of age, primary IOL implantation of a foldable acrylic IOL is the current standard of care. There is growing evidence in the literature to support the use of primary IOL implantation in children less than 2 years of age. While the surgical technique has mostly remained similar for all studies, the study methodology was variable. Most of the reports are retrospective, noncomparative series, while others have compared the outcomes of primary IOL implantation with the group receiving contact lens for visual rehabilitation. The number of patients in these series failed to provide the statistical power necessary to adequately assess the outcomes of IOL implantation. Four studies, depicted in Table 1, are prospective, and only one is a randomized trial comparing outcomes in the IOL with outcomes in a contact lens (CL) group. The reporting of results in the retrospective and nonrandomized studies could have been marred by fallacies of inadequate sample size and lack of standardized protocols.

The primary outcome measure in the majority of these studies has been the complication rate and overall safety profile of IOLs in young children. VAO has been the most frequent complication in all studies, but its incidence rate has differed between them. One glaring explanation could be the variable follow-up, as posterior capsular opacification (PCO) can have a delayed onset. Particularly, the use of acrylic foldable IOLs has been implicated with delayed and milder variety of PCO. At the start of the decade, many studies had some cases in which polymethyl-methacrylate (PMMA) IOLs were implanted. Research has conclusively established that IOL material and design has a direct influence on PCO rates. Acrylic foldable IOLs have been documented with lesser PCO rates and less severity of PCO compared to PMMA and silicone foldable IOLs. This trend has been corroborated by demonstrating increased biocompatibility and improved IOL design of acrylic foldable IOLs. Further, placement in-the-bag has been shown to be associated with lesser postoperative inflammation and PCO rates. Table 1 reveals that the information on type of fixation of IOLs, either in sulcus or bag, either has been missed or the exact number of cases with each type of fixation has not been mentioned. Moreover, the presence of associated ocular anomalies, like persistent fetal vasculature, in almost half of the studies could have influenced the VAO rates. Hence, the differential reporting of visual axis opacification and reoperation rates in various studies could partly be explained by the above-mentioned factors. It is worth noting that in the recent study by the Infant Aphakia Treatment Study (IATS) group, in spite of meticulous surgical protocols based on current knowledge, the reoperation rates in the IOL group were reasonably high. Possibly there are some
undetermined factors, particularly in infants, which may lead to visual axis opacification.

An attempt has been made to document the visual outcomes of primary IOL implantation in monocular aphakia in infancy, first by a pilot study and then by a randomized clinical trial by the IATS group. The results of the latter study, at 1 year of age, showed that there was no significant difference between the median visual acuity in the treated eyes with IOL and the CL group after cataract surgery during the first 6 months of life. This was in contrast to the pilot study, where visual outcomes in the IOL group were found to be better than the CL group. The median visual acuity in the treated eyes with IOL and the CL group after cataract surgery during the first 6 months of life.

Table 1. Studies on Primary IOL Implantation in Children Less Than 2 Years of Age: Methodology

<table>
<thead>
<tr>
<th>Authors, Year of Publication</th>
<th>Type of Study</th>
<th>Laterality of Cataract</th>
<th>Coexisting Ocular / Systemic Pathology</th>
<th>N (No. of eyes)</th>
<th>Age Group (Weeks)</th>
<th>Mean Follow-up (months)</th>
<th>Type of IOL</th>
<th>IOL Power Calculation Formula</th>
<th>Type of IOL Fixation</th>
</tr>
</thead>
<tbody>
<tr>
<td>Lambert SR et al, 1999&lt;sup&gt;6&lt;/sup&gt;</td>
<td>Prospective, nonrandomized</td>
<td>UL</td>
<td>Excluded</td>
<td>11</td>
<td>2-22</td>
<td>13 ± 6</td>
<td>PMMA (5) / AcrySof (6)</td>
<td>Hollay SRK II or I</td>
<td>Bag</td>
</tr>
<tr>
<td>Lambert SR, 2001&lt;sup&gt;1&lt;/sup&gt;</td>
<td>Prospective, nonrandomized</td>
<td>UL</td>
<td>Excluded</td>
<td>12</td>
<td>3-22</td>
<td>-</td>
<td>PMMA (6) / AcrySof (6)</td>
<td>Hollay I or SRK II</td>
<td>Bag</td>
</tr>
<tr>
<td>Plager D et al, 2002&lt;sup&gt;5&lt;/sup&gt;</td>
<td>Retrospective, comparative</td>
<td>UL, BL</td>
<td>Included</td>
<td>15</td>
<td>3-20</td>
<td>NA</td>
<td>AcrySof 3-piece</td>
<td>SRK II or SRK-T</td>
<td>Bag</td>
</tr>
<tr>
<td>Trivedi RH, 2004</td>
<td>Retrospective</td>
<td>UL, BL</td>
<td>Included</td>
<td>29</td>
<td>0.8-43.2</td>
<td>33.4 ± 16.1</td>
<td>AcrySof single piece</td>
<td>Hollay</td>
<td>Bag (86.2%) / Sulcus (13.6%)</td>
</tr>
<tr>
<td>Lundvall A, 2006&lt;sup&gt;8&lt;/sup&gt;</td>
<td>Retrospective</td>
<td>UL</td>
<td>Included</td>
<td>28</td>
<td>1-40</td>
<td>36</td>
<td>HSM-PMMA, AcrySof</td>
<td>NA</td>
<td>Bag</td>
</tr>
<tr>
<td>Ashworth JL&lt;sup&gt;9&lt;/sup&gt;, 2007</td>
<td>Retrospective</td>
<td>UL, BL</td>
<td>Included</td>
<td>25</td>
<td>1-51</td>
<td>44.36</td>
<td>NA</td>
<td>SRK-T</td>
<td>NA</td>
</tr>
<tr>
<td>Ram J et al, 2007&lt;sup&gt;10&lt;/sup&gt;</td>
<td>Prospective, nonrandomized</td>
<td>UL, BL</td>
<td>Excluded</td>
<td>45</td>
<td>3-104</td>
<td>18 ± 9.3</td>
<td>HSM-PMMA</td>
<td>NA</td>
<td>Bag, Sulcus</td>
</tr>
<tr>
<td>IATS, 2010&lt;sup&gt;11&lt;/sup&gt;</td>
<td>Prospective, randomized, comparative</td>
<td>UL</td>
<td>Excluded</td>
<td>57</td>
<td>4-24</td>
<td>12</td>
<td>AcrySof</td>
<td>Hollay I</td>
<td>Bag (93%) / Sulcus (7%)</td>
</tr>
<tr>
<td>Gupta A et al, 2011&lt;sup&gt;12&lt;/sup&gt;</td>
<td>Retrospective, noncomparative</td>
<td>UL, BL</td>
<td>Excluded</td>
<td>120</td>
<td>1-23 months</td>
<td>8.856 ± 7.73</td>
<td>PMMA (30)/ AcrySof (90)</td>
<td>SRK II</td>
<td>Bag, Sulcus</td>
</tr>
</tbody>
</table>

Abbreviations: UL indicates unilateral; BL, bilateral.

IOL Power Calculation and Myopic Shift

Other issues that have received attention in the literature are the need for accuracy in postoperative target refraction and a trend of emmetropization of refractive errors.

The need for pediatric IOL calculation formulae has been felt for a long time, primarily because all available formulas were derived from considerations regarding the adult eye. Unlike adults, pediatric eyes undergo rapid growth and significant refractive changes in the early years. Moreover, in most pediatric cases, the desired postoperative refraction is different from plano. All these factors add to the complexity of the IOL power calculations in children.

Various IOL formulae designed for adult eyes are being used to predict IOL power in pediatric eyes, which have shown varying degrees of accuracy. Neely et al found that the SRK II regression formula gave the least amount of variability, whereas the Hoffer Q gave the greatest amount, particularly among the youngest group of children with axial length (AL) < 19 mm. In a recent study, Nihalini et al concluded for eyes with significant deviation in prediction error (> 0.5 D) that there was usually an undercorrection, except with Hoffer Q, which was almost as likely to overcorrect as undercorrect. This may be explained by the higher proportion of short eyes in their series (AL < 22 mm; 69 eyes), and because Hoffer Q was formulated for shorter eyes.

A potential source of error in IOL power selection in children is inaccuracy of AL and/or keratometry power measurements. Most studies have used either anaplanation or immersion technique for AL calculation under general anesthesia.
Table 2. Studies on Primary IOL Implantation in Children Less Than 2 Years of Age: Results

<table>
<thead>
<tr>
<th>Authors, Year of Publication</th>
<th>Outcome Measures</th>
<th>Conclusion</th>
<th>Reoperation Rates</th>
</tr>
</thead>
<tbody>
<tr>
<td>Lambert SR et al, 1999</td>
<td>Adverse events, myopic shift</td>
<td>Increased adverse events, large myopic shift</td>
<td>72%</td>
</tr>
<tr>
<td>Lambert SR, 2001</td>
<td>Grating visual acuity at conclusion of study</td>
<td>Improved visual outcome compared to CL</td>
<td>83%</td>
</tr>
<tr>
<td>Plager D et al, 2002</td>
<td>Complication rate</td>
<td>VAO rate = 80%; higher reoperation rates</td>
<td>80%</td>
</tr>
<tr>
<td>Trivedi RH, 2004</td>
<td>Visual axis opacification (VAO)</td>
<td>VAO rate = 37.9%, greater VAO with associated ocular anomalies</td>
<td>37.9%</td>
</tr>
<tr>
<td>Lundvall A, 2006</td>
<td>Complications and visual results</td>
<td>VAO rate = 67%; better visual outcomes in BL cases compared to UL cases</td>
<td>70%</td>
</tr>
<tr>
<td>Ashworth JL, 2007</td>
<td>Refractive outcome</td>
<td>Satisfactory mean refractive outcomes, but wide range of errors</td>
<td>-</td>
</tr>
<tr>
<td>Ram J et al, 2007</td>
<td>Safety profile of IOLs</td>
<td>VAO rate = 13.3%; no major refractive surprises</td>
<td>28%</td>
</tr>
<tr>
<td>IATS, 2010</td>
<td>Grating visual acuity at 1 year of age, adverse events</td>
<td>Similar visual outcome in IOL and CL; VAO rate 72%; higher reoperation rates in IOL group</td>
<td>63%</td>
</tr>
<tr>
<td>Gupta A et al, 2011</td>
<td>Safety profile of IOLs</td>
<td>VAO rate = 6.7%; adverse events comparable in age group &lt; 6 months and beyond</td>
<td>NA</td>
</tr>
</tbody>
</table>

Abbreviations: CL indicates contact lens; UL, unilateral; BL, bilateral.

Applanation technique is thought to artificially reduce the AL and can contribute to the significant amount of additional error, especially for high-powered IOLs. Additionally, most studies except one failed to mention whether they tried to minimize the interobserver variability, by deploying single observers for biometry and refractive error measurements.

With the trend toward implanting IOLs in infants with shorter ALs (see Table 1), there is a felt need to understand the accuracy and the differences between prediction formulas at the lower extremes of AL and higher keratometry values. While in a few studies the age stratification of younger children was not done, in others an attempt was made to dichotomize children according to the age at the time of surgery. As most of the ocular growth occurs during the first 2 years of life, most studies have taken the same as the cutoff for grouping children. In two major studies, the number of eyes in < 2 year age group constituted about one-fifth of the total, the percentages being 22.7% in the study by Neely et al and 16.2% in the study by Nihalini BR et al. In other studies, the cutoff for “younger children” varies between 12 months, 18 months, and 36 months. This disparity and the smaller number of cases limits our understanding about refractive outcomes in the younger, especially the infantile, age group. However, these studies concur that there is a greater variability of refractive outcomes following cataract surgery in younger children compared to older children. In the work by Eibschitz et al, an analytical comparison of predicted implant power using in the pediatric range of AL and keratometry values was performed. Significant differences in IOL power prediction were found among the Hoffer Q, Holladay I, and SRK II formulas. This explains the higher degree of prediction error that was documented in the age group < 2 years in 2 recent studies.

It has been reported there is a trend toward larger postoperative prediction errors in younger children, compared to adults. The refractive outcome of pediatric patients depends on the assumed anterior chamber depth (ACD), effective lens position of the IOL, and the effects of axial displacement of the IOL. The effect is even more pronounced in pediatric eyes, as these short eyes require higher-power IOLs. The standardized assumptions about ACD and vaulting characteristics of an IOL within the bag may not be accurate for pediatric eyes owing to shallow anterior chambers and the particular postoperative dynamics of posterior capsule contraction, vitreous pressure, haptic angulation, effects of primary posterior capsulotomy and vitrectomy on lens position, and, later, the sometimes significant reproliferation of lens material. So it is imperative that studies evaluating IOL calculation formulae in children should include information on these parameters so as facilitate better comparison between them.

It has been documented that axial elongation of the globe in a pseudophakic eye leads to myopic shift, akin to what happens in normally developing eyes. Younger children are more prone to have larger and more unpredictable myopic shifts. Further, the myopic shifts have been shown to be variable, and no consistent correlations with preoperative axial length or IOL power have been found. One basic discrepancy in the reporting of myopic shifts in various studies has been the variable timing of the initial refraction and the length of the follow-up period. The initial refraction may vary from 2 weeks to 12 weeks, and the mean follow-up has ranged from 1 to 7 years. This has led to disagreement on the amount of myopic shift in different, largely retrospective studies. Further, the heterogeneity of study groups in terms of age groups and number of eyes included may be a determining factor.
It has been reported that pseudophakic eyes have tendency to grow longer than the fellow phakic eyes.\textsuperscript{13,17} Interestingly, in pseudophakic eyes, apart from the normal process of axial elongation with age, there can possibly be factors like secondary glaucoma or amblyopic visual deprivation that could influence the amount of myopic shift. Further, there could be other undetermined reasons, including genetic factors, that could contribute to the myopic shift. In the study by Greiner et al, the axial elongation in pseudophakic eyes was reduced in comparison to the phakic eye.\textsuperscript{18} It was worth noting that all eyes in this study had sulcus fixation of IOLs, compared to in-the-bag implantation in most other studies. However, the exact mechanism of sulcus fixation contributing to the myopic shift needs to be evaluated. Further, an interesting proposition was made by Nischal et al, who wondered whether the reduction of peripheral visual input by anterior capsular fibrosis in pseudophakic eyes might explain the refractive and axial length change following pediatric cataract surgery.\textsuperscript{17} It is possible that with improved understanding of IOL power calculation and prediction of postoperative refraction, improved results may be achieved in future prospective studies, in which children would be random-ized to grow longer than the fellow phakic eyes.\textsuperscript{13,17}

Glaucoma

Secondary glaucoma is one of the most vexing problems after congenital cataract surgery. It can emerge years later and can jeopardize vision. Some earlier studies had studied children and eyes with a coexistence of conditions associated with cataracts that will also develop glaucoma, such as Lowe syndrome, aniridia, and trauma.\textsuperscript{19} However, more useful information could be attained only after studying “uncomplicated” cases of surgical aphakia.

There is a wide variation in the reported incidence of glaucoma after congenital cataract extraction. It ranges between 6% and 58.7% of children, depending on the population studied and the length of follow-up.\textsuperscript{19} Initial reports by Chandler, Phelps, and Arafat drew attention to this problem.\textsuperscript{20} Magnusson et al studied Swedish children born with cataracts and found a 12% incidence of aphakic glaucoma (mean follow-up: 9.6 years).\textsuperscript{21} This figure is probably more reliable since the study was conducted within the confines of a fixed geographic location with centralized data collection. In the IATS, preliminary results showed that glaucoma developed in 5% of eyes in the CL group and in 12% of eyes in the IOL group.\textsuperscript{11} But the follow-up was too short to derive a true incidence.

Various factors have been postulated to influence the risk of developing postoperative glaucoma, such as age at detection of cataract, age at cataract surgery, cataract surgery procedure, primary IOL implantation, significant postoperative uveitis, and microphthalmia.\textsuperscript{19,22} However, such information is available from reports of selected individual case series, which may be subject to bias and confounding. The data, procured from the British Congenital Cataract Study, was an attempt to derive results from population-based research.\textsuperscript{23} Its finding suggested that detection of cataract was the only significant factor associated with the development of glaucoma after surgery for congenital cataract. It is imperative to believe that early detection of cataract would directly translate to early age of cataract surgery.

Literature review points out to a bimodal pattern of onset for aphakic glaucoma.\textsuperscript{19,22} The first onset peak is noticed within the first weeks to months following cataract surgery. This early-onset glaucoma is frequently associated with pupillary block, shallowing of the anterior chamber, and angle closure. It is well known that smaller eyes and eyes with reduced corneal diameter are more prone to develop angle closure in the postoperative period.\textsuperscript{22,23} However, the unavailability of data about preoperative gonioscopic findings in cases undergoing pediatric cataract extraction limits our understanding about the pre-existing state of the angle and its predisposition to insult in the postoperative period. Residual lens material contributes to the development of glaucoma by forming Elschnig pearls that may cause pupillary block and induce inflammatory adhesions in the angle and at the pupil edge, which can cause pupillary block and angle closure.

It has been observed that sulcus-fixation of the IOL is a risk factor for the development of pupillary block.\textsuperscript{19,22} But it is notable that most studies reporting the incidence of glaucoma following pediatric primary IOL implantation have failed to specify the differentiation of in-the-bag vs. sulcus fixation.\textsuperscript{22,23} The possible mechanism of glaucoma can occur due to the pupillary block, leading to extensive synechial closure or the forward displacement of iris tissue by the haptics of a sulcus-fixed IOL, causing crowding of structures in the angle.

The onset of delayed-onset open-angle glaucoma may occur years following the cataract. Because of the late onset occurring 5 or more years following the cataract surgery, it is not likely that inflammation, use of postoperative corticosteroids, or any other portion of the cataract surgery is the cause of this problem. Asrani et al, in a meta-analysis of 377 eyes with primary lens implantation, found only 1 eye with open-angle glaucoma.\textsuperscript{24} They have suggested that when a primary IOL is used, there is a reduced incidence of delayed-onset glaucoma. However, it is worth noting that implants are infrequently placed in eyes with microcornea and the surgery in this series was relatively delayed (mean age at surgery: 5.06 years). Hence, two important risk factors, microcornea and early age of surgery, which could have influenced the results otherwise, were conspicuous by their absence.

Although it is clear that pediatric cataract surgery places the eye at risk for glaucoma, the exact mechanism remains elusive. The volume of the lens and its dynamic role in accommodation are more prominent in younger eyes. Surgical removal of the lens early in life can alter normal development of the filtration angle. Morphologic studies of developing eyes of children have shown that the angle recess of the iridocorneal angle is expected to move toward the periphery, exposing the scleral spur and the ciliary body band.\textsuperscript{25} It has been speculated that the absence of the lens early in life alters or causes an arrest in development of the filtration angle, or it may be a lack of accommodation and pull of the ciliary muscles on the trabecular meshwork that in some way permits the meshwork to become compact and dysfunctional.\textsuperscript{26} It may be that some chemical substance may diffuse from the posterior eye into the anterior chamber and change the facility of outflow of the eyes. Clearly, there are some undetermined genetic and mechanical factors that may contribute to the development of glaucoma after pediatric cataract surgery.

On the one hand, early cataract surgery has been advocated to prevent amblyopia, and on the other hand, the possibility of a higher complication rate after early cataract surgery looms large. This leads to the dilemma, for which there are no clear answers. At present, the clinician may be best advised to do a balancing act of performing early cataract surgery, avoiding the neonatal period, followed by a careful surveillance to detect postoperative glaucoma. The latter must continue for the long term. Probably future prospective studies, in which children would be random-ized to cataract surgery at different ages within the critical period, would enlighten us about the issue of optimum timing.
In summary, pediatric cataract surgical outcome is affected by multiple factors. Among them, surgical technique, IOL power calculation, postoperative myopic shift, and occurrence of glaucoma play major roles in the final visual outcome. There is a need to study these factors in a robust way to find out possible answers to many issues, which are still not clear. When we offer surgery for these children, all these factors have to be incorporated in our decision making, keeping central the best interest of the child.

References

The Ciliopathies: What Are They?

Eduardo José Gil Duarte Silva MD

Ciliopathies are a recently defined group of rare genetic disorders characterized by dysfunction of a hair-like cellular organelle—the primary cilium. Cilia are microtubule-based structures found on almost all vertebrate cells. They originate from a basal body, a modified centrosome, which is the organelle that forms the spindle poles during mitosis. The cilium-centrosome complex represents nature’s universal system for cellular interaction, cellular detection, and management of external signals.

Primary cilium-related dysfunction can either affect a single tissue or organ, or lead to a full-blown syndromic spectrum of ciliopathy-related manifestations with simultaneous involvement of several organs. The retina is a good example of a single-tissue ciliopathy. Primary cilium dysfunction frequently affects photoreceptors (ciliated retinal cells) and causes retinal degeneration. Mutations in retina-specific ciliary genes lead to isolated nonsyndromic retinitis pigmentosa (RP). These mutations include the most common form of X-linked retinitis pigmentosa, linked to the RPGR gene, or subtypes of autosomal recessive Leber congenital amaurosis (LCA) linked to NPHP6/CEP290 and LCA5/lebercilin.

The retinitis pigmentosa phenotype is a common feature of syndromic ciliopathies. These include Usher syndrome (RP plus sensory-neural deafness with/without vestibular involvement), Bardet-Biedl syndrome (BBS), MORM (Mental retardation, truncal obesity, Retinal dystrophy and Microopenis), Alström syndrome, Senior-Loken syndrome (SLS), Joubert syndrome-related diseases (JSRD), Jeune syndrome, and Meckel-Gruber syndrome (MKS).

Most syndromic ciliopathies are inherited in an autosomal recessive fashion. However, more complex inherited mechanisms (trialelism, modifier effect) have been described. Allelic variability, defined as different mutations in the same gene giving rise to different clinical presentations or syndromes, is common among syndromic ciliopathies. It is well established that NPHP6/CEP290 mutations may cause a pure retinal phenotype (isolated LCA) to the lethal multisystemic MKS.

The cardinal features of Bardet-Biedl syndrome (BBS) are retinal dystrophy (RD), obesity, polydactyly, hypogonadism, cognitive impairment, and renal failure. Secondary clinical features such as anosmia, diabetes, cardiac anomalies, liver fibrosis, brachydactyly, and Hirschsprung disease may also be present. Different types of retinal dystrophy have been reported in BBS. These are mainly a rod-cone dystrophy or a cone-rod dystrophy; however, a choroidal dystrophy and a global severe retinal dystrophy have also been described.

BBS is a genetically heterogeneous condition with 16 genes identified to date. These account for about 80% of the cases. All BBS genes have been related to cilium biogenesis and/or function. BBS1 and BBS10 are the two most common culprits. Interestingly, several BBS genes are implicated in other ciliopathies: BBS13 is MKS1 (Meckel Gruber syndrome 1) and BBS14 is the CEP290/NPHP6 gene associated with LCA, JS, and MKS. In contrast, retinal-specific splice variants of BBS3 and BBS8 have been identified and mutations in these transcripts cause nonsyndromic RP.

Alström syndrome manifestations include RP in early childhood, hearing disability, and metabolic defects leading to hyperinsulinemia, type II diabetes mellitus, and obesity in childhood. However, these patients lack polydactyly and cognitive impairment, commonly seen in BBS.

Senior-Loken syndrome is a combination of nephronophthisis (NPH) and retinal degeneration. NPH is characterized by normal kidney size, tubulointerstitial nephritis, and a loss of cortico-medullary differentiation leading to cyst formation. The first symptoms are often polyuria and polydipsia caused by a defect in urinary concentration. Three forms of NPH can be distinguished based on end-stage of renal failure: infantile, juvenile, and early-adulthood. The occurrence of the retinal dystrophy is higher in the juvenile form of NPH. To date, 11 genes (named NPHP1e11) are known to be causative of SLS.

Joubert syndrome is a combination of cognitive impairment, ataxia, tachypnea, and eye movement abnormalities. Cerebellar vermis hypoplasia is a pathognomonic finding on MRI named molar tooth sign. Multiple other features can be associated with this midbrain-hindbrain malformation, leading to the denomination of Joubert syndrome-related disorders (JSRD). The retinal phenotype includes macular colobomas and a rod-cone dystrophy.

Within the spectrum of syndromic ciliopathies, Meckel-Gruber syndrome represents the most severe end of the disease spectrum, leading to prenatal or perinatal mortality. It is characterized by occipital encephalocele, kidney cystic dysplasia, hepatic ductal proliferation, liver fibrosis, and polydactyly. Seven genes have been found to be implicated in MKS: MKS1, 2 and 3, CEP290, NPHP3, RPGRIP1L, and CC2D2A.

Retinal degeneration due to primary cilium structure disruption has been put under the spotlight through the multispecies ciliopathy studies that have enabled the identification and dissection of the signaling pathways involving the connecting cilium. The retinitis pigmentosa phenotype secondary to dysfunction of the primary cilium can be associated with several other clinical manifestations (syndromic RP) and impacts on clinical practice. It is important to be aware of the target organs for ciliopathies defining well-known and new overlapping syndromes. Therefore, a child with RP/CRD should be assessed for associated features such as obesity, kidney impairment, polydactyly (asking for removal of extra digits during childhood), cognitive impairment, tested for anosmia and diabetes, and checked for bone changes, as the diagnosis of a ciliopathy will influence follow-up.

Identification of ciliopathy genes has improved both our biological understanding of these conditions and genetic counseling. The challenge is to dissect the mechanisms that underlie this retinal degeneration subgroup. Pharmacological therapy aimed at preventing connecting cilium dysfunction is still hypothetical. Understanding the mechanisms underlying biogenesis and function of the photoreceptor connecting cilium is essential to identifying therapeutic strategies.
Selected Readings


Retinal Repair by Transplantation of Photoreceptor Precursors

Stem Cell Therapy for Retinal Disease: Future Prospects for the Clinic

Jane C Sowden PhD

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Summary

New treatments are needed for retinal diseases involving the death of photoreceptor cells. Remarkable progress is being made in the research laboratory toward the development of novel stem cell strategies for retinal repair. I will review recent advances in generating new retinal neurons from pluripotent stem cells in vitro and describe our work demonstrating the feasibility of photoreceptor transplantation in animal models. Challenges for the development of clinical cell-replacement therapies for retinal disease will be discussed.

Selected Readings

Radiation Exposure to Children from Medical Imaging: Is There a Problem?

Michael Callahan MD, Marilyn Goske MD

Introduction

Computerized tomography (CT) has been recognized as one of the major scientific advances in patient care. This remarkable technology enables the radiologist to diagnose medical disease and life-threatening illness faster and more precisely, obviating the need for emergency and exploratory surgeries. Pediatric patients have been shown to be triaged more appropriately and faster in the emergency room while lowering the cost of patient care. Yet it has been shown that children are often imaged using adult technique, resulting in radiation doses that are higher than necessary. This presentation will focus on the challenges in discussions of radiation and will discuss educational interventions that can be used to promote safe care.

Reasons for Increased Attention in the Use of Ionizing Radiation in Imaging in Children

Both the scientific and the lay press have paid increasing attention to the use of radiation in medical imaging, particularly in children, for several reasons: The number of CT scans in children has risen dramatically in the United States. CT scans can be performed in less than a second. Many of the newer scanners (“helical scanners”) allow acquisition of a volume of data that can be “reconstructed” in many different scan planes. The number of CT scans in the United States has risen from 3 million in 1980 to 62 million in 2006, with 4-7 million of these scans performed annually in children. While the radiation dose from a single CT scan is usually relatively low, some studies suggest that the radiation dose to pediatric patients may be higher than necessary. The dose from a single CT scan is significantly higher than x-rays. Compared to one day of background radiation, for example, a single-view chest x-ray is similar to one day of background radiation, while a head CT is similar to up to 8 months of background radiation.

Children are more vulnerable to changes from radiation.

Children are more susceptible than adults to changes in their cells from a given dose of radiation. The primary risks to children from a CT scan are the slight increased risk in developing cancer and changes to their genes over their lifetime. Children’s cells are growing rapidly. This puts them at greater risk to many types of cell injury. The organs of a child’s body that are more susceptible to changes from radiation are the lens of the eye, bone marrow, thyroid, breast and the ovaries and testes. Strategies to decrease dose to the lens of the eye include angling the gantry to exclude the lens in brain CT and lead shielding.) Children have more remaining years of life during which radiation-induced cancer could develop. If a child’s cells change as a result of a radiation exposure, it may take 10, 20, or even 30 years for the cancer to develop. Finally, we know from phantom or simulation studies that if a child has a CT scan using an adult technique, the child’s dose is greater. Since children have less subcutaneous tissues, their core doses are significantly greater than an adult’s dose when incorrect “adult-sized” techniques are used.

Potential risk is a complex topic to discuss with families.

Discussion of risk from ionizing radiation used in medical imaging is complex, as it relates to population statistics, assessing risk compared to benefit, and the fact that cancer occurs in about 40% of all people in the United States over the course of their lifetime. The risk of developing a fatal cancer is about 20% over the course of a lifetime in the United States. Therefore, separating out the number of cases that arose from a specific medical radiation event is extremely challenging.

Comparison of Imaging Tests to Background Radiation

- Natural background radiation: 3 mSv/yr
- Airline passenger (cross-country): 0.04 mSv
- Chest x-ray (single view): up to 0.01 mSv
- Chest x-ray (2 view): up to .1 mSv
- Head CT: up to 2 mSv
- Chest CT: up to 3 mSv
- Abdominal CT: up to 5 mSv

Use of Alternate Imaging Strategies That Do Not Use Ionizing Radiation

When considering any medical imaging, evaluating the benefit to the patient relative to any potential risks from performing the study is first and foremost. Alternate strategies may include clinical follow-up or other consultation ultrasound and MR that do not use ionizing radiation. However, there are some instances when CT is the only test that provides the specific information. CT of the lung for suspected metastatic disease is an example of this situation.

The Image Gently Campaign

Image Gently is an education and awareness campaign to promote radiation protection for children worldwide. Sponsored by the Alliance for Radiation Safety in Pediatric Imaging, a consortium of more than 55 groups that represents over 700,000 health care professionals, the campaign hopes to change practice locally. Our website, www.imagegently.org, provides additional information.

References


Video: Fishtail Sign in Posterior Lenticous
Pre-existing Posterior Capsule Defect

Abhay Raghukant Vasavada MBBS FRCS

This video will highlight the diagnostic signs of a pre-existing posterior capsule defect (PPCD). A significant proportion of infants and small children with congenital cataracts have a PPCD in the posterior capsule. In classic cases, the PPCD is hidden behind a seemingly routine pediatric cataract in an undilated pupil. The defect looks like a total, white cataract. Preoperative evaluation of such a cataract under maximum dilatation is mandatory to unveil the important diagnostic signs. PPCDs have a visible demarcation by their thick margins, mainly as a result of capsule fibrosis. When the globe is moved with forceps, the degenerated vitreous with white granules moves like a fishtail, which is pathognomonic of a PPCD. Specific staining of the vitreous reveals calcium accumulation in the chalky white granules around the defect. Often, it is possible to identify the thick margins of the PPCD despite the “total” cataract. The cataract may not be uniformly white on dilatation, the density of whiteness being greatest in the center, with the periphery being semitransparent.
Section VIII: Have You Thought About . . .

Current Management Strategies for Blepharokeratoconjunctivitis

Mark E Jacobson MD

I. Blepharokeratoconjunctivitis is an eyelid margin disease with secondary conjunctivitis or corneal involvement.

A. Divided into anterior and posterior disease
   1. Anterior disease includes inflamed eyelids with debris and/or telangiectic vessels around cilia.
   2. Posterior disease includes inflammation and/or glandular debris around the meibomian orifices.

B. Not uncommon, 2 large centers found it accounted for 12%-15% of their referrals.1,2

C. Mean age of symptoms at age 4 and mean age of referral for treatment at age 6 (range: 7 months to 16 years).1-3

D. Symptoms noted
   1. Conjunctival erythema
   2. Lid margin erythema
   3. Eyelash anomalies
      a. Blepharitis
      b. Madarosis
      c. Styes
   4. Epiphora
   5. Photophobia
   6. Recurrent chalazion1,3,4

E. Clinical findings
   1. As noted in D above.
   2. Meibomian gland dysfunction
   3. Corneal/limbal changes
      a. Mild punctuate epithelial erosions to limbal phylctenules
      b. More severe: Corneal opacity, corneal scarring, or vascularization1-4

II. My Treatment of Blepharokeratoconjunctivitis

   Treatment regimen may vary, but all include 2 mainstays:
   A. Hot compresses
   B. Flaxseed oil

III. Hot Compress

   Method and duration most important:
   A. Use washcloth and small soft hot/cold gel pack.
   B. Preheat gel pack in hot water.
   C. Once hot, wring out washcloth in hot water.
   D. Wrap washcloth around hot gel pack and check on wrist to ensure it is not too hot.
   E. Place on eyes until cooled (8-plus minutes).
   F. Once removed, children will have a red flush (raccoon mask).
   G. Ingenuity and parent persistence are required.
      2. Listen to music or a book or tape.
      3. Parent read from a “chapter book” (ie, no pictures).
      4. Compress one eye at a time while watching favorite video (often the best way, especially if treating a chalazion and doing this 4 to 6 times a day).
      5. Do in the bathtub.
   H. Do every night at bedtime for 6-8 weeks until follow-up appointment. Watch skin for chapping or drying and apply facial moisturizer if noted.

IV. Flaxseed Oil

   A. Dosage varies by weight: About 1 teaspoon per 33 pounds up to a maximum of 1 tablespoon if 100-plus pounds.
   B. Flaxseed oil (α linolenic acid) is dietary source of omega-3 essential fatty acids.
   C. Two studies in women with omega-3 dietary supplement
      1. Improved dry eye symptoms
      2. Changed the meibomian glands secreted lipid profile5,6
   D. My personal experience mimics that reported by Kronemyer.7 Regular dietary intake of flaxseed oil thins and clears the meibomian gland secretions over time.
   E. Adding flaxseed oil to child’s diet:
      1. Mix in smoothie or yogurt.
      2. Mix in already cooked food to mask flavor and texture (parental experimentation). Note: Cooking flaxseed oil breaks down its nutritional benefit.
      3. Mix in old-fashioned peanut butter if child likes and will eat it.
         a. Measure and pour off peanut oil on top and replace with equal amount of flax seed oil.
b. A PB&J sandwich daily provides intake.
c. Note: You must refrigerate this peanut butter, as the flaxseed oil will become rancid.

V. Getting Parents to Buy Into Regimen
A. Explanation and understanding of purpose
B. Demonstration by digitally expressing the meibomian gland content and showing parent and child.
   1. Description of normal oil as thin clear olive oil.
   2. Showing thick, white, often wax-like substance that was expressed allows parent to understand:
      a. The need for prolonged hot compress to melt wax.
      b. The need for adding the gland’s dietary fuel to help it make new clear oil.
C. This treatment of blepharokeratoconjunctivitis by clearing out the meibomian glands (although done in a different manner) by Beauchamp et al in 1981 in his Case 1 was shown effective. The reference citings, however, only focus on Case 2 with its use of erythromycin.8

VI. Additional Arms of Treatment: Steroid Use
A. Limited to perilimbal vessels or corneal infiltrates
   1. My concern is exasperation with increased release of antigenic toxins with hot compresses.
   2. Generally use an antibiotic-steroid combination drop 3 to 4 times a day for 2 weeks then at bedtime after compress for 2 more weeks.
   3. Literature notes desire to use least amount in dosage and concentration through a tapering regimen.1-3,9
   4. For most cases steroid use is avoided because it quickly masks symptoms and parents discontinue the real treatment of heat and flaxseed oil.
B. When steroids are not used, always forewarn parents of “storm before the quiet,” noting that eyes will often look worse right after hot compress and release of toxin-filled debris.
C. Erythromycin ointment at bedtime
   1. Use with mild punctate epithelial erosions or other surface irritation.
   2. Use in cases of blepharitis.

VII. Follow-up
A. See patient back in 6-8 weeks.
   1. On initial visit always note to parent that this regimen does not always work and if no improvement noted may need systemic antibiotics.
   2. Review clinical signs and symptoms with parent and patient.
   3. Perform re-expression of the meibomian gland, if possible.
   4. If improvement noted, discuss continuing flaxseed oil.
   5. Taper the hot compresses to once or twice a week, depending on how clear and thin the oil is.
B. No improvement or desire to follow current controlled study literature
   1. Discussion centers on continuing present regimen and adding antibiotic.
   2. Erythromycin or doxycycline dependent on age and state of dentition.
   3. Erythromycin is given 15-25 mg/kg 2 times a day for 6 to 8 weeks (in literature may be longer).2,4,9,10
   4. Follow-up
      a. Signs and symptoms improved: Discontinue antibiotics, continue flaxseed oil, taper compresses, and review in 6-8 weeks.
      b. No change is noted: The oral course is repeated and recheck in 6-8 weeks.

VIII. Conclusion
A. Over the past several years, systemic antibiotics have rarely been needed.
   1. Occasionally a mild steroid like fluorometholone is needed for a short 2 times a day course while a nightly regimen of hot compress is reinstated.2
   2. Occasionally patient will have relapse when flaxseed oil/compresses have been discontinued. Reinstating both usually clears the eye.
B. Mainstay of treatment in my mind is omega-3/flaxseed oil.
   1. Essential need in developing infants is demonstrated by inclusion in baby formulas and recent studies.11,12
   2. Young children have no less need in their development but with lack of omega-3 included in packaged processed food the need for external supplements becomes apparent.

References


Evaluation of the Non-seeing Infant

Shira L Robbins MD

I. Identifying Poor Visual Behavior
   A. What is normal?
   B. Cerebral immaturity, foveal immaturity and lack of ciliary body control lead to hazy and intermittent vision, better in dim light.
   C. This typical poor newborn vision prevents sensory overload until the neurologic system is more mature.
   D. Visual acuity greatly improves several months after birth.
   E. Eye movement coordination develops by 3-4 months.
   F. Fine depth perception develops at 3-5 months (requires good eye movement coordination and adequate retinal nerve cell maturity).

II. Medical History (Development/Systemic Disease)

III. Family History (Ocular and Systemic)

IV. Physical Examination
   A. Exam style
      1. Talk to the infant before touching.
      2. Watch infant’s body response—not just eyes/visual response.
      3. No abrupt hand movements near or on infant as no visual cues to warn patient
   B. Parts of the physical exam

V. Additional Testing
   A. Preferential looking
   B. Vestibulo-ocular reflex
   C. Unequal nystagmus test
   D. Visual evoked potential
   E. Electroretinogram
   F. MRI
   G. EEG
   H. Eye movement recordings
   I. Pediatric neurology consult
   J. Metabolic workup
   K. Genetic workup

VI. Diagnosis: Ocular, Cerebral, or Both

Figure 1. Reprinted with kind permission from the author and Springer Science+Business Media: Pediatric Neuro-Ophthalmology, The Apparently Blind Infant, 2010, 2, Brodsky MC, Fig 1.1.

Fig. 1.1 Flow chart depicts simplified diagnostic algorithm to be used in evaluation of the apparently blind infant
Section VIII: Have You Thought About . . .

VII. Treatment

A. Philosophy of treatment: Patient is a baby/child first who happens to be non-seeing.

B. Medical: Diagnosis, prognosis

C. Psychiatric counseling
   1. Grieving process of parents
   2. Eventual grieving process of patient
   3. Advocacy
   4. Accommodations

D. Educational
   1. Braille learning readiness, similar to reading or kindergarten readiness: Braille learners need fine motor skills, tactile sensitivity, ability to recognize small shapes, identify difference between rough and smooth, follow a line with their finger across a page.
   2. Formal non-sighted education includes listening skills, orientation and mobility, daily living skills, art, physical education, and music, among others.
   3. Developmental associations
      a. Delay gross motor → need to teach jumping, hopping, and running
      b. Delay body image
      c. Delay sensory integration → need increased sensory exploration, especially tactile, and encourage to use any vision patient has
      d. Decreased imaginative play → can be healthy to be around sighted peers to encourage this

E. Communication assistance: Vision is not main communication or learning mode. Sign language and Braille can be positively framed as learning a foreign language. Braille can lead to faster reading efficiency instead of magnifying small portions of a page with assistive devices.

F. Local community resources

G. Global resources

Selected Readings


Additional Resource for Parents

Update on Oculoplastics

Don O Kikkawa MD

Eyelid/Face

The developing child undergoes dramatic transformation that includes both volumetric and 3-dimensional growth. Many anomalies that affect the pediatric populations arrest development, leading to deformity and functional deficit.

It is well recognized that one’s facial volume is full as a child and that with growth, certain areas of the face lose volume. Identical twin studies have shown that, in general, the heavier twin appears to be older in age until a certain age is reached (middle age). At that point, the heavier twin appears younger. Loss of facial volume is a noticeable characteristic in advanced age. The trend in aesthetic rejuvenation is volume replenishment.

This concept has been translated to the pediatric population. Craniofacial anomalies—in particular, clefting syndromes—lead to characteristic deficiencies in bone and soft tissue. Much of the previous reconstructive efforts have focused on rebuilding the facial skeleton, lacking emphasis on soft tissues. Examples will be shown where use of soft tissue augmentation leads to fully restored facial volume.

Vascular tumors continue to be a management challenge. Pharmacologic advances, such as propranolol use for capillary hemangioma, are proving to be first-line therapy. Surgical excision, however, remains an option for recalcitrant cases.

Orbital

The relationship of exophthalmos and ocular motility is nowhere better examined than in thyroid eye disease (TED). In TED, ocular motility may be limited due to tight restricted muscles. This relationship worsens when the eye is proptotic because this further worsens the situation by putting the already tight muscle on stretch. In such cases orbital decompression can dramatically improve ocular motility. In TED patients with large angle deviation, orbital decompression should be considered in preparation for strabismus surgery.

Restriction of ocular motility can also be found in previously operated eyes due to adhesions between the muscles and surrounding orbital tissues. The ability to alter the tissue biology of wound healing is often essential to optimizing the outcomes in difficult reoperations. The use of antimetabolite therapy intraoperatively and in the clinic setting can be very useful to prevent further scarring in these difficult cases.

Lacrimal

Small incision or endoscopic procedures are favored in lacrimal surgery because of faster healing times and minimized scarring. Endoscopic lacrimal procedures in the pediatric population are often challenging due to the size of the instrumentation and the nasal passage in children. In addition, permanent bypass tubes can be problematic in both children and adults. In the properly selected child, however, permanent bypass tubes are well tolerated and effective.

Selected Readings

Strabismus is a visible facial abnormality that has been shown to be associated with many adverse psychosocial consequences in adults. Adults often experience difficulties finding an employment and problems finding a partner. In children, it has been demonstrated that visible differences in general and also strabismus have a negative impact on how they are perceived. Even a negative social bias of teachers against schoolchildren with strabismus has been reported.

Unfortunately, an increasing number of health care insurances try to refuse reimbursement for strabismus surgery procedures despite the large body of literature showing that visible strabismus, even if there is no prospect for binocular vision, cannot be judged to be cosmetic.

In children, surgery should be performed before negative attitudes toward strabismus emerge. Until recently, it was unclear when such an attitude emerges. A recent study looking at which children are invited by peers to birthday parties showed that a negative attitude emerges at approximately 6 years. Children older than 6 years selected children with strabismus significantly less frequently than those with aligned eyes. Although until 6 years no preference for children with straight eyes was shown, a large percentage of children between 4 and 6 years of age already noted that something was strange with the eyes. The results show that corrective surgery for strabismus without prospects for binocular vision should be performed before the age of 6 years, when negative social attitudes may arise. The differences in the perception of strabismus between age groups can be explained by studies on the development of children in the recognition of facial features. These studies show that children older than 6 years of age mostly process faces holistically, like adults. Children younger than 4 years use part-based processing, meaning that faces are categorized in terms of piecemeal characteristics. Therefore, younger children may not be able to realize that two eyes are not straight.

What is the role of glasses on the perception of visible strabismus in children? A high percentage of children with strabismus wear glasses. Can glasses distract from visible strabismus? The results of studies about this topic are controversial. Terry and Stockton report that glasses have a negative effect; Walline et al report that they make children look smarter and more honest.

References


The Patient Has Thyroid Ophthalmopathy
Battle of the Bulge: Experience of the UCSD Thyroid Eye Center

David B Granet MD

I. Thyroid Eye Center
   In 1997 UCSD Multidisciplinary Thyroid Eye Center formed, combining Oculoplastics, Strabismus Specialist, Neuro-ophthalmologist, Fellows and Residents.
   The center improves care via improved communication, coordinated care, making treatment easier for patient and improving process/compliance, and research.

II. Graves Hyperthyroid
   A. Almost all have some eye signs.
   B. Half are clinically evident.

III. Thyroid Eye Disease: Clinical Spectrum of Disease
   A. Type I: Fat infiltration
   B. Type II: Extraocular muscle involvement

IV. Overall Approach: Five-Step Plan
   A. Medical treatment
   B. Botox / prism if needed
   C. Orbit (unless neuropathy)
   D. Strabismus repair
   E. Lid repair

V. Medical RX
   Lubricants, topical decongestants/anti-histamines/plugs, prisms, steroids (oral, IV, retrobulbar), Botulinum toxin, XRT (ORGO study)

VI. What have we learned?
   A. Depression study; 2 groups
      1. Severe/moderate eye signs vs. Mild/none
      2. POMS survey (profile of Mood States)
      3. Significant depression compared to non-ophthalmic Graves
      4. More associated with disfigurement than diplopia
      5. Psychological burden should be considered in planning treatment and prompt referral to mental health professionals.
   B. Orbital decompression (bony)
      1. Diplopia new onset: 10%
      2. Botulinum toxin A as an adjunct to decompression
      3. Intramuscular injection under direct visualization
      4. 10 to 15 units Botulinum A toxin
      5. Medial and/or inferior rectus
      6. Adjunct to orbital decompression
      7. Type II patients with large muscles or strabismus in primary gaze undergoing decompression
   C. Botulinum toxin A role in restrictive strabismus
      1. IOP effects, preop, intraoperative
      2. Dose 5-15 units per muscle
      3. Multiple injections per muscle
      4. IOP in injection patients
      5. Decrease in IOP on upgaze
      6. Primary gaze IOP also dropped
      7. Sustained after 3 months
   D. Strabismus surgery
      1. Primary position and reading position
      2. Our guidelines
         a. Undercorrect vertical: Aim for below primary
         b. Assymmetric surgery, recessions
         c. Use tendon shifts
      3. EOM surgery
         a. Defer until stabilizes after decompression
         b. Usually requires inferior rectus, medial rectus recession on adjustable
         c. ± Botulinum toxin
         d. Don’t forget lid issues

VII. Thyroid Strabismus Patterns: AAPOS 2005
   A. 45%: Inferior rectus OA pattern
   B. 22.5%: Superior rectus OA pattern
   C. 22%: Inferior oblique OA pattern
   D. 9.68%: Superior oblique OA pattern

VIII. Patient Education
   A. Use of adjustable sutures
   B. Delay adjustment
   C. Approach oblique muscles
   D. Other center approaches . . .
      1. May worsen proptosis
      2. May worsen lid position
E. Lower lid retraction repair

1. AAO 2003

2. For patients that go through the entire process, 33 months until finished from Dx, 9 months from start to finish for repair, RAI and thyroid orbitopathy

3. RAI and thyroid eye disease
I. First: Sort out if it is a paresis or paralysis.
   To help in the differential diagnosis of a paralysis vs. possible residual function of the lateral rectus, details of history can be useful and essential diagnostic maneuvers are essential.
   A. Cause
      1. Neurosurgery vs. closed head trauma
      2. Vascular insufficiency vs. progressive tumor-related compression
   B. Chronicity and delays
      1. Cause (see above)
      2. Stability of maximum angle: without any recovery
      3. Spread of commitance
   C. Full orthoptic assessment
      1. Motility
         a. Measures of the estotropia (ET): Lateral incomitancy, etc.
         b. Secondary deviations: Hess, Lees, Harms screens
      2. Fusion
         a. Confirm potential
         b. Eliminate motility barriers (eg, torsions – haploscope)
      3. Single binocular field
         a. Patient friendly
         b. Preop: Assessment, patient education, setting goals
         c. Postop: Results, planning
   D. Essential diagnostic tests
      1. Saccades by clinical optokinetic nystagmus or velocity recordings (essential): Limited by clinical experience, contractures, equipment
      2. Generated forces: Requires patient cooperation ++
      3. Forced ductions
         a. Useful in cases of small saccades amplitude
         b. Important to rule out contractures
II. Second: Complicating Factors
   A. Medial rectus contracture (with or without conjunctival shrinkage): Will demand a different surgical approach
   B. Other cranial nerve involvement (eg, CN IV): Will require additional surgical consideration (eg, IV in head trauma cases and hypertropia)
   C. Loss of fusional ability (brain injury): Free space and/or haploscope
   D. Patient expectations
      1. Field of single binocular vision
      2. Number of surgeries
      3. Motility vs. visual ultimate expectations
III. Third: Setting Up a Plan
   A. Three principles of Buckley
      1. Improve ocular rotation
      2. Balance yokes
      3. Anticipate problems
   B. Paresis = Recess-resect (RR); can be asymmetrical or large numbers
   C. Contracture = Release (Botox and timing vs. recession)
   D. Paralysis = Transposition(s)
   E. Other cranial nerves = Other muscles, other eye
   F. Larger field of binocular single vision = Balance yokes
      1. Weaken contralateral agonist (medial rectus [MR])
      2. . . . Weaken contralateral antagonist (lateral rectus [LR])
      3. Posterior fixation in small primary position deviations
   G. Good results in general
IV. The Interventions: Timing and Type
   A. Botox and contracture: Prevention vs. treatment
   B. 6-12 months of absolute stability. Unless . . .
   C. RR
      1. Complete recovery and residual ET = RR
      2. More often partial recovery = More resection
      3. Large chronic angle but good function = Large RR
         a. Watch for resulting lateral incomitancy
         b. Conjunctival recession
D. Transpositions
1. Full tendon: Knapp with or without Foster sutures
2. Partial tendon:
   a. Preserve one-half of vascular supply from the muscles
   b. Hummelsheim with or without Foster sutures
   c. Hummelsheim-Kaufmann
3. In all = Watch for tight inferior rectus and resulting hypotropia (HoT)

E. Prisms, and additional surgeries
1. Prisms: Small residual horizontal or vertical
2. Surgery on bigger vertical or missed contralateral CN IV
3. Surgery: Residual ET or increase binocular field – contralateral MR
   a. Recession
   b. Retro-equatorial myopexy (posterior fixation)
      i. Scleral
      ii. Pulleys

V. Complications
A. Anterior segment ischemia
1. Rare if only 2 muscles operated
2. Advantage of Botox on the homo-lateral antagonist MR
3. Advantage of partial transposition with MR surgery

B. Undercorrection
1. With misdiagnosis of complete paralysis
2. In untreated contracture of the homo-lateral antagonist (MR)
3. In partial transposition . . . role of Foster sutures/Kaufmann technique

C. Overcorrection
1. Most cases, short lived . . . adjustable?
2.Reported with MR recession/Botox when no contracture
3. Restriction of antagonist (MR) rotation (exotropia in opposite gaze)
4. Avoid excessive MR recession. Instead: Posterior fixation of contralateral agonist
5. Occurs with late recovery of the LR (hasty surgery)

D. Vertical deviation
1. Inferior rectus and Lockwood complex are too tight
2. Undiagnosed CN IV or partial/partially recovered CN III
3. Asymmetrical full transposition
4. Importance of technique: Forced and spring balance tests
5. Possible advantage of some form of adjustable . . .

Selected Readings
The Patient Has Adult-Onset CN III Palsy

David K Coats MD

I. Abstract

The treatment of a severe paresis or palsy of the third cranial nerve with onset after visual maturation is complex. Patients are almost always motivated to have surgical correction and frequently, if not usually, have unrealistic expectations as to their prognosis. This talk will provide an overview of 3 key components of the management of an adult with a third cranial nerve palsy: (1) the complex decision to recommend surgery or not, (2) surgical treatment options and expectations, (3) long-term follow-up needs.

II. Approach to the Patient Prior to Surgery (Assuming Etiology Is Known and Spontaneous Resolution Is Not Anticipated)

A. Is this patient a surgical candidate?
   1. Attitude and expectations of the patient
   2. Concurrent problems

B. Preoperative patient discussion
   1. Alignment objectives
   2. Diplopia
   3. Corneal exposure
   4. Residual ptosis
   5. Other potential complications
   6. Timing of ptosis surgery

III. Surgical Treatment Options

A. Traditional muscle surgery
   1. Large recess/resect operation
   2. Transposition procedures
   3. Mechanical fixation

B. Anticipated outcome
   1. Residual strabismus
   2. Duction limitations
   3. Abnormal head posture
   4. Residual ptosis

IV. Long-term Follow-up Needs

A. Strabismus
   1. Progressive recurrence possible
   2. Additional surgery may be warranted.

B. Anterior segment
   1. Corneal exposure concerns
   2. Protection of the cornea
You Are Faced With Partially Accommodative Esotropia ± High Accommodative Convergence-to-Accommodation Ratio

Monte A Del Monte MD

I. Treatment of Accommodative Esotropia (ET)

Full hypermetropic correction as soon as possible:
A. To straighten eyes by relaxing the overconvergence caused by accommodating
B. Delay can result in loss of binocular potential.
C. Child must wear optical correction full time.
D. Atropine relaxation: Short course of cycloplegia (atropine 1% O.U. x 7 days) if patient does not accept hypermetropic glasses
E. Continue full correction if:
   1. Eyes straightened to within 8 prism diopeters (PD) at distance and near
   2. Binocular vision and stereo present

F. After 2 to 4 weeks, surgery indicated if:
   1. Good compliance with glasses
   2. Residual esotropia of more than 10 PD for distance or near (with bifocal)
   3. No fusion or stereopsis with glasses

II. Surgical Dose Controversial

A. Bilateral medial rectus (MR) recession: procedure of choice
B. Controversy: Surgical dose
C. Standard surgery: Surgical tables
D. Residual deviation at distance after full hypermetropic correction
E. Result: Undercorrection (42%-65%)
F. Augmented surgery: Increase amount of surgery
   1. Parks: Add 1 mm to recession of each MR if high accommodative convergence-to-accommodation (AC/A) ratio
   2. Wright: Average of Ncc (near with correction) and Nsc (near without correction); or average of Nsc and Dcc (distance with correction) if AC/A ratio high
   3. Kushner, West and Repka: Use near deviation with correction

III. New Surgical Protocol

A. New surgical protocol for augmented surgery based on the average of Ncc and Dsc (distance deviation without correction). Used by author from 1985 to the present.

B. Rationale—Must correct:
   1. > Ncc: Prefer overcorrection, so can treat with decrease plus/remove glasses
   2. < Dsc: To reduce likelihood of overcorrection that cannot be treated with glasses reduction

IV. Methods

A. Retrospective review
B. Patients operated by MADM
C. Inclusion criteria
   1. Residual ET > 12 PD after full hypermetropia correction
   2. Surgery after 6 months of age
   3. Postop follow-up for at least 6 months
   4. No associated palsy
   5. A/V pattern, IO surgery, Down syndrome, developmental delay included

V. Results

A. 107 patients operated from July 1987 to Dec. 2008
B. 58 (55.2%) males, 49 females (44.8%)
C. Age at time of surgery: 4.77 years (0.64-15.9 years)
D. Duration of symptoms: 2.4 years (1 to 178 months)
E. 12.6% Medicaid, 87.3% private insurance

VI. Preop Characteristics

A. Hyperopia: +4.29 (+0.5 to +8.75)
B. Stereopsis: 55% none, 38% gross, 12% refined
C. 100% distance glasses, 35% bifocals
D. Near deviation without correction: 46.5 PD
E. Distance deviation without correction: 38.0 PD
F. Mean target angle: 38 PD
G. Mean target angle: 38 PD

H. Mean target angle: 38 PD
I. AC/A ratio: 8.9 (48.5% ≥ 10)

VII. Operative Details

A. Bilateral medial rectus recession
B. Mean 4.9 mm in each eye (3.25-6.75)
C. 2 supra-placement (for A-pattern), 1 infra-placement (for V-pattern)
D. 29.5% associated inferior oblique surgeries

VIII. Treatment Success: 2 Months Postop (Without Manipulation of Glasses), N = 92
A. Alignment at near and distance:
   1. Success if ET ≤ 10
   2. Any XT considered failure
B. Ortho: 62%
C. ET ≤ 10: 33%
D. ET > 10: 3%
E. XT: 2%
F. Stereo: 10% nil, 17.5% gross, 62.5% refined

IX. Results at 2 Months Postop
Success in subgroup (18 patients; 17%): Good alignment and good visual acuities without glasses at distance and near!
A. Mean spherical equivalent: +2.95 (range: +0.75 to +4.25, unaided visual acuity 20/25 or better)
B. Mean target angle: 30.25 PD (15 to 42.5 PD)
C. Mean preop near deviation without correction: 41.8 (28 to 52.5 PD)

X. Treatment Success 2 years Postop
A. 88 patients
B. 54% no glasses, 38% distance glasses, 8.0% bifocals (average +1.36)
C. Stable alignment
   1. Near deviation: 96% ortho/ ET ≤ 10
   2. Distance deviation: 94% ortho/ ET ≤ 10
D. 94% success

XI. Discussion (See Table 1)

XII. Summary
A. Full hyperopic correction for fully refractive accommodative ET; bifocals for high AC/A
B. Use new surgical protocol for augmented surgery for to treat partially accommodative esotropia. Physiologic target angle based on the average of Nsc and Dsc.
C. Restores binocular vision
D. Allows reduction or elimination of glasses or bifocals in selected patients
E. Safe, with low overcorrection/reoperation rates
F. Greater success than any other published surgical protocol

Selected Readings

Table 1.

<table>
<thead>
<tr>
<th>Author (Year)</th>
<th>Success Criteria</th>
<th>Success</th>
<th>Overcorrection</th>
<th>No. of Subjects</th>
<th>Target Angle</th>
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<tr>
<td>Jotterbrand, Isenbert (1988)</td>
<td>ET ≤ 10</td>
<td>65%</td>
<td>15%</td>
<td>20</td>
<td>Avg. of Dcc, Dsc</td>
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<tr>
<td>Multicenter Prism Adapt (1990)</td>
<td>ET/XT ≤ 8</td>
<td>83%</td>
<td>3.3%</td>
<td>61</td>
<td>Full prism adapted near angle</td>
</tr>
<tr>
<td>Wright (1993)</td>
<td>ET/XT ≤ 10</td>
<td>88%</td>
<td>12%</td>
<td>40</td>
<td>Avg. of Nsc, Ncc</td>
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<tr>
<td>West, Repka (1994)</td>
<td>ET/XT ≤ 10</td>
<td>80%</td>
<td>8%</td>
<td>25</td>
<td>Ncc + Aug (AC/A &gt; 10)</td>
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<tr>
<td>Kushner (2001)</td>
<td>ET &lt; 10</td>
<td>86%</td>
<td>4.5%</td>
<td>22</td>
<td>Ncc + Aug</td>
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<tr>
<td>Del Monte, Leo (2010)</td>
<td>ET ≤ 10</td>
<td>91.5%</td>
<td>3.4%</td>
<td>107</td>
<td>Avg. of Ncc, Dsc</td>
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Abbreviations: ET indicates esotropia; Dcc, distance deviation with correction; Dsc, distance deviation without correction; XT, exotropia; Nsc, near deviation without correction; Ncc, near deviation with correction.
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David B Granet MD
Alcon Laboratories: C

Ken K Nischal MBBS
Bausch + Lomb: C

AAO Staff
Ann L'Estrange
None

Melanie Rafaty
None

Debra Rosencrance
None
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<td>Monte A Del Monte MD</td>
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