Pediatric Ophthalmology 2019

San Francisco Sound Meets Science

Program Directors

Scott A Larson MD and Michael F Chiang MD

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

Moscone Convention Center San Francisco, California Saturday, Oct. 12, 2019

Presented by: The American Academy of Ophthalmology





DEDICATED TO THE HEALTH OF ALL CHILDREN



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On behalf of the American Academy of Ophthalmology, the American Association for Pediatric Ophthalmology and Strabismus (AAPOS), and the American Academy of Pediatrics (AAP), it is our pleasure to welcome you to San Francisco and Pediatric Ophthalmology Subspecialty Day 2019: San Francisco Sound Meets Science.



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None



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Clarity Medical Systems
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Genentech: S
Inteleretina: O
National Eye Institute: S
National Science Foundation: S
Novartis Pharmaceuticals Corp.: C

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National Eye Institute: S
Natus Medical Incorporated: L
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CME Credit

The Academy's CME Mission Statement

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

2019 Pediatric Ophthalmology Subspecialty Day **Meeting Learning Objectives**

This meeting will enable attendees to:

- Improve their ability to diagnose and manage pediatric ophthalmology, pediatric neuro-ophthalmology, and strabismus conditions
- Improve their outcomes in the management of pediatric ophthalmology, pediatric neuro-ophthalmology, and strabismus conditions
- Use AAO ophthalmic technology assessment reports to guide clinical practice
- Understand and apply emerging approaches to childhood anterior segment disease

2019 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.

Teaching at a Live Activity

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

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The American Academy of Ophthalmology is committed to ensuring that all CME information is based on the application of research findings and the implementation of evidence-based medicine. It seeks to promote balance, objectivity, and absence of commercial bias in its content. All persons in a position to control the content of this activity must disclose any and all

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Getting your badge scanned does not automatically grant CME credit. You still need to record your own educational activities. NOTE: You should claim only the credit commensurate with the extent of your participation in the activity.

CME Credit Reporting

Onsite, report credits earned during Subspecialty Day and/or AAO 2019 at CME Credit Reporting kiosks located in South Lobby, West Lobby, and the Academy Resource Center, West, Booth 7337.

Registrants whose attendance is verified at AAO 2019 receive an email on Monday, Oct. 14, with a link and instructions for claiming credit online. Attendees can use this link to report credits until Wednesday, Oct. 30.

Starting Thursday, Nov. 14, attendees can claim credits online through the Academy's CME web page, aao.org/ cme-cental

Academy Members

The CME credit reporting receipt is not a CME transcript. CME transcripts that include credits entered at AAO 2019 will be available to Academy members through the Academy's CME web page beginning Thursday, Nov. 14.

The Academy transcript cannot list individual course attendance. It will list only the overall credits claimed for educational activities at Subspecialty Day and/or AAO 2019.

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Proof of Attendance

The following types of attendance verification are available during AAO 2019 and Subspecialty Day for those who need it for reimbursement or hospital privileges, or for nonmembers who need it to report CME credit:

- CME credit reporting/proof-of-attendance letters
- Onsite registration receipt
- Instruction course and session verification

You must have obtained your proof of attendance at the CME Credit Reporting kiosks onsite, located in South Lobby, West Lobby, and in the Academy Resource Center, West, Booth 7337.

Faculty



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Joseph L Demer MD PhD Los Angeles, CA



George S Ellis Jr MD FACS New Orleans, LA



Edward G Buckley MD Durham, NC



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Aubrey L Gilbert MD Mill Valley, CA



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M Edward Wilson Jr MD Charleston, SC



Edward J Wladis MD Delmar, NY

Ask a Question and Respond to Polls Live During the Meeting Using the Mobile Meeting Guide

To submit an answer to poll or ask the moderator a question during the meeting, follow the directions below.

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- Select Program, Handouts & Evals
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- Choose "Answer Poll" or "Ask a Question"



Pediatric Ophthalmology Subspecialty Day 2019: San Francisco Sound Meets Science

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

SATURDAY, OCT. 12, 2019

7:00 AM	CONTINENTAL BREAKFAST	
8:00 AM	Welcome and Introductions	Scott A Larson MD Michael F Chiang MD*

Section I:	"Beat It on Down the Line"—Strabismus Enhancements/Reoperations					
	Moderators: Michael F Chiang MD* and Jeffrey S Hunter MD					
8:01 AM	Introduction	Jeffrey S Hunter MD				
8:03 AM	"Happy Together": Infantile Esotropes— The Art of Getting a Child's Eye Aligned	George S Ellis Jr MD FACS	1			
8:15 AM	Q&A					
8:22 AM	"Everybody's Talking": Exotropes— The Art of Managing Recurrent Exotropia	Edward G Buckley MD	2			
8:34 AM	Q&A					
8:41 AM	"Up Up and Away": Recurrent Dissociated Deviations— The Management of Recurrent DVD After Previous Surgery for DVD	David L Guyton MD*	4			
8:53 AM	Q&A					

Section II: "I Heard It Through the Grapevine"—Ophthalmic Technology Assessments

	Moderators: Gena Heidary MD* and Stacy L Pineles MD*		
9:00 AM	Introduction	Gena Heidary MD*	
9:01 AM	Atropine for the Prevention of Myopia Progression in Children	Stacy L Pineles MD*	6
9:11 AM	Q&A		
9:17 AM	Contact Lens Correction of Aphakia in Children	Scott R Lambert MD*	7
9:27 AM	Q&A		
9:33 AM	The Use of Beta-Blockers for the Treatment of Periocular Hemangiomas in Infants	Amy K Hutchinson MD	9
9:43 AM	Q&A		
9:49 AM	Orthokeratology for the Prevention of Myopic Progression in Children	Deborah K VanderVeen MD*	10
9:59 AM	Q&A		
10:05 AM	Balloon Dacroplasty for Congenital Nasolacrimal Duct Obstruction	Edward J Wladis MD*	11
10:15 AM	Q&A		
10:20 AM	REFRESHMENT BREAK and AAO 2019 EXHIBITS		

^{*} Indicates that the presenter has financial interest. No asterisk indicates that the presenter has no financial interest.

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Section III:	"Everyday People"—Controversies in Neuro-Ophthalmology	: Point-Counterpoint	
	Moderators: Gena Heidary MD* and Stacy L Pineles MD*		
10:45 AM	Are You AT the Table or ON the Menu?	Stacey J Kruger MD	12
10:50 AM	Introduction	Stacy L Pineles MD*	
10:51 AM	Do All Children With Neurofibromatosis Type 1 Require Routine Neuroimaging to Screen for Optic Pathway Glioma?	Stacy L Pineles MD*	
10:52 AM	Yes	Andrew G Lee MD	14
10:57 AM	No	Robert A Avery DO*	15
11:02 AM	Discussion		
11:07 AM	Is OCT Useful for the Diagnosis of Pseudopapilledema vs. Papilledema?	Gena Heidary MD*	
11:08 AM	Yes	Mays A El-Dairi MD	16
11:13 AM	No	Lynn K Gordon MD PhD*	17
11:18 AM	Discussion		
11:23 AM	Do All Children With Optic Neuritis Require Steroid Treatment at Presentation?	Stacy L Pineles MD*	
11:24 AM	Yes	Paul H Phillips MD	18
11:29 AM	No	Grant T Liu MD*	19
11:34 AM	Discussion		
11:39 AM	Do All Children With Acute Comitant Esotropia Require Neuroimaging?	Gena Heidary MD*	
11:40 AM	Yes	Aubrey L Gilbert MD	20
11:45 AM	No	Jane C Edmond MD	21
11:50 AM	Discussion		
11:55 AM	Q&A		
12:05 PM	LUNCH and AAO 2019 EXHIBITS		
Section IV:	"What a Long Strabismus Trip It's Been"— Surgical Lessons Learned Since I Began My Practice		
	Moderators: Gil Binenbaum MD* and Scott A Larson MD		
1:00 PM	Introduction	Gil Binenbaum MD*	
1:02 PM	Lessons From Boston	David G Hunter MD PhD*	23
1:14 PM	Lessons From DC	Mohamad S Jaafar MD FACS	24
1:26 PM	Lessons From Philadelphia	Brian J Forbes MD PhD	26
1:38 PM	Lessons From Melbourne	Lionel Kowal MBBS	27
1:50 PM	Q&A		
Section V:	"See Me, Heal Me"—Pediatric Cataract Surgery Pearls		
	Moderators: Scott A Larson MD and Nils K Mungan MD		
2:00 PM	Introduction	Nils K Mungan MD	
2:01 PM	How Many Roads Must a Doc Walk Down? Pearls From My Years in Practice	M Edward Wilson Jr MD*	28
2:11 PM	How Groovy Gadgets Can Improve Pediatric Cataract Surgery	Kanwal K Nischal MBBS*	29

^{*} Indicates that the presenter has financial interest. No asterisk indicates that the presenter has no financial interest.

Monte A Del Monte MD

David G Morrison MD

Federico G Velez MD

Malcolm R Ing MD

Sean P Donahue MD PhD*

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42

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4:13 PM

4:27 PM

4:41 PM

4:55 PM

5:00 PM

Third Nerve Palsy Surgery

Perforations and Infections

Q&A Adjourn

Complicated Strabismus After Non-strabismus Surgery

^{*} Indicates that the presenter has financial interest. No asterisk indicates that the presenter has no financial interest.

"Happy Together": Infantile Esotropes— The Art of Getting a Child's Eye Aligned

George S Ellis Jr MD FACS

NOTES

"Everybody's Talking": Exotropes— The Art of Managing Recurrent Exotropia

Edward G Buckley MD

- I. What Happened?
 - A. Incomplete or inaccurate preoperative evaluation
 - 1. Full deviation not uncovered
 - Refractive state not appreciated (especially myopia)
 - Accommodative factors not identified (high accommodative convergence-to-accommodation ratio)
 - 4. Incomitance not recognized
 - 5. Near distance disparity not apparent
 - 6. A or V pattern present
 - B. Insufficient surgical correction
 - 1. Poor technique (too little or too much)
 - 2. Inaccurate measurement
 - 3. Wrong surgical amounts
 - 4. Wrong surgical procedure
 - C. Poor or absent binocular function—stability barriers
 - 1. No fusion, no stereopsis
 - 2. Amblyopia
 - 3. Severe monocular vision loss
 - D. Type of original deviation
 - 1. Constant
 - 2. Intermittent
 - 3. Congenital
 - 4. Neurogenic
 - 5. Myopathic

- II. What's the Motility Look Like Now? What to Look For?
 - A. Exotropic everywhere
 - 1. Intermittent
 - a. Comitant: Best case, just a redo, good sensory assistance
 - Incomitant: Need to adjust surgical approach for different size deviation in different gaze positions
 - 2. Constant
 - a. Comitant: Usually insufficient amounts
 - b. Incomitant: Will require asymmetric surgery
 - B. Mixed exo/esodeviation
 - C. Ortho somewhere
- III. Presurgical Evaluation
 - A. Refraction: Look for myopia
 - B. Sensory status: Fusion possible?
 - C. Incomitance?
 - D. Near/distance disparity?
 - E. A or V pattern?
 - F. Muscle function: Most important!!!
- IV. What to Do: Nonsurgical Options
 - A. Prisms
 - B. OnabotulinumtoxinA, lateral rectus
- V. What to Do: Surgical Options (see Figure 1)

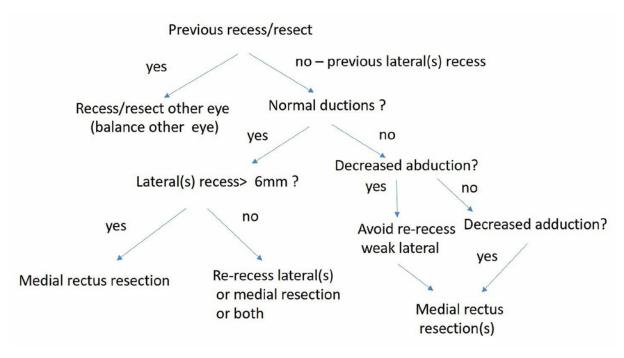


Figure 1. Surgical strategy, exotropia reoperation.

VI. What to Do for Patterns and Near Distance Disparity?

- A. A or V patterns
 - 1. Address A patterns with appropriate surgery
 - a. Medials up, laterals down
 - b. Avoid superior oblique weakening in fusing patients
 - 2. V patterns
 - a. Medials down, laterals up
 - b. Bilateral inferior oblique surgery if overacting
- B. Near-distance disparity
 - 1. Worse distance: Consider larger amounts
 - 2. Worse near: Consider medial resection(s)

"Up Up and Away": Recurrent Dissociated Deviations—The Management of Recurrent DVD After Previous Surgery for DVD

David L Guyton MD

- I. Cause of Dissociated Vertical Deviation (DVD)
 - A. Incomplete development of binocular vision in infancy
 - B. Latent nystagmus develops, including a cyclovertical component
 - C. DVD: A learned, sometimes anticipatory, vertical vergence/cycloversion combined with a supraversion, the vergence component of which damps the cyclovertical component of latent nystagmus to improve vision, each component obeying Hering's Law
- II. Muscles Involved in the Vertical Vergence Component of DVD
 - A. Superior oblique muscle of the fixing eye
 - B. Inferior oblique muscle of the hyperdeviating, non-fixing eye
- III. Possible Surgical Strategies
 - A. Eliminate latent nystagmus. But how? No known direct ways
 - B. Weaken muscles
 - C. Create restrictions
 - D. Transpose muscle attachments to alter the muscles' mode of action
- IV. Common Initial Surgeries
 - A. Bilateral superior rectus muscle recessions 8-14 mm, asymmetric if DVD is asymmetric
 - 1. Add inferior oblique muscle weakening if V pattern is present.
 - 2. Add superior oblique muscle weakening if moderate to large A pattern is present.
 - B. Bilateral inferior oblique muscle anterior transposition, especially if V pattern is present, but not if A pattern is present; inferior oblique nasal transposition is still stronger.
 - C. Bilateral posterior fixation procedures of superior rectus muscles, 12-16 mm posterior to the limbus
 - D. Bilateral inferior rectus muscle resections
 - E. Four-oblique-muscle weakening, avoids anterior segment ischemia if previous recession-resection

- V. Problems With Surgeries Mentioned Above
 - A. Bilateral superior rectus muscle recessions 8-14 mm
 - 1. Frequent problems with asymmetrical effects
 - Upper lid elevation if do not dissect far backward
 - B. Bilateral inferior oblique muscle anterior transpositions
 - 1. Some problem with asymmetric effects
 - 2. Risk of anti-elevation syndrome; should attach muscle adjacent to inferior rectus muscle corner
 - 3. Elevation of lower lids
 - 4. Long-term development of A-pattern
 - Long-term development of inverted Brown pattern
 - C. Posterior fixation procedures on superior rectus muscles, 12-16 mm posterior to the limbus
 - Not effective unless fixation is far backward, weakening muscles even in straight-ahead gaze
 - 2. Surgically difficult to obtain adequate exposure
 - D. Bilateral inferior rectus muscle resections
 - 1. Elevation of lower lids
 - 2. Recurrence of DVD
 - E. Four-oblique muscle weakening: As a symmetric procedure, only useful for symmetric cases
- VI. Exam Components When an Overcorrection or Recurrence Occurs
 - A. Restrictions, overactions, or underactions of muscles
 - B. New-onset A or V pattern
 - C. Asymmetries in misalignment, palpebral fissures, and fixation behavior
 - D. Fundus torsion (none in pure DVD, occurs primarily with oblique muscle abnormalities)
 - E. Assessment of anomalous head posture (usually to damp latent nystagmus; if so, must include surgery on fixing eye)
 - F. Lancaster red-green plot, with each eye fixing

VII. Reoperation Strategies

- A. If overcorrection, move overcorrecting muscle back toward its origin, if feasible.
- B. If undercorrection, perform more of the same surgery, on adjustable sutures if possible, or do judicious surgery on additional muscles.
- C. If new deviation, consider other muscle surgery to compensate.
- D. Adjust sutures where possible, but immediate postop DVD not fully manifest if fixation is poor.

VIII. Specific Suggestions for Reoperation

- A. Superior rectus muscle recession
 - 1. If asymmetric result, do more recession on higher eye, or advance muscle on lower eye, on adjustable sutures.
 - 2. Avoid strong double elevator weakening by adding only inferior oblique myotomy, not anterior transposition, to avoid upper lid retraction.
- B. Bilateral inferior oblique anterior transpositions
 - 1. If persistent hyperdeviation, judicious superior rectus muscle recession on adjustable suture.
 - If overcorrection, convert to an inferior oblique myectomy on the lower eye.
- C. Asymmetric hyperdeviation after posterior fixation procedures on superior rectus muscles: Consider inferior oblique myectomy on higher eye.
- D. Overcorrection from inferior rectus muscle resection: Loosen the inferior rectus muscle(s); helps correct lower lid elevation as well.

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Atropine for the Prevention of Myopia

Progression	in Children	•	
Stacy L Pineles MD			

NOTES

7

Contact Lens Correction of Aphakia in Children

Scott R Lambert MD

Introduction

Contact lenses have been used since the 1950s for optical correction in children after cataract surgery. They are particularly useful for children with monocular aphakia because of the marked aniseikonia induced by spectacles. Early contact lenses were made of polymethylmethacrylate or hydrogel, which have low oxygen permeability. In the 1970s, a major advance occurred with the introduction of contact lenses made of a silicone elastomer (SE) that has a high oxygen permeability (Dk = 340), which allows it to be worn on an extended-wear basis. Subsequently, rigid gas permeable (RGP) and silicone hydrogel contact lenses became commercially available; both also have increased oxygen permeability.

In the United States, the most commonly used contact lens to correct pediatric aphakia is an SE lens—the SilSoft Super Plus (Bausch + Lomb; Rochester, NY). However, SE lenses are manufactured in a limited range of powers and base curves, and the hydrophilic surface coating deteriorates over time. This allows

its underlying hydrophobic surface to be exposed, which results in reduced wettability and mucus buildup on its anterior surface. SE lenses also have a limited capacity to correct corneal-induced astigmatism that exceeds 2 D in magnitude.

In contrast, RGP lenses can be customized to achieve virtually any power, base curve, or diameter, and they are better able to correct corneal astigmatism (up to 6 D). In addition, they are more durable and more economical. The primary disadvantage of RGP lenses is that for the high plus powers needed to correct pediatric aphakia, their effective oxygen permeability is not optimal for extended wear.

Published Results

See Table 1.

Table 1. Level II Studies

Author(s), Year	Treatment	Cohort	Timing	Outcomes
Aasuri et al., 1999	• Retrospective review of	• N = 74 children (106	1 day, 1 mo, every 3 mos	• 27 eyes lost to FU
	aphakic eyes fitted with SilSoft CLs (Bausch +	eyes)		• 23 lens-related events, including epithelial erosions, keratitis,
	Lomb; Rochester, NY)	• Age 1 mo to 12 yrs		ulcers, corneal edema, and con-
	 Majority due to con- genital cataract 			junctivitis
Ozbek et al., 2002	Congenital cataract sur-	• <i>N</i> = 51 children (83	 Monthly FU first year 	• 11 eyes stopped CL wear
	gery followed by SilSoft CLs	eyes)	Bimonthly FU second	because of frequent loss, ocular irritation, and corneal infiltra-
		• 19 unilateral and 32 bilateral	year, 4 times/yr thereafter; mean FU 4.5 yrs	tion
		• Age at time of surgery:	,	• 2 cases of itching and photopho- bia due to conjunctival irritation
				• 1 case of recurrent corneal infiltration
Russell et al., 2012	Cataract surgery followed by CL wear with majority SilSoft	• <i>N</i> = 57 unilateral aphakes randomized to CL wear	Assessment of visual outcomes at 1 yr of age	 No difference between Sil- Soft and RGP in terms of VA improvement
		• 42 SilSoft only, 12 RGP, 3 both		• RGP lenses needed replacement more often, but not significant
				• 3 AEs in SilSoft group; all resolved (corneal abrasion, keratitis, and opacity)
Russell et al., 2017	Cataract surgery followed by CL wear with majority SilSoft	• <i>N</i> = 57 unilateral aphakes randomized to CL wear	Assessment of visual outcomes at 5 yrs of age	13 CL-related AEs, all but 1 in SilSoft group
		• 24 SilSoft only, 11 RGP, 17 both		

Future Research

Silicone hydrogel contact lenses with high oxygen permeability are currently available in the powers needed to correct pediatric aphakia. They have the comfort associated with hydrogel lenses and the oxygen permeability (Dk = 60) of RGP lenses. Compared with SE lenses, silicone hydrogel lenses are associated with fewer lens deposits and can be shaped to achieve a wider range of powers and base curves. However, custom soft lenses are not approved for overnight wear.

Parental education is critical to the success of contact lens wear in aphakic children. Research should be directed toward developing training modules for parents to enhance their proficiency at managing these lenses to improve consistency of lens wear, provide better lens hygiene, and reduce adverse events.

- Aasuri MK, Venkata N, Preetam P, Rao NT. Management of pediatric aphakia with SilSoft contact lenses. CLAO J. 1999; 25:209-212.
- 2. Ozbek Z, Durak I, Berk TA. Contact lenses in the correction of childhood aphakia. *CLAO J.* 2002; 28:28-30.
- 3. Russell B, Ward MA, Lynn M, Dubois L, Lambert SR. The Infant Aphakia Treatment Study contact lens experience: one-year outcomes. *Eye Contact Lens*. 2012; 38:234-239.
- 4. Russell B, DuBois L, Lynn M, Ward MA, Lambert SR. The Infant Aphakia Treatment Study contact lens experience to age 5 years. *Eye Contact Lens*. 2017; 43:352-357.

The Use of Beta-Blockers for the Treatment of **Periocular Hemangiomas in Infants**

Amy K Hutchinson MD

- I. Questions for Assessment
 - A. Are beta blockers safe and effective in inducing regression of periocular hemangiomas?
 - B. Do beta blockers reduce the amount of induced astigmatism and the incidence of amblyopia associated with periocular hemangiomas?
 - C. Does the effect of beta blockers or the frequency of adverse events vary with the route of administra-
- II. Background and Current Practice of Treatment for Periocular Hemangiomas
 - A. In 2008, Leaute-Labreze reported a serendipitous finding that propranolol could induce involution of infantile hemangioma (IH).
 - B. Current clinical practice guidelines¹⁻³
 - 1. There is "strong evidence" to recommend oral propranolol as first-line agent for IHs requiring systemic treatment.

Table 1. AHRQ Summary of Comparative Efficacy of **Various Treatments for IHs**

Drug	Mean Estimate of Expected Clearance (%)	95% Bayesian Credible Interval (%)
Propranalol	95	88-99
Topical timolol	62	39-83
Intralesional triamcinolone	58	21-93
Oral steroid	43	21-66
Control	6	1-11

- 2. Periocular IH > 1 cm considered "high risk" because of potential functional impairment including astigmatism, anisometropia, amblyopia, and proptosis.
- 3. Current recommendation is for propranolol 2-3 mg/kg/d unless there are comorbidities (PHACES) or adverse effects (sleep disturbance) that necessitate a lower dose.

- 4. Propranolol should be administered after feeding, and dosages should be held at times of diminished oral intake or vomiting to reduce the risk of hypoglycemia.
- 5. Initiation protocols (inpatient vs. outpatient)
- III. Current Approach to the Question (Panel Discussion, 3 mins.)
- IV. Summary and Overview of Evidence/Proposal of How Evidence Might Be Implemented/Potential Barriers⁴ (3 mins.)
 - A. Evidence for treatment of periocular hemangiomas is limited to Levels 2 (n = 3) and 3 (n = 13).
 - B. Question 1: Most common treatment regimen used was oral propranolol 2 mg/kg/d, but intralesional and topical beta blockers were used. Treatment effect was measured in terms of reduction of the size of the lesions, which occurred in most patients.
 - C. Question 2: Use of beta blockers was associated with reduced astigmatism but was statistically significant in only 2 series, and there was no control for the reduction of astigmatism typically seen in infants.
 - D. Question 3: Beta blockers were generally well tolerated, with mild side effects (fatigue, GI upset, sleep disturbances, minor wheezing, and cold extremities). Data were insufficient to determine whether route of administration had an effect on rate of complications.
- V. Panelist Comment and Audience Questions (6 minutes)

- 1. Krowchuk DP, Frieden IJ, Mancini AJ, et al. Clinical practice guideline for the management of infantile hemangiomas. Pediatrics 2019; 143:e20183475.
- 2. Drolet BA, Frommelt PC, Chamlin SL, et al. Initiation and use of propranolol for infantile hemangioma: report of a consensus conference. Pediatrics 2013; 131:128-140.
- 3. Darrow DH, Greene AK, Mancini AJ, et al. Diagnosis and management of infantile hemangioma. Pediatrics 2015; 136:e1060-1104.
- 4. Hutchinson AK, Kraker RT, Pineles SL, et al. Ophthalmic technology assessment: the use of beta-blockers for the treatment of periocular hemangiomas in infants. Ophthalmology 2019; 126(1):146-155.

Orthokeratology for the Prevention of Myopic Progression in Children

Deborah K VanderVeen MD

- I. Background
 - A. Myopic epidemic
 - B. Treatments studied to date
- II. Orthokeratology Basics
- III. Literature Search Through August 1, 2018
 - A. Level I evidence, 1 study; Level II evidence, 11 studies; Level III evidence, 1 study
 - B. Ortho-K vs. control (spectacles, soft contact lens, atropine)

IV. Results

- A. Review of Level I (randomized within subject crossover trial) and Level II (randomized and nonrandomized case comparison trials)
- B. Axial length studies (optical biometry) show approximately 50% (statistically significant) reduction in axial elongation over 2-year study period. Change in axial length values are approximately 0.3 mm for Ortho-K group, compared to 0.6 mm for control groups.

- C. Refractive outcomes: No significant increase in Ortho-K groups, statistically significant increase in myopia in control groups (clinically, mean/median about 1 D by final visit)
- D. Atropine (0.125%) vs. Ortho-K: Significant increase in atropine group at 3 years, but not statistically different from Ortho-K group
- V. Limitations of Studies
- VI. Safety of Ortho-K
- VII. Future Research

Balloon Dacroplasty for Congenital Nasolacrimal Duct Obstruction

Edward J Wladis MD

On behalf of the Oculoplastic Surgery Ophthalmology Technology Assessment Committee: Vinay Aakalu MD, Michael Yen MD, Jurij Bilyk MD, Rachel Sobel, and Louise Mawn MD

- I. In children with congenital nasolacrimal duct obstructions (NLDOs) that do not resolve spontaneously, probing remains the standard of care as a primary procedure.
- II. Multiple options have been proposed to address NLDOs that are refractory to an initial probing.
 - A. Repeat probing: low success rate (25%-64% in largest series)
 - B. Stenting of the duct
 - 1. High success rate
 - 2. May require return visit to the operating room to remove stent
 - 3. Risk of damage to the lacrimal system
 - 4. Risk of premature extrusion
 - C. Balloon dacryoplasty
 - D. Dacryocystorhinostomy
- III. The OTA sought to identify the benefits of balloon dacryoplasty after a failed initial probing.
 - A. A PubMed search was conducted in September 2017 and repeated in April 2018.
 - 1. 104 articles were reviewed to ensure that they fit with search criteria.
 - 2. 36 were selected for full review.
 - 3. Data were abstracted from studies that met cri-
 - 4. Methodologist assigned the level of evidence grade.

- IV. Results: Eight Articles Identified
 - A. Three studies rated as Level II
 - B. Five studies rated as Level III
 - C. Success rates ranged from 75% to 100%.
 - 1. Most studies defined success as complete resolution of symptoms and/or normal dye disappearance test.
 - 2. Success rates were comparable to those published for stenting.
 - D. Two complications reported, and both were selflimited emesis.
 - E. Two studies compared stenting with balloon dacryoplasty, and these investigations concluded that the techniques yielded similar outcomes.
- V. Future Investigations
 - A. Identify optimal duration between failed probing and a dacryoplasty
 - B. Determine optimal age for procedure
 - C. Perform studies that meet criteria for Level I evidence
 - D. Define role of this procedure in the adult population

Are You AT the Table or ON the Menu?

Pediatric Ophthalmology Subspecialty Day

Stacey J Kruger MD

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

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

Please join the dedicated community of ophthalmologists who are contributing to protect quality patient eye care for everybody.

The OPHTHPAC Committee is identifying Congressional Advocates in each state to maintain close relationships with federal legislators to advance ophthalmology and patient causes. At Mid-Year Forum 2019, we honored three of those legislators with the Academy's Visionary Award. This served to recognize them for addressing issues important to us and to our patients. The Academy's Secretariat for State Affairs is collaborating closely with state ophthalmology society leaders to protect Surgery by Surgeons at the state level.

Our mission of "protecting sight and empowering lives" requires robust funding of both the Surgical Scope Fund and OPHTHPAC. Each of us has a responsibility to ensure that these funds are strong so that ophthalmology can be represented "at the table."

OPHTHPAC®

OPHTHPAC represents the profession of ophthalmology to the U.S. Congress and operates to protect you and your fellow ophthalmologists from payment cuts, burdensome regulations, scope-of-practice threats, and much more. OPHTHPAC also works to advance our profession by promoting funding for vision research and expanded inclusion of vision in public and private programs—all of which provide better health-care options for your patients. OPHTHPAC is your federal voice in Washington, D.C., and we are very successful in representing your professional needs to the U.S. Congress.

Among OPHTHPAC's most recent victories are the following:

- Securing greater flexibility in the new Medicare Payment System
- Ensuring proper reimbursement of Medicare Part B drugs
- Blocking onerous administrative burdens on contact lens prescribers
- Preserving access to compounded drugs
- Preventing additional cuts to Medicare

However, ophthalmology's federal issues are a continuous battle, and OPHTHPAC is always under pressure to ensure we have strong political connections in place to help protect ophthalmology, its members, and their patients.

The support OPHTHPAC receives from invested U.S. Academy members helps build the federal relationships that advance ophthalmology's agenda on Capitol Hill. These relationships allow us to have a seat at the table with legislators

willing to work on issues important to us and our patients. We also use these congressional relationships to help shape the rules and regulations being developed by federal agencies. Help strengthen these bonds and ophthalmology's legislative support.

Right now, major transformations are taking place in health care. To ensure that our federal fight and our PAC remain strong, we need the support of every ophthalmologist to better our profession and ensure quality eye care for our patients. Invest with confidence in the strongest PAC working to ensure your success as an ophthalmologist.

Contributions to OPHTHPAC can be made here at AAO 2019, online at www.aao.org/ophthpac, or by texting MDEYE to 41444.

At Mid-Year Forum 2019, the Academy, the American Association for Pediatric Ophthalmology & Strabismus (AAPOS), and the American Academy of Pediatrics—Ophthalmology Section ensured a strong presence of pediatric ophthalmologists to support ophthalmology's priorities. Ophthalmologists visited members of Congress and their key health staff to discuss ophthalmology priorities as part of Congressional Advocacy Day. The AAPOS remains a crucial partner with the Academy in its ongoing federal and state advocacy initiatives.

Surgical Scope Fund

The Surgical Scope Fund (SSF) provides grants to state ophthalmology societies to support their efforts to protect patient safety from dangerous optometric surgery proposals. Since its inception, the Surgery by Surgeons campaign and the SSF, in partnership with state ophthalmology societies, have helped 40 state/ territorial ophthalmology societies reject optometric scope-ofpractice expansions into surgery.

Thanks to the 2019 SSF contributions from ophthalmologists just like you, SSF has had a successful year, preserving patient safety and surgical standards in state legislatures across the country, including six critical wins in Alabama, Texas, Vermont, Wyoming, Maryland, and Iowa. The 2019 battle is far from over, though. For example, Pennsylvania and Massachusetts are under attack, and California and Illinois are facing threats.

If you have not yet made a 2019 SSF contribution, contributions can be made at our booth at AAO 2019 or online at www.aao.org/ssf. If you already have made that 2019 contribution, please go to www.safesurgerycoalition.org to see the impact of your gift.

Dollars from the SSF are critical to building complete cutting-edge political campaigns, including media (TV, radio, and social media), educating and building relationships with legislators, and educating the voting public to contact their legislators. This work helps to secure success in protecting patient safety by defeating optometry's surgical initiatives.

Each of these endeavors is very expensive, and no one state has the critical resources to fight big optometry on their own. Ophthalmologists must join together and donate to the SSF at www.aao.org/ssf to fight for patient safety.

Surgical Scope Fund	OPHTHPAC® Fund	State EyePAC	
To protect patient safety by defeating opto-	Ophthalmology's interests at the federal level	Support for candidates for state House, Sen-	
metric scope-of-practice initiatives that threaten patient safety and quality surgical care	Support for candidates for U.S. Congress	ate, and governor	
Political grassroots activities, government relations, PR and media campaigns	Campaign contributions, legislative education	Campaign contributions, legislative education	
$\it No$ funds may be used for campaign contributions or PACs.			
Contributions: Unlimited	Contributions: Limited to \$5,000	Contribution limits vary based on state regu-	
Individual, practice, and organization		lations.	
Contributions are 100% confidential.	Contributions above \$200 are on the public record.	Contributions are on the public record depending upon state statutes.	

The Secretariat for State Affairs thanks the AAPOS, which joined state ophthalmology societies in already contributing to the SSF in 2019, and it looks forward to the association's continued financial support. These ophthalmic organizations complete the necessary SSF support structure for the protection of our patients' sight.

State Eye PAC

It is increasingly important for all ophthalmologists to support their respective State Eye PACs because campaign contributions to legislators at the state level must come from individual ophthalmologists and cannot come from the Academy, OPH-THPAC, or the SSF. The presence of a strong State Eye PAC providing financial support for campaign contributions and legislative education to elect ophthalmology-friendly candidates to the state legislature is critical, as scope-of-practice battles and many regulatory issues are all fought on the state level.

ACTION REQUESTED: Help Ophthalmology Ensure a "Seat at the Table"

Academy SSF contributions are used to support the infrastructure necessary for state legislative/regulatory battles and for public education. State PAC and OPHTHPAC contributions are necessary at the state and federal levels, respectively, to help elect officials who will support the interests of our patients. Contributions to *each* of these three funds are necessary and help us protect sight and empower lives. SSF contributions are completely confidential and may be made with corporate checks or credit cards, unlike PAC contributions, which must be made by individuals and are subject to reporting requirements.

Please respond to your Academy colleagues and be part of the community that contributes to OPHTHPAC, the SSF, and your State Eye PAC. Please be part of the community that ensures ophthalmology has a strong voice in advocating for patients.

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Do All Children With Neurofibromatosis Type 1 Require Routine Neuroimaging to Screen for Optic Pathway Glioma? Yes

Andrew G Lee MD

- I. Optic Pathway Gliomas (OPGs) in Neurofibromatosis Type 1 (NF1)
 - A. OPGs typically are grade 1, pilocytic astrocytomas.
 - B. About 15% of children with NF1 have OPG.
 - C. OPG in NF1 is often asymptomatic and more indolent than non-NF1 OPG.
 - D. Risk of symptomatic OPG is greatest in children under 7 years of age.
 - E. Older individuals are less likely to require OPG intervention.
 - F. Visual and endocrine presentations of OPG can be asymptomatic.
- II. The Additional Agreed-Upon Facts
 - Early detection has not been proven to reduce risk of visual loss.
 - B. Initial normal MRI does not exclude future OPG.
 - Treatment for an OPG is not required if asymptomatic OPG.
 - D. Routine screening often requires sedation (risk) in children.
 - E. Up to two-thirds of the OPGs would never become symptomatic.
 - F. But the gold standard for OPG detection is MRI.
- III. Thus, the "Party Line" for OPG in NF1
 - A. Until 8 years old clinical assessment for OPG: every 6 to 12 months
 - B. Routine MRI is not currently advised in asymptomatic NF1.
 - Scan symptomatic or "sign-omatic" (signs but no symptoms) kids with NF1 for OPG
 - D. Examining kids is tough.
- IV. The Problem: How Do We Define "Asymptomatic" in Young Kids With OPG NF1?
 - A. Young children do not/cannot complain of visual impairment until late.

- B. Sometimes only discovered after bilateral visual loss
- Visual assessment for symptoms and signs in kids is very difficult.
- D. Parents need to be alert to signs of visual problems (eg, failure to pick up small toys, bumping into objects).
- E. Visual assessment is problematic with NF1 cognitive deficits.
- F. OPG can present as endocrine disturbances (precocious or delayed puberty, increased growth velocity) or hydrocephalus.
- V. NF1 Is a Tumor Suppressor Gene
 - A. We scan people for less than this.
 - B. Not just neuroimaging but whole body MRI should be considered at transition to adult.
 - A single whole-body MRI should be considered at transition to adulthood.
 - To assist in determining approaches to longterm follow-up
 - 3. See Evans DGR, Salvador H, Chang VY, et al. Cancer and central nervous system tumor surveillance in pediatric neurofibromatosis 1. *Clin Cancer Res.* 2017; 23(12):e46-e53.
- VI. Do All Children With Neurofibromatosis Type 1 Require Routine Neuroimaging to Screen for Optic Pathway Glioma? Summary: YES
 - A. All NF1 children, consider MRI when transitioning to adulthood
 - B. Neuroimaging: Yes, but whole body MRI
 - C. Screen NF1: whole body tumor burden as well as OPG
 - D. MRI: symptomatic, "sign-omatic," but not automatic
 - E. NF1 is a tumor suppressor gene; we scan kids for less than this.

Do All Children With Neurofibromatosis Type 1 Require Routine Neuroimaging to Screen for Optic Pathway Glioma? No

Robert A Avery DO

No Position

Neurofibromatosis type 1 (NF1) is the most common cancer predisposition, occurring in nearly 1 in every 3000 births. Optic pathway gliomas (OPGs), low-grade gliomas involving 1 or multiple portions of the anterior visual pathway (optic nerve, chiasm, and tracts), can occur in upwards of 15%-20% of children with NF1. Interestingly, less than half of the children with NF1-OPGs will experience vision loss from their tumor and be candidates for treatment of their tumor.

NF1-OPGs are typically discovered for the first time using brain MRI during early childhood (ie, 1-8 years old), although discovery during the second decade of life has been reported even in the setting of previously normal neuroimaging results.²⁻⁴ Two separate studies attempted to determine whether performing a screening MRI improved clinical outcomes for children with NF1-OPGs.^{5,6} Despite having similar results, each manuscript arrived at a different conclusion about the utility of MRI screening.

Given the lack of data demonstrating a clinical benefit to screening MRIs, the potential risks to performing MRI under anesthesia, the risks of pursuing incidental findings, the extra costs, and the inability to capture a significant number of subjects whose care would potentially be improved, screening MRI should not be performed in children with NF1.^{7,8} Current guidelines clearly outline when MRI should be performed in children with NF1.¹

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- 2. Listernick R, Charrow J, Greenwald M. Emergence of optic pathway gliomas in children with neurofibromatosis type 1 after normal neuroimaging results. *J Pediatr.* 1992; 121:584-587.
- 3. Listernick R, Charrow J, Greenwald M, Mets M. Natural history of optic pathway tumors in children with neurofibromatosis type 1: a longitudinal study. *J Pediatr.* 1994; 125:63-66.
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Is OCT Useful for the Diagnosis of Pseudopapilledema vs. Papilledema? Yes

Mays El-Dairi MD

The following situation can give a lot of anxiety to many pediatric ophthalmologists, pediatricians, and parents: Bilaterally elevated optic nerves with blurred margins, no vessel obscuration, no visible drusen, and no visible spontaneous venous pulsations. Vision and visual fields are normal. Child with no headaches.

In a situation like this, the differential diagnosis is as follows:

- Pseudopapilledema. Most likely a benign scenario. There is no need for neuroimaging. Child to be followed over time.
- 2. Papilledema: High-risk scenario with need for extensive neuroimaging (MRI brain and orbits with contrast and magnetic resonance venography) and possible lumbar puncture

Missing the dangerous scenario can result in life- or visionthreatening consequences. Overdiagnosing the dangerous scenario can be very costly (76% of cases referred to a pediatric ophthalmologist).¹

To help discern the two scenarios, ancillary tests available to use include the following:

- Fundus photos (or funduscopy, identifying characteristic clinical appearance)
- 2. Visual fields
- 3. Brain CT scan looking for calcifications from drusen
- 4. Ocular ultrasound
- 5. Fluorescein angiogram
- 6. Fundus autofluorescence
- 7. OCT

Each modality carries its own sensitivity and specificity, and physicians will usually use the modality they are most comfortable with. Unfortunately, none of these modalities is currently a golden standard. (Lumbar puncture with intracranial pressure measure is the only current gold standard.)

Some physicians will use OCT as a personal preference for obvious reasons: ease of imaging and reproducibility; it's noncontact and does not require an intravenous injection.

Signs of papilledema vs. pseudopapilledema on OCT include the following:

- 1. **Upward bowing of Bruch membrane**: ^{2,3} Although not a very sensitive sign for a retrobulbar pathology, it is highly specific.
- 2. Thickening of the peripapillary nerve fiber layer (RNFL): This item in particular can be tricky to analyze because segmentation errors are very common and need to be corrected. Also, clinicians need to keep in mind that a highly hyperopic eye will have a falsely elevated RNFL, and a highly myopic eye will have a falsely thin RNFL.⁴
- 3. Enhanced depth imaging OCT of the optic nerve: Looking for the classic peripapillary hyper-reflective ovoid mass-like structures (PHOMSs) does not help distinguish papilledema from pseudopapilledema, as POHMS can be present in both entities. The presence of signal-poor core

- with a hyper-reflective cap is specific for optic nerve head drusen.⁴
- 4. Bruch membrane opening (BMO): Nerves with papilledema have large BMO that reverses as the swelling resolves. ⁵ Nerves with pseudopapilledema have a smaller Bruch membrane opening. ⁶
- 5. Other: Papilledema would be associated with decrease in RNFL after treatment, and an increase in RNFL with recurrence. Pseudopapilledema is expected to be stable over time. And OCT can be a great tool for following the resolution of swelling with treatment, especially since many pediatric patients end up with pseudopapilledema after treatment and remission.⁷

In conclusion, when acquired and interpreted properly, OCT can be highly useful in differentiating low grade papilledema from pseudopapilledema. The interpretation is a multistep analysis looking at more than one scan protocol. OCT should not, however, replace a clinical examination and judgment in any way and should be interpreted only in light of the history, visual function, including visual fields, and examination.

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Is OCT Useful for the Diagnosis of Pseudopapilledema vs. Papilledema? No

Lynn K Gordon MD PhD

Differentiating pseudopapilledema from papilledema is critically important in any patient, but perhaps it is even more important in the pediatric age group. In particular, one of the most important mimickers of papilledema is buried optic disc drusen. The question is: Which method best distinguishes or identifies papilledema in children? At this point, it is my opinion that although current techniques are improving, OCT continues to have limited reliability in terms of being able to definitively differentiate between papilledema and pseudopapilledema. Therefore, it is not sufficient to rely on OCT to rule out papilledema, in particular in the pediatric population.

The advantages of using OCT are the speed of acquisition, accessibility, availability in the ophthalmology practice, and low cost of imaging. Deeper imaging is available through enhanced depth imaging (EDI), spectral domain, and swept source (SS) OCT, putting them among the most sensitive methods for diagnosing optic disc drusen. Yet they remain imperfect. In a 2017 publication, pediatric patients with papilledema or with optic disc drusen were evaluated using fundus photography, autofluorescence, fluorescein angiography, and both SS and EDI OCT. Fluorescein angiography was the most accurate method of correctly diagnosing the patients with papilledema or pseudopapilledema. While there is certainly a role in using OCT to help in the diagnosis of optic disc drusen causing pseudopapilledema, it is not yet the "gold standard."

Selected Readings

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Do All Children With Optic Neuritis Require Steroid Treatment at Presentation? Yes

Paul H Phillips MD

Children with optic neuritis comprise a heterogenous group that includes postinfectious optic neuritis, postvaccination optic neuritis, optic neuritis associated with multiple sclerosis (MS), anti-myelin oligodendrocyte glycoprotein (MOG-IgG+) optic neuritis, and neuromyelitis optica (NMO).¹⁻³ It is often not possible to definitively distinguish which subgroup of optic neuritis is present in an individual child at presentation.

MOG-IgG + optic neuritis is frequently recurrent and steroid responsive. 3-4 Prolonged steroid treatment has been shown to prevent relapses. Acute attacks are often treated with methylprednisolone IV for 3-5 days followed by a prednisone taper of several weeks. Plasma exchange may improve outcomes if initiated early. 3,5 Patients that require a prolonged course of steroid treatment may be treated with steroid-sparing agents including mycophenolate, azathioprine, rituximab, and intravenous immune globulin. Patients may present with acute disseminated encephalomyelitis and transverse myelitis (simulating NMO). MS disease modifying agents are not effective treatment for prevention of future attacks.

Neuromyelitis optica (NMO) is associated with aquaporin-4-IgG + (AQP4-IgG) antibodies. Patients with NMO may have severe visual loss and poor visual recovery, as well as permanent neurologic disability.⁶⁻⁷ Aggressive treatment with steroids and plasma exchange may improve outcomes if initiated early.⁵ Chronic, aggressive, immunosuppressive treatment is often required to minimize visual and other neurological disability. MS disease modifying agents are harmful in this group of patients.

It is not possible to definitively distinguish children with MOG-IgG+ optic neuritis versus NMO versus other types of optic neuritis at presentation. Results of serologic testing for MOG-IgG and AQP4-IgG are not available until several days after presentation. Therefore, I initially treat children with optic neuritis with methylprednisolone IV for 5 days as I follow their clinical course and await results of serologic testing to guide further treatment.

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Do All Children With Optic Neuritis Require Steroid Treatment at Presentation? No

Grant T Liu MD

Introduction

Pediatric optic neuritis differs from adult optic neuritis because the condition in children is commonly bilateral, associated with optic disc swelling, and characterized by severe vision loss. The risk of developing multiple sclerosis (MS) is related to the presence of white matter lesions in the brain at presentation, as in adults, but it is also associated with older age at presentation. ²

Visual Outcome

Wan et al³ retrospectively analyzed 59 pediatric patients with first-episode optic neuritis seen at the Children's Hospital of Boston. Fifty-two percent had or developed an underlying diagnosis—(39% MS), 7% acute disseminated encephalomyelitis (ADEM), 7% neuromyelitis optica (NMO)—and 91% received some treatment (85% corticosteroids, 7% multimodal). At 1 year, 81% had visual acuity of at least 20/20 and 89% saw at least 20/40. A poor visual outcome at 1 year (<20/40) was associated with vision of <20/20 at 3 months. Visual acuity at presentation, sex, bilateral involvement, optic nerve edema, and underlying diagnoses were not associated with poor visual outcomes. They concluded that the majority of patients regained normal visual acuity at 1 year, regardless of baseline clinical characteristics.

Wan et al³ also included data on speed of recovery. Patients who regained normal visual acuity took an average of 61 days to do so, but the time to recovery depended on the presenting visual acuity. Those who presented with a mean of CF or worse took a mean of 97 days to recover normal vision, while those who presented with visual acuity better than CF took a mean of 35 days.

Treatment

Yeh et al⁴ stated that treatment of pediatric optic neuritis consists of IV methylprednisolone for 3-5 days, although they did state that there have been no clinical trials to establish the efficacy of this treatment modality. Children were not included in the Optic Neuritis Treatment Trial (ONTT). The optimal duration of oral steroids following IV is also not clear. One study⁵

found no difference in outcome between a shorter (2 weeks) and longer (>2 weeks) course of steroids in children with pediatric optic neuritis. Case reports and small series have described the success of intravenous immunoglobulin and plasmapheresis in patients with optic neuritis not responsive to corticosteroids. ^{6,7} The Pediatric Optic Neuritis Prospective Outcomes Study⁸ was not powered to compare steroid versus no-steroid treatment.

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Do All Children With Acute Comitant Esotropia Require Neuroimaging? Yes

Aubrey L Gilbert MD

A sudden-onset eye misalignment with an equal angle of inward deviation in all fields of gaze can be termed acute comitant esotropia (ACE). This entity is generally distinguished from more common forms of childhood esotropia, such as accommodative esotropia and infantile esotropia, based on lack of improvement with correction of refractive error and on a typically later age of onset. It is also distinct from paretic or restrictive strabismus, which are mostly incomitant. The potential etiologies underlying ACE are broad, but it may be associated with significant neurologic disease in some cases. Arguments are presented in favor of neuroimaging for children who present with acute comitant esotropia.

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Do All Children With Acute Acquired Comitant Esotropia Require Imaging? No!

Jane C Edmond MD

I. Introduction

When a child presents with acute comitant esotropia (ET) and no obvious associated risk factors, many pediatric ophthalmologists, and certainly all other ophthalmologists, pediatricians, and ER docs, are compelled to obtain STAT neuroimaging. However, the vast majority of the publications regarding acute acquired comitant esotropia (AACE) in the past decade have reported much lower incidences of intracranial pathology and brought to light additional possible non-intracranial risk factors.

The purpose of this presentation compel the audience to rethink neuroimaging for all children with AACE.

II. Reassuring Clues That There Is No Intracranial Pathology and No Need to Order an MRI

A. Exam

- 1. Moderate-high hyperopia
- 2. Near ET ≥ distance ET, comitant and no lateral rectus (LR) underaction. (Distance > near ET is a sign of intracranial pathology.)
- 3. No nystagmus, especially gaze evoked, or downbeat
- 4. No papilledema

B. History

- 1. Younger age: <6 years old, less likely to have intracranial pathology
- 2. Myopia
 - a. >6 D myopia is associated with AACE, but also lesser amounts of myopia.
 - b. Older children and adults
- 3. Smartphone addiction: spasm of the near reflex?
- 4. History of any previous episode of ET/double vision, or family history of ET
- 5. Monocular occlusion (patched, ptosis): decompensated esophoria?
- 6. No headache: Many/most intracranial causes of AACE are associated with HA (± papilledema)
- No ataxia or feeling of imbalance, dysarthria, dysphagia, or weak extremities: posterior fossa tumor (brainstem or cerebellum), Chiari malformation

- 8. Stable, expected eye alignment after strabismus surgery for ET
- III. Intracranial Pathology That Has Been Reported to Cause Otherwise Asymptomatic AACE

A. Chiari malformation

- Most commonly reported intracranial risk factor for AACE
- 2. Most patients have *additional* signs related to Chiari (nystagmus, dysmetria, etc.)
- 3. Without other Chiari-related symptoms, manage with occipital decompression or strabismus surgery?
 - a. Unknown if strabismus surgery or neurosurgery has better overall outcome in these otherwise asymptomatic patients
 - b. Strabismus surgery has less risk © and can be repeated.

B. Pontine glioma

- Rare, poor prognosis, high grade tumor, usually fairly rapid progression
- 2. Usual presenting signs: multiple cranial nerve palsies, hemiparesis

C. Thalamic glioma

- 1. Rare, poor prognosis regardless of tumor grade
- 2. Usual presenting signs: hemiparesis, dysmetria, unsteady gait, nystagmus
- IV. Frequency of Intracranial Pathology in Otherwise Asymptomatic AACE: *Rare*
 - A. Dotan et al¹
 - 1. 20 children, 19 were imaged
 - 2. All normal
 - B. Buch and Vinding²
 - 1. 48 children with AACE
 - 2. MRI/CT if + "neuro signs or symptoms", <3 D hyperopia, recurrent AACE
 - 3. Two of 48 had intracranial pathology with no other signs or symptoms of intracranial disease.

V. Conclusion

- A. Neuroimaging of *all* children with AACE is not indicated.
- B. Most patients with AACE do not have intracranial pathology.
- C. When imaging is indicated:
 - Worrisome histories or abnormal exam elements at presentation or follow-up
 - 2. Recurrence of AACE after strabismus surgery
- D. Consequences of undiagnosed intracranial pathology: Either nonfatal and treatable (Chiari), or tumor with poor prognosis regardless of timing of diagnosis and will develop additional symptoms soon (thalamic or diffuse intrinsic pontine glioma)

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Lessons From Boston

David G Hunter MD PhD

I will focus my presentation on one particularly challenging topic: surgery on the superior oblique (SO) muscle/tendon.

- I. Staying Out of the OR: Topical Treatment of SO Myokymia
- II. Preop Planning
 - A. How to anticipate when the SO has been wrapped around a scleral buckle
 - B. What to do about it
- III. Intraoperative Lessons
 - A. Quick pick: Finding the muscle from the superonasal approach
 - B. Assessing changes in torsion using a corneal marker
- IV. Weakening Procedures
 - A. Brown syndrome
 - Who is likely to get an overcorrection after surgery?
 - 2. Silicone-free SO weakening
 - B. Posterior 7/8 tenectomy for A patterns

V. Strengthening Procedures

There is more to life than SO tuck and Harada Ito.

- A. SO resection
- B. SO advancement

Selected Readings

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- 2. Chang MY, Yulek F, Pineles SL, Velez FG. Surgery for superior oblique tendon anteriorization and entrapment following scleral buckle. *J AAPOS*. 2016; 20(2):165.e1-165.e1652.
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Lessons From DCSafety Pearls in the Operating Room

Mohamad S Jaafar MD FACS

Introduction

Safety in the operating room and ensuring that we operate on the correct patient, correct eye, correct muscle and perform the correct surgery are every surgeon's concern, even more so in these days of EHRs, where pertinent data is buried within innumerable pages.

Presentation

Hospitals and ambulatory surgery centers have come a long way, adopting Universal Protocols and halting for time-outs. However, the ophthalmologist's role does not stop at identifying the correct side and the fire risk. We should not suffer from time-out burn-out; we need to keep vigilant to make sure we operate on the correct rectus muscle, and so on. The surgeon, not the circulating nurse or anesthesiologist, should clearly enunciate the procedure.

The use of a simple one-page summary sheet (see Figure 1) and of diagrams in the OR bring all members of the surgical team up to speed and keep everyone informed, involved, invested, and on the same page. A clinical photo of the patient with a head turn may prove to be quite beneficial, too.

The Surgical Plan Form will include the patient's surgically pertinent diagnosis (eg, consecutive A-exotropia), relevant systemic diagnosis (eg, asthma, seizures), previous ocular surgery (eg, recession of medial rectus 6 mm, bilaterally, and resection of lateral rectus 7 mm, bilaterally), previous general anesthesia (including possible complications in patient or family members), and bleeding tendency, present medications, allergies, etc. This is followed by a brief summary of the ocular findings that dictated the surgical intervention, which includes vision, refraction, motility, and anterior and posterior segment findings. The form ends with the Surgical Plan (eg, exploration and advancement of medial rectus muscle to original insertion, with upward transposition half a muscle width, bilaterally). Most importantly: This form is filled out by the attending surgeon on the day the patient is examined and the decision for and type of surgery were determined and discussed with the patient and family, when the surgeon has the full picture of the clinical situation.

Many other pearls will be shared, such as that the "count" should not be restricted to needles and gauze but should include all "extras" on the field (eg, tubes, corneal shields).



111 Michigan Ave NW Washington, DC 20010 (202) 476 – 3015

DEPARTMENT OF OPHTHALMOLOGY SURGICAL PLAN FORM This form to be completed by the Ophthalmologist

PLACE PATIENT LABEL HERE

NAME:		AGE:
		AUE.
OPHTHALMOLOGIC DIAGNOSES:		
NON OPHTHALMOLOGIC DIAGNOSES:		
ODUTAL LINOLOGIC MOTODY AND DESCRIPT HANDS		
OPHTHALMOLOGIC HISTORY AND PRESENT ILLNESS:		
ALLERGIES, DRUG SENSITIVITIES:		
BLEEDING TENDENCIES:		
PRESENT MEDICATIONS: (Drug – dose/day – Reason)		
PREVIOUS OPHTHALMOLOGIC SURGERY: (Type – Dates -	- Complications)	
PREVIOUS ANESTHESIA: (Type – Dates – Complications)		
Refraction: OD	Best Visual Acuity:	OD
OS	,	OS
Ocular Motility:	Anterior Segment:	OD OS
		OS
	Fundus:	OD
		OD OS
CAND CAGAA DA ANA		
SURGICAL PLAN:		
CICNIATUDE.	MD	Data
SIGNATURE:	WI.D.	Time:

Lessons From Philadelphia

Brian J Forbes MD PhD

There are many different ways of proceeding with strabismus surgery, all of which can be generally appropriate for a specific strabismus surgeon. I will be presenting what I have come to be comfortable with in my 20 years of strabismus surgery at the Children's Hospital of Philadelphia, as well as methods that Joe Calhoun of the Wills Eye Hospital developed and preached in his 45-year pediatric ophthalmology career. Certain techniques will be presented that can be utilized in a teaching institution to allow a safer indoctrination into surgery and ease the trainees into developing good surgical habits. While no one technique works for everyone, hopefully the attendee will be exposed to certain tricks of the business and walk away with some ideas about how to more easily and safely perform strabismus surgery.

Additional ideas presented by various pediatric ophthalmologists at the Joseph Calhoun conference at The Wills Eye Hospital (Wills Eye Hospital Pediatric Ophthalmology Forum) this fast past fall will also be presented.

- I. Comfortable with one muscle surgery in comitant if esotropia (ET) ≤ 20 or exotropia (XT) ≤ 16.
 - A. Double the angle and do a single muscle for the bilateral medial rectus (BMR) dose (or bilateral lateral rectus [BLR]) plus 1 mm.
 - B. For example, ET = 20. So $20 \times 2 = 40$. BMR for 40 = 5.5. So do 5.5 mm + 1.0 mm = 6.5 -mm recession.
- II. Can often do hangback surgery, unless:
 - A. Ischemic encephalopathy or other CNS disease where the child does not voluntarily fuse (converge) at least some. Consider BMR resect for these kids with constant XT.
 - B. Best if strabismus is intermittent
 - C. Can use hangback up to 6.5 mm for medial or 7.5 mm for lateral
- III. I do not use for larger than 6 for superior rectus (more concern with securely attaching to superior oblique tendon) or more than 7 mm for inferior rectus.

- IV. If strabismic after BMR (overcorrection or undercorrection), consider a re-recession (if moderate to begin with, 5 mm or less and still ET) or advance medials if XT.
- V. If strabismic after a BLR, go after the medials because a recessed lateral sticks to the inferior oblique and can be messy surgery to work on LR.
 - Exception: if initial surgery was BLR, then bilateral inferior oblique probably okay.
- VI. Don't overcomplicate simple patients:
 - A. Medium to larger horizontal strabismus with a small vertical component: Generally can ignore the vertical in kids who have an ability to fuse!
 - B. This is especially relevant for intermittent exotropia, because they frequently fuse at near.

VII. Nystagmus

- A. The fixating eye drives the head position.
- B. If child is also strabismic, adjust the surgical dose on the nonfixating eye only (as the fixating eye drives the head position), and the non-fixating eye is along for the ride.
- C. Null position surgery is more difficult if they already had strabismus surgery. Sit down and do the math for the surgical plan based on surgical tables, aware that you may need to modify intraoperatively.
- VIII. Use a Moody forceps after your first cut (start superiorly) of the muscle stump to keep the conjunctiva out of the way.

Cut horizontal muscles from superior to obviate the need for an assistant.

Lessons From Melbourne

Lionel Kowal MBBS

- Injecting OnabotulinumtoxinA
 - Mendonca forceps: a non-EMG technique¹
- Bimedial elevation (BME) in absence of A-pattern²
 - This creates a V pattern.
 - BME for CI: ... for the near XT'. LR recess OU for the distance XT
 - BMR with BME for ET, D>N if there is a threat of XT'
- Inferior oblique surgery
 - Recess: Use only 1 suture through anterior corner to reduce risk of anti-elevation.
 - Adjustable inferior oblique recession³
- Partial tenotomies to treat torsion^{3,4,5},
 - ... of vertical recti for in-/ex- cyclotropia
 - ... of ant ½ of IO insertion for excyclotropia
 - ... of ant ½ of SO insertion for incyclotropia
- A technique for topical surgery
- When you can (unexpectedly) do resection/plicate
 - Resection in thyroid eye disease⁶ and in Duane's⁷
- Sagging lateral rectus: Problems with superior myopexy of the LR
- When to do BMR in childhood nystagmus with orthotropia⁸
 - Look for convergence null for distance in infantile and periodic alternating nystagmus
- Consecutive XT⁹
 - Medial rectus stretched scar: Use nonabsorbable sutures
- Duane's^{4,10,11}
 - Superior rectus transposition/inferior rectus transposition
 - The medial rectus pulley suture for convergence excess^{12,13}
 - LR transposition and IR recess for monocular elevation deficiency¹⁴
 - Plication instead of resection 12

Thank you to:

- 1. Tomas Mendonca
- 2. Ed Buckley
- 3. Alan Scott
- 4. Federico Velez
- 5. Ken Wright
- 6. Del Monte & others
- 7. Stephen Kraft
- 8. Annette Spielmann
- 9. Irene Ludwig
- 10. Cumhur Sener
- 11. David Hunter
- 12. Joe Demer
- 13. Robert Clark
- 14. Ramesh Kekkunaya
- ...& many others

How Many Roads Must a Doc Walk Down? Pearls From My Years in Practice

M Edward Wilson MD

- I. Aphakic Contact Lens Management: Put the Lens On at the Completion of Surgery
 - A. When choosing to leave an infant aphakic at surgery, consider placing a silicone elastomer contact lens on the operating table at the conclusion of surgery and leaving it untouched for the first post-operative month. No patch or shield is needed since the contact lens acts as a bandage and the incisions are very small and are sutured.
 - B. To calculate the needed contact lens power, I perform A-scan ultrasound biometry in the operating room just before the surgery starts. I use 111.9 as the lens constant and use a common IOL formula like the Holladay, SRK/T, or Barrett.
 - C. The rule of thumb for base-curve of the silicone elastomer contact lens is to use a 7.5 base curve in infants and a 7.7 base curve after 18-24 months (modify based on K-readings when needed, but rarely is modification from the above rule of thumb needed).
 - D. Leave the contact lens in place day and night and use the postoperative drops as prescribed. At the 1-month postoperative visit, one of our technicians teaches the parents how to insert, remove, and clean the contact lens. After that visit, the parents remove the contact lens weekly.
- II. Secondary IOLs: Plan Ahead
 - A. Plan the first surgery with the second surgery in mind. The result is that most secondary IOLs can be placed within the capsular bag.
 - B. Placing an IOL in the ciliary sulcus is not bad, but late decentrations are more common than you think. Consider optic capture for stable centration when the sulcus is chosen.
- III. Vitrectomy: Highest Cut-Rate (7500 or More Cuts/ Minute) for Safety
 - A. Vitrectomy doesn't cause retinal holes and detachment; vitreoretinal *traction* causes retinal holes and detachment. Learn to use proper technique and the highest cut-rate available. I now use the same cut rate for cutting capsule and for cutting vitreous. There is no need to vary the cutting speed.
 - B. *Never ever* place a Weck-Cel sponge in contact with vitreous. If vitreous presents at the wound, hold the vitrectomy cutter up to the wound to safely amputate vitreous without traction. Do not pull on vitreous.
 - C. Learn to do bimanual surgery. Separate the infusion from the cutting handpiece; Venturi-pump machines work best for what we do.

- D. Use tight-fit incisions for less chamber bounce. This leads to less inflammation postoperatively.
- E. Become comfortable switching hands. Practice placing the vitrector or aspirator in your nondominant hand. When you need it in a tough case is not the time to learn.
- F. Become comfortable with both the anterior and the pars plana approach. Trocars are for tool transfer; just use an MVR blade and don't bother with a trocar.
- G. Check for vitreous using intracameral preservativefree triamcinolone whenever you can.
- IV. Pre-existing Incompetent Posterior Capsules Are Common in Children
 - A. When in doubt, don't hydrodissect.
 - B. Look for the "fishtail sign" intraoperatively.
 - C. Make a careful anterior capsulorrhexis; it may be the only support you have by the end of the surgery.
 - D. When doing bimanual irrigation and aspiration, substitute the vitrector for the aspiration handpiece if there is any chance you will encounter pre-existing lens/vitreous admixture.
- V. Wound Management: Use 10.0 Vicryl
 - A. Incisions often leak in pediatric eyes, even wellmade incisions.
 - B. Soft tissues mold into the round shape of the instrument placed through them.
 - C. Suture the operative wounds (10.0 Vicryl) and even add a sealant when needed.
 - The exceptions are wounds that only iris hooks or forceps have entered; these can often be left unsutured
- VI. Parental Compliance With Postoperative Drops Is Variable
 - A. For more consistent healing, consider using intracameral or intracanalicular delivery options for steroids to reduce reliance on compliance. These are rapidly becoming commonplace for cataract surgery in children (triamcinolone or dexamethasone). I no longer use subconjunctival injections.
 - B. My anti-inflammatory drop of choice is prednisolone acetate 1% given q.i.d. for 4 weeks. Taper if and when you prefer.
 - C. Intracameral preservative-free moxifloxacin is my preferred antibiotic, and my surgery pharmacy prepares a ready-to-inject syringe for each case: 500 µg in 0.1 cc using undiluted nonpreserved moxifloxacin (Vigamox).

How Groovy Gadgets Can Improve Pediatric Cataract Surgery

Kanwal K Nischal MBBS

The world of pediatric cataract surgery evolves continually. Recent advances have allowed for better teaching and understanding and ultimately for delivery of better outcomes. In this talk the use of intraoperative OCT will be discussed for teaching of pediatric cataract surgery and for improved outcomes by better understanding the anatomy of the cataract being dealt with. Specifically, the focus will be dealing with intumescent cataracts, understating why YAG laser capsulotomy can be so difficult in children under the age of 8 years, and visualizing vitreous without the use of intraocular air or other substances. Femtosecond laser and nanopulse capsulotomy techniques have been described with variable popularity and success. Lastly, the BIL (bag-in-the lens) has shown a much reduced visual axis opacification frequency after pediatric cataract surgery and will also be discussed.

"I've Got You, Babe": IOLs in Children Under 2 Years of Age

Ramesh Kekunnaya MD FRCS

IOL implantation in children under 2 years of age has been discussed extensively in different publications from around the world, including consensus statements arrived at through the Delphi process. While there is a fair agreement on IOL implantation in children under 2, IOL implantation in infants remains controversial.

The Infant Aphakia Treatment Study (IATS) studied the long-term outcomes of IOL implantation in unilateral cataract and favors no implantation in children with unilateral cataract. One should be aware that unilateral cataract eyes, in general, are small in size and mostly have associated anterior segment dysgenesis or angle or iris abnormalities and are thus prone to more intra/postoperative complications. One should be very selective when deciding to proceed with IOL implantation in unilateral cataract. However, the scenario in bilateral cataract is totally different.

Based on our experience and publications by fellow Indian ophthalmologists, it appears there is a preference for IOL implantation in children under 2 years, including infants. However, one should be able to identify circumstances where IOL implantation needs to be avoided. The criterion for IOL implantation should be based on axial length, corneal diameter, associated anterior segment or angle anomalies, intraoperative new findings/complications, follow-up compliance, socioeconomic status, and surgeon experience.

This talk will cover various determining factors for IOL implantation in patients under 2 years of age, long-term surgical outcomes, and intercontinental differences in preferred practice.

Selected Readings

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Traumatic Cataracts: Special Techniques for the Capsules

Kanwal K Nischal MBBS

When an intraocular implant can be placed in a case of traumatic pediatric cataract, the challenge is always to place the IOL in such a way that it does not move forward with time and cause lens-iris capture, at best, or lens dislocation into the anterior chamber, at worst. Techniques used to do this have involved the use of posterior capsule optic capture when the posterior capsule is intact but the anterior capsule is ruptured or penetrated. However, when both anterior and posterior capsules have been breached, the situation can be challenging.

The recently described "banded technique" involves manipulating the anterior capsule rrhexis in such a way as to create a band of the anterior capsule to hold the IOL in the bag. Familiarity with the 2-incision push-pull (TIPP) rrhexis technique is helpful but not essential to perform the banded technique. This technique will be demonstrated through schematics and videos to allow the audience to add this technique to their armamentarium for dealing with pediatric traumatic cataracts.

Persistent Fetal Vasculature Cataracts

M Edward Wilson MD and Rupal H Trivedi MD MSCR

- The extent of the vascular anomaly in an eye with persistent fetal vasculature (PFV) directly influences the surgical approach and prognosis.
- Any child with a unilateral cataract, especially when associated with a microphthalmic globe, should be suspected of having PFV.
- Vitrectomy instrumentation is used to remove the posterior lens capsule, abnormal membrane, and anterior vitreous as needed. Intraocular scissors and intraocular cautery are also used as needed.
- The rehabilitation of vision may be further facilitated using an IOL if PFV is not associated with stretched ciliary processes or tractional retinal detachment.
- Approximately 50% of patients undergoing surgery for PFV will achieve useful vision. Visual acuity outcomes in patients with PFV are correlated with the nature and extent of ocular risk factors. Some patients may not be candidates for surgery because of either minimal changes or advanced posterior disease that limits the potential for visual improvement.¹
- Aphakic infants with mild PFV treated with a contact lens had a greater incidence of adverse events after lensectomy compared with children with other forms of unilateral congenital cataract; nevertheless, similar visual outcomes at 1 year after surgery were obtained.²
- Favorable outcomes were more often achieved in anterior PFV, compared to when posterior involvement was seen. Surgical outcomes in eyes with PFV undergoing cataract surgery are limited by intraoperative and postoperative complications such as hyphema, vitreous hemorrhage, recurrent visual axis opacification, glaucoma, and retinal detachment.³

- PFV is a spectrum and if defined broadly, it is significantly more common than previously reported. Defined this way, overall outcomes are comparable, on average, to that for congenital and infantile cataracts overall.⁴
- Eyes operated for severe PFV with elongated ciliary processes are unlikely to have a final visual acuity greater than 20/200, and many will need additional surgery. Postoperative visual axis opacification occurred in 60%, and glaucoma developed in 18%.⁵

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- Warren N, Trivedi RH, Wilson ME. Persistent fetal vasculature with elongated ciliary processes in children. Am J Ophthalmol. 2019; 198:25-29.

Uveitis and Cataracts

Ramesh Kekunnaya MD FRCS

Cataract management in children with uveitis remains challenging and needs special attention. Chronic inflammation and steroid use are the two important causes for the development of cataract. There are conflicting reports with regard to timing of cataract surgery, techniques of surgery, IOL implantation, type of IOL, and long-term outcomes. This talk will cover these issues, with emphasis on consensus on timing of surgery and choice of IOL and the result of long-term surgical outcomes.

Selected Readings

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How to Handle Superior Oblique Weakening Procedures

Joseph L Demer MD PhD, David G Morrison MD

Advocacy of Preoperative Imaging for Complex Strabismus: Strabismus Following Melanoma Brachytherapy in Patient P.O.

Joseph L Demer MD PhD

Patient

A 63-year-old white woman complaining of vertical, binocular diplopia when looking up since plaque brachytherapy of superiorly located choroidal melanoma in her right eye that required "extirpation and hang back of the superior rectus muscle," according to the operative report.

Clinical examination reveals large right hypotropia increasing in sursumversion, but the patient is orthotropic in far deorsum version and so adopts a large chin-up compensatory head elevation that causes her chronic neck pain.

Differential Diagnosis

- Actual extirpation of significant part of right superior rectus muscle
- 2. Radiation necrosis to right superior rectus muscle, possibly from plaque
- 3. "Lost" or slipped right superior rectus muscle
- 4. Restriction by scar in the superior orbit
- 5. Something else

Surface coil MRI was repeated in multiple planes and in multiple gaze positions using fixation targets:

- 1. T1 imaging demonstrates contrast-enhancing atrophic choroidal melanoma just posterior to equator, best seen in the quasi-sagittal plane.
- 2. Right superior rectus muscle has normal size and is contractile with supraduction effort.
- 3. Right superior rectus tendon ends posteriorly compatible with disinsertion from sclera.
- 4. No muscle atrophy or radionecrosis is evident.

Strabismus Surgery

Findings

Exploration demonstrated disinsertion of the right superior rectus muscle, found adherent to the posterior aspect of the reflected superior oblique tendon. The superior rectus muscle was split longitudinally for about 10 mm posterior to its anterior termination.

Operation

- Recovery of disinserted right superior rectus muscle and reattachment on adjustable suture
- 2. Release of incarcerated right superior oblique tendon
- 3. Repair of longitudinal split right superior rectus muscle

Postoperative Result

- 1. Orthotropic in primary position at 1 week
- Orthotropic in primary position at 7 weeks, but moderately limited supraduction with some hypotropia in sursumversion

Teaching Points

- MRI can distinguish or reveal some complex alternative pathologies:
 - Atrophy, inflammation, or avascularity
 - Hypertrophy (thyroid eye disease)
 - Abnormal contractility (paralysis or mis-innervation)
 - Gross muscle damage (endoscopic entry, partial excision, etc.)
 - Gross muscle displacement (sag or heavy eye phenomenon)
- MRI *cannot* distinguish the following:
 - Scarring near the insertion or sclera
 - Exact location of tendon
 - Path of SO anterior SO tendon
- Irradiated extraocular muscles are very friable and may disintegrate on manipulation.

How to Handle Superior Oblique Weakening Procedures

David G Morrison MD

Case

A 14-year-old girl presented with A-pattern exotropia with right superior oblique overaction and right hypotropia. Past ocular history was significant for infantile esotropia of 45 PD. She underwent bilateral medial rectus recession of 6 mm at 1 year of age. She had done well with good alignment until recently. She had no other relevant medical history.

Surgery

She underwent right superior oblique Z-tenotomy with good result and resolution of abnormal head posture.

Strabismus Exam

Method: Alternate Cover

Correction: sc

Distance		Near		Near +3I	DS		N Bifocals	3
		XT' 12 LHT' 10)					
	0	0	0	XT 4 RhypoT 6-8	+	0	0	
X(T) 8 Rhypo 4	0		0	XT 8 Rhypo 10	0		0	X(T) 14 Rhypo 14
	0	0	+2	XT 12 Rhypo 12	-2	0	0	
R Tilt	L Tilt							

AHP: Yes: Right head

Figure 1.

Method: Alternate Cover

Correction: cc

Distance	Noar		N	Near +3DS		N Bifocal		
XT 5 RhypoT flick	Near XT' 7 DVD OU		IN.	eai +3DS		bilocal	5	
Tanype Timen	0	0	-1	XT 4	0	0	0	
XT 7 SmallDVD OU	0		0	XT 5	0		0	XT 4 Small DVD
	0	0	+	XT 6	0	0	0	OU
R Tilt	L Tilt							

AHP: small right head tilt

Figure 2.

Superior Oblique Z-Tenotomy

Marginal Z-tenotomy for superior oblique weakening was largely abandoned due to poor control of weakening when the tendon was cut using scissors. However, tendon weakening using a monopolar electrode microdissection needle was recently published. This technique has yielded improved results.

A total of 20 patients (mean age: 9.8 years; age range: 3-34) underwent bilateral superior oblique Z-tenotomy during the study period: 2 patients (10%) with esotropia and 18 with exotropia (90%). Average decrease in pattern was 16^{Δ} (range: 0^{Δ} to 32.5^{Δ}). Success rate for pattern collapse was 78%, with resolution of overdepression in adduction of 90%.

However, additional evidence in a basic model demonstrates that Z-tenotomy up to 50% progressively reduces extraocular tendon force transmission, but Z-tenotomy of \geq 50% is biomechanically equivalent in vitro to complete tenotomy.²

Pitfalls and pearls for this technique and alternative approached will be discussed.

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Imaging in Planning for Complicated Strabismus or Reoperation

Joseph L Demer MD PhD, Federico G Velez MD

Complicated Strabismus After Nonstrabismus Surgery

Federico G Velez MD

A 61-year-old man with a history of bilateral glaucoma more severe in the right eye. Past surgical history includes right eye trabeculectomy + mitomycin C, subsequent multiple needling, superotemporal Baerveldt drainage device implantation. Patient complains of constant vertical and torsional diplopia following the last surgery. Diplopia has been stable since the operation. Tried Fresnel prisms but patient could not tolerate them because he was still seeing torsional diplopia. Uncorrected visual acuity measured OD 20/60, OS 20/20.

RHT 20 RHT 20 RHT 14 RHT 25 RHT 4

Incyclotropia 7 degree

Figure 1.



Figure 2.

Patient was unable to fuse on free space prism test due to torsion.

Patient wanted surgery only in his right eye. He and his glaucoma treating physician did not want to remove the shunt due to his advanced glaucoma. Surgery consisted of a combined right eye superotemporal shunt scar tissue formation removal with lysis and removal of fibrotic tissue and right superior rectus muscle recession 5 mm to a position 12 mm posterior to the limbus.



Figure 3. One week postoperative follow-up right superior rectus muscle recession 5 mm.

Postoperative evaluation at 1 month demonstrated no improvement.

RHT 20 RHT 25 RHT 20 RHT 20 RHT 25

Incyclotropia 8 degree

Figure 4.



Figure 5.

Patient underwent a second strabismus surgery. Intraoperative forced duction test revealed no restriction to downward rotation. Surgery consisted of a right inferior rectus muscle resection 5.5 mm. Initial postoperative examination revealed some improvement and no diplopia up gaze.

RHT 8

RHT 20 RHT 12 RHT 25

RHT 12

Incyclotropia 8 degree

He underwent a third strabismus surgery. Intraoperative forced duction test was negative. Patient underwent an additional right inferior rectus muscle resection 5 mm.



Figure 7. One day postoperative follow-up right inferior rectus muscle resection 5 mm.

He was able to see single in extreme downgaze, but his diplopia was present everywhere else. He would like to proceed with additional surgery.

RHT 18 RHT 25 RHT 14 RHT 12 RHT 5

Incyclotropia 7 degree

Figure 8.



Figure 9.



Figure 10. Negative intraoperative forced duction test.

Surgery consisted of Trabectome, scar tissue removal, Baerveldt implant removal, right superior oblique muscle reposition, right superior rectus muscle recession 5 mm from 12 to 17 mm from the limbus, and superior symblepharon excision with amniotic graft.

At the patient's last postoperative follow-up 12 months after his last surgery, his diplopia was resolved. His motility examination measured orthotropia in all gazes, no torsional component on Maddox rods and stereopsis of 100 seconds of arc at near.

References

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- Methods: 195 adult patients with glaucoma prospectively enrolled. Results: Diplopia was reported in 41 of 195 medically and surgically treated patients (21%). Binocular diplopia 11 of 47 patients (23%) after glaucoma drainage device (GDD) (95% CI, 12-38), 2/61 [3%] after trabeculectomy (95% CI, 0.4-11), P = .002. The most common type of strabismus was hypertropia (10/11 GDD cases, 2/2 trabeculectomy cases). Conclusions: Diplopia may be under-recognized. Diplopia was more commonly seen after GDD than trabeculectomy, typically a noncomitant restrictive hypertropia. It is important to counsel patients on the higher occurrence of diplopia associated with GDD surgery.
- Osigian CJ, Cavuoto KM, Rossetto JD, et al. Strabismus surgery outcomes in eyes with glaucoma drainage devices. *J AAPOS*. 2017; 21(2):103-106.
 - Methods: Retrospective review of patients who underwent strabismus surgery after implantation of a GDD over a 13-year period. **Results:** 16 patients were included; 14 had exotropia $(34\Delta \pm 16\Delta)$, 11 had vertical deviations ($15\Delta \pm 7\Delta$), 9 had concurrent exotropia and vertical deviations. Nine patients had diplopia. The surgical approach was tailored to address the deviation most noticeable to the patient. Three patients underwent simultaneous horizontal and vertical surgery. All patients underwent strabismus surgery on an eye with a GDD. Surgical motor success (horizontal deviation $\leq 10\Delta$, vertical $\leq 4\Delta$) was achieved in 42% of horizontal and 57% of vertical deviations. Diplopia resolved in 50% of patients who presented with preoperative diplopia. Only 2 required a second strabismus surgery. Conclusions: Strabismus surgery with preservation of the filtering bleb following implantation of a glaucoma drainage device is a low-risk procedure that can improve ocular alignment and related symptoms, despite a low motor success rate by standard criteria.
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Vertical Kestenbaum Procedures

Monte A Del Monte MD and Sean P Donahue MD PhD

Surgical Management of Vertical Null Point Nystagmus

Monte A Del Monte MD

Introduction

Various surgical procedures have been presented to attempt to correct the sometimes very large chin-up or chin-down posture in patients with nystagmus and a vertical null zone. Most of the references in the literature report a combination of small vertical rectus surgery combined with various oblique muscle weakening procedures. There has been a fear of larger vertical rectus strengthening and weakening procedures because of a perceived risk of alteration of eyelid position or torsion.

Discussion

The author has found that patients with vertical null zone and chin-up or chin-down posture respond better to very large vertical rectus muscle surgery with minimal risk of overcorrection, changes in eyelid position, or subjective torsion. In the late 1980s we began to treat these patients with very large bilateral vertical rectus muscle recessions plus/minus very large bilateral vertical rectus muscle resections, often called a vertical Kestenbaum/Anderson procedure. In 2004 we published our initial experience with this approach, with excellent results and few complications. We concluded that combined large symmetrical bilateral vertical rectus muscle recession plus large symmetrical bilateral vertical rectus muscle resections (for postures greater than 25 degrees) successfully correct both chin-up and chindown postures secondary to vertical null zone nystagmus. We found that a total of up to 20-22 mm of total combined recession and resection on each eye was required to correct postures of 40 degrees or greater measured with a goniometer. In our series there were no cases of induced strabismus, no significant asymmetrical change in eyelid position, no loss of vision (3/21 patients improved 1-2 lines), no symptomatic torsion (<5 degrees of objective torsion in 6/21 patients measured), and no overcorrections. Results remained stable over an average followup period of 36 months (range: 12-66 months).

Selected Reading

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Vertical Kestenbaum Procedures

Sean P Donahue MD PhD

- I. Infantile Nystagmus Syndrome Characteristics
 - A. Typically horizontal nystagmus
 - B. Null position to blunt nystagmus
 - C. Planes for null position
 - 1. Horizontal (Yaw plane)
 - 2. Vertical (Pitch plane)
 - 3. Torsional (Roll plane)
 - 4. Multiplanar
 - D. All have associated torticollis to facilitate null.
- II. Surgical Options to Correct Torticollis
 - A. Horizontal
 - 1. Kesterbaum procedure
 - 2. Anderson procedure
 - B. Vertical (chin-up or chin-down)
 - 1. With horizontal nystagmus
 - 2. With vertical nystagmus
 - C. Torsional (not covered here)
 - D. Multiplanar: Do the most prominent (typically horizontal) and see what happens.
- III. Surgical Treatment of Chin-Down
 - A. Need to rotate eyes down
 - B. Options
 - 1. Bilateral IR resection with bilateral SR recession
 - a. I did 8 mm of each.
 - b. Typically corrected AHP initially
 - c. Began to see recurrence at 2-3 years
 - d. 50% incidence of V-pattern ET likely due to abducting effects of vertical recti
 - e. I no longer do this.
 - 2. BSR recession with bilateral IO myectomy
 - a. Lower recurrence
 - b. No induced pattern
 - c. Now my preferred procedure

- IV. Surgical Treatment of Chin-Up
 - A. Much rarer
 - B. Need to move eyes to upgaze
 - C. BIRc plus BSRs
 - 1. Prefer in fusing patients
 - 2. No pattern but small sample size
 - D. Consider BSO tenotomy in nonfusing but worry about causing V-pattern

Third Nerve Palsy Surgery

Monte A Del Monte MD and Sean P Donahue MD PhD

A Novel Technique to Manage Strabismus and Ptosis in Patients with Partial Third Nerve Palsy with Aberrant Regeneration

Monte A Del Monte MD

Case Report

A 7-year-old otherwise healthy girl presents with outward deviation of her left eye and drooping of her left eyelid since early infancy. Visual acuity: 20/20 OD and 20/50 OS. External exam revealed a moderate left ptosis with good lid crease and moderate levator function on attempted up gaze. The ptosis increases on left gaze and disappears on attempted right gaze. Ocular motility testing reveals a 30-35 PD left exotropia and 15 PD left hypotropia in primary gaze. The exotropia increases in right gaze to 50 PD and decreases to 20 PD in left gaze. The left hypotropia increases in left gaze and almost disappears in right gaze. Pupillary exam was normal without afferent pupillary defect. Ductions and versions reveal -3/-4 limitation of adduction OS as well as -1/-2 limitation to elevation and -1 limitation to depression. The remainder of the ophthalmic exam was normal.

Discussion

The history and exam presented above is most consistent with congenital partial left third nerve palsy with aberrant regeneration of the right medial rectus to the levator of the left eye. Mild to moderate left amblyopia is present from disuse. The most common treatment regimen for this patient would be to perform a recession of the left lateral rectus and resection of the left medial rectus for the exotropia and then either supra placement of the horizontal rectus muscles to near the superior rectus, plication of the superior rectus or recession of the left inferior rectus for the left hypotropia, with the increased risk of anterior segment ischemia. Then, after healing, a ptosis procedure can elevate the left eyelid to clear the pupil, with a risk of overcorrection with exposure or undercorrection.

We have described a novel surgical approach to this patient which can correct both the strabismus and ptosis with a simple single surgical procedure to correct both the ptosis and strabismus with a more predictable result and lower risk of complication. Using the aberrant regeneration to our advantage, instead of performing a recess/resect procedure on the left eye to correct the exotropia, we perform a recession of the right lateral rectus and resection of the right medial rectus to intentionally adduct the right eye to align with the left. Then, when the patient abducts the right eye to fixate with the preferred right eye in primary gaze, by Hering's law, the left eye will adduct as well and the left eyelid will elevate secondary to the aberrant regeneration. Often, as in this case, there is also some aberrancy of the

medial rectus to the superior rectus as well, which also corrects the left hypotropia. This combined correction can prove very useful in selected cases.

Reference

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Third Nerve Palsy Surgery

Sean P Donahue MD PhD

- I. Third Nerve Palsy
 - A. If complete, only 2 muscles work.
 - 1. Superior oblique
 - 2. Lateral rectus
 - 3. Very difficult to manage
 - B. Incomplete
 - 1. If versions full, one can do large R and R.
 - 2. If versions limited, operating on the fellow eye does *not* work.
 - C. If ptosis complete, consider not treating.
 - D. Surgical options for complete palsies
 - 1. Lateral rectus disinsertion
 - 2. Lateral rectus to periosteum
 - 3. Superior oblique transposition
 - 4. Lateral rectus splitting transposition
 - E. Lateral rectus extirpation with large medial rectus resection
 - 1. Good stability
 - 2. Quite powerful
 - 3. Allows some abducting force
- II. Technique
 - A. Large peritomy
 - B. Hook and clean LR to entrance to Tenon capsule
 - C. Suture through LR at insertion with locked poles (facilitates extirpation)

D. Disinsert muscle

- E. Lift muscle with suture, clamp posteriorly
- F. Remove cut stump and cautery
- III. Preliminary Data
 - A. Ten patients; 2 bilateral
 - B. One year follow-up
 - C. Two reoccurrences
 - D. No overcorrection
 - E. Able to fuse with small AHP

Complicated Strabismus After Non-strabismus Surgery

David G Morrison MD, Federico G Velez MD

NOTES

Perforations and Infections

Malcolm R Ing MD

The author will present his review of perforations and infections following strabismus surgery.

Perforation

Perforation of the sclera with or without perforation of the retina is believed to be the portal of entry of bacteria in endophthalmitis following strabismus surgery. There are two types of perforations, namely, occult perforations, which are found on inspection of the peripheral fundus with the indirect ophthalmoscope after the case has been completed, and recognized scleral penetrations, in which the strabismus surgeon realizes that the needle that was utilized to suture the muscle onto the sclera has penetrated too deep. Some scleral penetrations do not involve the retina. Retinal perforations are found to occur, but at a lower rate than scleral penetrations.

There is low incidence of recognized scleral penetrations: 1/1,000 as reported by Simon et al (1992). In contrast, the rate of occult perforations is reported to range from 9% to 12% of all strabismus surgeries in studies before 1990. Inadvertent penetration of the sclera was reported to be 2% by Cibis in 1992. A similar prospective study by Noel et al reported in 1997 that a scleral penetration occurred in 1.4% of cases. In this series, the incidence of retinal perforation was much lower, 0.4%. A prospective study by Dang et al in 2004⁴ found that scleral penetration occurred in 5% and perforation of the retina occurred in 3%. Thus, in these studies cited above, we have found that penetrations of the sclera, not necessarily with retinal penetration, occur in 2%-12% of all strabismus surgeries.

The incidence of endophthalmitis is far less than the incidence of scleral penetration, so it is obvious that not all penetrations are followed by endophthalmitis.

It is of note that most penetrations occur during a recession procedure, where the needle has to enter thinner sclera, rather than during a resection procedure, where the site of reattachment of a rectus muscle utilizes thicker sclera. Nevertheless, only 4 cases of the 22 endophthalmitis cases reported had known penetration.

Why should we be concerned about needle penetration?

Needles have been found to be contaminated in 15%-19% of cases despite the use of povidone iodine in the cul de sac before surgery. Furthermore, sutures have also been found to be culture-positive in 28% by Eustis et al despite the use of povidone iodine in the cul de sac. ⁵ This rate of contamination of needles and sutures can be reduced to 9% if the sutures are presoaked in povidone iodine.

Utilization of povidone iodine in the cul de sac before and after the insertion of the speculum is most efficient in reducing the bacterial count at the operative site. Although there has not been a prospective study to prove that povidone iodine is effective in reducing the infection rate following strabismus surgery, it would be reasonable to reduce the bacterial load at the operative site and on the needles and sutures utilized during the surgery at the incision site to the lowest percentage possible.

Endophthalmitis

An intraocular infection is the most feared complication, other than a cardiac arrest, following strabismus surgery. Ing found the estimated incidence to be 1/30,000 in his survey of 67 Costenbader Alumni members. Although rare, endophthalmitis is usually devastating to a good visual outcome. Signs and symptoms of endophthalmitis include malaise, loss of appetite, fever, eye pain, swelling of the lids (particularly when asymmetric), purulent discharge, hypopyon, cloudy vitreous, and decreased vision.

Indeed, in a recent review of 22 cases of endophthalmitis reported in the scientific literature, only 8 retained normal vision in the affected eye. The signs and symptoms of a possible complication developed during postoperative days 1-8 (mean: day 3). However, the definitive diagnosis of endophthalmitis was delayed in most cases and was usually made 3 days later in the postoperative period (day 1-14; mean: day 6-7). In this survey, all patients received intravitreal injections of antibiotics. Despite the intravitreal injections, by mean day 7, most of the eyes were blinded by the infection.

Since intraocular infections cannot be completely prevented, it is strongly recommended that the strabismologist do the following:

- Emphasize the signs and symptoms of infection with the patients or parents of patients, urging them to report any fever, swelling, excessive discharge, malaise or loss of appetite, and decrease in vision.
- Keep in touch with the parents, especially in out-of-town families. Since most parents have cell phones, they can send a photo to the surgeon for any questionable postoperative finding.

Orbital Cellulitis

Ing reported the incidence of orbital cellulitis to be 1 per 1000-1900.⁶

Twenty-five cases of this extraocular complication were investigated by the Periocular Infection Study Group.⁷ Lid swelling, especially; asymmetric proptosis; redness; and pain on attempted motion were signs present in all cases. Cultures revealed *Staphylococcus aureus* in most of the 25 cases.

Signs and symptoms developed in two-thirds of the patients by postoperative day 2, and patients were started on treatment after the development of signs and symptoms of orbital cellulitis. Three cases responded to oral antibiotics, but the majority required hospitalization and systemic antibiotics for resolution of the infection. Most of the cases were treated by IV cefuroxime or ceftriaxone.

With treatment, the prognosis reported in the PISG report, unlike that of endophthalmitis, was excellent. All infected eyes retained good vision, and the desired alignment was also achieved in 75% of the cases.

Subtenon Abscess

Another type of extraocular infection that can occur is a subtenon abscess. These infections are recognized by a painful lump developing over a sutured rectus muscle. The 6 cases reported in the literature were successfully managed by systemic antibiotics, with surgical drainage necessary in all cases for resolution.

Conjunctivitis Following Strabismus Surgery

Ing's study reported that the incidence of superficial infection was similar for those Costenbader Alumni who used antibiotics to that of those who did not.⁶ The author believes the majority of strabismologists still prefer to use a topical antibiotics in the postoperative management of their patients.

Summary

Infections cannot be completely prevented following strabismus surgery. However, the strabismus surgeon can minimize the risk of infection by reducing the bacterial load with a pretreatment of the surgical site, sutures, and needles with povodine iodine. The surgeon should try to avoid penetration of the sclera, and when this complication is suspected the surgeon needs to definitely examine the interior of the eye in the operative suite with a dilated pupil using the indirect ophthalmoscope. The key to successful management is early diagnosis and the practice of staying in touch with the parents and patients postoperatively, especially when families are from out of town.

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