Pediatric Ophthalmology 2018 Winds of Change in the Windy City

Program Directors Jonathan M Holmes MD and Scott A Larson MD

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

McCormick Place Chicago, Illniois Saturday, Oct. 27, 2018

Presented by: The American Academy of Ophthalmology



American Academy of Pediatrics



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2018 Pediatric Ophthalmology Subspecialty Day Planning Group

On behalf of the American Academy of Ophthalmology, the American Association for Pediatric Ophthalmology and Strabismus (AAPOS), and the American Academy of Pediatrics (AAP), it is our pleasure to welcome you to Chicago and Pediatric Ophthalmology 2018: Winds of Change in the Windy City.



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Planning Group iii

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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 in physician practices, resulting in the best possible eye care for their patients.

2018 Pediatric Ophthalmology Subspecialty Day Learning Objectives

This meeting will enable attendees to:

- Improve their ability to diagnose and manage pediatric ophthalmology and strabismus conditions
- Improve their outcomes in the management of pediatric ophthalmology and strabismus conditions
- Integrate new and existing clinical trial results into their management of childhood eye disease
- Apply new adult ophthalmology subspecialty developments to the care of children
- Apply the recent developments in therapeutic approaches to reducing myopia progression
- Use emerging developments in childhood anterior segment disease
- Apply new imaging and testing modalities in their care of children with eye conditions and patients with strabismus

2018 Pediatric Ophthalmology Subspecialty Day 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.

2018 Pediatric Ophthalmology Subspecialty Day CME Credit

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

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

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- Register in advance, receive materials in the mail, and turn in the *Subspecialty Day Syllabi* exchange voucher(s) onsite;
- Register in advance and pick up your badge onsite if materials did not arrive before you traveled to the meeting;
- Register onsite; or
- Scan the barcode on your badge as you enter an AAO 2018 course or session room.

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- 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, Level 2.5, and in the Academy Resource Center.

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

- Access at www.aao.org/mobile
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Pediatric Ophthalmology Subspecialty Day 2018: Winds of Change in the Windy City

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

SATURDAY, OCT. 27

7:00 AM	CONTINENTAL BREAKFAST		
8:00 AM	Welcome and Introductions	Jonathan M Holmes MD*	
		Scott A Larson MD	
Section I:	Weathering the Storm—Surgical Approaches to Vertica	l Strabismus	
	Moderator: Jonathan M Holmes MD*		
	Panelists: Sonal R Farzavandi FRCS, Jon Peiter Saunte MD*, Rosario Gomez De Liano MD, and Stacy L Pineles MD		
8:01 AM	Introduction	Jonathan M Holmes MD*	
8:06 AM	Drizzle or Downpour? Medium Angle Hypertropia	Jonathan M Holmes MD*	1
8:11 AM	Inferior Oblique Recession Alone	Sonal R Farzavandi FRCS	2
8:16 AM	Adding a Short-Tag Adjustable Inferior Rectus Recession	Jon Peiter Saunte MD*	3
8:21 AM	Panel Discussion		
8:26 AM	Low Pressure? Small-Angle Graves Hypotropia	Jonathan M Holmes MD*	4
8:31 AM	Topical Inferior Rectus Recession	Rosario Gomez De Liano MD	5
8:36 AM	Topical Marginal Tenotomy	Stacy L Pineles MD	6
8:41 AM	Panel Discussion		
8:46 AM	High Pressure? Small-Angle Hypertropia With Excyclotropia	Jonathan M Holmes MD*	7
8:51 AM	Superior Rectus Recession With Transposition	Rosario Gomez De Liano MD	8
8:56 AM	Leave That Superior Rectus Alone	Sonal R Farzavandi FRCS	9
9:01 AM	Panel Discussion		
9:06 AM	Twisting in the Wind: Residual Excylotropia After Harada-Ito	Jonathan M Holmes MD*	10
9:11 AM	Repeat Harada-Ito	Jon Peiter Saunte MD*	11
9:16 AM	Inferior Rectus Recessions	Stacy L Pineles MD	12
9:21 AM	Panel Discussion		
9:26 AM	Wrap-up	Jonathan M Holmes MD*	

Section II: Sudden Showers—Late Breaking RCTs and Observational Studies

	Moderator: Scott A Larson MD		
9:31 AM	Introduction	Scott A Larson MD	
9:32 AM	Glasses for Hyperopia	Donny Won Suh MD*	13
9:46 AM	Low-Dose Bevacizumab for ROP	Sharon F Freedman MD	15

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

10:00 AM	Natural History of Intermittent Exotropia	Christie L Morse MD*	16
10:14 AM	Binocular Treatment for Amblyopia	Lisa C Verderber MD	17
10:28 AM	Wrap-up	Scott A Larson MD	
10:29 AM	REFRESHMENT BREAK and AAO 2018 EXHIBITS		

Section III: Raining Cats and Dogs-Challenges of Applying Evidence-Based Medicine

	Moderator: Michael F Chiang MD*		
10:59 AM	Introduction: What Is an Evidence-Based Practice?	Michael F Chiang MD*	
11:04 AM	Evidence: Two vs. 6 Hours of Patching for Amblyopia	Michael X Repka MD MBA*	19
11:09 AM	Amblyopia in the Real World	Edward L Raab MD	20
11:14 AM	Rebuttal	Michael X Repka MD MBA*	
11:16 AM	Rebuttal	Edward L Raab MD	
11:18 AM	Evidence: Treating ROP Based on ETROP Guidelines	William V Good MD	21
11:23 AM	ROP in the Real World	R V Paul Chan MD*	22
11:28 AM	Rebuttal	William V Good MD	
11:30 AM	Rebuttal	R V Paul Chan MD*	
11:32 AM	Evidence: Treating Intermittent Exotropia Based on PEDIG and UK Studies	David K Wallace MD MPH*	24
11:37 AM	Intermittent Exotropia in the Real World	Burton J Kushner MD*	26
11:42 AM	Rebuttal	David K Wallace MD MPH*	
11:44 AM	Rebuttal	Burton J Kushner MD*	
11:46 AM	Conclusions: Value-Based Models and Outcome Measures	Michael F Chiang MD*	
11:53 AM	LUNCH and AAO 2018 EXHIBITS		

Section IV: Ice Breaker-What Our Adult Specialty Colleagues Can Teach Us

Moderator: Tammy L Yanovitch MD

12:53 PM	Introduction	Tammy L Yanovitch MD	
12:54 PM	Oculoplastics: Orbiting the Eye of the Storm	Julie A Woodward MD*	28
1:06 PM	Retina: Lightning Strikes	Michael M Altaweel MD*	29
1:18 PM	Neuro-Ophthalmology: A Head in the Clouds	Andrew G Lee MD	31
1:30 PM	Glaucoma: A High-Pressure System	Steven J Gedde MD*	32
1:42 PM	Cornea: The Tip of the Iceberg	Christina R Prescott MD	34
1:54 PM	Wrap-up	Tammy L Yanovitch MD	

Section V: Here Comes the Sun-Myopia Prevention

Moderator: Nils K Mungan MD 1:55 PM Advocating for the Profession and Patients Kenneth P Cheng MD 35 2:00 PM Introduction Nils K Mungan MD 2:01 PM The Myopia Epidemic Terri L Young MD MBA 38 2:06 PM Atropine for Myopia Donald Tan MD FRCS FRCOphth* 39

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

2018 Subspecialty Day Pediatric Ophthalmology		Program Schedule	xv
2:14 PM	Contact Lens Therapies for Myopia Control	Bruce H Koffler MD*	41
2:22 PM	Other Therapies for Myopia Prevention	Daniel Ian Flitcroft MD	42
2:30 PM	Wrap-up	Nils K Mungan MD	
2:31 PM	REFRESHMENT BREAK and AAO 2018 EXHIBITS		

Section VI: London Fog-What Am I Doing Differently in Pediatric Anterior Segment

	Moderator: Erick D Bothun MD		
3:01 PM	Introduction	Erick D Bothun MD	
3:03 PM	Newer Intraocular Instruments and IOLs	M Edward Wilson Jr MD*	44
3:13 PM	Genetics of Pediatric Cataracts	Ian Christopher Lloyd MBBS*	45
3:24 PM	Mainstreaming Corneal Crosslinking	Erin D Stahl MD*	46
3:35 PM	Topical Treatment for the Toughest Cornea Ulcer	Michael C Struck MD	47
3:46 PM	Use of IOL Optic Capture	Erick D Bothun MD	48
3:57 PM	Wrap-up	Erick D Bothun MD	

Section VII: Electrical Storm–Imaging in Pediatric Ophthalmology

	Moderator: Serena X Wang MD		
3:58 PM	Introduction	Serena X Wang MD	
3:59 PM	Corneal Topography in Children	Luca Buzzonetti MD	49
4:10 PM	Anterior Segment OCT: Corneal / Intraocular Surgery	Kanwal K Nischal MBBS*	50
4:21 PM	Anterior Segment OCT: Glaucoma and Other Indications	Yasmin Bradfield MD	51
4:32 PM	Retinal Fluorescein Angiography in Pediatric Uveitis	Jennifer Hsu Lee Jen Cao MD	52
4:43 PM	In-Office Handheld ERG in Pediatric Patients	Melanie A Schmitt MD	53
4:54 PM	Wrap-up	Serena X Wang MD	
4:55 PM	Closing Remarks and Adjourn	Jonathan M Holmes MD* Scott A Larson MD	

Drizzle or Downpour? Medium Angle Hypertropia

Jonathan M Holmes MD

History

- 72-year-old male patient
- Intermittent vertical diplopia, gradually worsening over last 15 years
- Fuses with prism but prefers not to wear glasses

Examination

- Visual acuity: 20/20 right eye, 20/30 left eye
- Mild right inferior oblique overaction
- Trace right depression deficit
- Distance prism and alternate cover test
 - 4 RHT 6 RHT 10 RHT
 - 8 RHT 14 RHT 14 RHT
 - 10 RHT 18 RHT 18 RHT
- Near prism and alternate cover test: 12 PD RHT 16 PD XT
- Double Maddox rod: 1 degree of excyclotropia
- Diagnosis: Right hypertropia due to presumed decompensated superior oblique palsy

Preoperative Testing

 In space, single with 10 PD BD (base down) over right eye at distance and near fixation

Intraoperative Testing

- Forced ductions: mild limitation of depression of right eye
- Torsional forced ductions:¹ symmetrical permissive excyclorotation 30 degrees in each eye
- Reduced permissive incyclorotation in right eye 15 degrees vs. 30 degrees left eye (tighter right inferior oblique by Guyton exaggerated traction test²)

Surgical Options (Audience Votes)

- 1. Right inferior oblique weakening alone
- 2. Left inferior rectus recession alone
- 3. Combined right inferior oblique weakening and left inferior rectus recession³
- 4. Right superior oblique strengthening
- 5. Other

Panel members present their recommended approaches, followed by discussion and audience revotes

- 1. Jung JH, Holmes JM. Quantitative intraoperative torsional forced duction test. *Ophthalmology* 2015; 122:1932-1938.
- 2. Guyton DL. Exaggerated traction test for the oblique muscles. *Ophthalmology* 1981; 88:1035-1040.
- 3. Nash DL, Hatt SR, Leske DA, et al. One- versus two-muscle surgery for presumed unilateral fourth nerve palsy associated with moderate angle hyperdeviations. *Am J Ophthalmol.* 2017; 182:1-7.

Inferior Oblique Recession Alone

Sonal Farzavandi FRCS(Edin)

The surgery of choice for a patient with decompensated unilateral superior oblique palsy and medium hypertropia (10-20 PD) in primary position is controversial. Nash et al¹ reported no clear advantage of 2-muscle surgery over 1-muscle surgery for motor outcomes, with success of diplopia correction being similar between the 2 groups. Rah and Kim² recently reported success on effect of isolated inferior oblique (IO) muscle recession for superior oblique palsy. Hence, with IO weakening surgery alone, IO recession or an IO graded anteriorization would be the procedure of choice.

- An IO recession acts by inducing muscle slack. This is achieved by moving the muscle insertion closer to the origin, up along the arc of contact of the muscle.³
- Graded anteriorization described by Wright⁴ changes the vector of forces by moving the muscle insertion anterior toward the inferior rectus (IR) insertion. The more anterior the new insertion, the greater the weakening effect. This changes the IO from an elevator to more of a depressor.

Surgical Steps and Points to Note

- Infero-temporal fornix incision
 - The lateral rectus is isolated on a Gass muscle hook. A 4-0 silk bridal suture is passed transconjunctivally through the hole in the toe of the Gass hook.⁵
 - The eye is rotated superonasally to expose the inferior temporal quadrant.
- The IO muscle is identified and hooked using the Stevens hook. The key is to hook the entire IO muscle by direct visualization of the posterior border of the muscle and to avoid the vortex vein by blind sweeping.
- The IO is delivered into the surgical field, keeping the tip of the hook up and the end of the hook down, to prevent the IO from slipping off the tip of the hook. Muscle slippage can split a muscle, a common cause of missed fibers and residual IO overaction. Hence it is important to check the quadrant for any missed fibers.
- The perimuscular fascial tissue and intermuscular septum are dissected, cutting close to the tip of the muscle hook. This exposes the edge of the IO muscle. Do not make deep cuts in the fornix as this will violate the posterior Tenon capsule, causing fat adherence and postoperative restriction.

- Dissect close to the muscle belly to remove intermuscular septum from above and below the IO muscle. The IO muscle is traced back to the insertion and disinserted.
- The IO is held with Moody forceps (one forceps is marked) to identify the anterior and posterior borders and secured with 6-0 Vicryl double arm suture.
- The IO is recessed or anteriorized based on the amount of IO overaction and the surgeon's choice.
- Recession is preferred in cases with small hypertropia (< 10 PD) in primary position and mild IO overaction (1+ to 2+).³
 - 1+ to 2+: IO recessed 3 mm posterior and 2 mm lateral to IR insertion
- Graded anteriorization is the method of choice for medium hypertropia (> 10 PD to < 20 PD) in primary position and moderate to severe IO overaction (3+ to 4+).³
 - 3+: 1 mm to 2 mm posterior to IR insertion
 - 4+: At IR insertion
- If there is residual hypertropia (more in downgaze) after the IO weakening surgery, the contralateral IR can be recessed at a later stage.

- 1. Nash DL, Hatt SR, Leske DA, et al. One- versus two-muscle surgery for presumed unilateral fourth nerve palsy associated with moderate angle hyperdeviations. *Am J Ophthalmol*. 2017; 182:1-7.
- 2. Rah SH, Kim JH. Invest Ophthalmol Vis Sci. 2018; 59: ARVO E-Abstract 2937.
- 3. Wright KW. Inferior oblique muscle weakening procedures. In: Color Atlas of Strabismus Surgery: Strategies and Techniques. 3rd ed. New York: Springer; 2007, ch. 17, 166-179.
- 4. Guemes A, Wright KW. Effect of graded anterior transposition of the inferior oblique muscle on versions and vertical deviation in primary position. *J AAPOS*. 1998; 2:201-206.
- Coats DK, Olitsky SE. Surgery on the inferior oblique muscle. In: *Strabismus Surgery and Its Complications*. Berlin: Springer-Verlag; 2007, ch. 11, 106-117.

Adding a Short-Tag Adjustable Inferior Rectus Recession

Inferior Oblique Weakening and Contralateral Recession of Inferior Rectus on a Hidden Adjustable Suture

Jon Peiter Saunte MD

Approach

In patients with IV nerve palsy and significant inferior oblique (IO) overaction, a weakening procedure of the ipsilateral IO is often indicated.¹ When the vertical deviation in primary position (PP) exceeds 15 PD, a single muscle procedure of IO transposition may not be sufficient, and a recession of the contralateral inferior rectus (IRc) on adjustable sutures may be added.² Late overcorrection of adjustable sutures in IRc can be avoided if a combination of nonabsorbable and absorbable sutures is used for the adjustable knots.

Details of Performing the Procedure

Different surgery techniques have been described. The IO graded transposition and the IRc can be performed in one combined procedure or staged. We prefer to perform the IO transposition under general anesthesia, and the IRc can be performed during the same procedure or under topical anesthesia at a later occasion.

In patients with \ge 15 PD hypertropia in PP, the combined IO transposition and IR recession may induce overcorrection.²

When performing the IO transposition, we use graded recession, suturing the IO muscle with double-armed 6-0 absorbable sutures. In recession of the contralateral IR, if the muscle is sutured with 6-0 absorbable sutures with a short-tag noose adjustable suture, 30% of patients in our cohort experienced late overcorrection at 6 weeks after surgery. Therefore we have changed to a double-armed nonabsorbable 6-0 hang-back pole suture (Mersilene) for the muscle; this is sutured to sclera 2-3 mm behind the original insertion in a double-diamond / crossed swords manner with an absorbable 6-0 (Vicryl) short-tag noose suture. The suture ends are kept long and pushed posteriorly along the IR muscle. The conjunctiva is closed with single 8-0 absorbable sutures with buried knots. The adjustable suture can be adjusted up to 7 days after surgery under topical anesthesia with eyedrops (oxybuprocaine and cocaine 4%) in the operation room with a small draping. We have not experienced any late overcorrections of IR adjustable surgeries after changing to a nonabsorbable pole suture, in concordance to Guyton's findings.3

- Chang MY, Coleman AL, Tseng VL, Demer JL. Surgical interventions for vertical strabismus in superior oblique palsy. Cochrane Database of Systematic Reviews 2017, Issue 11. Art. No.: CD012447.
- Nash DL, Hatt SR, Leske DA, et al. One- versus two-muscle surgery for presumed unilateral fourth nerve palsy associated with moderate angle hyperdeviations. *Am J Ophthalmol.* 2017; 182:1-7.
- 3. Awadein A, Marsh JD, Guyton DL. Nonabsorbable versus absorbable sutures in large, hang-back medial rectus muscle recessions. *J AAPOS*. 2016; 20(3):206-209.

Low Pressure? Small-Angle Graves Hypotropia Small Hypotropia Associated With Thyroid Eye Disease

Jonathan M Holmes MD

History

- 63-year-old female patient with history of thyroid eye disease
- Vertical diplopia with a twisting component
- Initially managed with Fresnel prism until stable (although struggling with torsion)

Examination

- Visual acuity: right eye 20/25, left eye 20/25
- Now no evidence of active thyroid eye disease
- Mild limitation of elevation of right eye, both in adduction and abduction
- Distance prism and alternate cover test:
 - 12 LHT 8 LHT 7 LHT
 - 12 LHT 8 LHT 8 LHT
 - 10 LHT 7 LHT 6 LHT
- Near prism and alternate cover test: 10 pd LHT
- Double Maddox rods: 10 degrees of excyclotropia¹

Preoperative Testing

- Forced ductions: limited elevation right eye
- To discuss role of intraoperative forced ductions, if planning surgical approach under topical anesthetic

Surgical Options (Audience Votes)

- 1. Recess right inferior rectus (RIR) under general anesthetic (with or without adjustable sutures)
- 2. Recess RIR under topical anesthetic with adjustable sutures
- 3. Temporal marginal tenotomy of RIR under topical anesthetic
- 4. Nasal marginal tenotomy of RIR under topical anesthetic
- 5. Recess both RIR and right superior oblique (RSO)
- 6. Other

Panel members present their recommended approaches, followed by discussion and audience revotes

Reference

1. Holmes JM, Hatt SR, Bradley EA. Masked superior oblique involvement in thyroid eye disease: avoiding postoperative A-pattern exotropia and intorsion. *J AAPOS*. 2012; 16:280-285.

Topical Inferior Rectus Recession Small Hypotropia Associated With Thyroid Eye Disease

Rosario Gomez de Liano MD

- Small amounts of vertical deviations in patients with thyroid eye disease (TED) often are fused spontaneously or by adopting head chin-up position or with small amounts of prisms. Bilateral surgery is often not necessary and may more likely result in overcorrection.
- Before surgery it is helpful to have an MRI or CT image to check involvement of all extraocular muscles to avoid overcorrections. In this case, although passive ductions were positive for left inferior rectus (IR) restriction, it is notable that the deviation does not increase significantly upward, nor in supra-abduction.
- Torsion in TED patients may result from multiple combinations of involvement of vertical and oblique muscles.
- A very large amount of excyclotorsion is suggestive of very tight bilateral IR, but it is not always present, particularly if the superior complex or the superior oblique muscles are enlarged. Medial wall decompression most often induces incyclotorsion and inferior wall excyclotorsion. It is important to measure it preoperatively and to evaluate how much it changes after recession of a vertical muscle.

Inferior Rectus Recession With Nasal Transposition Under Topical Anesthesia

- In this case we would perform a nasal transposition of the IR to improve excyclotorsion and recess it a very small amount (2-3 mm). Once the muscle is disinserted we can check for left inferior oblique tightness, which could also generate excyclotorsion.
- A transposition of half a tendon of the IR may correct 3-8° of excyclotorsion; and a full transposition, 8-15°.
- We use nonabsorbable suture to avoid slippage.
- We check changes in the torsion during surgery using limbal blue dots and adjust the amount of correction intraoperatively in the awake patient with double Maddox rods.

Selected Readings

- 1. Serafino M, Fogagnolo P, Trivedi RH, Saunders RA, Nucci P. Torsional diplopia after orbital decompression and strabismus surgery. *Eur J Ophthalmol.* 2010; 20(2):437-441.
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Topical Marginal Tenotomy Graded Vertical Rectus Tenotomy

Stacy Pineles MD

Approach

Graded vertical rectus tenotomy can be used in patients with small vertical deviations, less than 10 PD. The advantages of the procedure include the following:

- Can be performed easily under local anesthesia
- Fine adjustments can be made intraoperatively.
- No suturing, thus avoiding a possible foreign body reaction, eliminating the risk of globe perforation
- Overcorrections are extremely rare.
- Vessel sparing is often possible.
- Central partial rectus tenotomy has been performed transconjunctivally in the office.
- Graded temporal / nasal tenotomies can be exploited to correct laterally incomitant strabismus.

Details of the Procedure

- 1. A subconjunctival injection of lidocaine or eye-drop administration of lidocaine is administered.
- 2. A conjunctival incision is placed lateral and posterior to the muscle's insertion.
- 3. The muscle is engaged with a Jameson hook. A Stevens muscle hook is then used to reflect the conjunctiva and anterior Tenon capsule overlying the insertion of the muscle to expose the insertion.
- 4. Vessels overlying the portion of the muscle to be incised are cauterized.

- 5. An initial tenotomy of 30%-50% of the width of the tendon is performed.
- 6. The patient is helped to a sitting position. The patient's eyeglasses are positioned, and cover testing at distance and near is performed.
- 7. If the results of cover testing indicate undercorrection, further 5%-10% incremental cuts are made in the tendon, and then the alignment tested again. This is repeated until the patient has single binocular vision in the desired gaze positions.

Selected Readings

- 1. Yim HB, Biglan AW, Cronin TH. Graded partial tenotomy of vertical rectus muscles for treatment of hypertropia. *Trans Am Ophthalmol Soc.* 2004; 102:169-176.
- 2. Wright KW. Mini-tenotomy procedure to correct diplopia associated with small-angle strabismus. *Trans Am Ophthalmol Soc.* 2009; 107:97-103.
- Chaudhuri Z, Demer JL. Graded vertical rectus tenotomy for small-angle cyclovertical strabismus in sagging eye syndrome. *Br J Ophthalmol.* 2016; 100:648-651.
- 4. Chang M, Pineles SL, Velez FG. Adjustable small-incision selective tenotomy and plication for correction of incomitant vertical strabismus and torsion. *J AAPOS*. 2015; 19:410-416.

High Pressure? Small-Angle Hypertropia With Excyclotropia

Jonathan M Holmes MD

History

- 69-year-old female patient with intermittent vertical diplopia for many years
- Frustrated with prism glasses

Examination

- Visual acuity: right eye 20/20, left eye 20/20
- Mild limitation of depression of left eye, both in adduction and abduction
- Mild left inferior oblique (IO) overaction
- Distance prism and alternate cover test:
 - 10 LHT 10 LHT 8 LHT
 - 10 LHT 10 LHT 8 LHT
 - 10 LHT 10 LHT 10 LHT
- Near prism and alternate cover test: 10 PD left hypertropia
- Double Maddox rod: 3 degrees of excyclotropia

Intraoperative Testing

- Forced ductions; mild limitation to depression consistent with a slightly tight superior rectus (SR) muscle
- Qualitatively normal tension of left superior oblique (SO), assessed by Guyton exaggerated traction test¹
- Quantitative intraoperative torsional forced duction tests² revealed normal permissive excyclorotation and incyclorotation of 30 degrees each (right eye and left eye symmetric).

Surgical Options (Audience Votes)

- 1. Left SR recession alone
- 2. Left SR recession with temporal transposition
- 3. SO strengthening procedure
- 4. IO weakening procedure
- 5. Combined SO/IO procedure
- 6. Other

Panel members present their recommended approaches, followed by discussion and audience revotes

- 1. Guyton DL. Exaggerated traction test for the oblique muscles. *Ophthalmology* 1981; 88:1035-1040.
- 2. Jung JH, Holmes JM. Quantitative intraoperative torsional forced duction test. *Ophthalmology* 2015; 122:1932-1938.

Superior Rectus Recession With Transposition Small Angle Hypertropia With Excyclotropia

Rosario Gomez de Liano MD

CASE PRESENTATION

This 69-year-old female patient most likely initially had a left eye (LE) IV nerve palsy with contracture of the LE superior rectus (SR). Now the hypertropia is quite concomitant. It is important to note the LE superior oblique has normal tension and there is no restriction on rotatory torsional forced duction tests. We have evidence that the LE inferior oblique isn't tight, because she has a negative the rotatory passive incycloduction. Onset and course of diplopia, presence and magnitude of Bieslchowsky head tilt test, Hess screen, and passive ductions on the right eye (RE) will improve our diagnosis and help to differentiate from other clinical pictures.

It is important to check before surgery the patient's ability to fuse with prisms and synoptophore to determine whether torsion is a barrier to fusion. We would also like to evaluate objective torsion on both eyes (by fundus examination) and determine how much torsion is in the RE and how much in the LE when viewing binocularly.

Surgery

Because of the concomitance of this patient's deviation and small amount of torsion, we would perform surgery on a vertical rectus muscle under topical anesthesia that allows us to evaluate the vertical and torsional effects of the surgery intraoperatively.

In this case, one of several different surgeries may lead to a good result. Because passive duction reflects tight LE SR, we would perform a SR recession that will improve vertical deviation and Bielschowsky phenomenon. When excyclotropia is of small magnitude, we often do not need to address it. But because SR recession may increase the excyclotropia, we can diminish this effect by transposing the SR temporally half a tendon width. We use limbal blue dots to check changes in ocular rotation during surgery, once the SR is detached and also reinserted with transposition.

Selected Readings

- Khawam E, Ghazi N, Salti H. "Jampolsky syndrome": superior rectus overaction-contracture syndrome: prevalence, characteristics, etiology and management. *Binocul Vis Strabismus Q*. 2000; 15(4):331-342.
- Molinari A, Ugrin MC. Frequency of the superior rectus muscle overaction / contracture syndrome in unilateral fourth nerve palsy. J AAPOS. 2009; 13(6):571-574.
- 3. Kushner BJ. Multiple mechanisms of extraocular muscle "overaction." Arch Ophthalmol. 2006; 124(5):680-688.
- 4. Guyton DL. Exaggerated traction test for the oblique muscles. *Ophthalmology* 1981; 88:1035-1040.
- Jung JH, Holmes JM. Quantitative intraoperative torsional forced duction test. Ophthalmology 2015; 122:1932-1938.

Leave That Superior Rectus Alone

Sonal Farzavandi FRCS(Edin)

Small-Angle Hypertropia With Excyclotropia

In cases with unilateral superior oblique palsy, the ipsilateral hypertropia can transform into superior rectus contracture, giving rise to vertical restriction on down gaze.

This is a challenging problem, as the hypertropia is a compensatory mechanism of torsion. The contracture of the superior rectus gives rise to incyclotonus, which compensates and therefore leads to an underestimation of the real underlying torsion.

These patients with hypertropia and excyclotropia are often frustrated with prism glasses, as the torsion is the main barrier for fusion. The degree of excyclotorsion as determined by double Maddox rod may not be the exact amount of torsion present.

The presence of intolerance to prism glasses, elevation in adduction, limited depression in adduction, fundus photos and positive Bielchowsky head tilt test, positive forced duction test due to a slightly tight superior rectus muscle would support the presence of excyclotorsion.

Surgical Plan

Leave the superior rectus alone. If there was no torsion one could consider a small superior rectus recess (SRR).

In the presence of torsion, SRR is rarely useful and often dangerous. 1

In 1993 Quere et al² advised that SRR be associated with superior oblique resection in cases of decreased elongation of the superior rectus, while others observed induced objective paradoxical excyclotorsion after oblique muscle surgery, whether superior oblique strengthening or inferior oblique (IO) weakening was associated with SRR.³ Hence, leave that superior rectus alone. Graf and Weihs⁴ have shown that good results can be obtained with only oblique muscle surgery in cases of superior oblique palsy. They compared the surgical procedures in 51 acquired cases of superior oblique palsy: 28 with IO recess, 5 with superior oblique tuck, and 18 with combined superior oblique tuck and IO recession. They concluded that both Bielschowksy head tilt test and hypertropia on downgaze were more effectively reduced by SO tuck or combined oblique surgery than by IO recession alone.

My personal preference would be IO recession and if insufficient, adding a Harada Ito.

- Paris V. Unilateral superior rectus contracture related to extorsional syndrome: partner or enemy? Transactions of the 32nd European Strabismological meeting (ESA) 2008; 173-176.
- Quere MA, Pechereau A, Remy C, Bouchut P. Abnormal stretching of the ipsilateral superior rectus muscle in congenital superior oblique palsy. Transactions of the 21st ESA meeting. 1993; 193-198.
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- Graf M, Weihs J. Surgery for unilateral acquired trochlear nerve palsy: what's the best choice? Transactions of the 31st ESA meeting. 2007; 133-136.

Twisting in the Wind: Residual / Recurrent Excyclotropia After Harado-Ito

(After Previous Bilateral Superior Oblique Strengthening)

Jonathan M Holmes MD

History

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- 42-year-old female patient with history of neurosurgery at 20 years old, resulting in bilateral IV nerve palsies with torsional diplopia
- 14 years previous she had undergone bilateral superior oblique advancement (similar to Fells modification of Harada-Ito)¹ with dramatic improvement in her torsional diplopia.
- Over the last year, her torsional diplopia had returned and had been getting worse. She had now gone back to having to use a chin-down head posture.

Examination

- Visual acuity: right eye 20/20, left eye 20/20
- Minimal depression deficit in right eye in adduction
- Distance prism and alternate cover test:
 - 0 HT 0 HT 1 RHT
 - 0 HT 1 RHT 1 RHT
 - 1 RHT 4 RHT 6 RHT
- Near prism and alternate cover test: 1 RHT
- Double Maddox rod: 7 degrees excyclotropia in straight ahead and 12 degrees in downgaze

Preoperative Testing

Needed torsional correction to fuse house targets on synoptophore (at least 2 degrees in straight ahead and 5 degrees in downgaze)

Intraoperative Testing

Forced ductions: Mild limitation of elevation in adduction of each eye and mildly tight superior obliques by Guyton exaggerated traction tests² and by quantitative intraoperative torsional forced duction tests³ (20 degrees permissive excyclorotation)

Surgical Options (Audience Votes)

- 1. Unilateral Harada-Ito procedure
- 2. Bilateral Harada-Ito procedures
- 3. Bilateral inferior rectus recessions
- 4. Bilateral inferior oblique weakening procedures
- 5. Other

Panel members present their recommended approaches, followed by discussion and audience revotes

- Bata BM, Leske DA, Holmes JM. Adjustable bilateral superior oblique tendon advancement for bilateral fourth nerve palsy. *Am J Ophthalmol.* 2017; 178:115-121.
- 2. Guyton DL. Exaggerated traction test for the oblique muscles. *Ophthalmology* 1981; 88:1035-1040.
- 3. Jung JH, Holmes JM. Quantitative intraoperative torsional forced duction test. *Ophthalmology* 2015; 122:1932-1938.

Repeat Harada-Ito Residual/Recurrent Excyclotropia After Previous Harada-Itos

Jon Peiter Saunte MD

Approach

In patients with acquired bilateral IV nerve palsies with combined vertical and torsional diplopia, surgery strategy should include addressing the torsional component if this is a barrier to fusion. If torsional diplopia recurs after a successful initial Harada-Ito (H-I) procedure, prisms alone will not be sufficient, and repeat surgery to correct torsion should be considered. Several surgery strategies have been reported¹:

- 1. Bilateral repeat Harada-Ito procedures
- 2. Bilateral inferior rectus recession
- 3. Bilateral inferior oblique weakening procedure

In this outline, the repeat modified H-I procedure is discussed.

Details in Performing the Procedure

The preoperative exaggerated Guytons test² (Gt) combined with forced duction (FD) testing is important in planning of this type of surgery. In cases of tight superior oblique (SO) muscles resulting in < 5 degrees of excyclotorsion during Gt, a further tightening of SO may induce a limitation of elevation in adduction, mimicking a Brown syndrome. If Gt and FD are not particularly tight, a procedure very similar to the H-I procedure may be performed, by advancing the entire SO tendon. Any scar tissue restricting FD needs to be released. The SO tendon may be scarred to the superior-temporal quadrant, making identification of the previously operated SO difficult, and thus we approach the SO muscle on the nasal side of superior rectus (SR) and follow it temporally as far as possible. In this particular case we would perform a procedure very similar to the H-I procedure; advancing the entire SO tendon in both eyes. The sutures are placed at the superior border of the lateral rectus 8 mm posterior to the insertion, and the hang-back sutures are tightened until desired incyclotorsion is achieved. The repeated FD and Gt during surgery provide useful intraoperative information on how much to further tighten the SO muscles. We aim for 7 degrees of total overcorrection in incyclotorsion, and vertical alignment < 2 PD at adjustment, which can be postponed up to 7 days after surgery.³ Adjustment can be performed in the operation room with a small draping under topical anesthesia with oxybuprocaine and cocaine 4% eyedrops.

- Bata BM, Leske DA, Holmes JM. Adjustable bilateral superior oblique tendon advancement for bilateral fourth nerve palsy. *Am J Ophthalmol.* 2017; 178:115-121.
- Guyton DL. Exaggerated traction test for the oblique muscles. Ophthalmology 1981; 88(10):1035-1040.
- 3. Nihalani BR, Hunter DG. Adjustable suture strabismus surgery. *Eye (Lond)*. 2011; 25(10):1262-1276.

Inferior Rectus Recessions Bilateral Inferior Rectus Recession for Bilateral Superior Oblique Palsy

Stacy Pineles MD

Approach

In cases of V-pattern esotropia or excyclotropia in downgaze, patients with bilateral superior oblique palsies may benefit from bilateral inferior rectus recession. Weakening both inferior rectus muscles will improve excyclotorsion (more in downgaze), address small hypertropias (with asymmetric recessions), and address the V-pattern ET that is typically seen in these patients.

Potential Variations and Additions to the Procedure

- 1. Lower eyelid retractor dissection from the inferior rectus *muscle*: One can lyse lower eyelid retractors with assistance from oculoplastics, or simply use meticulous dissection from the inferior rectus. (Some have advocated for suturing of the capsulopalpebral head.)
- 2. *Fixed vs. adjustable vs. semiadjustable suture*: Adjustable suture has increased risk of slipped muscle in cases with tight inferior rectus. Some surgeons utilize a semiadjustable suture or fixed suture if there is concern for potential slippage. Semiadjustable suture can be advanced but not recessed at the time of adjustment.
- 3. Suture type—absorbable vs. nonabsorbable: Nonabsorbable suture is used by many surgeons to minimize the risk of slipped muscle, but long-term exposure of the nonabsorbable suture with adjustable suture can lead to chronic conjunctival inflammation or erosion.
- 4. Transposition can be considered to alter torsional effects.

Potential Advantages of This Procedure

- Adjustable
- Technically easier than alternative procedures involving oblique muscles
- Can address hypertropia in primary position and downgaze
- Weakening of the inferior rectus depression force may cause increased innervational input to the superior obliques, thereby improving their torsional function.

Potential Disadvantages of this Procedure

- Exotropia in downgaze (but typically these patients have a V-pattern esotropia). Surgeons can place the inferior rectus nasally if that is a concern. Preoperative free space prism test can help determine how much induced exotropia they can tolerate in downgaze.
- Could potentially exacerbate or cause hypertropia in down / side gaze; however, this may be balanced by the corresponding fellow eye superior oblique weakness.
- Lower eyelid changes
- Less torsional correction

Selected Readings

- 1. Souza-Dias C. Paresias y paralisis del IV nervio. In: Prieto-Diaz, Souza-Dias C, eds. *Estrabismo*. 2nd ed. São Paulo, Brazil: Roca; 1986:291-306.
- Kushner, BJ. 'V' esotropia and excyclotropia after surgery for bilateral fourth nerve palsy. *Arch Ophthalmol.* 1992; 110:1419-1422.
- 3. Bradfield YS, Struck MC, Kushner BJ, Neely DE, Plager DA, Gangnon RE. Outcomes of Harada-Ito surgery for acquired torsional diplopia. *J AAPOS*. 2012; 16:453-457.
- 4. Kushner BJ. An evaluation of the semiadjustable suture strabismus surgical procedure. *J AAPOS*. 2004; 8:481-487.

Glasses for Hyperopia

Donny W Suh MD

I. Background

- Moderate hyperopia in children is associated with the development of manifest strabismus and amblyopia.
- B. There is consensus that optical correction should be prescribed for moderate hyperopia when strabismus or amblyopia is present.
- C. There is no consensus regarding the prescription of optical correction in the absence of strabismus or amblyopia.
- II. Primary Objective

To compare visual acuity outcomes and development of strabismus after a 3-year follow-up period in children age 12 to < 36 months with moderate hyperopia (spherical equivalent +3.00 D to +6.00 D) who are prescribed glasses either immediately or only after confirmation of prespecified deterioration criteria (see Table 1).

- III. Study Design / Methods
 - A. Major eligibility criteria
 - 1. Cycloplegic refraction
 - a. +3.00 D to +6.00 D spherical equivalent refractive error in either eye
 - b. Astigmatism ≤ 1.50 D in both eyes
 - c. Spherical equivalent anisometropia ≤+1.50 D
 - 2. No prior treatment for refractive error with glasses (unless treatment was 1 week or less in duration and occurred more than 2 months prior to enrollment)
 - 3. No prior treatment for amblyopia or strabismus
 - 4. No measurable heterotropia at distance (3 meters) or at near (1/3 meter) by cover / uncover testing
 - 5. No known neurological anomalies

- B. Treatment groups
 - 1. Participants were randomized (1:1) to either:
 - a. Observation: glasses not prescribed unless participant had confirmation of 1 or more deterioration criteria (see Table 1)
 - b. Glasses: glasses prescribed at enrollment, to be worn for the duration of the study
- C. Sample size
 - 1. Planned, 246; to provide 90% power to detect a difference in failure rates at 3 years given expected 3-year failure rates of 10% and 25% in the glasses group and the observation group, respectively, and a type I error rate of 5%
 - 2. 130 participants were actually enrolled (due to slower than expected recruitment).
- D. Methods and testing procedures
 - 1. Participants were seen every 6 months for 3 years.
 - 2. The following were tested at each follow-up visit:
 - a. Monocular distance visual acuity using the ATS-HOTV (if 3 years of age or older)
 - b. Ocular alignment using the cover / uncover test
 - c. Near stereoacuity using the Randot Preschool Stereoacuity test (if 3 years of age or older)
 - 3. Primary outcome examination occurred 3 years (36 months) after randomization.
- E. Primary analysis

Proportion meeting failure criteria (Table 1) at 3 years postrandomization compared between treatment groups

F. Study results will be presented at the AAO 2018 Subspecialty Day Meeting.

Table 1. Failure and Deterioration Criteria

Failure (Primary Outcome)	Deterioration (Prior to 3 Years)	
The participant was considered to have met failure criteria if <i>any</i> of the following criteria (with the exception of strabismus surgery prior to the 3-year outcome exam) were met during testing by a masked examiner at the 3-year examination both with and without trial frames (without prism or bifocal), and the criteria was confirmed by a retest, with and without trial frames.	The participant was considered to have met deterioration criteria if any of the following criteria were met while wearing randomized correction by an unmasked examiner at a protocol-specified or non-protocol mandated visit after randomization but prior to the 3-year outcome exam, and confirmed by a retest performed by a masked examiner.	
1. Any measurable manifest strabismus in primary gaze at distance (3 meters) or at near (1/3 meter) not correctable with distance refractive correction alone	1. Any measurable manifest strabismus detected by cover / uncover test in primary gaze at distance (3 meters) or at near (1/3 meter)	
2. Distance VA below age norms in either eye	2. Distance VA below age norms in either eye	
3. ≥ 2 logMAR lines of IOD if VA is 20/25 or worse in the better- seeing eye (applies to IOD in either with or without correction but not one eye with and the other without)	3. ≥ 2 logMAR lines of IOD if VA is 20/25 or worse in the better- seeing eye (applies to IOD in either with or without correction but not one eye with and the other without)	
4. \geq 3 logMAR lines of IOD if VA is 20/20 or better in the better- seeing eye (applies to IOD in either with or without correction but not one eye with and the other without)	4. ≥ 3 logMAR lines of IOD if VA is 20/20 or better in the better- seeing eye (applies to IOD in either with or without correction but not one eye with and the other without)	
 Stereoacuity at near by Randot Preschool Stereoacuity test below age-normal values 	5. Stereoacuity at near by Randot Preschool Stereoacuity test below age-normal values	
6. Strabismus surgery prior to the 36-month outcome exam	6. Non-protocol treatment is received in the absence of meeting deterioration criteria	

Age-Normal Values for Stereoacuity and VA

Age range	Stereoacuity level needed to meet failure criteria (arcsec)	VA level needed to meet failure criteria
36-47 months (3 years)	800 or worse	20/63 or worse
48-59 months (4 years)	400 or worse	20/50 or worse
60-71 months (5 years)	400 or worse	20/40 or worse
72-83 months (6 years)	200 or worse	20/40 or worse
\geq 84 months (\geq 7 years)	100 or worse	20/32 or worse

Abbreviations: VA, visual acuity; logMAR, logarithm of minimum angle of resolution; IOD, interocular difference; arcsec, seconds of arc.

Low-Dose Bevacizumab for ROP

Sharon F Freedman MD, David K Wallace MD, and the Pediatric Eye Disease Investigator Group (PEDIG) / National Eye Institute

- I. Why Consider Low-Dose Bevacizumab for Severe ROP?
 - A. Antivascular endothelial growth factor (VEGF) drugs have shown efficacy for severe ROP.
 - B. Bevacizumab is affordable and has had widespread use for macular disease and ROP.
 - C. Standard-dose bevacizumab (0.625 mg intravitreal injection per eye) = ½ adult dose and is likely higher than necessary.
 - D. Bevacizumab at standard dose is known to lower circulating levels of VEGF for weeks in premature infants.
 - E. VEGF is important for continued normal development of premature infant in locations including brain, lungs, bones, kidneys, and retina.
 - F. Lower doses of intravitreal bevacizumab (0.25 mg) have been shown to be effective treatment for ROP.
- II. Phase 1 Dosing Study of Intravitreal Bevacizumab for ROP (PEDIG / NEI)
 - A. Purpose: To determine a dose of intravitreal bevacizumab that is lower than standard dose for severe (type I) ROP, is effective, and can be tested in future larger studies
 - B. Eligibility criteria: type I ROP (1 or both eyes), with no prior treatment for ROP
 - C. Dose de-escalation study in cohorts of 10-14 infants treated at each dose
 - 1. 0.25 mg, 0.125 mg, 0.063 mg, 0.031 mg
 - 2. One eye got lower dose, other eye got prior 2x dose (if both had type I ROP).
 - 3. Special dilution of drug by investigational drug pharmacy: Special 0.3-mL syringes allowed 0.01-mL volume to be injected.
 - D. Primary outcome = 4-week success or failure
 - 1. Success = improvement by 3-5 days; no recurrence of type I ROP, or severe neovascularization requiring additional treatment, within 4 weeks of injection
 - 2. Failure confirmed by second examiner, then treatment at investigator discretion
 - 3. Assessment of each cohort of 10-14 infants by data safety and monitoring committee
 - 4. Post-injection follow-up at 1 day, 4 days, weekly x 4 weeks, then as clinically indicated

5. Chart review at 6 months adjusted age, clinical exam at 12 months, eye exam + Bayley III at 24 months

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- III. Baseline Characteristics of Participants
 - A. 9 sites, 17 investigators, 61 infants enrolled / treated
 - B. 58 infants completed 4-week exam
 - C. Mean birthweight, 709 gm; mean gestational age, 24.9 weeks
 - D. 57 (93%) had type I ROP
- IV. Study Results: Success at 4 Weeks in Study Eye
 - A. 11/11 eyes at 0.25-mg dose
 - B. 14/14 at 0.125-mg dose
 - C. 21/24 at 0.063-mg dose
 - D. 9/9 at 0.031-mg dose
- V. Study Results: Retreatment of Study Eyes
 - A. No additional treatment: 39 eyes (64%)
 - B. Retreated (mean time 17.2 weeks after initial treatment):
 - 1. For early failure: 3 eyes (5%)
 - 2. For late recurrence of ROP: 11 eyes (18%); 7 laser, 4 bevacizumab
 - For persistent avascular retina: 8 eyes (13%) all lasered
 - C. Outcomes at ≥ 6 months
 - 1. Regressed ROP: 54 eyes
 - 2. Retinal detachment stage IVA: 1 eye
 - 3. Retinal detachment stage V: 1 eye
 - D. Adverse events: mild vitreous hemorrhage (1), death due to other conditions (5)
- VI. Conclusions: The Future
 - A. Dosage as low as 0.031 mg (5% of BEAT-ROP dosage) was successful in 9/9 eyes.
 - B. Some eyes required more treatment.
 - C. Study resumed at still lower dosages to find "lowest successful dose," and ongoing.

Natural History of Intermittent Exotropia

Christie L Morse MD

- I. Background
 - A. Intermittent exotropia (IXT) is the most common form of childhood-onset exotropia.
 - B. Conflicting literature regarding the natural history of this disorder
 - 1. Separate studies report worsening, improvement, or no change over time.
 - 2. Most are retrospective studies and only evaluate the angle of deviation.
 - 3. Buck and coauthors (BMC Ophthalmology, 2012)
 - a. 195 prospectively observed children with untreated IXT for 2 years
 - b. < 1% deteriorated to constant exotropia.
 - c. Mean control improved and the deviation showed no change.
- II. PEDIG IXT2 Natural History Study
 - A. Untreated observation arm of prospective randomized clinical trial (RCT) in children 3 to 10 years
 - B. Eligibility
 - 1. 3 to < 11 years
 - 2. Any form of IXT with ≥ 10 PD at distance (PACT)
 - 3. \geq 15 PD of IXT at distance or near (PACT)
 - 4. No previous IXT treatment, and family willing to forgo treatment during study

- C. Followed every 6 months to 36 months
- D. Received no treatment unless primary outcome was met
- E. Primary outcome (deterioration) is constant XT of 10 PD or more at distance and near or a drop in stereoacuity of 2 or more octaves.
- F. A third category of deterioration included patients started on treatment without meeting above criteria.
- III. PEDIG 3-Year Outcome Results (to be presented at the meeting)
 - A. Baseline characteristics
 - 1. 183 children enrolled, and 83% completed the study.
 - 2. Mean age of 6.1 years and 63% female
 - B. Deterioration by 3 years
 - C. Deterioration at 3 years
 - D. Secondary measures (change in angle, stereo, and control at both distance and near) assessed in patients who had not started any treatment.

Binocular Treatment for Amblyopia

Lisa C Verderber MD

I. Primary Study Objective

To compare the effectiveness of 1 hour/day of binocular game play 5 days per week plus spectacle correction, with continued spectacle correction only, for treatment of amblyopia in children 7 to < 13 years of age.

- II. Study Design/Methods
 - A. Major eligibility criteria
 - 1. Age 7 to < 13 years
 - Amblyopia associated with anisometropia, strabismus (< 5∆ at near measured by simultaneous prism and cover test [SPCT]), or both
 - 3. No amblyopia treatment (atropine, patching, Bangerter, vision therapy, binocular therapy) in the past 2 weeks
 - 4. Spectacle correction (if required) worn for at least 16 weeks, or until stability of visual acuity (VA) is demonstrated (< 0.1 logMAR change by the same testing method measured on 2 exams at least 8 weeks apart)
 - 5. Best-corrected amblyopic eye VA of 33 to 72 letters (E-ETDRS) (20/40 to 20/200)
 - 6. Best-corrected fellow-eye VA of ≥ 78 letters (E-ETDRS) (20/25 or better)
 - 7. Interocular difference ≥ 15 letters using the E-ETDRS (≥ 3 logMAR lines)
 - 8. No myopia greater than -6.00 D spherical equivalent in either eye
 - 9. Demonstrated ability to play the Dig Rush game under binocular conditions (with red-green glasses) on at least level 3, including ability to see red "diggers" and blue "gold carts" at 20% contrast in the nonamblyopic eye
 - B. Treatment groups

Participants randomly assigned with equal probability to either:

- 1. Binocular treatment: Binocular computer game play prescribed 1 hour per day, 5 days a week, with spectacles if needed
- 2. Continued spectacle correction, if needed
- C. Sample size:

116 children (with a maximum of 20% of enrolled participants having had previous binocular therapy)

- D. Visit schedule
 - 1. Randomized trial
 - a. Enrollment exam and randomization
 - b. 1-week phone call (7 to 13 days from randomization) to inquire about issues with the binocular game (if applicable) and to encourage compliance with treatment
 - c. 4 weeks ± 1 week: primary outcome
 - d. 8 weeks ± 1 week: secondary outcomes
 - Post-8-week phase: Spectacle group switched to binocular treatment and followed for 8 weeks; study ends for participants originally assigned to binocular treatment
 - a. 9-week phone call (7 to 13 days from 8-week exam) to inquire about issues with the binocular game (if applicable) and to encourage compliance with treatment
 - b. 16 weeks ± 1 week: final visit
- E. Testing procedures
 - 1. VA measured in each eye using the E-ETDRS at enrollment and all follow-up visits
 - 2. Near stereoacuity using the Randot Butterfly Stereoacuity test and Randot Preschool Stereoacuity test
 - 3. Ocular alignment (distance and near) by cover test, SPCT (if manifest deviation present), and prism and alternate cover test (PACT)
 - 4. History of diplopia (participant and parent questionnaires)
 - 5. Symptoms (parent-reported questionnaire)
- F. Analyses
 - 1. Primary analysis:

Compare mean change in amblyopic-eye VA from enrollment to 4 weeks between the binocular computer treatment group and the continued spectacle treatment group

- 2. Secondary analyses
 - a. Amblyopic-eye VA
 - i. Treatment group comparison of mean change in VA from enrollment to 8 weeks
 - ii. Improvement of ≥ 10 letters (≥ 2 logMAR) lines at 4 and 8 weeks after randomization

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b. Subgroup analyses at 4-week visit (exploratory)

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- c. Treatment group comparison of stereoacuity improvement at 4 and 8 weeks from enrollment
- d. Exploratory analyses evaluating the relationship between objective measures of binocular treatment (total hours of game play, felloweye contrast) with improvement in VA and stereoacuity
- e. Treatment group comparisons evaluating safety / adverse effects reported at 4- and 8-week visits
 - i. Change in fellow-eye VA from enrollment

- ii. Development of a new ocular deviation or worsening of a pre-existing deviation by ≥ 10∆ from enrollment
- iii. Presence / frequency of diplopia and symptoms
- f. Exploratory analyses at 16-week visit (participants assigned to continued spectacles who later received 8 weeks of binocular treatment)
- G. Study results will be presented at the AAO 2018 Subspecialty Day Meeting.

Evidence: Two vs. 6 Hours of Patching for Amblyopia

Michael X Repka MD MBA

- I. How Much to Patch?
 - A. Intuition and tradition
 - 1. More must be better!
 - 2. Full-time has been standard in texts, practice guidelines, and expert commentary.
 - B. Real practice
 - 1. Not always suggested
 - 2. Inconvenient for families
 - 3. Not always completed
- II. Pediatric Eye Disease Investigator Group (PEDIG)
 - A. Consortium of university- and community-based pediatric ophthalmologists and pediatric optometrists
 - B. Goal: to study common problems in a real-world setting
- III. Design
 - A. Randomized controlled trial
 - B. Children less than 7 years of age with amblyopia in the range of 20/40 to 20/80 were assigned to receive either 2 hours or 6 hours of daily patching combined with at least 1 hour per day of near visual activities while patching.
 - 1. Amblyopia from strabismus, anisometropia, or both combined
 - 2. Spectacles worn for a minimum of 4 weeks. At the time we did not understand that vision improves much longer with glasses, but this is a prerandomization factor so it does not affect the randomized outcome, other than some of the improvement seen in the study might have occurred with glasses alone.
 - 3. No patching or atropine therapy for at least 6 months prior to enrollment
 - 4. 189 children enrolled at 35 sites
- IV. Outcome Measure

Visual acuity using single-surrounded HOTV optotypes in the amblyopic eye after 4 months

- V. Results
 - A. Primary outcome
 - 1. Visual acuity in the amblyopic eye improved a similar amount in both groups. The improvement in the amblyopic eye acuity from baseline to 4 months averaged 2.40 lines in each group.
 - 2. Mean difference in logMAR acuity between groups = 0.001; 95% CI, -0.040 to 0.042 (*P* = .98).
 - B. Secondary visual acuity outcomes
 - The 4-month acuity was > 20/30 and/or improved from baseline by > 3 lines in 62% in each group (*P* = 1.00).
 - 2. 79% of patients in the 2-hour group and 76% of patients in the 6-hour group had improved by 2 or more lines from baseline.
 - C. Patient adherence
 - 1. Judged by the investigator
 - 2. Excellent in 58%, good in 25%, fair in 14%, and poor in 3% of patients in the 2-hour group
 - 3. Excellent in 37%, good in 37%, fair in 15%, and poor in 11% of patients in the 6-hour group.
- VI. Conclusion

When combined with prescribing near visual activities, 2 hours of daily patching produces an improvement in visual acuity that is of similar magnitude to the improvement produced by 6 hours of daily patching in treating moderate amblyopia in children 3 to less than 7 years of age.

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Amblyopia in the Real World

Edward L Raab MD

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The studies completed by the Pediatric Eye Disease Investigator Group and others have clarified several questions and have improved management and patient outcomes of both moderate and severe amblyopia. While these studies have provided a wealth of scientific information, practical considerations may limit what could be achieved through strict adherence to these advances in knowledge. This discussion highlights some of the constraints that ophthalmologists typically encounter and the need to find satisfactory approaches that modify what might be ideal practice. Reasoning and logic may dictate treatment modifications, whether or not their value has been scientifically determined. Selected medicolegal considerations, applicable as well beyond amblyopia management, will be considered.

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Evidence: Treating ROP Based on the ETROP Guidelines

William V Good MD

Guidelines for treating ROP have evolved over the last 30 years.

- I. Cryotherapy for ROP Treatment Trial
 - A. Treat infants with threshold disease within 72 hours of diagnosis. Threshold is zone I or II, plus disease, and 5 contiguous or 8 cumulative clock hours of stage 3.
 - B. Prognosis worsens with increasing amount (clock hours) of stage 3.
 - C. The success rate, as determined by visual acuity, has decayed over 20 years as these infants have been followed and measured.
 - D. No question that treatment is better than observation.
 - E. Myopia, strabismus, amblyopia are problems.
 - F. Used cryotherapy, not laser therapy.
- II. Early Treatment for Retinopathy of Prematurity Treatment Trial
 - A. Infants were randomized on the basis of risk for blindness, and this risk was based on an assessment of the Cryo-ROP cohort.
 - B. ≥ 15% risk; 1 eye randomized at prethreshold and the other observed until it developed "threshold ROP"
 - C. Prethreshold defined as zone I any ROP; zone II stage 2, or stage 3, plus
 - D. Many infants had zone I ROP in this trial, but a significant number of control zone I eyes never needed treatment.
 - E. Clinical algorithm = treat type I ROP (zone I, stage 3, or zone I plus, or zone II, stage 2 or 3 with plus disease)

- F. Caveats (often forgotten)
 - 1. Type II eyes do not do better with early prethreshold treatment. In fact, there is a trend for these eyes to actually do worse with early treatment. Therefore: wait to treat type II eyes.

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- 2. Although the type I, zone I eye subgroup did not have the same success rate as zone II, this is a subgroup. Caution is advised in treating zone I, type II eyes.
- 3. Myopia the same in ETROP as CRYO-ROP. So laser is not preventive.
- 4. Visual fields only slightly diminished.
- III. Anti-VEGF Treatments
 - A. Have they replaced laser therapy?
 - 1. Watch for systemic *and* local (ocular) side effects.
 - 2. The least effective dose is unknown.
 - 3. Follow-up requirements are unknown.
 - 4. There is much enthusiasm for reducing incidence of myopia, but it is not known whether anti-VEGF treatments help with this.
 - B. What is the optimal timing for treatment? Should this follow ETROP guidelines?
- IV. Conclusion

Only clinical trials can answer important questions about ROP management.

ROP Treatment in the Real World

RV Paul Chan MD

I. Introduction

The Early Treatment for Retinopathy of Prematurity (ETROP) study established the basis for treatment of ROP with peripheral retinal ablation at type I or worse ROP.¹ It has been shown, however, that cases milder than type I ROP may require treatment based on certain clinical findings.² Also, as anti-VEGF therapies are being used more frequently in clinical practice, the treatment algorithm for ROP is evolving and recurrence of disease is becoming an increasing concern.

- II. Treatment of ROP Milder Than Type I²
 - A. Study was performed to characterize the frequency of and clinical indications for which experts treat ROP milder than type I disease.
 - B. The findings from 1444 eyes of all babies screened for ROP in the i-ROP study were reviewed to identify any cases treated that had findings milder than type I ROP.
 - C. In 9.5% of treated eyes in this study, experts recommended treatment for disease less than Type 1 ROP.
 - D. Reasons for treatment outside of type I ROP included the following:
 - 1. Active ROP with the fellow eye being treated for type I ROP.
 - 2. Thick stage 3 membranes with anteroposterior traction concerning for progression to stage 4 ROP
 - 3. Tangential traction with temporal vessel straightening concerning for macular dragging
 - 4. Persistent active ROP disease at an advanced postmenstrual age
 - 5. Vitreous hemorrhage
- III. Treatment With Anti-VEGF
 - A. Treatment for ROP with cryotherapy and/or laser photocoagulation has been based on the Cryo-ROP and ETROP studies.^{1,3} More aggressive forms of treatment-requiring ROP (eg, aggressive posterior ROP) may progress despite laser or cryotherapy.
 - B. Since 2006 the number of published reports demonstrating the use of intravitreal anti-VEGF for ROP has increased significantly.
 - C. There is variability in published studies regarding indications for the use of anti-VEGF in treating ROP. These indications for treatment have not always followed strict type I ROP criteria.⁴⁻⁷

IV. Anti-VEGF vs. Laser Photocoagulation

In 2017, the Ophthalmic Technology Assessment Committee (OTAC) of the American Academy of Ophthalmology published a report discussing the evidence on anti-VEGF for ROP, compared to laser photocoagulation.⁴

- A. No Level I evidence available
- B. Level II evidence: 6 studies
- C. Level III evidence: 7 studies
- D. Results indicate:
 - 1. No clear advantage over laser for peripheral zone II
 - 2. No clear advantage for first-line combination therapy
 - 3. ROP recurrence is not insignificant.
- V. Recurrence of ROP and Incomplete Retinal Vascularization After Anti-VEGF Treatment⁸⁻¹¹
 - A. Variable recurrence rates with anti-VEGF therapy with majority of studies reporting higher recurrence rates
 - B. Delayed and incomplete retinal vascularization, with prolongation of follow-up period. Areas of avascular retina may be seen years after initial treatment of ROP with intravitreal bevacizumab.¹¹
 - C. There is no consensus on the definition of ROP recurrence after the treatment of ROP with anti-VEGF agents.
- VI. Summary
 - A. Seminal studies such as Cryo-ROP and ETROP provide a foundation for treatment that is critical for clinicians to understand as we introduce new therapies for treatment-requiring ROP.
 - B. Studies have shown that in real-world clinical care, there are cases of ROP where experts may recommend treatment even when ROP is milder than type I.
 - C. There is variability in the published literature on the indications for the use of anti-VEGF in treating ROP.
 - D. There is no consensus on the definition of ROP recurrence after anti-VEGF therapy.
 - E. Anti-VEGF for ROP can be associated with recurrence of ROP and delayed retinal vascularization. This may require treatment for ROP that does not specifically fit the definition of type I.

F. Management of ROP in clinical practice is evolving with the introduction of new therapies, imaging, and further understanding of plus disease. Therefore, there may be a need to rethink our classification of treatment-requiring ROP.

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Evidence: Treating Intermittent Exotropia Based on PEDIG and UK Studies

When to Do Surgery in Intermittent Exotropia

David K Wallace MD MPH

I. UK Cohort Study

- A. Prospective cohort study of 460 children aged < 12 years with previously untreated intermittent exotropia (IXT [but X(T) in references 1 and 2])
- B. Authors investigated whether the following factors were associated with choice of management
 - 1. Control
 - 2. Angle
 - 3. Visual acuity
 - 4. Stereoacuity
 - 5. Age
- C. Over a 12-month period after enrollment:
 - 1. 65% received no treatment
 - 2. 21% had treatment for impaired visual acuity
 - 3. 19% received treatment for strabismus
 - a. 12% nonsurgical
 - b. 8% eye muscle surgery
- D. Children who had no treatment were younger (mean age: 3.38 years) than those who were treated (mean age: 4.07 years) (*P* < .001).
- E. Stereoacuity and size of the angle of strabismus did not influence the type of management received.
- F. Two years after enrollment, data were available for 81% of the original cohort:
 - 1. 53% had no treatment.
 - 2. 17% had treatment for reduced visual acuity only.
 - 3. 13% had nonsurgical treatment.
 - 4. 17% had surgery.
 - 5. 0.5% developed constant exotropia.
- G. The surgically treated group had clinically significant improvements in angle and control.
- H. Eight percent (8%) of those treated surgically required second procedures for overcorrection within 6 months of the initial procedure.
- I. At 6-month follow-up after surgery, 21% (13) were overcorrected.

- J. Authors concluded that many children in the UK with IXT receive active monitoring only and that deterioration to constant exotropia is rare.
- II. PEDIG Patching vs. Observation RCTs
 - A. Multicenter, randomized clinical trials with older (3-10) and younger (1-2) cohorts
 - B. 358 children, 3 through 10 years of age, with previously untreated IXT (older cohort)
 - C. 201 children, 1 through 2 years of age, with untreated IXT (younger cohort)
 - D. Eligibility criteria
 - 1. Intermittent or constant exotropia at distance
 - 2. Intermittent exotropia or exophoria at near
 - 3. Exodeviation of at least 15 PD at distance or near and at least 10 PD at distance
 - 4. Near stereoacuity of 400 seconds of arc or better (older cohort only)
 - E. Random assignment to
 - 1. Observation (no treatment for 6 months)
 - 2. Patching for 3 hours daily for 5 months, then a 1-month washout period of no patching
 - F. Primary outcome: deterioration at either the 3-month or the 6-month follow-up visit, defined as:
 - 1. Constant exotropia measuring at least 10 PD at distance and near, or
 - 2. Near stereoacuity decreased by at least 2 octaves from baseline (older cohort only), or
 - 3. Any nonrandomized treatment
 - G. Deterioration: older cohort
 - 1. 6.1% of those in the observation group (10/165)
 - 2. 0.6% of those in the part-time patching group (1/159)
 - 3. P = .004 (difference, 5.4%; lower limit of 1-sided exact 95% CI, 2.0%)
 - H. Deterioration: younger cohort
 - 1. 4.6% of those in the observation group (4/87)
 - 2. 2.2% of those in the patching group (2/90)
 - 3. P = 0.27; 95% CI, -3.8% to +9.4%

- I. Six-month mean exotropia control scores: younger cohort
 - 1. 2.8 points for observation vs. 2.3 for patching at distance (*P* = .02)
 - 2. 1.4 points for observation vs. 1.1 for patching at near (*P* = .26)
- J. Authors concluded that deterioration of previously untreated childhood IXT over a 6-month period is uncommon with or without patching treatment and that both are reasonable options.

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Intermittent Exotropia in the Real World Or, Why I Do Not Always Follow the Recommendations of RCTs

Burton Kushner MD

- I. When I Do Not Follow the Guidelines of RCTs
 - A. They asked the wrong (or not the best) question.
 - B. I disagree with the data analysis.
 - C. Their conclusions are not what I think should follow from the data.
 - D. My patient does not quite fit the RCT's inclusion criteria.
 - E. The study did not take into account all the trade-offs.
 - F. Results are generalized, and my patient is unique (does not fit their demographics).
 - G. The RCT did not use the optimum treatment dose for the treatment group.

- II. With Respect to Intermittent Exotropia
 - A. The Buck studies imply that most patients can just be observed.
 - 1. Most of their patients were well controlled at the start.
 - 2. If my patient is not well controlled, I would not opt to just observe.
 - B. The PEDIG patching studies assessed its use to prevent deterioration.
 - 1. Most intermittent exotropias deteriorate slowly.
 - 2. Patching's real-world use is to defer or avoid surgery in patients needing intervention, in which case it is highly effective. So I reject the conclusion that it is of no value.

Cataract: Partly Cloudy

Thomas A Oetting MD

- Intracameral antibiotics are the standard of care for cataract surgery (CS). The United States was slow to adopt clear evidence from Europe. Amazingly convincing paper from Aravind demonstrating safety.
- Femto laser–assisted CS (FLACS) is neutral—not clearly safer, more expensive. Big studies from Europe do not show safety advantage for FLACS. While some centers have demonstrated efficiency gains, FLACS is slower for most surgeons. May be useful for complex cases. May lessen corneal endothelial damage with dense lenses, but cheaper options exist (eg MiLoop, extracapsular cataract extraction).
- Third-generation multifocal (MF) IOLs are less ambitious and less risky. Latest generation of multifocal IOLs emphasizes enhanced depth of focus and are less ambitious with near vision goals. This third generation of MF IOLs are less likely to create glare and halo than past generations of MF IOLs. Still, must be careful with surface issues or retina issues that are already limiting a patient's contrast sensitivity.
- Slight trend toward bilateral same-day cataract surgery. Kaiser published a review showing advantages to patients, with little risk. Veteran Affairs Medical Centers (such as ours) are following. Medicare pays for only half of the second eye on the same day, which has slowed acceptance in the United States. Eases travel burden to family and patient. Saves system money. Treat each eye as separate case.

- Structured training with simulation clearly makes early CS cases safer for residents. Ample evidence supports the finding that structured training with simulation attenuates the risk of early surgeons doing CS.
- Primary posterior capsulotomy. Nice YAG limiting strategy of doing centered primary posterior capsulotomy and then capturing optic with posterior capsule. Really old news described years ago by Gimbel and done in Peds forever. However, just catching on in adult CS world for idea of PCO and YAG prevention. Use of cohesive ophthalmic viscosurgical device to push anterior face of vitreous posterior to eliminate need for anterior vitrectomy. Very stable IOL.
- MiLoop for dense nucleus. Relatively inexpensive way to break up dense lens into 2-4 pieces. Can be used with nonphaco procedure through relatively small incision or with phaco but starting with smaller pieces. Quick learning curve.
- Zepto device for capsulotomy. Not sure about this device. Early study showed learning curve issues with incomplete rrhexis. Saves on Trypan blue.
- CustomFlex iris prosthesis approved by FDA. We have been waiting for years for U.S. artificial iris. Finally have an FDA-approved device. Plan is to roll this device in to centers spread over the United States, as insertion is tricky.

Oculoplastics: Orbiting the Eye of the Storm

Julie A Woodward MD

NOTES			

Retina: Lightning Strikes

Michael Altaweel MD

I. Applications of Retinal Imaging

- A. Oral fluorescein (5-10 mL of 10% fluorescein mixed in juice) for wide-field fluorescein angiography (FA) in children
 - 1. Quality of imaging can equal that obtained with IV administration.
 - 2. Retrospective study of 103 FAs (62 IV and 41 oral) in 82 patients: No difference in quality or clinical utility for assessment of familial exudative vitreoretinopathy, Coats disease, CNV, and uveitis.
- B. Spectral domain OCT (SD-OCT)
 - 1. High acquisition speed and resolution near histological level $(3-5 \ \mu m)$
 - 2. Eye tracking technology is useful for children and reduces motion artifact.
 - 3. Hand-held OCT is increasingly used to look at retinal changes in ROP, nonaccidental trauma, and optic nerve abnormalities.
 - 4. SD-OCT is helpful in identifying central pathology that is difficult to discern with biomicroscopy, such as foveal retinoschisis (it can also distinguish between retinoschisis and retinal detachment [RD] in the periphery), foveal hypoplasia, subtle macular edema.
 - 5. Excellent for noninvasively following response to treatment
 - 6. X-linked retinoschisis example
 - a. Progressive, bilateral disease; 1 in 10,000 males
 - b. Mutation in XLRS1 gene, codes for retinoschisin, protein involved in retinal cellular adhesion
 - c. Findings: foveal schisis (nearly 100%), peripheral schisis, spontaneous vitreous hemorrhage, giant retinal tears (up to 15%), average 20/70 as an adult
 - d. Medical management: topical carbonic anhydrase inhibitor for macular schisis
 - e. Case series: Dorzolamide 2% t.i.d., 29 eyes of 15 patients, 69% had positive response
 - 7. Von Hippel Lindau (VHL) example
 - a. 50% of retinal angiomas are associated with VHL; first manifestation of VHL in 43%.
 - b. Autosomal dominant, defect in chromosome 3.

- c. VHL protein suppresses VEGF production and is tumor suppressor gene.
- d. Associated with hemangioblastomas of CNS (cerebellum, spine), pancreas, liver, epididymis; cancer of kidneys (renal cell carcinoma) and adrenal glands (pheochromocytoma)
- e. OCT: macular edema, subretinal fluid; widefield FA: hemangiomas, with feeder and draining vessels in larger lesions, and leakage from active lesions
- f. Management options: Observation, cryotherapy, laser of lesion / feeder vessels, brachytherapy, combined anti-VEGF with laser for treatment (limits exudation)
- 8. Laser injury example: OCT identifies pathology of injury and prognosis.
- II. Coats Disease
 - A. Severity spectrum, management including laser ± anti-VEGF, surgical for severe cases (role of minimally invasive small-gauge vitrectomy)
 - B. Described by George Coats in 1908: retinal vascular anomaly
 - 1. Telangiectatic and aneurysmal vessels with capillary dropout and incompetence of retinal vessels (demonstrated by FA)
 - 2. Not hereditary; not infectious
 - 3. Males:females, 3:1
 - 4. Unilateral in 90%
 - 5. Bimodal distribution: age 4 to 10, and age 30 to 40
 - 6. Stages
 - a. Stage 1: Dilation of retinal blood vessels
 - b. Stage 2: Both telangiectasia and exudation
 - c. Stage 3: Exudative RD
 - d. Stage 4: Total RD
 - e. Stage 5: Complications, characterized by irreversible blindness (eg, neovascular glaucoma)
 - 7. Treatment
 - a. Laser vessels directly with large spots, moderate intensity, sectoral panretinal photocoagulation.
 - b. Fibrosis at fovea may limit vision.

- c. Bevacizumab as adjuvant can reduce edema / exudate in early stages but is associated with development of subretinal fibrosis in severe cases.
- d. Exudative RD has poor prognosis and requires surgery for most eyes. Minimally invasive vitrectomy surgery recommended; drainage of subretinal fluid from external approach, avoidance of retinotomy or elevation of hyaloid, and apply laser to telangiectatic vessels. This is effective and limits iatrogenic complications.
- III. Management of Pediatric RD / Stickler Syndrome
 - A. RD in children
 - 1. Incidence < 1 per 100,000 annual
 - 2. Etiology
 - a. Trauma, 40% (vs. 10% in adults), 70% male
 - b. 35%-56% congenital anomalies
 - c. 30% prior cataract surgery
 - d. Macula-off RD and proliferative vitreoretinopathy more common; giant retinal tears more common (15%-20%)
 - 3. Outcome
 - a. Final anatomic success rate of 70%-80%;60% for age 11 and younger
 - b. Only 30%-40% of patients reach final acuity of 20/200 or better.
 - B. Stickler syndrome
 - 1. Progressive, autosomal dominant connective tissue (collagen) disorder
 - 2. Features: Spondyloepiphyseal dysplasia, midline palatal clefting, facial dysmorphia and hearing loss, megaloglobus and congenital myopia, vitreous strands and condensations, oral giant retinal tear and detachment are common.
 - C. Treatment of RD
 - 1. Scleral buckle relieves vitreous base traction, 25-gauge vitrectomy
 - 2. IOP controlled through surgery, lens-sparing, silicone oil tamponade
 - 3. Prophylaxis with cryotherapy or laser
 - a. Reduces RD rate from 50% to 10% if initially unaffected O.U.
 - b. Reduces second eye involvement from 80% to 10% if one eye already has RD.

- D. Optic nerve pit
 - 1. 1 in 10,000, Male=Female
 - 2. 10% bilateral, 70% temporal
 - 3. Fluid enters through pit and between retinal layers, creating schisis and possible RD.
 - Natural history: present 20/40–20/60, reduction to ≤ 20/200 within 6 months
 - 5. Treatment options
 - a. Laser barricade between disc and macula; schisis may prevent adhesion
 - b. Pneumatic retinopexy ± laser
 - c. Vitrectomy + gas ± laser; allows release of traction, sealing of hole, and creates barricade to fluid movement

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Neuro-Ophthalmology: A Head in the Clouds Acute, Comitant Adult and Childhood Strabismus:

Finding the Wolf in Sheep's Clothing

Andrew G Lee MD

Overview

- Comitant, painless, diplopia without ductional deficit is usually a benign sheep in childhood, but beware ... the wolf in sheep's clothing.
- Acute acquired comitant esotropia
- Retinal hemifield slide phenomenon
- Central vs. peripheral rivalry
- The wolf in sheep's clothing

Case 1

- 12-year-old boy presents with blurred vision O.U.
- Refraction: -1.00 O.U., 20/20
- Pupils normal without relative afferent pupillary defect (RAPD)
- External, SLE, IOP, and fundus exam: within normal limits
- Comitant 25 PD esotropia (ET), no ductional deficit
- CT head: negative
- Undergoes strabismus surgery: initially straight but then ET recurs
- Final diagnosis: Acute acquired comitant ET (AACE), can be a brain tumor

Acute Comitant Esotropia of Childhood

- AACE can also be the Chiari malformation.
- Decompress the Chiari malformation *first*, strabismus surgery second
- Numerous series confirm same finding: Acute comitant esotropia can be neurologic in origin.
- AACE occurs in both adults and children.
- Intermittent binocular vertical diplopia

Case 2

- 12-year-old girl
- Headaches
- 20/20 vision +1.00 sphere O.U.
- Pupils normal without RAPD
- External, IOP, SLE, and fundus exam: normal O.U.
- CT head: negative
- Can a visual field defect cause binocular diplopia?
- Non-overlapping, juxtaposed nasal visual fields (no fusion)
- Decompensates prior phoria: nonparetic diplopia (no ductional deficit, comitant)

Case 3

- 35-year-old white male patient with binocular (not monocular) diplopia
- 20/50 O.D. and 20/20 O.S.
- Pupils normal without RAPD
- Motility: Comitant small angle 3 right hypertropia (RHT) but *cannot fuse with prism*
- Rest of exam is normal.
- Seen by retina: epiretinal membrane (ERM) O.D.
- CT and MRI head: normal O.U.
- Impression: Binocular diplopia is "not the retina."
- Plan: Send to strabismus: "We can't fix this."
- Can retinal disease cause binocular (rather than monocular) diplopia?
- Seen by neuro-ophthalmology: Central vs. peripheral rivalry due to ERM O.D.
- Impression: "It is the retina."
- Plan: "Back to retina"
- Bangerter filter

Summary

- Comitant, painless diplopia without ductional deficit is usually benign in childhood, but beware ...
- The wolf in sheep's clothing
- Acute acquired comitant esotropia: Brain tumors and Chiari
- Retinal hemifield slide phenomenon: Bitemporal hemianopsia
- Central vs. peripheral rivalry: Retina can cause monocular *or* binocular diplopia

Glaucoma: A High-Pressure System

Steven J Gedde MD

Introduction

The diagnosis and treatment of glaucoma is rapidly evolving. Recent innovations in glaucoma management are applicable to both adult and pediatric patients.

Perimetry

Data from glaucoma clinical trials have highlighted the importance of repeat visual field testing to confirm or refute any new field defects. The Advanced Glaucoma Interventions Study (AGIS), Collaborative Initial Glaucoma Treatment Study (CIGTS), Early Manifest Glaucoma Trial (EMGT), and Ocular Hypertension Treatment Study (OHTS) are multicenter randomized clinical trials sponsored by the National Eye Institute that required change from baseline be seen on 3 consecutive visual fields before progression was verified. In OHTS, only 14.1% of new visual field defects were confirmed on retest.¹ Computerized algorithms using event- and trend-based analysis assist in determining the probability that observed changes represent deterioration rather than physiologic fluctuation.

Imaging

Ophthalmic imaging is a routine part of clinical practice, including the management of glaucoma patients. Optical coherence tomography (OCT) provides reliable and accurate measurements of the retinal nerve fiber layer (RNF). The Cirrus OCT normative database includes heathy subjects age 19 to 85 years with refractive errors –12.00 to +8.00 D. Based on reproducibility studies of Cirrus OCT, intervisit thinning of the RNFL of at least 4 microns in overall average, 8 microns in a quadrant, and 12 microns in a clock hour is suspicious for progression.² Progression analysis software is useful in OCT interpretation.

Glaucoma Medications

Netarsudil (Rhopressa) and latanoprostene bunod (Vyzulta) were recently introduced as new agents in glaucoma medical therapy. Netarsudil is an inhibitor of rho kinase and norepinephrine transport, and it lowers IOP by increasing trabecular outflow, decreasing aqueous production, and decreasing episcleral venous pressure. Netarsudil 0.02% daily was found to produce IOP reduction similar to that of timolol 0.5% twice daily,³ but it was less effective than latanoprost 0.005% daily.⁴ Latanoprostene bunod is rapidly metabolized into latanoprost acid (a prostaglandin analogue) and butanediol mononitrate (a nitric oxide-donating moiety). Latanoprost increases uveoscleral outflow, while nitric oxide donors increase trabecular outflow. Latanoprostene bunod 0.024% daily demonstrated greater IOP reduction than timolol 0.5% twice daily⁵ and latanoprost 0.005% daily.⁶ Studies have shown that netarsudil and latanoprostene are both tolerated well in patients with ocular hypertension and glaucoma.

Laser Therapy

Cyclophotocoagulation (CPC) involves laser destruction of the ciliary body to reduce aqueous production and IOP. Contact transscleral CPC using continuous wave diode laser (CW-CPC) is the most common mode of delivery. However, micropulse CPC (MP-CPC) is a newer method of CPC that administers a series of short, repetitive pulses of laser energy. A small, randomized clinical trial found the cumulative probability of success was higher and the incidence of complications was lower with MP-CPC than with CW-CPC after 18 months of follow-up.⁷

Glaucoma Surgery

Incisional glaucoma surgery is generally indicated in adults when additional IOP reduction is needed despite the use of maximum-tolerated medical therapy and appropriate laser treatment. In contrast, congenital glaucoma is almost always managed surgically, with medical therapy used only as a temporizing measure. The surgical options for managing glaucoma have expanded exponentially in recent years, and they include traditional glaucoma surgery, nonpenetrating glaucoma surgery, endoscopic cyclophotocoagulation (ECP), and minimally invasive glaucoma surgery (MIGS).

Traditional Glaucoma Surgery

Trabeculectomy and aqueous shunt implantation have been the traditional approaches used in glaucoma surgery. Traditional glaucoma surgery is the most effective means of lowering IOP. Medicare claims data⁸ and surveys of the American Glaucoma Society membership⁹ demonstrate that aqueous shunts are being selected with increasing frequency as an alternative to trabeculectomy.

Nonpenetrating Glaucoma Surgery

Nonpenetrating glaucoma surgery involves excision of corneoscleral tissue under a scleral flap, leaving a thin window of trabecular meshwork and Descemet window to provide resistance to aqueous outflow. Deep sclerectomy, viscocanalostomy, and canaloplasty are types of nonpenetrating glaucoma surgery. These procedures appear to have a better safety profile than trabeculectomy, but they may be less effective in reducing IOP.

Endoscopic Cyclophotocoagulation

ECP involves laser treatment of the ciliary processes under direct visualization. ECP has been used in the management of refractory glaucoma and in pediatric patients, but it is more commonly performed in combination with phacoemulsification. The procedure produces modest IOP reduction. Cystoid macular edema is the most common cause of vision loss with ECP.

Minimally Invasive Glaucoma Surgeries

MIGS are a group of newer glaucoma procedures characterized by an ab interno approach, minimal tissue trauma, excellent safety, rapid postoperative recovery, and modest efficacy.¹⁰ Currently available MIGS include the trabecular micro-bypass stent (iStent), CyPass microshunt, Xen gel stent, Trabectome, Kahook dual blade, gonioscopy-assisted transluminal trabeculotomy (GATT), Trab360, ab interno canaloplasty (ABiC), and Visco360. MIGS are frequently performed in combination with phacoemulsification. Limited data are available describing the safety and efficacy of MIGS in children.

Conclusions

Recent advances in glaucoma management have improved the quality of care delivered to adult and pediatric patients. Perimetry and OCT are the major ancillary tests for diagnosing glaucoma and monitoring for progression. Repeat visual field testing should be performed to confirm or refute the development of a new field defect. A decrease in RNFL thickness on OCT of at least 4 microns in overall average, 8 microns in a quadrant, and 12 microns in a clock hour is suggestive of progression. Computerized algorithms can help in evaluating whether visual field and OCT changes are real.

Netarsudil (Rhopressa) and latanoprostene bunod (Vyzulta) are new medications to lower IOP. MP-CPC may offer advantages over CW-CPC. The surgical options for treating glaucoma have expanded in recent years. Trabeculectomy and aqueous shunts remain the most effective ways to lower IOP. MIGS provide modest IOP reduction but have a better safety profile than traditional glaucoma surgery. Limited information is currently available about MIGS in the pediatric population. Many techniques in glaucoma surgery may be applied in both adults and children.

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Cornea: The Tip of the Iceberg

Christina Rapp Prescott MD

Introduction

Children are not just little adults, but they can benefit from some of the advances pioneered in adult corneal surgery. Children suffer from unique corneal pathology, including congenital anomalies, as well as less age-specific corneal pathology including ocular surface disease, ectasia, and refractive errors. I will discuss the use of medical contact lenses, corneal crosslinking, refractive surgery, and selective corneal transplantation in pediatric patients.

Medical Contact Lenses

Scleral contact lenses, including the PROSE (prosthetic replacement of the ocular surface ecosystem) custom-fit lenses, can be used to treat a variety of ocular conditions, including keratoconus, Stevens-Johnson syndrome, vernal keratoconjunctivitis, and irregular astigmatism due to corneal scarring.

Corneal Crosslinking

Corneal crosslinking was approved in the United States in April 2016. Though crosslinking is most commonly performed in adults, it is likely to offer the most benefit to children, since keratoconus is particularly aggressive when it presents in childhood.

Refractive Surgery

Though laser refractive surgery is most commonly performed in adults to reduce or eliminate the need for glasses or contact lenses, children at risk of refractive amblyopia may also benefit from this treatment. Of the refractive surgery options, photorefractive keratectomy has the best safety profile in children, though it does require general anesthesia and close monitoring postoperatively because of increased risk of scarring and infection in children.

Selective Corneal Transplantation

Adult corneal transplantation has undergone an evolution to selective corneal transplantation, in that endothelial transplantation is now more common than full-thickness transplantation. However, this transition has been slower in pediatric corneal transplantation, likely due to differences in both patient and pathology. However, for pathology isolated to either the anterior or posterior aspect of the cornea, selective transplantation can be especially beneficial in children, due to the higher risk of graph failure, infection, trauma, and amblyopia in children.

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2018 Advocating for the Profession and Patients Pediatric Ophthalmology Subspecialty Day

Kenneth P Cheng 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[®] Fund
- Surgical Scope Fund (SSF)
- State Eye PAC

Please join the dedicated community of ophthalmologists who are contributing to protect quality patient eye care for everyone. The OPHTHPAC Committee is identifying Congressional Advocates in each state to maintain close relationships with federal legislators in order to advance ophthalmology and patient causes. At Mid-Year Forum 2018, we honored nine of those legislators with the Academy's Visionary Award. This served to recognize them for addressing issues important to us and to our patients. The 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 the OPHTHPAC Fund. Each of us has a responsibility to ensure that these funds are strong.

OPHTHPAC® Fund

OPHTHPAC is a crucial part of the Academy's strategy to protect and advance ophthalmology's interests in key areas, including physician payments from Medicare and protecting ophthalmology from federal scope-of-practice threats. Established in 1985, OPHTHPAC is one of the oldest, largest, and most successful political action committees in the physician community. We are very successful in representing your profession to the U.S. Congress.

Advocating for our issues in Congress is a continuous battle, and OPHTHPAC is always under financial pressure to support our incumbent friends as well as to make new friends among candidates. These relationships allow us to have a seat at the table with legislators who are willing to work on issues important to us and our patients.

The relationships OPHTHPAC builds with members of Congress is contingent on the financial support we receive from Academy members. Academy member support of OPHTHPAC allows us to advance ophthalmology's federal issues. We need to increase the number of our colleagues who contribute to OPH-THPAC and to the other funds. 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.

Among the significant impacts made by OPHTHPAC are the following:

- Secured relief from the burdens and penalties associated with the existing Medicare quality improvement programs for 2018
- Halted applications of MIPS penalties to Part B drug payments to physicians
- Convinced CMS to revisit drastic cuts to retina and glaucoma surgical codes
- Halted the flawed Part B Drug Demonstration
- Derailed an onerous global surgery payment data collection plan
- Continued efforts in collaboration with subspecialty societies to preserve access to compounded and repackaged drugs such as Avastin

Contributions to OPHTHPAC can be made here at AAO 2018, or online at www.aao.org/ophthpac by clicking "Join." You can also learn more by texting "OPHTH" to 51555.

Leaders of the American Association for Pediatric Ophthalmology and Strabismus (AAPOS) and the American Academy of Pediatrics – Ophthalmology Section are part of the American Academy of Ophthalmology's Ophthalmic Advocacy Leadership Group (OALG), which meets annually in January in Washington, D.C., to provide critical input and to discuss and collaborate on the Academy's advocacy agenda. At the January 2018 OALG meeting, panel discussions took place on the outlook for Medicare reimbursement and implementation of the Merit-based Incentive Payment System (MIPS), as well as specialty research related to the IRISTM Registry. In addition, meeting participants discussed the changing paradigm for optometric scope battles, held a roundtable to discuss challenges for surgical subspecialties, and considered how telemedicine could impact ophthalmology.

At Mid-Year Forum 2018, the Academy, AAPOS, and the AAP–Section on Ophthalmology 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 and the AAP–Section on Ophthalmology remain crucial partners with the Academy in its ongoing federal and state advocacy initiatives.

Surgical Scope Fund

Thanks to contributions to the 2018 Surgical Scope Fund (SSF) from ophthalmologists across the country, the Academy's Surgery by Surgeons initiative has had a successful year preserving patient surgical safety and surgical standards in state legislatures across the country. The SSF is key to the Academy's Surgery by Surgeons campaign. *If you have not yet made a 2018 SSF contribution*, visit our contribution booth at AAO 2018 or contribute online at www.aao.org/ssf. If you already have made that 2018 contribution, please consider making a crucially needed supplemental contribution.

The SSF provides grants to state ophthalmology societies in support of their efforts to derail optometric surgery proposals that pose a threat to patient safety. Since its inception, the Surgery by Surgeons campaign and the SSF, in partnership with state ophthalmology societies, has helped 34 state/territorial ophthalmology societies reject optometric scope-of-practice expansion into surgery.

To date in 2018, thanks to financial resources from the SSF, the Surgery by Surgeons campaign has netted patient safety and surgery standard preservation victories in the following battleground states:

- Florida
- North Carolina

Iowa

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- South Carolina
- Vermont
- Maryland Mississippi
- Nebraska
- Virginia

The 2018 battle is far from over, though. For example, California, Illinois, Massachusetts, and Pennsylvania are currently under assault. Furthermore, as of submission of this update in June 2018, the optometric surgery push had sprouted in six additional states.

Dollars from the SSF are critical in the state surgery campaigns. In each of these legislative battles, the benefits from SSF distributions are abundantly clear. The best lobbyists and public relations consultants are contracted as necessary. Additionally, media campaigns (including TV, radio, and social media) are launched to educate the voting public when needed. This helps to secure success in protecting patient safety by thwarting optometry's attempts at expanding its scope of practice to include surgery privileges.

Each of these endeavors is very expensive, and no one state has the resources to wage one of these battles on its own. Ophthalmologists must join together and donate to the SSF to fight for patient safety when a state faces a scope battle over optometric surgery.

The Secretariat for State Affairs thanks the AAPOS, which joined state ophthalmology societies in contributing to the SSF in 2017, and looks forward to its continued financial support. Subspecialty organizations like the AAPOS complete the necessary SSF support structure for the creation and implementation of successful Surgery by Surgeons campaigns.

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: Advocate for Your **Profession & Your Patients**

Academy SSF contributions are used to support the infrastructure necessary in state legislative / regulatory battles and for public education. State PAC and OPHTHPAC contributions are necessary at the state and federal level, 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 Surgical Scope Fund, and your State Eye PAC. Please be part of the community advocating for your patients now.

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

The Myopia Epidemic

Terri L Young MD MBA

I. General Epidemiology

Refractive errors are optical aberrations determined by mismatches in the focusing power of the cornea, the lens, and the axial length of the eye. The most frequent eye disorders worldwide, refractive errors are an increasingly common cause of blindness.

- A. The worldwide distribution is rapidly shifting toward myopia, or nearsightedness.
 - 1. The myopia epidemic is particularly prominent in urban East Asia, where up to 95% of 20-yearolds in cities such as Seoul, Hong Kong, and Singapore have this refractive error.¹⁻²
 - 2. The prevalence of myopia is also rising throughout Western Europe and the United States, affecting ~50% of young adults in these regions.³⁻⁴
- B. Refractive error consequences
 - 1. Although refractive errors can be optically corrected, even at moderate values they carry substantial risk of ocular complications with high economic burden.⁵⁻⁶
 - 2. One in 3 individuals with high myopia (-6 D or worse) develops irreversible visual impairment or blindness, primarily due to comorbidities of myopic macular degeneration, retinal detachment, or glaucoma.⁷⁻⁹
- C. At the other extreme, high hyperopia predisposes individuals to strabismus, amblyopia, and angle-closure glaucoma.^{7,9}
- II. Possible Causes
 - A. Refractive errors result from a complex interplay of lifestyle and genetic factors.
 - B. The most established lifestyle factors for myopia are high education, lack of outdoor exposure, and excessive near work.^{1,3}
 - C. Recent research has identified many genetic variants for refractive errors, myopia, and axial length.⁹⁻¹³

Two large study cohorts—the International Consortium for Refractive Error and Myopia (CREAM) and the personal genomics company 23andMe, Inc.—have provided the most comprehensive results.^{10,13}

1. This genome-wide association meta-analysis in 160,420 participants and replication in 95,505 participants increased the number of established independent signals from 37 to 161 and showed high genetic correlation between Europeans and Asians (> 0.78).

- 2. Expression experiments and comprehensive *in silico* analyses identified retinal cell physiology and light processing as prominent mechanisms, and they also identified functional contributions to refractive error development in all cell types of the neurosensory retina, retinal pigment epithelium, vascular endothelium, and extracellular matrix.
- 3. Newly identified genes implicate novel mechanisms such as rod-and-cone bipolar synaptic neurotransmission, anterior-segment morphology, and angiogenesis.

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Atropine for Myopia

Donald TH Tan FRCS FRCOphth

- I. Interventional Approaches to Reducing Myopia Progression (Myopia Control)
 - A. 2011 Cochrane review
 - 1. Network meta-analysis of myopia control therapies
 - 2. Meta-analysis of atropine treatment for childhood myopia
 - B. Spectacles (under / overcorrection, bifocals, peripheral defocus)
 - C. Contact lenses (soft, RGP, peripheral defocus, orthokeratology)
 - D. Pharmacological (antimuscarinics, IOP-lowering drugs)
- II. Putative Mode of Action of Atropine in Reducing Axial Elongation
 - A. Blocking accommodation refuted
 - B. M1/4 muscarinic receptors at retina / amacrine cell
 - C. Direct action on scleral fibroblasts; inhibition of GAG synthesis
- III. ATOM1 Randomized Controlled Trial (RCT)
 - A. 400 children, 6-12 years
 - B. 1% atropine vs. placebo drops
 - C. 3 years, 2 on treatment, 1 washout year
 - D. 77% reduction in myopia progression
 - E. Visual side effects present
 - F. Myopic rebound on treatment cessation
- IV. ATOM2 RCT
 - A. Dose-ranging RCT: 0.5%, 0.1%, 0.01%
 - B. 400 children, 6-12 years
 - C. Bilateral treatment
 - D. Five-year study: 2 years on treatment, 1 year washout, 2 years retreatment
 - E. Efficacy
 - 1. Dose-related but clinically insignificant differences
 - 2. 0.01% atropine: 50%-60% reduction in myopia progression
 - F. Myopic rebound phenomenon
 - 1. Dose related
 - 2. Minimal with 0.01% atropine

- G. Safety
 - 1. Dose-related
 - 2. Fewer side effects with lowest dose
- H. 0.01% atropine: 0.8-mm pupil dilation, no near vision loss
- V. Lessons Learnt From ATOM 1+2
 - A. Atropine eyedrops reduce myopia progression.
 - B. Dose-related but rebound phenomenon with higher doses
 - C. Safety established with atropine eyedrops; higher doses cause dilation and accommodation loss.
 - D. Best therapeutic index with 0.01% atropine
 - E. Nonresponders are more aggressive progressors.
- VI. Post-ATOM 1+2 Studies
 - A. Other RCTs using 0.01% atropine: ATOM-Japan
 - B. ATOM3:
 - 1. Prevention study
 - 2. Randomizing for premyopic children to prevent or retard onset of myopia
 - 3. Five years and above
- VII. Current Atropine Trials Registered with Clinicaltrials.gov
 - 28 clinical trials registered:
 - A. 2006-2008: 4 trials
 - B. 2012-2013: 2 trials
 - C. 2014: 6 trials
 - D. 2015: 3 trials
 - E. 2016: 3 trials
 - F. 2017: 9 trials
 - G. 2018: 1 trial to date
 - H. Trial countries: China, USA, Singapore, Taiwan
- VIII. Current Atropine Eyedrop Formulations Available
 - A. Myopine: Singapore, Malaysia, Japan
 - B. Myatro (Entod Pharma): India
 - C. MicroPine (Eyenovia): USA
- IX. Future Studies in Myopia Control
 - A. Combination therapy with OK, SCL, spectacles, behavioral therapy
 - B. Other pharmacological agents
 - C. Other atropine formulations

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Contact Lens Therapies for Myopia Control

Bruce H Koffler MD

I. Introduction

Significance of myopia control in light of myopia epidemic

- II. Classic Orthokeratology Contact Lenses
 - A. Definition and mechanism of action
 - 1. Using an RGP cornea mold
 - 2. Harnesses hydraulic forces under the mold to reshape the cornea overnight, reducing or eliminating refractive error for a period of time
 - B. Lens designs
 - 1. "Push/Pull" theory
 - a. Base curves "push" by central flattening
 - b. Reverse curves "pull" allow space for epithelial migration
 - 2. Landing or peripheral curves
 - C. How they work on hyperopic defocus: Dr. Earl Smith's work on theory of hyperopic defocus
 - D. Current ... past / present evidence
- III. Soft Lens Designs
 - A. Soft OK ArtMost strong peripheral inward focusing of 12-20 D, works on lens optics and not on corneal molding
 - B. True Ortho K style lens
 - C. Soft bifocal
 - 1. NaturalVue: daily multifocal
 - 2. All other soft bifocals

- IV. Safety
 - A. 1997-2001
 - 1. Multiple cases of microbial keratitis (MK), China / Taiwan
 - 2. Due to huge need, no protocols and no regulation
 - 3. Lack of patient training and compliance
 - 4. In our office since 2002, no cases of MK
 - B. 2002-present
 - 1. Bullimore MA, et al. Optometry and Vision *Science*, 2013.
 - a. Overall rate of MK: 7/10,000 years of wear
 - b. Children: 12/10,000
 - 2. Liu YM, et al. The safety of orthokeratology. *Eye and Contact Lens.* 2016.
 - 3. Recommended safety protocol to follow
 - V. Summary

Other Therapies for Myopia Prevention If Not Atropine or Contact Lenses, Then What?

Daniel Ian Flitcroft MD

- I. Behavioral Modification
 - A. Avoiding education
 - 1. Evidence now of a causal link
 - 2. More education = more myopia
 - 3. No education < 2% myopia
 - B. More time outdoors
 - 1. A confirmed effect in preventing or delaying myopia onset
 - 2. Most impact in young children
 - 3. Little or no impact on myopia progression
 - 4. Not the universal panacea it was supposed to be
 - C. Changing reading behavior
 - 1. 20:20:20
 - 2. Working distance
 - 3. Paper or screen?
- II. Other Pharmacological Therapies
 - A. Adenosine antagonists
 - 1. Oral 7-methylxanthine (caffeine and theobromine metabolite)
 - a. Some evidence from clinical studies
 - b. Lack of replication; no association with dietary intake (UK/USA)
 - c. Least effect in fastest progressors
 - d. In animal studies causes hyperopic shift in control eyes
 - e. Certain claims exceed evidence

- 2. Topical caffeine
 - a. Similar properties
 - b. Topical therapy in animals blocks lensinduced myopia.
 - c. Hyperopic shift in control eyes
- B. Glaucoma medications
 - 1. Timolol ineffective
 - 2. Latanoprost: impact on animal model of myopia (guinea pig)
 - 3. Brimonidine: impact on animal model of myopia (guinea pig)
- C. Other anti-muscarinics:
 - 1. Pirenzepine: no active development post Phase 2
- III. Other Optical Therapies
 - A. Undercorrection: No
 - B. Delayed correction: No
 - C. Executive bifocals: strongest optical effect
 - D. Varifocal: limited outside rare esophoric myopes
 - E. Peripheral defocus lenses: limited effect
- IV. The Era of Active Myopia Management
 - A. Is doing nothing acceptable? Probably yes, for now
 - B. Is saying nothing acceptable? No
 - C. It is time to talk to your patients about myopia and measure their axial length.

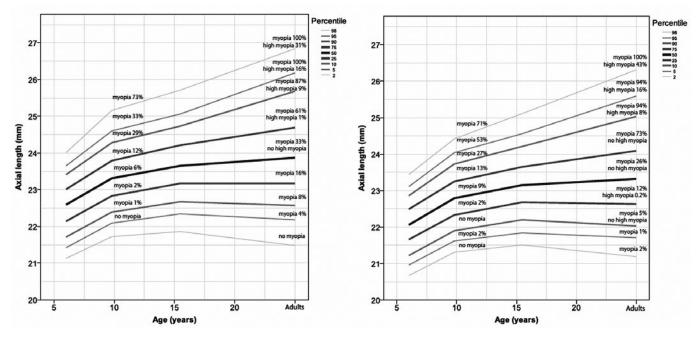


Figure 1. Growth chart depicting axial length (in mm) vs. age for European study subjects—males (left) and females (right)—with the risk of myopia in adulthood. The myopia percentage represents the proportion of myopia in halfway above and below the percentage line. Reproduced, with permission, from Tideman JWL, Polling JR, Vingerling JR, et al. Axial length growth and the risk of developing myopia in European children. *Acta Ophthalmol.* 2018; 96(3):301-309, figure 2.

Newer Intraocular Instruments and IOLs

M Edward Wilson MD

Using Now

- 25-gauge vitrector at 7500 cuts/minute: Leave it at the highest speed; no need to use 20-gauge even with tough membranes.
- Preloaded hydrophobic acrylic IOLs: Fewer loading errors
- Newer less-stress pupil expanders: Fewer pupil tears and peaks
- Iris-claw IOLs in the absence of capsular support: FDA compassionate use protocol has been active for 5 years and is ongoing.
- Microincision capsulorrhexis forceps: For anterior and posterior continuous curvilinear capsulorrhexis CCC
- Kloti radio-frequency diathermy capsulotomy: For white or scarred capsules

Under Evaluation

- 25- and 27-gauge bi-blade vitrector at 15,000 cuts/minute: Less traction
- Hypersonic vitrectomy: Potentially a new paradigm

- Hydrophobic acrylic IOLs with polyvinylidene fluoride (PVDF) haptics: For use with Yamane or Ararwal intrascleral fixation techniques. Concerns exist for soft sclera.
- Extended-depth-of-focus (EDOF) IOLs: Potentially better than multifocal IOLs for the second decade of life
- Zepto precision-pulse capsulotomy in children: Nitinol ring for shape precision; need a smaller size
- Femtosecond laser anterior and posterior capsulotomy: Logistics and cost are problems for pediatric hospitals.
- Bag-in-the-lens IOL: Impressive published European data in children, but compassionate use FDA protocol may be needed for use in the USA.

Already in the Pipeline for the Near Future

- Modular IOLs with exchangeable optics: Pediatric surgeons will be the primary users.
- Refractive index shaping of any IOL using a phase wrapping algorithm: Allows for multiple changes in power over the course of eye growth

Genetics of Pediatric Cataracts

Congenital and Pediatric Cataract: Genetics and Diagnostic Pathways

I Christopher Lloyd MB FRCS FRCOphth

Congenital and pediatric cataract is an uncommon but sightthreatening condition, largely caused by genetic mutations. Some of these mutations are also linked to inborn errors of metabolism. Appropriate early intervention in cases amenable to treatment can thus not only maximize visual function but prevent systemic disease progression.

Traditional diagnosis is a lengthy process that is costly to health services and is typically unsuccessful—particularly in congenital cataract. Next-generation sequencing (NGS) techniques have revolutionized the utility of genomics in aiding precise diagnosis and the subsequent provision of accurate prognosis, management, and treatment.

The speaker will present cases illustrating this utility and discuss revised models of care for children affected by these disorders.

Selected Readings

- Gillespie RL, Urquhart J, Anderson B, Williams S, Waller S, Ashworth J, Biswas S, Jones S, Stewart F, Lloyd IC, Clayton-Smith J, Black GC. Next-generation sequencing in the diagnosis of metabolic disease marked by paediatric cataract. *Ophthalmology* 2016; 123(1):217-220.
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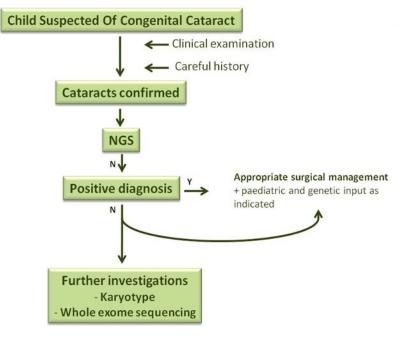


Figure 1.

Mainstreaming Corneal Crosslinking

Erin Stahl MD

- I. Introduction
 - A. Basic corneal crosslinking (CXL) history inside and outside the United States
 - B. Pediatric CXL: Literature review
- II. Current Situation
 - A. Insurance challenges: What is on the horizon?
 - B. Technical challenges in pediatric patients
- III. Medical Decision Making
 - A. Making the diagnosis
 - B. When to initiate treatment
 - C. Surgical tips
 - D. How to follow postoperatively
 - E. Visual rehabilitation
- IV. Conclusion

Topical Treatment for the Toughest Corneal Ulcer The Use of Topical Insulin to Treat Refractory Neurotrophic Corneal Ulcers

Michael C Struck MD, Angeline L Wang MD, Eric Weinlander MD, Brandon M Metcalf MD MPH, Neal P Barney MD, David M Gamm MD PhD, and Sarah M Nehls MD

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Introduction

Neurotrophic keratopathy is a degenerative disease of the corneal epithelium secondary to impaired corneal innervation by the trigeminal nerve. Standard treatment involves aggressive lubrication of the corneal surface, therapeutic contact lenses, amniotic membrane grafts, and tarsorrhaphy. Refractory neurotrophic corneal ulcers occur when treatment response is incomplete and are potentially blinding. Insulin is a widely available, relatively safe, and familiar medication that has been shown to improve corneal epithelial healing in vitro and in diabetic animal models. However, clinical experience with topical insulin in patients with nonhealing corneal wounds is minimal. We present our experience in patients with refractory neurotrophic corneal ulcers that were treated with topical insulin.

Outline

All patients were prescribed insulin drops as compassionate use for the treatment of neurotrophic corneal ulcers after standard treatment had failed. The risks, benefits, and alternatives of the treatment were discussed with all patients and/or their parents, and they verbally consented to the off-label use of insulin.

The drops were prepared by injecting regular insulin into a new bottle of artificial tears with a polyethylene glycol and propylene glycol base at a concentration of 1 unit per milliliter. Drops were prepared by pharmacy, the patients' providers, or the patients themselves with detailed written instructions. This was done with sterile technique. The drops were refrigerated and used up to 1 month after preparation.

We present 6 patients who developed neurotrophic corneal ulcers or epithelial defects that were refractory to a range of standard medical and surgical treatments. The addition of topical insulin resulted in rapid and complete corneal reepithelialization, ranging from 7 to 25 days following initiation of treatment. One patient developed crystalline keratopathy while on the treatment, although this was likely secondary to chronic topical steroid use. No other local or systemic side effects were noted, including change in corneal vascularization or opacity.

Topical insulin has been found to improve healing of decubitus ulcers1 and experimentally induced superficial skin wounds in diabetic and nondiabetic individuals.² The effect of topical insulin on corneal wound healing has been well studied in rodent models. Notably, in diabetic rats, topical insulin improves corneal sensation and improves wound healing after corneal abrasions.³ Experience with insulin in corneal wound healing in humans is limited to 2 case series. A 1945 study reported improved healing of corneal ulcers after systemic administration of insulin.⁴ A 2013 retrospective study looked at 5 patients who developed corneal epithelial defects during vitreoretinal surgery who were treated with topical insulin drops and reported faster re-epithelialization compared with 10 patients who were treated with lubrication.⁵ There have been no previous studies of topical insulin use in patients with neurotrophic corneal defects.

The mechanism of insulin in promoting corneal wound healing in our patients remains speculative, but data suggest that restoration of corneal nerves and/or improved epithelial cell migration may play key roles. In diabetic mice, topical insulin has been shown to slow the loss of sub-basal plexus corneal nerves. Furthermore, the addition of insulin promoted cell migration and closure of artificial wounds in cultured sheets of corneal epithelial cells in an in vitro model of corneal epithelial wound healing.

Topical insulin may be a simple and effective treatment for refractory neurotrophic corneal ulcers. Our case series is limited by the heterogeneity in the patient presentations, differences in treatment frequency and duration, and lack of a comparative control. Further study is needed to determine the clinical efficacy and side effect profile of topical insulin in corneal wound healing.

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Use of IOL Optic Capture IOL Optic Capture and New Capsular Management Tips and Tools for Long-term IOL Centration

Erick D Bothun MD

The long-term result of pediatric cataract surgery often depends on successful IOL placement within or around a viable lens capsule. IOL decentration or subluxation may occur with haptics located in the capsular bag or sulcus, and that risk only increases with poorly constructed capsulotomies. Traumatized anterior segments or membranes pose additional challenges to long-term IOL position. In addition, the standard placement of an optic within the capsular bag prevents anterior and posterior capsular ring fusion and increases the risk of cortical regrowth into the visual axis. Such secondary visual axis opacification with reoperation was a common adverse event with IOL implantation in the Infant Aphakia Treatment Study.

Various approaches have been developed to perform the anterior and posterior capsular opening and address these IOLcapsular bag challenges. Once such example includes novel IOL design such as the bag-in-the-lens IOL by Dr. Marie Jose Tassignon. This IOL uses optic capture within an interhaptic groove. The result is a sandwiching of the anterior and posterior capsule to seal the lenticular epithelial cells. Reduction in posterior capsular opacity and in number of reoperations and increased stability have been documented with such optic capture. I have found posterior optic capture of a standard 3-piece IOL useful in my pediatric anterior segment practice. This technique has been similarly reported with long-term visual axis clarity and IOL stability. This technique is also useful in traumatized capsular bags and secondary IOL insertion. In my limited number of cases, I have not detected visual axis opacification, capsular contracture, or late IOL decentration.

This presentation will offer techniques and tips of capsular management, including IOL optic capture. The tips and emerging tools toward capsular management will include the following:

- Microincision capsulorrhexis forceps (for anterior and posterior capsules)
- Kloti radio-frequency diathermy capsulotomy (for white or scarred capsules)
- Zepto precision-pulse capsulotomy in children (nitinol ring for shape precision)
- Femtosecond laser anterior and posterior capsulotomy (Logistics and cost are problems for pediatric hospitals.)

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Corneal Topography in Children

Luca Buzzonetti MD

Corneal topography is currently one of the most important tools for corneal imaging evaluation. This noninvasive technique, based on a quantitative detection system, is routinely used for clinical diagnosis of pathologies related to an alteration of corneal morphology. The more common available instruments are based on the light reflection and on the projection of a slit light on the cornea.

Children are generally challenging patients. However, actual corneal topography devices provide fast acquisition, suitable for any less cooperative patient, as well as those in the pediatric age group. Depending on individual patient behavior, in our daily practice corneal topography can be performed in the majority of children, starting from 5 years of age.

In our experience there are three main fields for applying corneal topography in the pediatric age group:

- 1. Medium-to-high astigmatism screening
- 2. In cases of systemic disease, examination related to corneal ectasia development risk
- 3. Clinic evaluation of surgical indications in case of corneal morphology irregularities

The medium-to-high astigmatism screening must represent an ordinary indication for corneal topography: we suggest the exam in cases of greater than 3 D. In our department the youngest patient with a diagnosis of keratoconus was 6 years old, and without this periodic examination an early diagnosis would not be achieved.

Several systemic diseases can induce cornea ectasia: Down syndrome,¹ atopy,² Marfan syndrome,³ intellectual disability,⁴ and so on. Rarely, patients affected by these pathologies undergo an ophthalmological evaluation and then corneal topography before ectasia develops, sometimes because of the serious general condition.

Very special cases include congenital, postinfectious, or traumatic cornea scars. In these patients, corneal topography, combined with altitudinal and aberrometric data result as necessary, is performed, mainly in order to evaluate the opportunity for and the strategy of a surgical treatment. For example, we treated a 2-year-old patient who had a history of penetrating corneal trauma with lens removal and cornea wound suture. The patient wore contact lens and did left eye patching 5 hours daily. Two months after the emergency operation we implanted an IOL into the sulcus, and 6 months after, distance-corrected visual acuity (DCVA) was 0.2 and higher-order abberations (HOAs) for a 5.0-mm pupil were 3.39 µm. An arcuate keratotomy (AK) was then performed by femtosecond laser according to topographic elevation map inferior asymmetric steepening. Four years after AK, the patient is continuing with the contact lens and 5 hours daily patching, DCVA is 0.9, while corneal HOA error is 1.72 µm.

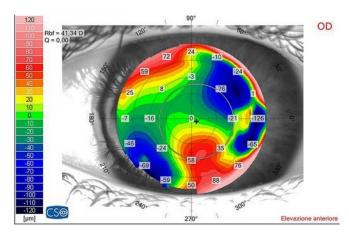


Figure 1. The right eye anterior elevation map (Sirius Scheimpflug camera, CSO; Italy) shows the inferior steepening area corresponding to the corneal scar.

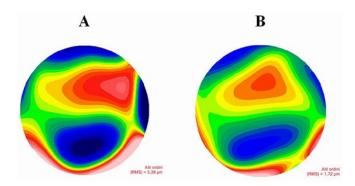


Figure 2. Corneal high-order aberrations for 5.0-mm pupil (Sirius Scheimpflug camera, CSO; Italy) measured before (A) and after (B) femtosecond laser arcuate keratotomy performed according to corneal topographic result.

In selected pediatric patients, refractive topographically guided surgical approaches could be evaluated in order to correct high irregular astigmatism, to improve anti-amblyopia treatment, and to avoid more complex procedures as well as corneal keratoplasty.

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Anterior Segment OCT: Corneal / Intraocular Surgery

Kanwal K Nischal MBBS

Intraoperative OTC for pediatric anterior segment surgery has developed quickly in the past 10 years. Initially standalone OCT units could be used in the operating room, but this meant moving the diagnostic equipment into use and moving the microscope away. Various centers developed adaptations to help integrated intraoperative OCT move forward. In the past 2 years, 2 commercially available integrated intraoperative OCT microscopes have become available.

This talk discusses the evolution of intraoperative OCT for pediatric anterior segment surgery and discusses its use in pediatric corneal surgery (deep anterior lamellar keratoplasty, penetrating keratoplasty, and Descemet-stripping automated endothelial keratoplasty) and in complex cataract surgery and its use in glaucoma surgery, both as a tool to flatten the learning curve in such cases but also as a teaching tool.

Anterior Segment OCT: Glaucoma and Other Indications

Yasmin Bradfield MD

- I. Leica (Previously Bioptigen) Anterior Segment OCT (AS-OCT)
 - A. Novel imaging
 - B. Potentially allows better understanding of the anatomic differences of eyes with glaucoma compared to nonglaucomatous eyes
 - C. Strengths of handheld AS-OCT in a pediatric population
 - 1. Easier utility in the operating room
 - 2. Ability to image very high resolution structures as small as 3 µm; and its noncontact design
 - 3. Scanning speed of 32,000 A scans per second, scan depth of 3.4 mm, very high resolution (VHR) light source for improved axial resolution without increase in patient irradiant exposure
 - D. Costs
- II. Can this imaging modality identify differences of anatomic size and structure of the Schlemm canal, even within the clinical spectrum of pediatric glaucoma?

Finding differences in quality and size of the Schlemm canal by OCT may explain why some primary congenital glaucoma patients achieve IOP control with a single surgical intervention, while others require multiple interventions.

III. Pilot Study

Comparison of ocular anterior segment features using AS-OCT in pediatric patients with and without glaucoma

A. To better understand anatomical differences in Schlemm canal and anterior segment structures in pediatric patients with and without glaucoma using handheld AS-OCT

- B. Data obtained from this study can potentially be used to guide the type of surgical intervention in infants and children with glaucoma.
- C. Potentially useful in determining the severity of glaucoma and thus aiding family counseling regarding how many surgical interventions might be required for eye pressure control
- D. Presentation of data
- IV. Other Clinical Uses
 - A. Deposits in long-standing Ahmed tube shunt in patient with Axenfeld-Reiger glaucoma
 - B. Episcleral lesion: differential diagnosis of hemangioma, lymphangioma, coagulopathy disorder with cysts, malignant tumor (rhabdomyosarcoma)
 - C. Placement of IOL behind pupillary membrane
 - D. Recurrent esotropia in patient status post transposition procedure

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Retinal Fluorescein Angiography in Pediatric Uveitis

Jennifer Hsuleejen Cao MD

- I. Uveitis Is a Potentially Blinding Disease
 - A. The pediatric population has unique challenges in presentation and examination.
 - B. Due to chronicity of disease, pediatric patients can have a greater absolute duration of disease than adults, resulting in increased lifetime risk of accumulating complications from undertreated / uncontrolled uveitis.
- II. The Presence of Retinal Vasculitis Is a Risk Factor for Poor Prognosis in Uveitis
 - A. Fluorescein angiography (FA) is routinely used for screening / monitoring for retinal vasculitis in adult intermediate and posterior uveitis.

- B. FA can be used to detect occult retinal vasculitis in patients who appear quiet based solely on clinical examination.
- C. If additional uveitis activity is detected on FA, titration of medications is warranted.
- III. FA Can Also Be a Useful Tool in Monitoring Disease Activity in Pediatric Patients

The advent of noncontact, wide-angle retinal imaging systems has facilitated the use of FAs on pediatric patients in the clinical setting without the need for general anesthesia.

In-Office Handheld ERG in Pediatric Patients

Melanie A Schmitt MD

- I. Conventional Full-Field Electroretinogram (ERG): Disadvantages
 - A. Sedation may be required in young children and infants.
 - B. High cost
 - C. Lack of availability
 - D. Technically more difficult
 - E. Time consuming
 - F. Maximum dilation preferred
- II. In-Office Handheld ERG (such as RETeval): Advantages
 - A. Less invasive
 - B. Sedation rarely necessary
 - C. Lower cost
 - D. Simple to operate
 - E. Short test time
 - F. May use on undilated pupils
 - G. Easily transportable
- III. Conditions in Which Handheld ERG Is Particularly Useful
 - A. Diagnosing early-onset inherited retinal degenerations
 - B. Monitoring drug (eg, vigabatrin) toxicity
 - C. Noncooperative patients
 - D. Patients with nystagmus

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