

015 A prospective observational study of adult divergence insufficiency esotropia.

Eric R. Crouch, Trevano W. Dean, Jonathan M. Holmes, Raymond T. Kraker, Aaron M. Miller, Courtney Kraus, Kammi B. Gunton, Michael X. Repka, Justin D. Marsh, Monte A. Del Monte, Paula A. Luke, Jason H. Peragallo, David K. Wallace

Introduction: This study was designed to assess treatment outcomes for adult onset divergence insufficiency distance esotropia.

Methods: In a nonrandomized observational study, we prospectively enrolled adults with divergence insufficiency (defined as distance esodeviation of 2^{Δ} - 30^{Δ} at least 1.25 times larger than near esodeviation by prism and alternate cover test, and distance diplopia with frequency of "sometimes," "often," or "always" in primary gaze by diplopia questionnaire). Participants were enrolled when a new treatment was being initiated (either prism, orthoptic exercises, or surgery). The primary 12-month outcome was "symptom success" defined as diplopia "rarely" or "never" straight ahead in the distance.

Results: A total of 114 participants were enrolled and initiated treatment: surgery ($n = 76$, 67%), prism ($n = 34$, 30%), or exercises ($n = 4$, 4%). Prior treatment was reported primarily in the surgery group ($n = 61$, 80%). Success criteria were met for 55 (89%; 95% CI, 78%-95%) with surgery and 17 (65%; 95% CI, 44%-83%) with prism. Success rates were high for both major types of surgery (bilateral medial rectus recession: 32 of 35 [91%]; bilateral lateral rectus resection: 10 of 11 [91%]).

Discussion: Although success rates cannot be compared directly in this nonrandomized study because there were important differences in baseline characteristics, success was common.

Conclusions: When assessed 12 months after initiating treatment, strabismus surgery or prism often successfully improves symptoms in divergence insufficiency. These data may be useful for counseling patients and for future RCTs.

016 The medial rectus is the bad actor in intermittent esotropia.

Joseph L. Demer, Robert A. Clark

Introduction: Fusional convergence controls exophoria, but failure of fusional divergence in esodeviations has been enigmatic. Magnetic resonance imaging (MRI) can clarify muscle function during divergence.

Methods: Orbital MRI was performed during binocular fusion of centered targets. Nine orthophoric controls fused monocular 4^{Δ} base-in prism at 400 cm and 10 fused 8^{Δ} base-in at 20 cm. Four patients fused acquired, intermittent esotropia averaging $24^{\Delta} \pm 4^{\Delta}$. Changes in compartmental posterior partial volumes of the horizontal rectus muscles quantified contractility.

Results: In patients and controls, both diverging lateral rectus (LR) compartments contracted symmetrically for near and distant targets, although contractility was much larger in patients ($P < 0.002$). At near in controls, only the diverging medial rectus (MR) superior compartment relaxed, while the inferior compartment remained contracted ($P < 0.03$). The normal MR did not relax during far divergence. At near in patients, the MR superior relaxed significantly more than the inferior compartment ($P = 0.005$). For near targets, MR and LR co-relaxed in the aligned eye in both patients and controls.

Discussion: The diverging MR inferior compartment co-contracts against the LR, resisting fusional divergence at near, and the entire MR fails to relax as much as the LR contracts for far targets. The MR and LR co-relax in the aligned eye when its fellow diverges to fuse intermittent esotropia.

Conclusions: The inferior compartment of the MR actively opposes fusional divergence in intermittent esotropia, not reciprocating the

LR. Poorer MR relaxation accounts for lower fusional divergence for far than near targets. Selective weakening of the inferior MR compartment may treat acquired intermittent esotropia.

017 How long does gene therapy last? 4-year follow-up of phase 3 voretigene neparovec trial in RPE65-associated LCA/inherited retinal disease.

Arlene V. Drack, Jean Bennett, Stephen Russell, Katherine A. High, Zi-fan Yu, Amy Tillman, Daniel Chung, Kathleen Z. Reape, Thomas Ciulla, Albert Maguire

Introduction: Voretigene neparovec (VN) gene therapy improves ambulatory navigation, light sensitivity, and visual field in subjects with RPE65-associated Leber congenital amaurosis /inherited retinal disease. We report Year 4 results for original intervention (OI) subjects, year 3 for delayed intervention (DI), and Y1 results for all subjects stratified by age < 10 , 11-17 or > 18 years at treatment for the primary endpoint.

Methods: Subjects were randomized to either original intervention (OI: bilateral subretinal VN at baseline) or delayed intervention (DI: VN after 1 year). Primary endpoint was change in bilateral performance on the multi-luminance mobility test (MLMT).

Results: There were no significant differences in MLMT between subjects aged ≤ 10 ($n = 13$), 11-17 ($n = 7$), and ≥ 18 years ($n = 9$) at Year 1. Mean changes in MLMT at Year 1 were maintained at Year 4 for OI and Year 3 for DI (1.7 and 2.4 light levels, respectively). At year 4, 5/20 OI subjects (ages at treatment 4, 6, 11, 11 and 34 years) showed a decrease of one light level. Three of the 5 remained stable compared to year 2 or 3. No subject declined below baseline, and 1/20 (age at treatment 16 years) gained a light level. One subject had a retinal detachment detected at year 4.

Discussion: Amblyopia may not be a major hindrance to gene therapy treatment but loss of photoreceptors in a progressive disease may affect outcome.

Conclusions: Functional vision is stable in 24 of 28 patients (86%) from one year post-treatment through 3-4 years of follow-up.

018 Stepped strabismus surgery.

Amr A. Elkamshoushy, Ahmed Kassem

Introduction: To present and evaluate a new intraoperative technique in strabismus surgery that allows further operation on additional extraocular muscles or to be skipped if the immediate intraoperative alignment is satisfactory.

Methods: This is a retrospective chart review of cases of stepped strabismus surgery from 2010 until 2018. In stepped surgery the first muscle is done under propofol IV infusion general anesthesia (GA). The anesthesia technique is modified to allow full recovery within 30 minutes in the OR. Patient is assessed in the OR. If deemed necessary, GA is given again and another muscle is operated. No adjustable sutures were utilized.

Results: A total of 56 cases were included (22 superior oblique palsy [SOP], 29 horizontal deviations and 5 thyroid eye disease). The technique was used in SOP cases with angles ranging 12^{Δ} - 25^{Δ} . Inferior oblique myectomy was done as first step in all cases and 5 cases needed additional muscle surgery. Horizontal deviations ranged from 12^{Δ} to 20^{Δ} and all cases underwent a single horizontal rectus recession. Thirteen cases required another muscle surgery. Overall reoperation rate was 9%.

Discussion: The technique was utilized in borderline cases where the decision to operate on one or two muscle was difficult to make. It obviated the need for adjustable sutures in such cases with comparable success rate.

Conclusions: Stepped strabismus surgery is a useful technique for borderline cases with the potential for reducing the number of extra ocular muscles operated on without compromising the surgical outcome.

019 Incidence of symptomatic torsional and vertical diplopia after superior rectus transposition for esotropic duane syndrome and 6th nerve palsy. Anna G. Escuder, Melanie A. Kazlas, Gena Heidary, David G. Hunter, Linda R. Dagi

Introduction: To describe the incidence of symptomatic vertical and torsional strabismus after superior rectus transposition (SRT) for esotropic Duane syndrome (DS) and 6th nerve palsy.

Methods: Retrospective chart review of pre- and postoperative sensorimotor exams on patients with 6th nerve palsy or esotropic DS treated with SRT with or without medial rectus recession (2000-2018). Patients with bilateral SRT, or treatment with additional rectus or oblique surgery were excluded.

Results: 66 patients met inclusion criteria, including 32 patients with sixth nerve palsy and 34 patients with DS. Average follow up was 2.4 years and age at surgery, 22.8 years. Average preoperative esotropia was 42^Δ (95% CI, 38.5- 46.2) and postoperative was 10.2^Δ (95% CI, 7.76-12.7). Average pre- and postoperative vertical deviation in primary gaze was 1.78^Δ (95% CI, 0.95-2.62) and 2.62^Δ (95% CI, 1.48-3.63), respectively. Abduction enhancement was performed with SR-LR loop myopexy in 47 and scleral-fixated myopexy in 7 patients. Symptomatic vertical diplopia occurred in 4 of 47 treated with loop myopexy and in 1 of 7 with scleral-fixated. None of the 66 patients developed symptomatic torsion.

Discussion: Superior rectus transposition has been advocated as an alternative to balanced vertical rectus transposition. In this largest-to-date retrospective review, 7.5% of patients developed symptomatic vertical diplopia and none developed symptomatic torsional diplopia.

Conclusions: Superior rectus transposition with or without medial rectus recession provides a muscle-sparing alternative to balanced vertical rectus transposition with similar rates of induced vertical and torsional diplopia.

Conclusions: Superior rectus transposition with or without medial rectus recession provides a muscle-sparing alternative to balanced vertical rectus transposition with similar rates of induced vertical and torsional diplopia.

020 Home tonometry redefines glaucoma drainage device management in childhood glaucoma. Michelle S. Go, Navajyoti R. Barman, Robert J. House, Sharon F. Freedman

Introduction: The postoperative management of the nonvalved Baerveldt glaucoma drainage device (GDD) presents challenges in children due to widely variable intraocular pressure (IOP) often occurring perioperatively. We evaluated the use of home tonometry in the management of Baerveldt GDDs for refractory childhood glaucoma.

Methods: As part of an ongoing prospective study involving home rebound tonometry, the families of patients receiving Baerveldt GDDs were trained to use the Icare® rebound tonometer (Ta01, Finland, Oy) and asked to document IOP, relevant symptoms, and medication changes onto a web-based data application or Excel spreadsheet. Data were analyzed for time to tube opening, multiple-day fluctuations, and various IOP trends. Clinician response to IOP fluctuations detected by home tonometry was also evaluated.

Results: Included were 19 patients (mean age, 16.1 ± 9.6 years) having Baerveldt implantation from 2015-2018 by one attending. Home tonometry detected 100% (12/12) of spontaneous tube openings, which occurred at 6.0 ± 0.5 weeks. Mean IOP decreased 32.8% (25.1 vs 16.9 mm Hg; $P < 0.01$) and 5-day IOP fluctuation decreased

from 14.5 to 6.2 mm Hg ($P < 0.05$) after tube opening. Preoperative, post-implantation, and post-opening IOP range was 11-59, 3-61, and 1-50 mm Hg, respectively. Home tonometry corroborated clinical hypotony in 5 eyes and early hypertensive phase in 9. It prompted 75 documented medication changes among 14 patients.

Discussion: Home rebound tonometry accurately detected tube opening and alarming IOP fluctuations, allowing clinicians to promptly and appropriately respond to these events.

Conclusions: Home tonometry-augmented GDD management in childhood glaucoma may improve care of these challenging patients.

021 High prevalence of sagging eye syndrome in adults with binocular diplopia. Toshiaki T. Goseki, Suh Soh Youn, Laura Robbins, Stacy L. Pineles, Federico G. Velez, Joseph L. Demer

Introduction: Sagging eye syndrome (SES), horizontal and/or vertical strabismus caused by orbital connective tissue degeneration, was first defined 10 years ago. While SES is increasingly recognized as a cause of acquired diplopia, its prevalence is unknown. We investigated SES prevalence in diplopic adults.

Methods: We reviewed all new adults over age 40 years, presenting to the UCLA strabismus division with binocular diplopia between August 2017 and September 2018. Age, gender, and type of strabismus were analyzed.

Results: We reviewed 208 total patients of mean ± SD age 67 ± 11 (range, 40-91) years of whom 113 (54%) were female. The most common cause of diplopia was SES (28.8%), followed by exotropia (10.1%), thyroid ophthalmopathy (8.2%), trochlear palsy (8.2%), abducens palsy (7.7%), decompensated esophoria (4.8%), orbital trauma (3.4%), scleral buckling (2.9%), and skew deviation (2.4%). The 63 patients with SES were older at 71 ± 9 years (range, 52-91 years, $P < 0.0001$) and more predominantly female at 63% than other patients (49%, $P = 0.02$). SES caused 15% of all diplopia in patients from ages 50-59 years, 33% from ages 60-69 years, 37% from ages 70-79 years, and 33% over age 79 years, but no diplopia under age 50 years.

Discussion: SES is the most common cause of acquired binocular diplopia in adults over 50 years old, comprising about 30% of all cases, easily surpassing cranial neuropathies and thyroid eye disease. However, SES was not encountered in patients under age 50 years.

Conclusions: It is important to recognize that SES is a very common cause of adult binocular diplopia.

022 Deep learning for monitoring rop progression. Kishan Gupta, Stanford Taylor, J. Peter Campbell, Jayashree Kalpathy-Cramer, James M. Brown, R. V. Paul Chan, Sang J. Kim, Michael F. Chiang

Introduction: To evaluate the clinical utility of quantitative image analysis using a deep learning plus disease severity score to monitor disease progression and response to treatment in patients with retinopathy of prematurity (ROP).

Methods: Images from clinical exams performed between July 2011 and December 2016 of infants in the multicenter Imaging and Informatics in ROP study were reviewed to identify babies with treatment-requiring disease, and scored by an automated deep learning algorithm with from 1 (normal retinal vasculature) to 9 (severe plus disease). Severity scores for treated and untreated eyes were compared longitudinally. The 4-week pre- and post-treatment scores with either laser or anti-vascular endothelial growth factor (anti-VEGF) were assessed.

Results: A total of 1692 eyes were analyzed. 91 eyes progressed to treatment-requiring disease. Mean severity scores of the two groups