

What's New and Important in Pediatric Ophthalmology and Strabismus for 2019

The Abridged 'All-Star' Handout

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1. AMBLYOPIA

Home use of binocular dichoptic video content device for treatment of amblyopia: a pilot study.

Mezad-Koursh D, Rosenblatt A, Newman H, Stolovitch C. *JAAPOS*. April 2018;22(2):134-138.e4.

The goal of this study was to evaluate the efficacy of the BinoVision home system as measured by improvement of visual acuity in the patient's amblyopic eye. This study was an open-label prospective pilot-trial of the system was conducted with amblyopic children aged 4-8 years at the pediatric ophthalmology unit, Tel-Aviv Medical Center, January 2014 to October 2015. Participants were assigned to the study or sham group for treatment with BinoVision for 8 or 12 weeks. Patients with amblyopia from anisometropia or strabismus were included. Patients were instructed to watch animated television shows and videos at home using the BinoVision device for 60 minutes, 6 days a week. The BinoVision program incorporates elements at different contrast and brightness levels for both eyes, weak eye tracking training by superimposed screen images, and weak eye flicker stimuli with alerting sound manipulations. Patients were examined at 4, 8, 12, 24, and 36 weeks. A total of 27 children were recruited (14 boys), with 19 in the treatment group. Median age was 5 years (range, 4-8 years). Mean visual acuity improved by 0.26 logMAR lines in the treatment group from baseline to 12 weeks. Visual acuity was improved compared to baseline during all study and follow-up appointments ($P < 0.01$), with stabilization of visual acuity after cessation of treatment. The sham group completed 4 weeks of sham protocol with no change in visual acuity ($P = 0.285$). The average compliance rate was $88\% \pm 16\%$ (50% to 100%) in treatment group. Overall, this pilot trial of 12 weeks of amblyopia treatment with the BinoVision home system demonstrated significant improvement in patients' visual acuity and appeared to be associated with good compliance. The BinoVision home system may provide an option for future treatment of amblyopia and requires further study.

Multiple-Choice Answer Form Completion Time in Children With Amblyopia and Strabismus

KR Kelly, RM Jost, A DeLaCruz, EE Birch. *JAMA Ophthalmol*. August 2018 136(8):938-41.

In this cross-sectional study completed between 2014 and 2017 at a nonprofit eye research institute to assess for a time difference in academic tasks in chil-

dren with amblyopia and/or strabismus. At the research institute, there were enrollment of 47 children with amblyopia treated for strabismus, anisometropia, or both, 18 children with non-amblyopic strabismus, and 20 normal controls. In particular, children were asked to transfer the correct answers from a standardized reading achievement test booklet to a multiple-choice answer form as quickly as possible without making mistakes or reading the text. Of the 85 included children, 40 (47%) were female, the mean (SD) age was 10.09 (0.91) years, and the last mean (SD) grade completed was 3.42 (0.92). Compared with children in the control group (mean [SD] time to completion, 230 [63] seconds), children with amblyopia (mean [SD] time to completion, 297 [97] seconds; difference, 63 seconds; 95% CI, 24-102; $P = .001$) and children with non-amblyopic strabismus (mean [SD] time to completion, 293 [53] seconds; difference, 68 seconds; 95% CI, 21-115; $P = .002$) required approximately 28% (95% CI, 20-37) more time to fill out a multiple-choice answer form. Completion time was not associated with etiology, visual acuity, or stereoacuity. In summary, this study found that longer completion time in children with amblyopia or strabismus may affect a child's performance on tests using multiple-choice answer forms and may hinder academic success.

Improved Binocular Outcomes Following Binocular Treatment for Childhood Amblyopia

Kelly KR, Jost RM, Whang Y-Z, Dao L, et al. *Invest Ophthalmol Vis Sci*. Mar 2018;59:1221-8.

The purpose of this study was to evaluate binocular outcomes in children who had received binocular treatment for childhood amblyopia. Binocular games or movies that rebalance contrast between the eyes are thought to reduce the depth of interocular suppression in children with amblyopia so they can experience binocular vision. While visual acuity gains have been reported following binocular treatment, previous studies rarely reported gains in binocular outcomes (i.e., stereoacuity, suppression) in amblyopic children. Data for amblyopic children enrolled in two ongoing studies were pooled. The sample included 41 amblyopic children (6 strabismic, 21 anisometropic, 14 combined; age 4–10 years; ≤ 4 prism diopters [PD]) who received binocular treatment (20 game, 21 movies; prescribed 9–10 hours treatment). Mean amblyopic eye visual acuity ($P < 0.001$) and mean Randot Preschool Stereoacuity improved ($P = 0.045$), and mean extent ($P = 0.005$) and depth of suppression ($P = 0.003$) were reduced from baseline at the 2-week visit (87% game adherence, 100% movie adherence). Depth of suppression was reduced more in children aged < 8 years than in those aged ≥ 8 years ($P = 0.004$). Worse baseline depth of suppression was correlated with a larger depth of suppression reduction at 2 weeks ($P = 0.001$). Although not all children had improved binocular outcomes after 2 weeks, the group as a whole did experience improved visual acuity and stereoacuity, as well as a reduction in the extent and depth of suppression. Binocular treatments that rebalance contrast to overcome suppression are a promising additional option for treating amblyopia. This study had a reasonable sample size, but the binocular outcomes will be better eluci-

dated with even larger studies, and it would be interesting to see how binocular treatments fare against traditional patching treatment with regards to binocular outcomes.

Amblyopia in High Accommodative Convergence / Accommodation Ratio Accommodative Esotropia. Influence on Bifocals on Treatment Outcome.

Tejedor J and Gutierrez-Carmona FJ *Am J Ophthalmol.* 2018; 191: 124-128.

This is a retrospective comparative case series of 61 children with high ac/a accommodative esotropia between 2011 and 2016. All patients were in single vision glasses for 2 months and were evaluated at that time as a baseline exam. At that time, 46 patients were changed to bifocals. There were 27 patients who had amblyopia at diagnosis, 21 of those still had amblyopia at the 2 month baseline exam in single vision hyperopic spectacles. 13 of amblyopic patients were placed in bifocals. Overall, the decision to add the bifocal was based on the clinician and the family. Most clinicians were in favor of the bifocals, but parents were given opportunity to opt out of the bifocal after recent studies suggested that this practice was controversial since it is unclear if this provided long term benefit. No patient required surgery for decompensated esotropia during this study. The authors demonstrated that there was faster short term improvement in amblyopia in the group of patients who used the bifocal, but that the acuities in the two groups were similar at 1 year. Similar to previous studies, the patients in the bifocal group of this study did not demonstrate improved stereoacuity compared to the non bifocal group. The conclusion of this paper is that bifocals can provide a transient advantage in the rate of improvement in vision of amblyopic eyes in patients with high ac/a accommodative esotropia. The authors point out that this difference could be due to hypoaccommodation in amblyopic eyes and not because of the alignment at near in these glasses.

2. VISION SCREENING

A comparison of Three Different Photoscreeners in Children.

Kuddusi Teberik, Mehmet Tahir Eski, Murat Kaya, Handan Ankarali. *J Ped Ophthalmol & Strabismus.* 2018;55(5):306-311

The purpose of this study is to compare the results obtained from three non-cycloplegic handheld photorefractometers with cycloplegic autorefractometry (Topcon KR-8100; Topcon Corporation, Tokyo, Japan) measurement in children. The refractive status of 238 eyes in 119 healthy children was assessed. The values acquired using photorefractometry with the non-cycloplegic Plusoptix A12 (Plusoptix GmbH, Nuremberg, Germany), Retinomax K-plus 3 (Righton, Tokyo, Japan), and Spot Vision Screener (Welch Allyn, Skaneateles Falls, NY) devices were compared with those obtained from the cycloplegic Topcon KR-8100. The

agreement between the measurements was assessed using the intraclass correlation coefficient. The mean age was 10.1 ± 3.2 years (range: 6 to 17 years). The mean spherical value for the right eyes was 0.38 diopters (D) (range: -4.50 to 6.25 D) for the Plusoptix A12; 0.45 D (range: -4.50 to 6.25 D) for the Spot Vision Screener; -1.15 D (range: -8.75 to 6.50 D) for the Retinomax K-plus 3; and 0.62 (range: -4.50 to 6.00) for the Topcon KR-8100. The mean spherical equivalent value for the right eyes was 0.41 D (range: -4.50 to 7.90 D) for the Plusoptix A12; 0.18 D (range: -4.75 to 6.13 D) for the Spot Vision Screener; -1.30 D (range: -10.50 to 6.38 D) for the Retinomax K-plus 3; and 0.67 D (range: -4.00 to 6.00 D) for the Topcon KR-8100 (for the right eyes). The authors conclude that the photorefractometer method was found to be beneficial in the measurement of refractive errors of school-aged children. However, its disadvantages are a limited measurable refractive error range and being affected by mydriatic pupils. The Plusoptix A12 photorefractometer may eliminate the need for cycloplegia in the detection of refractive errors in children. Further studies examining more cases with an extreme range of refractive errors may be needed to confirm the outcomes of this study.

Oregon Elks Children's Eye Clinic vision screening results for astigmatism.

Vaughan J., Dale T., Herrera D. and Karr D. *J AAPOS* June 2018; 22(3): 207-210.

In the Elks Preschool Vision Screening program, which uses the plusoptix S12 to screen children 36-60 months of age, the most common reason for over-referral, using the 1.50 D referral criterion, was found to be astigmatism. The goal of this study was to compare the accuracy of the 2.25 D referral criterion for astigmatism to the 1.50 D referral criterion using screening data from 2013-2014. Vision screenings were conducted on Head Start children 36-72 months of age by Head Start teachers and Elks Preschool Vision Screening staff using the plusoptix S12. Data on 4,194 vision screenings in 2014 using the 2.25 D cutoff and 4,077 in 2013 using the 1.50 D were analyzed and compared. Area under the curve (AUC) and receiver operating characteristic curve (ROC) analysis were performed to determine the optimal referral criteria. A t-test and scatterplot analysis were performed to compare how many children required treatment using the different criteria. A total of 136 (2.25 D) and 117 children (1.50 D) who were referred by the plusoptix screening for potential astigmatism and received dilated eye examinations from their local eye doctors were included. Mean subject age was 4 years. Treatment for astigmatism was prescribed to 116 of 136 using the 2.25 D setting compared to 60 of 117 using the 1.50 D setting. The authors concluded that changing the astigmatism setting from 1.5 D to 2.25 D has increased the percentage of referrals requiring treatment to 85%, reducing false positives by 34%. Of note, the negative predictive value of this screening test using the the 2.25 D cutoff is relatively low (64%).

3. REFRACTIVE ERROR

Prevalence and Risk Factors

Environmental Risk Factors Can Reduce Axial Length Elongation and Myopia Incidence in 6- to 9-Year-Old Children

Tideman JW, Polling JR, Jaddoe, VV, Vingerling JR, et al. *Ophthalmology*. 2019 Jan;126(1):127-136

It is becoming increasingly clear that an important cause of the myopia rise in the world is the changing lifestyles of school children. The goal of this study was to identify the risk factors for eye growth at a young age that may help to characterize children at risk for whom lifestyle advice and interventions could be beneficial. This study was embedded in the Generation R Study, population-based prospective cohort study of pregnant women and their children in Rotterdam the Netherlands. Children born between April 2002 and January 2006 were invited at age 6 and 9 years of age for examination which included axial length (AL) and corneal radius (CR) measured with an IOLMaster 500. Corneal radius was obtained from average of K1 and K2 from IOL master. Also, daily life activities and demographic characteristics were obtained by questionnaire. Among 4,734 children who completed examination at age 6 and 9, 3,362 children (71%) were eligible for cycloplegic refractive error measurements. Of these, 2,175 children had ocular biometry data at 9 years of age and cycloplegic refractive error. Linear regression models on AL elongation were used to create a risk score based on the regression coefficients resulting from environmental and ocular factors. The predictive value of the prediction score for myopia (≤ -0.5 diopter) was estimated using receiver operating characteristic curves. To test if regression coefficients differed for baseline AL-to-CR ratio, interaction terms were calculated with baseline AL-to-CR ratio and environmental factors. The results show that from 6 to 9 years of age, average AL elongation was 0.21 ± 0.009 mm/year and myopia developed in 223 of 2,136 children (10.4%), leading to a myopia prevalence at 9 years of age of 12.0%. Seven parameters were associated independently ($P < 0.05$) with faster AL elongation: parental myopia, 1 or more books read per week, time spent reading, no participation in sports, non-European ethnicity, less time spent outdoors, and baseline AL-to-CR ratio. The discriminative accuracy for incident myopia based on these risk factors was 0.78. Axial length-to-CR ratio at baseline showed statistically significant interaction with number of books read per week ($P < 0.01$) and parental myopia ($P < 0.01$). Almost all predictors showed the highest association with AL elongation in the highest quartile of AL-to-CR ratio; incidental myopia in this group was 24% (124/513). The authors concluded that determination of a risk score can help to identify school children at high risk

of myopia and suggest that behavioral changes can offer protection particularly in these children. Also notable in this study is that the highest effect of the environmental factors was found for those children with the highest risk of myopia.

Dim Light Exposure and Myopia in Children

Landis EG, Yang V, Brown DM, Pardue MT, et al. *Invest Ophthalmol Vis Sci.* Oct 2018;59:4804-11.

Experimental myopia in animal models suggests that bright light can influence refractive error and prevent myopia. Additionally, animal research indicates activation of rod pathways and circadian rhythms may influence eye growth. In children, objective measures of personal light exposure, recorded by wearable light sensors, have been used to examine the effects of bright light exposure on myopia. The effect of time spent in a broad range of light intensities on childhood refractive development is not known. This study evaluated dim light exposure in myopia. The authors reanalyzed previously published data to investigate differences in dim light exposure across myopic and nonmyopic children from the Role of Outdoor Activity in Myopia (ROAM) study in Queensland, Australia. The amount of time children spent in scotopic (<1–1 lux), mesopic (1–30 lux), indoor photopic (>30–1000 lux), and outdoor photopic (>1000 lux) light over both weekdays and weekends was measured with wearable light sensors. Significant differences were found in average daily light exposure between myopic and nonmyopic children. On weekends, myopic children received significantly less scotopic light ($P = 0.024$) and less outdoor photopic light than nonmyopic children ($P < 0.001$). In myopic children, more myopic refractive errors were correlated with increased time in mesopic light ($R = -0.46$, $P = 0.002$). These findings suggest that in addition to bright light exposure, rod pathways stimulated by dim light exposure could be important to human myopia development. Optimal strategies for preventing myopia with environmental light may include both dim and bright light exposure. Limitations of the study include relatively small sample size (102 patients), limited age range of children (10-15 years), and evaluating light exposure only during waking hours. Future studies should include younger children, be designed to determine causation (interventional), and also evaluate light exposure during sleep.

Myopia Prevention and Outdoor Light Intensity in a School-Based Cluster Randomized Trial.

Wu PC, Chen CT, Lin KK, Sun CC, et al. *Ophthalmology* 2018 Aug;125(8):1239-1250

This is a report from Taiwan's school-based Recess Outside Classroom Trial program to increase the outdoor time for school aged children to reduce myopia progression. The program includes recess outside the classroom, incentive-based outdoor homework and other assignments. The authors investigated the

effectiveness of this program and aimed to identify the protective light intensities necessary for such measure. A light meter was used to measure the light intensity. This is a multi-area, cluster-randomized controlled trial including 16 schools in four geographic areas with various weather conditions. A total of 693 students in grade 1 (age 6- to 7- years old) were enrolled. Two hundred sixty-seven schoolchildren were in the intervention group and 426 were in the control group. In the intervention group schoolchildren were encouraged to go outdoors at least 11 hours weekly. The intervention also incorporated near work breaks (10 minute break for every 30 minutes of near work). Data collection included eye examinations, cycloplegic refraction, noncontact axial length measurements, light meter recorders, diary logs, and questionnaires. Of note, the control group already received some intervention to minimize myopia, but not as intensive or deliberate as the intervention group. After 1 year of intervention, the authors found that the intervention group showed significantly less myopic shift and axial elongation compared with the control group (0.35 diopter [D] vs. 0.47 D; 0.28 vs. 0.33 mm; $P = 0.002$ and $P = 0.003$) and a 54% lower risk of rapid myopia progression (odds ratio, 0.46; 95% CI, 0.28-0.77; $P = 0.003$). The myopic protective effects were significant in both nonmyopic and myopic children compared with controls. Regarding spending outdoor time of at least 11 hours weekly with exposure to 1000 lux or more of light, the intervention group had significantly more participants compared with the control group (49.79% vs. 22.73%; $P < 0.001$). Schoolchildren with longer outdoor time in school (≥ 200 minutes) showed significantly less myopic shift (measured by light meters; ≥ 1000 lux: 0.14 D; 95% CI, 0.02-0.27; $P = 0.02$; ≥ 3000 lux: 0.16 D; 95% CI, 0.002-0.32; $P = 0.048$). The school-based outdoor promotion program effectively reduced the myopia change in both nonmyopic and myopic children. Thus, outdoor activities with strong sunlight exposure may not be necessary for myopia prevention. Relatively lower outdoor light intensity activity with longer time outdoors, such as in hallways or under trees, also can be considered. Despite a short duration of follow-up and weakness in gathering light exposure data outside of school, this study suggests that school based interventions can help control myopia and that less than expected light intensity may be necessary to achieve this outcome.

Reducing the Progression of Myopia

Low-Concentration Atropine for Myopia Progression (LAMP) Study: A Randomized, Double-Blinded, Placebo-Controlled Trial of 0.05%, 0.025%, and 0.01% Atropine Eye Drops in Myopia Control

Yam JC, Jiang Y, Tang SM, Law AKP et al. *Ophthalmology*. 2019 Jan;126(1):113-124

This study aimed at answering the question of efficacy and optimal concentration of low dose atropine in preventing myopia progression and comparing them to

placebo. This double-masked, placebo-controlled trial was conducted in Hong Kong. The concentrations of atropine studied were 0.05%, 0.025%, and 0.01% compared with placebo over a 1-year period. A total of 438 children aged 4 to 12 years with myopia of at least -1.0 diopter (D) and astigmatism of -2.5 D or less were included in the study. Participants were randomly assigned in a 1:1:1:1 ratio to receive 0.05%, 0.025%, and 0.01% atropine eye drops, or placebo eye drop (0.9% sodium chloride) once nightly to both eyes for 1 year. Cycloplegic refraction, axial length (AL), accommodation amplitude, pupil diameter, and best-corrected visual acuity were measured at baseline, 2 weeks, 4 months, 8 months, and 12 months. The purpose for the 2 week visit was to determine the hyperopic shift that has been reported in higher concentration of atropine in the ATOM 1 and 2 studies. Visual Function Questionnaire was administered at the 1-year visit. After 1 year, the mean SE change was -0.27 ± 0.61 D, -0.46 ± 0.45 D, -0.59 ± 0.61 D, and -0.81 ± 0.53 D in the 0.05%, 0.025%, and 0.01% atropine groups, and placebo groups, respectively ($P < 0.001$), with a respective mean increase in AL of 0.20 ± 0.25 mm, 0.29 ± 0.20 mm, 0.36 ± 0.29 mm, and 0.41 ± 0.22 mm ($P < 0.001$). The accommodation amplitude was reduced by 1.98 ± 2.82 D, 1.61 ± 2.61 D, 0.26 ± 3.04 D, and 0.32 ± 2.91 D, respectively ($P < 0.001$). The pupil sizes under photopic and mesopic conditions were increased respectively by 1.03 ± 1.02 mm and 0.58 ± 0.63 mm in the 0.05% atropine group, 0.76 ± 0.90 mm and 0.43 ± 0.61 mm in the 0.025% atropine group, 0.49 ± 0.80 mm and 0.23 ± 0.46 mm in the 0.01% atropine group, and 0.13 ± 1.07 mm and 0.02 ± 0.55 mm in the placebo group ($P < 0.001$). Visual acuity and vision-related quality of life were not affected in each group. The authors concluded that the concentrations studied did reduce myopia progression along a concentration-dependent response. All concentrations were well tolerated without an adverse effect on vision-related quality of life. Of the 3 concentrations used, 0.05% atropine was most effective in controlling SE progression and AL elongation over a period of 1 year. However, there was no difference in axial length, between the 0.01% and placebo group. There were no treatment-related adverse events. This study is significant as this is the first placebo-controlled trial looking at the effect of atropine.

Genepin-Crosslinked Donor Sclera for Posterior Scleral Contraction/ Reinforcement to Fight Progressive Myopia

Xue A, Zheng L, Tan G, Wu S, et al. *Invest Ophthalmol Vis Sci*. July 2018; 59:3564-73.

In East Asia, myopic retinopathy has become one of the leading causes of blindness and visual impairment in the elderly population. This prospective self-controlled interventional case series evaluated the efficacy of posterior scleral contraction/ reinforcement (PSCR) surgery on controlling the progressive elongation of axial length of highly myopic eyes in young patients. Forty young patients (<18-years old) with progressive high myopia received PSCR with a genepin-crosslinked donor scleral strip for one eye and the fellow eye served as concurrent control without surgery. The main outcome measurement was the change of axial length over 2 to 3 years of follow-up. Immediately after the surgery, axial

length was shortened and subsequently increased by 0.32 mm over the follow-up period. In contrast, axial length of the fellow eyes increased by 0.82 mm over the same period ($P < 0.001$, paired t-test). PSCR delayed axial elongation in eyes with or without staphyloma. No significant change of visual acuity, cornea refractive power, or retina thickness was noted between the surgery and fellow eyes. None of the patients lost visual acuity compared with the baseline. The procedure was well tolerated with only temporary corneal refractive axis shifts that recovered by the 6-month postsurgical visit. This study concluded that PSCR with genipin-crosslinked sclera is safe and effective to restrain eye globe elongation in young patients within a 2- to 3-year follow-up period. A larger study sample size with longer follow-up data should shed more light on the safety and efficacy of this PSCR procedure. In this study, the surgery was performed on the more myopic eye in each case at patient's/ parent's request, which could introduce bias.

Miscellaneous

4. VISION IMPAIRMENT

Frequency of Visual Deficits in Children With Developmental Dyslexia.

Apart Raghuram, Sowjanya Gowrisankaran, Emily Swanson, David Zurakowski, et al. *JAMA Ophthalmology* October 2018; 136 (10): 1089-1095.

This was a prospective, uncontrolled observational study from May to October 2016 in an outpatient ophthalmology clinic in 29 children with developmental dyslexia (DD) and 33 typically developing (TD) children. The authors wanted to assess the frequency of visual deficits (vergence, accommodation, and ocular motor tracking) in children with DD compared with a control group. Demographics included mean age of 10.3 years for the DD children and mean age of 9.4 years for the TD children. The authors report statistically significant accommodation deficits in the DD group compared to the TD group (55% versus 9%, respectively) and statistically significant ocular motor tracking abnormalities in DD group compared to the TD group (62% versus 15%, respectively). Overall, children in the DD group were diagnosed with more than 1 visual deficiency (79%) as compared to children in the TD group (33%) especially abnormalities in ocular motor tracking and accommodation. Reporting statistically significant visual deficiencies in children with DD as compared to TD children, the authors suggest that it is important to evaluate visual function in children with dyslexia, including an evaluation of vergence, accommodation, and ocular motor tracking. Limitations of the study is the small sample size and the unmasked examiners. Furthermore, the authors suggest that additional studies should assess if treatment of particular visual deficiencies will help improve visual symptoms for children with DD.

5. NEURO-OPHTHALMOLOGY

Detection and characterisation of visual field defects using Saccadic Vector Optokinetic Perimetry in children with brain tumours

Murray IC, Schmoll C, Perperidis A, Brash HM, et al. *Eye*. Oct 2018;32:1563-73.

The purpose of this study was to determine the ability of Saccadic Vector Optokinetic Perimetry (SVOP) to detect and characterise visual field defects in children with brain tumours using eye-tracking technology, as current techniques for assessment of visual fields in young children can be subjective and lack useful detail. This was a case-series study of children receiving treatment and follow-up for brain tumours at the Royal Hospital for Sick Children in Edinburgh from April 2008 to August 2013. Patients underwent SVOP testing and the results were compared with clinically expected visual field patterns determined by a consensus panel after review of clinical findings, neuroimaging, and where possible other forms of visual field assessment. Sixteen patients participated in this study (mean age of 7.2 years; range 2.9–15 years; 7 male, 9 female). Twelve children (75%) successfully performed SVOP testing. Of the 4 children in which SVOP failed due to poor eye tracking, one was due to heavy mascara use, one had congenital glaucoma with buphthalmos and cloudy cornea in one eye, one had extremely poor vision and unsteady fixation due to severe optic atrophy, and reason for failure in the fourth patient was not elucidated. SVOP had a sensitivity of 100% and a specificity of 50% (positive predictive value of 80% and negative predictive value of 100%). In the true positive and true negative SVOP results, the characteristics of the SVOP plots showed agreement with the expected visual field. Six patients were able to perform both SVOP and Goldmann perimetry; these demonstrated similar visual fields in every case. SVOP is a highly sensitive test that may prove to be extremely useful for assessing the visual field in young children with brain tumours, as it is able to characterise the central 30° of visual field in greater detail than previously possible with older techniques. The automated nature of SVOP requires minimal experience to operate the system, children find the test easy with engaging animations and it takes only ~5 minutes to perform. Future studies on longitudinal follow-up of children with visual pathway tumors will help determine repeatability and reliability of the test and demonstrate changes of visual field defects in relation to changes in tumour size over time and response to medical or surgical interventions.

Prevalence of Strabismus Among Children With Neurofibromatosis Type 1 Disease With and Without Optic Pathway Glioma.

Gad Dotan, Hanya M.Qureshi, Hagit Toledano-Alhadeif, Nur Azem et al. *J Ped Ophthalm & Strabismus*.2019;56(1):19-22

The purpose of this study is to evaluate the prevalence of strabismus in Neurofibromatosis type 1 (NF-1) by comparing children with normal neuroimaging to those with optic pathway glioma. A retrospective data collection of all children with NF-1 with neuroimaging studies examined at a single medical center between 2000 and 2016. Of the 198 children with NF-1 reviewed, 109 (55%) were male, 121 (61%) had normal neuroimaging, and 77 (39%) had an optic pathway glioma. Mean age at presentation was 6.3 ± 4.7 years and mean follow-up was 4.8 ± 3.1 years. Strabismus was present in 29 (15%) children and was significantly more prevalent in children with NF-1 with optic pathway glioma (21 of 77 [27%]) than in those with normal neuroimaging (8 of 121 [7%], $P < .001$). Sensory strabismus was only found in children with optic pathway glioma, accounting for most cases (12 of 21 [57%]). A strong association between strabismus and optic pathway glioma is demonstrated by an odds ratio of 5.29 ($P < .001$). Children with NF-1 with optic pathway glioma have a 4.13 times higher relative risk of developing strabismus than children with NF-1 without it ($P = .001$). The direction of ocular misalignment in children with NF-1 with optic pathway glioma was not significantly different than that observed in children without optic pathway glioma ($P = .197$, Fisher's exact test). Only 5 (17%) children with NF-1 with strabismus (3 with optic pathway glioma) underwent corrective surgery to align their eyes. The authors concluded that optic pathway glioma in children with NF-1 is associated with an increased risk of strabismus, especially sensory strabismus. Although exotropia is the most common ocular misalignment associated with optic pathway glioma, the direction of strabismus cannot be used as an accurate predictor for the presence of optic pathway glioma. Many children with NF-1 with strabismus do not undergo corrective surgery. This study's results should be interpreted within the context of its limitations. Because data were collected retrospectively based on chart reviews, it is subject to variability depending on the accuracy and completeness of records. Furthermore, because all children with NF-1 included were examined in a tertiary referral medical center, they may not accurately represent the entire pediatric population of patients with NF-1.

Visual outcomes after chemotherapy for optic pathway glioma in children with and without neurofibromatosis type 1: results of the International Society of Paediatric Oncology (SIOP) Low-Grade Glioma 2004 trial UK cohort

Falzon K, Drimtzias E, Picton S, Simmons I. *Br J Ophthalmol*. October 2018;102:1367-1371.

Although survival rates are good, children with optic pathway gliomas (OPG) can experience significant visual impairment. Management decisions are sometimes difficult due to the variable natural history of these tumors. This study aimed to report visual outcomes following chemotherapy for OPG in children with or without neurofibromatosis type 1 (NF1). The authors performed a prospective, multi-center study between 2004 and 2012. 90 children (180 eyes) with complete follow-up and visual acuity outcomes were included. 46 children had NF1 associated OPG and 44 had sporadic OPG. Visual acuity loss was the most frequent indication to initiate therapy in both groups. Average follow-up was 6.5 years. At the start of chemotherapy, 26% and 49% of eyes in NF1 and sporadic groups respectively had VA ≥ 0.7 logMAR. At completion, in the NF1 group 49% had acuity ≤ 0.2 , 23% 0.3-0.6, and 28% had VA ≥ 0.7 logMAR. In the sporadic group, 32% had ≤ 0.2 , 11% 0.3-0.6, and 57% had VA ≥ 0.7 logMAR. Children with sporadic OPG were significantly less likely to have visual outcomes ≤ 0.6 logMAR compared to the NF1 group. Overall the two groups had about the same rate of visual acuity improvement, stabilization, or worsening, but the children with sporadic OPG had a poorer visual outcome. Better initial visual acuity, increasing age, absence of post-chiasm tumor, and presence of NF1 were associated with better visual acuity outcomes. Overall timely treatment arrested the decline in VA in most children and some children regained vision.

Retinal and optic nerve changes in microcephaly: An optical coherence tomography study

Eleni Papageorgiou, Anastasia Pilat, Frank Proudlock, Helena Lee, et al.
Neurology. Aug 7 2018;91:e571-e585.

In this case-controlled prospective study, the authors seek to characterize the optic disc and retinal morphology in 27 patients with microcephaly using hand held OCT. The hypothesis is that given the relationship between ocular and brain development, there may be abnormalities in ocular development in the setting of microcephaly. With respect to the retina, 85% of patients had abnormalities on OCT, 70% with abnormalities of the fovea, and 15% with abnormalities of the retinal periphery. Findings included abnormal foveae, disruption of the ellipsoid zone, and parafoveal thinning. These findings were detectable on funduscopy in only 1/3 of patients. With respect to the optic nerve, 4 patients were noted to have optic nerve hypoplasia both by funduscopy and by OCT. rNFL thinning was also noted. The study highlights the use of OCT to identify ophthalmic changes which may not be readily detectable on clinical exam in patients with microcephaly. However the etiologies which contributed to microcephaly in these patients were heterogeneous and therefore the utility of applying these OCT findings in differentiating amongst different causes of microcephaly appears to be limited.

Idiopathic Intracranial Hypertension

Perimetry

Detection and characterization of visual field defects using Saccadic Vector Optokinetic Perimetry in children with brain tumors.

Murray IC, Schmoll C, Perperidis A, Brash HM(1), McTrusty AD, Cameron LA, Wilkinson AG, Mulvihill AO, Fleck BW, Minns RA. *Eye (Lond)*. 2018 Jun 7. doi: 10.1038/s41433-018-0135-y. [Epub ahead of print]

This study evaluated the ability of Saccadic Vector Optokinetic Perimetry (SVOP) to detect and characterize visual field defects in children with brain tumors. SVOP testing and results were compared with clinically expected visual field patterns determined by a consensus panel after review of clinical findings, neuroimaging, and where possible other forms of visual field assessment. Sixteen patients participated in this study with a mean age of 7.2 years (2.9-15 years). Twelve children (75%) successfully performed the test with a sensitivity of 100% and a specificity of 50% (positive predictive value of 80% and negative predictive value of 100%). Similar visual fields were found in 6 patients who were able to perform both SVOP and Goldman perimetry. SVOP is a highly sensitive test able to characterize the central 30° of visual field.

Optic Nerve Imaging

Retinal and optic nerve changes in microcephaly: An optical coherence tomography study

Eleni Papageorgiou, Anastasia Pilat, Frank Proudlock, Helena Lee, et al. *Neurology* August 2018;91:e571-e585

This was a prospective case-control study to investigate the morphology of the retina and optic nerve (ON) in microcephaly. The study included 27 patients with microcephaly and 27 healthy controls. All participants underwent ophthalmologic examination and handheld optical coherence tomography (OCT) of the macula and ON head. The thickness of individual retinal layers was quantified at the foveal center and the parafovea (1,000 µm nasal and temporal to the fovea). For the ON head, disc diameter, cup diameter, cup-to-disc ratio, cup depth, horizontal rim diameter, rim area, peripapillary retinal thickness, and retinal nerve fiber layer thickness were measured.

Results showed seventy-eight percent of patients had ophthalmologic abnormalities, mainly nystagmus (56%) and strabismus (52%). OCT abnormalities were found in 85% of patients. OCT revealed disruption of the ellipsoid zone, persistent inner retinal layers, and irregular foveal pits. Parafoveal retinal thickness was significantly reduced in patients with microcephaly compared to controls, nasally

(307 ± 44 vs 342 ± 19 μm , $p = 0.001$) and temporally (279 ± 56 vs 325 ± 16 μm , $p < 0.001$). There was thinning of the ganglion cell layer and the inner segments of the photoreceptors in microcephaly. Total peripapillary retinal thickness was smaller in patients with microcephaly compared to controls for both temporal (275 vs 318 μm , $p < 0.001$) and nasal sides (239 vs 268 μm , $p = 0.013$). The authors conclude that retinal and ON anomalies in microcephaly likely reflect retinal cell reduction and lamination alteration due to impaired neurogenic mitosis. OCT allows diagnosis and quantification of retinal and ON changes in microcephaly even if they are not detected on ophthalmoscopy

Myasthenia Gravis

Optic Neuritis

6. NYSTAGMUS

Long-Term Follow-up of Spasmus Nutans

Rupin N. Parikh, John W. Simon, Jitka L. Zobal-Ratner & Gerard P. Barry
Journal of Binocular Vision and Ocular Motility, 2018; 68:4, 137-139

Spasmus nutans is an acquired asymmetric, fine amplitude, high frequency nystagmus commonly accompanied by head bobbing and torticollis. The majority of cases present within the first year of life with spontaneous resolution by 4 years of age. Rarely spasmus nutans is associated with CNS lesions and patients commonly undergo neuro-imaging. Twenty-two patients with spasmus nutans were included in the study with an equal number of females and males. The series investigated the evolution of clinical findings. The authors found that the nystagmus associated with spasmus nutans does not resolve as quickly as reported in the literature. The authors only encountered the classic triad in 4 of the 22 patients. There was an association with developmental abnormalities and Down syndrome with spasmus nutans. Strabismus occurred in 64% of patients in this study, which has not been defined as a classic characteristic. Despite finding zero incidence of a space occupying lesion on neuro-imaging the authors still recommend scanning all patients with spasmus nutans.

7. PREMATURITY.

Ophthalmic Features of Premature Infants

Prematurity and Outcomes

8.ROP

ROP and Telemedicine/Screening

Development of Modified Screening Criteria for Retinopathy of Prematurity: Primary Results From the Postnatal Growth and Retinopathy of Prematurity Study.

Gil Binenbaum, Edward F. Bell, Pamela Donohue, Graham Quinn, et al. for the G-ROP Study Group. *JAMA Ophthalmology*. September 2018; 136 (9): 1034-1040.

This is a retrospective multi center cohort study of the incidence and early course of retinopathy of prematurity (ROP) from infants having ROP screening from 29 hospitals in the United States and Canada from 2006 to 2012. The authors performed a secondary analysis of the G-ROP study data. Of note, the data collection was standardized with a rigorous certification process for interpretation of medical records. To be enrolled in the study, the infant had to meet 1 of 2 conditions: (1) either eye met criteria for the ETROP type 1 or type 2 ROP or underwent treatment for ROP or (2) both eyes had mature retinal vasculature, immature vasculature in zone III with no prior ROP, or a regression of ROP of less than type 1 or type 2 ROP. Among the 7,483 infants included, 947 (12.7%) had birth weight (BW) of 1500g or more and 1440 (19.2%) had a gestational age (GA) of older than 30 weeks. Regarding the demographics, almost half the infants were white and more than 30% were African American. The authors reported that 43.1% (3224 infants) developed ROP, 6.1% (459 infants) developed type 1 ROP and 6.3% (472 infants) developed type 2 ROP. Furthermore, only 514 infants (6.9%) underwent treatment in 1 or both eyes and 147 infants (2%) had zone 1 disease. In infants with BW of less than 1251g, most had type 1 or 2 ROP (98.1%) and only approximately half of the eyes (49.4%) had retinal vasculature into zone III by 37 weeks postmenstrual age. One critical finding in this study is that these multi center cohort study involved ROP screenings of all eligible infants and not only high-risk infants. The authors remind us that for infants with BW less than 1251g, there is a higher risk of developing severe ROP and they reported 12.5% of severe ROP from low BW infants. Limitations of the study include the retrospective analysis and retinal photography was not used to confirm ROP zone or the presence of plus disease. However, study strengths include the large sample size from ROP screening programs from 29 hospitals with a

rigorous data abstraction procedure. The authors suggest that this study is helpful for ophthalmologist, neonatologists, and care coordinators by providing ROP risk profiles across GA and BW groups for these infants.

ROP and imaging

ROP and Anti-Vascular Endothelial Growth Factor Treatment

A Dosing Study of Bevacizumab for Retinopathy of Prematurity: Late Recurrences and Additional Treatments

Wallace DK, Trevano, WD, Hartnett ME, Kong L, et al. *Ophthalmology*. 125; 12 Dec 2018: 1961-1966.

Intravitreal bevacizumab (IVB) is increasingly used to treat severe retinopathy of prematurity (ROP), but it enters the bloodstream, and there is concern that it may alter development of other organs. Previously, this study group reported short-term outcomes of 61 infants enrolled in a dose de-escalation study, where one eye was injected with 0.25 mg, 0.125 mg, 0.063 mg or 0.031 mg of IVB. The lowest dosage of 0.031 mg was effective after 4 weeks in 9 out of 9 infants. There was a concern that these low doses, however, will lead to higher recurrence of the disease. Here, the authors report the late recurrences and additional treatments and structural outcomes for infants receiving lower doses of IVB. This was a masked, multicenter, dose de-escalation study including 61 premature infants with type 1 ROP. If type 1 ROP was bilateral at enrollment, then the study eye was randomly selected. In the study eye, bevacizumab intravitreal injections were given at de-escalating doses of 0.25 mg, 0.125 mg, 0.063 mg, or 0.031 mg; if needed, fellow eyes received 1 dose level higher: 0.625 mg, 0.25 mg, 0.125 mg, or 0.063 mg, respectively. After 4 weeks, additional treatment was at the discretion of the investigator. Of 61 study eyes, 25 (41%; 95% confidence interval [CI], 29%–54%) received additional treatment: 3 (5%; 95% CI, 1%–14%) for early failure (within 4 weeks), 11 (18%; 95% CI, 9%–30%) for late recurrence of ROP (after 4 weeks), and 11 (18%; 95% CI, 9%–30%) for persistent avascular retina. Re-treatment for early failure or late recurrence occurred in 2 of 11 eyes (18%; 95% CI, 2%–52%) treated with 0.25 mg, 4 of 16 eyes (25%; 95% CI, 7%–52%) treated with 0.125 mg, 8 of 24 eyes (33%; 95% CI, 16%–55%) treated with 0.063 mg, and 0 (0%; 95% CI, 0%–31%) of 10 eyes treated with 0.031 mg. By 6 months corrected age, 56 of 61 study eyes had regression of ROP with normal posterior poles, 1 study eye had developed a Stage 5 retinal detachment, and 4 infants had died of preexisting medical conditions. The fellow eyes treated with IVB had similar outcome. Due to small sample size, the study was not powered to address the relationship between the dose and recurrence rate. The authors concluded that retinal structural outcomes are very good after low-dose, as low as 0.031 mg, bevacizumab treatment for ROP, although many

eyes received additional treatment. This is the first study aimed at determining the ideal anti-VEGF dosing in infants with ROP.

Foveal Development in Infants Treated with Bevacizumab or Laser Photocoagulation for Retinopathy of Prematurity

Vogel RN, Strampe M, Fagbemi OE, Visotcky A et al. *Ophthalmology* 2018 Mar;125(3):444-452.

This study uses OCT to study the early foveal development in preterm infants and to compare this development between eyes treated with intravitreal bevacizumab or laser photocoagulation (LPC), and untreated eyes. This is an observational case series of 131 preterm infants undergoing retinopathy of prematurity (ROP) screenings, while they were inpatient. Using a handheld OCT, thickness measurements of the inner and outer retinal layers were obtained at the foveal center and the nasal and temporal foveal rims. Comparisons between treated and untreated eyes were adjusted for age and other confounding variables. They also measured weekly changes in inner and outer retinal thickness and presence of inner retinal layers, ellipsoid zone (EZ), and cystoid macular changes (CMCs). The authors found that the outer retinal thickness at the foveal center increased by 3.1 $\mu\text{m}/\text{week}$ in untreated eyes and 7.2 $\mu\text{m}/\text{week}$ in bevacizumab-treated eyes ($P = 0.038$). Eyes treated with LPC had a lower probability of having all inner retinal layers present at the foveal center (OR, 0.04; $P = 0.001$) and a lower probability of having the EZ present at the foveal center (OR, 0.07; $P = 0.024$) compared with untreated eyes. Cystoid macular changes were found in 53% of patients and 22% of imaging sessions. However, the age-adjusted incidence of CMCs was not significantly different for bevacizumab or LPC-treated eyes. The authors concluded that intravitreal bevacizumab therapy for ROP is associated with more rapid outer retinal thickening at the foveal center, whereas LPC is associated with earlier extrusion of the inner retinal layers and delayed development of the EZ at the foveal center. Limitations of the study include the fact that infants undergoing inpatient ROP screenings were included in the study hence those with more severe ROP or a more unstable clinical course were represented in the cohort. Long-term follow-up is needed to determine the visual significance of these findings.

Medi-cal and developmental outcomes of bevacizumab versus laser for retinopathy of prematurity

Kennedy KA, Mintz-Hittner HA; BEAT-ROP Cooperative Group
J AAPOS. 2018 Feb;22(1):61-65.e1.

Infants with stage 3+ retinopathy of prematurity (ROP) in zone I or zone II posterior were randomized to initial treatment with bevacizumab or laser in a multicenter trial (BEAT-ROP). The purpose of this study was to assess the effects of bevacizumab on medical and neurodevelopmental outcomes of infants enrolled in the BEAT-ROP trial at one Houston site, where very preterm (<27 weeks

gestation at birth) inborn infants are routinely seen at 18-22 months' corrected age for medical and neurodevelopmental evaluations.. Inborn infants of <27 weeks' gestational age underwent medical and standardized neurologic and developmental assessments at 18-22 months' corrected age (age after expected date of full-term delivery). Of the 18 infants enrolled at our site, 16 (7 bevacizumab, 9 laser) were evaluated for medical and neurodevelopmental outcomes at 18-28 months' corrected age. For each of the groups, the medians and ranges of growth percentiles were low compared with norms for healthy infants. The ranges for Bayley III developmental scores were also low relative to expected norms for healthy infants. There were no significant differences between the bevacizumab and laser therapy groups in weight (median percentile: bevacizumab, 18; laser, 7), length, head circumference, cerebral palsy, or Bayley scores (median Cognitive Composite Score: bevacizumab, 85; laser, 65). There was a significant difference in length of hospital stay (median days, 98 vs 140 days) favoring the bevacizumab group. In this patient cohort 2-year follow-up evaluation of infants treated with bevacizumab versus laser therapy for retinopathy of prematurity showed no adverse effects on medical or neurodevelopmental outcomes. Further randomized trials are being conducted and a 5-year developmental follow-up is planned for the one of these trials; the authors also point out that there is increasing recognition that neurodevelopmental testing at 18-24 months can overestimate severe disability and cannot detect limitations in more sophisticated domains, such as executive functioning.

ROP Epidemiology and Outcomes

ROP - Other Topics

9. STRABISMUS

Strabismus – double vision, binocular vision and visual perception

Anatomy

Strabismus – Cranial Nerve palsy

Strabismus – Childhood XT and ET

Time and Factors Affecting the Direction of Re-drift in Essential Infantile Esotropia.

Kun-Hoo Na, Yoonae A.Cho, Seung-Hyun Kim *J of Ped Ophth & Strabismus*.2018; 55(2): 93-99

The purpose of this study is to investigate the development pattern of post-operative re-drift in patients with infantile esotropia and identify factors associated with the re-drift. A total of 112 patients with infantile esotropia who underwent surgery before the age of 3 years were included. Surgical outcomes were divided into (1) consecutive exotropia: more than 8 prism diopters (PD) of exodeviation; (2) recurrent esotropia: more than 8 PD of esodeviation; and (3) monofixation syndrome: maintenance of deviations within 8 PD. The occurrence rate, time of onset and associated factors of the re-drift were evaluated. At a mean follow-up of 9.5 years, consecutive exotropia developed in 37 patients (33.0%) and recurrent esotropia in 43 patients (38.4%). Whereas 76.7% of total recurrent esotropia cases were identified within postoperative 1 year, consecutive exotropia occurred constantly over 10 years postoperatively. The mean time to consecutive exotropia development from surgery was 78.6 months, greater than that of recurrent esotropia development (8.9 months) ($P < 0.001$). In multinomial logistic regression using monofixation syndrome as the reference category, fixation preference before surgery (odds ratio [OR]: 6.64, 95% confidence interval [CI]: 2.07 to 21.32) and the rate of myopic progression (OR: 15.07 per -1.00 D/year, 95% CI: 1.23 to 184.86) were associated with consecutive exotropia, whereas increase in the angle of esodeviation on postoperative day 1 (OR: 1.15, 95% CI: 1.04 to 1.26) was correlated with recurrent esotropia. The study showed that re-drift after surgery occurred in more than 70% of patients with infantile esotropia during a long-term observation period. There was a clear difference in the development pattern between exotropic and esotropic drift; most recurrent esotropia cases appeared within postoperative 1 year, whereas consecutive exotropia occurred constantly over a long period of time. Detailed evaluation before surgery and close observation of postoperative deviations and changes in refractive status will help to determine the surgical prognosis in patients with infantile esotropia. The study has several limitations such as retrospective nature and small number of cases. Also there was an interval between the onset of infantile esotropia and the time of operation.

Strabismus – Convergence / Divergence insufficiency

Strabismus – Acquired

Adult-onset nonparalytic, small-angle hypertropia

Shah SM, Martinez-Thompson JM, Diehl NN, Mohny BG. *JAAPOS*. Dec 2018;22(6):438-440.

The purpose of this paper is to describe the prevalence and clinical features of a common but underrecognized disorder of adult vertical strabismus. The medical records of all adult (≥ 19 years of age) residents of Olmsted County, Minnesota, diagnosed with nonparalytic, small-angle hypertropia (NPSAH) from January 1, 1985, through December 31, 2004, were retrospectively reviewed for demographic and clinical features. Of 753 patients diagnosed with adult-onset strabismus, 99 (13.1%) were found to have NPSAH, yielding an annual incidence of 7.50 per 100,000 patients >18 years of age and a cumulative incidence of 1.28%. The median age at diagnosis was 71 years (range, 27-98 years); 63 (64%) of the patients were women. Diplopia was reported at the initial diagnosis in 91 patients (93.8%), with 90 (92.8%) having the diplopia in primary or reading position. The median initial angle of hypertropia was 2^Δ (range, 1^Δ - 22^Δ) at near and 2^Δ (range, 0^Δ - 12^Δ) at distance. Only 3 patients had an initial deviation of at least 11^Δ . After a median follow-up of 10.8 years (range, 6.2 months to 23.7 years), the final median angle of vertical deviation was 4^Δ (range, 0^Δ - 20^Δ) at near and 4^Δ (range, 0^Δ - 16^Δ) at distance for all 99 patients. 84.8% of the patients received prisms during the follow up period and only one underwent surgery. NPSAH is a relatively common but infrequently recognized disorder among adults. NPSAH was more prevalent among elderly and female patients in this study cohort and the vast majority presented with diplopia and a hypertropia of $\leq 10^\Delta$ that progressed over time.

Strabismus – Misc

Parent attitudes toward resident involvement in their child's strabismus surgery

Andrews H, Soni A, Green M, Quillen D. *JAAPOS*. Aug 2018;22(4):262–265.e3

This paper sought to explore patterns in parents' understanding and preferences related to ophthalmology resident participation in their child's strabismus surgery. Over a 4-week period, a survey was distributed at a suburban, academic eye center to English-speaking parents of children with strabismus who have not previously undergone, or were not being scheduled for, strabismus surgery. All of the 64 eligible parents participated in the survey. 80% and 97% of parents, respectively, indicated it was important or extremely important to be asked permission beforehand if a resident was going to assist or perform the surgery; 69% of the patients also indicated the attending surgeon should ask permission for the resident to perform the surgery, whereas only 11% believed a standard written consent was sufficient. Of the 64 respondents, 77% of the patients indicated that they would agree to a resident assisting with their child's operation while 36% of the patients stated they would agree to a resident performing the surgery. Nearly all parents in this study indicated that they would want to be informed of resident

involvement by the attending surgeon. The vast majority would consent to having an ophthalmology resident assist in their child's strabismus surgery, and more than one-third would consent to having the resident perform their child's strabismus surgery. The study was small and completed at a single site but emphasizes the importance of communication with the patient regarding resident involvement. Surgeons should keep in mind that obtaining informed consent prior to resident involvement increases transparency and highlights the importance of ophthalmology residency education.

Abnormal fixational eye movements in strabismus

Ghasia F, Otero-Millan J, Shaikh A. *Br J Ophthalmol*. February 2018;102:253-259.

Prior studies have reported greater fixation instability in patients with amblyopia as well as strabismus. In particular patients with strabismus can have disconjugate horizontal saccades. This study examined the stability of eye position during fixation with strabismus in order to correlate the severity of the instability with strabismus angle and vision. They recorded movements in 13 patients with strabismus and 16 controls using a high-resolution video eye tracker. The authors found that patients with strabismus had greater fixation instability in the deviating eye, higher intersaccadic drift velocity and greater disconjugacy in fixational saccades. Patients with small-angle strabismus and preserved binocular vision had better fixational stability than those with large-angle strabismus and absent stereopsis. Therefore they conclude that strabismus alone is sufficient to disrupt the fixational stability even in the absence of amblyopia and latent nystagmus, and fixational instability could be a screening tool to diagnose strabismus.

Long-term Surgical Outcomes in the Sagging Eye Syndrome.

Chaudhuri Z, Demer JL. *Strabismus*. 2018 Jan 3:1-5.

Sagging eye syndrome is due to degeneration of the connective tissues supporting the extraocular muscles, elongation of the muscles and anomalies in the pulley positions. The lateral rectus is the most affected of all the muscles. In order to compare the various procedures to treat this form of strabismus, the authors performed a retrospective chart review of consecutive patients who underwent strabismus surgery for small angle horizontal or vertical strabismus over a ten year period. Of the 103 cases included, 93 underwent surgery. 84 used prisms prior to surgery. Except for imbrication of the LR muscle to the SR muscle combined with superior LR transposition, all other procedures had a recurrence rates between 14 and 25%. The average age of the 15 patients with recurrences was 72 ± 7.5 years (five males), significantly higher than the 72 patients who maintained orthotropia at 66 ± 12 years ($p = 0.02$). The authors conclude that the recurrence of post-operative diplopia in patients with SES was due to progression of the age-related dehiscence of orbital connective tissue, not surgical overcorrection or un-

dercorrection. This is supported by the fact that there were no cases of surgical overcorrection, only undercorrection which manifested as symptomatic diplopia over a period of time. They recommend that counseling patients that strabismus surgery can provide relief from diplopia for an interval of time but the diplopia and strabismus may recur.

10. STRABISMUS SURGERY

Strabismus, Strabismus Surgery, and Reoperation Rate in the United States: Analysis from the IRIS Registry

Repka MX, Lum F, Burugapalli B. *Ophthalmology* 125; 10 Oct 2018: 1646-1653.

Recent population-based estimates of the prevalence of strabismus are available for children younger than 6 years of age in the United States, but are lacking for older age groups. Data are even more limited for rates of strabismus surgery in adults. The purpose of this study was to determine the prevalence of strabismus, the rate and types of strabismus surgery and the 1-year reoperation rates among all ages in the US population using the IRIS (Intelligent Research in Sight) Registry. The IRIS Registry is the nation's first comprehensive clinical registry of eye disease. It collects real-world practice patterns from electronic health records of ophthalmology practices across the United States. This study includes encounters from January 1, 2013, to December 31, 2016. As of December 31, 2016, there were 7200 ophthalmologists in 2307 electronic health record-integrated practices participating in the IRIS Registry. Of these, self-designated pediatric ophthalmologist specialists numbered 258. Based on the diagnosis codes and procedures codes, the study identified types and rates of strabismus and strabismus surgery from 2013 to 2016 with subgroups by age, sex, race/ethnicity, and region of the United States. The 1-year reoperation rate was determined for strabismus surgery performed during 2013–2015 for all age groups. A total of 30,827,185 unique patients were identified; 846,477 (2.75%) had a diagnosis of strabismus: 3.02% of male patients and 2.55% of female patients (difference = 0.47%, 95% confidence interval [CI], 0.46–0.48, $P < 0.0001$). Strabismus surgery was performed in 40,780 (0.13%) unique patients during the 4 years. The rate of surgery decreased from 1.99% for children from birth to 5 years of age to 0.05% for adults 40 years of age and older. Horizontal surgical codes were reported 38,813 times, vertical surgery codes were reported 9,304 times, and superior oblique codes were reported 711 times. Adjustable sutures were used for 3,027 patients (7.42%). Cases with a code for repeat eye muscle surgery or restrictive myopathy were reported for 6,098 patients (14.9%). Esotropia accounted for 30.06% and exotropia in 21.77% of diagnoses reported for surgery. The rate of reoperation within 1 year of a strabismus surgery was 6.72%, lowest for the group 6 to 9 years of age (3.95%) and increasing with age ($P < 0.001$) to 11.5% for patients 65 years of age and older. Overall, approximately 1 in 750 patients in the IRIS Registry received strabismus surgery (1 in 20 with a strabismus diagnosis) during a 4-year period. Reoperations during the first year after

surgery were performed for 1 in 15 patients, increasing with age at surgery. “Big” data from clinical data registries represent real-world care that can be used to develop benchmarks for clinical outcomes and to identify areas for practice improvement and training program design.

Infection following strabismus surgery

Bruce Michael Schnall and Anat Feingold *Curr Opin Ophthalmol* Sept 2018, 20:407-411

The incidence of infection following strabismus surgery is rare and estimated to be between 1/1100 to 1/1900. Most commonly the causative organisms are MRSA, *S. aureus*, *S. pneumoniae*, *S. epidermidis*, Group A strep, and *H. flue*. In children, early symptoms may be systemic such as fever and lethargy. Other signs include increasing lid edema, eye redness and pain. Infection usually occurs in the first week and symptoms often begin to appear on postoperative days 1-5. The infections can manifest in a number of ways. Sub-tenon’s abscess usually had discharge, painful eye movements and fever associated with it and is often the site of muscle reattachment which can weaken the tendon. IV antibiotics and abscess drainage are used to treat. Orbital cellulitis and abscess usually occurs within 5 days and has systemic symptoms such as fever as well as lid swelling, chemosis, restricted EOMs, and proptosis. Aggressive treatment with IV antibiotics and surgical drainage is warranted if abscess is present. Endophthalmitis occurs in 1:3500 to 1:185,000 cases. Usually visual outcome is poor and can occur with or without globe perforation. It also presents with systemic signs initially in children and then with local pain, redness and swelling. Preoperative measures can reduce the risk of post-operative infection. Preoperative topical antibiotics do not reduce the risk or severity of infection. There may be a role for IV antibiotics as they do achieve adequate levels in the conjunctiva. Children who have excessive eye rubbing, poor hygiene and are of preschool age have a higher risk of infection. Currently, the most effective method to reduce bacterial colony count prior to surgery is a drop of povidone-iodine 5% in the eye. A second application of two drops after placement of the speculum helps maintain the concentration. And a single dose of povidone-iodine 5% at the conclusion of surgery is as effective as a post-operative course of topical antibiotic/steroids. Soaking sutures in povidone-iodine 5% reduced the suture contamination rate from 28% to 9%. There is NO evidence that postoperative topical antibiotics reduces the incidence of post-op infection.

The authors undertake a review of the frequency of post-operative infection in strabismus surgery and steps to prevent its occurrence. Most notably the use of povidone-iodine 5% before and after surgery is the most effective method of infection prevention.

Horizontal muscle surgeries

Long-term outcomes After Same Amount of Bilateral Rectus Muscle Recession for Intermittent Exotropia With the Same Angle of Deviation.

Haeng-Jin Lee, Seong-Joon Kim, Young Suk Yu. *J Ped Ophthal & Strabismus*.2018;55(5):319-325

The purpose of this retrospective review is to evaluate the long-term outcomes of homogenous bilateral rectus recession in patients with the same preoperative angle of deviation in intermittent exotropia and investigate factors associated with surgical outcomes. Patients with the same preoperative angle of deviation who underwent bilateral 6-mm lateral rectus recession were observed for 2 or more years. Patients were classified into two groups based on deviation angle: success (orthophoria or exodeviation < 10 prism diopters [PD]) or recurrence (exodeviation ≥ 10 PD). Preoperative and postoperative ophthalmologic factors were compared between groups. The success and recurrence groups contained 50 and 49 patients, respectively. Preoperative maximum angle of deviation was 29.0 ± 1.8 PD at distance in the success group and 28.9 ± 1.8 PD in the recurrence group. Deviation at the 2-year follow-up was 3.7 ± 3.7 and 18.3 ± 5.3 PD in the success and recurrence groups, respectively ($P < .001$). Preoperative factors were not significantly different between groups except for presence of lateral incomitance; success group patients presented more lateral incomitance ($P = .035$). The success group also presented more esodeviation just after the operation and showed a more stable course during follow-up. Surgical outcomes of patients with 10 PD or more of esodeviation 1 week postoperatively were significantly more favorable than patients with less than 10 PD of esodeviation ($P = .027$, log-rank test). The authors conclude that the presence of lateral incomitance and early postoperative overcorrection were significantly associated with favorable surgical outcome and should be considered when planning intermittent exotropia surgery. There were some limitations in this study. First, this was a retrospective study. Second, the authors assigned groups based on 2-year postoperative deviation values because the final follow-up period was different for all patients. Even if the minimum follow-up was 2 years, a prospective study including a long-term follow-up exceeding 2 years would be necessary to validate the results of the study.

Long-term motor and sensory outcomes after surgery for the nonaccommodative component of partially refractive accommodative esotropia.

Mohan, K. and S. K. Sharma. *J AAPOS*. Oct 2018; 22(5): 356-360.

The purpose of this retrospective study was to assess the long-term motor and sensory outcomes after surgery for the nonaccommodative component of partially refractive accommodative esotropia (PRAET). A total of 47 consecutive patients ≤11 years old (median age, 3.0 years) operated for the

nonaccommodative component of PRAET and follow-up of at least 10 years, were included in the study. Excluded from the study were patients with high AC/A ratio. The mean cycloplegic refraction was $+4.22 \pm 1.65$ D (range, +1.75 to +9.00 D). Forty patients (85%) underwent unilateral medial rectus recession combined with lateral rectus resection, and 7 (15%) underwent unilateral medial rectus recession alone. The mean postoperative follow-up was 12.15 ± 2.05 years (range, 10.00-17.50 years). Overall, 23 patients (49%) had surgical success (an orthophoria or alignment within 10 PD of esotropia at near and distance); 10 (21%), decompensation (an increase of a previously controlled esotropia to >10 PD at near and distance); and 7 (15%), esotropia with a high ratio of accommodative convergence to accommodation (AC/A) or consecutive exotropia. None of the patients had residual esotropia. Kaplan-Meier survival analysis showed probabilities of surgical success of 57% at 5 years, 51% at 10 years, and 47% at 15 years postoperatively. Surgical success was achieved in 22 of 40 patients (55%) who underwent recession-resection surgery, compared to 1 of 7 patients (14%) with unilateral medial rectus recession alone ($P = 0.008$). The median age at surgery, mean cycloplegic refraction, median near and distance deviation, presence of binocular vision, and amblyopia did not predict long term outcome (decompensation, a high AC/A ratio esotropia or consecutive exotropia). Eight patients (18%) achieved stereopsis. Patients with an older age at onset (2.87 ± 1.31 years) and a shorter duration of strabismus (≤ 4 years) achieved better stereopsis. The authors concluded that in their cohort of patients with PRAET nearly half achieved a successful ocular alignment after surgery for the nonaccommodative component. Few patients achieved stereopsis. Older age at onset and a shorter duration of strabismus predicted a better stereopsis outcome. Despite its relatively small sample size, this study provides us with some insights into the of the long-term effects of surgery in partially accommodative esotropia.

Vertical muscle surgeries

Consecutive superior oblique palsy after adjustable suture spacer surgery for Brown syndrome: incidence and predicting risk.

Sharma M, MacKinnon S, Zurakowski D, Dagi LR. *J AAPOS*. Oct 2018; 22(5): 335-339.e332.

The aim of this retrospective study was to determine the incidence of significant superior oblique palsy (SOP) after adjustable superior oblique suture spacer surgery for treatment of Brown syndrome and to identify characteristics predicting its development. A total of 19 patients treated for unilateral Brown syndrome with adjustable suture spacers (2005-2016) were reviewed to identify possible association of age at surgery, spacer length,

surgeon performing procedure, severity of Brown syndrome, preoperative hypotropia in primary position and affected side gaze, and reduction in Brown restriction on postoperative superior oblique function. "Good" postoperative superior oblique function was defined as absence of hyper-tropia and diplopia in primary position and no more than intermittent di-plopia in downgaze comfortably fused with ≤ 4 PD base-down or head tilt of $< 10^\circ$. Of 19 patients, 16 (84%) achieved sufficient resolution of Brown syndrome (defined as Brown restriction of ≤ -2), but 6 (32%) developed significant SOP. Using logistic regression modelling, preoperative minimal hypotrophia on contralateral gaze was shown as the only predictor of significant SOP (likelihood ratio test = 7.11; $P = 0.008$). The authors concluded that suture spacer surgery can result in significant SOP. Risk may be predicted by magnitude of preoperative contralateral side gaze hypotropia. In their discussion the authors suggest that given the potential for spontaneous resolution of Brown syndrome and the high risk of SOP associated with minimal preoperative hypotropia in affected side gaze, conservative management would have been preferable for these patients. On the other hand, patients with > 16 PD of hypotropia in affected side gaze have $\geq 80\%$ estimated probability of retaining good postoperative superior oblique function. These patients benefit the most from surgery. This study makes an important observation regarding adjustable suture spacers in Brown. The discussion outlines the possible approach to patients with Brown drawn from this study's data.

Dose Effect and Stability of Postoperative Cyclodeviation After Adjustable Harada-Ito Surgery

Liebermann L, Leske DA, Holmes JM, et al. *Am J Ophthalmol*. 2018 December; 196: 91-95.

A retrospective cohort study of one surgeon's patients over a 20-year period was performed with the goal of reporting the dose-response relationship of the adjustable Harada-Ito surgery. The secondary goals of this study were to report the changes in the cyclodeviation over time and to recommend a target angle in the immediate postoperative period (adjustment target). There were 20 patients who underwent a unilateral adjustable advancement of the anterior fibers of the superior oblique tendon. Double Maddox rod was used to measure the cyclodeviation. The pre op measurements were compared to the 1 day and 6-week post op measurements in all patients and to the 1- and 5- year measurements when available. The authors found that there was a dose effect of 1.3 degrees per mm of advancement (\pm resection). There was a regression towards excyclodeviation between adjustment and the 6 week post op of 6.5 ± 2.6 degrees, and to a lesser extent after that. The authors recommend an initial overcorrection target of 7 degrees of incyclotorsion after adjustment. The authors point out the limitations, which include lack of complete follow up data in all patients and continued debate about the need for an adjustable procedure for torsion. This paper's most im-

portant contributions are the reminder of the cyclodeviation regression with time and the dose effect calculations.

Transposition surgeries

Sutures / Adjustables

Strabismus surgery - Misc

A randomized controlled trial comparing the efficacy of topical antibiotic steroid combination versus no treatment after fornix-incision strabismus surgery.

Elkamshoushy AA, Soni A, Alsanousy A. *J AAPOS*. Oct 2018; 22(5): 344-347.

The purpose of this prospective, randomized single-masked study was to compare comfort and inflammation in patients treated with postoperative topical antibiotic-steroid combination in one eye versus no treatment in the other eye. The study included all patients with planned symmetrical strabismus surgery via fornix incision. One eye was randomly assigned to topical postoperative tobramycin-dexamethasone and the other eye was used as control and was not treated. Patient and parent questionnaires were administered, and two masked observers assessed conjunctival injection over the muscle and wound site. A total of 70 patients completed at least 1 postoperative visit and were included. There was no statistically significant difference between the treated eye and the untreated eye in any of the studied parameters. The authors concluded that post-operative topical antibiotic steroid was not superior to no treatment in uncomplicated fornix surgery regarding patient comfort and inflammation. The authors suggest that in uncomplicated fornix strabismus surgery, consideration should be given to sparing the patient the inconvenience, cost, and potential complications of the topical medication. The study was not designed to draw any conclusions regarding the possible risk of infections, when antibiotic drops are avoided post-operatively.

Strabismus surgery outcomes without removal of scleral buckle in patients with previous retinal detachment repair

Olsigian CJ, Rothfield L, Rabina G, Cavuoto KM, et al. *JAAPOS*. Aug 2018;22(4):272-275.e1.

The goal of this paper was to report the motor and sensory outcomes of strabismus surgery following scleral buckle procedure for retinal detachment (RD) without removal of the scleral buckle. The medical records of patients who underwent strabismus surgery without removal of the scleral buckle following RD surgical repair at a tertiary referral center between 2002 and 2015 were reviewed retrospectively. Demographic data were recorded, and rates of surgical motor success (defined as horizontal deviation of $\leq 10^\Delta$ and vertical deviation of $\leq 4^\Delta$) and sensory success (resolution of diplopia) were calculated. A total of 23 patients (mean age, 58.4 ± 24.4 years; 12 males) were included in this study. The average time between the RD surgery and onset of strabismus was 11.05 ± 10.95 months (range, 1-42 months) in this patient group. The strabismus was horizontal in 6 patients, vertical in 2 patients, and combined in 15 patients. Eighteen patients (78%) presented with diplopia prior to surgery. Adjustable sutures were used in 18 patients. Final motor surgical success was achieved in 17 of 23 patients (74%), and diplopia improved in 17 of 18 patients (94%) who had preoperative fusional capability. There was no statistically significant difference in age, number of RD surgeries, macular status, time to strabismus surgery, visual acuity in the worse eye, or magnitude of preoperative horizontal and vertical deviation with regard to motor success rate and with persistence of diplopia post-operatively. In this study cohort, strabismus surgery without removal of the scleral resulted in motor success and alleviated diplopia in the majority of patients with prior RD. Strabismus surgery can be considered without removing the buckle in patients with previous RD repair with anticipation of a good success rate.

11. ANTERIOR SEGMENT

Herpetic Eye Disease Study: lessons learned

Tanja Kalezic, Mostafa Mazen, Eric Kuklinski et al Curr Opin Ophthalmol 2018, 29: 340-346 (July)

HSV has two manifestations of stromal disease: necrotizing stromal keratitis due to direct viral invasion and immune stromal keratitis (interstitial) due to immune reaction in the stroma. The authors review the HEDS study in this paper. In stromal keratitis, steroid use can reduce the progression and speed recovery with no effect on visual outcome compared to a placebo group. The use of oral acyclovir did not significantly improve the outcome either in time to resolution or visual outcome. The use of oral acyclovir may be beneficial in iritis but the trend was only noted after 3 weeks of treatment. Patients with a history of stromal keratitis and iritis were more likely to experience a recurrence than those without a

history. No benefit with a 3-week course of oral acyclovir was noted in preventing HSV stromal keratitis or iritis in the 1 year of follow up. However, the long-term use of oral acyclovir reduces the rate of recurrent HSV epithelial and stromal keratitis by 50%. The reported rates of epithelial keratitis was similar in patients who had previous disease than those who did not. In contrast, previous stromal keratitis increased the risk of recurrence by 10-fold and the risk was strongly related to the number of previous episodes. There have been new treatments since the HEDS study. Two new topical antivirals are available: gancyclovir and acyclovir. A comparative study between the two topical agents found similar efficacy and healing rates. Gancyclovir was better tolerated by patients. At this time acyclovir ointment is not FDA approved. Oral antivirals can be used instead of topical treatment for keratitis in patients with significant ocular surface disease or who cannot use topical medications. Oral acyclovir has been reported to be as effective as topical antivirals for epithelial keratitis. In addition, there are new oral antivirals such as valacyclovir and famciclovir that simplify dosing regimens, although the optimal dose for ocular disease has not been determined. History of ocular HSV is considered a relative contraindication for excimer treatment to the eye. In patients undergoing corneal crosslinking prophylactic antiviral therapy may decrease the possibility of recurrence after CXL therapy. The authors review the HEDS study results and recommendations and discuss new treatment modalities and recommendations as well.

Adenoviral keratitis: a review of the epidemiology, pathophysiology, clinical features, diagnosis, and management

Amra A. Omari and Shahzad I. Mian *Curr Opin Ophthalmol* 2018, 29: 365-372 (July)

The authors review adenoviral keratitis which is a fairly common issue for ophthalmologists. EKC is most common in adults between 20 and 40 while pharyngoconjunctival fever (PCF) is most common in children. Infections tend to spread in places where patients have close contact and especially in ophthalmic units. Overall a nosocomial infection can cost nearly \$30,000. Adenovirus 3 is most commonly implicated in PCF. The virus replicates in epithelial cells once inoculated. Patients have a follicular reaction and in EKC may have significant photophobia, with keratitis occurring 80% of the time. In PCF only 30% develop keratitis. Keratitis usually starts with epithelial microcysts leading to punctate epithelial keratopathy which can lead to subepithelial and stromal infiltrates. Chronic keratoconjunctivitis can lead to symblepharon and pseudomembrane formation. Corneal opacities can lead to astigmatism and refractive errors. Diagnosis can be made with cultures of conjunctival swab with almost 100% specificity. Rapid detection immunoassays are a promising alternative which is sensitive and specific, inexpensive and rapid. Management is achieved with hygiene and prevention of transmission, symptomatic measures such as warm compresses and lubrication as well as anti-inflammatory agents. Topical steroids may be beneficial in patients with photophobia and decreased vision.

A newer possible treatment is with povidone-iodine and dexamethasone, which provides antiseptic and symptomatic relief. Topical cyclosporine can be employed for persistent SEI's that are resistant to steroids and is safer with fewer side effects. Cidofovir was in the midst of clinical trials but was terminated due to the risk of lacrimal canalicular obstruction. PTK with mitomycin can be used for persistent corneal infiltrates. The authors summarize the approach to management of adenoviral keratitis as well as some newer treatment modalities.

12. CATARACT

Contact Lens Correction of Aphakia in Children: A Report by the American Academy of Ophthalmology

Lambert SR, Kraker RT, Pineles SL, Hutchinson AK, et al. *Ophthalmology* September. 125(9): 1452-1458.

In this review, the authors evaluated the use of 2 most commonly used contact lenses for treating aphakia in children (silicone elastomer and rigid gas permeable) to assess the visual outcome and adverse events associated with these lenses. Literature searches were conducted in January 2018 in the PubMed, Cochrane Library, and ClinicalTrials.gov databases with no date or language restrictions. These combined searches yielded 167 citations, 27 of which were reviewed in full text. Of these, 10 articles were deemed appropriate for inclusion in this assessment and subsequently assigned a level of evidence rating by the panel methodologist. The literature search identified 4 level II studies and 6 level III studies. There were insufficient data to compare visual outcomes for eyes treated using SE lenses versus RGP lenses. Silicone elastomer lenses have the advantage that they can be worn on an extended-wear basis, but they were associated with more adverse events than RGP lenses. These adverse events included microbial keratitis, corneal infiltrates, corneal edema, corneal scars, lenses adhering to the cornea, superficial punctate keratopathy, lid swelling, and conjunctival hyperemia. The lens replacement rate was approximately 50% higher for RGP lenses in the only study that directly compared SE and RGP lenses. The authors concluded that there is limited evidence in the literature on this topic. Both silicone elastomer and RGP contact lenses were found to be effective for treating aphakia in children. Silicone elastomer lenses are easier to fit and may be worn on an extended-wear basis. Rigid gas permeable lenses must be removed every night and require a more customized fit, but they are associated with fewer adverse events. The choice of which lens a practitioner prescribes should be based on the particular needs of each patient.

13. CATARACT SURGERY

Pediatric cataract surgery outcomes

Outcome of pediatric cataract surgery with intraocular injection of triamcinolone acetonide: Randomized controlled trial.

Allam G, Ellakkany R, Ellayeh A, Mohsen T, et al. *Eur J Ophthalmol*. Nov 2018 Nov;28(6):633-638.

The authors performed a randomized controlled trial evaluating the impact of intraocular injection of triamcinolone acetonide during pediatric cataract surgery in 44 eyes of 22 children. Their hypothesis was that this agent would serve to highlight vitreous as a “vitreous dye” and facilitate better anterior vitrectomy during surgery thereby reducing post operative complication. Outcomes evaluated included IOP, post op inflammation, development of PCO and post op infection. The main difference in outcome was with respect to PCO where it was noted in 1 eye which had triamcinolone and 9 eyes without injection. The authors suggest that this agent is a useful tool and should be considered in pediatric cataract surgery to improve inflammation and reduce PCO formation.

Five-Year Postoperative Outcomes of Bilateral Aphakia and Pseudophakia in Children up to 2 years of Age: A Randomized Clinical Trial

Vasavada BR, Vasavada V, Shah SK, et al. *Am J Ophthalmol*. 2018 September;193: 33-44.

This is a randomized clinical trial of 60 children (120 eyes) undergoing bilateral congenital cataract surgery. Half of the patients were randomized to receive intraocular lens (IOL) implantation and the other half were randomized to aphakia (n=30 in each group). The authors compared the outcomes of visual acuity, glaucoma, visual axis obscuration requiring surgery, and inflammation. The median age of surgery was 5 months in the aphakic group and 6 months in the pseudophakic group (p=0.56). At 5 years, the incidence of glaucoma was not different between the groups: 16% in the aphakic group and 13.8% in the pseudophakic group (p=0.82). Visually significant obscuration requiring surgery was also not different between the two groups (p= 0.79) with 10.3% of eyes in the pseudophakic group and 8% of eyes in the aphakic group needing surgery. The one significant difference was the incidence of posterior synechiae, which was significantly higher in the pseudophakic group (27.6% vs. 8%; p=0.004). In regards to vision, mean LogMAR acuity was 0.59 +/- 0.33 and 0.5 +/- 0.23 (p=0.79) with a trend toward better vision in the pseudophakic group, and more eyes in that group giving documentable vision earlier in their postoperative course. The authors point out one of the major limitations, which was that the aphakic group

had very poor compliance with contact lenses and aphakic spectacle correction vs IOL may not be a fair comparison. There is a letter to the editor from the authors of the Infant Aphakia Treatment Study (IATS) highlighting some of the differences between this study and IATS, which is an important corollary to this paper.

Pediatric cataract surgery complications

Pediatric cataract surgery – other topics

14.GLAUCOMA

Pediatric glaucoma - surgical management

Long-term efficacy of endoscopic cyclophotocoagulation in the management of glaucoma following cataract surgery in children.

Cantor A. J., Wang J., Li S., Neely D. E. and Plager D. A. *J AAPOS* June 2018; 22(3): 188-191.

The purpose of this retrospective study was to report the long-term efficacy of endoscopic cyclophotocoagulation (ECP) in pediatric glaucoma following cataract surgery (GFCS). ECP was performed on 35 eyes of 25 patients <16 years of age with GFCS. Patients were followed for a minimum of 2 years. Treatment failure was defined as consecutive postoperative intraocular pressure (IOP) of >24 mm Hg, alternative glaucoma procedure following ECP, or occurrence of visually significant complications. Analysis was performed to estimate risk factors for failure. A total of 27 aphakic and 8 pseudophakic eyes were included. Pretreatment IOP averaged 33.9 ± 7.9 mm Hg. Final IOP after a mean follow-up period of 7.2 years was 18.9 ± 8.8 mm Hg ($P < 0.001$). The success rate was 54% (19/35 eyes). The failure rate was not increased in pseudophakic patients relative to aphakic patients. Eyes requiring multiple ECP had a higher failure rate. Patients with single ECP demonstrated preserved visual acuity from baseline to final follow-up. The authors concluded that in their patient cohort, with average follow-up period of 7.2 years, ECP was useful in the treatment of pediatric GFCS. The authors also state that failed eyes demonstrated an elevated IOP at 6 months after ECP; thus an elevated IOP at this time may be a good predictor of whether a patient is likely to fail ECP.

Pediatric glaucoma – corneal biometry, OCT and visual field

Pediatric glaucoma – other topics

A Long-term Safety Study of Latanoprost in Pediatric Patients with Glaucoma and Ocular Hypertension: A Prospective Cohort Study.

Younus M, Schachar RA, Freedman SF, et *Am J Ophthalmol.* 2018 December; 196: 101-111.

This prospective cohort study aims to assess the safety of long-term latanoprost use in children. The authors prospectively studied 175 patients (102 in the latanoprost group) for 3 years. The primary endpoint of this study was a change in best-corrected visual acuity (BCVA) over the study period and secondary endpoints included corneal thickness and ocular hyperpigmentation. The authors found no significant difference in the change of BCVA in the two groups. Additionally, there were no differences found in corneal thickness or hyperpigmentation between the two groups. The authors looked at the longest eyelash and found no significant difference in lash growth in the two groups over the study time period. Of note, they did have one patient in the latanoprost group that developed iris hyperpigmentation. There was also not an appreciable difference between the two groups in terms of conjunctival hyperemia. The authors concluded that latanoprost has an acceptable safety profile in the pediatric patient. The important limitations were highlighted by the authors including the possibility that more adverse effects could be seen with a longer follow up period, specifically the possibility of developing enophthalmos and hyperpigmentation. This is an important study to the pediatric ophthalmologist who treats patients with glaucoma and is helpful when explaining the (low) risks of this therapy to parents and patients.

Steroid-induced ocular hypertension in the pediatric age group.

Al Hanaineh AT, Hassanein DH, Abdelbaky SH, El Zawahry OM¹
Eur J Ophthalmol. Jul 2018 Jul;28(4):372-377.

This prospective study evaluated the impact of a new topical corticosteroid rimexolone in pediatric patients undergoing bilateral strabismus surgery. Two cohorts of 20 patients each (40 eyes) were created. In one group, IOP post operatively in children receiving dexamethasone in one eye and rimexolone in the other was compared; in the second group, IOP post operatively in children receiving FML in one eye and rimexolone in the other was compared. Overall, the authors found that there was a statistically significant rise in IOP in the dexamethasone group. There was no significant difference in the cohort receiving FML versus rimexolone. Although ultimately the IOP rise was transient in all but 1 pa-

tient, this study highlights the need to carefully consider choice of steroid, frequency and duration in order to avoid ocular hypertension in pediatric patients.

15. REFRACTIVE SURGERY

16. GENETICS

Genome-Wide Association Study Identifies a Susceptibility Locus for Comitant Esotropia and Suggests a Parent-of-Origin Effect.

Shaaban S, MacKinnon S, Andrews C, Staffieri SE et al. *Invest Ophthalmol Vis Sci.* 2018 Aug 1;59(10):4054-4064.

This study aims to identify genetic variants conferring susceptibility to esotropia, the most common form of comitant strabismus (with highest incidence in European ancestry populations). Esotropia is believed to be inherited as a complex trait.

White European American discovery cohorts with nonaccommodative (826 cases and 2991 controls) or accommodative (224 cases and 749 controls) esotropia were investigated. White European Australian and United Kingdom cohorts with nonaccommodative (689 cases and 1448 controls) or accommodative (66 cases and 264 controls) esotropia were tested for replication. A genome-wide case-control association study was performed using a mixed linear additive model. Meta-analyses of discovery and replication cohorts were then conducted. A significant association with nonaccommodative esotropia was discovered (odds ratio [OR] = 1.41, $P = 2.84 \times 10^{-9}$) and replicated (OR = 1.23, $P = 0.01$) at rs2244352 [T] located within intron 1 of the WRB (tryptophan rich basic protein) gene on chromosome 21 (meta-analysis OR = 1.33, $P = 9.58 \times 10^{-11}$). This single nucleotide polymorphism (SNP) is differentially methylated, and there is a statistically significant skew toward paternal inheritance in the discovery cohort. Meta-analysis of the accommodative discovery and replication cohorts identified an association with rs912759 [T] (OR = 0.59, $P = 1.89 \times 10^{-8}$), an intergenic SNP on chromosome 1p31.1.

This is the first genome-wide association study (GWAS) to identify significant associations in esotropia and suggests a parent-of-origin effect. Additional cohorts will permit replication and extension of these findings. Future studies of rs2244352 and WRB should provide insight into pathophysiological mechanisms underlying comitant strabismus.

Choroideremia Gene Therapy Phase 2 Clinical Trial: 24-Month Results.

Lam BL, Davis JL, Gregori NZ, et al. *Am J Ophthalmol.* 2019 Jan;197:65-73.

Choroideremia is a rare X-linked recessive disorder in which gradual vision loss results from a mutation or deletion of the *CHM* gene and absence of the *CHM* gene product, Rab escort protein 1 (REP1), essential for intracellular trafficking. Vision loss progresses from nyctalopia in children to visual field constriction in early adulthood and ultimately to near complete blindness by age 40-50 years. There are no current treatments for choroideremia. The authors report the final results of a phase 2 high dose gene therapy clinical trial in choroideremia. Six men (aged 32-72 years) with genetically-confirmed advanced choroideremia were included in the study. Patients received subfoveal injection of AAV2-REP1 (10^{11} genome particles in 0.1 mL) in the worse-sighted eye. Primary measure was best-corrected visual acuity (BCVA) change from baseline in the treated eye compared to the untreated eye. Secondary endpoints included change from baseline in microperimetry, fundus autofluorescence, and spectral-domain optical coherence tomography (OCT). Safety evaluations included adverse events, viral shedding in body fluids, and vector antibody responses. Baseline mean ETDRS BCVA was 65.3 ± 8.8 (SD, range 56-77, 20/32-20/80) letters in the treated eyes and 77.0 ± 4.2 (69-81, 20/25-20/40) letters in the untreated eyes. At 2 years, 1 treated eye improved by 10 letters and another by 5 letters, while 1 untreated eye improved by 4 letters. All other eyes were within 2 letters of baseline. Baseline microperimetry sensitivities in the treated eyes were poor (1.2 ± 2.1 (0, 5.1) dB) and showed no significant change. No serious adverse event occurred. Two patients developed an atrophic retinal hole in a nonfunctioning macular area where baseline OCT showed preexisting thinning. Intraoperative microscope-integrated OCT allowed proper subretinal injection with avoidance of excessive foveal stretching and macular hole formation. In conclusion, the study provides evidence that treatment of choroideremia with high-dose subfoveal gene therapy has the potential to maintain BCVA, as well as improve BCVA in some cases, indicating that improvement in BCVA could be used as a viable primary outcome for future choroideremia gene therapy trials for patients with advanced choroideremia. Choroideremia gene therapy safety is enhanced with automated injection guided by real-time MIOCT. Larger-scale studies are required to ascertain the significance of these initially encouraging result

Joubert Syndrome: Ophthalmological Findings in Correlation with Genotype and Hepatorenal Disease in 99 Patients Prospectively Evaluated at a Single Center.

Brooks BP, Zein WM, Thompson AH, Mokhtarzadeh M, et al. *Ophthalmology*. 2018 Jul 25.

This article describes the ophthalmic manifestations of Joubert syndrome (JS) and draws correlations with the underlying genotype and systemic findings. JS is caused by mutations in >34 genes that encode proteins involved with primary (nonmotile) cilia and the cilium basal body. Ninety-nine patients with JS were

systematically and prospectively examined at the National Institutes of Health (NIH) Clinical Center in the setting of a dedicated natural history clinical trial. All patients underwent genotyping for JS, followed by complete age-appropriate ophthalmic examinations at the NIH Clinical Center, including visual acuity (VA), fixation behavior, lid position, motility assessment, slit-lamp biomicroscopy, dilated fundus examination with an indirect ophthalmoscope, and retinoscopy. Color and fundus autofluorescence imaging, Optos wide-field photography (Dunfermline, Scotland, UK), and electroretinography (ERG) were performed when possible. Main outcome measures included VA (with longitudinal follow-up where possible), ptosis, extraocular muscle function, retinal and optic nerve status, and retinal function as measured by ERG. Among patients with JS with quantifiable VA (68/99), values ranged from 0 logarithm of the minimum angle of resolution (logMAR) (Snellen 20/20) to 1.5 logMAR (Snellen 20/632). Strabismus (71/98), nystagmus (66/99), oculomotor apraxia (60/77), ptosis (30/98), coloboma (28/99), retinal degeneration (20/83), and optic nerve atrophy (8/86) were identified. The authors recommend regular monitoring for ophthalmological manifestations of JS beginning soon after birth or diagnosis. Result analysis demonstrates delayed visual development and the article notes that the amblyogenic time frame may last significantly longer in JS than is typical. In general, patients with coloboma were less likely to display retinal degeneration, and those with retinal degeneration did not have coloboma. Severe retinal degeneration that is early and aggressive is seen in disease caused by specific genes, such as CEP290- and AHI1-associated JS. Retinal degeneration in INPP5E-, MKS1-, and NPHP1-associated JS was generally milder. Finally, ptosis surgery can be helpful in a subset of patients with JS; decisions as to timing and benefit/risk ratio need to be made on an individual basis according to expert consultation.

17. TRAUMA

Comparison of the characteristics of retinal hemorrhages in abusive head trauma versus normal vaginal delivery.

Kim SO, Morgan LA, Baldwin AJ, Suh DW. *JAAPOS*. April 2018;22(2):139-144.

Retinal hemorrhage (RH) is one of the hallmarks of abusive head trauma (AHT); however, RH is also encountered with normal vaginal deliveries (NVD) and thus presents the clinician with a diagnostic dilemma. The purpose of this study was to compare RHs in AHT with those of NVD. Records of with AHT and NVD infants with RH evaluated from 2013 to 2015 were reviewed retrospectively. Pattern, size, extent, and severity were compared using RetCam images. Severities

were calculated using the RH grading scale. A total of 20 patients with AHT and 200 NVD infants were included. RH size was significantly larger in AHT patients compared to the NVD group (3.1 ± 0.512 vs 0.96 ± 0.046 disk diameters, resp.). The AHT group also demonstrated a higher RH incidence involving all three retinal layers compared to the NVD group (60% vs 0.6%, resp. [$P < 0.001$]). Vitreous hemorrhages were more common in the AHT group compared to the NVD group (54.3% vs 1.5% [$P < 0.001$]). Also, the grading scale demonstrated higher scores in the AHT group than the NVD group (7.15 ± 0.948 vs 3.59 ± 0.274 , resp.). Subdural hematoma was found in >95% of the AHT group as well. The authors conclude that AHT and NVD share similar retinal findings, but they also have unique differentiators. Clinicians can conclude from this study that AHT presented with more severe retinal findings than NVD, including larger RH size, a higher percentage involving all three retinal layers, a higher percentage of vitreous hemorrhages, and higher RH grading scale scores. Also, NVD retinal hemorrhages resolved quickly, within 4 weeks of birth in 95% of the patients.

Airsoft gun-related ocular injuries: long-term follow-up.

Khalaily S, Tsumi E, Lifshitz T, Kratz A, et al. *JAAPOS*. April 2018;22(2):107-109.

This paper seeks to describe the long-term ocular effects of airsoft gun pellet injuries. This study extends by 7-10 years the results of a 2010 study on the acute ocular findings related to airsoft gun pellet injuries in 59 patients, wherein the authors found a variety of anterior and posterior segment injuries, including hyphema (66%), corneal edema (61%), corneal erosions (59%), and traumatic mydriasis (25%), as well as retinal edema in (22%), retinal hemorrhages and mild vitreous hemorrhage in (2.1%), and, in 1 patient, elevated intraocular pressure and traumatic cataract. Of the 59 patients in the original study, up-to-date medical records were available for 26 (44%; 20 males). The mean follow-up time was 8 years (range, 7.2-10.3 years); the mean age, 17.1 years. Persistent abnormal findings included traumatic cataract in 3 cases (11.5%) and iris dialysis in 1 case (3.8%). In all traumatic cataract cases, cataract was not present at the time of initial examination after injury. Final mean best-corrected visual acuity was 0.92 (range 0.67-1.0), logMAR 0.03 (range 0.18-0). The authors conclude that while most acute airsoft gun-related ocular injuries are transient and do not require surgical intervention, some patients may develop significant and potentially sight-threatening ocular damage, even in the absence of significant pathologic findings at the time of the injury. They note that long-term follow-up on these patients is advisable and that use of safety goggles should be emphasized. Patients with airsoft gun-related injuries should be evaluated and followed closely due to the potential of sight-threatening damage and parents should be educated of the importance of goggles.

Visual acuity recovery following traumatic hyphema in a pediatric population.

Boese EA, Karr DJ, Chiang MF, Kopplin LJ. *JAAPOS*. April 2018;22(2):115-118.

The purpose of this paper is to determine the rate of visual recovery following hyphema caused by traumatic blunt force injury in children. The medical records of patients evaluated between July 2008 and July 2014 were reviewed retrospectively. Primary outcome measures included presenting and follow-up visual acuities. The most common injuries were sports related injuries with small projectiles being the second most common reason for injury. A total of 56 eyes of 55 children (<18 years of age) were diagnosed with hyphema following blunt force non-penetrating injury. The average patient age was 10.3 ± 3.2 years. The majority of subjects were male (78%). Presenting visual acuities ranged from logMAR 0.0 (Snellen equivalent, 20/20) to light perception. Rebleeding occurred in 4 subjects (7.1%). Visual acuity demonstrated improvement over the first 28 days following injury, with 59% achieving visual acuity of logMAR 0.0 (Snellen equivalent, 20/20) and 82% recovering vision to logMAR 0.2 (Snellen equivalent 20/30) by day 28. All but 1 patient (43 of 44 eyes, 98%) had a best-corrected visual acuity of better than or equal to logMAR 0.2 at their last recorded follow-up. The authors conclude that there is good potential for visual recovery following uncomplicated traumatic hyphema in children although the rate of recovery varies between individuals. In this patient cohort, the majority of patients had significant improvement in visual acuity within the first 28 days with the most visual recovery occurring in the first 14 days; in some children visual acuity continued to improve beyond the first month.

NON-ACCIDENTAL HEAD TRAUMA

Predictors of long-term neurological outcomes in non-accidental head injury.

Chong CF(1)(2), Misra SL(2), Escardo-Paton JA(3), Dai S(1)(2). *Eye* (Lond). 2018 Mar;32(3):608-614. doi: 10.1038/eye.2017.266. Epub 2017 Dec 8.

This study aimed to investigate the predictive values of acute findings, especially ocular, for long-term neurological outcomes. A total of 38 patients (24 males, 14 females) were included. Twelve children died acutely from the head injury. A younger age of injury ($P=0.004$) was the only statistically significant predictor of good neurological outcome as compared with absence of macular retinoschisis, unilateral retinal hemorrhage, and unilateral subdural hemorrhage. Retinoschisis was seen in 17/38 children. Nine children with macular retinoschisis died acutely, 4 suffered a degree of developmental delay. Only 4 children with retinoschisis were developmentally normal. Long-term visual acuity data was available for 18/26. Visual acuity ranged from NPL to Snellen 6/5. Retinoschisis was significantly associated with worsened visual acuity ($P<0.05$).

Conclusions. Bilateral macular retinoschisis on acute presentation of NAI is associated with a seven-fold and unilateral with a four-fold increase in the development of a poor neurological outcome and eventual death.

MISCELLANEOUS

18. RETINA

Visual Acuity in Patients with Stargardt Disease after Age 40.

Collison Ft, Fishman GA. *Retina* 2018 Dec;38(12):2387-2394.

Stargardt disease is an inherited retinal disease with a prevalence of approximately 1 in 8,000 to 1 in 10,000, making it the most common juvenile onset form of macular dystrophy. Onset of symptoms in Stargardt disease occurs most often between the ages of 8 and 16 years, but onset can also occur in adulthood and even late into adulthood. In the conventional characterization of Stargardt disease, visual acuity loss often stabilizes around Snellen acuity of 20/200 to 20/400, but this observation has been expanded in studies that have demonstrated that some proportion of patients progress to worse than 20/400 vision. Some subsets of patients with Stargardt disease have been found to maintain good acuity later in life as well. The authors sought to better define visual acuity loss in patients with Stargardt disease later in life. The most recent best-corrected visual acuities in the better-seeing eye of 221 patients with Stargardt disease over 40 years of age were recorded. Also included were the age at subjective onset for symptoms and duration of symptoms. Juvenile onset was defined as onset before age 21; adult onset was defined as onset between 21 and 40 years; and late onset was defined as onset at age 41 or later. The median age of the patients with Stargardt disease was 53.1 years. Twenty-four patients (10.9%) had worse than 20/400 best-corrected visual acuity, and none had either light perception or no light perception vision. Whereas 17 of the 52 juvenile onset patients had best-corrected visual acuity worse than 20/400, only 4 of 80 adult-onset patients and 1 of 70 late-onset patients reached this level of acuity loss. Although many patients with Stargardt disease lose visual acuity to the 20/200 to 20/400 range, and some lose visual acuity beyond 20/400, none of these patients reached either light perception or no light perception. The numbers found in this study will be valuable in counseling patients with Stargardt disease and could have value in planning treatment trials.

Association of Vitamin A Supplementation With Disease Course in Children With Retinitis Pigmentosa

EL Berson, C Weigel-DiFranco, B Rosner et al. *JAMA Ophthalmol.* May 2018;136(5):490-95.

This was a retrospective, nonrandomized comparison of vitamin A and control cohorts followed up for a mean of 4 to 5 years by the Electroretinography Service of the Massachusetts Eye and Ear Infirmary. The study included children with different genetic types of typical retinitis pigmentosa: 55 taking vitamin A and 25 not taking vitamin A. The dates for patient evaluations ranged from June 1976 to July 2016. Of note, the age-adjusted dose of oral vitamin A palmitate was $\leq 15\,000$ IU/d. Of the 55 children in the vitamin A cohort, 38 (69%) were male; the mean [SD] age was 9.1 [1.9] years; and 48 (87%) were white, 6 (11%) were Asian, and 1 (2%) was black. Of the 25 members of the control cohort, 19 (76%) were male; the mean [SD] age was 9.2 [1.7] years; and 25 (100%) were white. The estimated mean rates of change with the unadjusted model were -0.0713 loge unit/y (-6.9% per year) for the vitamin A cohort and -0.1419 loge unit per year (-13.2% per year) for the control cohort (difference, 0.0706 loge unit per year; 95% CI for the difference, 0.0149 - 0.1263 loge unit per year; $P = .01$). The adjusted model confirmed a slower mean rate of decline in the vitamin A cohort (difference, 0.0771 loge-unit per year; 95% CI for the difference, 0.0191 - 0.1350 loge-unit per year; $P = .009$). With respect to ocular safety, the mean exponential rates of change of visual field area and visual acuity and the incidences of falling to a visual field diameter of 20° or less or a visual acuity of 20/200 or less in at least 1 eye did not differ by cohort. In summary, a vitamin A palmitate supplement was associated with a slower loss of cone electroretinogram amplitude in children with retinitis pigmentosa. These findings support consideration of an age-adjusted dose of vitamin A in the management of children with the common forms of retinitis pigmentosa.

Funduscopy examination and SD-OCT in detecting sickle cell retinopathy among pediatric patients.

Jin J., Miller R., Salvin J., Lehman S., et al. *J AAPOS* June 2018; 22(3): 197-201.e191.

The purpose of this prospective study was to compare the results of fundus examination and spectral domain optic coherence tomography (SD-OCT) in detecting retinal changes in pediatric patients with sickle cell disease. Over a period of 19 months, consecutive African American patients with sickle cell disease underwent complete ophthalmologic examination including SD-OCT images of the maculas of both eyes, these were compared to age-matched African American healthy controls. A total of 69 patients (37 males, mean age 12.89 ± 4.09 , 5-20 years) with sickle cell disease (SC, 26; SS, 36; S β +, 5; S β 0 thalassemia, 2) were examined. Patients' visual acuity range was 20/20 to 20/40. On funduscopy examination, 11 of 69 showed signs of retinopathy, whereas 47 of 68 showed inner retina thinning in the watershed zone temporal to the fovea on SD-OCT. On average, SD-OCT diagnosed disease 1.78 years earlier than fundus examination. Of patients <10 years of age, 1 was diagnosed with retinopathy by

funduscopy, whereas retinal changes were evident on SD-OCT in 12 of 22 (54.5%). Fundus examination showed no significant difference in retinal findings between SS/S β 0 and SC genotypes. On SD-OCT, SS/S β 0 showed worse disease process than SC in frequency of diagnosis (82% vs 56%), bilateral involvement (87% vs 43%), and foveal involvement (18% vs 0). The authors concluded that peripheral retina could be visualized on fundus examination but not easily imaged on SD-OCT, which, however, had a higher detection rate and offered earlier diagnosis. In their patient cohort SD-OCT showed that the severity of retinal change was associated with more severe sickle cell disease genotypes (SS and S β 0). Current NIH guidelines recommend annual or biennial eye examination for patients with SCD beginning at age 10 years. This study suggests that for the purpose of early diagnosis and close monitoring of disease process, beginning routine retinal screening examinations by 10 years of age in children with SCD as recommended by the current NIH guideline may not be adequate.

Investigation of the Effect of Dietary Docosahexaenoic Acid (DHA) Supplementation on Macular Function in Subjects with Autosomal Recessive Stargardt Macular Dystrophy.

MacDonald IM, Sieving PA. *Ophthalmic Genet.* 2018 Aug;39(4):477-486.

Currently, there is no treatment available to individuals affected by Stargardt macular dystrophy. Docosahexaenoic acid (DHA) is the major very long chain polyunsaturated fatty acid of the retina and is found in high concentration in the photoreceptor cells. The major source of DHA for humans is directly through the diet. The North American diet is rich in fats, but it appears that our diets are relatively poor in omega-3 fatty acids, including DHA. Deficiency of DHA has been implicated as a factor in macular degeneration. The purpose of this study was to test the effect of DHA dietary supplementation on macular function in patients with Stargardt disease. It was a single center, double-masked, randomized placebo-controlled trial of 11 subjects (2 males, 9 females) with Stargardt disease in a crossover design. Six participants were randomized to two sequences of three month periods of DHA supplementation (2000 mg/day) followed by three months of placebo. Five participants were randomized to the opposite sequence. All participants were evaluated with a food frequency and NEI-VF25 questionnaires, complete ophthalmic examination, multifocal electroretinography (ERG, primary outcome), 30-Hz flicker ERG, Humphrey 10-2 visual field, D15 color tests and serum lipid analysis. During periods of DHA supplementation, serum rose and then fell with transition to periods of placebo. None of the participants experienced greater than 20% change from baseline values of the mfERG during periods of DHA supplementation or placebo, while the average change in peak amplitude and phase angle of the flicker ERG remained similar at all visits. No significant change was observed for any of the secondary outcome measures. Eight adverse events occurred but these were not considered to be due to the treatment. The author found no perceived effect of DHA supplementation on macular

function. This study will help design future studies of the effect of DHA supplementation on retinal function in cohorts with retinal dystrophies.

Retinal Detachment Surgery in a Pediatric Population

Read SP, Aziz HA, Kuriyan A, et al. *Retina* July 2018; 38:1393-1402.

Pediatric retinal detachments (RDs) are unique in etiology, anatomy, and prognosis compared to those in the adult population. Mechanisms include tractional (TRD), rhegmatogenous RD, traumatic, and other types such as exudative or hemorrhagic. This retrospective consecutive case series of patients clinically diagnosed and undergoing surgery for RD between birth and 15 years of age during an 11 year period at a single academic institution examined visual and anatomic outcomes. 206 patients (231 eyes) were included in the study; 25 (12%) had bilateral RDs. 67 patients (29%) had TRD (ROP, PFV, or FEVR), 51 (22%) had rhegmatogenous RD (myopia, X-linked retinoschisis, or Stickler syndrome), 60 (26%) had traumatic RD, and 52 (23%) were due to other types of RD such as Coats disease or coloboma. Presenting BCVA better than 20/200 correlated with better final BCVA. Anatomical success was strongly correlated with visual acuity outcome and was significantly more likely in rhegmatogenous RD versus TRD. The likelihood of obtaining a final BCVA > 20/200 was poorer in TRD (10%) compared to rhegmatogenous RD (39%) or traumatic RD (28%).

This study is somewhat limited by its retrospective nature, lack of long-term follow-up in some patients (average time of post-operative follow-up was 48 months with range 3 months to 12.9 years), and missing or incomplete records in some patients, often as a byproduct of the tertiary nature of pediatric RD referrals coming from distant locales. Long-term sequelae of repair and involvement of the contralateral eye could often not be assessed. In addition, grouping the etiologies of the RDs was subjective, especially in cases with possible combined etiology. However, this large series of pediatric RDs confirms ROP and trauma as the most common etiologies. Although visual and anatomic outcomes vary among categories of RD, rhegmatogenous RDs were associated with the best anatomical success and globe conservation outcomes, and TRDs generally had poorer outcomes.

OCT IMAGING IN DISEASE

OCT IMAGING – DATA ON NORMAL EYES

COAT'S DISEASE **MISCELLANEOUS**

19.RETINOBLASTOMA / INTRAOCULAR TUMORS

RETINOBLASTOMA

Ophthalmic Vascular Events after Primary Unilateral Intra-arterial Chemotherapy for Retinoblastoma in Early and Recent Eras

Dalvin LA, Ancona-Lezama DA, Lucio-Alvarez JA, Masoomian B, et al. *Ophthalmology* 125;11 Nov 2018: 1803-1811.

The purpose of this study was to assess risk factors for ophthalmic vascular events after intra-arterial chemotherapy (IAC) for retinoblastoma. Although IAC is efficacious in achieving tumor control, it can lead to thromboembolic or hemorrhagic events due to the technique itself or secondary to chemotherapy-induced toxicity. Ophthalmic vascular events include choroidal ischemia, branch or central retinal artery occlusion, ophthalmic artery spasm or occlusion, vitreous hemorrhage and others. Although early series reported ophthalmic vascular event rates as high as 35%, more recent series have described a lower rate of 1%. In this study, the authors conduct a retrospective, consecutive, comparative analysis to describe ophthalmic vascular events at a single center during two time periods: early IAC era (2009–2011) compared with the recent era (2012–2017). The study population included patients who received unilateral IAC as primary treatment for retinoblastoma from January 1, 2009, to November 30, 2017, at Wills Eye Hospital. All patients underwent complete eye exam under anesthesia prior to administration of IAC and then monthly exams were performed, which included anterior and posterior indirect ophthalmoscopy, B-scan ultrasonography, RetCam fundus photography, Fluorescein angiography, and OCTS as needed. After tumor control was achieved, the interval between examinations under anesthesia was extended. Records were reviewed for patient demographics, tumor features, IAC parameters, and treatment-related vascular events. Change in event rates over time were assessed using Poisson regression analysis, with Spearman's rho used to test correlation. There were 243 chemotherapy infusions in 76 eyes of 76 patients, divided into early (22 eyes, 57 infusions) and recent (54 eyes, 186 infusions) eras. Intra-arterial chemotherapy consisted of melphalan (243 infusions), topotecan (124 infusions), and carboplatin (9 infusions). A comparison (early vs. recent era) revealed fewer mean number of infusions (2.6 vs. 3.4, $P = 0.02$) with similar mean patient age and presenting tumor features. Event rates decreased over time ($P < 0.01$), with fewer ophthalmic vascular events (early era vs. recent era) in the recent era (59% vs. 9% per eye, 23% vs. 3% per infusion, $P < 0.01$), including peripheral retinal nonperfusion (5% vs. 2% per eye, $P = 0.50$), vitreous hemorrhage (9% vs. 2%, $P = 0.20$), subretinal hemorrhage (0% vs. 2%, $P = 0.99$), branch retinal vein occlusion (5% vs. 0%, $P = 0.29$), choroidal ischemia (14% vs. 4%, $P = 0.14$), and ophthalmic artery spasm/occlusion (27% vs. 0%, $P < 0.01$). Event rates did not correlate with patient age ($P = 0.75$), tumor diameter ($P = 0.32$), tumor thickness ($P = 0.59$), or

cumulative dosage of melphalan ($P = 0.13$) or topotecan ($P = 0.59$). There were no IAC-induced vascular events in 72 infusions of 21 consecutively treated eyes in 2016 to 2017. This study shows that the ophthalmic vascular events after IAC have decreased from the early era (2009–2011) through the current era (2012–2017) at this center. Experience performing this highly specialized procedure could be an important factor predicting IAC-related vascular events. There were technical changes made between the two time periods. This includes pulsatile delivery of the infusion, eliminating the guide wire and advancing the catheter only to the ostium of the ophthalmic artery. This alone does not explain the observed change. The later time period also eliminated the use of carboplatin but it does not show more toxicity compared to melphalan in non-human primate studies. The authors surmise that the technique and experience is what accounts for the reduced rate of the ophthalmic vascular events.

Strabismus in retinoblastoma survivors with long-term follow-up

Fabian ID, Stacey AW, Naeem Z, Onadin Z, et al. *J AAPOS*. Aug 2018;22(4):276.e1–276.e7

The goal of this paper was to report the long-term strabismus rate in salvaged retinoblastoma (Rb) patients and investigate possible risk factors leading to strabismus in these patients. The medical records of patients with Rb presenting at a single institution over a 9-year period were reviewed retrospectively with regard to ocular alignment outcomes after long-term follow-up. A total of 64 eyes of 42 patients (22 bilateral cases which consisted of 52% of the patients) were included, presenting with International Intraocular Retinoblastoma Classification (IIRC) in the worse eye as follows: group A ($n = 1$), B ($n = 16$), C ($n = 12$), D ($n = 11$), no Rb ($n = 2$). Fifteen patients (36%) were initially referred because they had no family history of Rb. Mean age at presentation was 8.2 months (range, 0.3–58.3 months). Overall treatments in this group of patients included intravenous chemotherapy (62 eyes), intraophthalmic artery chemotherapy (10 eyes), brachytherapy (11 eyes), transpupillary thermotherapy (22 eyes), cryotherapy (47 eyes), and external beam radiotherapy (4 eyes). At final follow-up (mean, 93.7 months), 69% of patients had strabismus, with exotropia being the most common type ($n = 18$), followed by esotropia ($n = 8$), and alternate exotropia/esotropia ($n = 3$). On univariate analysis, the worse eye group IIRC and cTNMH, sporadic cases, strabismus, and foveal tumor at presentation were found to be significantly associated with the presence of strabismus at the final follow-up ($P \leq 0.043$). On multivariate analysis, only foveal involvement was found to be significant ($P < 0.001$). The authors find that strabismus, exotropia in particular, is a common adverse sequela following successful conservative treatment for Rb, with 69% of the present cohort having some type of deviation after long-term follow-up, for which foveal tumor at presentation was found to be a significant risk factor. This information can be helpful in guiding parental expectations in this group of patients.

Screening Children at Risk for Retinoblastoma: Consensus Report from the American Association of Ophthalmic Oncologist and Pathologists

Skalet AH, Gombos DS, Gallie BL, Kim JW et al. *Ophthalmology* 2018 Mar;125(3):453-458.

This is a consensus statement for surveillance guidelines for children at risk for development of retinoblastoma. A patient "at risk" was defined as a person with a family history of retinoblastoma in a parent, sibling, or first- or second-degree relative. Of note, the majority of at-risk relatives who do not carry the RB1 mutation do not require specific retinoblastoma screening. Key recommendations are as follows: (1) Dedicated ophthalmic screening is recommended for all children at risk of retinoblastoma above the population risk. (2) Frequency of examinations is adjusted on the basis of expected risk for RB1 mutation. (3) Genetic counseling and testing clarify the risk for retinoblastoma in children with a family history of the disease. (4) Examination schedules are stratified on the basis of high-, intermediate-, and low-risk children. (5) Children at high risk for retinoblastoma require more frequent screening, which may preferentially be examinations under anesthesia. Refer to the risk stratification in the paper based on the RB1 screening.

NON-RETINOBLASTOMA

20. ORBIT

Intravenous Steroids With Antibiotics on Admission for Children With Orbital Cellulitis

Lena Chen, Nora Silverman, Andrew Wu, Roman Shinder *Ophthal Plast Reconstr Surg* 2018;34:205–208

This prospective comparative interventional study compared the outcomes of children with orbital cellulitis treated with intravenous (IV) dexamethasone and antibiotics on admission to patients treated with antibiotics alone. The study looked at forty-three children admitted to a tertiary institution with orbital cellulitis. On admission, all patients were started on broad spectrum IV antibiotics and parents were offered IV dexamethasone (0.3 mg/kg/d every 6 hours for 3 days). Patients whose parents refused steroid treatment served as the control group. Twenty-eight (65%) patients received IV steroids and antibiotics on admission while 15 (35%) received IV antibiotics alone. Children who received IV steroids had significantly shorter hospital stays than those who did not receive steroids (3.8 ± 0.2 days vs. 6.7 ± 0.3 days; $p < 0.001$). This was true both for children who underwent surgery for subperiosteal abscess (5/28 with steroids, 3/15 without; 5.0 ± 0.7 days vs. 7.3 ± 1.2 days; $p = 0.011$) and for those who did not require

surgical intervention (23/28 with steroids, 12/15 without; 3.6 ± 0.6 and 6.5 ± 1.0 days; $p < 0.001$). Side effects of steroid treatment were considered mild (hyperactivity and insomnia) and did not require termination of therapy. Children who received steroids had a shorter hospital stay than those who did not. During follow up, all study patients had returned to their baseline health without any cases of decreased vision or disease recurrence. The results of the current study give additional evidence to the relative safety and efficacy of systemic steroid use concurrently with IV antibiotics in children with orbital cellulitis. This is the first study to recommend IV steroids on hospital admission and a standardized dosing regimen.

21. OCULOPLASTICS

Balloon Dacryoplasty for Congenital Nasolacrimal Duct Obstruction: A Report by the American Academy of Ophthalmology: Ophthalmic Technology Assessment

Wladis EJ, Aakalu VK, Yen MT, Bilyk JR, Sobel RK et al. *Ophthalmology* 125; 10 Oct 2018: 1654-1657.

Balloon dacryoplasty has emerged as a popular option to address recalcitrant nasolacrimal duct obstructions. This technique involves passing a lubricated, inflatable balloon along a guide wire into the nasolacrimal duct and through the level of obstruction. The balloon is then inflated for 90 seconds to dilate the obstruction, deflated, and reinflated a second time, or removed. The goal of this study was to determine the efficacy and adverse events of balloon dacryoplasty. A literature search was last performed in September 2017 in the PubMed database to identify all reports of balloon dacryoplasty. All searches up to and including the last search were limited to the English language, and they yielded 104 articles that were assessed for relevancy. Thirty-six articles were selected for full review, and 8 of these were selected for inclusion in this assessment and assigned a quality of evidence rating by the panel methodologist. Three of the 8 studies included in this assessment were rated level II, and 5 were rated level III. Success rates varied from 75% to 100%. Only 2 complications were identified, and these were cases of self-limited postoperative emesis. The 2 studies that compared balloon dacryoplasty with lacrimal stenting reported that outcomes were comparable between the 2 techniques. Although level I evidence was not available, the studies that were included in the literature review indicate that balloon dacryoplasty is a safe, effective procedure to address congenital nasolacrimal duct obstruction that persists after standard probings. The outcomes of this intervention are similar to those of lacrimal stenting, and the absence of an implanted stent theoretically reduces the risk of complications. This review did not examine the age at initial probing or balloon dacryoplasty. The optimal time for balloon dacryoplasty is also not addressed. It is important to note that there was no level 1 evidence in the literature for this review.

Ophthalmic Pyogenic Granulomas Treated With Topical Timolol—Clinical Features of 17 Cases

Lauren N. DeMaria, Nora K. Silverman, Roman Shinder *Ophthalm Plast Reconstr Surg* Nov/ Dec 2018;34:579–582

Topical timolol has been increasingly demonstrated to be an effective treatment for pyogenic granulomas (PG). The authors review the treatment outcomes of 17 patients with ocular PG treated with topical timolol. Retrospective interventional study of 17 patients with ocular PGs treated with timolol 0.5% solution. Patient demographics, clinical features, treatment response, and recurrence were noted. Nine females and 8 males with a mean age of 23 years (range, 3–67 years) were included. Mean duration of disease prior to treatment was 3.81 months (range, 0.25–11 months). Etiologies included chalazia (12 cases, 71%), postsurgical (4, 24%) and trauma (1, 6%). Five patients (29%) had treatment with topical steroids prior to presentation. Fifteen patients (88%) had PG located on the palpebral conjunctiva and 2 (12%) involving the bulbar conjunctiva. Mean lesion size was 5.06 × 6.06 mm (range, 3–8 × 3–18 mm). Fifteen patients (88%) had complete lesion resolution with a mean treatment duration of 3.07 weeks (range, 2–5 weeks) and no adverse events or recurrences with a mean follow up of 9.47 months (range, 6–27 months). Two patients (12%) underwent lesion excision after 6 weeks of timolol failed to yield resolution. Topical timolol appears to be a well-tolerated nonsurgical treatment of ocular PG in both children and adults. Clinicians may wish to consider topical timolol to treat PG as opposed to topical steroids, given the inherent risk of steroid response ocular hypertension and the difficulty to measure intraocular pressure in younger children who require general anesthesia for excision.

22.INFECTIONS

Randomized, Controlled, Phase 2 Trial of Povidone-Iodine /Dexamethasone Ophthalmic Suspension for Treatment of Adenoviral Conjunctivitis

Pepose JS, Ahuja A, Liu E, et al. *Am J Ophthalmol*. 2018 October;194:7-15.

The authors of this multicenter, randomized, vehicle-controlled, double-masked trial aimed to evaluate the efficacy and safety of a 0.6% povidone-iodine (PVP-I) and 0.1% dexamethasone suspension in patients with acute adenoviral conjunctivitis. Adults with a positive rapid adenovirus screening test were randomized to PVP-I 0.6%/dexamethasone 0.1%, PVP-I 0.6% alone, or vehicle, bilaterally, four times per day for 5 days. Patients were examined on days 3, 6, and 12 with the end points of clinical resolution and adenoviral eradication. The authors found

that the proportion of eyes with adenoviral eradication at day 3 and day 6 were higher in the PVP-I/dexamethasone group than in the other two groups. Additionally, the PVP-I/dexamethasone group had a higher proportion of clinical resolution at day 6 than the other two groups (31.3% vs. 10.9% (vehicle) and 18.0% (PVP-I)). The authors concluded that the PVP-I/dexamethasone treatment was safe and improved eradication of the adenovirus and clinical resolution. They discussed that the drop tolerability was as good in the PVP-I/dexamethasone group compared to the vehicle group. And one of the major limitations of this study is that it was only tested on adults (though included here because it certainly is relevant to the pediatric ophthalmologist).

23.PEDIATRICS/ INFANTILE DISEASE/ SYNDROMES

The Phenotypic Spectrum of Albinism.

Kruijt CC, de Wit GC, Bergen AA, Florijn RJ et al. *Ophthalmology*. 2018 Dec;125(12):1953-1960.

This retrospective cohort study aims to describe the phenotypic spectrum of a large cohort of albino patients, to investigate the relationship between the ocular abnormalities and the visual acuity (VA), and to define diagnostic criteria for the white population. The authors also estimated the prevalence of albinism in The Netherlands. They investigated the phenotype of 522 patients with albinism from the databases of Bartiméus (452 patients), Leiden University Medical Center (44 patients), and the Academic Medical Center Amsterdam (26 patients).

Collected data included clinical, genetic, and electrophysiologic data of patients with albinism. Grading schemes for iris translucency, fundus hypopigmentation, and foveal hypoplasia were utilized. The main outcome measures were visual acuity, nystagmus, iris translucency, fundus pigmentation, foveal hypoplasia, and misrouting.

Nystagmus was absent in 7.7% (40/521), iris translucency could not be detected in 8.9% (44/492), 3.8% (19/496) had completely normal fundus pigmentation, 0.7% (3/455) had no foveal hypoplasia, and misrouting was not established in 16.1% (49/304). The VA varied from -0.1 to 1.3 logarithm of the minimum of angle of resolution (logMAR). The foveal hypoplasia grading correlated best with the VA ($r = 0.69$, $P < 0.001$), whereas iris translucency, fundus pigmentation, and misrouting did not predict the VA significantly. The authors estimated a prevalence of albinism in The Netherlands of at least 1:12 000.

The authors conclude that none of the characteristics of albinism were consistently present in this cohort. To be able to distinguish albinism from other conditions with similar ocular features, especially in northern and western European countries, they propose major and minor clinical criteria. Major criteria would be

(1) foveal hypoplasia grade 2 or more, (2) misrouting, and (3) ocular hypopigmentation, either iris translucency or fundus hypopigmentation grade 2 or more. Minor criteria would be (1) nystagmus, (2) hypopigmentation of skin and hair, (3) grade 1 fundus hypopigmentation, and (4) foveal hypoplasia grade 1. They propose that 3 major criteria or 2 major and 2 minor criteria are necessary for the diagnosis. In the presence of a molecular diagnosis, 1 major criterion or 2 minor criteria will be sufficient.

Adrenal Suppression in Infants Treated with Topical Ocular Glucocorticoids

Bangsgaard R, Main KM, Boberg-Ans G, Monten La Cour et al. *Ophthalmology* 125; 10 Oct 2018: 1638-1643.

Increasingly, clinical evidence suggests that the any treatment with glucocorticoids (GC) may suppress adrenal function and cause Cushing's syndrome irrespective of administration route. Adrenal suppression after topical ocular GCs is not well documented. The main objective of the study was to analyze the incidence of adrenal suppression and the glucocorticoid (GC) dose per kilogram body weight in infants treated with standard protocol for topical ophthalmic GCs after congenital cataract surgery. The authors analyzed retrospectively collected data from patients younger than 2 years of age who underwent operation for congenital cataract between January 2011 and May 2015 in a single center. Standard regimen after cataract surgery was subconjunctival injection at the time of surgery 0.5 to 1.0 mL methylprednisolone acetate 40 mg/ml (Depo-Medrol, Pfizer, Belgium). This was followed by topical administration of dexamethasone 1 mg/ml (Maxidex, Alcon, UK) eye drops 6-8 x/day for the first week, then 4 to 6 drops for the second week then tapering by one drop per week, hence up to 6 weeks of administration of drops. A standard ACTH provocation test was scheduled approximately 1 month post-operatively whenever possible. Among 26 consecutive infants, 15 (58%) were tested while they were still on GC treatment. Ten of these 15 infants (67%) had adrenal suppression, 2 of whom had obvious clinical signs of Cushing's syndrome and 1 of whom had signs of Addisonian crises during general anesthesia. Eleven of the 26 infants (42%) were tested at a median time of 21 days (range, 6–89) after treatment cessation, and they all had normal test results. Infants with suppressed adrenal function had received cumulative GC doses per body weight that were significantly higher the last 5 days before testing compared with children with normal test results. Infants with adrenal suppression were treated with hydrocortisone replacement therapy. Adrenal function recovered after a median of 3.1 months (range, 2.3 months to 2.3 years). In conclusion two thirds of the infants tested during treatment with a standard GC protocol after congenital cataract surgery showed adrenal suppression. There was a significant association between the cumulative daily dose of GCs and the test result. Because adrenal suppression is a serious but treatable condition, the authors recommend a systematic assessment of adrenal function in infants treated with doses of topical ocular GCs comparable to our regimen and careful evaluations of other treatment regimens.

24.UVEITIS/ SYSTEMIC

What is New in Paediatric Uveitis

Kara C. LaMattina and Anjum F. Koreishi *Curr Opin Ophthalmol* 2018, 29:412-418 (Sept 2018)

Pediatric uveitis is most commonly an anterior, nongranulomatous, chronic uveitis that is noninfectious (67.2-93.8% of cases). JIA is the most common systemic association with VKH, Behcet disease and TINU common in Asian and European studies. Pars planitis is common in the Middle East. Infection uveitis is less common (6.2-32.9 % of cases) with toxoplasmosis one of the most widely reported. In addition toxocariasis, viral and TB infections occur.

Treatment includes a number of modalities. Steroids should be used in the initial control of uveitis, flares and as a bridge to steroid sparing agents (SSAs). Topical prednisolone acetate 1% most commonly used. Chronic use of steroids can lead to cataract and glaucoma. The use of steroid implants has been shown in some case reports to provide good control although there is an increased risk of glaucoma and cataract. Antimetabolites such as methotrexate have been used for a long time with good long-term data on safety and efficacy. Mycophenolate sodium is also effective at controlling inflammation. Biologic agents such as TNF-alpha are another class of medication. Adalimumab is 80% effective in controlling refractory uveitis although adverse events such as infection were a concern. Interleukin-1 and interleukin-6 blockade is a newer area of treatment but there are only small number of studies looking at the efficacy. This paper reviews the different treatment modalities currently being used for the management of pediatric uveitis.

Ocular complications in a young pediatric population following bone marrow transplantation.

Hoehn M. E., Calderwood J., Gannon E., Cook B., et al. *J AAPOS* April 2018; 22(2): 102-106.e101.

The purpose of this retrospective study was to investigate ocular complications associated with bone marrow transplant and associated continued maintenance therapy in a preschool population. The medical records of patients <7 years of age were reviewed. Patient charts were screened for cataract formation, dry eye, and other anterior and posterior segment disease.

A total of 270 cases were reviewed, 91 met inclusion criteria. Mean age at diagnosis was 3.17 years. Average follow-up was 5.8 years (range, 1.9 months-14.1 years). Of the 91, 37 patients developed cataracts (41%) over a 14-year period. Cumulative incidence corrected for competing event (death before cataract) for the study population was found to be 58.4% after 14 years. Univariate analysis for cataract formation showed statistical significance for total body irradiation dose, age at diagnosis, race, donor type (related vs unrelated), product type, diagnosis type, survival status, calcineurin inhibitor use, and bisulfan, cytarabine, and thiotepa use. Multivariate analysis for competing event (death), showed that total body irradiation dose was not statistically significant; however, when studied in a binary logistic regression model, total body irradiation dose was statistically significant. Notably, steroid use and presence of graft-versus-host disease did not show statistical significance for cataract development. No other ocular complication was found in sufficient quantities to allow statistical analyses. The authors proposed that screening examinations by a pediatric or general ophthalmologist be completed at least annually. due to the high incidence of cataract formation in this population, especially those enduring a treatment regimen with total body irradiation. They also urged a low threshold for treatment of dry eye syndrome. This study presents a relatively large cohort of young children who underwent allogeneic BMT with a relatively long follow-up period.

Intravenous dexmedetomidine augments the oculocardiac reflex.

Arnold R. W., Biggs R. E. and Beerle B. J. *J AAPOS* June 2018; 22(3): 211-213.e211.

Dexmedetomidine is a selective alpha-2 adrenergic agonist affording sedation with minimal respiratory depression. It is helpful for dose-dependent prevention of emergence agitation and postoperative nausea. A previous retrospective report had indicated that premedication with nasal dexmedetomidine was associated with a more intense oculocardiac reflex (OCR). This case-control interventional study was conducted to test the authors' hypothesis that IV dexmedetomidine potentiates OCR. A total of 33 patients were enrolled. Oculocardiac reflex (greatest change heart rate/baseline heart rate) was prospectively monitored with 10-second, square-wave 200g tension on the inferior rectus or other muscles during strabismus surgery. Between the first and second muscle, intravenous (IV) dexmedetomidine 0.5 µg/kg was delivered. Intrasubject comparison was performed before and after exposure to dexmedetomidine. All patients had no anticholinergic agents. A total 842 historic control patients (median age, 5.5 years) with no dexmedetomidine exposure, experienced an average first OCR percentile of 75% ± 24% (SD) and the second OCR percentile of 77% ± 22%. The 33 study patients (median age, 5.6 years) experienced the first OCR 84% ± 16% and the post dexmedetomidine second OCR of 66% ± 25% for a bradycardia

augmentation of $18\% \pm 19\%$ ($P < 0.01$ [Mann-Whitney]). The authors concluded that intravenous push of dexmedetomidine augmented the bradycardia associated with extraocular muscle traction. Despite some weaknesses with the study's design and its control group, it increases the reader's awareness to the possible detrimental effect that the alpha-2 agonist sedative dexmedetomidine may have on the oculocardiac reflex.

Blau Syndrome – Associated Uveitis: Preliminary Results from an International Prospective Interventional Case Series

Sarens IL, Casteels I, Anton J, et al. *Am J Ophthalmol.* 2018; 187:158-166.

The authors of this study sought to look at baseline and follow up eye findings in a large multi centered trial of the rare disease Blau Syndrome. There were 25 centers worldwide who provided baseline data for 50 patients. These patients were also followed for 1, 2, or 3 years after enrollment and their data recorded, when available. The median age of the onset of eye disease was 60 months and most of these patients had uveitis (78%) (almost always bilateral) and 21% of patients had moderate to severe visual impairment. Over half of patients had panuveitis with multifocal choroidal infiltrates. Other findings included optic disc pallor, peripapillary nodules, and anterior chamber inflammation. Patients who had panuveitis had a longer disease duration. Most patients were on topical steroid and most received systemic steroids and immunomodulatory therapies. A large percentage of patients had complications of chronic inflammation (band keratopathy, cataract, synechiae, retinal detachment, macular edema, etc). Many patients had persistent eye inflammation despite topical and systemic treatments. The authors point out the important conclusion with this paper is that the uveitis in this rare disease is found frequently and is very severe, warranting close eye follow up.

Safety of Oral Propranolol for Infantile Hemangioma

Catherine Droitcourt, Sandrine Kerbrat, Caroline Rault, et al. *Pediatrics* June 2018; 141(6); e20173783

In 2010, oral propranolol was approved for proliferative infantile hemangiomas and a risk management plan was implemented. The risk factors included cardiovascular disorders including atrioventricular block, bradycardia, hypotension; respiratory disorders including bronchospasm and/or bronchial hyperactivity; metabolic disorders including hypoglycemia or related seizures, and hyperkalemia for ulcerative infantile hemangiomas. The objective of the study was to assess the safety of oral propranolol using observational data from the French national medico-administrative exhaustive database. The researchers performed an observational study on existing data comparing subjects <3 years of age who had and who had not used propranolol identified in the Systeme National Inter-Regime de l'Assurance Maladie (SNIRAM). 1934 children had at least one deliv-

ery of Hemangioli (oral propranolol) between July 2014 and June 2016. The mean age of medication delivery was 5.7 months. 1753 children had at least 2 deliveries of Hemangioli with 1484 being healthy children and 269 having an underlying disorder. These disorders included 133 with cardiovascular disease, 49 children with respiratory disease, 139 with metabolic disease. The study calculated the standardized morbidity ratios (SMRs) for patients on Hemangioli. The main outcome was hospitalizations for cardiovascular, respiratory, and metabolic events identified through ICD coding. In the healthy population, the authors found 2 cardiovascular events (SMR = 2.8), 51 respiratory events (SMR = 1.7), and 3 metabolic events (SMR = 5.1). In the unhealthy group, they observed 11 cardiovascular events leading to an SMR of 6.0. SMRs were not significantly raised for respiratory or metabolic events in the nonhealthy population. The increased cardiovascular SMR is likely due to the usage of Hemangioli in patients with Tetralogy of Fallot that could not be excluded with the given database because Hemangioli is given to this population as well. The strengths of the study include a very large study population, virtually no selection bias, and a quality comparative pediatric population. The weaknesses include an inability to relate side effect cause to the propranolol, understand true usage of medication consumption, and no recording of medical events outside of hospitalization. The data confirm the overall good safety profile of oral propranolol in children with hemangiomas. It is advised to avoid propranolol in children with lower respiratory infections or low food intake. The study also recommended a cardiovascular monitoring system during the titration phase to identify cardiovascular issues. The study provides valuable information regarding the safety profile of oral propranolol.

25.PRACTICE MANAGEMENT/ HEALTH CARE SYSTEM

Outbreak of Adenovirus in a Neonatal Intensive Care Unit: Critical Importance of Equipment Cleaning During Inpatient Ophthalmologic Examinations.

Sammons JS, Graf EH, Townsend S, Hoegg CL, et al. *Ophthalmology* 2019. Jan;126(1):137-143.

Adenovirus is a common cause of respiratory infections and conjunctivitis in children and adults. Although these infections are often benign and self-limited, they can have severe complications and even death in vulnerable populations. In this report, the authors describe an outbreak of adenovirus in neonatal intensive care units (NICUs) due to contaminated handheld ophthalmologic equipment used during retinopathy of prematurity (ROP) screening and describe the investigation, response, and successful containment of an adenovirus outbreak in a NICU. A total of 23 hospitalized neonates, as well as NICU staff and parents of affected infants were included in this epidemiologic investigation. In August 2016, a routine surveillance identified an adenovirus outbreak in a level IV NICU. Epidemiologic investigation followed, including chart review, staff interviews, and observations. Cases were defined as hospital-acquired adenovirus identified from any clinical specimen (NICU patient or employee) or compatible illness in a family

member. Real-time polymerase chain reaction (PCR) and partial- and whole-genome sequencing assays were used for testing of clinical and environmental specimens. A total of 23 primary neonatal cases and 9 secondary cases (6 employees and 3 parents) were identified. All neonatal case-patients had respiratory symptoms. Of these, 5 developed pneumonia and 12 required increased respiratory support. Less than half (48%) had ocular symptoms. All neonatal case-patients (100%) had undergone a recent ophthalmologic examination, and 54% of neonates undergoing examinations developed adenovirus infection. All affected employees and parents had direct contact with infected neonates. Observations revealed inconsistent disinfection of bedside ophthalmologic equipment and limited glove use. Sampling of 2 handheld lenses and 2 indirect ophthalmoscopes revealed adenovirus serotype-3 DNA on each device. Sequence analysis of 16 neonatal cases, 2 employees, and 2 lenses showed that cases and equipment shared 100% identity across the entire adenovirus genome. Infection control interventions included strict hand hygiene, including glove use; isolation precautions; enhanced cleaning of lenses and ophthalmoscopes between all examinations; and staff furlough. The authors recommended that ophthalmologists performing inpatient examinations take measures to avoid adenoviral spread from contaminated handheld equipment.