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What's New and Important in Pediatric Ophthalmology and Strabismus for 2013

Jitka L Zobal-Ratner MD
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What's New and Important in Pediatric Ophthalmology and Strabismus for 2013

American Academy of Ophthalmology

New Orleans, Louisiana
Tuesday, November 19, 2013
12:45-3:00 PM
Instruction Course 569

Presented by the
American Association for Pediatric Ophthalmology and Strabismus – Professional Education Committee

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1. AMBLYOPIA

A comparison between the amblyopic eye and normal fellow eye ocular architecture in children with hyperopic anisometropic amblyopia.

Wang BZ and Taranath D. J AAPOS 2012;16:428-430

The authors examined the anterior and posterior ocular architecture of hyperopic children with anisometropic amblyopia. Fourteen Caucasian children were included in the study. OCT and Pentacam were performed. There was no difference between the amblyopic eye and the fellow eye in regards to anterior or posterior corneal curvature, corneal thickness and volume, anterior chamber depth and volume, retinal nerve fiber layer thickness, macular thickness and macular volume. This study enrolled a small number of patients and only included patients with hyperopic, anisometropic amblyopia. Fellow eyes were used as the 'normal' comparison, but this may not be valid.

Characterization of Bangerter Filter Effect in Mild and Moderate Amblyopia Associated with Strabismus.

Carlos Laria, and David P. Pinero Binocular Vision And Strabology Quarterly, Simms-Romano's. 2012;27(3):174-86.

This was a prospective study of 30 children with unilateral mild to moderate amblyopia associated with esotropia and hyperopia. Inclusion criteria included the presence of mild or moderate unilateral amblyopia associated with esotropia and hyperopia, 11 years of age or younger, best spectacle-corrected visual acuity of 20/25 or worse in the amblyopic eye, at least a difference of one LogMAR line of visual acuity between the eyes, and unsuccessful outcome with refractive correction combined with other amblyopia treatment. All children were treated with a Bangerter filter in the non-amblyopic eye combined with refractive correction. The children were evaluated monthly for a 12 month period. The filters were used to reduce the visual acuity 2 lines below the best spectacle-corrected visual acuity (BSCVA) of the amblyopic eye. A statistically significant improvement in BSCVA was observed at 3 months in amblyopic eyes ($p<0.01$). At 6, 9, and 12 months additional small improvements were found. Filter density had to be adjusted in 40% of the eyes. The authors conclude that Bangerter filters seem to be useful for treating mild or moderate amblyopia due to strabismus. They also feel ocular dominance inversion should be maintained during treatment for obtaining an optimized outcome

Frequent Evaluation To Improve Compliance In Patients Treated With Occlusion For Amblyopia: A Randomized Controlled Trial.

Hernan Iturriaga, Mario Zanolli, Constanza Damm, Jorge Oporto, et alBinocular Vision And Strabology Quarterly, Simms-Romano's. 2012;27(3):195-204.

Prospective comparative blind trial where 30 children with amblyopia were randomly assigned to be followed up more frequently (every 4-6 weeks) or on their routine basis of every few months (x month intervals, x = age in years). The primary outcome was treatment compliance as assessed by the physician. Survey results with adherence were also evaluated to assess concordance between compliance and adherence. Study group compliance was 83% + 27 versus 76% + 26 in the control group (p=0.5). Compliance in the study group improved if the intention to treat analysis was removed. Pearson correlation between negative responses to a parental survey after treatment (of the percentage of adherence and compliance) was -0.57 (p=0.049). The authors conclude that there was no difference in patient compliance comparing routine evaluation and more frequent follow-up. In general there was a high compliance with occlusion therapy in this group.

The effect of amblyopia treatment on stereoacuity

Stewart CE, Wallace MP, Stephens DA, et al J AAPOS 2013;17:166-173

The Monitored Occlusion Treatment for Amblyopia Study (MOTAS), looked at dose-response function of occlusion with respect to visual acuity and some secondary outcomes. This paper reports the stereoacuity changes during and after amblyopia treatment. Amblyopia caused by anisometropia, strabismus or mixed was treated with a run-in phase of spectacles followed by occlusion therapy in 85 children. Two-thirds of patients had nil stereoacuity at study entry. During the refractive phase, stereoacuity improved from 2400 to 600 log arc sec overall and from 170 to 110 log arc sec in those with some stereoacuity at the outset of the study respectively. During the occlusion phase, stereoacuity improvement was exactly the same overall and similar in those with some stereoacuity at outset (170 to 75 log arc sec). At least one octave of stereoacuity is needed to exceed test-retest variability. This was achieved in 38% of refractive adaptation patients, 28% who received occlusion, and 45% who underwent refractive adaptation and/or occlusion. 44% of patients had nil stereoacuity throughout the study. Poor stereoacuity was associated with poor visual acuity in the amblyopic eye. Poor stereoacuity was also associated with severe strabismus. Anisometropes were more likely than strabismic patients to have better stereopsis at study entry. The authors did not distinguish manifest from latent strabismus. Smaller angle strabismus is generally more likely to be latent or intermittent, skewing stereoacuity to be markedly better as compared to larger angle strabismus, which is more likely to be constant.

Effect of Occlusion Amblyopia After Prescribed Full-Time Occlusion on Long-Term Visual Acuity Outcomes

Susannah Longmuir,; Wanda Pfeifer William Scott, Richard Olson J Pediatr Ophthalmol Strabismus 2013; 50:94-101 (March/April)

This study was a retrospective study evaluating the incidence of occlusion amblyopia when full time patching was prescribed and determine its effect on long term visual outcomes. Occlusion amblyopia was found to be more common when the child was younger and if occlusion amblyopia did develop, the study found that this actually was beneficial and allows for the development of better vision in the originally amblyopic eye.

Anisometropic amblyopia: factors influencing the success or failure of its treatment

Sonia S. Toor , Anna M. Horwood , Patricia M. Riddle Br Ir Orthopt J. August 2012;9:9-16.

This review article attempts to provide insight on why treatment for anisometropic amblyopia isn't more successful than currently reported. Variations in how anisometropia is defined, duration of anisometropia prior to diagnosis, depth of amblyopia, undiagnosed microtropias, undiagnosed ocular structural defects or CNS abnormalities, asymmetric deficit in accommodation, aniseikonia and poor compliance may be factors in treatment failure.

Anisometropic amblyopia: factors influencing the success or failure of its treatment

Sonia S. Toor, Anna M. Horwood , Patricia M. Riddle
Br Ir Orthopt J. August 2012;9:9-16.

This review article attempts to provide insight on why treatment for anisometropic amblyopia isn't more successful than currently reported. The paper compares and contrasts the methods and results of 25 previous studies published during the preceding 10 years. The authors conclude that there is no consensus on which factors limit treatment success in patients with anisometropic amblyopia but patient age, depth of amblyopia and compliance are the most commonly cited reasons. The authors note that less commonly cited reasons for treatment failure variations in how anisometropia is defined, duration of anisometropia prior to diagnosis, undiagnosed microtropias, undiagnosed ocular structural defects or CNS abnormalities, asymmetric deficit in accommodation and aniseikonia. This review article provides a summary and discussion the results of other published studies but the does not provide any new data or statistical data analysis.

Internet-Based Perceptual Learning In Treating Amblyopia

Wenqiu Zhang, Xubo Yang, Meng Liao, Ning Zhang
Eur J Ophthalmol July August 2013; 23(4): 539 – 545

The aim of this study was to address the efficacy of Internet-based perceptual learning in treating amblyopia. A total of 530 eyes of 341 patients with amblyopia were retrospectively reviewed. Internet-based perceptual learning proved better than conventional treatment in ametropic and strabismic amblyopia older than 7 years. The mean cure time was also shorter for this group (3.06 ± 1.42 months vs 3.52 ± 1.67 months). Internet-based perceptual learning can be considered as an alternative to conventional treatment. It is especially suitable for ametropic and strabismic patients with amblyopia who are older than 7 years and can shorten the cure time of amblyopia.

Efficacy of split hours part-time patching versus continuous hours part-time patching for treatment of anisometropic amblyopia in children: a pilot study

Virender Sachdeva, Vaibhev Mittal, Ramesh Kekunnaya, Amit Gupta, et al. Br J Ophthal 2013; 97: 874-878.

This prospective, non-randomized, study attempted to evaluate the efficacy of split time patching versus continuous patching in a specific group of 4-11 year old children with anisometropic amblyopia. Inclusion criteria included 2.5 diopters or more of anisomteropia, vision between 20/40 to 20/400, and no amblyopia treatment in the previous month. The total hours of patching was determined by severity of amblyopia and varied between children. 44 children were enrolled in the continuous wear and 24 children were enrolled in the split hour wear. After 3 and 6 months of therapy, both groups showed improvements in vision. The difference between the two groups was not statistically significant. Compliance with split patching was only slightly better. Disadvantages to the study include small sample size, non-randomized, non-masked, with parents deciding which group the child would be enrolled in.

The effect of simulated normal and amblyopic higher-order aberrations on visual performance.

Dominguez-Vicent A, Perez-Vives C, Ferrer-Blasco T, et al
J AAPOS 17:3;269-275

The mechanism of idiopathic amblyopia is unclear. Some investigators have hypothesized higher-order aberrations between left and right eyes may be the cause. Lower-order aberrations are corrected with spectacles,, contact lenses or refractive surgery. Higher-order aberrations can only be corrected by adaptive optics. This study simulated amblyopic and normal higher-order aberrations and their effect on visual function. An adaptive optics visual simulator was used to compensate volunteers' ocular aberrations and simulate the wavefront aberration

patterns found in healthy and amblyopic eyes in 7 healthy adults. No statistically significant differences in visual acuity and contrast sensitivity were found between both groups for any analyzed contrast level, spatial frequency, and pupil size values. Based on the results it does not appear that higher-order aberrations are a major factor in compromising visual performance of idiopathic amblyopic eyes. Sample size was small in this study. The aberration patterns used were based on pediatric data but this study enrolled adult patients.

Fixation instability in anisometropic children with reduced stereopsis

Birch EE, Subramanian V and Weakley DR
J AAPOS 17:3;287-290

Why is microstrabismus often a result of foveal suppression despite normal peripheral fusion? Why is the microstrabismus usually an esotropic one? The authors tested the hypothesis that bifoveal fusion disruption by anisometropia directly affects ocular motor function. A Nidek MP-1 microperimeter was used with 94 children with anisometropic hyperopia between the ages of 5 and 13. Forty-three healthy controls were also included. This was performed prospectively over a 2.5-year period. Stereoacuity was correlated with fixation instability. All children with normal stereoacuity had stable fixation. Children with nil stereoacuity displayed the most instability. Visual acuity of the poorer seeing eye was correlated to a lesser degree. The eye movement recordings were of sufficient quality in 81/94 children. The flick microesotropic eye movements were actually found to be fusion maldevelopment nystagmus movements (FMNS) (nasalward slowdrift with a temporalward refoveating fast-phase microsaccade). Binocular decorrelation was found to be critical for the development of FMNS; visual acuity impairment is not required. The presented data support the hypothesis that anisometropia disrupts ocular motor development and can cause reduced stereoacuity and abnormal binocular visual experience.

Clinical Translation of Recommendations from Randomized Clinical Trials on Patching Regimen for Amblyopia

Ya-Ping Jin, Amy H.Y. Chow, Linda Colpa, Agnes M.F. Wong *Ophthalmology*
April 2013;120:657-662

This is a retrospective cohort study involving children with amblyopia seen from 2007 through 2009 by academic and community ophthalmologists in Toronto, Ontario, Canada. Using PEDIG criteria for defining moderate amblyopia as a visual acuity between 20/40-20/80 and severe amblyopia is defined between 20/100-20/400.
The main intervention was patching of the sound eye. The main outcome measures included the number of prescribed patching hours per day and the

amblyopic eye visual acuity expressed as logarithm of the minimal angle of resolution (LogMAR).
Conclusions: The evidence-based recommendations for amblyopia management have not been widely adopted by practicing academic and community ophthalmologists who practice in large urban center in North America (namely, Toronto, Ontario, Canada).
Comments: This article stresses that although a tremendous amount of medical research has been conducted, one of the greatest current challenges of medicine is that there is a gap in applying research evidence to the process of making informed clinical decisions. This is evident across all groups of decision makers including health care providers, patients, managers and policy makers. In both primary and specialty care across all disciplines.
The article discusses various strategies including the Knowledge-to-Action Cycle framework as a proposed mechanism to translate gains from randomized clinical trials into actual clinical practice.

Quantitative Measurement of Interocular Suppression in Anisometropic Amblyopia
A Case-Control Study

Jinrong Li, Robert F. Hess, Lily Y.L. Chan, Daming Deng, *Ophthalmology* August 2013;120:1672-1680

This is a case control study of 45 participants with anisometropic amblyopia and 45 matched controls with a mean age of 8.8 years for each group. The purpose of the study is to assess 1) the relationship between interocular suppression and visual function, 2) whether suppression can be simulated in matched controls by using neutral density filters, and 3) to determine the effects of spectacles or rigid gas-permeable contact lenses on suppression in patients with anisometropic amblyopia. This study was performed in New Zealand.
Conclusions: Interocular suppression plays a key role in the visual deficits associated with anisometropic amblyopia and can be simulated in controls by inducing a luminance difference between the eyes. Accurate quantification of suppression using the dichoptic motion coherence threshold technique may provide useful information for the management and treatment of anisometropic amblyopia.

Interactive binocular treatment (I-BiT) for amblyopia: results of a pilot study of 3D shutter glasses system

N Herbison; S Cobb; R Gregson; I Ash; for the I-BiT study group

Eye; September 2013; 27(9):1077–1083

A computer-based interactive binocular treatment system (I-BiT) for amblyopia has been developed, which utilizes commercially available 3D ‘shutter glasses’. The purpose of this pilot study was to report on visual acuity in amblyopic children undergoing iBit. Ten children with anisometropic, strabismic, or

strabismic anisometropic amblyopia ages 4-8 years underwent thirty minutes of treatment once weekly for 6 weeks. Treatment sessions consisted of playing a computer game and watching a DVD through the I-BiT system. The system allows both eyes to see certain parts of the image (for example, the player in the computer game) but only presents other parts of the image to the amblyopic eye (for example, the obstacles in the computer game). Six out of nine patients (67%) who completed the treatment showed a clinically significant improvement of greater than 0.1LogMAR (more than one line) of visual acuity. All had previously worn glasses based on cycloplegic refractions for a minimum of eighteen weeks. The study was small and uncontrolled. It is unknown whether visual acuity gains were influenced by bias or learning. The possibility of using video games in the treatment of amblyopia, which is more appealing to children and families than patching or penalization, is intriguing.

The Regional Extent of Suppression: Strabismics Versus Nonstrabismics

Babu JR, Clavagnier SR, Bobier W, Thompson B, Hess RF. The Regional Extent of Suppression Invest Ophthalmol Vis Sci. 2013;54:6585–6593.

Suppression may be the cause of amblyopia rather than a secondary consequence of mismatched retinal images. Furthermore, the measurement of suppression may have prognostic value for patching therapy. The authors describe a new method for delineating the regional distribution of suppression. This method provides a more global, quantitative rather than binary measure of suppression. The new method is novel in that it does not rely on threshold measures, but on suprathreshold matching. The technique is well suited for use in the clinic as it is relatively fast to administer and provides quantitative information on the distribution of suppression within the central visual field. They used this new method to address two questions. First, is suppression limited to the fovea of strabismic amblyopes or does it involve all of the central 20°? Second, does the strength and regional distribution of suppression differ between strabismic and nonstrabismic forms of amblyopia?
The method involves a dichoptic perceptual matching procedure at multiple visual field locations. They compared a group of normal controls (mean age: 28 + 5 years); a group with strabismic amblyopia (four with microesotropia, five with esotropia, and one with exotropia; mean age: 35 + 10 years); and a group with nonstrabismic anisometropic amblyopia (mean age: 33 + 12 years).
The extent and magnitude of suppression was similar for observers with strabismic and nonstrabismic amblyopia. Suppression was strongest within the central field and extended throughout the 20° field measured, corresponding to the fovea of the fixing eye.

Does amblyopia have a functional impact? Findings from the Dunedin Multidisciplinary Health and Development Study.

Wilson GA, Welch D. Clin Experiment Ophthalmol. 2013 Mar;41(2):127-34

One thousand thirty-seven children born in Dunedin, New Zealand, between April 1972 and March 1973 were assessed from ages 3 to 32 years. Comparison of study members with no amblyopia, recovered amblyopia, possible amblyopia or amblyopia was done. The authors evaluated childhood motor development, teenage self-esteem and adult socioeconomic status (assessed by occupation, education, reading ability and income).

RESULTS: There was no evidence of poorer motor development, lower self-esteem or reduced adult socioeconomic status in study members with amblyopia or recovered amblyopia when compared with those with no amblyopia.

CONCLUSIONS: Amblyopia or having recovered amblyopia does not functionally impact on childhood motor development, teenage self-esteem or adult socioeconomic status within this cohort. The wide range of visual deficits and adaptations that are known to occur in amblyopic vision do not translate into important 'real life' outcomes for the study members with amblyopia or recovered amblyopia. The age-related cumulative lifetime risk of bilateral visual impairment in amblyopia will be assessed in future studies.

2. VISION SCREENING

Predictive value from pediatrician plusoptix screening: impact of refraction and binocular alignment.

Robert W. Arnold, Daniel Tulip, Erin Mcarthur, Janet Shen, et al. Binocular Vision And Strabology Quarterly, Simms-Romano's. 2012;27(4):227-32

The positive predictive value (PPV) of conventional preschool acuity screening is about 50% whereas previous Polaroid photoscreening with experienced interpretation can achieve PPV greater than 85%. The Plusoptix photoscreener has computer interpretation and a CPT code for pediatricians. This study looked at the results of photoscreening by two Plusoptix machines set up in pediatric offices. 675 children were screened and 84 were referred for follow-up examination. 39 (47%) had AAPOS gold-standard exams. The PPV from strabismus referrals was 17% and 96% for refractive referrals, resulting in an overall PPV of 69%. The authors report that by the end of the study, over half of the insurance companies were reimbursing photoscreening with the 99174 CPT code. The authors report that the manufacturer's settings for the ocular alignment criteria were too sensitive and resulted in the low PPV for strabismus referrals.

They report that this has since been relaxed. Given the high PPV for refractive referrals and the increasing reimbursement for photoscreening they strongly encourage the use of photoscreening in pediatric practices.

Vision Screening Across the World

Noelle S. Matta ,David I. Silbert
Am Orthopt J. August 2012;62:87-89.

The authors report the results of a survey of 18 members of the International Orthoptic Association from 18 different countries regarding international vision screening practices. Almost 90% of countries provide vision screening, approximately half are government funded but only about one quarter are mandatory. All include the use of optotypes whereas fewer than half assess ocular alignment or stereopsis. Screening is performed by a variety of individuals including orthoptists, ophthalmologists, other physicians, optometrists, nurses, parents and opticians.

Visual Acuity Assessment of Children with Special Needs

Sarah E. Morale , Dianna K. Hughbanks-Wheaton , Christina Cheng; Vidhya Subramanian Am Orthopt J. August 2012;62:90-98.

The authors prospectively surveyed parents and referring pediatric ophthalmologists of children with special needs referred to a single center specifically for visual acuity testing with Teller Acuity Cards. The authors conclude from their parent surveys that Teller Acuity Card testing tend to reduce parental stress about their child's vision. The physician survey revealed that referring pediatric ophthalmologists most frequently valued Teller Acuity Card test results for assistance in diagnosis and treatment as well as document eligibility for school or social services.

Comparison of the iScreen and the MTI photoscreeners for the detection of amblyopia risk factors in children

Silbert DI, Arnold RW and Matta NS J AAPOS 2013;17:34-37

The iScreen photoscreener (IS) is handheld, acquires images faster than the MTI photoscreener (MTI), and the images can be viewed immediately. Both screening devices were used to evaluate 169 children between the ages of 6 months and 17 years (average age- 7 years). Sixty-two of the children (37%) had normal pediatric ophthalmology examinations and the rest were found to have amblyopic risk factors. The accuracy, sensitivity, and specificity of the MTI were all 81%. The IS had rates of 81%, 88%, and 71% respectively. These results may not be applicable to all settings. First, two-thirds of the patients had amblyopic risk factors, which would not be consistent with a routine screening population.

Secondly, the testing was performed in a pediatric eye care practice by experienced technicians. This is very different than a community-based setting were a lay person may be administering the screening.

The challenges to ophthalmic follow-up care in at-risk pediatric populations

Williams S, Wajda BN, Alvi R, et al J AAPOS 2013;17:140-143

Give Kids Sight Day and the Eagles Eye Mobile are two non-governmental programs providing comprehensive vision care accessible to low-income, underinsured and uninsured children. The authors attempted to identify the barriers impeding follow-up in these children. With both programs only 30% of children's families were successfully contacted. Telephone numbers were out-of-service, invalid, or unavailable in 42%. Extenuating circumstances or voice-mail only, limited communication in 22%. Some families had more than one barrier. All families who could not be reached by telephone were sent a letter and a questionnaire. The response rate from this was 0%. Overall 71% of children did not receive a scheduled follow-up appointment. This study focuses on an inner-city population with low socioeconomic status. Cell phones have increased call screening. Clearly better communication protocols need to be established. Increasing contact information and education at the time of the initial contact may help.

Plusoptix photoscreening may replace cycloplegic examination in select pediatric ophthalmology patients

Silbert DI, Matta NS and Andersen K J AAPOS 2013;17:163-165

The standard of care for a pediatric ophthalmic examination has included a cycloplegic refraction and a dilated funduscopic examination. The authors sought to determine whether or not a normal acuity, normal alignment/motility check combined with a normal autorefractor/photoscreener would suffice. A retrospective evaluation of children who had an undilated plusoptiX S04 or plusoptiX A09 photoscreening was performed, including half for review. Children who were referred for a medical or ophthalmic condition requiring dilation were excluded. Children with acuity <20/30 or alignment/motility problems were excluded as well. Two hundred and twenty-two children out of 451 (49%) who were photoscreened/autorefracted had normal results. Of the 222, 32 were excluded for not meeting inclusion criteria. Of the remaining 190 patients, 186 had no abnormal findings on dilated examination and cycloplegic refraction. Optic nerve cupping with normal intraocular pressures, moderate hyperopia, a non-visually significant cataract, and refractive amblyopia from moderate astigmatism were the causes of the 4 abnormal examinations. One must remember when reviewing this study, that children with strabismus, amblyopia, or medical pathology would most likely be excluded based on inclusion criteria so the

applicability of the information is confined to children with normal acuity and alignment with no serious medical or ophthalmic conditions. This group would be expected to have very rare instances of pathology, but if autorefractor/photoscreener alone eliminates dilated exam and cycloplegic retinoscopy, those children would be missed. Most children referred to a pediatric ophthalmologist do not have both normal motility and acuities so evaluation of this group of patients is not modified by the results of this paper. Rather the children who often do not see an eye care provider (routine, normal screenings), probably do not need a cycloplegic evaluation or dilated fundus exam.

Calibration and Validation of Nine Objective Vision Screeners With Contact Lens-Induced Anisometropia

Robert W. Arnold, Bethanne Davis; Laura E. Arnold; Kayla S. Rowe; Jodi M. Davis J Pediatr Ophthalmol Strabismus 2013; 50:184-190 (March/April)

Nine objective vision screeners were used on 2 patients who were emetropic. The patients were placed in various power contact lenses and the screeners were put to the test to validate them in terms of being able to diagnose amblyopia. Screeners were found to be valid including the iphone (although the article did say the iphone was found to be too sensitive)

Reliability and Validity of an Automated Computerized Visual Acuity and Stereoacuity Test in Children Using an Interactive Video Game

Dae Joong, Hee Kyung Yang, Jeong-Min Hwang. Am J Ophthal June 2013; 156 (1): 195-201.

This retrospective study evaluated 102 children between the ages of 3-7 years reliability on automated interactive video game as compared to Snellen acuity results. Results showed the average Snellen acuity was 0.22+/- 0.19 log MAR, while the computerized results were 0.18+/- 0.14 logMAR on the initial test, and 0.22+/- 0.17 logMAR on the second test. The results seem to reliability and validity compared to the Snellen visual acuity chart and distance Randot Stereotest.

Test-retest reproducibility of accommodation measurements gathered in an unselected sample of UK primary school children

Paul Adler, Andrew Scally, Brendan Barrett. Br J Ophthal 2013; 97:592-597.

This study determined the reproducibility of accommodation measurements gathered in primary school children as well as determining intra observer variability. The near point of accommodation was measured on 137 children with average age of 8.1 years. Testing was performed on 3 different occasions. Binocular amplitudes and monocular amplitudes were determined. Results showed that children aged 4-12 years exhibit monocular accommodation of 19

diopters. Larger accommodative amplitude was measured with binocular testing. Seventy-four percent of children who showed an amplitude less than 12 diopters will exhibit a normal measurement on retesting. In addition, great intra-observer variability was noted.

Findings of perinatal ocular examination performed on 3573, healthy full-term newborns

Li-Hong Li, Na Li, Jun-Yang Zhao, Guo-ming Zhang, et al. Br J Ophthal 2013; 97: 588-591.

This study prospectively looked at 3573 babies born and examined their eyes with in the first week of life to determine what ocular pathology may be detected early after birth. Only healthy babies born after 37 weeks gestation with apgars of 7 were evaluated. During these examinations, 871 (24.4%) of abnormal cases were detected. The majority were retinal hemorrhages, 769 cases (21.52%). Of these cases, 215 were significant retinal hemorrhages, representing 6.02%. One case of retinoblastoma was detected. The long term impact of these abnormalities at birth and significant retinal hemorrhages is unknown; the authors raised concern that these events may lead to the development of amblyopia and/or anisometropia.

Optic disc measurements in full term infants.

Yogavijayan Kandasamy, Roger Smith, Ian Wright, Leo Hartley. Br J Ophthal 2012; 96: 662-664.

This study attempted to measure optic disc size and see if there was variance with sex or birth weight. This was a cross-sectional study, Australia. All assessments were performed with in the first 7 days of life. Babies were classified by birth weight, <2500 gm, 2500-4499 gm, and >4500 gm. The posterior pole was photographed with a RetCam. 35 full-term babies were enrolled. Mean optic disc size was 1.26+/- .23 mm2; mean vertical diameter was 1.37 +/- .15 mm2; mean horizontal size was 1.14 +/- .12 mm2. The vertical diameter was significantly longer than the horizontal diameter. There appeared to be no significant difference between males and females or between birth weights. This study provides some normative data or optic nerve size.

Normative data for the crowded logMAR Kay's pictures vision test in children

Teresa Saul BSc and Kate Taylor BSc Br Ir Orthopt J 2012; 9: 36–43

This prospective study involving 110 children with normal vision aged 3-4 years provides normative date for the logMAR crowded Kay's pictures used for vision screening of pre-literate children in the United Kingdom. Mean and median normal visual acuity in this population using logMAR crowded Kay's pictures was

0.100. Test/retest reliability indicated that ≥ 5 pictures was significant. There was no significant difference between the responses of boys and girls. The authors recommend referral of screened children with acuity score of ≥ 0.150 or interocular difference of ≥ 6 pictures to be referred for further evaluation.

The prevalence of visual problems in stroke patient and the effectiveness of the current screening tool used

Zishan Naeem BSc Br Ir Orthopt J 2012; 9:55-58

In this prospective study, the screening tool used by occupational therapists (OT) at the Royal London Hospital to detect visual deficiencies in patients following acute stroke was compared with orthoptic evaluation. The OT screening tool is comprised of questioning the patient about symptoms of diplopia and visual field loss as well as an assessment of convergence, smooth pursuits, saccades, visual field to confrontation and visual neglect. The OT tool does not include measurement of visual acuity. The orthoptic assessment used as a control in this study included more comprehensive patient history and formal assessment of visual acuity, ocular alignment, binocular function, convergence, smooth pursuits, saccades, visual field to confrontation, Albert's test for neglect and the line bisection test for neglect. The methods of the Albert's test and line bisection test were not described or cited in the paper. A total of 28 patients with mean age of 67 years were assessed within 72 hours of admission for stroke using both methods. In this small study, the OT screening tool yielded false positive rate of 25% and false negative rate of 7%. The authors conclude that the OT tool is effective but deserves some improvement to reduce the number of over-referrals.

Practical Community Photoscreening in Very Young Children

Susannah Q. Longmuir, Erin A. Boese, Wanda Pfeifer, Bridget Zimmerman
Pediatrics February 2013; 131:3 764-769

In response to the US Preventive Services Task Force report citing insufficient data to recommend vision screening in children <3 years of age, the authors retrospectively analyzed their Iowa photoscreening program data which included 210,695 photoscreens on children over age 6 months. The under age 3 year-old group was compared to the age 3 – to 6-year-old group. There was no statistically significant difference was found in screening children from 1 to 3 years old compared with screening children >3 years old. These results confirm that early screening, before amblyopia is more pronounced, can reliably detect amblyogenic risk factors in children younger than 3 years of age.

Unmet Visual Needs Of Children With Down Syndrome In An African Population: Implications For Visual And Cognitive Development

Ada E. Aghaji, Linda Lawrence, Ifeoma Ezegwui, Ernest Onwasigwe
Eur J Ophthalmol May – June 2013; 23(3): 394 – 398

A group of 30 Nigerian students with Down syndrome aged 5-15 were evaluated in a school for the mentally challenged. Uncorrected refractive errors were identified in 76% of these children. Also, one-third had ptosis, one-third strabismus, 13% nystagmus and 3% cataracts. The authors stressed the unmet visual needs of such patients and recommended early screening and treatment.

Test-retest variability of Randot stereoacuity measures gathered in an unselected sample of UK primary school children. Paul Adler, Anfrew Scally, Brendan Barrett. Br J Opthal 2012; 96: 656-661.

Randot stereoacuity is performed as part of vision screening in schools. This study evaluated the likelihood of a normal result on retesting after initial abnormal result. Stereoacuity was tested 3 times by 5 examiners who were not clinically qualified. Children were between the ages of 4-10 years. Results show that older children performed better on the Randot testing than younger children. In addition, stereoacuity improved at the second test, but not much improvement by the third test. In addition, it appears that stereoacuity improves upto age 8 years.

Fetal ocular measurements by three-dimensional ultrasound

Bojikian KD, de Moura CR, Tavares IM, et al
J AAPOS 17:3;276-281

The goal of this study was to obtain normative data for fetal eye volume and evaluate reproducibility. This was a prospective, longitudinal observational study, involving 71 eyes of 37 fetuses between 17 and 40 weeks gestational age. The normative data can be helpful when assessing fetal eyes with risk of genetic diseases that affect ocular growth. Three-dimensional ultrasound was performed. A strong correlation between orbital measurements and gestational age was found. Both manual and sphere mode methods were performed. The two measurements were highly correlated. However, the sphere measurement was consistently greater than the manual measurement. The authors comment that as gestational age advanced, it became difficult to measure both eyeballs, because the fetal head is frequently engaged in the pelvis. Also this data only comments on normal gestational growth; and does not help if abnormal growth occurs after gestation. Finally, any abnormalities found in-utero are not treatable at this time.

The accuracy of the plusoptiX Ao8 photoscreener in detecting risk factors for amblyopia in central Iowa

Bloomberg JD and Suh DW J AAPOS 17:3;301-304

This was a retrospective review of consecutive patients aged 5 month-5 years seen in one practice over a two-month period. All patients were screened with the plusoptiX Ao8 photoscreener (POAP) and received a complete pediatric ophthalmology examination. Of the 290 children examined, 190 (66%) were found to hae amblyopia or amblyogenic risk factors during the examination, based on AAPOS referral criteria guidelines. POAP offered a testability rate of 98%, sensitivity of 87%, specificity of 88%, positive predictive value of 93%, and negative predictive value of 78%. The sensitivity of detecting smaller angle strabismus (<= 20 prism diopters) was 52%. POAP showed high sensitivity for detecting amblyopia and amblyogenic risk factors. Also, it does not require expert interpretation of the results, which allows more widespread use. The low rate of detecting smaller angle strabismus caused the authors to recommend adding a cover-test or stereotesting. In this study, photoscreening and the examination could have been performed as far apart as 6 months, which could induce different results and alter the data. Also the POAP detects risk factors for amblyopia but does not detect the condition directly. The patients screened are not representative of the pediatric population at-large, as the prevalence of amblyopia or risk factors was much higher than the general population. This may inflate the positive predictive value reported.

Prevalence and Causes of Visual Impairment in Asian and Non-Hispanic White Preschool Children
Multi-Ethnic Pediatric Eye Disease Study

Kristina Tarczy-Hornoch, Susan A. Cotter, Mark Borchert, Rohit Varma, *Ophthalmology* June 2013;1220-1226

This is a population-based cross-sectional study designed to determine the prevalence and cause of decreased visual acuity in a multi-ethnic sample of children 30 to 72 months of age currently living in Los Angeles. All eligible children underwent comprehensive ophthalmic evaluation including monocular visual acuity testing, cover testing, cycloplegic autorefraction, fundus evaluation, and visual acuity retesting with refractive correction.
Conclusions: Seventy percent of all decreased visual acuity in both Asian and non-Hispanic White populations occurred as a result of uncorrected refractive error or amblyopia resulting from uncorrected refractive error.

Higher Order Aberrations in Children with Down Syndrome.

McCullough SJ, Little J-A, Saunders KJ. Invest Ophthalmol Vis Sci. 2013; 54:1527-1535.

Even with optimal refractive management, good vision care, and in the absence of pathology children with down syndrome (DS) show reduced visual function compared with their age-matched, typically developing peers not attributable to subject selection, attention, or motivational factors. Studies report that an optical deficit may explain much of the reduced visual performance, as there is a 4-fold improvement in visual performance when a visual stimulus that bypassed the eye optics (interferometric sinusoidal gratings) was used to assess resolution visual acuity. Previous studies have reported differences in structural optical eye components in DS of thinner, steeper corneae, thinner crystalline lenses, and reduced accommodative function, and variations in tear film composition. There is also increased prevalence of high astigmatism, keratoconus, and cataracts. The aim of the study was to quantify the optical quality of the eyes in DS children beyond refractive errors by measuring their higher order aberrations (HOA) (following cycloplegia using Shack-Hartmann aberrometry) and to compare these with a typically developing reference group.
HOAs were analyzed over a 3mm and 5 mm pupil using Zernike polynomials from third-sixth order. Optical quality was explored using Visual Strehl ratios (VSX) and equivalent defocus values. HOAs were measured successfully from 68% of the DS group (n= 44) and 95% of controls (n=209) (ages 6-16 years). Root mean square of total combined HOAs, third, and sixth orders and coma were significantly greater in the DS group ($p<0.005$). Significant differences were found between groups for Zernike coefficients

i^{-3}_3, i^3_3, i^4_0 (P, 0.013). VSX and equivalent defocus values indicated significantly poorer optical quality in DS eyes ($P< 0.02$). DS children have greater HOAs and reduced central optical quality compared with typically developing children. The subtle reduction in optical quality may compound the visuocortical deficits previously reported in DS.

Low-Contrast Acuity Measurement: Does It Add Value in the Visual Assessment of Down Syndrome and Cerebral Palsy Populations?

Little J-A, McCullough S, McClelland J, Jackson AJ, Saunders KJ Invest Ophthalmol Vis Sci. 2013;54:251–257

Children with Down syndrome (DS) and cerebral palsy (CP) often have reduced visual acuity (VA). This study assessed VA and low-contrast acuity (LCA) with Lea symbols in DS and CP populations to explore whether LCA measures provide useful additional information about visual performance. VA and LCA were also measured in a large group of typically developing young people.
The authors determined high-contrast VA and LCA performance measured monocularly using crowded Lea symbols with 45 young people with CP (mean age 11.8 ± 4 years), 44 with DS (mean age 10.5 ± 3 years), and 211 controls (mean age 11.4 ± 3 years). Refractive status was confirmed with cycloplegic retinoscopy. The DS and CP groups had significantly lower acuities than controls at all contrasts ($P < 0.001$). Mean (±SD) high-contrast VA was as follows: DS =

+0.39 ± 0.2 logMAR; CP = +0.18 ± 0.2 logMAR; controls = −0.04 ± 0.1 logMAR. Mean 2.5% LCA was as follows: DS = +0.73 ± 0.2 logMAR; CP = +0.50 ± 0.2 logMAR; controls = +0.37 ± 0.1 logMAR. For controls, the mean difference between VA and 2.5% LCA was 0.40 logMAR (95% limits of agreement, ±0.22 logMAR). While there was a positive relation between VA and 2.5% LCA scores (linear regressions, $P < 0.0001$), considerable variation existed, with VA explaining only 36% of the variance in LCA performance for control data. VA and LCA performance was significantly poorer in DS and CP groups than in controls, and high-contrast VA did not reliably predict low-contrast performance. Therefore both high- and low-contrast acuity assessment are valuable to fully describe an individual's visual function, and this may be particularly relevant in DS and CP in cases in which patients are unable to articulate visual difficulties. Age-specific reference data from a large sample of typically developing young people across a broad age range are presented for clinicians using high- and low-contrast Lea symbols.

CORTICAL VISUAL IMPAIRMENT IN CHILDREN: IDENTIFICATION, EVALUATION, AND DIAGNOSIS

Sharon S. Lehman Current Opinion in Ophthalmology Sept. 2012 23(5) p. 384-387

Cortical visual impairment (CVI) is a major cause of visual loss in children worldwide. The description of this condition is constantly evolving with respect to definition, identifying those at risk, and technology for diagnosis. These advances can be used for early diagnosis, design of accommodations and services, as well as future therapies and prevention strategies. Clinical questionnaires are being developed to aid in the early identification of those at risk for CVI. The definition is constantly being modified and now includes deficits in vision-guided motor planning and higher level executive functions. These patients may have normal visual acuity. Neuroimaging techniques, such as MRI, functional MRI, and diffusion tensor imaging; electrophysiologic testing, such as sweep visual-evoked potentials; and perceptual testing, allow for further refinements in correlating structural defects and deficits in function. Recent developments will allow identification of those children at risk for CVI and earlier interventions for specific deficits.

3. REFRACTIVE ERROR

A New Method for Quantifying Ocular Dominance Using the Balancing Technique

Tomoya Handa; Kimiya Shimizu , Hiroshi Uozato ; Nobuyuki Shoji Am Orthopt J. August 2012;62:77-86.

The authors describe a simplified method for quantifying sensory ocular dominance by modifying a traditional “balancing” technique based on binocular rivalry. The modified technique was compared with the traditional technique using the BET screener and found to yield similar measurement results in much less time. The authors propose this sensory test as a method of pre-operative evaluation of patients interested in surgical monovision.

A Comparison of Different Autorefractors With Retinoscopy in Children Yesim Oral, Nesrin Gunaydin, Ozlen Ozgur, Aysu Karatay Arsan, Sibel Oskan, J Pediatr Ophthalmol Strabismus 2012;49:353-358 (Nov/Dec)
The purpose of this article was to compare different autorefractors to the gold standard of checking for refractive errors in children and detecting amblyopia (cycloplegic refraction). Multiple autorefractors were used with and without cycloplegia and tested for accuracy when compared to cycloplegic refraction by one senior resident. The table mounted autorefractor was found to have values significantly more myopic than cycloplegic refraction. When cycloplegia was achieved and the table mounted autorefractor was again tested, the results were found to be more hyperopic than the cycloplegic refraction results. The handheld autorefractors and cycloplegic refraction values were closer. Videoretinoscopy was also tested and results were found to be slightly lower than cycloplegic refraction. The autorefractors diagnosed hyperopia and astigmatism much more accurately than myopia.

Amblyopia and Refractive Errors among School-Aged Children with Low Socioeconomic Status in Southeastern Turkey

Ihsan Caca, Abdullah Kursat Cingu, Alparslan Sahin, Seyhmus Ari, et al J Pediatr Ophthalmol Strabismus 2013;50:37-43 (Jan/Feb)

A cohort of over 21,000 children was examined for refractive errors and amblyopia risk factors. Risk factors for myopia included older age, female gender, higher parental education. Only 22.7% needed spectacles with just over 10% of patients being unaware of their spectacle needs. A little over 2% of the children were diagnosed with amblyopia – anisometropia (1.2%) and strabismus (0.9%) being the most common causes of amblyopia.

Why do words jump? An exploration of visually symptomatic readers

Nadia Northway Br Ir Orthopt J. August 2012;9:3-8.

This review article highlights symptoms associated with reading such as blur, text movement, ocular fatigue and asthenopia which may be caused by deficiency in convergence, fusional vergence or accommodation. Treatments discussed include optical correction for near fixation, monocular occlusion, colored lenses

and colored overlays. Two figures included in the paper provide helpful visual examples of “pattern glare” and “visual stress” which are terms not commonly found in the ophthalmic literature. The author advises that most reading issues are phonological but that ocular etiologies should be diagnosed using objective measures. Although the precise methods involving use of colored lenses and overlays was not described in the paper, the author notes that these techniques should be considered controversial. This helpful review article provided detailed discussion of past published studies relating to visual issues that may affect reading; however the methods used for the literature search were not described. In this review article, new data and statistical analyses were not presented.

Effect of Day Length on Eye Growth, Myopia Progression, and Change of Corneal Power in Myopic Children

Dongmei Cui, Klaus Trier, Soren Munk Ribel-Madsen, *Ophthalmology* May 2013;120:1074-1079

This is a cross-sectional study of 235 children ages 8 to 14 years of age found to have myopia during screening for a government sponsored clinical trial in Denmark. The purpose of this study was to investigate whether axial eye growth, nearsighted progression, or corneal power change in Danish myopic children varies with the length of day (between 7 and 17.5 hours of daylight during the calendar year).
Two hundred and thirty-five children ages 8 to 14 years of age were screened. Cycloplegic refraction was measured using an autorefractor, axial eye length, and corneal power using an automatic combined noncontact partial coherence interferometer and keratometer. An astronomical table was used to calculate the number of daylight hours during the measurement period.
Conclusions: Eye elongation and myopic progression seem to decrease in periods with longer days and to increase in periods with shorter days. Children should be encouraged to spend more time outside during daytime to prevent nearsightedness.
Comments: The first article from European investigators supports numerous articles from Asian investigators who reported a decrease in myopic progression when children spend more time outdoors. This is the first study that I have reviewed that correlates daylight hours with myopic progression.

Outdoor Activity during Class Recess Reduces Myopia Onset and Progression in School Children

Pei-Chang Wu, Chia-Ling Tsai, Hsiang-Lin Wu, Yi-Hsin Yang, *Ophthalmology* May 2013;120:1080-1085

This is a prospective, comparative, consecutive, interventional study of 571 students recruited from 2 nearby schools located in the suburban area of Southern Taiwan. School students were 7 to 11 years of age. The aim of the study was to investigate the effect of outdoor activity during class recess on the

nearsighted changes among elementary school students in the suburban area of Taiwan.
Outcome measures: Data were obtained by means of parental questionnaires, ocular evaluations including axial length, and cycloplegic autorefraction at the beginning and at the end of 1 year.
Conclusions: Outdoor activities during class recess and school have a significant effect on nearsighted onset and nearsighted/myopic shift. Such activities have a prominent effect on the control of nearsighted shift especially in nonmyopic children.

Prevalence and 5- to 6-Year Incidence and Progression of Myopia and Hyperopia in Australian Schoolchildren

Amanda N. French, Ian G. Morgan, Paul Mitchell, Kathryn A. Rose, *Ophthalmology* July 2013;120:1482-1491

This study was performed in South Wales, Australia. The purpose of the study was to determine the prevalence, incidence, and change in refractive errors for Australian schoolchildren and examine the impact of ethnicity and sex. This is a population-based cohort study.
Conclusions: In Sydney, myopia prevalence (14.4%) and incidence (2.2%) and was thought to be lower than other locations. However, in European Caucasian children at age 12 there was a significantly higher prevalence of nearsightedness in a younger sample suggest a rise in prevalence consistent with international trends.
Progression of myopia was similar for children of East Asian and European Caucasian ethnicity, but lower than reported in children of East Asian ethnicity. This suggested that environmental differences may have some impact on progression. Another paper correlated myopia progression with environment, daylight hours, and activity out-of-doors.

Parental smoking and childhood refractive error: the STARS study

J V Iyer; W CJ Low; M Dirani; S-M Saw Eye. October 2012;26(10):1324-8.

This population based study assessed the relationship between parental smoking and childhood refractive errors in Singapore Chinese children aged 6–72 months.

A total of 4164 children were recruited, with a positive response rate of 72.3% (n=3009). Cycloplegic refraction measurements were obtained from all children. Parents underwent an interviewer-administered questionnaire with information on demographics, lifestyle, and parental smoking history being obtained.

The overall prevalence of myopia (at least −0.5 D) was 11.0%. Overall, 37.1% of the fathers interviewed gave a history of smoking. Among the mothers interviewed, 9.2% gave a history of smoking, 6.6% had smoked during the child's life, and 2.2% had smoked during the pregnancy. Maternal history of ever smoking, smoking during the child's life, and smoking during pregnancy were associated with decreased odds ratio (OR) of childhood myopia (OR 0.50 (P=0.01), OR 0.39 (P=0.01), and OR 0.3 (P=0.14), respectively). Paternal history of smoking was associated with decreased OR of childhood myopia (OR of 0.72 (P=0.02)).
Because of the inverse association between parental smoking and childhood myopia, the authors recommend additional research to better understand the role of nicotinic acetylcholine receptor pharmacology in ocular development.
Although this study controlled for multiple confounders, including the education of the parents and the family's income level, there may be other confounders that explain the inverse relationship between parental smoking and myopia.

Prescription of atropine eye drops among children diagnosed with myopia in Taiwan from 2000 to 2007: a nationwide study

Y-T Fang; Y-J Chou; C Pu; P-J Lin, et al. *Eye.* March 2013;27(3):418-24

This population-based cross-sectional study from Taiwan was conducted to examine the atropine eye drop prescription trend for children diagnosed with myopia, and to determine the factors associated with the prescription of atropine eye drops. The study was conducted using a national representative sample from the National Health Insurance claims data. All school children between 4 and 18 years of age who had visited an ophthalmologist and were diagnosed with myopia between 2000 and 2007 were included. The main outcome measure was the proportion of subjects who were prescribed atropine eye drops in each year. Logistic regression was used to identify the factors associated with atropine eye drops being prescribed.

Atropine eye drop prescriptions for children diagnosed with myopia increased significantly from the school years 2000 (36.9%) to 2007 (49.5%). There was also a shift from prescribing high concentrations (0.5 and 1%) of atropine eye drops to lower concentration ones (0.3, 0.25, and 0.1%) within this period. Atropine eye drops were more frequently prescribed to 9–12-year-old children (OR=1.26–1.42), compared with those 7–8 years old, and to children from families with a high socioeconomic status (OR=1.19–1.25). They were prescribed less often to those living in mid to low urbanized areas (OR=0.65–0.84) compared to highly urbanized areas.

In Taiwan, which has a high rate of myopia, 49.5% of school-aged children are being prescribed atropine eye drops, primarily at low concentrations. Lower concentrations of atropine may decrease myopia progression without causing as much pupillary dilation and photophobia. However, this study was not designed to assess efficacy or side effects of atropine use in myopia.

Profile of Anisometropia and Aniso-Astigmatism in Children: Prevalence and Association with Age, Ocular Biometric Measures, and Refractive Status.

O'Donoghue L, McClelland JF, Logan NS, Rudnicka AR, Owen CG, Saunders KJ. Invest Ophthalmol Vis Sci. 2013; 54:602-608.

The authors describe the profile and associations of anisometropia and aniso-astigmatism in the population based Northern Ireland Childhood Errors of Refraction (NICER) study. They used a stratified random cluster design to recruit schoolchildren. The examinations consisted of cycloplegic autorefractions, and measures of axial length, anterior chamber depth, and corneal curvature. χ^2 tests were used to assess variations in the prevalence of anisometropia and aniso-astigmatism by age group, with logistic regression used to compare odds of anisometropia and aniso-astigmatism with refractive status (myopia, emmetropia, hyperopia). The Mann-Whitney *U* test was used to examine inter-ocular differences in ocular biometry. 661 Caucasian children aged 12 to 13 years (50.5% male) and 389 children aged 6 to 7 years (49.6% male) were used. The prevalence of anisometropia ≥1 diopters sphere (DS) did not differ statistically significantly between 6- to 7-year-old (8.5%; 95% confidence interval [CI], 3.9–13.1) and 12- to 13-year-old (9.4%; 95% CI, 5.9–12.9) children. The prevalence of aniso-astigmatism ≥1 diopters cylinder (DC) did not vary statistically significantly between 6- to 7-year-old (7.7%; 95% CI, 4.3–11.2) and 12- to 13-year-old (5.6%; 95% CI, 0.5–8.1) children. Anisometropia and aniso-astigmatism were more common in 12- to 13-year-old children with hyperopia ≥+2 DS. Anisometropic eyes had greater axial length asymmetry than non-anisometropic eyes. Aniso-astigmatic eyes were more asymmetric in axial length and corneal astigmatism than eyes without aniso-astigmatism.
In this population, there is a high prevalence of axial anisometropia and corneal/axial aniso-astigmatism, associated with hyperopia, but whether these relations are causal is unclear.

New Cases of Myopia in Children

RN. Kleinstein, L. Sinnott, L A. Jones-Jordan, J Sims, et.al. Arch Ophthalmol. 012;130(10):1274-1279.

Studies have revealed different prevalences of refractive error, especially myopia in different countries. Much work has been conducted in Asia, documenting high rates of myopia in these populations. Not much research has been done in American populations, with its varied demographics, on the onset of myopia and prevalence in Caucasian, Hispanic, Asian and African American communities. The authors report the percentage of new cases of myopia in 4927 children aged 5 to 16 years who participated in the Collaborative Longitudinal Evaluation of Ethnicity and Refractive Error Study between 1989 and 2009. Sixteen percent of children enrolled in the Collaborative Longitudinal Evaluation of Ethnicity and

Refractive Error Study developed myopia of at least 0.75 D or greater, during their school-aged years. The percentage increased yearly until age 11 years, after which it decreased. New cases of myopia varied by ethnic/racial group Each racial/ethnic group had 500 children, making it a powerful study. Asians had the largest percentage of new cases and whites the least.

4. VISION IMPAIRMENT

An update on progress and the changing epidemiology of causes of childhood blindness worldwide

Kong L, Fry M, Al-Samarraie M, et al. J AAPOS 2-12;16:501-507

The World Health Organization (WHO) definition of visual impairment has 4 levels of visual function. 'Low vision' = moderate and severe visual impairment. Low vision + blindness = 'visual impairment. This study presents up-to-date data on various causes of childhood blindness worldwide and in the United States (since 1999). A systematic search of world literature was performed. In economically developed countries, the rate of childhood blindness is 0.3 to 0.4 per 1,000 children. In the Western Pacific, the rate is 0.2 to 0.7 per 1,000; in Asia 0.9 per 1,000 and in underdeveloped countries, the rate is 1.2 per 1000. In low-income countries, corneal disease was the major anatomic cause of visual impairment, while retinal disease was the major cause in medium and high-income countries. In developing countries 51% of childhood blindness is avoidable, 19% is preventable and another 27% is treatable. ROP is the most prevalent avoidable or treatable cause n middle-income countries. Hereditary factors are important in about 1/3 of the cases in America, Europe, Asia and the Western Pacific region. InAfrica and the Eastern Mediterranean, child blindness often occurs after the prenatal period due to corneal opacification (measles, Vitamin A deficiency, and traditional eye medicines). Vitamin A cases are on the decline and trauma-induced blindness is common in regions affected by civil war. In the United States, the leading causes of visual impairment are cortical visual impairment, optic nerve hypoplasia, and ROP. The leading treatable causes of childhood blindness worldwide are cataracts, glaucoma, and ROP.

Prevalence and Causes of Monocular Childhood Blindness in a Rural Population in Southern India

Parasappa Bandrakalli, Sunil Ganekal, Vishal Jhanji, Yuan Bo Liang, Syril Dorairaj, J Pediatr Opthalmol Strabismus 2012; 49:303-307 Sept/Oct

Monocular blindness is a common cause of ocular morbidity in all age groups; this is especially important in children because of the impact it has in their overall development, mobility, education and employment opportunity and can affect their quality of life. This article focuses on the major causes of monocular

blindness as well as their prevalence in randomly selected villages. Children were initially screened with visual acuity and those with poor vision were sent to a tertiary facility to determine the cause of blindness. Refractive amblyopia was found to be the most common cause of monocular visual impairment followed by trauma, congenital disorders and vitamin A deficiency. Considering the most common cause of monocular blindness is refractive amblyopia and is preventable, improved screening methods may decrease the prevalence.

The Short- and Long-term Effects on the Visual System of Children Following Exposure to Maternal Substance Misuse in Pregnancy.

Kurt Spiteri Cornish, Monica Hrabovsky, Neil W. Scott, Elizabeth Myerscough, et al. Am J Ophthal July 2013; 156(1): pg. 190-194.

This retrospective study evaluated 301 children born to mothers misusing substances during pregnancy. The incidence of substance abuse in pregnancy is on the rise and this study attempted to evaluate the prevalence of ophthalmic morbidities over a 5 year period. Out of the 301 children, 96 (31.9%) were referred for ophthalmic evaluation. Forty-six (15.3%) were diagnosed with strabismus, and eleven (3.7%) were diagnosed with nystagmus. The control group of 1035 children demonstrated 218 (2.8%) with strabismus and 3 (0.004%) with nystagmus. This study suggests that this subgroup of children should be monitored more carefully for ophthalmic problems.

The orthoptic role and experiences in breaking bad news

Nana Theodorou Br Ir Orthopt J 2012; 9:59-62

This small survey of 9 orthoptists indicates that orthoptists may frequently help patients who have received bad news from their ophthalmologists. The orthoptists surveyed report that they receive very little training in this area. The respondents indicated that they are interested in and frequently do answer patient questions and provide emotional support. The authors advocate for orthoptists involvement in this aspect of patient care recommend additional training of orthoptists to help patients who receive bad news.

Mechanisms compensating for visual field restriction in adolescents with damage to the retro-geniculate visual system

L Jacobson; F Lennartsson; T Pansell; G Öqvist Seimyr, et al.
Eye. November 2012; 26(11):1437-45.

In this case series, the authors describe three adolescents with damage to the optic radiation who may have developed compensatory mechanisms to address the resultant visual field loss. The patients under**went** Goldmann perimetry, and several types of computerized visual field perimetry: Rarebit, Humphrey Visual

Field Analyzer and Esterman. Eye movements were tracked with video oculography during the Rarebit examination.
Two boys had spastic cerebral palsy with bilateral periventricular leukomalacia due to prematurity. The first subject had early-onset exotropia, and a relative homonymous VF defect, but a normal binocular VF. The second subject showed an inferior right quadrantanopia, confirmed by the binocular field. He had horizontal scanning eye movements during visual field testing. The third subject had an almost total left homonymous hemianopia after resection of a brain tumor in the right temporal lobe. This patient also exhibited horizontal scanning eye movements during his binocular visual field test.

The authors conclude that in young subjects, homonymous visual field defects may be compensated for by development of exotropia and by scanning eye movements

FUNCTIONAL VISION LOSS

John Pula. Current Opinion in Ophthalmology November 2012 23(6) p. 460-465

Functional disorders are a major category of vision loss not uncommonly presenting to an ophthalmologist or neuro-ophthalmologist. There are constant advances in the discrimination of causes for functional vision loss, as well as testing and treatment of these patients. New tests for diagnosing functional vision loss have been reported. These include techniques for evaluating both decreased visual acuity and visual field loss, allowing the physician to objectively state if a patient's vision is better than what he is claiming. Newer imaging modalities such as functional MRI suggest that certain areas of the brain show impaired modulation in persons with conversion disorder. There are few clinical trials for the treatment of persons with functional vision loss, and therefore no conclusive treatment recommendations. This article discusses the recent advances regarding functional vision loss relevant to the ophthalmologist.

Cerebral Damage May Be the Primary Risk Factor for Visual Impairment in Preschool Children Born Extremely Premature

Carina Slidsborg, Regitze Bangsgaard, Hans Callø Fledelius, Hanne Jensen, et.al. Arch Ophthalmol. 2012;130(11):1410-1417.

To examine which is a greater risk factor for visual impairment in children born extremely premature, retinopathy of prematurity or cerebral damage. A cohort of 178 infants born extremely premature, that is, less than 28 weeks gestation, were examined. A control group of 56 children born term were also studied. All participants were identified through the National Register of Denmark. Visual acuity, foveal abnormalities, maximum ROP stage and global developmental deficits were recorded. Global developmental delay was used as a surrogate for cerebral damage and assessed through the Ages and Stages questionnaire.

Global developmental delay and foveal sequelae occurred more frequently in extremely premature children compared with controls, and increased with increasing severity of ROP. Cerebral damage and ROP appear to be independent risk factors for visual impairment and cerebral damage may be the primary risk factor.

5. NEURO-OPHTHALMOLOGY

A 2-year prospective surveillance of pediatric traumatic optic neuropathy in the United Kingdom

Ford RL, Lee V, Xing W, and Bunce C J AAPOS 2012;16:413-417

This was an observational study of etiology, presenting signs and symptoms, initial and final visual acuity, investigations and treatments for traumatic optic neuropathy. Data was collected from the British Ophthalmic Surveillance Unit. New cases of traumatic optic neuropathy (TON) were prospectively documented over a 2-year period.
Clinical data was obtained from 121 cases, of which, 26 were under 18 years old. The reported incidence of TON was roughly 1 per million in both adult and pediatric groups. The most common causes of injury were falls, sports and traffic accidents. Sports injuries were more common in children and assault was more common in adults. Visual acuity at presentation was <=20/200 in 15 of the 24 cases where acuity was measurable. Treatment efforts were more likely to occur with worse presenting acuity and if the patient was seen by an ophthalmologist. Steroid treatment regimens varied. Treatment did not improve final acuity. Approximately 1/3 of patients showed some improvement in final acuity compared to baseline.
The authors note that true incidence of TON may be higher than reported. The authors recommend fine-section CT imaging to look for orbital fractures and bony damage to the optic canal.

Newborn thyroid-stimulating hormone in children with optic nerve hypoplasia: Associations with hypothyroidism and vision

Fink C, Vedin AM, Garcia-Filion, et al.J AAPOS 2012;16:418-423

Optic nerve hypoplasia (ONH) can be associated with pituitary hormone deficiencies. Central hypothyroidism (CH) can be associated with developmental delays. In the United States, there is mandated newborn screening for congenital **primary** hypothyroidism. However, CH cases can display low, normal, or high levels of TSH. This study assessed whether screening TSH levels are predictive of CH diagnosis in children with ONH. The authors also attempted to determine whether thyroid status correlates with visual function or visual development in children with ONH. The authors hypothesize that lower circulating levels of

thyroid hormone may lower the chance of some spontaneous visual improvement which is often seen in young children with ONH.
Data was obtained from an ONH registry. TSH levels in CH patients were significantly lower than in euthyroid subjects. Initial vision was not affected by TSH level but final acuity and vision improvement rates (37% vs 18%) were better in those with normal TSH levels. Central hypothyroidism rates were higher in bilateral ONH (54%) versus unilateral (15%). Children with or without vision had equal rates of hypothyroidism. Larger relative optic disk size was seen in euthyroid patients compared to hypothyroid.
Early diagnosis of ONH and associated CH is important because of the high associated risk of developmental delay. Children with ONH may have late onset central hypothyroidism and therefore would be missed with current newborn screening. Current screening is to detect elevated TSH levels as would be seen with congenital primary hypothyroidism. However, CH patients may have low TSH levels. The authors admit that further studies are needed to determine whether early detection and treatment of hypothyroidism in children with ONH may improve visual and neurologic outcomes. Visual acuity categories were not precise, nor validated. Also, developmental delays which could affect acuity testing performance, were more common in the hypothyroid group.

Acquired bilateral myellinated retinal nerve fibers after unilateral optic nerve sheath fenestration in a child with idiopathic intracranial hypertension

Prakalapakorn SG and Buckley EG. J AAPOS 2012;16:534-538

A 17-month old girl with idiopathic intracranial hypertension (IIH) underwent unilateral optic nerve sheath fenestration and later developed bilateral myellinated retinal nerve fibers (MRNF) adjacent to the optic nerve. This occurred a few months later. Her vision was unaffected. Including this case, there are 11 cases of reported acquired MRNF in the literature. 8/11 had previous optic nevre abnormalities, 4/11 had a history of IIH, 4/11 had bilateral optic nerve head drusen, and 3/11 had associated optic nerve glioma. Damage to the lamina cribosa is postulated as the cause which allows oligodendrocytes access into the eye.

Retinal Nerve Fiber Layer Thickness Using Spectral-Domain Optical Coherence Tomography in Patients with No Light Perception Secondary to Optic Atrophy

Syliva L Goth, Andrew Harrison, Alana L Grajewski, and Michael S Lee . *J Neuro-Ophthalmol* 2013; 33:37-39.

Spectral domain OCT is used to evaluate eyes with longstanding no light perception (NLP) visual acuity secondary to optic atrophy in order to ascertain the lower limit of measured mean retinal nerve fiber layer (RNFL). A retrospective cross-sectional chart review identified 10 patients with NLP acuity for a mean of

3.72 (+/- 1.2) years secondary to optic atrophy. Etiologies of optic atrophy included optic nerve sheath meningioma, autoimmune optic neuropathy, other meningioma, neuro-myelitis optica, trauma and glaucoma. The Spectralis Spectral Domain OCT(Heidelberg Engineering, Heidelberg Germany) was used to quantify mean RNFL thickness. The range was 28-40 um with a mean of 34.18. This is thinner than the lower limit previously reported with the Stratus Time Domain OCT (45.42 um) however the latter is an older technology capable of less resolution. The study validates the clinically important fact that “complete” optic atrophy leaves an anatomically measurable RNFL of about 34 um. This layer is thought to be comprised of retinal blood vessels, glial cells and dead or non-functioning ganglion cell axons.

Differentiation of Optic Disc Edema from Optic Nerve Head Drusen with Spectral-Domain Optical Coherence Tomography

Ozge Srarc, Yelda Y Tasci, Canan Gurdal, and Izzet Can, *J Neuro-Ophthalmol 2012; 32:207-211.*

This prospective clinical study was designed to assess how Spectral Domain OCT (RTVue, software version 2.7, Optovue, Fremont CA) distinguishes patients with optic nerve head drusen (n=25) from those with optic disc edema (n=25) and from normal controls (n=25). Those studied with optic disc edema were a mixed group including patients with papilledema, non-arteritic ischemic optic neuropathy and optic neuritis. Diagnosis of optic nerve head drusen required a minimum of 2 of the following 4 characteristics: anticipated autofluorescence on fundus photography; calcification on B scan ultasonogaphy or on CT scan, or a normal opening pressure on lumbar puncture. Average RNFL was greater in patients with disc edema than in controls or patients with optic nerve head drusen. In particular, patients with disc edema had thicker RNLF temporally and nasally than those with drusen and thicker RNFL superiorly (temporal and nasal) and inferio-temporally than in normal controls. Patients with drusen had thinner RNFL temporally and nasally than controls, but thicker RNFL superonasally and infero-temporally than controls. In addition this group studied the thickness and the area of the sub-retinal hyporefective space (SHYPS) and the optic nerve head α angle. The SHYPS represents the hyporeflective region between the sensory retina and the RPE and choriocapillaris, --the region presumed to house the edema in patients with disc edema, as well as the “lumpy bumpy” hypoechoic regions attributed to drusen . The optic nerve head α angle is the measure angle between the RNFL and the edge of the optic nerve in the region where the optic nerve has the highest configuration. The SHYPS area as well as the optic nerve head α angle were notably greater in patients with optic disc edema than in those with drusen. Practically speaking the take home message is that, in addition to looking for the “lumpy-bumpy” configuration associated with optic nerve head drusen, the RNFL nasally and temporally will likely be thinner, the mean RNFL will likely be thinner, the SHYPS smaller, and the optic nerve head α angle more acute in patient with optic nerve head drusen when compared to patients with

disc edema. The patient mix included in the disc edema group was more diverse than ideal in a study of this nature.

Novel Treatment for Radiation Optic Neuropathy with Intravenous Bevacizumab

Osman Farooq, Norah S Lincoff, Nicolas Saikali, Dheerendra Prasad, *et al J Neuro-Ophthalmol 2012;32:321-24.*

A case report of radiation induced optic neuropathy and necrosis of the right temporal lobe (secondary to external beam irradiation for a tectal pilocytic astrocytoma) demonstrates nearly full reversal with the use of IV Bevacizumab. MRI T1 contrast enhanced images and PET scanning confirmed the diagnosis of radiation induced neuropathy. The patient presented with associated 20/80 acuity right eye and 20/40 left eye and partial bitemporal hemianopsia iin addition to color desaturation, all starting 5 years after her radiation treatment. Vision loss progressed during treatment with IV dexamethasone. When the patient's acuity declined to NLP right eye and 20/100 left eye a course of IV Bevacizumad (7.5 mg/kg), 3 doses every three weeks, in addition to dexamethasone 8 mg daily and and pentoxifylline 400 mg daily (unstated duration) reversed the optic neuropathy. There was complete resolution of associated enhancement on MRI. By 6 weeks acuity had returned to 20/20 right eye, 20/25 left eye, and the visual field deficit was nearly resolved. Her post treatment examination remained stable for 3 years. The report details other successful attempts at reversing or halting radiation necrosis of the CNS with bevacizumab including one placebo double blind study documenting complete reversal of MRI finding s of CNS necrosis in all patients treated with Bevacizumb and no reversal in the control group. Although a randomized trial to determine the best dosing and duration of treatment is indicated, bevacizumab should be considered in cases of radiation induced optic neuropathy

Idiopathic Intracranial Hypertension: Relation between Obesity and Visual Outcomes

Aimee J Szewka, Beau B Bruce, Nancy J Newman, and Valerie Biousse, *J Neuro-Ophthalmol 213; 33:4-8.*

The retrospective chart review was designed to determine whether patients with IIH and severely elevated body mass index (BMI ≥ 40) (n=158) had worse visual outcomes than those with IIH and BMI 30-39.9 (n=172). Patients with BMI ≥ 40 were more likely to present initially with severe papilledema (p-0.02) and there was a trend to more sever vision loss in one or both eyes of these patients, after controlling for sex, race, hypertension and sleep apnea. The 10 unit increase in BMI increased the odds ratio of sever vision loss by 1.4 (95% confidence interval 1.03-1.91 P-0.03)

The Clinical Validity of the Spontaneous Retinal Venous Pulsation

Sui H Wong; MCP and Richard P White, MD FRCP
J Neuro-Ophthalmol 2013; 33:17-20.
The investigators determine the specificity and positive predictive value of the presence of spontaneous venous pulsations (SVP) in indicating normal intracranial pressure. Opening pressure with lumbar puncture was assessed to determine the possibility of elevated intracranial pressure in 106 patients with relevant clinical indications. Fundus examinations by two distinct observers were carried out prior to the spinal tap Thirteen of these patients had a documented opening pressure ≥30 cmwater; SVP were present in 11 of these 13. Of the remaining 93 patients with ICP≤30 cm water, 83 had SVP and 10 did not . The sensitivity and positive predictive value of the presence of SVP indicating the presence of normal ICP were 0.89 and 0.88 respectively. However, noting that 11 of 13 patients with elevated ICP had document SVP; the presence of this clinical sign clearly does not rule out the possibility of elevated ICP

Mechanical Oscillopsia after Lower Eyelid Blepharoplasty with Fat Repositioning

Sumeer Thinda, Michael S Vaphiades, and Louise Mawn, *J Neuro-Ophthalmol 2013; 33:71-73.*

A patient who underwent transcutaneous lower eyelid blepharoplasty with fat repositioning developed oscillopsia with any mechanical movement of the face. The movement was most notable with chewing, infraducting, moving the mouth or pressing in the check or lower eyelid. Exploration of the surgical site revealed cicatricial tissue between the inferior oblique muscle and the anterior superficial musculo-aponeurotic complex; severing of these adhesions relieved the oscillopsia.

Visual Field Improvement after Pituitary Surgery in Patients with McCune-Albright Syndrome

Jin Ma, Chan Zhao, Renzhi Wang, Feng Feng, *et al J Neuro-Ophthalmol 2013; 33 26-29.*

McCune Albright Syndrome is a sporadic congenital disorder associated with café-au-lait spots, polyostotic fibrous dysplasia and endocinopathies. Pituitary tumor is present in 66% of cases that have documented growth hormone excess. Optic canal narrowing associated with the fibrous dysplasia has been assumed to be the major source of vision loss, yet other investigations have found that the presence of growth hormone excess is the factor statistically associated with vision decline. In this report, three patients with McCune Albright Syndrome were found to have growth hormone secreting pituitary tumors with chiasmal compression in addition to marked optic canal narrowing with thickening and deformity of craniofacial bones. Notable improvement in acuity and reduction in

visual field loss resulted from simple transsphenoidal pituitary resection without surgery to the optic canals. In a prior study by Lee, JS, et al NEJM 2002; 347:1670-76, a large cross sectional analysis of a case controlled cohort failed to demonstrate that narrowing of the optic canals was associated with visual loss; at this time optic canal decompression is only indicated in patients with fibrous dysplasia and acute or progressive visual impairment clearly from this source. The purpose of this report is to point out the importance of remembering the potential contribution of the highly treatable optic chiasmal compression that may be found in patient s with McCune Albright Syndrome.

Prism adaptation: is this an effective means of rehabilitating neglect?

Kirsty Lavery, Fiona J. Rowe Br Ir Orthopt J 2012;9:17–22

This review article sites 9 small studies (3 studies without control groups) and suggests that prism adaptation inducing a binocular optical shift of the visual axes by 6-10 degrees using prisms is the most effective intervention to improve the clinical measures of visual neglect as seen most commonly in patients following a right parietal lobe stroke. Although the details of prism adaptation methods were not described in this paper, this method involves having the patient wear bilateral yoked prisms for a period of time while pointing at a target that appears to be directly ahead. After the prism are removed there appears to be some expansion of visual awareness relative to the target. The authors recommend further study to determine whether the improvements in clinical measures translate into significant improvements in daily function before routinely offering prism adaptation therapy to patients with visual neglect. In this review article, new data and statistical analyses were not presented.

Comparison of the uniocular field of fixation assessed objectively and subjectively using the Goldmann perimeter

Anne Bjerre and Lucy Baldwin Br Ir Orthopt J 2012; 9:49-54

This study evaluates the technique of ocular motility assessment by measuring (in degrees) the field of uniocular excursion whereby foveal fixation is maintained. This prospective study evaluated subjective and objective measures of uniocular extraocular muscle excursions in 6 axes in 24 subjects while varying fixation target size. Each subject was asked to fixate on and then follow the Goldmann perimeter stimulus as it moved peripherally in 1 axis. The subject reported when fixation was lost (subjective endpoint) and the examiner observed when fixation was lost (objective endpoint). The objective and subjective mean total extraocular muscle excursions using the IIe targets were 288.2 degrees and 290.7 degrees, respectively. The authors recommend using the subjective method with the same fixation target in serial examinations of an individual patient with oculomotor dysfunction.

Grey and white matter changes in children with monocular amblyopia: voxel-based morphometry and diffusion tensor imaging study.

Qian Li, Qinying Jiang, Mingxia Guo, Chunquan Cai, Xiaohui Yin. Br J Ophthal 2013; 97: 524-529.

VBM provides whole brain analysis of group differences in the grey and white matter volume. The DTI provides structural and functional information about white matter. The authors looked at these techniques to see if global brain change is peculiar to amblyopia. 40 children were evaluated, 20 normal and 20 with amblyopia. Compared with the normal children, grey matter volume was reduced in the left inferior occipital gyrus, bilateral parahippocampal gyrus, and left supramarginal/postcentral gyrus in the monocular amblyopes. The white matter was decreased in the left calcarine and the bilateral inferior frontal and right percuneus areas. This study supports previous suggestions that amblyopia affects higher levels of the visual pathway as well as the primary visual cortex.

Cyclic (alternate day) vertical deviation- possible forme fruste of ocular neuromyotonia

Roper-Hall G, Cruz OA, Espinoza GM, and Chung SM
J AAPOS 17;3:248-252

The authors report 3 consecutive adult patients with 48-hour cyclic hypotropia. Sustained contraction of a vertically acting extraocular muscle lasted 24 hours and then was absent for 24 hours. Two patients had thyroid eye disease and the third had a cavernous sinus schwannoma. Two patients improved with medications (carbamazepine and gabapentin), while the other improved spontaneously. Based on the behavior of the strabismus and the response to membrane-stabilizing medications, the authors posit a relationship of the cyclic vertical strabismus to ocular neuromyotonia. The authors state the importance of early recognition, especially when patients report ‘good’ and ‘bad’ days. Acquired incomitant cyclic deviations require neuroimaging to rule out a neurologic cause.

Optic Disc Crescent and Tilt – Letter to the Editor, General Correspondence

Chiara Morini, Amit Gupta, Ramesh Kekunnaya, *Ophthalmology* September 2012;119:1942-1943

THE AUTHORS OF THE LETTER to the editor commented on the article by Kim TW, Kim M, and Weinreb RN, et al. from *Ophthalmology* September 2012;119:21-26.

The letter to the editor comments on Kim, et al. article which proposes a new theory regarding the development of structural change in the optic papillae during childhood. The 2 structural changes were temporal crescent and tilting of the disc. Both of these are common findings in a pediatric ophthalmology practice. The authors of the original article propose that when myopic shift occurs (due to axial elongation) scleral growth is not followed by corresponding development of the choroid retina, which causes a thrust of the disc away from the area of the scleral growth causing both a peripapillary scleral show or crescent on one side and an elevation of the optic disc in the opposite direction (tilt).
Not all patients with progressive myopia demonstrate a crescent/tilt development. The authors of the original article explain that this is due to a predisposition to the development of crescent tilt and would be present in some subjects and not others.
The authors of the letter bring up 3 points, namely the localization of the crescent tilt is not always observed in opposite direction and may not be proportionate. An area of 360^{o} peripapillary retinal choroidal atrophy may be appreciated in some patients and may also be accompanied by the optic tilt in some patients (but not in all patients). There may be some limitations to the age group study by Kim et al., namely 7.8+2.6 years. The authors of the letter feel that this is “not the average age of maximum myopic tilt occurring to the eye in life”.
The authors of the letter suggest that “it would be interesting to look at more data in support of the proposed theory before it could definitely be accepted”. They suggest studies utilizing measurement of axial length as well as possible confirmation by optical coherence tomography (OCT) for evaluation of “tilting of the optic nerve head”.
The letter to the editor was commented by the original authors of the article, namely Robert N Weinreb and Eun Ji Lee “further study is needed to understand the mechanism of disc tilt development and peripapillary atrophy/scleral crescent in myopic eyes”. They agree that their original study “may provide a platform for subsequent studies”.

Tuberous Sclerosis Complex: Genotype/Phenotype Correlation of Retinal Findings

Mary E Aronow , Jo Anne Nakagawa , Ajay Gupta , Elias I Traboulsi
Ophthalmology September 2012;119:1917-1923

This article comes from the Cole Eye Institute, Cleveland Clinic, Cleveland, Ohio, Department of Pediatric Epilepsy, Neurological Institute, Cleveland Clinic, Cleveland, Ohio.
This is a retrospective consecutive case series involving 132 patients enrolled in the Cleveland Clinic Foundation Tuberous Sclerosis Program (CCF-TSCP) and 907 patients from the tuberous sclerosis alliance (TSC-A). The purpose of this study is to evaluate the genotype/phenotype correlation in individuals with

astrocytic hamartoma (AH) and retinal achromic patch over AH or AP in the setting of tuberous sclerosis complex (TSC).
Tuberous sclerosis complex is autosomal dominant inherited disease characterized by hamartomas affecting multiple organ systems. Genetic studies recently identified 2 distinct variants of tuberous sclerosis complex (TSC). The first variant results from the mutation of TSC1 gene on chromosome 9q34 and the second resulting from mutations on the TSC2 gene on chromosome 16p13. Ocular manifestations of tuberous sclerosis complex were first noted by VanderHoeve in 1921 and were primarily limited to the retina. The presence of astrocytic hamartoma is considered one of the major diagnostic criteria for the disease. In some patients, retinal achromic patches (APs) have also been observed. Less commonly hamartomas of the iris, ciliary body, and pigment epithelium have been described in individuals with tuberous sclerosis. Patients were studied with the following parameters: age, gender, presence of TSC1 or TSC2 mutations, detailed ophthalmologic findings, detailed systemic findings, the presence or absence of epilepsy, and the presence or absence of cognitive impairment.
Individuals with retinal findings are more likely to have concomitant subependymal giant cell astrocytomas, renal angiomyolipomas, cognitive impairment, and epilepsy. TSC2 mutations are more frequent in patients with retinal findings than those without retinal findings.
Genetic testing for TSC1 or TSC2 mutations are now available. The retinal findings are more likely to have concomitant subependymal giant cell astrocytomas, renal angiomyolipomas, and cognitive impairment as well as epilepsy. TSC2 mutations are more frequent in patients with retinal findings than those without retinal findings.

Dominant Optic Atrophy: Novel OPA1 Mutations and Revised Prevalence Estimates

Patrick Yu-Wai-Man, Patrick F. Chinnery, *Ophthalmology* August 2013;120:1712

Autosomal-dominant optic atrophy (DOA) is classified as a mitochondrial disorder and is the most common inherited optic nerve disorder seen in clinical practice. Two causative genes have been identified in patients with dominant optic atrophy; OPA1, which accounts for 50%-60% of cases, and OPA3, which is relatively rare having been identified in only isolated families in association with premature cataracts. OPA1 mutations have a high penetrance rate, but the disease phenotype is characterized by marked intra- and interfamilial variability. The purpose of this paper is to report the identification of additional 8 families of confirmed pathogenic OPA1 mutations.
Conclusions: The authors reported that their updated epidemiologic data has firmly established dominant optic atrophy as an important cause of inherited visual failure and the urgent need for effective treatments to help patients with this disabling form of mitochondrial blindness.

Incidence of pupillary involvement, course of anisocoria and ophthalmoplegia in diabetic oculomotor nerve palsy. Dhume KU, Paul KE. Indian J Ophthalmol 2013;61:13-7.

In this prospective analytical study, 35 consecutive patients with diabetes-associated oculomotor nerve palsy who were subjected to a comprehensive ocular examination. Standardized methods were used to evaluate pupil size, shape, and reflexes. Results: Pupillary involvement was found to be present in 25.7% of the total number of subjects with diabetic oculomotor nerve palsy. The measure of anisocoria was < 2 mm, and pupil was variably reactive at least to some extent in all cases with pupillary involvement. Majority of patients in both the pupil-involved and pupil-spared group showed a regressive pattern of ophthalmoplegia. Ophthalmoplegia reversed much earlier and more significantly when compared to anisocoria. Conclusions: Pupillary involvement in diabetes-associated oculomotor nerve palsy occurs in about 1/4th of all cases. Certain characteristics of the pupil help us to differentiate an ischemic insult from an aneurysmal injury to the 3rd nerve. Ophthalmoplegia resolves much earlier than anisocoria in diabetic oculomotor nerve palsies.This is useful information who deal with oculomotor nerve palsy, but imaging is necessary when pupil is invoved.

Dural Puncture Induced Intracranial Hypotension Causing Diplopia

Padmaja Sudhakar, Jonathan D. Trobe, and Jeffrey Wesolowski, J Neuro-ophthalmol 213; 33:106-112

The authors present six cases where diplopia from 6th nerve palsy resulted from intracranial hypotension; The hypotension occurred after typically after removal of an epidural catheter that had been placed, initially, for medication delivery. Dural puncture upon removal of the catheters resulted in unintended CSF leak, postural headache and in these rare cases, 6th nerve palsy. The 6th nerve palsy is thought to result from traction on the brainstem as it sinks downward because of lowered intracranial pressure. Resolution of the leak results in recovery of 6th nerve function. The paper is important as new onset 6th nerve palsy is typically seen as an ominous sign requiring a significant evaluation to detect the cause. Awareness of this rare but relatively benign etiology fact can avert unnecessary workup and concern.

Visual and Neurological Outcomes following Endovascular Stenting for Pseudotumor Cerebri Associated with Transverse Sinus Stenosis

Martin G Radvany, David Solomon, Satnam Nijjar, Prem S. Subramanian, Neil R Miller, et al. *J Neuro-Ophthalmol 2013 33:117-122.*

A subgroup of patients with presumed idiopathic intracranial hypertension (IIH) have been noted to have cerebral dural sinus stenosis, particularly involving the

transverse sinuses. It remains unclear whether these stenoses are caused by, or are responsible for, the associated elevation in intracranial pressure. The authors present a series of 12 patients with transverse sinus stenosis and IIH who had failed medical therapy and were subsequently treated with endovascular venous stenting. This procedure proved to be effective in reducing intracranial pressure and treating the associated visual symptoms as well as reducing headache and pulsatile tinnitus in nearly all cases.

Drug Related Mitochondrial Optic Neuropathies

Michelle Y Yang; Alfredo A. Sadun, *J Neuro-Ophthalmol 2013 33:172-178*

Mitochondrial optic neuropathies typically affect the papillo-macular bundle as the associated fibers are small, unmyelinated, and exercise high energy demand. A variety of medications cause “mitochondrial optic neuropathy” as they interfere with mitochondrial function. These medications include chloramphenicol, ethambutol, linezolid, erythromycin, streptomycin and anti-retroviral drugs. Awareness of the possible undesirable side effect resulting from using these medications can help reduce morbidity if the association is recognized and the patient treated by discontinuing the medication; early recognition can permit recovery and may help prevent permanent loss.

Cranial autonomic symptoms in pediatric migraine are the rule, not the exception **Gelfand AA, Reider AC, Goadsby PJ. Neurology July 30, 2013; 81(5):431-6.**

The presence of cranial autonomic symptoms often leads to a misdiagnosis of “sinus headache” in adult migraineurs, leading to unnecessary treatments and delaying appropriate migraine therapy. This cross-sectional study examined the frequency of cranial autonomic symptoms in pediatric/adolescent patients with migraine at 4 different sites over the course of the study period. Of 125 pediatric migraineurs, 62% had at least one cranial autonomic symptom based on current International Classification of Headache Disorders, second edition (ICHD-II) criteria, and 70% based on proposed ICHD-III criteria. The majority had more than one cranial autonomic symptom and the symptoms tended to be bilateral. Age, sex, laterality of headache, presence of aura, and whether migraine was episodic vs chronic did not influence the likelihood of having cranial autonomic symptoms. The authors concluded that in pediatric/adolescent migraine, the presence of cranial autonomic symptoms appears to be the rule rather than the exception. They recommended that clinicians should be careful to consider migraine when evaluating a child with headache and associated ocular or nasal symptoms so as to avoid giving a misdiagnosis of sinus headache.

Morning Glory Disc Anomaly With Peripheral Retinal Nonperfusion in 4 Consecutive Cases

Duangnate Rojanaporn, Swathi Kaliki, Carol L. Shields, Jerry A. Shields, Arch

Ophthalmol. 2012;130(10):1327-1330.

Peripheral retinal nonperfusion has been described in familial exudative vitreoretinopathy (FEVR), Coats' disease, incontinentia pigmenti, retinopathy of prematurity, and fascioscapulohumeral muscular dystrophy. The authors describe the clinical characteristics and diagnostic findings in 4 patients with morning glory disc anomaly with peripheral retinal nonperfusion. All cases were unilateral and brain MRI was normal. The extent of nonperfusion was 360 degrees in 3 and 90 degrees in one patient. Three patients exhibited "brushfire" retinal vessels at the border of perfused and nonperfused retina and 1 patient showed vascular looping. There were no cases of telangiectasia, neovascularization of disc, retina, or choroid or progression of peripheral nonperfusion over a mean follow-up period of 20.3 months.

Visual Function and Optic Pathway Glioma: A Critical Response

David H. Gutmann, Robert Avery, Rosalie E. Ferner, Robert Listernick, JAMA Ophthalmol. 2013;131(1):

The authors raised concerns with an article by Dr. Parsa. They want to emphasize that in the literature only 13 cases over a 20 year period regressed, an uncommon occurrence. Emphasizing theses lesions as hamartoass can give a false impression that no harm is done to the patient. Loss of vison as well as endocrinologic dysfunction is seen in up to 50% of NF-1 associated optic pathway gliomas., However, non NF-1 optic pathway gliomas are rarely symptomatic.

Visual Function and Optic Pathway Glioma: A Critical Response In Reply

Cameron Parsa, JAMA Ophthalmol. 2013;131(1):122-124.

Optic pathway gliomas are type 1 juvenile pilocytic astrocytomas and not-mitotically active. The effect of the tumor on vision cannot be correlated with tumor size. The tumor is intrinsic to the optc nerve and alters the glial neuronal interactions necessary for neuronal function. Optic nerve dysfunctioni is not from compression. Dr. Parsa cites that spontaneous regression is not rare. Also, he emphasizes that the use of chemotherapy and radiotherapy for such lesions based on worsening vision may just reflect the effect of natural regression of the lesion with its attendant altered neuronal glial interactions. "Success" with chemotherapy or radiation may be spurious since these are not mitiotically active lesions. Caution should be used with such agents because of the risk for "chemo brain" and risk of secondary tumors in these young patients. Less toxic agents, but which inhibit protein synthesis, like rapamycin, should be considered for treatment.

Ophthalmic Artery Ischemic Syndrome Associated With Neurofibromatosis and Moyamoya Syndrome

Matthew T. Witmer, Richard Levy, Kaleb Yohay, Szilard Kiss, JAMA Ophthalmol. 2013;131(4):538-539.

The authors present the first description of a patient with evidence of retinal and choroidal infarction, and consequent necrosis, from ophthalmic artery ischemia associated with moyamoya syndrome and neurofibromatosis type 1. Moyamoya syndrome predisposes patients to cerebrovascular ischemia as the result of stenosis of the intracranial portion of the internal carotid arteries and their proximal branches. Stenosis of intracranial blood vessels lead to collateral circulation which on cerebral angiography produce an appearance of puff of smoke or MoyaMoya in Japanese. This finding is seen in patients with NF-1, Down Syndrome and sickle cell disease. Their patient underwent a neurosurgical procedure, pial synangiosis, a cerebral revascularization procedure in which a donor scalp artery is sutured to the surface of the brain. The authors recommend close ophthalmic and neurological follow-up for these patients.

Optic Pathway Gliomas:Neoplasms, Not Hamartomas

Grant T. Liu, James A. Katowitz, Lucy B. Rorke-Adams, Michael J. Fisher, JAMA Ophthalmol. 2013;131(5):646-650.

Optic pathway gliomas (OPG) are a significant neuroophthalmic cause of visual loss in children. Whether OPG are hamartomas or neoplasms is an important distinction with regard to visual prognosis and treatment. The objective of this article was to provide evidence, from review of the literatre, that OPG are neoplasms. Histopathology and growth patterns suggest they are slow growing neoplasms. Chemotherapy should be administered if progression can be documented in order to prevent visual loss.

Incidence and Associated Endocrine and Neurologic Abnormalities of Optic Nerve Hypoplasia

Brian G. Mohney, Ryan C. Young, Nancy Diehl, JAMA Ophthalmol. 2013;131(7):898-902.

This paper determined the incidence of optic nerve hypoplasia in a population-based cohort in Olmstead County, Minnesota, as well as the rate of neurologic, endocrine and developmental abnormalities. In this 25 year retrospective review, the incidence was calculated as 2.4/100,000 patients age less than 19 years old or 1/ 2287 live births. Mean age at diagnosis was 2.1 years and 53% were male. Eighty four percent had bilateral optic nerve hypoplasia. Associated conditions included primiparity in 42%, prematurity in 32% and maternal diabetes in 16%. Ocular findings included reduced visual acuity in 47%, strabismus in 42% and nystagmus in 26% . Associated systemic findings included developmental delay

in 63%, neurologic deficits in 53% and endocrine dysfunction in 26%.

6. NYSTAGMUS

Smooth-Pursuit Changes After the Tenotomy and Reattachment Procedure for Infantile Nystagmus Syndrome: Model Predictions and Patient Data

Zhong I. Wang, Louis F. Dell'Osso, Surachi Prakash, Xin Chen, J Pediatr Opthalmol Strabismus 2012; 49:295-302 Sept/Oct

There are 2 elements to visual function – static vision (visual acuity) and dyamic vision (the ability to locate new visual targets or track moving targets. It is important to be able to evaluate both aspects of visual function especially in a patient with ocular motor dysfunction such as infantile nystagmus syndrome (INS). In a previous study it was established that visual acuity can be improved with a tenotomy and reattachment procedure (T&R). T&R was also found to broaden the high acuity gaze angle range. The authors in this study wanted to evaluate whether T&R improved the smooth pursuit of patients with INS. The hypothesis was that although T&R improves static visual function, the steady state pursuit would not improve. This was based on personal experience by one of the authors. The conclusion of the study proved the hypothesis to be true; although T&R can improve peak visual acuity, broaden the high acuity gaze angle range, and reduce target acquisition time to static targets, this is not the case with moving targets.

Reading Performance in Infantile Nystagmus

Niraj Barot, Rebecca J. McLean, Irene Gottlob, Frank A. Proudlock, *Ophthalmology* June 2013;120:1232-1238

This is a prospective cross-sectional study involving 71 participants with infantile nystagmus (37 idiopathic, 34 with albinism). These 71 participants were compared with 20 age-matched controls. The purpose of the study was to characterize reading deficits in infantile nystagmus to determine optimal font size for reading in infantile nystagmus and investigate whether visual acuity and severity of nystagmus are good indicators of reading performance in infantile nystagmus.
Methods: Reading performance was assessed using Radner reading charts and was compared with near logarithm of minimal angle resolution (logMAR) visual acuity (VA), nystagmus intensity, and foveation characteristics quantified by using eye movement recordings. The eXpanded Nystagmus Acuity Function (XNAF) was used to evaluate foveation.

Conclusions: Maximum reading speeds can be near normal in infantile nystagmus when optimal font sizes are provided even in individuals with poor visual acuity or intense nystagmus. However, reading performance in infantile nystagmus is acuity sensitive to font size limitations. Font sizes for optimal reading speeds in infantile nystagmus may be as much as 6 logMAR lines worse than the near visual acuity.
Comments: This article emphasized the importance of teacher consultants for visually impaired (TCVI) and evaluation of all children with nystagmus regardless of etiology. Furthermore, ophthalmologists should test reading in their office to see what font size is the most affective in optimizing reading proficiency. These findings should be communicated with the teacher consultant who will make the ultimate decision as the most appropriate font size.

Anatomic Features and Function of the Macula and Outcome of Surgical Tenotomy and Reattachment in Achiasma

Avery H. Weiss, James O. Phillips, John P. Kelly, *Ophthalmology* July 2013;120:1469-1475

Achiasma is a rare condition characterized by failure of the nasal retinofugal fibers to decussate at the optic chiasm. As a result, nasal and temporal retinofugal fibers of each eye are routed to the ipsilateral visual cortex. Monocular visual fields are normal in spatial extent, suggesting that each brain hemisphere maps the entire visual field.
This is a case series performed in Seattle, Washington. It consists of 2 children with isolated achiasma. Ophthalmologic examinations, brain magnetic resonance imaging, full field and multi-focal electroretinography (MERG), visual evoked potentials, spectral-domain optical coherence tomography (OCT), and eye-movement recordings were performed. Bilateral tenotomy and resection was performed in 1 patient.
Visual acuity before and after surgery, macular anatomic features and function, and eye velocity before and after tenotomy and replacement (T&R) surgery were considered in the main outcome measures.
Conclusions: The findings that the macula is normal and achiasma suggest that reduced central acuity is the result of retinal image motion from nystagmus. Two-muscle tenotomy and replacement (T&R) procedures reduce horizontal retinal image motion and can improve visual acuity in achiasma or patients with infantile nystagmus.
Comments: This is an article supportive of tenotomy and replacement surgery not only achiasma but for patients with infantile nystagmus. The eye movement recordings can be seen on page 1471. It should be mentioned that the conclusion and study are based on the evaluation of 2 patients, but surgery was only performed on 1 of the patients.

Investigating the Relationship Between Foveal Morphology and Refractive Error in a Population With Infantile Nystagmus Syndrome

Healey N, McLoone E, Mahon G, Jackson AJ, Saunders KJ, McClelland JF.. Invest Ophthalmol Vis Sci. 2013;54:2934–2939.

The authors explored associations between refractive error and foveal hypoplasia in infantile nystagmus syndrome (INS). 50 participants with INS (albinism n = 33, nonalbinism infantile nystagmus [NAIN] n=17) aged 4 to 48 years were recruited for study. Cycloplegic refractive error and logMAR acuity were obtained. Spherical equivalent (SER), most ametropic meridian (MAM) refractive error, and better eye acuity (VA) were used for analyses. SD-OCT was used to obtain foveal scans, which were graded using the Foveal Hypoplasia Grading Scale.

Participants with more severe foveal hypoplasia had significantly higher MAMs and SERs (P = 0.005 and P = 0.008, respectively). There were no statistically significant associations between foveal hypoplasia and cylindrical refractive error (P = 0.144). Analyses demonstrated significant differences between participants with albinism or NAIN in terms of SER and MAM (P = 0.001). There were no statistically significant differences between astigmatic errors between participants with albinism and NAIN. Controlling for the effects of albinism, results demonstrated no significant associations between SER, and MAM and foveal hypoplasia (partial correlation P > 0.05). Poorer visual acuity was associated statistically significantly with more severe foveal hypoplasia (P = 0.001) and with a diagnosis of albinism (P = 0.001).

Increasing severity of foveal hypoplasia is associated with poorer VA, reflecting reduced cone density in INS. Individuals with INS also demonstrate a significant association between more severe foveal hypoplasia and increasing hyperopia. However, in the absence of albinism, there is no significant relation between refractive outcome and degree of foveal hypoplasia, suggesting that foveal mal-development in isolation does not impair the emmetropization process.

Visual Deprivation and Foveation Characteristics Both Underlie Visual Acuity Deficits in Idiopathic Infantile Nystagmus

Felius J, Muhanna ZA**.** Invest Ophthalmol Vis Sci. 2013; 54:3520–3525.

Previously, the attained level of visual acuity (VA) was modeled in terms of foveation characteristics of the nystagmus waveform, that is, the patient's fixation stability at the time of testing. The authors present evidence for an additional component of VA loss associated with the (partial) binocular visual deprivation experienced during the sensitive period of visual development. The premise of their approach is that pendular nystagmus waveforms are typically associated with poor foveation characteristics and thus may lead to binocular visual deprivation, whereas jerk-like waveforms with extended foveation periods usually have much better foveation.

Binocular grating VA and eye movement recordings were obtained from 56 children with IIN and documented waveform history from longitudinal visits. VA was modeled in terms of foveation characteristics (Nystagmus Optimal Fixation Function, NOFF) and of each child's time course of pendular nystagmus during the sensitive period.
Mean VA was 0.25 + 0.19 logMAR below age norms, and the mean foveation fraction was 0.28 (NOFF_ -0.9 + 2.3 logits). Nystagmus had a median onset at age 3 months and transitioned to waveforms with extended foveation at age 35 months. The best fit of the model showed the following: Poor foveation (0.01 foveation fraction) was associated with 0.60 logMAR acuity deficit; this deficit gradually reduced to zero for increasingly better foveation; pendular nystagmus during each decile of the sensitive period was associated with an additional 0.022 logMAR deficit. The model accounted for 57% of the variance in VA and provided a better fit than either component alone.
VA in IIN is explained better if, besides the child's foveation characteristics, an additional component is taken into account representing the nystagmus induced visual deprivation during the sensitive period. These findings may have implications for the timing of treatment decisions in children with IIN.

Dalfampridine in patients with downbeat nystagmus—an observational study Classen J, Feil K, Bardins S, et al. Journal of Neurology August 2013; 260(8):1992-6.

The authors investigated the effects of dalfampridine, the sustained-release form of 4-aminopyridine, on slow phase velocity (SPV) and visual acuity (VA) in patients with downbeat nystagmus (DBN) and the side effects of the drug. In this proof-of-principle observational study, ten patients received dalfampridine 10 mg bid for 2 weeks. Recordings were conducted at baseline, 180 min after first administration, after 2 weeks of treatment and after 4 weeks of wash-out. Mean SPV decreased from a baseline of 2.12 deg/s ± 1.72 (mean ± SD) to 0.51 deg/s ± 1.00 180 min after first administration of dalfampridine 10 mg and to 0.89 deg/s ± 0.75 after 2 weeks of treatment with dalfampridine ($p < 0.05$; post hoc both: $p < 0.05$). After a wash-out period of 1 week, mean SPV increased to 2.30 deg/s ± 1.6 ($p < 0.05$; post hoc both: $p < 0.05$). The VA significantly improved during treatment with dalfampridine. Also, 50 % of patients did not report any side effects. The most common reported side effects were abdominal discomfort and dizziness. The authors concuded that Dalfampridine is an effective treatment for DBN in terms of SPV and that it was well-tolerated in all patients.

7. PREMATURITY

Characteristics of Peripapillary Retinal Nerve Fiber Layer in Preterm Children Jingyun Wang, Rand Spencer, Joel N. Leffler, Eileen E. Birch. Am J Ophthal May 2012; 153 (5); pg. 850-855.

This study attempted to correlate the type and severity of peripapillary RNFL abnormalities seen in infants born less than 32 weeks gestation to normal full term controls. Twenty-five preterm children were enrolled in the study, all with regressed ROP or no ROP, normal appearing posterior poles on exam, and greater than 5 years of age. All patients were scanned with the same Spectralis FD-OCT. Results found that preterm infants had a global average thickness of RNFL lower than normal children (8%). The mean distribution profile was flatter in preterm infants. The peri-papillary RNFL termporal superior and nasal inferior sectors were thinner than normal children (9-13%). However, data from this study was collected from preterm children who underwent laser treatment (9/16) and with visual acuities ranging to as low as 20/80.

Tear Osmolarity in Premature Infants

Andrea Ryan, Bernadette Lanigan; Michael O'Keefe, J Pediatr Ophthalmol Strabismus 2012;49:348-352 (Nov/Dec)

The purpose of this article was to report any abnormalities in tear production in premature infants at birth (gestational age), at term (if the infant were born at 40 weeks) and 3 months after term (52 weeks). Previous articles have found contradictory results such that there is no clear answer. The authors found that mean tear osmolarity is within the expected range and shows no significant change as the infant matures; therefore, most preterm infants do not have dry eye based on assessment of tear osmolarity.

Congenital Nasolacrimal Duct Obstruction in Premature Children

Silvia Helena Tavares Lorena, João Amaro Ferrari Silva, Marinho Jorge Scarpi, J Pediatr Ophthalmol Strabismus 2013; 50:239-244 (July/August)

The purpose of this study was to compare premature babies to full term babies and to determine the incidence of congenital nasolacrimal duct obstruction (CNLDO). A retrospective chart review was performed that evaluated 400 infants – 200 premature and 200 full term infants. Of the 400 infants, 53 NLD in 32 premature children and 9 NLD in 7 full term infants were diagnosed with CNLDO. The study concluded that there was a higher incidence of CNLDO in premature infants than in full term infants.

8. ROP

The rate of change in retinal vessel width and tortuosity in eyes at risk for retinopathy of prematurity.

Ghodasra DH, Thuangtong A, Karp KA, et al. J AAPOS 2012;16:431-436

This study describes the change in retinal vessel width and tortuosity in at-risk eyes from the first imaging session to the session at which the most severe ROP

was noted. A digital fundus camera was used and there were two masked graders. 10 eyes (10 patients) with type 1 ROP and 31 eyes (31 patients) that did not develop ROP were included. Type I ROP eyes had a greater rate of change in venule width and for the 3 widest vessels. Arteriole width was not significantly different. However the change in arteriole vessel tortuosity and for the 3 most tortuous vessels was greater in the type I ROP group. Venular tortuosity was not significantly different. These findings were independent of birth weight and gestational age. The study had a relatively small sample size, and should be considered a pilot study.

Glaucoma in the Early Treatment for Retinopathy of Prematurity (ETROP) study

Bremer DL, Rogers DL, Good WV, et al.J AAPOS 2012;16:449-452

401 infants participated in the ETROP study. The authors report the clinical features of glaucoma in this group of patients. Within the first 6 years of life, 12/718 eyes developed glaucoma (5 in the early treatment group, 7 in the conventional group). Only 9 of the 12 had reported elevated IOP, the rest were diagnosed by other findings (optic nerve head appearance. Anterior chamber depth was shallow in 7/12 and questionably shallow in 1/12. At the time of glaucoma onset, 8/12 infants had stage 4B retinal detachments or worse. At the six-year follow-up examination, only one eye of one patient had a recordable acuity (20/400). The authors are unclear as to the mechanism of glaucoma in those eyes that did not have shallow chambers and postulate an inflammatory cause associated with previous ocular surgeries. In this study, glaucoma diagnosis was based on best clinical judgment rather than strict criteria, which could induce interexaminer variability.

Time to normalization of intracranial pressure secondary to intraventricular hemorrhage and the need for retinopathy of prematurity treatment in infants diagnosed with both conditions

Kella YR, Snir M, Ehrlich R, et al. J AAPOS 2012;16:515-517

The authors sought to determine whether lower rates of ROP treatment in premature infants was achieved by earlier control of increased ICP secondary to IVH. This was a retrospective study of consecutive patients diagnosed over a 5-year period who had both ROP (any stage) and IVH (any grade). 63 infants were diagnosed with both conditions, but those who were only followed for both conditions (n=27) or those who were not treated for increased ICP (n=15), were excluded. Of the remaining 21 patients, 10 were treated for both increased ICP and for ROP. ICP levels normalized at a significantly later postnatal age in the infants treated for both elevated ICP and ROP than the infants treated for ICP alone (100 days vs 45 days). This difference remained significant after adjustment for birth weight and gestational age. IVH-induced increased ICP was

positively correlated with advance stage of ROP. The reduced ocular blood flow secondary to elevated ICP and poor vascular autoregulation may be more susceptible to ROP.

Pediatric ophthalmology fellowship training in laser ablation for retinopathy of prematurity

Bradley MH and Motley WW. J AAPOS 2012;16:539-542

This study quantifies ROP laser surgery training received by recent pediatric ophthalmology fellowship graduates, explores potential learning modalities, and assesses the effect of training on physician willingness to treat ROP. A questionnaire was created and given to recent fellowship graduates. An on-line survey was also e-mailed to all AAPOS candidates-in-training. The mean number of ROP laser procedures reported to have been performed during fellowship training was 6.4. Just over half of the included respondents felt that they had been at least adequately trained for this procedure in fellowship. Respondents who averaged fewer procedures were more likely to report uncertain or inadequate training. Interest in performing ROP laser procedures in practice, showed a moderately strong correlation with better reported training in ROP laser treatment during fellowship. Better reported training also correlated with the number of cases performed annually in practice. The authors express concern because more recently trained pediatric ophthalmologists were more likely to report poor preparation for ROP treatment while in training. The study is limited by a relatively small sample size.

Long-term Visual Outcomes Following Lens-sparing Vitrectomy for Retinopathy of Prematurity

Ruhi Singh, Deepthi M. Reddy, Andrew Jonathan Barkmeier, Eric R. Holz et al. Br J Ophthal 2012; 96: 1395-1398.

This study is a retrospective chart review looking at the long term outcomes of lens-sparing vitrectomy for stage 4 and 5 ROP. Thirty-seven eyes of 30 patients were reviewed. Anatomic success rate for stage 4A was 91%, 4B was 88%, 5 was 40%. Mean age at last follow up was 7.1 years. Long term visual outcomes in the stage 4A group included 21 eyes. Only 16 eyes had a measureable Snellen visual acuity and 12 eyes had vision of 20/400 or better. Three eyes exhibited ability to fix and follow but remaining four were LP or NLP. Four of the 9 eyes witn stage 4B ROP had measurable visual acuity of 20/800 or better. Three of the 9 showed some ability to fix and follow, and two had LP or NLP vision. No differences were found between patients who had scleral buckling simultaneously with the vitrectomy vs. only vitrectomy.

Impact of Changing Oxygenation Policies on Retinopathy of Prematurity in a Neonatal Unit in Argentina Julio A. Urrets-Zavalia, Nicolas Crim, Erna G. Knoll, Fernando A. Esposito et al. Br J Ophthal 2012; 96: 1456-1461.

This study examined rate of ROP in Argentina where ROP is the commonest cause of blindness due to high birth rates and lack of resources and trained personnel after new policies on oxygen administration were introduced. Total of 1532 infants were examined over a period of 3 years. The first year, saturation rates of 88-96% were used; the second year, saturation rates were 83-93%. Results demonstrated that 22.5% of babies developed ROP of any stage. During the higher saturation period, 6.9% of babies developed type 1 ROP which declined to 3.6% with lower saturation rates. Fifty-four of the 65 babies developing type 1 ROP in the lower saturation group were treated. The mortality rate between the two groups was the same. Other factors also seem to be contributing to ROP development, since the rate of ROP was still higher than in developed countries.

Intravitreal Pegaptanib Combined With Diode Laser Therapy For Stage 3+ Retinopathy Of PrematurityIn Zone I And Posterior Zone II *Rudolf Autrata, Inka Krejčířová, Kateřina Šenková, Marie Holoušová,* Eur J Ophthalmol September – October 2012; 22 (5): 687-694

ROP is the leading cause of blindness in developing countries, The prevalence of worldwide blindness due to ROP is 50,000. Treatment has included cryotherapy, diode laser and now anti-VEGF therapy. This is the second published clinical trial of anti-VEGF treatment for ROP. The authors prospectively divided 152 eyes with zone I or zone II posterior ROP 3+ into two groups. Group 1 received intravitreal anti-VEGF (pegaptanib) with conventional diode laser. Group 2 received laser therapy combined with cryotherapy. There was a follow-up of 20 months post treatment. Treatment success was defined as absence of recurrence of stage 3+ ROP. There was a significant treatment success in group 1 (89.7% compared to 60.8%). Regression of plus disease and peripheral retinal vessels developed occurred more rapidly in group 1 (2.23 weeks compared to 3.57 weeks). The recurrence of neovascularization (stage 3+ROP) was also lower in group 1 (14.6% compared to 50%). However, these recurrences occurred much later in group 1 (15.1 weeks compared to 5.9 weeks), requiring a longer and more aggressive follow-up schedule. The authors recommend additional clinical trials are needed to determine the dose, frequency and timing of the injection and to study its long-term ocular and systemic side effects.

Correlation Between Periventricular Leukomalacia And Retinopathy Of Prematurity Hsiu-Mei Huang, Sue-Ann Lin, Ying-Chao Chang, Hsi-Kung Kuo Eur J Ophthalmol November – December 2012; 22(6): 980 – 984

One hundred and ninety-five patients were retrospectively studied for any correlation between ROP severity requiring treatment and/or PVL. The patients, born less than 30 weeks gestation or less than 2000g, were compared also for correlation to birthweight, gestational age, and APGAR scores There was no correlation with the presence of PVL in the severity of ROP. Only low birthweight, gestational age, and APGAR scores correlated with ROP severity requiring treatment.

An Updated Study of the Use of Bevacizumab in the Treatment of Patients with Prethreshold Retinopathy of Prematurity in Taiwan

Wei-Chi Wu, His-Kung Kuo, Po-Ting Yeh, Chung-May Yang, et al. Am J Ophthal January 2013, 155 (1)

This study monitored the effectiveness and complications of Bevacizumab, a humanized anti-VEGF monoclonal antibody, in ROP. This study was a multicenter retrospective review of patients in 4 cities of Taiwan. Each child was injected with 0.625 mg of bevacizumab intravitreally. All patients treated were found to have prethreshold disease excluding stage 4 and 5. In total, 162 eyes of 85 patients were evaluated. In this study, use of bevacizumab resulted in 88% of eyes regressing. Only 9% showed no response to the medication and required laser treatment. 1% of eyes required a second injection. Also, 2% of eyes progressed to stage 4 disease and required vitrectomy. No systemic toxicity was found. This study showed better results than the BEAT-ROP study, however, this study was retrospective, consisted of higher birth weight babies, and repeat treatment was allowed as deemed necessary by the treating physician.

Aggressive Posterior Retinopathy of Prematurity:Risk Factors of Retinal Detachment Despite Confluent Laser Photocoagulation

Gaurav Sanghi, Mangat R. Dogra, Deeksha Katoch, Amod Gupta. Am J Ophthal January 2013; 115 (1): pg. 159-164.

This study was a retrospective review of aggressive posterior ROP treated with laser photocoagulation to search for risk factors for retinal detachment. This study was performed in India looking at 109 eyes. Ninety eyes had favorable outcome and 19 had unfavorable outcome. All eyes with posterior zone 1 disease and all eyes with fibrovascular traction after laser treatment demonstrated retinal detachment. On logistic regression, the most significant factor to predict retinal detachment was gestational age younger than 29.5 weeks, hemorrhages, need for repeat laser treatment, and limited fibrovascular traction after laser.

A Cloud-Based Electronic MEdical Record for Scheduling, Tracking and Documenting Examinations and Treatment of Retinopathy of Prematurity

Robert W. Arnold, Jack Jacob, Zinnia Matrix J Pediatr Ophthalmol Strabismus 2012;49:342-346 (Nov/Dec)

The authors discuss a new method to following Retinopathy of Prematurity (ROP) patients to ensure that no infant is lost to follow up or discharged without proper follow up instructions. They discuss the use of a cloud-based system which means the software is provided as a internet based service rather than a locally installed and maintained product, having advantages of secure, backed-up storage and converged infrastructure. The concept os an interesting concept and would definitely help avoid any lost patients but there is no discussion on how to obtain such a software if one were to attempt to utilize this system. In addition, if you already have a system in place that seems to be working, putting a new system into place may complicate things and you may end up getting more lost to follow up patients and more patients who have no clue how to proceed with examinations after discharge initially. In a disease as devastating as ROP, this initial rough patch may not be worth the risk of losing even one patient.

Incidence of Retinopathy of Prematurity and Risk Factors among Premature Infants at a Neonatal Intensive Care Unit in Canada

Gloria Isaza, Sourabh Arora, Manpartap Bal, Varun Chaudhary, J Pediatr Ophthalmol Strabismus 2013;50:27-32 (Jan/Feb)

This article discusses the incidence and risk factors of Retinopathy of Prematurity (ROP) in a Canadian intensive Care Unit. In addition to low birth weight and gestational age, the study found that oxygen supplementation and necrotizing enterocolitis were independently associated with ROP. The article also reviews other studies that have discussed ROP incidence and risk factors and compares the current study with the previous studies and outlines reasons for variations in the results of the studies.

Incidence of cataract development by 6 months' corrected age in the Early Treatment for Retinopathy of Prematurity study

Davitt BV, Christiansen SP, Hardy RJ, et al J AAPOS 2013;17:49-53

The authors report the incidence of cataract development in patients up to 6 months' corrected age following randomized treatment in the ETROP trial. Of the 401 infants enrolled, 366 survived. Six hundred fifty-six eyes of the 366 subjects are included. Conventional treatment (CM) was used in 326 eyes and 330 were assigned to early treatment (ET). Eight eyes (3 ET and 5 CM) developed cataracts by 6 months. Of the 5 CM eyes, only 3 received laser treatment. Four

of the six eyes that developed a cataract after laser treatment had persistent pupillary vessels. Only 1 eye developed the cataract within ten days of treatment. There is some evidence that anterior segment ischemia played a role in the development of the cataracts (all had plus disease, 4 had persistent papillary vessels at the time of treatment, 1 developed corneal opacification, 1 developed iris atrophy, 2 displayed shallow anterior chambers, and 3 had iris synechiae). Unlike prior studies, the authors did not find that earlier treatment increases the risk of cataract development. The fact that two patients developed cataracts even though they were not treated reinforces that it is not always the laser treatment that causes the cataract. Close follow-up of these patients is warranted.

Progression of myopia and high myopia in the Early Treatment for Retinopathy of Prematurity Study: Findings at 4 to 6 years of age

Quinn GE, Dobson V, Davitt BV, et al J AAPOS 2013;17:124-128

Children who participated in the ETROP study had their refractive errors recorded between ages 4 and 6. There were 401 enrolled infants, all of whom developed prethreshold ROP in one or both eyes. Infants were either treated at an earlier stage of ROP (ET) or conventionally managed (CM) with treatment or not if regression occurred. Myopia and high myopia were defined as spherical equivalent of >=0.25 D of myopia and >=5.00 D of myopia respectively. Both ET and CM patients had >60% myopia and more than a third of eyes displayed high myopia. In the CM group, >75% of infants who required treatment developed myopia, but <50% of infants whose ROP regressed and did not require treatment developed myopia. Rates of myopia increased early in life, but stabilized at the 4-,5-, and 6-year examinations. There was no difference in median monthly rate of change between ET and CM eyes. More than 20% of infants had myopia >8.00 D by age 4 years. Based on this study, patients who required treatment for their ROP had higher rates of myopia, but there was no difference between the ET and CM groups. Most of the myopic progression occurred by age 3 years. Plus disease and the zone of the ROP also affected prevalence of myopia and high myopia. This information is helpful when counseling patients on risks of myopia and the course of progression.

Clinical characteristics of children with severe visual impairment but favorable retinal structural outcomes from the Early Treatment for Retinopathy of Prematurity (ETROP) study

Siatkowski RM, Good WV, Summers CG, et al J AAPOS 2013;17:129-134

This study describes visual function abnormalities in children who were enrolled in ETROP, with visual impairment despite relatively normal ocular structure. This cohort was selected from children who had completed the 6-year examination. Evaluations occurred for 342 of the 370 surviving children. Thirty-nine (11%) met

inclusion criteria. Normal bilateral fundus exams were present in 25/39 (64%). Macular ectopia or straightening of temporal vessels was seen in one or both eyes of 11 patients. One patient had a stage 4B detachment in one eye. Eighteen children had optic atrophy in one or both eyes. Three patients had isolated increased cupping of the optic disk, and another nine had cupping associated with optic atrophy. Thirty children had nystagmus. About half of those with nystagmus had optic atrophy and half did not. Of the 39 included patients, 25 had definite CVI, 12 had combined anterior and posterior pathway disease, and 2 had indeterminate cause of visual loss. Optic atrophy is not specific for subcortical/cotical visual loss and is often present in ROP and other retinal disease. Hypoxic prenatal brain damage or damage to the lateral geniculate nucleus can cause optic atrophy. PVL can cause nonglaucomatous cupping of the optic disk. Many PVL patients often have nystagmus as well, which may indicate combined or isolated posterior visual pathway disease. The authors found that postgeniculate disease as the only or main factor to cause visual impairment was uncommon (25/342). Despite occurring infrequently, it is still a higher rate than that seen in the CRYO-ROP study, which may be due to increased survival of younger infants in the current study. Manifest nystagmus can be seen in the setting of anterior visual pathway disease or with posterior visual pathway disease. Therefore manifest nystagmus in isolation, cannot help localize the etiology of visual loss. Also, the absence of cupping or optic atrophy does not rule out posterior pathway involvement as the cause of decreased visual acuity. Combining an evaluation of neurologic status, developmental status and visual function can help determine possible posterior pathway involvement in vision loss.

Comparison of fentanyl and morphine in laser surgery for retinopathy of prematurity

Orge FH, Lee TJ, Walsh M, and Gordon K J AAPOS 2013;17:135-139

There is no consensus as to the best choice of anesthesia for laser photocoagulation in the treatment of ROP. Fentanyl's action is more rapid and briefer with better CNS penetration. It is more potent and can cause adverse respiratory effects. This study provides pilot data on intravenous sedation during ROP laser ablation. The medical records of consecutive preterm neonates undergoing laser treatment of ROP over a 3-year period were reviewed retrospectively. Morphine was used for the first two years and then fentanyl was substituted. The primary outcome was rate of change in ventilation status after anesthesia administration during ROP surgery. A total of 40 neonates (53 treatments) were reviewed and 35 treatments were analyzed (no more than one per infant). Seventeen subjects were in the morphine group and 18 were in the fentanyl group. The rate of any change in ventilation status, as well as the rate of change in ventilation status greater than one level, was greater in the morphine group. More patients (5/17) in the Morphine group experienced any worsening of ventilation status than in the Fentanyl group (1/18). Ventilation status declined two or more levels in 3/17 in the Morphine group and 0/18 in the Fentanyl group.

None of these differences were statistically significant because of the small group sizes. The Morphine group also had 1 episode of temperature instability, 6 apneic events, and 7 bradycardic events, versus 0, 2, and 5 respectively. Fentanyl caused slightly more desaturation events (12 versus 9). True change in ventilation status of one level or greater, was 5.30 times more common in the Morphine group. Overall there was a trend towards fewer adverse events with Fentanyl, but the sample sizes are small. Also the two agents were used sequentially and there may have been other factors in the NICU that changed as well. A larger prospective study would be needed to truly determine whether the use of one of these agents is less likely to cause side effects.

Patent Ductus Arteriosus and Indomethacin Treatment as Independent Risk Factors for Plus Disease in Retinopathy of Prematurity

Irena Tsui, Edward Ebani, Jamie B. Rosenberg, Juan Lin, Robert M. Angert, Umar Mian, J Pediatr Ophthalmol Strabismus 2013; 50:88-92 (March/April)

The authors discuss factors that may realted to causing plus disease or retinopathy of prematurity requiring disease. A retrospective, cross-sectional study evaluated patients who were on indomethacin and had patent ductus arteriosus. The main outcome measure was increased rate of plus disease or ROP requiring treatment. The study concluded that PDA and indomethacin were associated with plus disease and ROP requiring treatment but this was not significant after adjusting for other risk factors. PDA was strongly related to bronchopulmonary dysplasia and blood transfusions which may explain the results.

Intravitreal Bevacizumab for Retinopathy of Prematurity: Refractive Error Results

Bjorn C. Harder, Frank C. Schlichtenbrede, Stefan Von Baltz, Waldemar Jendritza et al. Am J Ophthal June 2013; 155 (6): pg. 1119-1124.

This retrospective study compared babies with ROP treated with laser to those babies treated with intravitreal Bevacizumab. The outcomes of treatment were reported as well as refractive outcomes after the ROP regressed. The study group consisted of 23 eyes of 12 children with acute posterior stage of ROP or stage 3+ with neovascularization in 5 adjacent sectors. These children received an injection of either 0.375 mg or 0.625 mg (depending on treating physician). The control group consisted of 26 eyes of 13 children with similar treatment criteria. These children were monitored for one year. No children in the study group required a second injection. One child in the control group received intravitreal Bevacizumab and another child developed a partial retinal detachment. At the end of the follow up period, refractive error was evaluated. The study group showed a mean refractive error of -1.04+/- 4.24 diopters versus

the control group with a mean refractive error of -4.41+/- 5.50 diopters. Refractive astigmatism was monitored as well with the study group showing a mean of -1.0+/- 1.04 diopters and the control showing 1.82+/- 1.41 diopters.

Retinopathy of Prematurity in Extremely Low Birth Weight Infants in Turkey

Selim Demir, Osman Sayin, Canan Aygün, Hüseyin Ortak, et al J Pediatr Ophthalmol Strabismus 2013; 50:229-233 (July/August)

The purpose of the study was to determine the incidence of retinopathy of prematurity in extremely low birth weight infants (less than 1000g) in Turkey. 225 infants were found to have ELBW from January 2003 to September 2011. Low birth weight infants have a higher incidence of retinopathy of prematurity (ROP). With advances in neonatology, ELBW infants are surviving and as such, screening, diagnosis and treatment of ROP is even more important now.

The effectiveness of policy changes designed to increase the attendance rate for outpatient retinopathy of prematurity (ROP) screening examinations

Barry GP, Tauber K, Emmanuel G, et al J AAPOS 17:3;296-300

The authors reviewed records of consecutive neonatal intensive care unit patients at one hospital, before and after the implementation of policy changes, to determine attendance rates for the first outpatient appointment after discharge. The policy changes included education forms, streamlined scheduling, and a patient log. Attendance on the recommended date was increased from 22/52 (42%) prior to policy change, to 46/57 (81%) after policy implementation. The number of patients who met the criteria for conclusion of the acute retinal screening examination also improved significantly, from 90% pre- to 100% post-implementation. Exclusion criteria included fetal demise, hospital transfer, NICU discharge before first ROP screening, and completion of ROP screening as an in-patient. The groups had similar rates of mean round-trip travel for the appointments, and percentage of infants whose surname changed after discharge. Because multiple intensive policies were instituted simultaneously, it cannot be determined which of these policies were most effective. This was a retrospective study and the patients in the post-implementation arm of the study were all enrolled during the first year of the new policy changes. It is possible that over time the implementation of the policy and enthusiasm for its enforcement will wane, and results might worsen. However, ROP examination follow-up compliance is critical, and the authors are to be commended for attempts to increase parental compliance.

Visual Acuity, Optical Components, and Macular Abnormalities in Patients with a History of Retinopathy of Prematurity

Wei-Chi , Rey-In Lin , Chia-Pang Shih , et al
Ophthalmology September 2012;119:1907-1916

This is a perspective case-controlled study from the Chang Gung Memorial Hospital, Department of Ophthalmology, Taoyuan, Taiwan.
There were 133 patients in the study to examine the optical components and spectral domain optical coherence tomography (SDOCT) findings in children with a history of retinopathy of prematurity (ROP) and identify any associations between the OCT findings and the visual acuities of patients. The authors concluded that patients who were treated using laser therapy or cryotherapy for retinopathy of prematurity (defined as threshold ROP) were more likely to show abnormal foveal development and have a poorer visual prognosis than other patient groups despite a fundus with no macular dragging, disc dragging, or retinal detachment. A steeper corneal curvature, shallower anterior chamber, and greater lens thickness are the main changes in optical components in these patients.

Mechanisms and Management of Retinopathy of Prematurity

M. Elizabeth Hartnett; John S. Penn *N Engl J Med*; December 2012;

367:2515-2526

This is a bench to bedside review of retinopathy of prematurity. The authors describe the two phases of retinopathy of prematurity: (1) delayed physiologic vascular development and (2) vasoproliferation in terms of two animal models. The mouse model of oxygen induced retinopathy is easy to work with and study, but imperfectly mimics the human disease, whereas the rat model better approximates the human disease but is harder to manipulate and study. On the basis of molecular mechanisms identified in animal models of oxygen-induced retinopathy, some translational considerations for retinopathy of prematurity management are presented. Antioxidants, such as vitamin E, may be beneficial for reducing severe retinopathy of prematurity, but broad antioxidant inhibition may not be systemically safe. Erythropoietin supplementation is still inadequately understood. Intravitreal bevacizumab improves outcomes for zone 1 stage 3 ROP, but can cause progression to stage 5 in some patients, can lead to persistent retinal avascularity and recurrent late angiogenesis, and has an uncertain systemic safety profile. Nutritional factors, including postnatal weight gain, can be considered in ROP screening algorithms to decrease the number of babies requiring screening. However, algorithms developed in certain populations do not behave in the same way when applied to other populations (for example, an American algorithm performed more poorly in Mexico).

Letter to the editor: mechanism and management of retinopathy of prematurity.

Rajesh Rao; Brian Diouhy *N Engl J Med*; March 2013; 368(12):1161

The authors urge additional study of the VEGF-fibronectin pathway as a potential target for medical and surgical therapy for patients with ROP. Cleavage of fibronectin induces posterior vitreous detachment, which decreases vitreoretinal traction and reduces the need for vitreoretinal surgery. Use of autologous plasmin enzyme, which cleaves fibronectin, during vitreoretinal surgery was associated with improved outcomes in stage 4 and 5 ROP. Ocriplasmin was recently approved by the FDA as an agent to cleave fibronectin and modulate the vitreoretinal interface. We may hear about ocriplasmin in the treatment of stage 4 and 5 ROP in the near future.

Letter to the editor: mechanism and management of retinopathy of prematurity.

Bonnie Jasany; Ruchi Nanavati; Nandkishor Kabra *N Engl J Med*; March 2013; 368(12):1161-2

The authors discuss the growing interest in propranolol as a treatment for stage 2 retinopathy of prematurity. The interest in this drug arose from observations that it may reduce VEGF levels in infantile hemangiomas. Mouse models have not shown promise for propranolol as an ROP drug, but there is a human trial currently ongoing (Propranolol in Newborns with Retinopathy of Prematurity: PROP-ROP).

Alternative methods for the screening of retinopathy of prematurity: binocular indirect ophthalmoscopy *vs* wide-field digital retinal imaging

M A Sekeroglu; E Hekimoglu; H T Sekeroglu; U Arslan *Eye*; September 2013; 27(9):1053–1057

The study compared wide-field digital retinal imaging using the Retcam shuttle with binocular indirect ophthalmoscopy for retinopathy of prematurity (ROP) screening. 58 infants of gestational age 24 to 32 weeks (median 30 weeks) and birth weight 760 to 2000 g (median 1335 g) in a Turkish hospital were enrolled in the study. They had a total of 124 bilateral eye examinations performed. Two ophthalmologists experienced in ROP exams performed the exams: one did the indirect ophthalmoscopy with scleral depression, whereas the other (masked to the results of indirect ophthalmoscopy) performed the imaging. The authors calculated the sensitivity and specificity of wide-field digital imaging, compared to the “gold standard” indirect ophthalmoscopy in the detection of any stage ROP and treatment requiring ROP. The sensitivity for any stage ROP was 58.6 and for treatment requiring ROP was 100%. The specificity was 100% for any stage ROP and for treatment requiring ROP, meaning that the wide-field digital retinal imaging did not “overcall” ROP. This study showed that wide field retinal imaging

was excellent for the diagnosis of severe and treatment-requiring ROP. However, it was suboptimal for detecting any ROP because of difficulty imaging the periphery, which becomes problematic in terms of deciding when to terminate ROP screening. A potential statistical critique of the study is using the individual eye, rather than the patient, as a unit of analysis. Another limitation is that an experienced ophthalmologist acquired the images, and perhaps acquired better images than would be acquired by a technician. One of the purported benefits of wide field digital retinal imaging is having non-ophthalmologists screen babies in settings with insufficient ophthalmologist screeners.

Posterior to the ridge laser treatment for severe stage 3 retinopathy of prematurity

A L Ells; G A Gole; P Lloyd Hildebrand; A Ingram; et al. *Eye.* April 2013; 27(4):525-30

The authors report a case series from Alberta Children's Hospital in which additional laser treatment, called 'posterior laser', was delivered posterior to the neovascular ridge, for eyes with severe stage 3 ROP in zone II. Eighteen eyes of 11 infants were treated with posterior laser. These were babies with mean birth weight of 688 grams (552-930 g) and mean gestational age of 24 weeks (23-28). Fourteen of these eighteen laser treatments were performed after standard anterior laser had failed, and four were performed as a primary treatment. The authors did not specify why this was performed as primary treatment in four eyes. Sixteen of the eighteen treated eyes had regression of the neovascular ridge, and two eyes went on to stage 4A detachments requiring vitrectomy. The authors presented morphological criteria for posterior laser treatment, including thick stage 3 in four confluent temporal clock hours, but acknowledge that the morphological criteria have not been validated. Studies are necessary in which a control arm receives standard anterior laser treatment.

Astigmatism and biometric optic components of diode laser-treated threshold retinopathy of prematurity at 9 years of age

C-S Yang; A-G Wang; Y-F Shih; W-M Hsu *Eye.* March 2013;27(3):374-81.

The study assessed the prevalence of astigmatism and described ocular biometry among nine year-old children who had received diode laser treatment for threshold ROP as neonates. 24 children (46 eyes) were included in the study. They underwent cycloplegic refraction using an autorefractor, keratometry, and A scan ultrasound biometry. Their refractive errors and biometric data were compared with data obtained from a national survey of full-term age-matched controls.
The laser-treated eyes had a mean astigmatism of 3.47 D, whereas control eyes had 0.08 + 0.90 D (P<0.001). Laser-treated eyes had a mean spherical equivalent of −4.49 D., whereas control eyes had -0.44 + 1.48 D (P<0.001).

50% of the laser-treated eyes had high astigmatism (>3.0 D). Most laser-treated astigmatic eyes (97.7%) showed with-the-rule astigmatism, with the mean plus cylinder axis at 89 degrees. The astigmatism in the refraction was highly correlated with the corneal astigmatism. The corneas were significantly steeper vertically (P=0.003) and flatter horizontally (P=0.031) in eyes with laser-treated ROP when compared with age-matched full-term controls. The eyes with laser-treated ROP also show significantly thicker lens (3.93 mm) and shallower anterior chamber depth (ACD; 2.92 mm) than full-term controls (P<0.001).
The authors conclude that there is a higher prevalence and greater magnitude of astigmatism in eyes with laser-treated threshold ROP compared with full-term controls, due to vertical corneal steepening. They recommend continued follow up of ROP treated children to detect amblyogenic refractive errors.

Insulin-like growth factor binding protein-3 in preterm infants with retinopathy of prematurity.

Gharehbaghi MM, Peirovifar A, Sadeghi K, Mostafidi H. Indian J Ophthalmol 2012;60:527-30.

This study was conducted to determine whether insulin-like growth factor bindingprotein -3 (IGFBP-3) is associated with proliferative ROP and has a role in pathogenesis of the disease in premature infants. Materials and Methods: A total of 71 preterm infants born at or before 32 weeks of gestation participated in this study. Studied patients consisted of 41 neonates without vaso-proliferative findings of ROP as the control group and 30 preterm infants with evidence of severe ROP in follow up eye examination as the case group. Blood samples obtained from these infants 6-8 weeks after birth and blood levels of IGFBP-3 were measured using enzyme-linked immunosorbent assay (ELISA). Results: The mean gestation age and birth weight of the studied patients were 28.2±1.6 weeks and 1120.7±197 gram in the case group and 28.4±1.6 weeks and 1189.4±454 gram in the control group (**P**=0.25 and **P**=0.44 respectively). The infants in the case group had significantly lower Apgar score at first and 5 min after birth. Insulin-like growth factor binding protein -3 (IGFBP-3) was significantly lower in the patients with proliferative ROP than the patients without ROP [592.5±472.9 vs. 995.5±422.2 ng/ml (**P**=0.009)]. Using a cut-off point 770.45 ng/ml for the plasma IGFBP-3, we obtained a sensitivity of 65.9% and a specificity of 66.7% in the preterm infants with vasoproliferative ROP. Conclusion: Our data demonstrated that the blood levels IGFBP-3 was significantly lower in the patients with ROP and it is suspected that IGFBP-3 deficiency in the premature infants may have a pathogenetic role in proliferative ROP.

Utility of Large Spot Binocular Indirect Laser Delivery for Peripheral Photocoagulation Therapy in Children

Saranya C. Balasubramaniam, Brian G. Mohney, Genie M. Bang, Thomas P.

Link, et.al. Arch Ophthalmol. 2012;130(9):1213-1217.

Ablation of the avascular retina, first with cryotherapy and more recently with laser therapy, has been a mainstay in the treatment of advanced retinopathy of prematurity. Trese in the 1990's advocated that the avascular retina be densely treated with laser with no skip areas. In order to achieve complete ablation of the avascular retina, the authors propose to study large spot size laser delivery through a binocular indirect laser delivery system. One patient with bilateral retinopathy of prematurity received photocoagulation
with standard spot size burns placed adjacently to LSS burns. Using a pixel analysis
program called Image J on the Retcam picture, the areas of each retinal spot size were determined in units of pixels, giving a standard spot range of 805 to 1294 pixels and LSS range of 1699 to 2311 pixels. Additionally, fluence was calculated using theoretical retinal areas produced by each spot size: the standard spot setting was 462 mJ/mm_2 and the LSS setting was 104 mJ/mm_2. For eyes with retinopathy of prematurity, this study shows that LSS laser indirect delivery halves the number of spots required for treatment and reduces fluence by almost one-quarter, producing more uniform spots. The importance of this finding is that the time it takes to treat a child is reduced with attendant reduction of time under anesthesia which is beneficial for these infants who often have multiple medical problems.

Improved Clinical Assessment of a Mouse Model of Retinopathy of Prematurity

Marco Zarbin, Arch Ophthalmol.2012;130(11):1461.

The author comments on a study published in Translational Vision Science and Technology about an in vivo method that can provide real time imaging of the effects of oxygen on retinal vascularization in a mouse model of ROP. The device is an endoscopic camera which applanates the cornea to image the retina. In mice with oxygen-induced retinopathy, topical endoscopy fundus imaging demonstrated retinal vascular incompetence with a high degree of interobserver and intraobserver agreement. This technique permits one to follow the course of the disease quantitatively in vivo, at relatively low cost. The traditional, but invasive, methods of scoring the severity of disease, such as retinal vaso-obliteration and retinal neovascularization, in postmortem tissue is time-consuming, but facilitates the assessment of novel drugs to treat ROP. The search for safe, effective drugs to treat ROP remains a high priority, and the oxygen-induced retinopathy model plays an important role in this search. Introduction of topical endoscopy fundus imaging into this paradigm may improve the quality of data analysis and the speed of drug screening.

Improving Preterm Ophthalmologic Care

in the Era of Accountable Care Organizations

C. Jason Wang, Alison A. Little, Karen Kamholz, Jaime Bruce Holliman, et.al. Arch Ophthalmol.2012;130(11):1433-1440.

This study seeks to understand current barriers to preterm ophthalmologic care as well as opportunities for improvement in the context of the chronic care model and within the structure of accountable care organizations. The study design included interviews and focus groups consisting of parents of VLBW infants and eye care providers and coordinators from 6 hospitals, representative of urban/suburban communities from the North and South in the United States. The authors conclude that under accountable care organizations, hospitals and ophthalmologic care providers should share responsibility for care and follow-up of VLBW infants at risk and with ROP in order to mitigate the medico legal liability and promote seamless care. Health information systems can promote communication among providers to facilitate care. Self-management tools can help parents meet the challenge of complying with recommended follow-up appointments for their infants who often have multiple medical issues.

Swedish National Register for Retinopathy of Prematurity (SWEDROP) and the Evaluation of Screening in Sweden

Gerd E. Holmstro¨m, Ann Hellstro¨m, Peter G. Jakobsson, Pia Lundgren, et.al. Arch Ophthalmol. 2012;130(11):1418-1424.

The purpose of this study was to evaluate screening for ROP in Sweden and consider revision of current guidelines. At the start of the study, infants 31 weeks +6 days or less were screened for ROP. Data from the Swedish National Register for ROP (SWEDROP) was extracted for the year 2008-2009 and compared with a national perinatal quality register. SWEDROP has a national coverage rate of 96%. Treatment was performed in 4.4%, none of whom had a gestational age at birth of greater than 28 weeks. Nine infants with gestational age greater than 28 weeks developed Stage 3 ROP, but regressed spontaneously. The total number of exams in their cohort of 1784 babies was 9286, with almost one thousand exams occurring in infants born at 31 weeks gestation. The authors conclude that it may be possible to reduce the upper limit of age for screening by one week to 30 weeks plus 6 days or less but cautioned that sicker and more mature infants may still need to be screened.

The CHOP Postnatal Weight Gain, Birth Weight, and Gestational Age Retinopathy of Prematurity Risk Model

Gil Binenbaum, Gui-shuang Ying, Graham E. Quinn, Jiayan Huang, et.al. Arch Ophthalmol. 2012;130(12):1560-1565.

This is a retrospective review with the goal of developing a postnatal weight gain, gestational age and birth weight prediction model for developing ROP in a cohort

of babies eligible for screening. Infants with birth weight less than 1501 g or 30 weeks or less gestational age were eligible for study. Daily weight gain rate was recorded and risk for developing ETROP Type 1 or 2 ROP was determined. If the infant went beyond a certain risk, examination was indicated. 524 infants were studied. 4% developed Type 1 ROP and were treated with laser. 5% had Type 2 ROP. The model accurately predicted all infants who developed Type 1 and all but one with Type 2. The infant who developed Type 2 ROP, which was not predicted by the model, did not require laser. Using the daily weight gain rate risk model would have reduced the number of screenings by 49%. The authors caution that prior to adopting this model for wide scale clinical use, larger population studies should be conducted to determine sensitivity.

Correlation of Recognition Visual Acuity With Posterior Retinal Structure in Advanced Retinopathy of Prematurity

David K. Wallace, Don L. Bremer, William V. Good, Rae Fellows, et.al. Arch Ophthalmol. 2012;130(12):1512-1516.

Form and function usually correlate well. This study compares the appearance of posterior retinal structure with recognition visual acuities obtained at the 6-year follow-up exam of the children who participated in the ETROP study. Concordant outcomes were found in 72%, that is good visual function correlated with good structural outcome. Discordant outcomes occurred in 15%, with 86 eyes having favorable structural outcome, but poor vision, and 6 had unfavorable structural outcome, but good vision. When posterior retinal structure is normal, but vision is poor, consider other diagnoses, such as optic atrophy or cortical visual impairment.

Signaling Pathways Triggered by Oxidative Stress That Mediate Features of Severe Retinopathy of Prematurity

Haibo Wang, Sarah X. Zhang, Mary Elizabeth Hartnett, JAMA Ophthalmol. 2013;131(1):80-85.

Reactive oxygen species can trigger signaling pathways that have been implicated in the pathogenesis of ROP. In this study, the authors examine the current role oxidative stress plays in activating signaling pathways that cause pathologic features in severe retinopathy of prematurity. Rather than broadly proposing antioxidant therapy, it is helpful to study the signaling cascades activated by reactive oxygen species that mediate pathologic features in ROP to find potential safer therapies.

Efficacy of the Screening Algorithm WINROP in a Korean Population of Preterm Infants

Jung-Han Choi, Chatarina Lo¨fqvist, Ann Hellstro¨m, Hwan Heo, JAMA

Ophthalmol. 2013;131(1):62-66

WINROP is an algorithm based on serial measurements of neonatal body weight to predict proliferative retinopathy of prematurity. Records of preterm infants with gestational age less than 32 weeks body weight in a Korean hospital were studied.
Infants were weighed weekly and entered into a computer- based surveillance system, WINROP, and the outcome was analyzed. 314 preterm infants participated and in 166 of 314 infants (52.9%), a high risk alarm was noted. In the high-risk alarm group, 36 infants developed type 1 ROP requiring treatment. In this Korean population, the WINROP algorithm had a sensitivity of 90% for identifying infants with type 1 ROP.

Need for Revised Screening Protocol for Early Detection of Retinopathy of Prematurity in Infants Born Before 25 Weeks

Kamal Kishore, Kamlesh S. Macwan, JAMA Ophthalmol. 2013;131(4):546.

Screen infants less than 25 weeks gestation no later than 29 weeks gestation. So not to miss treatable ROP.

Need for Revised Screening Protocol for Early Detection of Retinopathy of Prematurity in Infants Born Before 25 Weeks In Reply.

Michael P. Blair, Michael J. Shapiro, JAMA Ophthalmol. 2013;131(4):546-547.

The authors reiterate that the aim of their study was to describe reactivation patterns and progession to RD after the 54 week BEAT-ROP endpoint. Treatment is suggested when plus disease returns or extraretinal fibrovascular tissue growth is seen, either posteriorly or anteriorly. Additionally, to reduce the follow-up burden and prevent late reactivation, the authors recommend laser treatment of persistent avascular retina past 60 weeks' postmenstrual age since one cannot predict which eyes may reactivate months or years later.

Recurrence of Retinopathy of Prematurity Following Bevacizumab Monotherapy:Is It Only the Tip of the Iceberg?

Kamiar Mireskandari, Gillian G. W. Adams, Nasrin N. Tehrani JAMA Ophthalmol. 2013;131(4):544-545.

Comment on Berrocal and Moshfeghi's paper on late recurrence of ROP after Avastin injection. Their average time to recurrence after injection (up to 35 weeks). So in BEAT_ROP endpoint was 55 weeks PMA. This endpoint needs to be extended. However in Berrocal study, questions about the study patients, a 32 week larger infant who developed APROP and did poorly. The definition of

recurrence as not just ROP.
Only complete vascularization to the ora and no active disease can be considered successful.

Plus Disease in Retinopathy of Prematurity Qualitative Analysis of Diagnostic Process by Experts

Nina J. Hewing, David R. Kaufman, R. V. Paul Chan, Michael F. Chiang, JAMA Ophthalmol. 2013;131(8):1026-1032.

Plus disease, the most significant diagnostic finding in defining severe treatment -requiring ROP, has definable clinical features, but diagnostic consistency among practioners is a problem. This study examines the cognitive process 6 experts used in their analysis of infants with Plus, pre-Plus or no Plus disease. The experts were asked to use a "think out loud" model and were videotaped describing their reasoning. They also were asked the type and location of anatomical details, such as arterial and venous dilation and tortuosity, which contributed to their decision making. The study found that experts differed in their reasoning process, retinal features which were focused on, and interpretation of the same features.

Bilateral Effect of Unilateral Bevacizumab Injection in Retinopathy of Prematurity

Cagatay Karaca, Ayse Ozturk Oner, Ertugrul Mirza, Osman Ahmet Polat, et.al. JAMA Ophthalmol. 2013;131(8):1099-1101.

The authors describe 4 infants with aggressive posterior ROP who were administered intravitreal bevacizumab 0.625mg in one eye and whose Plus disease and extraretinal neovascularization regressed significantly in the contralateral uninjected eye. All uninjected eyes eventually received laser ablation to the avascular retina since the response to bevacizumab was insufficient to halt disease activity sufficiently. The authors highlight that this anti-VEGF agent must cross the blood retinal barrier since there were visible effects in the contralateral uninjected eye. Other studies demonstrate detectable serum levels of bevacizumab after unilateral or bilateral intravitreal injection, as well as depressed serum levels of VEGF. Systemic effects on the developing vasculature of other organs can only be speculated, but serve as a cautionary tale in the use of this powerful agent, which suppresses vascular activity in the retina and probably beyond.

9. STRABISMUS

Exotropia in children with high hyperopia.

Kassem IS, Rubin SE, and Kodsi SR. J AAPOS 2012;16:437-440

This retrospective study of children with exotropia and hypertropia (at least 4 diopters) culled patient data from a 20-year period. Patients were divided into those who had received full hyperopic correction, partial hyperopic correction, or no correction. 26 patients (age 2.5 months- 9 years at first exam) were followed for an average of 4.2 years. 23 had intermittent exotropia. 15/26 patients had developmental delays, or other associated medical conditions. 86% (19 of the 22 patients whose acuity could be measured) displayed amblyopia. 23 of the 26 patients were treated with glasses. Ten of the 15 treated with full correction experienced improvement or resolution of their exotropia. Average deviation decreased from 26 PD to 13.5 PD. Three of the 8 children treated with partial correction experienced improvement or resolution of their exotropia (ave 21 PD pre- and 16 PD post). All 5 patients who underwent strabismus surgery developed postoperative accommodative esotropia that was controlled with spectacles.
This study found children with exotropia and hyperopia had a high incidence of developmental delays and a greater rate of amblyopia than typically seen in exotropes. Full hyperopic correction seemed to improve the exotropia more frequently in these patients. However, study groups were small so statistical analysis could not be performed. Children with developmental delays often have impaired accommodation, so these results may not be applicable to the general population of exotropes. Also, the exotropes were not uniform and the study included both constant exotropes and intermittent exotropes (basic, true- and pseudodivergence excess and convergence insufficiency types). The patients were seen at a tertiary center and therefore developmental delay rates and amblyopia rates may not be generalizable. The study was retrospective so the decision on what type of glasses to give each child was not randomized or uniform. Reduced plus glasses were also not uniform in the amount of reduction prescribed.

Discrepancies between parental reports and clinical diagnoses of strabismus in Korean children

Han KE and Lim KH. J AAPOS 2012;16:511-514

The authors compared parental reports and clinical diagnoses regarding ocular deviation and to analyze concordance rates based on several factors. Medical records of children <15 years old seen over a 5-year period were retrospectively reviewed. An in-house questionnaire was also performed. 45% of children with a reported esotropia were orthophoric and another 21% actually had intermittent exotropia. In contrast, 94% of children with a reported exotropia were exotropic (3% were esotropic and 3% were orthophoric). Overall the concordance rate was 67%, with lower rates for reported esotropia, younger children, and smaller angles. This paper dealt with Asian children so results may have been skewed by pseudoesotropia. This was a tertiary referral center so there is potential bias (falsely elevated concordance).

Possible association of congenital Brown syndrome with congenital cranial dysinnervation disorders

Ellis FJ, Jeffrey AR, Seidman DJ, et al. J AAPOS 2012;16:558-564

This study explored the etiology of congenital Brown syndrome by reporting several patients with congenital Brown syndrome associated with other ocular motility abnormalities in the ipsilateral or contralateral eye consistent with known congenital cranial dysinnervation syndromes (CCDDs). This was a retrospective chart review over a four year period. Nine cases were identified. Three patients had Brown syndrome in one eye and contralateral superior oblique palsy. Two patients had a Brown syndrome in one eye and Duane syndrome type 1 in the contralateral eye. One patient had Brown syndrome and congenital ptosis in the same eye. Three patients had Brown syndrome and ipsilateral hypoplasia of the superior oblqie muscle. The authors propose that abnormalities in the development of the trochlear nerve or its nucleus may lead to abnormal development of the superior oblique-muscle-tendon-trochlea complex. The authors also propose that some cases of congenital Brown syndrome are associated with or in the spectrum of CCDDs.

Monovision Correction for Small-Angle Diplopia

Matthew D. Bujak, Andrea K. Leung, Mila Kisilevsky, Edward Margolin. Am J Ophthal September 2012; 154(3): pg. 586-592.

This study attempted to see if symptoms of small-angle binocular diplopia could be relieved in presbyopic adult patients with monovision correction. Twenty adults greater than 45 years of age with less than 10 prism diopters of acquired deviation were evaluated. The Diplopia Questionnaire was used to quantify symptoms. This questionnaire has been validated against the Goldman diplopia field. Secondary outcome was subjects' specific quality of life after monovision correction, the standardized Amblyopia and Strabismus Questionnaire (ASQE). Monovision was performed with contact lenses in 4 patients, glasses in 14 patients, and both in 2 patients. Testing of binocular contrast sensitivity has shown suppression when one eye is defocused a +2.50 add. Results demonstrated no significant difference between the +2.50 or +3.00 monovision. The Diplopia Questionnaire showed 85% improvement, 10% unchanged, 5% worsening. Statistically significant improvements were observed in the double-vision and social contact/appearance portion of the improved quality of life scale. Monovision shows promise as an alternative treatment for patients with diplopia.

Binocular Visual Acuity in Intermittent Exotropia: Role of Accommodative Convergence

Song Joon Ahn, Hee Kyung Yang, Jeong-Min Hwang Am J Ophthal December 2012; 154(6): pages 981-986.

This study evaluated binocular vision in patients with intermittent exotropia. Sixty-three patients were between 8-15 years of age with intermittent exotropia and no other ocular abnormalities. Binocular visual acuity was measured first and then monocularly with best correction. Accomodative responses were also measured with the WAM-5500 binocular autorefractor/keratometer. This study showed that binocular interaction of visual acuity is associated with accommodative response during binocular vision in patients with exotropia, helping maintain control. Angle of exodeviation at distance was seen to increase as accommodative response increased. This study suggests that binocular inhibition in a patient with intermittent exotropia may indicate diminishing control.

Nonsurgical Management of Diplopia

Lisa Fraine, Am Orthopt J. August 2012;62:13-18

This review article focuses on benefits and limitations of nonsurgical management of diplopia due to incomitant strabismus associated with restrictive strabismus after various ocular and orbital surgeries. Nonsurgical methods may be used indefinitely or to temporize until surgery is performed. Practical methods prism correction and monocular occlusion are described.

Diplopia Following Cosmetic Surgery

Marlo Galli, Am Orthopt J. August 2012;62:13-18

This review article sites small case series and single case reports to conclude that diplopia is a rare complication but is likely due to aggressive excision of fat especially from the lower lids. It is recommended that the eyelid surgeon visualize and isolate the fat pad before excising and to identify the inferior oblique in order to avoid violating it. If post-operative diplopia persists for 8 weeks following blepharoplasty then consultation with a strabismologist is recommended for nonsurgical or surgical management.

Nonsurgical Management of Diplopia After Orbital Decompression Surgery

Jorie L. Jackson, Am Orthopt J. August 2012;62:29-33

This review article sites the incidence of new onset diplopia with orbital decompression ranging from 2.6% with deep lateral wall decompression to 30% with medial wall and floor decompression. The authors report that most surgeons wait at least six months after orbital decompression but also mention that some operate sooner if there is evidence of disease inactivity on imaging. In addition

prism and occlusion in the nonsurgical management of restrictive strabismus, the authors suggest lowering the bifocal segment in spectacle to permit some patients to use chin up head position and to consider botulinum toxin injection in patients with acute thyroid eye disease. Patients with thyroid eye disease may require higher than typical botulinum toxin dosage and may have shorter duration of treatment effect.

Nonsurgical Management of Diplopia after Retinal Surgery

David J. Hodgetts, Am Orthopt J. August 2012;62:38-43.

This review article covers pathophysiology of diplopia due to strabismus or nonstrabismus causes such as macular disease and optical issues caused by retinal surgery. Post-operative strabismus is discussed including local anesthetic myotoxicity, scaring of extraocular muscles, changes in one or more force vectors due to an encircling band placed under extraocular muscle(s), cryotherapy, and trauma to muscle by cleaning or disinsertion and reattachment. Diagnosis and management of diplopia due to nonstrabismus causes such as metamorphopsia, image size distortion associated with stretching or compression of retinal cells and refractive aniseikonia are discussed. Patients may benefit from treatment of strabismus with prisms, maculopathy with partial occlusion and aniseikonia with optical correction. Some patients may require various combinations of these options.

Where the Wild Things Are: When Esotropia Misbehaves

Christopher Lyons, Am Orthopt J. August 2012;62:61-69.

This publication of the John Pratt-Johnson Annual Lecture highlights clinical pearls in the assessment of patients with esotropia using 12 case vignettes. The challenging diagnostic scenarios described include consecutive esotropia unmasking superior oblique palsy, over-corrected myopia, fixation switch diplopia, comitant esotropia with cerebellar tumor, sixth nerve palsy due to pontine tumor or cavernous pathology and thyroid strabismus mimicking sixth nerve palsy.

Positive Angle Kappa: A Possible Sign of Aniridia

Kimberly S. Merrill, C. Gail Summers Am Orthopt J. August 2012;62:70-76.

The authors prospectively evaluated the corneal light reflexes of 16 patients with congenital aniridia. While fixating on a penlight monocularly, all eyes were found to have at least 1 mm of medial displacement of the light reflex. The authors conclude that although some features of aniridia are variable, a positive angle kappa is commonly present and therefore should be included in the assessment of patients with nystagmus or decreased vision.

Prisms Are Effective In Resolving Diplopia From Incomitant, Large, And Combined Strabismus

Madhura A. Tamhankar, Gui-Shuang Ying, Nicholas J. Volpe Eur J Ophthalmol November – December 2012; 22(6): 890 – 897

Symptomatic diplopia due to incomittant strabismus may be alleviated by use of prisms, even oblique ones. In this retrospective study, 64 patients with restrictive strabismus were treated by prisms. Although vertical diplopia was more successfully treated (83 %) than horizontal diplopia (70%), 73% of all patients continued using prisms as opposed to 23% who underwent surgery. The authors recommend managing diplopia with prisms initially or in the interim.

Factors Associated With Strabismus in Spina Bifida Myelomeningocele

Heather A. Anderson, Karla K. Stuebing, Ray Buncic, Malcolm Mazow, Jack M. Fletcher, J Pediatr Opthalmol Strabismus 2012; 49:284-289 Sept/Oct

Spina bifida myelomeningocele (SBM) is a developmental disorder in which the spinal cord and meninges protrude through a defect in the vertebral column. The lesions can cause a variety of abnormalities such as lower limb motor paralysis, sensory deficits, and bowel/bladder difficulties below the lesion. Strabismus is commonly associated with SBM which has been linked to the fact that patients with SBM often have hydrocephalus. This article sort to link strabismus and SBM regardless of hydrocephalus. This study found a relationship between strabismus and SBM as well as between lesion location and strabismus. The authors found that higher level lesions were more likely to be associated with strabismus than lower level lesions

Ocular Torsion Among Patients with Intermittent Exotropia: Relationships with Disease Severity Factors

Kwang Hoon Shin, Hye Jin Lee, Hyun Taek Lim Am J Ophthal January 2013; 155(1): 177-182.

This study evaluated 150 patients with intermittent exotropia between the ages of 4 and 15 years and evaluated the distribution of ocular torsion. Torsion was measured with photography, the disc fovea angle was calculated using digital software. Ocular torsion was discovered in 30% of patients with exotropia. The mean disc foveal angle was 12.32+/-2/78 degrees. Controls had only 5.13+/-2.79 degree angle. Ocular torsion was only seen in 10.7% of controls. In addition, there appeared to be a correlation with low stereopsis and torsional presence.

Clinical Course and Characteristics of Acute Presentation of Fourth Nerve Paresis

Ayman Khaler, Emma Dawson, John Lee, J Pediatr Ophthalmol Strabismus 2012;49:366-369 (Nov/Dec)

Superior Oblique palsy is the most frequent acquired vertical and tortional diplopia, as well as anomalous head position. A retrospective review of 32 patients was performed to follow the typical course of a fourth nerve palsy. Microvascular disease is a very common cause of acquired fourth nerve palsy and following the course of this revealed that 89% of cases completely resolved within 10 months. Only one patient needed surgery and 3 patients continued with prisms.

Functional magnetic resonance imaging of horizontal rectus muscles in esotropia

Schoeff K, Chaudhuri Z, and Demer JL
J AAPOS 2013;17:16-21

Many ideas about the biological mechanisms of esotropia exist. Medial rectus excessive innervation +/- deficient lateral rectus innervation, hypercontractility, and mechanical shortening have all been proposed. Information about strabismus in animals is reported. The authors obtained functional MRI data on size and contractility of human horizontal rectus muscles in concomitant esotropia. Adult non-strabismic control subjects (n=13) and patients with esotropia (n=12) were enrolled. Four of the esotropic subjects had undergone previous ocular surgery (1 orbital fracture repair and 3 strabismus surgery). Medial rectus muscle cross sections averaged up to 39% larger in esotropic patients, and lateral rectus muscles were larger as well but not significantly so. Medial rectus contractility was significantly increased up to 60% in esotropic subjects whereas lateral rectus muscle contractility was also increased but not significantly so. The authors felt that these findings imply an element of central gaze co-contraction. The anatomic findings may be a consequence of abnormal innervational patterns. Nine of the esotropic subjects were myopic and no comment is made on the control group. Duration of strabismus varied widely from a few months to many years and this could also affect muscle size and contractility but this was not addressed.

Fusion can mask the relationships between fundus torsion, oblique muscle overaction/underaction, and A- and V- pattern strabismus

Deng H, Irsch K, Gutmark R, et al J AAPOS 2013;17:177-183

The purpose of this study was twofold. The authors analyzed the correlation and relationship between the grade of objective ocular torsion, the grade of oblique

muscle overaction/underaction, and the amplitude of A and V pattern. They also looked at the role of residual fusion in controlling these correlations. Patients were obtained from a 30-year retrospective chart review based on a diagnosis of abnormal fundus torsion or an A- or V-pattern strabismus. Three hundred ninety-six patients were included with a roughly equal distribution between esotropes and exotropes. A strong correlation existed between the size of the pattern, the degree of fundus torsion, and the degree of oblique over/underaction. The presence of stereopsis decreased the correlation while the absence of stereopsis increased it. The relationship between superior oblique overaction and an A-pattern, and inferior oblique overaction and a V-pattern were statistically significant. Lack of stereopsis increased the percentage of patients with a pattern strabismus who had oblique overaction and the presence of steropsis diminished it. Fundus excyclotorsion was correlated with inferior oblique muscle overaction, and incyclotorsion was correlated with superior oblique muscle overaction. These correlations were statistically significant and increased or decreased based on the absence or presence of stereopsis, respectively. There was a strong correlation between obique overaction and fundus torsion. There was a statistically significant correlation between pattern strabismus and fundus torsion. This relationship was lessened to a statistically significant degree if stereospsis was present in the V-pattern but not the A-pattern group. The correlation between pattern strabismus, oblique overaction, and fundus torsion has been well known, but the impact of the presence or absence of stereopsis on these relationships is of interest to the reader.

Diplopia after glaucoma drainage device implantation

Abdelaziz A, Capo H, Bannitt MR, et al J AAPOS 2013;17:192-196

The authors estimate the incidence of diplopia after glaucoma drainage device (GDD) implantation using 15-year financial claims data of a large university hospital-based glaucoma practice. The accuracy of the claims data was verified through a retrospective review of the medical records. Of 2661 patients who underwent GDD surgery, 59 were coded as having diplopia or undergoing strabismus surgery. Diplopia developed in 1.4% of GDD patients. All cases developed within one year of GDD surgery., with the majority having an onset 2 weeks to 3 months after surgery. The majority of superotemporal GDD surgery cases developed exotropia and hypertropia; of the inferonasal GDD surgeries, almost half developed hypotropia. Slightly more than half of the diplopia cases were treated with prisms. Only 3 patients underwent surgery (2- altered drainage device or bleb, 1- strabismus surgery). Five patients had spontaneous resolution of their diplopia. This study is limited by its retrospective nature and reliance on financial claims data. The true incidence of diplopia was most likely underestimated.

A computerized version of the Lancaster red-green test Awadein A J AAPOS 2013;17:197-202

Changes in vergence can cause fluctuations in the results of measured ocular misalignment on the Lancaster red-green test. Accurate plotting of the results on paper is subject to operator error. A new software program based on the Lancaster red-green test is compared to the original. Consecutive adult patients who complained of diplopia caused by incomitant strabismus were enrolled over a 9-month period. The software-based test was performed on a 40-inch monitor at a distance of 50cm (monitor version) and a second time with a projector and a screen at a working distance of 1 meter (projector version). Eighty-two patients were enrolled with a mean age of 34.3 years. Conventional testing results were comparable to computerized results for vertical and torsional deviations. For horizontal deviations, agreement was better between the traditional testing and the projector version than with the monitor version. Measured deviations for both computerized versions tended to be smaller than with conventional testing. Test timing averaged between 7 and 8 minutes for all three versions of testing. The computer monitor testing substituted blue for green because blue targets are more efficiently filtered on a computer monitor. Computerized results also have the advantage of being immediately stored and they can be transmitted electronically.

Ocular Torsion Among Patients With Intermittent Exotropia: Relationships With Disease Severity Factors

Kwang Hoon Shin, Hye Jin Lee, Hyun Taek Lim Am J Ophthal Jan 2013; 155 (1); pg. 177-182.

This prospective study evaluated 600 eyes of 300 children to determine the distribution of ocular torsion amongst intermittent exotropes and whether this correlates with severity of exotropia. One hundred fifty patients with intermittent exotropia were compared to normals. Torsion was determined by using a digital nonmydriatic fundus camera. The disc-fovea angle was calculated off of these photos. The median age in the exotropia group was 6 years. The average amount of exotropia was 28.29 diopters. The disc-foveal angle of extorted eyes was 12.32 on average. Ocular torsion was found on at least 30% of patients with exotropia. The control group, the mean disc-foveal angle for extorted eyes was 11.33 on average. Ocular torsion was only seen in 10.7%. In addition, there was a positive correlation between the amount of ocular torsion and the severity of exotropia. In addition, there was a positive correlation between ocular torsion degree and level of stereoacuity.

Quantifying Diplopia with a Questionnaire

Jonathan M. Holmes, Laura Liebermann, Sarah R. Hatt, Stephen J. Smith, *Ophthalmology* July 2013;120:1492-1496

This is a cross-sectional study. The purpose of this is to report a diplopia questionnaire (DQ) with a data-driven scoring algorithm. One hundred and forty-seven adults with double vision associated with strabismus completed both the diplopia questionnaire and the Adult Strabismus-20 quality-of-life questionnaire (HRQOL).
Conclusions: The authors state that they developed a data-driven scoring algorithm for the diplopia questionnaire. The diplopia questionnaire has excellent test-retest reliability and responsiveness and may be useful in both clinical and research settings.

Amblyopia and sensory features at initial presentation of Brown syndrome: an issue to recognize

H T Sekeroglu; E Muz; A S Sanac; E C Sener; et al. *Eye.* April 2013; 27(4):515-8

The study investigated the frequency of amblyopia and sensory features at initial presentation in patients with unilateral congenital Brown syndrome. This was a retrospective chart review from one Turkish institution, identifying 44 patients with Brown syndrome of ages 4-21 years, with median age of 5 years. The frequency of amblyopia was 15.9% (seven patients) at initial presentation. The amblyopic eye was not necessarily the Brown syndrome eye. Absence of sensory fusion, assessed by the Worth 4 dot test, was associated with amblyopia.

Automated Analysis of Binocular Alignment Using an Infrared Camera and Selective Wavelength Filter.

Yang HK, Seo J-M, Hwang J-M, Kim KG. Invest Ophthalmol Vis Sci. 2013; 54:2733-2737.

This study is a comparison of a computerized software that automatically quantifies the angle of strabismus from photographs based on a biometric 3-dimentional eye model with good reproducibility and minimal inter-observer variability. The software shows excellent agreement with the Krimsky test, but did not measure the latent component of strabismus, resulting in less correlation with the prism cover test. The authors developed an occluder made with a filter that blocks the subject's view and all visible light, but selectively transmits infrared light with wavelengths above 720nm. The photographs taken with the selective wavelength filter in front of the eye visualize the details of the eye completely behind the occluder, while blocking the subject's view. These infrared images may reveal the latent components of strabismus that are manifest only after disruption of fusion. They evaluated the efficacy of this system to estimate binocular alignment with the gold standard of the prism and alternate cover test (PCT). The 95% limit of agreement of inter-observer variability was + 4.8 prism diopters (D) for the PCT and + 4.3 PD for the selective wavelength filter analysis. The 95% limit of agreement of test-retest reliability between the PCT and

selective wavelength filter analysis was + 8.5PD. Results of the PCT and selective wavelength filter analysis showed a strong positive correlation (R=0.900, P<0.001). This automated method is an accurate and reliable tool for measuring ocular deviation with minimal observer dependency.

PRISM USE IN ADULT DIPLOPIA

Kammi B. GuntonCurrent Opinion in Ophthalmology Sept. 2012 23(5) p. 400-404

Prismatic correction to restore binocularity in adult diplopia can be challenging. This review summarizes the results of prismatic correction in adults based on the cause of diplopia and gives guidelines for prescribing prisms. Satisfaction with prismatic correction is achieved in approximately 80% of all adult patients with diplopia from all causes. Of patients with vertical diplopia, skew deviation and fourth nerve palsy have the highest satisfaction rates, 100 and 92%, respectively. Patients with thyroid eye disease and orbital blowout fractures associated with diplopia had the lowest satisfaction rates, 55 and 8%, respectively. With regard to horizontal deviations, patients with decompensated childhood strabismus with a combination of horizontal and vertical deviations and patients with convergence insufficiency had satisfaction rates of 71 and 50%, respectively. Careful selection of patients for prismatic correction, management of patient's expectations, and continued follow-up to monitor the symptoms are critical to the successful use of prisms.

Inferior rectus palsy as an isolated ocular motor sign: acquired etiologies and outcome Kwang-Dong Choi, Jae-Hwan Choi, Hee Young Choi, et al. Journal of Neurology January 2013; 260(1):47-54.

This retrospective study identified 44 patients with acquired inferior rectus (IR) palsy between April 2006 and May 2011 from four Neurology and two Ophthalmology Clinics in Korea. The aim of the study was to elucidate underlying etiologies, lesion locations, and outcomes of acquired IR palsy. They analyzed clinical features, the results of radiological and laboratory evaluation, and prognosis. The most common causes were vascular (*n* = 16, 36 %) and trauma (*n* = 12, 27 %). Vascular disorders included microvascular ischemia (*n* = 10, 23 %), cerebral infarction (*n* = 5, 11 %), and dural arterio-venous fistula (*n* = 1, 2 %). Other causes were inflammation (*n* = 7, 16 %), myasthenia gravis (*n* = 5, 11 %), and thyroid ophthalmopathy (*n* = 1, 2 %). They were unable to determine the etiology in the remaining three patients (7 %). Most patients (95 %) showed a complete recovery with or without treatment. The authors concluded that acquired IR palsy mostly occurs with brainstem or orbital lesions, and has an excellent prognosis for spontaneous recovery.

An Expanded View of Infantile Esotropia:Bottoms Up!

Michael C. Brodsky, Arch Ophthalmol. 2012;130(9):1199-1202.

The pathogenesis of congenital esotropia is poorly understood. The debate goes back to Alexander Chavasse who proposed that esotropia occurs in response to an abnormal oculomotor environment, a theory termed the reflexogenic theory. Claude Worth supposed an innate defect in the fusion faculty. This debate continues. Brodsky sheds new light on our understanding of the cause of congenital esotropia. Instead of a defect in the visual cortex, Brodsky contends that the problem leading to infantile esotropia arises in the cortico-mesencephalic cerebellar area. Binocular cortical maldevelopment permits atavistic subcortical visual pathways to remain operational. This integrated neuroanatomical model predicts that primary neurodevelopmental disorders involving the accessory optic system or its connections to the cerebellum can also give rise to infantile esotropia. Infantile ET reflects reactivation of primitive subcortical systems.

Torsion and Pattern Strabismus:Potential Conflicts in Treatment

Burton J. Kushner, JAMA Ophthalmol. 2013;131(2):190-193

In this study, Dr. Kushner highlight the potential adverse affects of rectus muscle transposition on torsion or pattern strabismus. This was a retrospective nonblinded medical record review of patients treated by the author between January 1, 1990, and June 30, 2009, in whom rectus muscle transposition to address pattern strabismus worsened torsion, or in whom transposition to address torsion worsened pattern strabismus. Eight patients were identified, 5 in whom torsion developed because of transposition to address pattern strabismus and 3 in whom pattern strabismus developed after transposition to address torsion. The presence of bifoveal fusion and/or Graves orbitopathy were risk factors for these adverse outcomes.

Esotropia Greater at Distance:Children vs Adults

Erin P. Herlihy, James O. Phillips, Avery H. Weiss, JAMA Ophthalmol. 2013;131(3):370-375.

Esotropia greater at distance than at near can be related to abducens palsy or to divergence insufficiency. This retrospective study of 32 patients examined the clinical and eye movement findings that distinguish abducens palsy from divergence insufficiency. Details regarding age, medical history, oculomotor and neurological examinations, and result of any neuroimaging studies were recorded. Eye movements were recorded in 2 subjects using binocular video-oculography. Fifteen children and 17 adults were identified; 93.3% of the children had an underlying central nervous system disorder that coincided with the onset of their esodeviation, and 23.5% of the adult patients had an underlying central nervous system disorder. Eye movement recordings in 2 pediatric patients revealed lateral incomitance suggestive of abducens palsy not detected by clinical examination. Esotropia greater at distance pattern in an otherwise healthy

adult is more likely due to age-related reduction in accommodation, increased ratio of accommodative vergence to accommodation, and relative divergence insufficiency.

Inferior Oblique Myokymia:A Unique Ocular Motility Disorder

Nicholas D. Chinskey, Wayne T. Cornblath, JAMA Ophthalmol. 2013;131(3):404-405.

The authors describe a unique form of myokymia involving monocular, high frequency,
low-amplitude contractions causing excyclotorsion, not incyclotorsion, induced by supraduction, suggesting an inferior oblique myokymia. This is the opposite of what is expected for superior oblique myokymia, in which infraduction triggers incyclotorsion. The etiology of superior oblique myokymia is uncertain, with reports suggesting vascular compression of the trochlear nerve, direct involvement of the muscle, and brainstem disorders. In our case, there was no abnormality of other oculomotor nerve functions, perhaps lending support to this being a primary muscle problem

Thromboembolism and Congenital Malformations From Duane Syndrome to Thalidomide Embryopathy

Cameron F. Parsa, Matthieu P. Robert, JAMA Ophthalmol. 2013;131(4):439-447.

Fibrin emboli and focal hypoperfusion may explain the development of many sporadic congenital malformations. In their study, the authors apply inductive and deductive reasoning to study a pathophysiologic mechanism to unify a variety of disparate sporadic congenital malformations such as Duane syndrome, Peters anomaly, unilateral congenital cataracts, and the morning glory disc anomaly. All these share a vascular territory of the carotid arteries and a propensity for left-sided involvement in girls. Most aberrant misinnervation phenomena such as jaw-winking syndrome, crocodile tear syndrome, Brown syndrome, and congenital fibrosis syndrome and, by extrapolation, the hypoplasia or dysgenesis of noncephalic anatomical structures (including limbs) may be similarly explained. Such malformations will occur more frequently under thrombogenic conditions, such as those induced by thalidomide.

Divergence Insufficiency Esotropia Is a Misnomer

David Mittelman, JAMA Ophthalmol. 2013;131(4):547.

In a commentary on Dr. Chaudhuri and Dr. Demer’s article on sagging eye syndrome which describes the pathophysiolocy of non-neurologic causes of divergence paralysis esotropia, Dr. Mittelman emphasizes that the term divergence insufficiency esotropia is a misnomer and that the designation of divergence insufficiency esotropia should be reserved only for those patients with

serious neurological disease. Instead, he proposes that the term “adult-onset age-related distance esotropia” should be used to describe patients with acute onset esotropia in the distance which results from aging changes of orbital tissues and disruption of the LR-SR band.

Divergence Insufficiency Esotropia Is a Misnomer In Reply

Zia Chaudhuri, Joseph L. Demer, JAMA Ophthalmol. 2013;131(4):547-548.

Dr. Chaudhuri and Dr. Demer agree with Dr. Mittelman that “age-related distance esotropia” (ARDE) should enter the lexicon to describe this non-neurologic condition

No, Not More Talk About Duane Syndrome

Creig S. Hoyt, JAMA Ophthalmol. 2013;131(4):522-524.

In this editorial Dr. Creig Hoyt comments on the article by Parsa and Robert which suggests that thromboembolism during the perinatal period may be a causative factor in a number of congenital malformations, for example, Duane syndrome and benign sixth nerve palsy in childhood. The absence or hypoplasia of the sixth cranial nerve and innervation of the lateral rectus muscle by a branch of the third cranial nerve. characterizes Duane syndrome. How this specific and stereotypic miswiring comes about in Duane syndrome remains a mystery. Rather than seeing it come about because of a genetic abnormality resulting in impaired axonal guidance, they assert that “Duane syndrome may develop following a focal vascular insult to the sixth nerve trunk with axonal degeneration, allowing for substitutive innervation from third nerve axons to the lateral rectus muscle.” Parsa and Robert suggest the possibility that the benign sixth nerve palsy of childhood and Duane syndrome have a common pathogenesis differentiated only by the timing of insult, being prenatal in Duane syndrome and postnatal in the benign sixth nerve palsy of childhood. Dr. Hoyt encourages further study of the proposal by Parsa and Robert that these features could be explained by emboli from the heart.

Sagging Eye Syndrome: Connective Tissue Involution as a Cause of Horizontal and Vertical Strabismus in Older Patients

Zia Chaudhuri, Joseph L. Demer, JAMA Ophthalmol. 2013;131(5):619-625.

To avoid unneeded neurologic evaluation and neuroimaging, the authors stress the importance of recognizing the clinical features of this syndrome. Acquired vertical and horizontal strabismus may be the result of rupture of the LR-SR band causing horizontal rectus pulley displacement and EOM elongation. This is suggested on clinical exam by blepharoptosis and superior sulcus deficit.

Congenital Bilateral Aplasia of Medial Recti in a Family

Anirudh Singh, Anudeepa Sharma, Pradeep SharmaJAMA Ophthalmol. 2013;131(6):798-800.

The authors present a father and 2 sons (non-sanguinous) with impaired adduction OU and large angle exotropia with preserved vertical ductions and no ptosis. MR of the orbits revealed hypoplastic medial rectus. At time of surgery, only a fibrous capsule was visualized in the location where the medial rectus should be found. The authors hypothesize that the orbital layer of the medial rectus was intact, but the global layer was aplastic. A large lateral rectus recession and split vertical transposition with Foster modification gave improved alignment of less that 20 Δ exotropia on post-op exam. Genetic analysis was not performed to check for mutations in genes associated with cranial dysinnervation syndromes.

10. STRABISMUS SURGERY

Diplopia After Strabismus Surgery

Sandra Holgado, Am Orthopt J. August 2012;62:5-8

This review article concludes that persistent diplopia following eye muscle surgery for multiple forms of strabismus is uncommon based on the findings of two previously published retrospective cases series. Patients with pre-operative comitant strabismus are less likely to have diplopia than those with pre-operative incomitant strabismus. Pre-operative prism adaptation specifically for patients with exotropia is described but does not seem to predict whether patients will have post-operative persistent diplopia.

Diplopia After Glaucoma Surgery

Stephen P. Christiansen, Am Orthopt J. August 2012;62:9-12

The author presents a single case report and discussion of post-operative strabismus associated with glaucoma surgery. While siting 4 previously published studies, the author concludes that the implantation of a tube shunt device (verses filtering surgery without device) and older patient age each increase the risk of new strabismus and diplopia following glaucoma surgery. In addition to a typical ocular alignment and motility measurements for strabismus surgical planning, the strabismus surgeon is advised to consider the stability of intraocular pressure, location of tube shunt device and bleb (especially in proximity to extraocular muscles), forced duction testing and double Maddox rod testing. Intraopertive issues may include need to revise the filtering bleb, repositioning of the device, trimming the device plate, temporary ligature of the drainage tube to manage intraoperative hypotony, meticulous manipulation of the

conjunctiva and water-tight conjunctival closure. Risk of endophthalmitis should be considered.

Diplopia Secondary to Orbital Surgery

David I. Silbert, Noelle S. Matta , Eric L. Singman Am Orthopt J. August 2012;62:22-28

The authors present a single case report, describe pathophysiology of diplopia following repair of orbital floor fracture and make recommendations help avoid and/or manage this clinical entity. Recommendations include careful strabismus measurements prior to orbital floor fracture repair, use of transconjunctival approach instead of subciliary approach, infracturing at the edges of the floor fracture if necessary to ensure that all orbital tissue is freed from the fracture site, blunt dissection rather than sharp, non-adherent implant surface such as nylon or polyethylene instead of bare titanium, avoid attempting to cover posterior fractures with implant, repeat forced ductions intra-operatively after placement of implant. Diplopia in downgaze is more likely to warrant surgery than diplopia in upgaze and may often benefit from a Faden procedure on the contralateral inferior rectus muscle.

Surgical management of bilateral esotropic Duane syndrome.

Sachdeva V, Kekunnaya R, Gupta A and Bhoompally VR.
J AAPOS 2012;16:445-448

This paper reports surgical results with bilateral medical rectus recessions in the treatment of bilateral esotropic Duane syndrome. Consecutive patients over a 10-year period were retrospectively reviewed. There was a minimal follow-up of 3 months (average of 11.7 months), and all patients underwent standard bilateral medial rectus recessions, except two patients (1 hangback technique, and 1 had simultaneous lateral rectus recession to treat globe retraction). There were 14 patients with an average age of 11.9 years. 12/14 patients achieved surgical success in the primary position (86%) with an average reduction of esotropia from 38.1 prism diopters preoperatively to 5.1 prism diopters postoperatively. The four patients with torticollis all experienced improvement to <10'. Overshoots and globe retraction were also improved. No patient had substantial limited adduction or a substantial exotropia postoperatively. The authors increased their surgical dosages slightly higher than conventional surgery because of the expected tightness of the medial rectus muscle. This study is limited by small patient numbers, its'retrospective nature, and a lack of objective measures of torticollis, globe retraction, and field of single binocular vision.

Outcomes of Harada-Ito surgery for acquired torsional diplopia

Bradfield YS, Struck MC, Kushner BJ, et al. J AAPOS 2012;16:453-457

This study describes outcomes of modified Harada-Ito surgery in a large series of patients. This was a retrospective study of consecutive patients over 7 years. Surgical success was defined as the absence of diplopia in the primary position at distance and downgaze at near with or without the use of prisms. Partial success was the same criteria as success but with new onset restrictive strabismus in the secondary gaze positions. Twenty-six patients ranging in age from 13-89 years were included. Mean duration of follow-up was 2 years. 14 of the 26 had bilateral Harada-Ito surgery (2 with inferior rectus muscle recession) and 12 had unilateral surgery (5 with inferior rectus recession). The mean torsion correction was 10.3'. Bilateral surgery produced more correction than unilateral (12.0' versus 8.4'). Adding vertical recessions did not enhance surgical effectiveness. Surgery was successful in 73% of patients and partially successful in 7%. Lower success was associated with greater preoperative torsion. Better outcomes were associated with the presence of preoperative fusion. Surgical success was not affected by the type of suture used (absorbable versus nonabsorbable). Postsurgical complications included Brown syndrome (n=4, 2 resolved) and restrictive esotropia (n=2, 1 resolved). Six patients underwent reoperations and one patient had regression of torsion correction. Prisms were required in 4/19 success patients.
This study was retrospective and surgery was not uniform as some patients also had inferior rectus recessions. Also surgical success included the use of postoperative prisms.

Strabismus among fee-for-service Medicare beneficiaries

Repka MX, Yu F, and Coleman A J AAPOS 2012;16:495-500

This study describes the prevalence of a strabismus diagnosis and the rate of strabismus surgery in the United States aged Medicare population participating in Part B (data 2002 to 2010). This was a 5% random sample (over 1 million beneficiaries). Strabismus was diagnosed in 0.68% of beneficiaries. 92% were Caucasian and 37% were >=80 years of age. The most common types of reported strabismus were paralytic, exotropia and esotropia. Strabismus diagnosis was more common in Caucasians and the more elderly. Only 0.016% of patients received strabismus surgery and surgical intervention decreased with age. The rate of diagnosis and surgery were less in non-Whites. This data is dependent on proper coding, so mis-coding or undercoding could skew data. Paralytic strabismus would be more likely to be coded, because it would be more likely to be the chief complaint, rather than a secondary finding.

Surgical management of vertical ocular misalignment in thyroid eye disease using an adjustable suture technique

Volpe NJ, Mirza-George N, and Binenbaum G. J AAPOS 2012;16:518-522

The authors used adjustable sutures in patients with thyroid eye disease (TED). Surgical decision making was based on preoperative deviation and intraoperative

forced duction testing. Patients underwent vertical strabismus surgery for restrictive hypotropia by a single surgeon over a decade, and the results were retrospectively reviewed. Surgical plans varied based on the size of the hypotropia and limitation of upgaze. Records of 55 patients were reviewed and 54 were included (1 did not have adequate follow-up). A minimum of 4 weeks of postoperative follow-up was required. Almost half of patients had 6 months of follow-up. 59% of patients required postoperative adjustment, which allowed all of them to have single binocular vision in slight downgaze in the immediate postoperative period. Over follow-up 52/54 (96%) of subjects had an excellent or good result after a single surgery (of whom about 1/3 required prisms postoperatively to relieve diplopia) . 30/54 (55%) were orthotropic in primary position. Two patients underwent reoperation for continued diplopia. The rate of overcorrection was 20% and most commonly developed 1 to 8 weeks after surgery. Patients with abnormal thyroid function tests actually had greater surgical success than those who were euthyroid, but the euthyroid group was small. There was a trend towards lower success with greater preoperative deviations, but this was not significant. The authors propose a surgical algorithm for the treatment of these patients, but the data is retrospective, surgeries were performed by a single surgeon, and some patients had short follow-up.

The efficacy of asymmetric bilateral medial rectus muscle recession surgery in unilateral, esotropic, type 1 Duane syndrome

Dotan G, Klein A, Ela-Dalman N, et al. J AAPOS 2012;16:543-547

The authors report their results with asymmetric bilateral medial rectus recession in 28 patients with unilateral, esotropic, (type 1) Duane syndrome. This was a retrospective data collection of patients from a 19-year period. There were 28 cases (18 female) with a mean age of surgery of 8.2 years. The mean follow-up was 2.8 years. In forced primary position, the esotropia decreased from 32 PD preoperatively to 6 PD postoperatively. There was a mean 22' improvement in torticollis with 86% of patients having complete resolution of their head turn. There was a small improvement in abduction and a small worsening of adduction in the Duane eye. The authors point out that recessing the medial rectus in the unaffected eye creates a 'fixation duress' which helps decrease contracture of the affected eyes medial rectus muscle. This was a retrospective review at a tertiary referral center, with potential selection bias.

Surgical Intervention in Childnood Intermittent Exotropia: Current Practice and Clinical Outcomes from an Observational Cohort Study

Deborah Buck, Christine J. Powell, John J. Sloper, Robert Taylor et al. Br J Ophthal 2012; 96: 1291-1295.

This was an observational study of 460 patients in the United Kingdom that were followed with the diagnosis of intermittent exotropia between the years of 2005 and 2006. Only 18 of the 26 enrolled centers performed surgery on 87 of the 460 followed children (19%). Since this was an observational study, no criteria were provided for time or type of surgery. Nine (12.5%) of patients underwent additional surgery for over-correction. Three additional patients (4%) underwent additional surgery for under-corrections. Surgical success was excellent in 36% but poor alignment was seen in 36% as well (XT >15 or ET >4). At last evaluation, 11 children were esotropic. No difference was seen in outcome when comparing the type of surgical procedure performed. This study demonstrates our poor understanding and ability to still correct intermittent exotropia.

Surgical Management of Strabismus Following Vitreo-Retinal Surgery

Michael C. Struck, Am Orthopt J. August 2012;62:44-49.

This review article on strabismus surgery following viteo-retinal surgery highlights eight key management principles: the ability to regain sensory fusion must be determined preoperatively, all three axes of deviation must be addressed, lysis of adhesions and buckle removal alone may not be sufficient, muscle weakness may accompany restrictions, the functional insertion of the muscle may become the posterior edge of the buckle piece, strabismus surgery on the fellow eye may be necessary or sufficient for best outcome, undercorrection of strabismus may result from nonmuscular restrictions and ocular misalignment may have multiple causes. Three cases are presented to illustrate these points.

Medial Rectus Surgery for Convergence Excess Esotropia with an Accommodative Component: A Comparison of Augment Recession, Slanted Recession and Recession with Posterior Fixation

George S. Ellis, Cindy H. Pritchard , Leonard Baham ; Allison Babiuch Am Orthopt J. August 2012;62:50-60.

This retrospective chart review of compares the outcomes of 131 pediatric patients who had undergone one of three surgeries for esotropia measured pre-operatively greater with near fixation than with distance fixation. The authors compare the results of bilateral medial rectus recession 1) without augmentation, 2) with augmentation, 3) with posterior fixation suture and 4) with slanted re-attachment of the insertion. The authors conclude that slanted re-attachment of the insertion yielded the largest decrease in the distance-near incomitance and is technically easier to perform than posterior fixation sutures. Raw data are presented without statistical analysis.

Comparison of Preoperative and Postoperative Anterior Segment Measurements With Pentacam in Strabismus Surgery

Jae Ho Jung, Hee Young Choi J Pediatr Opthalmol Strabismus 2012; 49:290-294 Sept/Oct

There have been reports of refractive errors after ordinary strabismus surgery for various extraocular movement disorders. Most of these are transient but some have reported persistent changes after strabismus surgery. This article evaluates changes in anterior segment measurements using a Pentacam. The study found the nether a recession nor a recession/resection caused any permanent changes in anterior segment measurements; however, there was a transient change in anterior chamber depth during the early post operative period which resolved within 3 months of surgery.

Transposition Surgery for Internuclear Ophthalmoplegia

Niraj R. Nathan, Sean P. Donohue, J Pediatr Ophthalmol Strabismus 2012;49:378-381 (Nov/Dec)

Manifestations of internuclear ophthalmoplegia (INO) include adducting weakness, contralateral abducting nystagmus and a slowed adducting saccade. The authors reviewed cases of a surgical procedure to correct large exotropias associated with bilateral INO. The surgical procedure is a variation based on the Jensen transpositional position. The article reviews 5 patients. 4 of 5 patients had orthophoria after the transpositional procedure and one had a small angle exodeviation. Most of the patients required an added lateral rectus recession either unilateral or bilateral. The article also discussed the risk of anterior segment ischemia since essentially all 4 rectus muscles were operated on in some cases, they found no cases of anterior segment ischemia but their sample size was very small and in a larger study there may be more cases of anterior segment ischemia.

Surgical Results of Patients With Unilateral Superior Oblique Palsy Presenting With Large Hypertropias

Mitra Nejad, Neepa Thacker, Federico G. Velez, Arthur L. Rosenbaum, Stacy L. Pineles, J Pediatr Ophthalmol Strabismus 2013;50:44-52 (Jan/Feb)

Superior oblique palsy is one of the most common reasons for vertical strabismus. There are several different surgical procedures that can be performed to achieve orthophoria which include single muscle surgery on the superior oblique muscle, the inferior oblique muscle, the superior retuc muscle or the inferior rectus muscle. For smaller deviations less than 20 PD of hypertropia in primary gaze, the article found that one muscle surgery was adequate to schieve orthophria after one surgery; however, if there was a greater than 20 PD

hypertropia, 2 muscle procedures done at the same time were more likely to achieve orthophoria with little risk of overcorrection which tends to be the most common fear when one is planning multiple muscle procedures.

The Prevalence of Reoperation and Related Risk Factors Among Patients With Congenital Esotropia

Zhale Rajavi, Ahmad Ali Ferdosi, Mina Eslamdoust, Mehdi Yaseri, et al. J Pediatr Ophthalmol Strabismus 2013;50:53-59 (Jan/Feb)

The article discusses prevalence and reoperation rates and risk factors for the above. Risk factors being amount of deviation (over 30 PD has a higher rate of failure and needing reoperation vs. under 30 PD), age less than 15 months (this may be because the accuracy of measurements is more difficult in younger children), and lateral rectus muscle underaction. Achievement of gross stereopsis was also evaluated and the study found that in cases where surgery was performed prior to 3 years of age, gross stereopsis was more likely to be achieved.

Rectus muscle resection in Graves' ophthalmology

Yoo SH, Pineles SL, Goldberg RA, and Velez FG J AAPOS 2013;17:9-15

Traditional teaching eschews rectus muscle resections in cases of Graves strabismus. However the authors point out that in certain situations, such as functionally monocular patients, prior maximal recessions, or no obvious restrictions, resections may prove useful.
A 20-year retrospective chart review was performed. Eight patients were found who had muscle resections. The average age of the patients was 51.1 years. Six of the eight had prior decompression surgery, but their strabismus was present before this procedure. Half had prior strabismus surgery. Five were esotropic and 3 were exotropic preoperatively. Five also had a vertical strabismus. Seven lateral recti (5 patients) and 3 medial recti were resected. Seven of the 8 were orthotropic in primary position postoperatively. One medial rectus muscle that had been resected slipped but was recovered. This patient required additional surgery. No overcorrections occurred and there were no cases of atypical inflammation. Mild limited horizontal eye movements were noted in 2 patients. Other risk factors were looked at for successful results (ie. Steroid use, smoking, prior history of radioactive iodine), but the number of patients in this manuscript limit the value of this data.
Some of the lateral recti which were resected were not enlarged on CT imaging, so applying this data to resections on enlarged muscles, is of questionable benefit. The authors recommend avoidance of resections in the presence of positive forced ductions or enlargement of the muscle on imaging studies.

Improved sensory status and quality-of-life measures in adult patients after strabismus surgery

Dickmann A, Aliberti S, Rebecchi MT, et al J AAPOS 2013;17:25-28

Strabismus can affect quality of life. This study evaluated adults with long-standing strabismus and the effect of surgery on their sensory status and quality of life. The Amblyopia and Strabismus Questionnaire (A&SQ) and the Short Form Health Survey (SF-36) were used. Twenty consecutive patients were enrolled. Thirteen of the twenty achieved satisfactory postoperative alignment and an increase in binocular function. Overall mean score on both questionnaires only improved in the 13 patients with successful surgery. The A&SQ improved particularly for contact and cosmesis, estimation of distance, and visual disorientation. The SF-36 improved particularly for physical function, general health, vitality, social function and mental health domain. These results provide further support to the concept that adult strabismus surgery is not merely 'cosmetic'. When successful alignment was obtained, binocular function improved, and many quality of life indicators improved.

A comparison of ocular alignment success of hang-back versus conventional bilateral lateral rectus recession for true divergence excess intermittent exotropia

Mohan K and Sharma A J AAPOS 2013;17:29-33

Motor results of conventional lateral rectus recessions were compared with results with hang-back procedures for patients with intermittent exotropia. Patients had true divergence excess intermittent exotropia. This study was a retrospective chart review of patients over a 16-year period. Hang-back technique was performed for the first half of the 16-year period and conventional recessions were performed for the second half. Forty-two patients were included (13 hang-back, and 29 conventional). Some patients in each group also had simultaneous inferior oblique surgery. The two groups were comparable for mean age at surgery, mean preoperative deviation, mean amount of recession performed, and mean follow-up. The mean recession of the lateral rectus was 7.5mm in each group. The success of the conventional surgery was significantly better than the hang-back technique (83% vs 31% at last follow-up). Within the hang-back group, better results occurred in patients who required smaller hang-backs but the difference was not statistically significant. Some prior studies show no difference between these surgical options but this study shows a marked difference. The authors comment on the possibility that forward creep of the muscle occurred in the hang-back group. There are a number of problems with this paper. The surgical technique used by the single surgeon was consecutive and improvement in surgical skill and surgical pre-operative plan may have occurred over time, skewing success towards the conventional group. Also many of the recessions were greater than 7mm, and it is known that hang-back recessions greater than 7mm on the lateral rectus muscle may be inaccurate and less predictable. The study was retrospective and nonrandomized with a small

number of patients in the hang-back group. The pre- and postoperative measurements were taken by a single, unmasked observer. If a non-inferiority claim for hang-back lateral rectus recessions is desired, a randomized study would be more appropriate.

Adjustment versus no adjustment when using adjustable sutures in strabismus surgery

Liebermann L, Hatt SR, Leske DA and Holmes JM J AAPOS 2013;17:38-42

This study compares outcomes in a cohort of patients, all of whom underwent adjustable strabismus surgery. The decision to adjust or not, was based on a predefined target range alignment. The authors hypothesized that there should be no difference in surgical success rates between those who required adjustment and those that did not, since they were both placed into the predefined target alignment range. All surgeries were performed by one author over a ten-year period and reviewed retrospectively. Esotropes and exotropes, with and without fusion potential, were included. Eighty-nine adult patients were included (mean age 42 years). Sixty percent received an adjustment. At both the six-week and one year follow-up, the adjusted patients achieved greater success rates, but the difference was not statistically significant. The authors feel that outcomes in the adjusted group would have been worse if they had not been adjusted but there is no way to know that this is the case. Also, the target angles for immediate postoperative alignment were those used by the authors and do not represent any standard table or universally accepted measures. Some patients did not complete one-year follow-up and this may skew data. The authors comment on the need for a randomized clinical trial of adjustable suture strabismus surgery versus fixed sutures and point out the difficulties with such a protocol.

Clinical characteristics of patients that experience different rates of exodrift after strabismus surgery for intermittent exotropia and the effect of the rate of exodrift on final ocular alignment

Park KH and Kim SY J AAPOS 2013;17:54-58

There is a lack of consensus on the ideal amount of initial overcorrection after strabismus surgery for intermittent exotropia. The authors evaluate factors related to the amount of exodrift over time and the relationship between the rate of drift and final alignment. This study was a retrospective review of consecutive patients over a two-year period who underwent strabismus surgery for intermittent exotropia. Patients had to have at least one year of follow-up. Exodrift was compared between one week and one year postoperatively. Surgery techniques were not uniform as some patients had bilateral lateral rectus recessions (n=104), while others had unilateral lateral rectus recessions combined with medial rectus resection (n=26), or medial rectus resections. Of 159 patients operated on, 130 (81.76%) experienced exodrift at 12 months

follow-up. These are the included patients. The mean preoperative deviation at distance was 28.82 prism diopters (PD). At one-year postop, the mean deviation was 5.96 PD. The bulk of the exodrift occurred in the first 3 weeks after surgery. Based on the speed of exodrift, patients were divided into fast and slow groups. These groups were similar in every respect except the fast group had significantly more amblyopic patients. The fast group had slightly larger average preoperative deviations and also had a larger degree of day 1 postoperative esotropia. At 12 months, there was no statistically significant difference in ocular alignment between the two groups. Therefore rate of exodrift had no effect on final ocular alignment.

Severe complications of strabismus surgery

Bradbury JA, and Taylor RH J AAPOS 2013;17:59-63

The authors determine severe complications of strabismus surgery and outline clinical outcomes based on 2-year prospective data from the British Ophthalmic Surveillance Unit (BOSU). Surgical complications occurred in 1 per 400 operations. Sight-threatening complications occurred in 1.6 per 1000 operations; motility-related complications occurred in 0.9 per 1000 operations. Surgically induced scleritis was significantly more common in adult strabismus cases. There were 19 cases (0.08%) of globe perforation. Of these 17 were recognized at the time of surgery. There were 5 ‘lost’ muscles, all in adult strabismus surgery cases. Four involved the medial rectus muscle. This study has significant limitations. Results are based on a questionnaire design and complications may have been underreported. Some key data is missing, such as reoperation rates and resident surgical rates. Data regarding strabismus surgery complication rates, if accurate, is critical in the counseling of patients preoperatively.

Surgical results after one-muscle recession for correction of horizontal sensory strabismus in children

Hopker LM and Weakley DR J AAPOS 2013;17:174-176

The authors evaluate one-muscle recession for the treatment of sensory strabismus in children. This was a retrospective review of surgeries over a 7-year period. Thirty-three patients met inclusion criteria. Approximately 1/3 (n=12) were esotropic. The mean follow-up was 39 months. Successful postoperative alignment was achieved in 75% of the esotropes and 90% of the exotropes. The authors did not specifically look to see if lateral incomitance was induced. No patients reported postoperative diplopia. The dose-response curve was similar between esotropes and exotropes. The authors felt this procedure has utility up to a preoperative deviation of 25-30 prism diopters.

Choice of conjunctival incisions for horizontal rectus muscle surgery- a survey of American Association for Pediatric Ophthalmology and Strabismus members

Mikhail M, Verran R, Farrokhyar F, and Sabri K J AAPOS 2013; 17:184-187

This paper reports results of a worldwide questionnaire on conjunctival incision preference for adult and pediatric strabismus surgery (primary and reoperation). The questionnaire was distributed to all AAPOS members. The net overall response rate was 27.8% (301/1022). For primary surgeries, the fornix incision was the most popular with both pediatric (58%) and adult (53%) usage. For reoperations, the limbal incision was preferred in both children (58%) and adults (63%). Respondents cited less pain and inflammation postoperatively as the reasons to use the fornix incision. Better exposure and better teaching opportunity were cited for the use of the limbal incision. Limbal incision was preferred for adjustable sutures 3:1. This study is limited by a low response rate and no evaluation of risk of ASI in the decision-making process.

The effect of previous orbital decompression on results of strabismus surgery in patients with Graves' ophthalmopathy

Kim MH, Park K and Oh SY J AAPOS 2013;17:188-191

The authors evaluated the effect of strabismus surgery on proptosis and compared the surgical outcomes of patients with and without previous orbital decompression. A retrospective review of 14 years of consecutive patients with surgery for Graves ophthalmopathy was performed. A minimum of 6 months of follow-up was required postoperatively (mean followup was 25 months). Fifty-six patients were included. There was a statistically significant difference in the age between decompression patients (48.7 years) and nondecompression patients (40.2 years). Preoperative strabismus measurements were comparable between the groups. Exophthalmometric measurements improved minimally post-strabismus surgery in both groups. Strabismus surgical success (defined as no diplopia in primary and downgaze or no diplopia with prisms within 8PD horizontally and 4PD vertically) was achieved in over 90% of both the decompression and nondecompression groups. There was a 10.7% reoperation rate. Other studies have shown strabismus surgery success is lower after decompression surgery but this is not validated in this study. This discrepancy may be because in this study the indication for decompression was cosmetic in 22 of 27 patients (not for neuropathy). The medial bony strut was preserved in all patients, which may have improved strabismus surgery outcomes. Recessions of 1 or 2 muscles did not significantly affect proptosis results. This study was retrospective with no control group.

Self-Grading Effect of Inferior Oblique Recession

Jun Ho Yoo, Seung-Hyun Kim, Ji Won Seo, Hae Jung Paik, et al J Pediatr Ophthalmol Strabismus 2013; 50:102-105 (March/April)

The study determine the effects of a 10mm inferior oblique recession versus a 14mm inferior oblique recession. A retrospective study reviewed 43 patients with inferior oblique overaction associated with congenital unilateral superior oblique palsy. 17 patients had a 10mm IO recession whereas 26 had a 14mm IO recession. The study found that both IO recession procedures were self grading and no significant differences were evident at 3 months postoperatively.

Clonidine Premedication Versus Placebo: Effects on Postoperative Agitation and Recovery Time in Children Undergoing Strabismus Surgery

Laura J. Heinmiller, Leonard B. Nelson, Marc B. Goldberg, Adam R. Thode, J Pediatr Ophthalmol Strabismus 2013; 50:150-154 (March/April)

The purpose of this article is to evaluate the effects of using clonidine preoperatively to avoid postoperative agitation. The article also discusses recovery time when using clonidine versus placebo postoperatively. The article is well written and does a very good job to keep the study double blinded. The study shows that although clonidine does reduce postoperative agitation, the recovery time for patients who receive clonidine prior to surgery is longer and patients remain in the post-anesthesia care unit later than placebo. In addition, parents are also questioned to evaluate parental satisfaction. Parents were overall more satisfied with a reduction in postoperative agitation and were happier to take home a slightly more sleepy child.

Determinants of Ocular Deviation in Esotropic Subjects Under General Anesthesia

Vincent Daien, Chloé Turpin, François Lignereux, Riadh Belghobsi, et al J Pediatr Ophthalmol Strabismus 2013; 50:155-160 (March/April)

The study aimed to identify determinants of ocular deviation in patients with esotropia while under general anesthesia. Forty esotropic patients were evaluated for their ocular deviation in the awaked stated as well as under general anesthesia. The study concluded that the ocular position under general anesthesia was most influenced by preoperative ocular deviation and patient age; the older the patient, the less the ocular deviation under general anesthesia.

Changes in Sagging Extraocular Muscle Following Surgical Recession of the Superior Rectus Muscle in Rabbit Eyes

Won Yeol Ryu, Jeong Bum Bae, J Pediatr Ophthalmol Strabismus 2013; 50:162-168 (March/April)

The study evaluates whether central muscle sag has an effect on recession on rabbit models. The authors created central muscle sag in one eye purposely and in the other eye recessed the superior rectus muscle without any central muscle sag. The purpose was to determine if the effects of a muscle recession are increased when central muscle sag is created. The authors concluded that the effects of a recession were exaggerated in an eye where central muscle sag was created because there seems to be posterior migration of the nasal and temporal edges.

Decrease in the rate of esotropia surgery in the United Kingdom from 2000 to 2010 S. Jacob Heng, Caroline J. MacEwen. Br J Ophthalmol 2013; 97: 598-600.

This study evaluated the annual incidence of strabismus surgery in children aged 0-14 years in the United Kingdom over 10 years. There appeared to be a steady decline in both the incidence and frequency of strabismus surgery from 2000 to 2006. England showed a decrease of 19.7%, Scotland 29.7%, and Wales 17.3%. 2006 to 2010, the incidence of surgery only decreased by 2.1% in England, 3.3% in Scotland, and 4% in Wales. Authors suggest that the decrease in surgery may be due to a true decrease in the incidence of strabismus, earlier detection with more successful conservative treatment or an increased threshold in performing surgery.

Postoperative minimal overcorrection in the surgical management of intermittent exotropia Yoonae A. Cho, Seung-Hyun Kim. Br J Ophthalmol 2013; 97: 866-869.

This was a retrospective study evaluating the outcome of 111 patients who underwent surgery for exotropia. All patients were followed for 5 years. Patients may have had unilateral or bilateral surgery. The mean age was 7.3 years. 16 patients underwent unilateral surgery, and 95 underwent bilateral surgery. Mean preoperative deviation was 27.7 diopters. The patients were divided into 4 groups based on postoperative outcome at day 1: ortho group, minimally overcorrected group, usually overcorrected group, and highly overcorrected group. The success rate in the ortho group was 43%, 60% in the minimally overcorrected, 58% in the usually overcorrected, and 56% in the highly overcorrected. There was no difference statistically in the success rate of the 4 groups. However, the overcorrection rate was 0% in the ortho group and minimally overcorrected, and was 8% and 16% respectively for the other two groups. It appeared that the minimally overcorrected group had a lower recurrence rate than the ortho group and also had a lower overcorrection rate.

Contralateral lateral rectus recession versus recess-resect for recurrent exotropia after unilateral recess-resect. Joo Hyun Kim, Hae Jin Kim, Dong Gyu Choi. Br J Ophthal 2013; 97: 752-756.

This retrospective study looked at 39 subjects who underwent recurrent surgery for exotropia. Initially, the patient underwent primary surgery for exotropia with a unilateral recess-resect. The second procedure was than assigned to either a contralateral lateral rectus recession (9-10 mm recession) vs. contralateral recess-resect (5-6 mm recess, 4-5 mm resect). Mean follow up after the reoperation was 32 months. Success rate was 73% in the LR group and 80% in the RR group. The difference was not significant. Rate of stereopsis was similar in both groups. The author suggests that lateral rectus recession is adequate to correct 20-25 diopters of recurrent exotropia.

Preoperative Factors Predicting the Surgical Response of Bilateral Lateral Rectus Recession Surgery in Patients With Infantile Exotropia

Jason C. S. Yam, Gabriela S. L. Chong,Patrick K. W. Wu, Ursula S. F. Wong, et al J Pediatr Ophthalmol Strabismus 2013; 50:245-250 (July/August)

A retrospective study of 50 patients with infantile exotropia who had bilateral lateral rectus recessions was performed. The study analyzed preoperative parameters such as age of onset, age at surgery, interval between onset and surgery, preoperative deviation, refractive error, anisometropia, amount of surgery performed, intermittent or constant exotropia, presence of A or V pattern. The authors concluded that surgical response decreases over time due to an exotropic drift. When applying the surgical dose, one should consider both the exotropic drift and the preoperative deviation.

Intraoperative Assessment Of Medial Rectus Pulley Location In Strabismus

Dominique Thouvenin, Olivier Norbert Eur J Ophthalmol Jan-Feb 2013; 23(1): 13 – 18

The authors assessed the pulley location of the medial rectus muscle (MR) intraoperatively in 194 consecutive patients with all types of strabismus aged 2 to 64 years. The distance between scleral insertion of MR and anterior part of the pulley was measured in 357 muscles. Median location of the anterior part of the MR's pulley measured at 12.03 mm from scleral insertion, varying from 8 to 15 mm. The MR's pulley tends to be more anterior in hyperopic cases and esotropias, and posterior in myopias and exotropias. This may explain some unexpected effects of strabismus surgery.

Comparison of bilateral lateral rectus recession and unilateral recession resection for basic type intermittent exotropia in children.

Lihua Wang, Qizheng Wu, Xiangyun Kong, Zhiwei Li. Br J Ophthal 2013; 97: 870-873.

This study retrospectively evaluated 85 patients between the ages of 3-15 years with basic type exotropia who underwent surgery, eiter bilateral lateral rectus recession or recess resect surgery. Based on binocular fixation preference testing, patients with alternative fixation were selected for BLR surgery; otherwise the non-fixating eye was selected for R&R surgery. Thirty eight patients had BLR surgery and 47 patients in the R&R group. Outcomes early did not vary between the two groups. Long term success was higher in the R&R group; the undercorrection rate was lower in the R&R group; te overcorrection rate was the same between the two groups. The limitations of this study include the fact that it is retrospective and the sample size is small as well a short followup period.

Principles and general strabismus surgical rules in cyclovertical eye muscle palsies.

E. Khawam, M. Abdulaal, V. Massoud, M. Joroudi, Binocular Vision And Strabology Quarterly, Simms-Romano's. 2012;27(4):249-63.

The authors summarize their approach to cyclovertical muscle palsies. They propose that evaluation of the deviation in the cardinal directions of gaze is sufficient and oblique gazes may be omitted. The Beilschowsky forced head tilt test and double Maddox rod test are still performed. With regards to oblique muscle palsies with a vertical deviation of less than 15 prism diopters, they recommend weakening of the direct oblique antagonist muscle. For larger deviations, they recommend weakening the antagonist oblique muscle and the contralateral inferior rectus muscle for superior oblique palsies or superior rectus muscle for inferior oblique palsies. Despite careful initial measurements they report that bilateral superior oblique palsies may be overlooked initially, but surgical overcorrection must also be considered as an etiology. In vertical rectus muscle palsies, they advocate weakening the direct antagonist when the deviation is less than 15 PD. For larger deviations, they recommend a recess/resect procedure on the vertical rectus muscles. In the setting of a complete palsy, they recommend horizontal rectus muscle transposition. They report that cyclotropia is typically managed by selecting the correct vertical muscle for surgery and isolated cyclotropia is extremely rare. For isolated cyclotropia they advocate the use of sagittalisation/ desagittalisation of the vertical rectus muscle (temporal or nasal transposition of the vertical rectus muscle).

Localising rectus muscle insertions using high frequency wide-field ultrasound biomicroscopy.

Hayat Ahmad Khan, David Smith, Stephen Kraft. BJO

This prospective double masked observational study examined 27 patients undergoing muscle surgery. The location of the insertion was measured with calipers and also with the ultrasound biomicroscope. This information appears to be useful in measuring the location of a muscle after having undergone previous strabismus surgery. This study was performed using the Sonomed VuMax II UBM. 50 rectus muscles were evaluated. The degree of agreement between the two measurements was very good. All measaurements were within 1 mm. In addition, this method seems to be able to differentiate a pseudo tendon from the true muscle insertion.

Extraocular muscle insertion positions and outcomes of strabismus surgery: correlation analysis and anatomical comparison of Western and Chinese populations.

Yu-Hung Lai, Wen-CHuan Wu, Hwei-Zu Wang, Hsin-Tien Hsu. Br J Ophthal 2012; 96: 679-682.

This study attempted to analyze anatomical differences in the the eye muscle insertions. This was a retrospective study of 123 strabismus patients. This study showed that the distances from the insertion to the limbus of the IR, LR and SR observe in the Western populations was significantly longer than those observed for the Taiwanese. The MR was the most reliable regardless of the ethnicity. The larger the eyeball, the further back the muscle insertions happen to be. The Taiwanese population has a higher incidence of myopia and increased axial length. However, the LR was more anterior in the Taiwanese population compared to Western population.

Inferior oblique myectomy for upshoots mimicking inferior oblique overaction in Duane retraction syndrome

Awadein A J AAPOS 17;3:253-258

Typically the upshoot seen in the adducting eye of a patient with Duane syndrome is not treated with inferior oblique surgery. This study was a prospective, interventional study of consecutive patients over a 5-year period with Duane syndrome, who underwent an inferior oblique myectomy to treat upshoot. Eleven patients were included. The patients had to display a gradual elevation of the eye in adduction or have a hypertropia in primary position, to be included. Patients who displayed abrupt movements, which suggest possible muscle slippage, were not included. Minimum followup was six months (mean 8.6 months). Two patients underwent bilateral inferior oblique surgery and four

simultaneously had medial rectus recessions. At final follow-up, 10/11 (91%) displayed no residual upshoot. No patient developed prolonged inferior oblique underaction. V-patterns also improved. Mean pre- and postoperative hypertropia measured 5 prism diopters and 1 prism diopter respectively. It is important to note that only 17 of 59 (29%) of patients with Duane syndrome and upshoot who were seen during the study period met the inferior oblique criteria for inclusion (six families declined surgery). Therefore preoperative selection is critical when considering this surgical technique to correct Duane syndrome upshots.

Bilateral lateral rectus muscle recession with medial rectus pulley fixation for divergence excess intermittent exotropia with high AC/A ratio

Choi HY and Jung JHJ AAPOS 17:3;266-268

Over a four-year period, seven consecutive patients with an exodeviation >=10 prism diopters (PD)more at distance than at near and with a high AC/A ratio underwent bilateral lateral rectus muscle recessions and pulley posterior suturing on both medial rectus muscles. Five of the seven patients achieved successful results (shrinking of the distance-near disparity to <10 PD and a near or distance angle <10 PD). One patient required a bifocal to correct a postoperative esotropia at near. Followup was at least 1 year. The preoperative evaluation included a 1 hour patch test, and near measurements with +3.00 D lenses. This study had a small sample size and was nonrandomized. Also all patients had a residual exophoria or intermittent exotropia. These could potentially worsen over time.

Changes in refractive error and anterior segment parameters after isolated lateral rectus muscle recession

Noh JH, Park KH, Lee JY et alJ AAPOS 17:3;291-295

The authors evaluated the short-term effect of isolated lateral rectus muscle recession on refractive error, corneal measurements, anterior chamber depth, and volume. This was a 9-month prospective study of consecutive patients. Measurements were performed before surgery, and 1 week and 1 month after surgery. Patients with prior eye surgery were excluded, as were those with sensory strabismus or an inability to maintain reliable fixation. The study includes 24 eyes of 24 patients with a mean age of 8 years. The article states that the conjunctival incision was on the muscle insertion. One week after surgery, there were statistically significant changes in spherical equivalent, horizontal and mean keratometry, corneal astigmatism, anterior chamber volume, and both central and peripheral anterior chamber depth. At the one month followup, these changes became progressively smaller, with the exception of spherical equivalent which persisted. Flattening in corneal power in the

horizontal meridian with a myopic shift was found at 1 week after surgery. At one month after surgery there was subsequent steepening of corneal power in the vertical meridian. The authors speculate these changes are based on scleral malleability but that this would not explain the transient nature of the findings. Anterior chamber depth and volume were shallowed. Alteration of ciliary body circulation and lenticular curvature are postulated as the cause. The authors suggest longer followup to see if the myopic shift completely returned to baseline. Future studies might also look at the type of conjunctival incision to see if this plays any role in these changes. Muscle reattachment technique, and the tightness of suture tying might also play a role.

Surgical Outcomes of Medial Rectus Recession in Esotropia with Cerebral Palsy

Dae Joong Ma, Hee Kyung Yang, Jeong-Min Hwang, *Ophthalmology* April 2013;120:663-667

This is a retrospective cohort study involving 30 patients with esotropia and cerebral palsy and 60 age-matched esotropes without cerebral palsy (CP) who underwent a unilateral or bilateral medial rectus muscle resection.
The purpose of the study is to determine the outcome of a reduced amount of medial rectus muscle recession in esotropes with cerebral palsy and to compare the surgical outcomes with that of normal controls. The surgical amount of medial rectus muscle recession was reduced by 1mm per muscle in patients with CP. Research was performed at the Seoul National University College of Medicine, Seongnam, Korea.
Conclusions: The main outcome measures were success rates, surgical response, cumulative probabilities of success, and factors affecting surgical responses evaluated by generalized linear mixed models. Even with reduced amount of recession, esotropes with CP show a greater surgical response to medial rectus muscle recession than did those without cerebral palsy. The incidence of late overcorrection was significantly higher compared with that of patients without CP.

Comparing Outcome Criteria Performance in Adult Strabismus Surgery

Sarah R Hatt, David A Leske, Laura Liebermann, Jonathan M Holmes, *Ophthalmology* September 2012;119:1930-1936

This is a prospective, nonrandomized cohort study that evaluated 159 adults who underwent 171 strabismus surgeries. The purpose of the study was to evaluate the performance of motor, diplopia, health-related quality of life (HRQOL) criteria when analyzing the outcome of adult strabismus surgery.
All patients underwent a clinical assessment preoperatively and 6 weeks postoperatively including completion of the adult strabismus (HRQOL)

questionnaires. Preoperatively strabismus was classified as either diplopic (n = 117), nondiplopic (n = 38), or atypical diplopia (n = 116). Success criteria was described prior to the investigation for motor criteria: less than 10PD by simultaneous prism cover test (SPCT).
The main outcome measures: surgical success rate when applying motor, double vision, and HRQOL criteria alone or in combinations. The success rate for motor criteria alone was 90%. The success rate for diplopia/double vision criteria alone was 74% and the success rate for HRQOL was 60%. COMBINING THE ABOVE CRITERIA THE HIGHEST SUCCESS RATE WAS FOR MOTOR CRITERIA PLUS DIPLOPIA CRITERIA (67%). The lowest success rate using combined criteria methods was when combining motor, double vision, and HRQOL criteria (50%).
The authors concluded the following: 1) motor criteria alone yields the highest success rate when evaluating outcomes in adult strabismus surgery, 2) motor criteria does not fully represent the patients postoperative status,
3) combining double vision criteria with motor criteria provides for a more clinically relevant standard for judging the success of adult strabismus surgery. The main message of this study is that success cannot be defined by motor criteria alone and information obtained from the health-related quality of life questionnaire is important in evaluating the successful outcome of strabismus surgery.

Outcomes of surgery in children with early-onset exotropia

S Y Suh; M J Kim; J Choi; S-J Kim *Eye*; July 2013; 27(7): 836–40

This was a retrospective chart review o f o evaluate the sensory and motor outcomes of patients undergoing surgery for early-onset exotropia, and to compare differences between constant (XT) and intermittent exotropia (X(T)) at presentation. The medical records of 45 patients with a reported onset of exotropia before 1 year of age were reviewed. The mean age of onset of exodeviation was 9.3±3.8 months. Mean age at first visit was 3.7±2.3 years and mean age at first surgery was 4.5±2.4 years. The mean postoperative follow-up was 17.3 months (range, 6–37 months). Of the 45 patients, 67% showed alignment within ±10 PD at the final visit. Gross stereopsis on the Titmus fly was achieved in all 34 testable patients, and stereopsis of at least 60 seconds of arc was achieved in 10 patients (29%). Eleven patients were included in the XT group and 34 patients in X(T) group. Of the seven XT patients who could have sensory testing performed, 1 of 7 (14%) had stereopsis greater than 60 seconds of arc. The authors conclude that patients with a history of exotropia presenting in infancy can achieve reasonable motor and sensory outcomes after strabismus surgery. Please note that these patients may have had intermittent exotropia in infancy, not (constant) infantile exotropia, and patients with neurological disease were excluded. A critique of this study was that parental reporting was used to determine age of exodeviation onset.

Botulinum toxin injections combined with or without sodium hyaluronate in the absence of electromyography for the treatment of infantile esotropia: a pilot study

J Chen; D Deng; H Zhong ; X Lin, et al.*Eye.* March 2013;27(3):382-6.

In this prospective randomized study, the authors evaluate the feasibility and safety of botulinum toxin type A (BTA) injections with or without sodium hyaluronate for the treatment of infantile esotropia. Forty-seven patients with infantile esotropia who were ages 12-81months were randomly divided into two groups. In both groups, the patients received 2.5- 3.75 units BTA into both medial rectus muscles, injected 5-10 mm posterior to the muscle insertion. Injections were performed under general anesthesia without electromyography. In one group, the BTA was mixed with sodium hyaluronate. Alignment and complications were compared at 6 months. Alignment within 10 PD of orthotropia was present in approximately one-third of patients 6 months after injections (30.4% in the BTA+ sodium hyaluronate group *vs* 37.5% in the BTA group). Ptosis of 2-3 mm was present more often in the BTA group (20.8%) than in the BTA + sodium hyaluronate group (2.2%; P=0.008). The authors conclude that the addition of sodium hyaluronate decreases the frequency of complicated ptosis, and they speculate that sodium hyaluronate decreases diffusion of BTA to other muscles.

ADJUSTABLE SUTURES: AN UPDATE

J. Mark Engel Current Opinion in Ophthalmology Sept 2012 23(5) p. 373-376

In theory, the ability to adjust the surgical dose in the immediate postoperative period and thereby improve surgical alignment sounds appealing. However, adjustable sutures have not been embraced by a majority of surgeons, particularly in children, as they require considerable extra time, manipulation, and sedation along with the difficulty of judging postoperative drift. Recently, several studies have compared the results of patients undergoing strabismus surgery performed by the same surgeons using adjustable sutures versus nonadjustable sutures. These demonstrated slightly improved outcomes with adjustable sutures in the short term. Long term alignment has not been compared. Modifications of the adjustable suture technique allow for easier use in children. One such modification is to bury the suture underneath the conjunctiva. This allows the child to be discharged without further manipulation if no adjustment is needed, and provides easier access if adjustment is needed. The author reviews the adjustable suture technique and advocates for the more widespread use of adjustable sutures especially with complex strabismus, such as reoperations, in which surgical outcomes with nonadjustable sutures are less predictable.

Medial Rectus Recession Is as Effective as Lateral

Rectus Resection in Divergence Paralysis Esotropia

Zia Chaudhuri, Joseph L. Demer,Arch Ophthalmol. 2012;130(10):1280-1284.

Work by Archer and others have dispelled the notion that operating the lateral rectus affects distance deviations greater than operating the medial rectus. Further support for the assumption that operating the lateral and medial rectus affects distance and near measurements equally is provided in this study. In this retrospective review, the authors examine the clinical features and pre and post op measurements of patients who carry a diagnosis of divergence paralysis esotropia (DPE). Twenty-four patients with DPE underwent surgery. Six patients underwent bilateral LR resection and 2 underwent unilateral LR resection (group L), while 13 underwent bilateral MR recession and 3 underwent unilateral MR recession, with the target angle double the distance ET (group M). One of 8 patients in group L and 15 of 16 patients in group M underwent intraoperative adjustable surgery under topical anesthesia. Mean (SD) preoperative central gaze ET measured 15.0 (7.7) _ at distance and 4.1 (3.4) _ at near in group L, but 10.4 (6.8)_ at distance and 0.6 (1.7) _ at near in groupM(**P=**.15; distance, 0.003, near). Postoperatively, no patient in either group had symptomatic diplopia or convergence insufficiency in follow-up from 8.5 to 40 months. Twice the usual surgical dose of MR recession per prism diopter was required to achieve correction of the distance deviation in DPE as compared with that recommended for ET generally and also for LR resection in the same condition. The authors measured convergence amplitudes at near preoperatively so that MR recession would not be of sufficient magnitude to induce convergence insufficiency. No patient post-operatively demonstrated convergence insufficiency.

Errors in Strabismus Surgery

Elizabeth Shen, Travis Porco, Tina Rutar ,JAMA Ophthalmol. 2013;131(1):75-79

Little is known about wrong eye, wrong muscle or wrong procedure error rates in strabismus surgery. A survey was sent to AAPOS members asking to report number of surgical errors and circumstances surrounding such errors. 47% responded. 33.5% reported an error during their training or as an attending. The error rate was 1 in 2506 surgeries. Inexperienced surgeons, defined as less than 1500 career strabismus surgeries, were 5.9 times more likely to experience an error than more experienced surgeons. 35% of errors were wrong procedures, 22% were wrong muscles and 9% were wrong eyes. Contributing factors included confusion about pre-op deviation and type of procedure, torsion or other anatomical abnormalities, and distraction or inattention. A strabismus specific modification to the Universal Protocol is suggested.

Three Horizontal Muscle Surgery for Large-Angle Infantile or Presumed Infantile Esotropia

Long-term Motor Outcomes

Klio I. Chatzistefanou; Ioannis D. Ladas, Konstantinos D. Droutsas, Cryssanthi Koutsandrea, et.al. JAMA Ophthalmol. 2013;131(8):1041-1048.

In this study, the authors analyzed the short and long term motor outcomes of bimedial rectus recession and one lateral rectus resection for large angle esotropia, defined as 50 Δ or greater. The short-term (8 weeks) and long-term motor outcomes of one hundred ninety four consecutive patients with presumed congenital esotropia were studied. Motor success was defined as within +/- 10 Δ of orthotropia. The median age of patients at surgery was 2.7 years and the median follow-up was 4.5 years. The mean preoperative deviation was 68.2Δ. Seventy nine percent were successfully aligned at the 8-week postoperative evaluation and successful alignment decreased to sixty two percent at the last follow-up visit or prior to reoperation. Early versus late outcomes were compared and revealed a higher rate of late overcorrections (5.15%vs 24.1%, respectively; P = .001) but the same rate of undercorrections (15.4%vs 15.1%; P = .85). The presence of amblyopia, high hyperopia, or the total amount of millimeters of surgery did not influence surgical outcome but the presence of inferior oblique overaction and the magnitude of the preoperative esodeviation were associated with poor outcome. Delayed consecutive exotropia was more prevalent in the 50Δ to 69Δ range of preoperative esodeviation.

11. ANTERIOR SEGMENT

Two-Year Corneal Cross-Linking Results in Patients Younger Than 18 Years With Documented Progessive Keratoconus

Paolo Vinciguerra, Elena Albe', Beatrice E. Frueh, Silvia Trazza, et al. Am J Ophthal September 2012; 154(3): pg. 520-6.

Keratoconus in children can be a dramatically progressive disease in some and intraconal ring segments have failed to induce a permanent flattening of the cornea. Thirty eyes of 30 children defined to have progressive disease between the ages of 9-18 years (average age 14.2 years) were treated with corneal cross linking. Progression was defined as change of myopia or astigmatism of at least 3 diopters over previous 3 months, or a mean central K reading change of at least 1.5 diopters observed in 2 consecutive topographies during the preceding 3 months, or a mean central corneal thickness decrease of at least 5% in 3 consecutive tomographies performed in previous 3 months. The procedure was performed after epithelium was removed with an Amoils Brush. The patients were treated with riboflavin every minute for 30 minutes and then ultraviolet source emitted light at wavelength of 370 nm, lasting for 30 minutes. Postoperative results included UCVA (expressed in logarithm of the minimal angle of resolution units) improving from 0.79 +/- 0.21 to 0.58 +/- 0.10. BSCVA

improved from 0.39+/- 0.10 to 0.20 +/- 0.09. Spherical equivalent reduced by 1.57 diopters. Topography demonstrated a flattening effect of the CXL treatment. Most clinical indices improved over a 24 month time period.

Childhood Pterygium: A Descriptive Study of 19 Cases Presented to a Tertiary Eye Care Center Goyal et al. Am J Ophthal November 2012; 154 (5): pages 859-864.

This study presents the largest series of pediatric pterygium to date. Nineteen children presenting with pterygium were studied. Average age 10.63 years, and 7 of the 19 had bilateral involvement. One child was as young as 2 years. Only four cases were surgically managed, the rest were cared for with conservative therapy. Surgical management was excision with conjuctival-limbal autograft. Histopathology was performed on the surgical specimens. Results showed epithelial metaplasia and hyperplasia. Two-thirds of patients showed some refractive astigmatism. No direct evidence was found for the etiology of the pterygium through histopathology or history. This study demonstrates that pterygium is a rare but definitive possibility in children under 16 years of age.

Pasteurella canis and Granulicatella adiacens Conjunctivitis Outbreak Resistant to Empirical Treatment in a Child Welfare Agency Melike Balikoglu-Yilmaz, Tolga Yilmaz, Ayse Banu Esen, Kaya N. Engin, Muhittin Taskapili, J Pediatr Opthalmol Strabismus 2012; 49:314-319 Sept/Oct

This article discusses 28 eyes of 14 children with conjunctivitis resistant to empirical treatment. The article stresses that use of empirical antibiotics is not the best approach. The approach the authors recommend is to take conjunctival cultures and switch antibiotics accordingly. The message of the article is worthwhile – that use of specific antibiotics is better than empirical antibiotics to reduce the risk of resistance but it speaks of very specific pathogens in a very specific environment which is one of its limitations I believe.

Deep Anterior Lamellar Keratoplasty in Children Jatin N. Ashar, Shivani Pahuja, Muralidhar Ramappa, Pravin K. Vaddavalli, et al. Am J Ophthal January 2013, 155 (1)

This retrospective study evaluated the surgical outcome of deep anterior lamellar keratoplasty in 26 children who underwent the procedure for keratoconus, keloid, keratitis, scar, chemical injury and dermoid. Recommendation for this procedure over penetrating keratoplasty is based upon less rejection and less endothelial cell loss. Results demonstrated that 75% of grafts remained clear for keratoconus and achieved visual acuities of 20/80 or better. Grafts remained clear in 18 of 26 eyes. Visual acuity overall improved with 10 eyes achieving

20/40 or better postoperatively; preoperatively, no patients saw better than 20/60. Deep anterior lamellar keratoplasty seems to be a viable alternative to PK.

Endothelial keratoplasty without Descemet's stripping in congenital hereditary endothelial dystrophy

Ashar JN, Ramappa M and Chaurasia S J AAPOS 2013;17:22-24

DSEK is an alternative to PK for the management of CHED. However in children DSEK can be difficult because of limited visualization from corneal edema, difficult positioning of the lenticule, or difficult stripping of Descemet's membrane. It is possible to perform endothelial keratoplasty without stripping Descemet's membrane. The authors compare outcomes of non-Descemet's stripping endothelial keratoplasty (nDSEK) (n=3) with standard DSEK (n=3) in patients with CHED. This was a retrospective interventional case series in children with corneal edema since birth. The average age in both groups was 5.6 years. One patient in the nDSEK group presented with a partially detached graft 1 week postoperatively, which was successfully secured after a repeat air injection. The final outcome was a clear cornea in all six patients. This study had a limited number of patients but showed comparable results with the two surgical techniques.

Intraobserver reliability of contact pachymetry in children

Weise KK, Kaminski B, Melia M, et al J AAPOS 2013;17:144-148

This study evaluated intraobserver reliability of a handheld contact measurement of central corneal thickness. The cohort of children was normal and ranged in age from newborn to 17 years. There were 1933 subjects (3494 eyes) with a mean age of 7.5 years. The mean CCT was 559 ≒m with a mean absolute value of the test-retest difference of 6.1 ≒m. In-office measurements were more variable than those taken under anesthesia, but this difference did not persist, after adjusting for the difference in CCT magnitude in the two groups. More younger children, (with thinner OCTs) were measured under anesthesia. As the CCT increased, the test-retest difference increased, as did the coefficient of repeatability. There was no evidence of an effect on testing order for the 3 measurements taken. Age, sex, race, and examination setting did not affect the magnitude of test-retest differences, after adjusting for laterality. Thinner cornea measurements were more reliable, probably because of off-center measurements. Off-center measurements are more likely to be thicker, causing greater variability. Thicker measured CCT values had larger test-retest differences than thinner measured CCT values. The authors recommend disregarding measurements when the reported standard deviation is >5≒m. Also when the first measurement is >575≒m, a second measurement should be taken.

Corneal thickness measured by Scheimpflug imaging in children with Down syndrome

Aslan L, Aslankurt M, Yuksel E, et al. J AAPOS 2013;17:149-152

Corneal changes are more common in Down syndrome patients. Accurate corneal thickness measurements can be obtained with the Pentacam Scheimflug imaging system. This system captures dozens of images quickly. The entire cornea is measured. This system was used to measure full corneal thickness, including central and paracentral areas in healthy individuals and Down syndrome patients. Prospective evaluation of 31 Down syndrome and healthy age-matched controls was performed. Four Down syndrome patients were excluded because of poor image results. CCT values were thinner in Down syndrome patients. CCT values of <500≒m (59.2%) and <450≒m (18.5%) in Down syndrome patients were obtained, versus 37.8% and 5.4% respectively in controls. Thinnest point (TP) values were also less in the Down syndrome patients (62.9%/21.1% versus 43.2%/10.8%). Down syndrome patients had CCT measurements, which averaged 45 ≒m less and TP values which averaged 51≒m less than controls. Mean corneal volume was less as well (56.2 ≒m^3 versus 61.3 ≒m^3). Pentacam software analysis revealed subclinical keratoconus in 21.1% of Down syndrome eyes versus 1.35% of eyes in the control group. Down syndrome eyes showed the TP most commonly centrally (52%) and inferotemporally (38%). CCT measurements are important because they can affect IOP measurements. Not only are CCT measurements often abnormal in Down syndrome patients, but these patients are at higher risk for keratoconus. Early detection can lead to potential earlier cross-linkage treatment. This rapid non-contact method of corneal evaluation may have utility, especially in low cooperation individuals, such as Down syndrome patients.

Clinical Correlations of Dry Eye Syndrome and Allergic Conjunctivitis in Korean Children

Tae hyung Kim, Nam ju Moon, J Pediatr Ophthalmol Strabismus 2013; 50:124-127 (March/April)

Children with dry eyes and allergic conjunctivitis were investigated their clinical patterns were evaluated. The study found that children with allergic conjunctivitis had shorter tear breakup time, and those with multiple allergens had shoter times than those with single allergens. Children do not often complain of dry eyes so its important to remember that this can occur and treat children appropriately.

Pathologic Epithelial and Anterior Corneal Nerve Morphology in Early-Stage Congenital Aniridic Keratopathy

Ulla Eden, Per Fagerholm , Reza Danyali , Neil Lagali, *Ophthalmology* September 2012;119:1803-1810

This study comes from the Department of Ophthalmology, Institute for Clinical Experimental Medicine, Faculty Health Sciences, Linkoping University, Linkoping, Sweden.
This is a prospective observational comparative case series of 16 eyes in 16 subjects with congenital aniridia keratopathy and clear central cornea. This study also studied 6 eyes from 6 healthy controls (unaffected relatives). 9 of the 16 eyes with aniridia came from 5 families with documented family history of aniridia. The purpose of the study was to document the clinical and morphologic corneal findings in the early stages of congenital aniridia keratopathy in Swedish families.
Conclusions: Early-stage aniridia keratopathy is characterized by the development of focal opacities in the basal epithelium, altered sub-basal nerves, infiltration of the central epithelium of dendritic cells, tear film instability, and increased corneal thickness and degradation of limbal cellular architecture. These findings characterize the pathogenesis of aniridic keratopathy.
Detailed ophthalmic examinations including best corrected visual acuity, tear film production, tear break-up time, ultrasound pachymetry, slit lamp biomicroscopy, and laser scanning in vivo confocal microscopy (IVCM) were performed.
COMMENT: The article has excellent photographs of dendritic cell analysis determined by in vivo confocal microscopy. The photograph in figure 7 documents the prominent sub-basal nerve whorl pattern in a case of aniridia. The photographs in figure 8 show “in vivo images of the inferior limbal palisades region in healthy controls compared to patients with aniridia.” The difference is quite striking.

Cellular Changes of the Corneal Epithelium and Stroma in Herpes simplex Keratitis:An In Vivo Confocal Microscopy Study

Pedram Hamrah , Afsun Sahin , Mohammad H Dastjerdi , et al
Ophthalmology September 2012;119:1791-1797

This is a prospective, cross-sectional, controlled, single-center study performed in Barcelona, Spain. This study included 31 eyes in affected patients with unilateral herpes simplex keratitis as well as evaluation of the contralateral, non-clinically affected eye were compared with normal controls (n=15).
The main outcome measures were changes in morphologic features and density of the superficial and basal epithelial cells as well as stromal keratocytes as determined by 2 masked observers. Changes were correlated to corneal sensation, number of nerves, and total length of nerves. In vivo confocal microscopy and corneal esthesiometry of the central cornea were performed in all patients in controls.
Conclusion: In vivo confocal microscopy reveals profound herpes simplex keratitis induced changes in the superficial epithelium as demonstrated by increase in cell size, decrease in cell density and squamous metaplasia. This

study also demonstrated these changes correlate strongly with changes in corneal innervation.
COMMENT: Figure 1, page 1794, show images of in vivo confocal microscopy images taken in patients with herpes simplex keratitis. There was comparison of a normal cornea and anterior stroma found in the photographed eye. A contrast in the changes of cell size, cell density, and squamous metaplasia is quite clear from these photographs.

Corneal Cross-Linking as a Treatment for Keratoconus Four-Year Morphologic and Clinical Outcomes with Respect to Patient Age

Riccardo Vinciguerra, Mario R. Romano, Fabrizio I. Camesasca, Paolo Vinciguerra, *Ophthalmology* May 2013;120:908-916

This is a retrospective, single-center, nonrandomized clinical study that takes place in Italy. The purpose of the study is to report the four-year outcome of corneal cross-linking (CXL) for progressive keratoconus in a population of different age groups. Four hundred consecutive eyes treated with corneal cross-linking for progressive keratoconus in April 2006 and April 2010 are included in this study.
Intervention: After removal of corneal epithelium, the cornea was irrigated for 30 minutes with a solution of 0.1% riboflavin and 20% dextran, followed by irradiation with an ultraviolet A light for 30 minutes.
Comparative analysis included 400 eyes of 301 patients. The patients studied were stratified in 3 groups; Group A: patients younger than 18 years of age, group B: patients 18-29 years of age, group C patients 30-39 years of age, group D patients greater than 40 years.
Conclusions: Outcomes stratified by age indicate the efficacy of corneal cross-linking in stabilizing the progression of ecstatic disease and keratoconus in all age groups. The results indicated better functional and morphologic results in the population between 18 and 39 years of age.
Comment: Today, most corneal cross-linking research has been done in Europe and South America. Randomized trials are being developed in the United States.

Evaluation of Moxifloxacin 0.5% in Treatment of Nonperforated Bacterial Corneal Ulcers:A Randomized Controlled Trial

Namrata Sharma, Manik Goel, Shubha Bansal, Rasik B. Vajpayee, *Ophthalmology* June 2013;120:1173-1178

This is a randomized, controlled, equivalence clinical trial performed primarily in New Delhi, India. The purpose of the study is to compare the equivalence of Moxifloxacin 0.5% with a combination of fortified cefazolin sodium 5% and

Tobramycin sulfate 1.3% eye drops in the treatment of moderate bacterial corneal ulcers.
Microbiologically proven cases of bacterial corneal ulcers were enrolled in the study and were allocated randomly to 1 of 2 treatment groups. Group A was given a combination of therapy of fortified cefazolin and tobramycin. Group B was given monotherapy of moxifloxacin 0.5%. The primary outcome variable for this study was percentage of the ulcers healed at 3 months. The secondary outcome variables were best-corrected visual acuity and resolution of infiltrates. Conclusions: Corneal healing using 0.5% moxifloxacin monotherapy is equivalent to that of combination therapy using fortified cefazolin and Tobramycin in the treatment of moderate bacterial corneal ulcers.

Long-term clinical course and visual outcome associated with Peters' anomaly

J W Chang; J H Kim; S-J Kim; Y S Y *Eye.* September 2012;26(9):1237-1242

This was a retrospective review of 90 eyes in 65 patients from Seoul National University Children's Hospital with Peters anomaly who had more than 5 years of follow up. Eyes were managed medically (mydriatics and amblyopia treatment), surgically, or observed. Final visual outcomes were presented with respect to disease severity and treatment modalities. Severe disease was defined as corneal opacity >50% of the cornea, corneal opacity with lenticular adhesion, or corneal opacity + other ophthalmic abnormality such as microphthalmos. Mild disease was defined as corneal opacity <50% and normal corneal diameter. 61% of patients had bilateral disease and 39% had unilateral disease. The mean final visual acuity of all Peters eyes was 20/2240. Not surprisingly, the visual outcomes of the eyes with mild disease in each treatment group were significantly better than the eyes with severe disease. The visual acuities in the medical group were better than in the surgical or observation group, but the surgical and observation groups included more severe cases of Peters anomaly. Of the six patients who underwent trabeculectomy, all six had final visual acuity of no light perception. There was a 29% rate of systemic anomalies, which occurred in both bilateral and unilateral Peters cases. The most common systemic anomalies were developmental delay and cardiopulmonary anomalies. Overall, this study gives a sobering picture of the visual outcome in Peters anomaly cases, especially among those with severe disease.

Transepithelial corneal collagen crosslinking for progressive keratoconus in a pediatric age group

Abdelrahman G. Salman *J Cataract Refract Surg.* August 2013;39(8):1164-70

The purpose of the study was to evaluate the effectiveness of transepithelial corneal collagen crosslinking (CXL) in children with keratoconus. In this prospective study, the right eye was treated with transepithelial corneal collagen crosslinking whereas the comparison left eye was treated with topical lubricants and standard care. Transepithelial riboflavin administration spares the patient corneal epithelial debridement, but requires a special riboflavin formulation to penetrate into the corneal stroma. After riboflavin application, the treated eye was exposed to UVA light for 30 minutes. Twenty-two patients were enrolled, ranging in age from 13 to 18 years, with a mean age of 15.7 years ± 2.1 (SD). Eighteen of the 22 patients were boys. Five patients required conscious sedation for treatment. Prior to the study, the right and left eyes were well matched in terms of visual acuity and keratoconus parameters. Mean follow up was 12.0 +/- 3.4 months. After transepithelial CXL, the mean uncorrected distance visual acuity improved by 2.7 lines (from 0.95 ± 0.34 logMAR to 0.68 ± 0.45 logMAR) (P<.05). There was no improvement in the control group (P>.05). The mean simulated keratometry (K) decreased by a mean of 2.03 diopters (D); P<.05), whereas the simulated K increased by a mean of 0.59 D (P>.05) in the control group. Preliminary results of transepithelial CXL in children with keratoconus were encouraging, with no evidence of progression of keratoconus over 12 months in the treated eyes.

Management of pediatric keratoconus – Evolving role of corneal collagen cross-linking: An update

Vardhaman P Kankariya, George D Kymionis, Vasilios F Diakonis, Sonia H Yoo

Indian Journal of Ophthalmology Aug 2013 Vol. 61 No. 8

The above article is a review article detailing the role of corneal collagen cross-linking.

Table 1: Chief characteristics of published studies regarding outcomes of corneal collagen cross linking in pediatric keratoconus

Authors (year)	Subjects (eyes)	Age range (years)	Design	Documented progression	CXL technique	Outcomes	Follow-up
Soeters *et al.* (2011)[11]	4 (5)	10-16	Case series	Yes	Standard protocol	4 eyes were stabilized, 1 eye ended up in PKP	1 year
Arora *et al.* (2012)[12]	15 (15)	10-15	Prospective	No (other eye with advanced keratoconus)	Standard protocol	Stabilization	1 year
Bakshi *et al.* (2012)[13]	9 (9)	11-17	Retrospective	Yes	Standard protocol	Stabilization	2 years
Vinciguerra *et al.* (2012)[14]	40 (40)	9-18	Prospective	Yes	Standard protocol	Stabilization	2 years
Caporossi *et al.* (2012)[15]	77(152)	10-18	Prospective	Yes	Standard protocol	Stabilization	3 years
Chatzis *et al.* (2012)[16]	52 (59)	9-19	Retrospective	Yes	Standard protocol	Initial stabilization, and late progression	3 years
Magli *et al.* (2013)[17]	29 (37)	12-18	Retrospective, comparative	Yes	Epi on and Epi off (2 groups)	Stabilization for both groups	1 year
Buzzonetti *et al.* (2012)[18]	13 (13)	8-18	Prospective	No	Transepithelial CXL	No stabilization	18 months
Zotta *et al.* (2012)[19]	4 (8)	11-16	Case series	Yes	Standard protocol	Stabilization	3 years

With limited evidence CXL can be considered in children with progressive keratoconus. Standard epithelium of CXL protocol should be followed. Parents should be informed about the off-label nature, possibility of short lasting effect and need for re-treatment especially in very aggressive forms.

A NEW APPROACH TO THE CLASSIFICATION OF NEONATAL CORNEAL OPACITIES

Ken K. Nishcal Current Opinion in Ophthalmology Sept. 2012 23(5) p.344-354

Review of corneal opacities with presentation of a new classification system. Neonatal corneal opacification (NCO) describes the loss of corneal transparency at or soon after birth (>4 weeks). Traditionally, the differential diagnosis for NCO is remembered by the pneumonic 'S.T.U.M.P.E.D.' (Sclerocornea, Tears in Descemet's membrane, Ulcers, Metabolic, Peters, Endothelial dystrophy, and Dermoid). Although this serves as an aid for differential diagnosis, it does not help define phenotype accurately.The literature is filled with terminology that is misleading and is a hindrance to selecting appropriate treatment and accurate genotype-phenotype correlation. The designation 'sclerocornea' is unhelpful when alluding to total NCO. The term Peters Anomaly has also become a 'waste paper basket' diagnosis for anterior segment developmental anomalies. A new classification of NCO is suggested by the author, which allows a better understanding of etiology and the likely prognosis of therapeutic intervention. The author classifies NCOs as either primary or secondary. Primary NCOs are always congenital. Secondary NCOs are further subdivided into congenital and acquired. The decision whether to intervene surgically, and procedure selection is best determined with a clear understanding of NCO causation.

CURRENT OPHTHALMOLOGIC TREATMENT STRATEGIES FOR ACUTE AND CHRONIC STEVENS JOHNSON SYNDROME AND TOXIC EPIDERMAL NECROLYSIS

Jessica B. Ciralsky et al Current Opinion in Ophthalmology July 2013 24(4) p.321-328

Review of the ophthalmologic treatments for acute Stevens-Johnson syndrome (SJS) as well as the emerging treatment options for patients with chronic, severe ocular surface damage from the disease. Amniotic membrane transplantation (AMT) applied to the eyes and eyelids in the acute phase, preferably within the first week, of SJS can prevent scarring and visual problems that characterize the chronic phase of the disease. The severity of ocular inflammation in the acute phase does not always correlate with severity of skin and systemic involvement. It is crucial that all patients with SJS be urgently evaluated by an ophthalmologist familiar with the current management of the disease, and offered the option of AMT. Although challenging, the severe, chronic ocular problems of SJS can be at least be partially alleviated with autologous serum drops, mucous membrane grafting to replace scarred tarsal conjunctiva, specialized contact lenses (PROSE), conjunctival replacement surgery (COMET), limbal stem cell transplantation and kerotoprostheses. Emerging treatments offer increased hope for those who have already suffered damage from SJS, but emphasis on preventing damage in the acute phase is most crucial.

UPDATE ON MANAGEMENT OF HERPES KERATITIS IN CHILDREN

Karen Revere et al Current Opinion in Ophthalmology July 2013 p. 336-342

Summary of the articles published in 2012 pertaining to the clinical presentation, diagnosis, and treatment of herpetic keratitis, with specific attention to the pediatric population. Herpetic keratitis has higher recurrence rate in children than adults. Recurrences are more likely to occur as stromal disease. Vision loss in children is from corneal scarring leading to deprivation and/or refractive amblyopia. Acyclovir is safe and well tolerated in children, and preferable to difficult and toxic eye-drop regimens. Immunochromatographic assay is an effective diagnostic tool to confirm diagnosis of herpes simplex virus-1 (HSV) in corneal scrapings with high specificity but poorer sensitivity. Real time PCR can be employed to follow changes in HSV viral load in patients where resistance is suspected. Delays in treatment related to misdiagnosis, as well as resistance to current antiviral therapeutics, can lead to visually devastating corneal opacification. In the pediatric population, already at risk for amblyopia, this can be especially damaging. Children are unique with regards to the way in which they manifest herpetic keratitis, making rapid diagnosis and treatment even more challenging.

Conjunctival Papilloma:Features and Outcomes Based on Age at Initial Examination

Swathi Kaliki, Sruthi Arepalli, Carol L. Shields, Kendra Klein, et.al. JAMA Ophthalmol. 2013;131(5):585-593.

Conjunctival papilloma is a benign epithelial tumor occurring in both children and adults with varying clinical features and outcomes. This study evaluated the clinical features, treatment, and outcomes in patients with conjunctival papilloma based on age at initial examination. Ten children and adolescents and 63 adults with conjunctival papilloma. Interventions included excisional biopsy, cryotherapy, oral cimetidine, topical or injection interferon alfa-2b, and photodynamic therapy. Conjunctival papillomas are larger and more likely to be multiple in children and adolescents than in adults. Excisional biopsy and cryotherapy with or without adjuvant oral cimetidine and/or topical interferon alfa-2b provide satisfactory tumor control. Papilloma recurrence is more common in children and adolescents than in adults.

12. CATARACT

Juvenile Cataract Morphology in 3 Siblings Not Yet Diagnosed with Cerebrotendinous Xanthomatosis

Arif O. Khan, Mohammed A. Aldahmesh, Jawaher Y. Mohamed, Fowzan S. Alkuraya, *Ophthalmology* May 2013;120:956-960

This is a prospective case series involving 4 siblings and their 2 parents who are first cousins. The purpose of the study is to evaluate the treatment with oral chenodeoxycholic acid as a prevention of cataracts and neurologic deterioration associated with cerebrotendinous xanthomatosis.
Children with clinically undiagnosed cerebrotendinous xanthomatosis (CTX) frequently demonstrate early lenticular changes heralding the disease. This study highlights the morphology of lens opacities in a family with genetically confirmed disease. CYP27A1 is a candidate gene associated with cerebrotendinous xanthomatosis.
Conclusions: An unusual pattern of fleck lenticular deposits along the Y-sutures characterized the lenticular findings in this condition. Findings are best seen on page 958, volume 120, number 5, May 2013. FLECK OPACITIES AND POSTERIOR CAPSULAR CATARACT OPACITY CAN BE APPRECIATED IN MANY OF THESE PATIENTS. FURTHERMORE, THE PUNCTATE OPACITIES TEND TO FOLLOW ALONG THE Y-SUTURE.
The authors emphasize that such juvenile lenticular findings should raise suspicion of this treatable metabolic storage disease especially when in the context of recurrent diarrhea during early childhood. Asymptomatic fleck-like opacities at or near the anterior Y-suture may be assigned a carrier status.

Comment: This is a rare disorder not frequently seen in the United States. It is caused by a recessive CYP27A1 mutation that is characterized by abnormal deposition of cholestanol and cholesterol in multiple tissues including the lens and brain.

13. CATARACT SURGERY

Congenital cataract surgery with intracameral triamcinolone: Pre- and postoperative central corneal thickness and intraocular pressure.

Ventura MC, Ventura BV, Ventura CV, et al.J AAPOS 2012;16:441-444

Children with congenital cataracts who underwent cataract surgery with insertion of a primary intraocular lens at <2 years of age received intracameral triamcinolone at the end of the procedure to control post-operative inflammation. This retrospective case series assessed IOP and central corneal thickness (CCT). Measurements were taken within the first 4 minutes of general anesthesia, and then 2 months postoperatively and 12 months postoperatively. 53 eyes of 34 children were included. The mean age at the time of surgery was 11 months. No patient experienced a secondary obstruction of the optical zone or needed additional intervention. Pre- and postoperative IOP values were not significantly different. No patient developed ocular hypertension or glaucoma. Two patients had >5mm Hg elevation in intraocular pressure at the 2 month exam, but this resolved by the 12 month examination. CCT mean values were not significantly different over the course of the study. The study is somewhat limited by its retrospective nature. Young children have a greater inflammatory reaction to intraocular surgery than adults. Evidence supporting the lack of effect on IOP or CCT of steroid injection into the anterior chamber is important in considering this regimen at the end of cataract surgery in younger children.

Relationship between the timing of cataract surgery and development of nystagmus in patients with bilateral infantile cataracts

Young MP, Heidary G, and VanderVeen DK. J AAPOS 2012;16:554-557

The purpose of this study was to evaluate whether the timing of cataract surgery in patients with bilateral infantile cataracts influences the development of nystagmus. The authors also look at the effect of nystagmus on visual acuity outcomes. A retrospective chart review of patients over an 18 year period was performed. Mean follow-up was 92 months. 38% of the patients who underwent cataract removal before 8 weeks of life (10/26) had manifest nystagmus at their last follow-up versus 50% (15/30) who underwent surgery after 8 weeks of life. There was no statistical difference. 39/56 (73%) patients were able to complete Snellen visual acuity testing. 20/40 or better acuity was seen in 33 of 39 (85%)

patients. There was no statistically significant differencein final acuity based on timing of surgical intervention. There was also no significant difference in acuity testing results based on the presence or absence of preoperative nystagmus. However there was a significant difference in final acuity based on the presence of manifest nystagmus at that visit. A significant confounding factor in this study is the possibility that some patients had developmental cataracts (rather than congenital) which would have a different effect on the visual system.

Accuracy of Intraocular Lens Power Calculation Formulae in Children Less Than Two Years

Ramesh Kekunnaya, Amit Gupta, Virender Sachdeva, Harsha L. Rao et al. Am J Ophthal. July 2012; 154(1): pg. 13-19.

This was a retrospective study looking at 128 eyes of 84 children who underwent cataract extraction with primary lens implantation less than 2 years of age. Fifteen eyes had axial length less than 18 mm. Across the board, predictive errors were large (0-14.3 diopters) with all formulas. SRK II formula showed the minimum prediction error (+2.27+/- 1.69 diopters). 57% of eyes were within 2 diopters of absolute prediction error for the SRK II formula. The prediction error with the SRK II was not influenced by age, keratometry, or axial length. Axial length did adversely affect the Holladay and Hoffer Q formulas. This study suggests that, unlike adults with short axial lengths undergoing IOL calculation, the SRK II may be a better formula for children less than 2 years of age undergoing lens implantation. This pediatric age group remains an extremely difficult group to anticipate lens implantation. Even this formula, however, had an accuracy of only 50% within 1 diopter.

Central Corneal Thickness and Intraocular Pressure in Children Undergoing Congenital Cataract Surgery: A Prospective, Longitudinal Study

Graziela Massa Resende, Alvaro P.C. Lupinacci, Carlos Eduardo Leite Arieta, Vital P. Costa. Br J Ophthal 2012; 96: 1190-1194.

This study prospectively analyzed the central corneal thickness and intraocular pressure in 37 eyes of 26 children undergoing cataract extraction. Twenty-two eyes were pseudophakic and 15 eyes were aphakic. Mean follow up was 30.5 months. Among the 22 pseudophakic eyes, only 4 were younger than 2 years of age. Among the aphakic group, 13 eyes were younger than 2 years of age. Mean IOP did significantly increase from 12 mm Hg to 13.89 mm Hg; no statistical difference existed between aphakic or pseudophakic eyes. Central corneal thickness (CCT) also showed a significant increase in the aphakic group from baseline and inversely correlated with age at the time of surgery. IOP change did not correlate with age of surgery. The CCT change could be directly correlated with the IOP change in the aphakic group; this fact could be explained by the higher CCT. This series suggests that age may be the most important predictor of CCT changes postoperatively.

Refractive Changes After Pediatric Intraocular Lens Implantation in Hong Kong Children

Jason C.S. Yam, Patrick K.W.Wu, Simon T.C. Ko, Ursula S.F. Wong, Clement W.N. Chan, J Pediatr Opthalmol Strabismus 2012; 49:308-313 Sept/Oct

Following cataract surgery with intraocular lens (IOL) implantation, refractive errors continue to occur in children. The challenge is always to find a balance and determine the amount of refractive error a child will have so that one is able to closely approximate emmetropia. This article reports refractive changes after cataract surgery and IOL implantation in Hong Kong Chinese children. Looking at children who had cataract surgery prior to age 18 and at least 2 years of follow up, the authors were able to chart refractive errors in the different age ranges. They found that post operative myopic changes are greatest in younger children and continue until 8 years of age. They also found that the refractive development of the pseudophakic eyes was not significantly different from the fellow phakic eyes.

Reanalysis of refractive growth in pediatric pseudophakia and aphakia

Whitmer S, Xu A, and McClatchey S J AAPOS 2013;17:153-157

The logarithmic model of the rate of refractive growth for aphakic children is flawed at the youngest ages. A newer model (RRG2) was developed with 'adjusted age' to account for in utero eye growth. RRG2 is based on refractions at the spectacle plane. In highly hyperopic young children, this produces artificially large differences between an aphakic refraction at the natural lens plane versus the spectacle plane. The authors have developed a new model the RRG3 based on an aphakic refraction at the natural lens plane. This was a retrospective observational case series to develop and test the model. The primary outcome measures were the mean values of rate of refractive growth for pseudophakic and aphakic eyes calculated with the RRG3. Factors that might affect the RRG3 were also evaluated.
Sevetny-eight pseudophakic eyes and 70 aphakic eyes were included. There was a mean follow-up time of 9.5 years. RRG3 values did not differ for pseudophakic or aphakic patients, when comparing surgery before or after 6 months of age. This stands in contradistinction to when the RRG or RRG2 model is applied. Therefore all further analysis grouped ages. The mean RRG3 value for pseudophakic and aphakic eyes was -13 +/-6 D and -16+/-10 D respectively. Other factors such as sex, presence of glaucoma, and bilateral or unilateral surgery were not found to contribute significantly. LogMAR best corrected visual acuity, presence of an IOL, and calculated initial adjusted aphakic refraction had a mild effect on the RRG3. The RRG3 eliminates the optical effect of vertex and distance therefore eliminates previously observed age-related differences when

using the RRG or RRG2. This study was retrospective and not randomized. All children in this study had surgery at or after 3 months of age so the data may not be applicable to earlier surgeries. A large, prospective, randomized study could help validate these results.

Glaucoma after pediatric cataract surgery in a population with limited access to care

Baden C, Shija F, Lewallen S et al J AAPOS 2013;17:158-162

This study evaluates the incidence of postoperative glaucoma after pediatric cataract surgery in a resource-constrained setting in Africa. This paper was a retrospective three-year chart review. Inclusion criteria, were met by 136 patients (222 eyes). Primary IOLs were placed in 211/222 but data was not available on the remaining 11 eyes. Mean diagnosis of the cataract was at 6 months of age. Surgery was performed at a median age of 6.6 years. Median follow-up was 7.8 months. The short follow-up may lower secondary glaucoma estimates. Eyes operated on prior to 9 months of age were more likely be microphthalmic, less likely to have received an IOL, more likely to have bilateral cataracts, more likely to have preoperative nystagmus, and have received longer follow-up. Six eyes of 5 patients developed glaucoma during follow-up. The study cohort had a 2 cases per 100 operated annual glaucoma incidence rate. Primary IOL implantation was associated with a lower risk of glaucoma development, whereas longer followup was associated with a higher risk. Long-term followup rates were improved if the patient presented for their one-year followup and if spectacles were prescribed at the most recent visit. Only ¼ of patients were still attending follow-up by 3 years post surgery. The low rate of glaucoma in this study could be due to underdiagnosis, older average age at the time of surgery, patient loss to follow-up, or small sample size of glaucoma cases, preventing the use of multiple logistic regression analysis. The small number of glaucoma cases prevents comment on risk factors. Efforts must be made in the future to increase followup rates.

Visual Outcome and Changes in Corneal Endothelial Cell Density Following Aphakic Iris-Fixated Intraocular Lens Implantation in Pediatric Eyes With Subluxated Lenses

Sorath Noorani Siddiqui, Ayesha Khan, J Pediatr Ophthalmol Strabismus 2013; 50:178-182 (March/April)

The study determines the safely of artisan lenses in children with subluxed lenses. A prospective study followed children who had an artisan lens placed in their eye after lensectomy for subluxed lenses. The children were followed for approximately a year and the lens was found to have minimal side effects. The corneal endothelial density was not adversely affected in the first year following lens implantation and the vision was good postoperatively. The article does state that (an it is obvious) that one year is not a sufficient amount of time to determine efficacy of the lens as this lens will be in the child's eye for a very long time, so

although the concept is interesting and definitely worth studying, more time is needed to determine the safely of these lenses.

Long-term Risk of Glaucoma After Congenital Cataract Surgery

Scott Lambert, Amitabh Purohit, Hillary M. Superak, Michael J. Lynn, et al. Am J of Ophthal August 2013; 156(2): pg. 355-360.

This retrospective study analyzed the occurrence of glaucoma after cataract extraction on children less than 7 months of age. Sixty-two eyes of 37 children who had undergone surgery with a minimum of 3 years follow up were reviewed. Six eyes had IOL implantation whild 56 were left aphakic. Nine eyes developed open angle glaucoma. The probability of developing glaucoma was estimated to be 19.5% by 10 years. If patients classified as glaucoma suspect were included in the statistics, 2/3 of patients would have developed disease. It is important to monitor children in this young age group for a prolonged time for glaucoma development.

Posterior Iris-claw Aphakic Intraocular Lens Implantation in Children

Johannes Gonnermann, Necip Torun, Matthias Klamann, Ann-Karina Maier, et al. Am J Ophthal August 2013; 156(2): pg. 382-386.

This study reviewed the implantation of iris-claw lens implantation (Artisan PCIOL, Verisyse VRS54) in 7 eyes in 4 children who were aphakic without adequate posterior capsular support as a viable alternative to angle supported IOL, scleral fixated PCIOL, iris fixated PCIOL, or intrascleral fixated PCIOL. Mean follow up was 31 months. There was mean endothelial cell loss of 6.4% (3013 cells/mm3 to 2831 cells/mm3). One dislocation occurred. Six of the 7 eyes achieved BCVA of 0.1 logMAR or better. Postoperative IOP was similar to preoperative measurements. The authors suggest that iris-claw IOL is a viable option for lens implantation in children without capsular support.

Corneal Endothelial Cell Characteristics After Pediatric Cataract Surgery

Aparna Ramasubramanian, Iason Mantagos, Deborah K. Vanderveen,
J Pediatr Ophthalmol Strabismus 2013; 50:251-254 (July/August)

The study described corneal endothelial cell characteristics after pediatric cataract surgery. Using specular microscopy, endothelial cell features were correlated with clinical and surgical features. The authors concluded that there is endothelial cell loss along with polymegathism and pleomorphism in children who underwent cataract surgery and that this is an important aspect of the ophthalmic examination when the child is old enough to performa specular microscopy to determine corneal changes and if there is a risk for corneal decompensation.

One-Year Strabismus Outcomes in the Infant Aphakia Treatment Study

Erick D. Bothun, Julia Cleveland, Stephen P. Christiansen, Scott R. Lambert, *Ophthalmology* June 2013;120:1227-1231

This is a secondary outcome analysis in a prospective, randomized clinical trial known as the Infant Aphakia Treatment Study (IATS). All participants in this study were randomized in multiple institutions. The purpose of the study was to compare treatment of aphakia with primary intraocular lens or contact lenses in 114 infants with unilateral congenital cataract.
The main outcome measures included a portion of patients in whom strabismus developed during the first 12 months of follow-up comparing the intraocular lens group versus the contact lens group.
Conclusions: Intraocular lens placement does not prevent the early development of strabismus after congenital cataract surgery. However, strabismus was less likely to develop in infants whose cataract was removed at an early age.

Combined pars plana and limbal approach for removal of congenital cataracts

Xin Liu; Yi Luo; Xingtao Zhou; Lin Jiang, et al. *Journal of Cataract & Refractive Surgery*. December 2012; 38(12):2066-70

The authors describe the removal of bilateral congenital cataracts in a five month old boy with 25-gauge vitrectomy instrumentation. 25-gauge trochars were used to make the incisions. The infusion cannula was placed at the limbus, entering the anterior chamber, and the vitrector was placed through a transconjunctival sclerotomy 2 mm posterior to the limbus. An anterior capsulotomy (after entering the lens at the equator), lens removal, posterior capsulotomy and anterior vitrectomy were performed with the pars plana vitrector. No sutures were placed.

Uncorrected visual acuity in children with monofocal pseudophakia

Bharti R. Nihalani; Deborah K. VanderVeen *J Cataract Refract Surg*. March 2013;39(3):419-24

In this single institution retrospective chart review, the authors report on uncorrected distance and near visual acuity in pediatric eyes treated with primary monofocal intraocular lens (IOL) implantation. Records of children older than 5 years who had uneventful monofocal IOL implantation targeted for emmetropia within ±1.00 diopter (D) spherical equivalent were reviewed. Eyes with secondary IOL placement and sulcus-fixated IOLs were excluded. The main outcome measure was uncorrected visual acuity at distance and near. Good visual acuity was defined as 20/40 or better. Forty-one eyes of 25 children had uncorrected

distance and near visual acuity recorded in the early postoperative period. The mean age was 11.2 years ± 3.6 (SD). Twenty eyes (49%) had good visual acuity at distance and near, 11 had good visual acuity at distance only, 6 had good visual acuity at near only, and 4 had worse than 20/40 at distance and near. In children with bilateral pseudophakia, 12 (75%) of 16 had 20/40 or better uncorrected acuity at distance and near, with approximately symmetric refractive error (0.2 +/- 1.2 D), meaning that monovision was neither targeted nor present.

In summary, monofocal IOL placement resulted in good uncorrected distance and near visual acuity in almost 50% of pediatric eyes and in 75% of patients with bilateral pseudophakia. This study brings into question whether multifocal IOLs are truly needed to obtain good uncorrected distance and near vision in young eyes. Young eyes may pseudoaccommodate better than older eyes.

Evaluation of whether intracameral dexamethasone predisposes to glaucoma after pediatric cataract surgery

Asimina Mataftsi; Ahmad Dabbagh; Will Moore; Ken K. Nischal *J Cataract Refract Surg*. October 2012; 38(10):1719-23

The authors describe a single institution case series of infants undergoing cataract extraction with intraocular lens implantation, who received intracameral dexamethasone during surgery. The authors report on the percentage of eyes developing glaucoma. There was no comparison group. During a two year period, eighteen patients (24 eyes) were included. The patients underwent lensectomy, acrylic IOL implantation in the bag, posterior capsulotomy and anterior vitrectomy. In addition to intracameral dexamethasone, the patients were treated with subconjunctival dexamethasone, orbital floor triamcinolone, and postoperative topical Maxitrol hourly for the first 24 hours and then tapered. The median age at surgery was 3 months (mean 4 months ± 3 (SD); range 1 to 11 months). The median follow-up was 38 months (mean 34 ± 10 months; range 20 to 48 months). Glaucoma was defined as IOP higher than 21 on two occasions or increased digital IOP with ocular signs of glaucoma, such as myopic shift or increase in cupping. No eye developed glaucoma during the follow-up period. In 4 eyes, transient postoperative antihypertensive medication was used. There was a 58% rate of posterior visual axis opacification, similar to the high rates seen in other studies on infantile cataract surgery with IOLs.

The title of the study is somewhat misleading, as it is not possible to attribute the nil rate of glaucoma to the use of intracameral dexamethasone, versus other surgical and medical techniques employed by the authors. But certainly, these results are reassuring that intracameral dexamethasone is not causing high rates of glaucoma.

Subtle signs of an intact anterior vitreous face during pediatric cataract surgery Aditya A. Sudhalkar; Mamidipudi R. Praveen; Viraj A. Vasavada; Sajani K. Shah; et al. *J Cataract Refract Surg*. September 2012; 38(9):1690-3.

The authors describe the triamcinolone-stained appearance of the anterior vitreous face in three children, ages 4-6 years, who underwent cataract extraction with manual posterior continuous curvilinear capsulorrhexis (PCCC). The authors also describe their technique in creating the manual PCCC and avoiding disturbing the anterior vitreous face. Surgeons who are contemplating cataract extraction in children with PCCC but without anterior vitrectomy should review the photographs provided by the authors, which show the appearance of the intact vitreous face as a central triamcinolone-stained disc, without extension of vitreous anteriorly.

Vitrectorhexis versus forceps posterior capsulorhexis in pediatric cataract surgery. Kochgaway L, Biswas P, Paul A, Sinha S, Biswas R, Maity P, et al. Indian J Ophthalmol 2013;61:361-4.

Fifty eyes with congenital and developmental cataract were included in this study. The posterior capsulorhexis was created using utrata forceps in 17 eyes or through a vitrector in 33 eyes. Forceps capsulorhexis was performed before IOL implantation, while vitrectorhexis was performed after IOL implantation in the bag. The results of both the surgery were compared using the following criteria: incidence of extension of rhexis, ability to achieve posterior rhexis of appropriate size, ability to implant the IOL in the bag, the surgical time, and learning curve. Vitrectorhexis after IOL implantation was an easy to learn alternative to manual posterior continuous curvilinear capsulorhexis in pediatric cataract surgery. It was more predictable and reproducible, with a short learning curve and lesser surgical time.

PEDIATRIC INTRAOCULAR LENS POWER CALCULATIONS

Mary A. O'Hara Current Opinion in Ophthalmology Sept. 2012 23(5) p. 388-393

Accurate determination of appropriate intraocular lens (IOL) power is still a major challenge in pediatric cataract surgery. As pediatric cataract surgery techniques evolve and the age at IOL implantation becomes younger, the need for more accurate IOL power calculations increases. More studies on the ideal postoperative refractive state and new approaches to a variable refractive solution are warranted. Formulas for IOL power calculations are mainly derived from studies in adults and therefore pediatric IOL power calculations suffer from significant prediction error. These formulas are most accurate in the older children. The smallest eyes have the most prediction error with all available formulas. To implant an appropriate IOL in a child, along with accurate ocular

measurements and IOL power calculations it is necessary to predict the refractive change that comes with growth of the pseudophakic eye to maturity. Individual circumstances and parental concerns must be factored into the choice of a postoperative refractive target.

14. GLAUCOMA

Goniotomy versus circumferential trabeculotomy with an illuminated microcatheter in congenital glaucoma.

Girkin CA, Rhodes L, McGwin G, et al. J AAPOS 2012;16:424-427

This study compared visual outcomes of circumferential trabeculotomy performed with an illuminated microcatheter versus a standard goniotomy in the treatment of congenital glaucoma. This retrospective study defined complete success as a similar level of intraocular pressure (IOP) reduction without the use of antiglaucoma medications, and qualified success as with medications. Twelve eyes had trabeculotomy (6-360', 3-270', 3-180'). Four eyes in the trabeculotomy group and 2 eyes in the goniotomy group had prior surgery. Significantly lower average final postoperative IOP was obtained in the trabecuolotomy group. Neither group had long-term complications. Complete 360 degree trabeculotomy showed the greatest mean IOP reduction. The trabeculotomy group showed 91.6% qualified and 83.3% unqualified success rates versus 53.8%/46.2% in the goniotomy group. Prior surgery lowered success, but all of these groups had small numbers of patients. Follow-up was short and groups were not randomized. Finally, the groups were not contemporaneous.

The use of the Icare tonometer reduced the need for anesthesia to measure intraocular pressure in children

Grigorian F, Grigorian P, and Olitsky SE. J AAPOS 2012;16:508-510

The authors evaluate the reliability of the Icare tonometer and whether or not it decreased the need for examinations under anesthesia (EUA). There were 2 groups of patients: 1.diagnosis of glaucoma, 2.procedures in operating room, which included EUAs. The authors found that in-office IOP measurements increased significantly, and the need for EUAs decreased significantly during the study period as the Icare tonometer was introduced in their practice. This was a retrospective study, and the authors comment that the % of new versus established patients during the course of the study (which was not evaluated) could also influence the rates of EUAs that were needed.

Longitudinal reproducibility of optical coherence tomography measurements in children

Prakalapakorn SG, Freedman SF, Lokhnygina Y, et al. J AAPOS 2012;16:523-528

This study was a prospective evaluation of the longitudinal reproducibility of OCT in children with known or suspected glaucoma or healthy eyes. The first study setting included 8 normal eyes of 8 healthy children (average age 9.1 years) followed for over 3 years. There was good reproducibility of RNFL despite increased axial length over time. The second study group included normal eyes (27 eyes of 27 children), mild glaucoma (23 eyes of 23 children) and advanced glaucoma (14 eyes of 14 children). These children were also followed for over 3 years. At the initial visit the average macular volume was sequentially lower, and the average RNFL was sequentially thinner as one compared normal to mild glaucoma to advanced glaucoma. There was good reproducibility. Only one patient in the advanced glaucoma group showed progression of disease by optic nerve appearance during the study period. Postoperatively, there was reversal of cupping, and optic nerve pallor, as well as decreased macular volume and average RNFL. Sample sizes in this study were small, and since only one patient in the study had progression of disease, it is difficult to comment as to whether average RNFL thickness or total macular volume are helpful in measuring progressive glaucoma.

A Clinical and Molecular Genetics Study of Primary Congenital Glaucoma in South Korea

Wool Suh, Changwon Kee. Br J Ophthal 2012; 96: 1372-1377.

This paper studied the clinical manifestations of human cytochrome P450 (CYP1B1) and myocilin (MYOC) genes in the South Korean patients with primary congenital glaucoma (PCG). Eighty-five patients with PCG were studied and grouped according to presence of mutations and they were surgically treated for their glaucoma. All clinical data was compared between the two groups. The group with the mutations presented at a younger age (0-3 years) compared to the non-mutation group (0-6 years). The proportion of early onset, involvement of both eyes, and severity index were higher in the mutation group. However, the number of surgical interventions did not differ. Fifty percent of the group which possessed the mutation required supplementary medication to keep IOP below 21. Final visual outcome showed no difference between the two groups. In conclusion, no consistent correlation existed between clinical findings at initial presentation and genotype, however, treatment response was associated with number of CYP1B1 mutations.

Surgical Outcomes in Childhood Uveitic Glaucoma

Brenda L. Bohnsack, Sharon F. Freedman. Am J Ophthal January 2013, 155 (1): pgs. 134-42.

This was a retrospective study reviewing 36 patients with juvenile uveitic glaucoma who underwent surgical therapy over a 15 year period. The majority of

the patients had juvenile rheumatoid arthritis. Thirty-one eyes had goniotomy while 5 underwent a drainage device. In the goniotomy group, 1 went on to trabeculectomy and 6 went on to a filtering device. Despite surgical procedures for glaucoma and cataracts, these children went on to have a good prognosis for vision preservation. Goniotomy was found to be a good first choice surgery, preserving conjunctival tissue and controlling IOP in 24 eyes.

Trabeculectomy Versus Combined Trabeculotomy-Trabeculectomy in Pediatric Glaucoma

Scott D. Lawrence, Peter A. Netland, J Pediatr Ophthalmol Strabismus 2012;49:359-365 (Nov/Dec)

The management of congenital glaucoma in children is primarily surgical – either goniotomy or trabeculotomy. In some instances, one can perform a combination of surgical procedures such as trabeculotomy – trabeculectomy. This article discusses the success rate of a trabeculotomy versus a combined trabeculotomy – trabeculectomy. A retrospective review of 40 eyes in 33 patients showed that the 2 procedures were equally effective at lowering IOP in children. The combined procedure was associated with a higher long term success rate.

Late-Recognized Primary Congenital Glaucoma

David S. Walton, Karina Nagao, Helen H. Yeung, Steven A. Kane, J Pediatr Ophthalmol Strabismus 2013; 50:234-238 (July/August)

The purpose of the study is to describe a cohort of children with later recognized primary congenital glaucoma (LRPCG). The medical records of children were examined who were diagnosed with primary congenital glaucoma after age 1 were reviewed. The study determined the age of onset, diagnostic signs, intraocular pressures, visual acuity. Interestingly enough, corneal clouding was not an initial sign for any of the patients. The authors concluded that an awareness of the diagnostic signs can assist in early diagnosis of LRPCG and prevent vision loss.

Caregiver Burden Assessment In Primary Congenital Glaucoma

Tanuj Dada, Ashutosh Aggarwal, Shveta Jindal Bali, Meenakshi Wadhwani Eur J Ophthalmol May-June 2013; 23(3): 324 – 328

Caregivers of patients with primary congenital glaucoma have significant emotional and psychological burden. One-third of these primary care givers suffer from moderate to severe depression. This study used a Caregiver Burden Questionnaire (CBQ) and the depressive symptomatology was evaluated using a Patient Health Questionnaire–9 (PHQ-9) on 55 primary caregivers, of which 2 were males.

Online-Only Article: Ophthalmic Technology Assessment: Rebound Tonometry in Children: A Report by the American Academy of Ophthalmology

Scott R. Lambert, Michele Melia, Angela N. Buffenn, Michael F. Chiang, *Ophthalmology* April 2013;120:657-662

Rebound tonometry in children is better tolerated by young children than applanation tonometry.

A case of arrested primary congenital glaucoma

M Shaw; S Handley; H Porooshani; M Papadopoulos *Eye*. January 2013;27(1):100.

A healthy 14 month old boy was noted to have unilateral buphthalmos with an ispilateral Haab striae, myopia, longer axial length and larger optic disc cup. Infantile glaucoma was suspected, but two exams under anesthesia showed normal intraocular pressure. The authors speculate that this may be the third published case of spontaneous resolution of infantile glaucoma. One mechanism that would explain such findings is postnatal maturation of the drainage angle.

Polypropylene vs silicone Ahmed valve with adjunctive mitomycin C in paediatric age group: A prospective controlled study

Y El Sayed; A Awadein *Eye.* June 2013; 27:728–34.

The authors prospectively compared silicone and polypropylene Ahmed glaucoma valves implanted in children ages 10 and under who had already failed other glaucoma procedures. 50 eyes of 33 patients with pediatric glaucoma were followed for two years after implanting either a polypropylene or silicone Ahmed valve. In eyes with bilateral glaucoma, one eye was implanted with polypropylene and the other eye was implanted with silicone, and this assignment was made at random. In eyes with unilateral glaucoma, patients were matched on the basis of glaucoma type and the number of prior operations. Twenty five eyes received a polypropylene valve, and 25 eyes received a silicone valve. Eyes implanted with silicone valves achieved a significantly lower intraocular pressure (IOP) compared with the polypropylene group at 6 months, 1 year, and 2 years postoperatively. The average survival time was significantly longer (P=0.001 by the log-rank test) for the silicone group than for the polypropylene group and the cumulative probability of survival by the log-rank test at the end of the second

year was 80% (SE: 8.0, 95% CI: 64–96%) in the silicone group and 56% (SE: 9.8, 95% CI: 40–90%) in the polypropylene group. The difference in the number of postoperative interventions and complications between both groups was statistically insignificant. The authors conclude that silicone Ahmed valves can achieve better IOP control, and longer survival with fewer antiglaucoma drops compared with polypropylene valves in children younger than 10 years. They suspect that there is less encapsulation with the silicone valves than the polypropylene valves. There were more smaller eyes and thus more pediatric sized implants placed in the silicone group than in the polypropylene group, but if anything, this makes the better results in the silicone group more convincing.

Based on the results of this study and the authors' review of other retrospective literature, silicone rather than polypropylene Ahmed valves seem like the better choice for pediatric glaucoma cases that have failed other surgical procedures.

Deep sclerectomy in pediatric glaucoma filtering surgery

N H L Bayoumi *Eye*. December 2012;26(12):1548-53

This study was a prospective, randomized case series of twenty eyes with primary congenital glaucoma evaluating the addition of a deep sclerectomy to planned combined trabeculotomy—trabeculectomy with mitomycin C (CTTM). The single author/surgeon performed the operations. He fashioned a 4 x 4 x 4 mm triangular scleral flap carried forward into clear cornea. A nurse then randomized patients intraoperatively to undergo standard CTTM, or CTTM with the addition of an additional 2 x 2 mm deep scleral flap. In the ten patients randomized to deep sclerectomy, the author used the deeper scleral flap to deroof Schlemm's canal to facilitate the trabeculotomy.

Despite randomization, the patients were not perfectly matched by age. The CTTM group was younger than the CTTM + deep sclerectomy group: 4.7 months (±2.0, 2–8) vs 7.0 (±3.8, 3–13) months. The mean (±SD, range) preoperative intraocular pressure in the CTTM and CTTM + deep sclerectomy groups was 16.7 (4.3, 10–26) and 16.4 (8.4, 8–36), respectively, and these dropped at 12 months of follow-up to 4.9 (2.0, 2–8) and 5.6 (3.3, 2–10), respectively. Two eyes (20%) in the CTTM+ deep sclerectomy group developed hypotony disc edema, which resolved spontaneously after two months.

The author concludes that the addition of deep sclerectomy to the procedure of CTTM in pediatric glaucoma surgery facilitates the finding of Schlemm's canal, and this could be a helpful surgical strategy even when performing trabeculotomy alone. A critique of the study is limited data regarding the patients' visual function, so it is unclear whether there was an effect on vision in the two patients who developed transient hypotony disc edema. Another observation was the relatively low intraocular pressures measured under sevoflurane general anesthesia preoperatively, which the author attributes to sevoflurane effect.

15. REFRACTIVE SURGERY

Diplopia Associated with Refractive Surgery

Burton J. Kushner, Am Orthopt J. August 2012;62:34-37

This review article describes etiologies for post-refractive surgery binocular diplopia including monovision (with and without fixation switch) and pre-existing strabismus with incorrect optical outcome resulting in an increase or decrease in accommodative convergence. Management algorithm based on clinical findings is described and includes options such as elimination of monovision, corrective lenses to manage accommodation as well as prisms and eye muscle surgery.

16. GENETICS

Severity of Familial Isolated Retinitis Pigmentosa Across Different Inheritance Patterns Among an Asian Indian Cohort

Purva Bende, Krupa Natarajan, Thennarasu Marudhamuthu, Jagadeesan Madhavan, J Pediatr Ophthalmol Strabismus 2013;50:34-36 (Jan/Feb)

Retinitis Pigmentosa is an inherited disease with various inheritance modes including x-linked recessive, autosomal dominant and autosomal recessive. In this article, retinitis pigmentosa is discussed in great detail and each mode of inheritance is studied in terms of its effects on visual function. Which mode of inheritance is most likely to lead to blindness and how quickly. Many forms of RP are associated with myopia; however, upon breakdown into the various inheritance modes, its found that higher myopia is associated with the x-linked form whereas the autosomal dominant form is more associated with no refractive error.

Mutations in the CYP1B1 gene may contribute to juvenile-onset open-angle glaucoma

C-C Su; Y-F Liu; S-Y Li; J-J Yang, et al. *Eye.* October 2012; 26(10):1369-77.

The study investigated whether mutations in *CYP1B1* gene may be responsible for juvenile-onset open-angle glaucoma (JOAG) among Taiwanese patients. CYP1B1 is a causative gene in primary congenital glaucoma. 61 unrelated probands with JOAG were compared to 100 healthy control subjects. JOAG was defined as onset before age 35 years, intraocular pressure greater than 22, open angle, glaucomatous visual field defect and optic disc cupping. Controls were subjects older than 50 years who had no glaucoma. The *CYP1B1* gene was sequenced for all subjects. Two *CYP1B1* mutations were identified in three of

the 61 JOAG subjects (4.9**%) but none of the controls**. The mutations included a missense mutation and a mutation in the 5′-untranslated region, which is near the promoter for the CYP1B1 gene. The missense mutation is in a part of the gene that is highly conserved across species. Using modeling, the authors demonstrated that their missense mutation affects protein structure and function.

The authors conclude that the CYP1B1 gene is occasionally responsible for JOAG. In a previous study, they found that 12.5% of the same JOAG subjects enrolled in this study had mutations in the MYOC or OPTN genes. However, the gene(s) responsible for the majority (>80%) of JOAG cases in Taiwan remain to be found.

Null CYP1B1 Genotypes in Primary Congenital and Nondominant Juvenile Glaucoma

Maria-Pilar Lopez-Garrido, Cristina Medina-Trillo, Laura Morales-Fernandez, Julian Garcia-Feijoo, *Ophthalmology* April 2013;120:716-723

This is a CYP1B1 genotyping, segregation analysis, and functional evaluation of mutations in a cohort 177 probands clinically diagnosed with primary congenital glaucoma (PCG) and nondominant juvenile glaucoma (ndJG).
Methods: Automatic DNA sequencing of the promoter gene region and 3 CYP1B1 exons. This study was performed in Madrid and Albacete, Spain. The main outcome measures were based on slit lamp examinations, measurements of intraocular pressures, gonioscopy, and fundus examinations.
Conclusions: This is reported to be the largest of analysis of CYP1B1 mutations performed in European patients with primary congenital glaucoma. The study shows that complete absence of the CYP1B1 gene activity frequently lead to severe phenotypes of glaucoma. The results of this study support CYP1B1 glaucoma is not a simple monogenic disease and that CYP1B1 activity levels could influence the clinical/phenotypic expression of the disease.

A Homozygous Frameshift Mutation in LRAT Causes Retinitis Punctata Albescens

Karin W Littink, Maria M van Genderen , Mary J van Schooneveld
Ophthalmology September 2012;119:1899-1906

This article comes from the Rotterdam Eye Hospital, Rotterdam, Netherlands.
This is a case series/observation study of 13 patients from 8 families with retinal dystrophy characterized by tiny, yellow-white dots on fundoscopy typical for fundus albipunctatus (FAP) or retinitis punctata albescens (RPA).
Thirteen patients from 8 families with retinal dystrophy characterized “tiny, yellow-white dots on fundoscopy”, underwent full ophthalmologic evaluation including visual field assessment. Fundus photography and electroretinography

was performed in 12 of the 13 patients. Optical coherence tomography and fundus autofluorescence were performed in 4 patients.
DNA samples for all patients were screened for mutations of RLB1 (retinitis punctata albescens) retinaldehyde-binding protein 1. For those patients that did not carry mutations with RLBP1, DNA was analyzed for RDH5 (associated with fundus albipunctatus [FAP]), (encoding 11-cis-retina dehydrogenase 5 [RDH5]). Both of these enzymes are responsible in retinal metabolism and particularly the retinoid cycle. The retinoid cycle is a crucial process in which 11-trans-retinal is recycled into 11-cis-retinal to provide a constant rate of 11-cis-retinal for the phototransduction cascade. Defects in the retinoid cycle have recently become a subject of new pharmaceutical therapy trials in mouse models and humans. Treatment with 9-cis-retinoids rescued visual function and retinal structure in RPE65 and LRAT deficient mice. This study is significant in a sense that it identified a mutation of LRAT in 4 patients with retinitis punctata albescens (RPA). Mutations in RLBP1 were identified in 7 patients with retinitis punctata albescens and in 1 patient with fundus albipunctata (FAP) with mild maculopathy).

Conclusion of the study was a genetic defect was identified in LRAT and thus represented a novel cause of retinitis punctata albescens. LRAT is therefore the fourth gene involved in the visual cycle that may cause a white dot retinopathy. Mutations in RLB1 may lead to fundus albipunctatus with cone dystrophy.

In order to understand the significance of this paper, a review is in order. Fundus albipunctatus and retinitis punctata (RPA) are hereditary retinal diseases characterized by the presence of tiny, yellow-white dots throughout the retina. Both are inherited as autosomal recessive patterns. This disease is characterized by decreased night vision and is classified as different entities because of the course of the disease and the electroretinogram abnormality.

Fundus albipunctatus (FAP) is a stationary disorder that has initially been classified as a form of congenital stationary night blindness (CSNB). Retinitis punctata albescens (RPA) is a progressive disorder and is a classified form of retinitis pigmentosa. Fundus albipunctatus is characterized by a mutation in the RDH5 gene. Retinitis punctata albescens is caused by mutations in the RLBP1 gene. The significance of this study is that mutations of the LRAT gene have classically been associated with Leber's congenital amaurosis and early-onset severe retinal dystrophy (EOSRD). This study suggests that mutations in LRAT typically involve the development of white dot retinopathy, particularly retinitis punctata albescens (RPA) and fundus albipunctatus (FAP).

Genome engineering using the CRISPR-Cas9 system.

Ran FA, Hsu PD, Wright J, Agarwala V, Scott DA, Zhang F. Nat Protoc. 2013 Nov;8(11):2281-308. doi: 10.1038/nprot.2013.143. Epub 2013 Oct 24

A revolution in treating genetic diseases may have just started! CRISPR stands for **c**lustered **r**egularly **i**nterspaced **s**hort palindromic **r**epeats. It is a part of the bacterial immune system which has been discovered to be useful in correcting DNA mutations; with this system it may be possible to excise and correct many errors in DNA that cause human disease. The authors describe the tools needed for RNA-guided Cas9 mediated genome editing. They are not the first authors to describe this (although the first reports are also very recent), but they are the first to modify it to minimize off-target cleavage. In brief, with the CRISPR technique an RNA-guide is used to get a Cas9 nuclease to a specific spot in the genome where there is a disease-causing mutation. The nuclease cuts the DNA at this spot and the DNA can then repair itself using a template. Unlike gene therapy, now used for one type of LCA, in which a viral vector with a new copy of the defective gene must be delivered with only genes small enough to be packaged eligible for the most well studied vector, with CRISPR even large genes could be amenable to "gene therapy" by directly fixing the defective region of DNA.

Characterisation of retinoblastomas without RB1 mutations: genomic, gene expression, and clinical studies.

Rushlow DE, Mol BM, Kennett JY, Yee S, et al. Lancet Oncol. 2013 Apr;14(4):327-34. doi: 10.1016/S1470-2045(13)70045-7. Epub 2013 Mar 13.

Retinoblastoma (Rb) is a malignant ocular tumor of childhood. When a child presents with Rb it is of critical importance to determine whether it is caused by a germline mutation, present in all of the child's cells, predisposing them to other ocular and non-ocular tumors and also implying risk to other family members, or a spontaneous mutation which is an isolated event carrying no other risks. For many years this has been determined by sending a portion of the tumor and the patient's blood for genetic testing to look for mutations in the RB1 gene, the only known cause of Rb. In this extremely important paper, Brenda Gallie's group describes their discovery of another gene that can cause Rb, the MYCN gene. This is of vital importance in the work up of children with retinoblastoma.
The authors analysed 1068 samples from unilateral Rb patients and studied genomic copy number, RB1 gene expression and protein function, retinal gene expression, histological features, and clinical data, and they compared this information between those in which RB1 mutations had been found versus those with no mutation found. They had 29 tumors in which no RB1 mutation was found. 15 of the 29 RB1(+/+) tumours had high-level MYCN oncogene amplification (28-121 copies; RB1(+/+)MYCN(A)), whereas none of 93 RB1(-/-) primary tumours tested showed MYCN amplification ($p<0{\cdot}0001$). Two more MYCN amplification patients were subsequently identified. Median age at diagnosis of the 17 children with RB1(+/+)MYCN(A) tumours was 4·5 months (IQR 3·5-10), compared with 24 months (15-37) for 79 children with non-familial unilateral RB1(-/-) retinoblastoma.
The take-home message: A portion of children with unilateral retinoblastoma develop it as a result of MYCN amplification, not sporadic RB1 mutation. These children have a very young age of onset (such as we usually associate with

hereditary Rb) and aggressive histologic features. This new cause of retinoblastoma should be considered in patients with retinoblastoma.

Pax6 downregulation mediates abnormal lineage commitment of the ocular surface epithelium in aqueous-deficient dry eye disease.

Chen YT, Chen FY, Vijmasi T, Stephens DN, Gallup M, McNamara NA. PLoS One. 2013 Oct 15;8(10):e77286. doi: 10.1371

Keratinzing squamous metaplasia of the ocular surface can cause blindness and is found in systemic autoimmune disease. It is also a feature of aniridia. The authors had previously established the autoimmune regulator-knockout (Aire KO) mouse as a model of autoimmune keratoconjunctivitis sicca and identified an essential role for autoreactive CD4+ T cells in squamous metaplasia pathogenesis. They had also noted the down-regulation of paired box gene 6 (Pax6) in both human patients with Sjögren's syndrome and Aire KO mice. Pax6 encodes a pleiotropic transcription factor guiding eye morphogenesis during development. They hypothesized that the role of Pax6 in maintaining ocular surface homeostasis was disrupted in the inflamed eye and that loss of Pax6 played a functional role in the development and progression of surface disease. Adoptive transfer of autoreactive T cells from Aire KO mice to immunodeficient recipients confirmed CD4+ T cells as the cause of Pax6 downregulation in Aire KO mice. They then treated the mice with adenovirus vector forcing Pax6 expression in corneal epithelial cells, and it reversed the squamous metaplasia! This could be a novel treatment for inflammatory ocular surface disease, and possibly for aniridia related cornea opacity.

Small-interfering RNAs (siRNAs) as a promising tool for ocular therapy.

Guzman-Aranguez A, Loma P, Pintor J. Br J Pharmacol. 2013 Oct;170(4):730-47

Genetic eye diseases which are caused by lack of a normal protein are the best targets for subretinal gene replacement therapy. This encompasses many autosomal recessive disorders. But what about autosomal dominant disorders where the problem is not the lack of enough normal protein but instead the toxicity of an abnormal protein product? For these disorders small-interfering RNAs are promising. This review nicely discusses how interfering RNAs work, and why the eye seems to be a very good target for this type of treatment for glaucoma, retinitis pigmentosa, and neovascular eye diseases, among others.

Patient-specific iPSC-derived photoreceptor precursor cells as a means to investigate retinitis pigmentosa.

Tucker BA, Mullins RF, Streb LM, Anfinson K, et al Elife. 2013 Aug 27;2:e00824.

The authors demonstrate that it is possible to take a skin biopsy from patients with retinitis pigmentosa, grow them in a dish, de-differentiate them into

pluripotent stem cells, then drive differentiation to photoreceptor precursor cells. These cells can be studied to determine which gene is causing the RP. Then a normal copy of that gene can be placed into a viral vector and used to treat the cells in vitro. In this study these treated cells were then injected subretinally into a mouse without photoreceptors and photoreceptors formed from the transplanted cells. This is proof of concept that a strategy might be developed for humans in which patients with retinal degeneration could have their own skin cells made into photoreceptors, corrected by gene therapy in a dish, then transplanted into their own eyes to restore vision when all photoreceptors have been lost.
This study describes using next-generation and Sanger sequencing to identify disease-causing USH2A mutations in an adult patient with autosomal recessive RP. Induced pluripotent stem cells (iPSCs), generated from the patient's keratinocytes, were differentiated into multi-layer eyecup-like structures with features of human retinal precursor cells. The inner layer of the eyecups contained photoreceptor precursor cells that expressed photoreceptor markers and exhibited axonemes and basal bodies characteristic of outer segments. Analysis of the USH2A transcripts of these cells revealed that one of the patient's mutations causes exonification of intron 40, a translation frameshift and a premature stop codon. Western blotting revealed upregulation of GRP78 and GRP94, suggesting that the patient's other USH2A variant (Arg4192His) causes disease through protein misfolding and ER stress. Transplantation into 4-day-old immunodeficient Crb1 (-/-) mice resulted in the formation of morphologically and immunohistochemically recognizable photoreceptor cells, suggesting that the mutations in this patient act via post-developmental photoreceptor Degeneration.

Subretinal gene therapy of mice with Bardet-Biedl syndrome type 1.

Seo S, Mullins RF, Dumitrescu AV, Bhattarai S, Gratie D, Wang K, Stone EM, Sheffield V, Drack AV. Invest Ophthalmol Vis Sci. 2013 Sep 11;54(9):6118-32. doi: 10.1167/iovs.13-11673.

Bardet Biedl syndrome usually presents with loss of vision in the first decade and may produce complete blindness due to retinal degeneration between the second and third. There are 17 known genetic subtypes, but the most common is BBS1 and the most common mutation is M390R. A mouse model of this disorder has been created and recapitulates the human phenotype. Subretinal gene therapy was attempted in this mouse as a pre-clinical study for humans. An AAV2/5 vector with a normal copy of the gene was created, and was injected subretinally in the right eye of affected mice, while the same viral vector with a reporter molecule, Green Fluorescent Protein (GFP), which can be seen as a green fluorescent glow, was injected subretinally in the fellow eye. Unaffected mice were also treated. It was discovered that while the treatment rescued formation of the protein complex of BBS proteins, called the BBSome, it did not rescue the ERG of treated animals. The reason was over-expression toxicity of

the BBS1 protein. That is, just as too little of the BBS1 gene protein product is toxic to the retina, so is too much. In normal mice receiving a subretinal injection of the viral vector there was retinal degeneration. In BBS1 mice there was no worsening of retinal function, but neither was there improvement in most cases. None of the eyes treated with the same vector carrying the reporter molecule showed toxicity. One small group of BBS mice did show a small improvement in ERG function—this was the group given the smallest volume of vector. The conclusion is that for this gene the expression of the delivered normal gene will have to be titrated carefully, but that treatment of BBS1 retinal degeneration by subretinal gene therapy may be possible.

AAV-mediated gene therapy for choroideremia: preclinical studies in personalized models.

Vasireddy V, Mills JA, Gaddameedi R, Basner-Tschakarjan E, Kohnke M, et al PLoS One. 2013 May 7;8(5):e61396. doi: 10.1371

The authors report that Choroideremia (CHM) is an X- linked retinal degeneration that is symptomatic in the 1(st) or 2(nd) decade of life causing nyctalopia and loss of peripheral vision. The disease progresses through mid-life, when most patients become blind. It may be amenable to subretinal gene therapy because it is due to loss of function of a protein, REP1, and the gene is of the size that can be packaged in AAV which is already in a successful clinical trial for treating RPE65 LCA. There is no animal model to test it, however. Therefore the authors used cells from CHM patients, namely lymphoblasts and induced Pluripotent Stem Cells (iPSCs) from human patients and treated them in vitro. It restored the REP1 enzymatic activity and restored protein trafficking! They also did subretinal gene therapy in normal mice with their vector, and it was not toxic. This bodes well for a future treatment of human choroideremia with gene therapy.

Gene therapy for blindness.

Sahel JA, Roska B Annu Rev Neurosci. 2013 Jul 8;36:467-88. doi: 10.1146/annurev-neuro-062012-170304. Epub 2013 May 31.

This is an excellent review of gene replacement and neuroprotection and also new avenues such as optogenetic therapies.

Cone Structure in Patients With Usher Syndrome Type III and Mutations in the Clarin 1 Gene

Kavitha Ratnam, Hanna Va¨stinsalo, Austin Roorda, Eeva-Marja K. Sankila, et.al. JAMA Ophthalmol. 2013;131(1):67-74.

The authors studied macular structure and function in three patients with Usher syndrome type III caused bymutations in the Clarin 1 gene (CLRN1). Adaptive optics scanning laser ophthalmoscopy and spectral domain optical coherence tomography were used to study the macula in these patients and compared with

those of age-similar control subjects Vision function measures included best-corrected visual acuity, kinetic andstatic perimetry, and full-field electroretinography. Coding regions of the CLRN1 gene were sequenced Cones were observed centrally but not in regions with scotomas, and retinal pigment epithelial cells were visible in regions without cones in patients with CLRN1 mutations. High-resolution measures of retinal structure demonstrate patterns of cone loss associated with CLRN1 mutations.These findings provide insight into the effect of CLRN1 mutations on macular cone structure, which has implications for the development of treatments for Usher syndrome type III.

Visual Acuity Changes in Patients With Leber Congenital Amaurosis and Mutations in CEP290

J. Jason McAnany, Mohamed A. Genead, Saloni Walia, Arlene V. Drack, et.al. JAMA Ophthalmol. 2013;131(2):178-182

Leber's congenital amaurosis is characterized by severe visual loss early in life with nystagmus, a normal appearing fundus and a severely reduced or absent photopic and scotopic responses on electroretinography. To date, 18 LCA genes have been reported. The most commonly mutated genes in the European population are CEP290 (15% of cases) and GUCY2D (12% of cases) In this study, the authors evaluate changes in visual acuity over time in patients with Leber congenital amaurosis caused by mutations in the CEP290 gene. Visual acuity was determined at the initial and most recent visits of 43 patients. At the initial visit, 14 patients had measurable chart visual acuity in the better-seeing eye, 25 patients had nonmeasurable chart visual acuity, and 4 young patients did not have visual acuity assessed. At the most recent visit, 15 patients had measurable chart visual acuity and 28 had nonmeasurable chart visual acuity. The average interval between the 2 visits was 10.4 years Patients with LCA and CEP290 mutations had a wide spectrum of VA that was not related to age or length of follow-up. Severe VA loss was observed in most, but not all, patients in the first decade.

Novel Mutation in BEST1 Associated With Retinoschisis

Ruwan A. Silva, Audina M. Berrocal, M,Byron L. Lam, Thomas A. Albini, JAMA Ophthalmol. 2013;131(6):794-798.

Best vitelliform macular dystrophy is caused by mutations in BEST1 on the long arm of chromosome 11. An array of BEST1 phenotypes have been characterized, including microcornea, rod cone dystrophy, early-onset cataract, posterior staphyloma, vitreoretinochoroidopathy, and adult-onset foveomacular vitelliform dystrophy.
BEST1 encodes bestrophin, a 585–amino acid protein with more than 120 described mutations. The authors present 2 siblings with bilateral retinoschisis and electroretinography consistent with Best's disease. The siblings

demonstrated bilateral symmetric multifocal macular lesions and a normal full-field ERG with an abnormal electrooculogram. The children also had subretinal fluid and retinoschisis associated with a novel mutation in BEST1.

De Novo Splice Mutation in the Versican Gene in a Family With Wagner Syndrome

Pierre-Raphae¨l Rothschild, Isabelle Audo, Brigitte Nedelec, Tiffany Ghiotti, et.al. JAMA Ophthalmol. 2013;131(6):805-807.

Wagner syndrome (WS) is a rare inherited vitreoretinopathy caused by mutations in the versican gene (VCAN). Clinically, Wagner syndrome is characterized by an optically empty vitreous with no systemic features. The risk of retinal detachment is high.The authors present a patient with asymptomatic parents and de novo VCAN mutation with clinical features typical for Wagner syndrome. De novo VCAN mutations have not been reported to date. This case report highlights the importance of VCAN screening in isolated individuals with a WS phenotype.

Mutation of GNAQ in a Cytologically Unusual Choroidal Melanoma in an 18-Month-Old Child

Steve Daniel Levasseur, Katherine E. Paton, Catherine D. Van Raamsdonk, ,Manraj K. S. Heran, JAMA Ophthalmol. 2013;131(6):810-812.

Fewer than 1% of uveal melanomas occur in patients younger than 20 years. The authors report a histologically unusual uveal melanoma in an 18 month old girl that was confirmed by the presence of a GNAQ somatic mutation. The authors note that only 5 cases of congenital uveal melanoma have been reported. Their patient presented with an abnormal red reflex at 6 months of age, so the author believe this case may be congenital. The clinical and histopathologic features of pediatric uveal melanoma are usually similar to those found in adults. They are typically unilateral, occur in the choroid with no sex predilection, and are usually of spindle cell type.

17. TRAUMA

Visual Outcome Of Traumatic Cataract In Pediatric Age Group

Mehul A. Shah, Shreya M. Shah, Adway H. Appleware, Krunal D. Patel, Eur J Ophthalmol November – December 2012; 22(6): 956 – 963

This retrospective study analyzed the visual outcome of open-globe and closed-globed traumatic pediatric cataracts that underwent surgical extraction, lens implantation and amblyopia therapy. Of the 354 cases, 287 eyes were open-globe injury and 67 were closed globe injury. At 6 weeks post-operative

evaluation, visual acuity =20/40 was present in 35.3% and VA > 20/200 was present in 61.3%. There was no statistical difference between the groups.

Effectiveness of Protective Eyewear in Reducing Eye Injuries Among High School Field Hockey Players

Peter K. Kriz, R. Dawn Comstock, David Zurakowski Pediatrics November 2012; 130:6 1069-1075

The authors analyzed field hockey exposure and injury data for two scholastic seasons (2009 – 2011) from national and regional databases in states that mandated protective eyewear versus states that had no eyewear mandate. All head and face injuries were significantly higher in the non-mandating states (0.69 vs. 0.47 injuries per 1000 athletic exposures. The eye injury rate was 5.33 fold higher in the non-mandating states as was the longer recuperation (>10 days) period (32% in non-mandating states vs. 17%). There was no significant difference in concussion rates.

Prediction of Inflicted Brain Injury in Infants and Children Using Retinal Imaging

Robert A. Minns, Patricia A. Jones, Anamika Tandon
Pediatrics October 2012; 130:5 e1227-e1234

This is a prospective study showing that through retinal imaging of all potential causes of retinal hemorrhages, a young age and increased number of hemorrhages are strong predictors of non-accidental trauma

Prediction of visual outcomes after open globe injury in children: A 17-year Canadian experience

Bunting H, Stephens D and Mireskandari K J AAPOS 2013;17:43-48

The ocular trauma score (OTS) has existed for a decade and provides information regarding visual prognosis based on specific findings at initial assessment after an ocular injury. More recently, a pediatric ocular trauma score (POTS) was published. The authors retrospectively evaluated a large cohort of Canadian children with open globe injuries over an 18-year period. Three-quarters of the 131 identified patients were boys, and almost half of the injuries occurred in children under age 6 years. Indoor injuries were more common. The most common causes for injuries were glass, knives and scissors. Patients were followed for a mean of 24.8 months. Slightly over half of the patients achieved a final best-corrected acuity of >=20/40. Independent risk factors predictive of a poor final acuity included: age <5 years, retrolimbal involvement, blunt injury causing rupture, would length >=6mm, vitreous hemorrhage, and retinal detachment. Sex, lens rupture, iris prolapse, vitreous in the anterior chamber, need for cataract surgery, and IOL implantation were not found to be predictive of

visual outcome. There was a significant difference between predicted final visual outcomes when comparing OTS and POTS. POTS predicted lower final acuities. The POTS places less emphasis on initial visual acuity and more emphasis on patient age and wound location. In children inaccurate or absent visual acuities are more likely at initial evaluation. However, POTS variables lack the statistical evidence that OTS is based on. Amblyopia may also play a role in the final visual outcome of younger children, which is not a component of the trauma score. The authors comment that OTS, remains the most useful tool in assessing prognosis after pediatric globe injuries. Finally, the data on methods of injuries may not have the same distribution in other countries and is probably location-specific.

Protective Eyewear Use as Depicted in Children's Television Programs

Michael E. Korchak, Robert U. Glazier,Rebecca S. Slack, Michael W. Plankey, Hylton R. Mayer, J Pediatr Ophthalmol Strabismus 2013; 50:102-105 (March/April)

The study analyzes popular television programs to evaluate use of protective eye wear and depiction of eye injuries and found that for the most part children's television programs do not treat ocular injuries as seriously as it should be treated. There were several scenes where eye protection should have been used and was not. In addition, when eye injuries did occur (mostly chemical injuries), the consequences were not major and were often resolved by itself in the following scene. As many eye injuries can be prevented by better education, appropriate protective eyewear use and avoidance of risk factors, children's television programs are at a unique position to assist in this as many children watch an average of 3-5 hours of television a day. The study found that the use of protective eyewear and eye inuries are rarely shown on scripted children's television programs

Incidence of Retinal Hemorrhages in Abusive Head Trauma

Majida A. Gaffar, ; Debra Esernio-Jenssen, Sylvia R. Kodsi, J Pediatr Ophthalmol Strabismus 2013; 50:169-172 (March/April)

The authors did a retrospective review of 48 cases of abusive head trauma to determine whether there is a correlation between retinal hemorrhages and cases where the perpetrator was identified versus if the perpetrator was not identified. In addition, if the perpetrator was identified, the authors further evaluated if the perpetrator confessed versus when the perpetrator did not confess. The study concluded that there was a statistically significant difference if the perpetrator was identified (whether there was a confession or not) versus if there was no identified perpetrator.

Patterns of Retinal Hemorrhage Associated With Increased Intracranial Pressure in Children

Gil Binenbaum, David L. Rogers, Brian J. Forbes, Alex V. Levin
PEDIATRICS Vol. 132 No. 2 August 2013 pp. 430 -434.

This is a study that analyzes the incidence and patterns of retinal hemorrhages associated with non-traumatic raised intracranial pressure (> 20 cm H_2O). This incidence and pattern is compared to retinal hemorrhages noted in raised intracranial pressure seen in abusive head trauma.
One hundred children were evaluated prospectively and retrospectively (mean age: 12 years; range: 3–17 years). Mean opening pressure was 35 cm H_2O (range: 20–56 cm H_2O); 68 (68%) children had opening pressure >28 cm H_2O. The most frequent etiology was idiopathic intracranial hypertension (70%). Seventy-four children had papilledema. Sixteen children had retinal hemorrhages: 8 had superficial intraretinal peripapillary retinal hemorrhages adjacent to a swollen optic disc, and 8 had only splinter hemorrhages directly on a swollen disc. All had significantly elevated OP (mean: 42 cm H_2O).
In this study, only a small proportion of children with nontraumatic elevated ICP have retinal hemorrhages. When present, retinal hemorrhages are associated with markedly elevated OP, intraretinal, and invariably located adjacent to a swollen optic disc. This peripapillary pattern is distinct from the multilayered, widespread pattern of retinal hemorrhages in abusive head trauma. When retinal hemorrhages are numerous, multilayered, or not near a swollen optic disc (eg, elsewhere in the posterior pole or in the retinal periphery), increased ICP alone is unlikely to be the cause.

Epidemiology of infant ocular and periocular injuries from consumer products in the United States, 2001-2008

Chen AJ, Linakis JG, Mello MJ, and Greenberg PBJ AAPOS 17;3:239-242

Data for this study was derived from the National Electronic Injury Surveillance System. One hundred sample hospitals are representative of the estimated 5300 hospitals with 24-hour emergency departments in the United States. Over a seven-year period, there were an estimated 21,000 visits to US emergency departments by patients aged 0-12 months for consumer product related eye injuries. The majority (63%) involved infants 9-12 months old and there was a slight preponderance of males (54%). Chemical (46%) and household items (24%) were the most likely causative agents. Over ¾ of the injuries occurred in the home. The leading injuries were contusions and abrasions, followed by conjunctivitis. Patient acuity and long-term follow-up data were not determined. Also this data only represents emergency room visits., and may not represent across the board injury percentages.

Visual Outcomes after Blunt Ocular Trauma

Richard J. Blanch, Peter A. Good, Peter Shah, Jon R.B. Bisho *Ophthalmology* August 2013;120:1588-1591

This is a study from Birmingham, United Kingdom. The purpose of the study is to describe the prognosis and retinal location in patients presenting with acute traumatic maculopathy and extramacular retinal injuries. This is a retrospective, noninterventional case series. All patients presented with commotio retinae or sclopetaria retinae to the Birmingham Midland Eye Centre Eye Casualty from October 1, 2007, to February 23, 2011. The main outcome measures were assessed by visual acuity.
Conclusions: After macular injury, 26% of patients were left with a visual acuity of less than 20/30. Reduced visual acuity after extramacular commotio retinae may represent occult macular injury or previously undiagnosed visual impairment in the affected eye. Extramacular commotio occurs mostly in an inferotemporal to temporal location, consistent with direct trauma to the sclera overlying the injured retina.

Retinal haemorrhages and related findings in abusive and non-abusive head trauma: a systematic review

S A Maguire; P O Watts; A D Shaw; S Holden, et al.*Eye.* January 2013;27(1):28-36

An exhaustive and systematic review of the medical literature, 1950-2009, was conducted to find publications related to head trauma and specific retinal findings among children ages 0 to 10 years. The authors included only studies with confirmation of abuse (such as admission by a perpetrator, or by multidisciplinary assessment, not relying on medical findings alone) or of accidental injury leading to head trauma. Thirteen studies involving 908 children were analyzed for prevalence data of retinal hemorrhages. Overall, retinal hemorrhages were found in 78% of the 504 abusive head trauma cases vs 5% of the 404 accidental head trauma patients. In a child with head trauma and retinal hemorrhages, the odds ratio that this is abuse is 14.7 (95% confidence intervals 6.39, 33.62) and the probability of abuse is 91%. Retinal hemorrhages were bilateral in 83% of abuse cases compared with 8.3% in accidental cases. These statistics can be useful in the medico-legal arena. The authors believe a standardized ophthalmic examination record of all children with possible abusive head trauma would aid in future meta-analyses on this topic.

18. RETINA

Maturation of the Human Fovea: Correlation of Spectral-Domain Optical Coherence Tomography Findings with Histology Lejle Vajzovic, Anita E. Hendrickson, Rachelle V. O'Connel, Laura A. Clark et al. Am J Ophthal November 2012; 154(5): pg. 779-789.

This study attempts to correlate human foveal development with histologic specimens using spectral-domain optical coherence tomography for the first time. Measurements were taken in 22 premature infants, 30 term infants, 16 children and 1 adult. The study divided the results according to age of the child; seven phases existed starting with 20-32 weeks gestation through 6-16 years of age. Results showed a progressive increase in neurosensory retinal thickness across the 7 phases of development. Total neurosensory thickness was consistently least in the fovea and greatest in the periphery. NFL is consistently thin in the fovea and changes little with age. Correlation was also made between OCT findings and histopathologic comparison. This study found extreme immaturity of the outer retina before and after birth, especially in the fovea. At 13-15 months, the foveal inner segment/outer segment length begins to pass the periphery. By 5 years, the foveal inner segment and outer segment are adult like both in histology and OCT.

Histologic Development of the Human Fovea From Midgestation to Maturity Anita Hendrickson, Daniel Possin, Lejla Vajzovic, Cynthia A. Toth. Am J Ophthal November 2012; 154(5): pg.767-778.

This study describes the histologic development of the human central retina around birth and provides detailed histologic descriptions of the human central retina from midgestation to early adulthood. All histologic specimens were enucleated between 2- 8 hours after death of expired children or aborted fetuses. Results showed that after fetal week 25, outward displacement of the inner retinal layer begins to form a foveal pit. Pit formation begins shortly after fetal week 24-25 when the foveal avascular zone forms. By 13-15 months, pit formation is complete. In addition, dramatic changes occur in the outer retina after birth. Foveal ONL is made of only one cell layer of cones until after birth but jumps to 10-12 cones deep by 6-8 years. Cone density is 18,472 cones/mm2 at fetal week 22 and doubles at birth. By 15 months, it increases to 52,787 and by 3.8 years, it increases to 108,439. The human fovea develops over a long period of time.

Reproducibility of Optical Coherence Tomography Measurements in Children Irene Altemir, Victoria Pueyo, Noemi Elia, Vicente Polo, et al. Am J Ophthal January 2013, 155 (1)

This was a prospective cross-sectional study looking at the repeatability of OCT measurements of the RNFL and macular measurements in healthy pediatric eyes. 358 children were evaluated with a full ophthalmic examination and OCT (Humphrey-Zeiss). Only 100 children were included in the study. Age was average 9.15 years with the youngest being 6.22 years old. Twenty-nine of the

children demonstrated ocular motility anomalies. The OCT appeared to give highly reproducible measurements in children between ages 6-12 years of age.

Visual Outcome in Isoametropic Amblyopic Children with High Hyperopia and the Effect of Therapy on Retinal Thickness

Wuhe Chen, Jie Chen, Fang Zhang, Xixi Zhu, Fan Lu Am J Ophthal January 2013; 155(3): pg 536-543.

This study evaluated the the change in retinal thickness with isoametropic amblyopia. The criteria for selection included hyperopia greater than +5.00, interocular difference less than 1.00 diopter, amblyopia in both eyes, no previous treatment of amblyopia. 217 children were included in the study. Out of this group, only 27 could perform OCT (Zeiss-Humphrey) testing; they were compared to 31 normal vision control subjects. In Final visual acuity was negatively correlated with amount of hyperopia and positively correlated with length of treatment. OCT findings demonstrated that the outer ring macular and RNFL thickness were positively correlated with refraction. The initial vision did not correlate with thickness of the macular or RNFL. There was a slight thinning of the foveal thickness in the amblyopic group.

Normative Reference Ranges for the Retinal Nerve Fiber Layer, Macula, and Retinal Layer Thicknesses in Children

Susan E. Yanni, Jingyun Wang, Christina S. Cheng, Kelly I. Locke, et al. Am J Ophthal February 2013; 155(2): pg. 354-360.

This study evaluated 83 healthy children between the ages of 5- 15 years to try to establish normative peripapillary retinal nerve fiber layer thickness, macular thickness, and retinal layer thickness. The measurements were made with the SD OCT (Heidelberg Engineering). Average macular thickness among all children was 271.2 +/- 2 microns. This was similar to data achieved in adult patients. Mean peripapillary RNFL thickness was higher than normative data for adults, running 107.6+/- 1.2 microns. The outer segment layer was thinner in the children than adults, due to outer segments are shorter in children.

Clinical Phenotypes and Prognostic Full-Field Electroretinographic Findings in Stargardt Disease

Sarwar Zahid, Thiran Jayasundera, William Rhoades, Kari Branham et al. Am J Ophthal March 2013; 155(3): pg 465-473.

This retrospective review of 198 patients with Stargardt's Disease evaluated the relationship between clinical and full-field ERG findings with progressive loss of visual function. This study found that the majority of patients with Stargardt's do not exhibit progressive scotoma. A smaller subgroup did progress more quickly and they appeared to have significantly worse scotopic B-wave amplitudes at

presentation. In addition, there was a correlation between full field ERG values at presentation and clinical stage with respect to funduscopic examination.

Surgical Outcomes Following Repair of Traumatic Retinal Detachments in Cognitively Impaired Adolescents With Self-Injurious Behavior

Robert A. Sisk, W. Walker Motley, III, Michael B. Yang, Constance E. West, J Pediatr Ophthalmol Strabismus 2013;50:20-26 (Jan/Feb)

Children with severe intellectual disability and autism spectrum disorders are associated with self injurious behavior which can be triggered by ohysical or emotional pain, frustration and stress. These behaviors include head banging, striking the face, biting or scratching oneself. The authors review cases of patients who sustained retinal detachments from this self injurious behavior. All 9 patients (9 eyes) had successful reattachment of their retinas. Despite these initial successes, recurrent retinal detachments occurred due to proliferative vitreoretinopathy and continued self injurious behavior. In some cases, the recurrent retinal detachment occurred once the silicone oil was removed which needed to happen secondary to corneal decomposition or glaucoma.

Vitreous hemorrhage in children and adolescents in India

Rishi P, Rishi E, Gupta A, et al J AAPOS 2013;17:64-69

The authors report the etiologies, demographic profiles, clinical findings, treatment modalities, and treatment outcomes of vitreous hemorrhages (VH) in patients <18 years of age. This was a 10-year retrospective chart review. The study includes 261 eyes of 246 patients. The overwhelming majority (82%) were male. The most common cause was trauma (68.5%). Of the trauma cases, 2/3 were blunt trauma and 1/3 were penetrating trauma. Eales disease was the next most common cause of VH. Twenty-two of the 25 patients with Eales disease were male. The most common cause of VH in children under age 1 was retinopathy of prematurity. Visual outcome data was available for 223 eyes. Vision improved in 52.9% of eyes. Risk factors for severe visual loss included: male, age > 8 years, unilateral presentation, and surgical intervention. Blunt trauma cases did worse than penetrating trauma cases. Eales disease is endemic to south Asia and therefore more commonly reported in this study. This study was retrospective and took place in a tertiary care center. This might skew data. Interestingly, shaken baby syndrome is rarely reported in south Asia.

Evaluation of Retinal Nerve Fiber Layer Thickness in Patients With Anisometropic and Strabismic Amblyopia Using Optical Coherence Tomography

Ismail Ersan, Nazmi Zengin, Banu Bozkurt, Ahmet Özkagnici J Pediatr Ophthalmol Strabismus 2013; 50:113-117 (March/April)

Retinal nerve fiber layer thickness was evaluated in strabismic amblyopic eyes, anisometropic amblyopic eyes and control eyes using optic coherence tomography. OCT measurements found no significant difference in RNFL thickness between strabismic, anisometropic and control eyes; however, the RNFL thickness seemed to be related to refraction.

Macular and Retinal Nerve Fiber Layer Thickness in Unilateral Anisometropic or Strabismic Amblyopia

Dima Andalib, Alireza Javadzadeh, Reza Nabai, Yashar Amizadeh, J Pediatr Ophthalmol Strabismus 2013; 50:218-221 (July/August)

The purpose of the article is to compare macular and nerve fiber layer (NFL) thickness in amblyopic versus nonamblyopic eyes by optical coherence tomography (OCT). A prospective cross sectional descriptive study was performed. Fifty patients were evaluated and the macular thickness as well as the peripapillary nerve fiber layer thickness. In anisometropic amblyopia, the macula was found to be thicker. This was not the case in strabismic amblyopia. There was no significant difference in NFL thickness in strabismic or anisoometropic amblyopia.

Hypertensive Retinopathy in Severely Hypertensive Children: Demographic, Clinical, and Ophthalmoscopic Findings From a 30-Year British Cohort

Katie M. Williams, Anish N. Shah,; Danny Morrison, Manish D. Sinha, J Pediatr Ophthalmol Strabismus 2013; 50:222-228 (July/August)

A retrospective chart review of 53 British children with severely hypertensive disease with or without renovascualr disease was performed. 39 children received an ophthalmic exam; of these, only 7 had hypertensive retinopathy, 6 were severe disease. The trend was for children with renovascular disease to have more severe retinopathy but the difference was not significant. Also, children with hypertensive retinopathy had higher blood pressure than those without.

Comparison Of Anatomic And Functional Results After Retinotomy For Retinal Detachment In Pediatric And Adult Patients

Marcin Stopa, Jaroslaw Kociecki, Piotr Rakowicz, Anna Gotz-Wieckowska, Eur J Ophthalmol May June 2013; 23(3): 410 – 416

This was a retrospective, nonrandomized, interventional study involving 20 pediatric patients and 25 adult patients operated with vitrectomy and retinectomy due to retinal detachment. The study observed that in the pediatric population, reattachment was less (60%) than adults (88%), a higher number of further reoperations (p=0.008) was required and postoperative proliferative vitreoretinopathy was significantly more frequent (p=0.003). Statistically significant improvement in visual acuity was observed in adults (p<0.001) but not in children (p=0.360) due to large proportion of anatomic failure. The clinical features and prognosis for pediatric retinotomies and retinectomies are different from those for adults. Even though the overall visual acuity was worse in children than in adults, 25% of pediatric eyes achieved 5/50 or better vision.

Characteristics of Peripapillary Retinal Nerve Fiber Layer in Preterm Children Jingyun Wang, Rand Spencer, Joel N. Leffler, Eileen E. Birch. Am J Ophthal May 2012; 153 (5); pg. 850-855.

This study attempted to correlate the type and severity of peripapillary RNFL abnormalities seen in infants born less than 32 weeks gestation to normal full term controls. Twenty-five preterm children were enrolled in the study, all with regressed ROP or no ROP, normal appearing posterior poles on exam, and greater than 5 years of age. All patients were scanned with the same Spectralis FD-OCT. Results found that preterm infants had a global average thickness of RNFL lower than normal children (8%). The mean distribution profile was flatter in preterm infants. The peri-papillary RNFL termporal superior and nasal inferior sectors were thinner than normal children (9-13%). However, data from this study was collected from preterm children who underwent laser treatment (9/16) and with visual acuities ranging to as low as 20/80.

Normal reference ranges of optical coherence tomography parameters in childhood. Noemi Elia, Victoria Pueyo, Irene Altemir, Daniel Oros, Luis Emilio Pablo. Br J Ophthal 2012; 96: 665-670.

This study reported normal values for peripapillary retinal nerve fiber layers thickness and optic disc parameters in children by the application of OCT. This was an observational cross-sectional study. The age of the children was 6-13 years.358 children were included in the study. The Cirrus OCT was utilized. 96% of the children were Caucasian. Non-caucasian children were excluded from the study due to small sample size. The mean RNFL thickness was 98.46 mm. The distribution of the quadrants showed the double hump in the inferior quadrant followed by the superior, nasal, and temporal quadrants. The mean rim area was 1.59 mm2 and the mean disc area was 2.05 mm2. The average cup to disc ratio was .43. NO difference was found between boys and girls. This study provided normative data for the RNFL and optic nerve in the Caucasian group of children.

Topographic Correlation between β-Zone Parapapillary Atrophy and Retinal Nerve Fiber Layer Defect

Bum-Joo Cho, Ki Ho Park, *Ophthalmology* March 2013;120:528-534

This is a retrospective, cross-sectional study involving 128 eyes from 128 consecutive patients with primary open angle glaucoma (POAG) and a single localized retinal nerve fiber layer (RNFL) defect performed in Seoul, Korea. This involved the use of digital optic disc photographs involving topographic measurements of localized retinal nerve fiber layer (RNFL) and β-zone parapapillary atrophy (PPA). (This is very well-defined in figure 1, page 529.) The main outcome measures were angular location and angular extent of β-zone parapapillary atrophy and retinal nerve fiber layer defect, angular location of the point of maximal radial extent (PMRE) of the β-zone parapapillary atrophy and the β-zone parapapillary atrophy-to-disc ratio.
Conclusions: 64.1% of the 120 eyes with single localized retinal nerve fiber layer defect had β-zone parapapillary atrophy. The angular location of the retinal nerve fiber layer defect showed a linear correlation of those with the β-zone parapapillary atrophy. Comments: In primary open angle glaucoma a localized retinal nerve fiber layer defect is correlated spatially with β-zone of parapapillary atrophy. This study involved some new concepts including β-zone parapapillary atrophy. This is an extension of traditional analysis of cup-to-disc measurements utilizing digital photographs.

Peripheral Nonperfusion and Tractional Retinal Detachment Associated with Congenital Optic Nerve Anomalies

Michael J. Shapiro, Clement C. Chow, Michael P. Blair, Lawrence M. Kaufman, *Ophthalmoscopy* March 2013;120:607-615

This is a retrospective observational case series of 15 patients with congenital optic nerve anomalies referred for pediatric retinal consultation. Sixteen eyes of the 9 patients with optic nerve hypoplasia and 8 eyes of 6 patients had congenital optic nerve anomalies, including optic nerve coloboma, morning glory disc deformity and peripapillary staphyloma, were included.
All patients underwent examination under anesthesia. Wide angle retinal photographs and fluorescein angiograms were reviewed. The severity of nonperfusion was graded. The presence of fiber vascular proliferation, vitreous hemorrhage, and traction retinal detachment were documented. The anatomic outcome was also recorded.
Conclusions: Congenital optic nerve anomalies may be associated with peripheral retinal nonperfusion and secondary complications of vascular proliferation, vitreous hemorrhage, and traction retinal detachment. In this select group of patients and nonperfusion associated with optic nerve hypoplasia seem to be more severe and associated more frequently with secondary complications.

Peripheral retinal examination and eyes with optic nerve anomalies may identify nonperfusion or fibrovascular proliferation. The author suggests that laser treatment to the avascular retina may help prevent complications of proliferative retinopathy in the future.
Comments: This article strongly suggests the use of fluorescein angiograms to detail the peripheral area of the retinal nonperfusion or fibrovascular proliferation. These suggestions, if adopted by consensus with other investigation, may change the standard of care for patients with congenital anomalies of the optic nerve head.

Persistent Fetal Vasculature – Letter to the Editor, General Correspondence

Wisam J Muen, Clare Roberts, Mandeep S Sagoo, M. Ashwin Reddy
Ophthalmology September 2012;119:1944-1945

This letter to the editor comments on the article written by Lambert SR, Buckley EG, and Lenhart PD, et al. Congenital Fibrovascular Pupillary Membranes: Clinical and Histopathologic Findings, from *Ophthalmology* 2012; 119: 634-41.

THE AUTHORS OF THE LETTER report on a similar case with vascular anomalies of the iris, retina, and orbit in a 2 year old, white girl. Fluorescein angiography of this child highlighted the prominent iris vessels traversing the pupil. The left fundus revealed markedly tortuous retinal, arterial, and venous vessels. The macula and retina were otherwise normal without leakage of fluorescein angiography.
B-mode ultrasound showed no other structural abnormalities. The axial lengths were 21mm in the right eye and 22.2mm in the left eye. Despite the longer axial length of the left eye, the left eye had microcornea. No systemic abnormalities were reported.
Magnetic resonance angiogram (MRA) demonstrated “normal intracranial appearance with no evidence of vascular malformations.” There were abnormally enlarged vessels within the left orbit, particularly at the supramedial corner where a cystic mass was defined. The cystic mass demonstrated flow characteristics on magnetic resonance angiogram sequences. This cystic mass was closely related to the optic nerve but not involved. There was a second similar extraconal abnormality in the inferomedial orbit with an abnormal bridging vessel communicating with the more superior cystic lesion. The left superior and inferior ophthalmic veins were enlarged. The lesions had the features of lymphatic-arteriovenous malformation.
According to the authors of the letter, the presence of flow within the lesion suggested that the cystic mass had a vascular component that predominated. Magnetic resonance imaging of the contralateral right orbit was normal. A 24 month follow-up showed no progression or vascular complication.
The purpose of this letter was to underscore the importance of looking for an associated orbital malformation in patients with persistent fetal vasculature.

Cases of persistent fetal vasculature are not associated with orbital vascular malformations. The authors of the letter state that "our case represents a new combination of persistent fetal vasculature with iris, retinal, and orbital vascular anomalies." The authors emphasize the following: THE PRESENCE OF VASCULAR ABNORMALITIES (EITHER INSIDE THE EYE OR THE ORBIT) CAN BE ASSOCIATED WITH SYSTEMIC CONDITIONS IN PARTICULAR WYBURN-MASON'S AND KLIPPEL-TRENAUNAY-WEBER SYNDROMES. BOTH OF THESE SYNDROMES CAN CONTRIBUTE TO BLINDNESS. ORBITAL VASCULAR MALFORMATIONS MAY BE CONFINED TO THE ORBIT (AS THEY WERE IN THIS CASE) OR CAN EXTEND TO INTRACRANIAL COMPARTMENTS. THE AUTHORS OF THE ARTICLE STRESS THAT IT IS POSSIBLE THAT INTRACRANIAL OR INTRAORBITAL VASCULAR ABNORMALITIES CAN ACCOMPANY PERSISTENT FETAL VASCULATURE AND THESE CHILDREN NEED TO BE FOLLOWED FOR THESE CONDITIONS.

Three-Year Follow-up after Unilateral Subretinal Delivery of Adeno-Associated Virus in Patients with Leber Congenital Amaurosis Type 2

Francesco Testa, Albert M. Maguire, Settimio Rossi, Eric A. Pierce, *Ophthalmology* June 2013;120:1283-1291

This is a clinical trial involving 5 patients with Leber congenital amaurosis and 2 patients with RPE65 gene mutations. The aim of the study was to show the clinical data of long-term (3-year) follow-up of 5 patients affected by Leber congenital amaurosis type 2 (LCA2) treated with a single unilateral injection of adeno-associated virus AAV2-hRPE65v2. This study included 5 LCA2 patients with RPE65 mutations.
Methods: Subjects were evaluated before and after surgery at designated follow-ups visits by complete ophthalmic examination. Each evaluation included best-corrected visual acuity, kinetic visual acuity, nystagmus testing, and pupillary light reflex.
Conclusions: The 3 year follow-up on 5-patient Italian cohort involved in the LCA2 gene therapy clinical trial showed a stability of improvement in visual and retinal function that had been achieved a few months after treatment. Longitudinal data analysis showed that the maximum improvement was achieved within 6 months after treatment, and the visual improvement was stable up to the last observed time point (3 years).
Comment: Photographs on page 1288, 1289, along with visual field assessment showed stability in all eyes and in some cases improvement in the Goldman visual field. This report describes a 3 year follow-up of 5 Italian subjects enrolled in the RPE65 gene therapy clinical trial performed at Children's Hospital of Philadelphia in conjunction with Second University of Naples.

A Phenotype-Genotype Correlation Study of X-Linked Retinoschisis

Ajoy Vincent, Anthony G. Robson, Magella M. Neveu, Genevieve A. Wright, *Ophthalmology* July 2013;120:1454-1464

This is a retrospective, comparative study of 57 patients with molecularly confirmed X-linked retinoschisis at Moorfields Eye Hospital in London, United Kingdom. The purpose of the study is to compare the clinical phenotype in detail with electroretinographic parameters and X-linked retinoschisis (XLRS). The clinical and electrophysiologic data associated with different types of mutation of the RS1 gene were used as main outcome measures. Pattern electroretinography (PERG) and full field electroretinography (ERG), incorporating international standard recordings, were performed in 44 cases. Thirteen cases were tested using a simplified ERG protocol. This autofluorescence imaging and optical coherence tomography were performed in most cases.

Conclusions: There is a profound phenotypic variability in patients with X-linked retinoschisis. Most patients have abnormal ERG with inner retinal dysfunction. Generalized cone system dysfunction is commonly associated with an abnormal "On-Response" and less commonly with an "Off-Response". Mutations in the RS1 gene consistently caused eletronegative bright-flash electroretinogram responses.

Metastasis from Uveal Melanoma Associated with Congenital Ocular Melanocytosis A Matched Study

Arman Mashayekhi, Carol L. Shields, Jerry A. Shields, Neelema Sinha, *Ophthalmology* July 2013;120:1465-1468

This is a matched retrospective study of 57 patients with uveal melanoma associated with ocular melanocytosis. The purpose of this study was to determine the rate of metastasis resulting from uveal melanoma associated with congenital ocular melanocytosis (COM) and to compare it with the rate of metastasis resulting from uveal melanoma not associated with congenital ocular melanocytosis. This study was performed at the Wills Eye Institute in Philadelphia.

Each patient in the melanocytosis group was matched with 2 patients with uveal melanoma not associated with ocular melanocytosis (nonmelanocytosis group) for age, gender, location of anterior tumor margin, location of tumor epicenter, tumor basal diameter, and tumor thickness were measured.

Conclusions: In this matched study, patients with uveal melanoma associated with congenital ocular melanocytosis were twice as likely to have systemic metastasis compared with patients with uveal melanoma not associated with congenital ocular melanocytosis.

Comments: This study reinforces the importance of taking careful photo images of both the sclera as well as the fundus in children affected with ocular

melanocytosis. It also stresses the importance of following this patient throughout life.

Subclinical Macular Findings in Infants Screened for Retinopathy of Prematurity with Spectral-Domain Optical Coherence Tomography

Adam M. Dubis, C. Devika Subramaniam, Pooja Godara, Joseph Carroll, *Ophthalmology* August 2013;120:1665-1671

This is a prospective, observational case series of 49 prematurely born neonates born in Milwaukee, Wisconsin. Forty-nine infants were imaged using a handheld spectral-domain OCT. Images were acquired in nonsedated infants in the neonatal intensive care unit. Some patients were followed and reimaged over the course of several weeks.
Main Outcome Measures: This included determination of foveal retinal lamination, image analysis, and clinical observation.
Conclusions: Data from this study suggested that there is persistence of the inner retinal layers in premature infants regardless of what stage of ROP was present. Subclinical cystic macular edema is seen in premature infants; however, cystic macular edema does not appear to be correlated with the stage of retinopathy of prematurity. The study shows that hand-held spectral-domain OCT imaging is a viable technique for evaluating subclinical macular findings in premature infants.

Retinal nerve fiber layer and optic disc measurements by spectral domain OCT: normative values and associations in young adults

Y M Tariq; H Li G Burlutsky; P Mitchell *Eye.* December 2012; 26(12):1563-70.

The study purpose was to determine normative values and associations of retinal nerve fiber layer (RNFL) and optic disc parameters in normal eyes measured by spectral domain optical coherence tomography (OCT).
In a population-based study, 1521 students from Sydney, Australia of ages 16-19 were examined. Patients with visual acuity less than 20/40 were excluded. The average RNFL was found to be 99.4±9.6 μm. RNFL thickness was least for the temporal quadrant (69.9±11.2 μm), followed by the nasal (74.3±12.8 μm), superior (124.7±15.7 μm) and inferior (128.8±17.1 μm) quadrants. The mean disc area in this population was 1.98±0.38 mm^2 and the mean cup/disc ratio was 0.44±0.18. Multivariate-adjusted analyses showed that optic disc parameters varied according to gender and race. The RNFL was thinner with greater axial length (P<0.0001).
This study documents normative values for the RNFL thickness and other optic disc measures using Cirrus spectral domain OCT in young adults. Such data can help better delineate normal variation from pathological changes. The authors caution that normative values differ between Stratus and Cirrus OCT due to difference in the scanning pattern and optic disc and cup delineation algorithm.

Wide-field spectral-domain optical coherence tomography in patients and carriers of X-linked retinoschisis.

Gregori NZ, Lam BL, Gregori G, Ranganathan S, Stone EM, Morante A, Abukhalil F, Aroucha PR Ophthalmology. 2013 Jan;120(1):169-74

6 patients with X-linked retinoschisis and 3 carrier females hasd wide field OCT performed. All 6 of the affected male patients had macular schisis cavities on OCT; 55% had only central schisis while the rest had schisis that also extended outside the macula. None of the carriers had any schisis seen.
OCT is very useful for helping to diagnose X-linked retinoschisis in affected people but this study does not support its use for screening carriers.

Surgical Removal of an Atypical Macular Epiretinal Membrane in Neurofibromatosis Type 2: Clinicopathologic Correlation and Visual Outcome

Dennis P. Han, Melody Chin, Kenneth B. Simons, Daniel M. Albert, Arch Ophthalmol. 2012;130(10):1337-1339.

Epiretinal membranes, when located in the macula, can impair visual acuity. When an epiretinal membrane is discovered in a child, deprivation amblyopia often compounds the visual loss, making timely detection and treatment important. Macular epiretinal membranes (ERMs) are common manifestations in children with neurofibromatosis
type 2 (NF2). The ERMs in NF2 have been speculated to be hamartomatous in nature. This study is a case report of one child with NF-2 and a visually significant epiretinal membrane. The report describes the clinical and histopathologic findings of the epiretinal membrance. Interestingly, fluorescein angiography revealed and absent foveal avascular zone. Spectral domain optical coherence tomography confirmed the dense ERM. Histology revealed a highly cellular membrane with weak staining for glial fibrillary acidic protein. Improvement in visual acuity in the operated eye is likely limited secondary to the abnormal fovea.

Retinal Glioneuronal Hamartoma in Neurofibromatosis Type 1

Eleonora M. Lad, Jason R. Karamchandani, Deborah M. Alcorn, Darius M. Moshfeghi, et.al. Arch Ophthalmol. 2012;130(10):1335-1337.

Retinal tumors occasionally arise in patients with neurofibromatosis type 1 (NF1). There have been reports of astrocytic hamartomas, capillary hemangiomas, and combined hamartomas of the retina and retinal pigment epithelium (CHRRPE)—typically without pathologic confirmation of the diagnosis. This is a case report of a child with NF1 with an unusual retinal tumor, a glioneuronal hamartoma. The

child was born with buphthalmos and subsequently noted to have numerous plexiform neurofibromas of the eyelid and face in a trigeminal distribution and right sphenoid wing dysplasia. A diagnosis of neurofibromatosis type 1 was made. Glaucoma was treated with a Baerveldt valve and a retinal lesion was thought to be combined hamartoma of the retina and retinal pigment epithelium. The eye became blind and painful and was removed and examined. The uvea was thickened by a diffuse neurofibroma typical for eyes involved with NF1. The predominant spindle cells within the neurofibroma reacted positively with S-100 protein and microtubule-associated protein 2 but were negative for glial fibrillary acidic protein, CD56 (neural cell adhesion molecule—a marker of neurons, astrocytes, and nonmyelinating Schwann cells), neurofilament, and Ki-67. Scattered clusters of larger neuronal cells with comparatively more abundant cytoplasm and large round nuclei with prominent nucleoli were also present and were positive for synaptophysin, microtubule associated protein 2, and neurofilament.

Evaluation of Normal Human Foveal Development Using Optical Coherence Tomography and Histologic Examination

Adam M. Dubis, Deborah M. Costakos, C. Devika Subramaniam, Pooja Godara, et.al. Arch Ophthalmol. 2012;130(10):1291-1300.

From the work of Mintz-Hittner, among others, we know that there is post-natal maturation of the fovea. In this study the authors wish to assess outer retinal layer maturation during late gestation and early postnatal life using optical coherence tomography and histologic examination. Longitudinal imaging offers the opportunity to track postnatal foveal development among preterm infants in whom poor visual outcomes are anticipated or to follow up treatment outcomes in this population. The foveal avascular zone (FAZ) is characterized by increased cone density, and excavation of inner retinal neurons characterize. The most salient feature of the human fovea is the shallow pit that is left behind by the lateral displacement of the inner retinal layers.

Spectral-Domain Optical Coherence Tomographic Characteristics of Autosomal Recessive Isolated Foveal Hypoplasia

Norman SaffraSwati Agarwal, John Pei-Wen Chiang, Robert Masini, , et.al. Arch Ophthalmol. 2012;130(10):1324-1327.

Foveal hypoplasia, also referred to as foveal planum, is a congential condition that can be associated with other ocular abnormalities such as aniridia, albinism, microphthalmos, and achromatopsia. Isolated foveal hypoplasia (IFH) is an even rarer disorder, with similar clinical findings in the fovea. Patients with foveal hypoplasia exhibit nystagmus, poor visual acuity, absent or abnormal maculofoveal reflexes on ophthalmoscopy, and variable and incomplete filtering

of the choroidal fluorescence in the macular area on fluorescein angiography. No single hereditary pattern has been established for patients with IFH. The authors present the clinical and SD-OCT features of 5 affected family members with IFH who tested negative for mutations in the PAX-6 gene.

Bietti Crystalline Retinopathy: Report of Retinal Crystal Deposition in Male Adolescent Siblings

Bita Manzouri, Panagiotis I. Sergouniotis, Anthony G. Robson, Andrew R. Webster, et.al.Arch Ophthalmol.2012;130(11):1470-1473.

Bietti crystalline dystrophy (BCD) is a genetically determined disorder characterized by progressive chorioretinal degeneration, nyctalopia, visual field constriction, and multiple intraretinal yellow-white crystalline deposits. Most cases present after the second decade of life and legal blindness is common in the fifth to sixth decade. This report identifies 2 adolescent siblings, one 16 and the other 13 with this disease. Lipid crystals can be seen at the corneoscleral limbus. Retinal crystals are observed predominantly at the posterior pole and in the superficial and deep retinal layers. They are associated with multiple, sharply demarcated areas of atrophy in the retinal pigment epithelium and loss of choriocapillaris; the crystals are less obvious as the disease advances. Bietti crystalline dystrophy is inherited as an autosomal recessive trait and is associated with mutations in the CYP4V2 gene (cytochrome P450). The CYP4V2 gene maps to chromosome 4q35, is expressed in a wide variety of tissues, including retina and cornea. The gene product is a protein associated with fatty acid metabolism.

Characteristics of Myelinated Retinal Nerve Fiber Layer in Ophthalmic Imaging: Findings on Autofluorescence,Fluorescein Angiographic, Infrared, Optical Coherence Tomographic, and Red-free Images

Julie B. Shelton, Kathleen B. Digre, James Gilman, Judith E. A. Warner, et.al. JAMA Ophthalmol. 2013;131(1):107-109.

Myelinated nerve fibers are a striking but benign finding. Myelination of axons proceed from the optic chiasm to the optic nerve. The anatomy of the lamina cribosa provides a natural barrier to the myelin. Although myelinated retinal nerve fibers are considered congenital, acquired cases have been described as well as occasional regression of myelinateed fibers after optic nerve injury..Using the latest diagnostic techniques the authors describe the features of retinal myelinated nerve fibers seen on color fundus photos, infrared photos, Spectralis OCT and autofluorescence. The authors emphasize that it is important to describe the typical features of this benign entity to distinguish it from more serious diagnoses like cotton wool spots, branch retinal artery occlusion, retinal infiltrate and even retinoblastoma.

Rhegmatogenous Retinal Detachment and Bilateral Optic Disc Coloboma in Organoid Nevus Syndrome

J. Fernando Arevalo, Andres F. Lasave, Fernando A. Arevalo, Jerry A. Shields, JAMA Ophthalmol. 2013;131(1):111-113.

Organoid nevus syndrome is characterized by cutaneous sebaceous nevus of Jadassohn, seizures and mental retardation. The nevi are found typically on the scalp, back of the neck, retroauricular area or trunk.Eye findings are seen in up to 50% and include epibulbar dermoids, posterior scleral cartilage, and colobomas of lid, iris, ciliary body, optic nerve and retina. Patients with chorioretinal colobomas are predisposed to retinal detachments, up to 42%. The authors present the first case of a rhegmatogenous retinal detachment associated with optic dis colobomas and organoid nevus syndrome.

Phenotypic Conservation in Patients With X-Linked Retinitis Pigmentosa Caused by RPGR Mutations

Sarwar Zahid, Naheed Khan, Kari Branham, Mohammad Othman, et.al. JAMA Ophthalmol. 2013;131(8):1016-1025.

The purpose of this study was to evaluate the clinical findings in patients with X-linked retinitis pigmentosa with 13 distinct RPGR mutations and assess for phenotypic concordance or variability. This retrospective review, spanning 25 years, identified 42 such patients with a median follow up of 5.5 years. Clinical data assessed for concordance included visual acuity, Goldmann visual fields, and full-field electroretinography. Visual acuity and ERG phenotypes are concordant in some patients carrying identical mutations but assessment of Goldmann visual field phenotypes revealed stronger phenotypic conservation. Phenotypic concordance is important for establishing proper counseling of patients as well as for establishing accurate patient selection and efficacy monitoring in therapeutic trials.

Varicella-Zoster Virus–Associated Multifocal Chorioretinitis in 2 Boys

Christina Meenken, Aniki Rothova, JAMA Ophthalmol. 2013;131(7):969-970.

The authors report on 2 boys, one aged 17 years and the other, aged 14 years, with unilateral multifocal chorioretinitis that was proven to be of varicella zoster virus (VZV) origin by intraocular fluid analysis. VZV may cause severe posterior eye segment inflammation and represents a major cause of the clinical syndrome called acute retinal necrosis (ARN). Acute retinal necrosis is a sight-threatening condition and continues to have a poor visual outcome. Typically, visual acuity

rapidly deteriorates, and the retinal inflammation may progress within several days to a full-thickness necrosis affecting the entire retinal periphery and often late retinal detachment.

Association of Ocular and Oculodermal Melanocytosis With the Rate of Uveal Melanoma Metastasis Analysis of 7872 Consecutive Eyes

Carol L. Shields, Swathi Kaliki, Michael Livesey, BriannaWalker, et.al. JAMA Ophthalmol. 2013;131(8):993-1003.

The risk for metastases from uveal melanoma is greater in patients who have associated ocular melanocytosis. This could be ocular or oculodermal melanocytosis which is a congenital periocular dermal pigmentary disorder. Uveal melanoma can develop in 1/400 patients and the risk for metastases is 1.6 times greater than those patients with uveal melanoma who did not have melanocytosis.

19. RETINOBLASTOMA

Direct Cost of Retinoblastoma Treatment During the First Year of Comprehensive Therapy in China

Xunda Ji, Yi Xuan, MD; Jing Li, Junyang Zhao, et al J Pediatr Ophthalmol Strabismus 2012;49:353-358 (Nov/Dec)

This article focusing on the monetary and non monetary costs of retinoblastoma on a family during the first year of treatment. The authors breakdown the costs in terms of which treatment option is chosen - chemotherapy (systemic or intra-arterial), external beam radiation treatment, enucleation. The costs was found to be $9,422 +/- $3,809. This article speaks mainly of countries where resources for treatment of retinoblastoma are sparse. Furthermore, non monetary costs are high because there are limited areas where appropriate treatment can be performed and so families travel long distances and have to stay locally for long periods of time while the child is undergoing treatment. Reduction of this burden on the families can occur if better medical insurance systems are put in place and if there is a broad availability of medical expertise on pediatric tumor therapy.

Clinical Profile, Management, and Outcome of Retinoblastoma in Singapore

Fiona Pin Miao Lim, Shui Yen Soh, Jayant Venkatramani Iyer, Ah Moy Tan, et al J Pediatr Ophthalmol Strabismus 2013; 50:106-112 (March/April)

A review of the various clinical manifestations of retinoblastoma in Singapore. The article reviews 51 patients diagnosed with retinoblastoma collecting data such as laterality, genetics, presentations, disease severity, treatment and prognosis. The study found a comparable 5-year survival rate to international data but did find that the severity of disease was worse which made globe preservation rate lower than more developed countries. Better education to the public as well as healthcare professionals may help to improve the rate of globe preservation.

Differentiation in Retinoblastoma and Histopathological Risk Factors in Mexico

Ricardo Gómez-Martínez, María Elena Rosales-Gradilla, Noemí Silva-Padilla,, Adrián Corona-Macías, J Pediatr Ophthalmol Strabismus 2013; 50:174-177 (March/April)

This study describes histopathological risk factors (massive choroidal infiltration, postlaminar optic nerve invasion, tumor in optic nerve cut, scleral invasion, and involvement of orbital soft tissues) and differentiation of retinoblastoma using enucleated eyes. The study found that advanced age is associated with moderately differentiated tumors as well as increases the risk of having a tumor with histopathological risk factors.

Study On Clinical Therapeutic Effect Including Symptoms, Eye Preservation Rate, And Follow-Up Of 684 Children With Retinoblastoma

Dongsheng Huang, Yi Zhang, Weiling Zhang, Yizhou Wang
Eur J Ophthalmol July Aug 2013; 23(4): 532 – 538

A retrospective analysis was made of 684 children (885 eyes) with advanced retinoblastoma diagnosed in Beijing, China. The average age at first diagnosis was 2.2 ± 1.7 years with overall median age 1.91 years. Leucocoria was the most common sign at the initial diagnosis. Three percent had positive family history. 551 cases (80.57%, 723 eyes) were A-E stage and 81.47% (589/723) were D-E stages of retinoblastoma; extraocular stage was present in 101 cases (120 eyes, 14.76%); metastatic stage was present in 32 cases (44 eyes, 4.67%). Pathology diagnosis was performed in 494 cases of unilateral or bilateral enucleation; most cases were grade II (260), 10% (49) were grade IV. Total survival rate was 95.13%. Key factors of clinical treatment and long survival rate were diagnosis and treatment at the early stage with multidisciplinary methods.

Sporadic unilateral retinoblastoma or first sign of bilateral disease?

Petra Temming, Anja Viehmann, Eva Biewald, Dietmar Lohmann. Br J Ophthal 2013; 97: 475-480.

This study looked at the clinical and genetic characteristics of children with sporadic unilateral retinoblastoma to see if factors could be identified for the development of metachronous bilateral disease. About 4% of children with initial monocular retinoblastoma will develop a new tumor in the other eye after treatment. Retrospectively, 480 records were reviewed. No child with a positive family history or treatment at another facility was included. The most decisive factor indicating high risk of second eye involvement was the presence of the oncogenic RB1 mutation in DNA of the blood. Early age at diagnosis was also considered high risk. In total, 9 (22.5%) of the 40 children with positive RB1 mutation developed bilateral retinoblastoma. None of the 155 children without a mutation in the blood developed bilateral disease.

Retinoblastoma presenting with orbital cellulitis

Walinjkar J, Krishnakumar S, Gopal L, et alJ AAPOS 17:3;282-286

In developing nations, an orbital cellulitis-like presentation of retinoblastoma is more common. Over a three-year period, retinoblastoma-associated cellulitis was seen in 14/260 (5.39%) patients in this article out of India. All had large intraocular tumors and showed endophytic growth. The authors excluded 4 patients for having received neoadjuvant chemotherapy, and another patient was excluded because the cellulitis was not the initial presentation sign. All of the remaining patients were enucleated, but 5 received pre-enucleation systemic steroids. This produced a marked reduction in inflammation. The reduced inflammation facilitated the enucleation procedure, by reducing vascularity and soft-tissue edema. No direct comparison was performed with those patients who did not receive systemic steroids.

Ophthalmic Artery Chemosurgery for Retinoblastoma Prevents New Intraocular Tumors

David H. Abramson, Jasmine H. Francis, Ira J. Dunkel, Y. Pierre Gobin, *Ophthalmology* March 2013;120:560-565

This is a single-center retrospective review of all genetic retinoblastoma cases managed at Memorial Sloan-Kettering Cancer Center/Weil-Cornell Medical School since May 2006.
This study evaluates 81 patients (80 with bilateral disease and 1 with unilateral disease) with genetic retinoblastoma. There were a total of 116 eyes treated with ophthalmic artery chemosurgery (OAC) since May 2006.
Results: 41 eyes were treated primarily with ophthalmic artery chemosurgery and 75 eyes were treated with ophthalmic artery chemosurgery after prior treatment with systemic chemotherapy, external beam radiation, or both.
Conclusions: The eyes receiving ophthalmic artery chemosurgery demonstrated fewer, new intraocular retinoblastomas after radiation or systemic chemotherapy than has been reported in the literature. This suggests that ophthalmoscopically

the undetectable tumors are present at the initial diagnosis are eliminated as a result of ophthalmic artery chemosurgery.

High-Risk Retinoblastoma based on International Classification of Retinoblastoma: Analysis of 519 Enucleated Eyes

Swathi Kaliki, Carol L. Shields, Jerry Shields, Ralph C. Eagle, Jr., *Ophthalmology* May 2013;120:997-1003

This is a retrospective study of 519 patients who underwent primary enucleation for retinoblastoma. The purpose of the study is to determine the correlation between the International Classification of Retinoblastoma (ICRB) and histopathologic high-risk retinoblastoma.
Conclusions: On the basis of the International Classification of Retinoblastoma, 17% of group D and 24% of group E eyes are at increased risk for metastatic disease. In this study, 8% of patients developed metastasis. There was no metastasis in any patient classified with a non-high-risk retinoblastoma.
Comments: Group D and group E eyes have the most severe pathology and therefore are at an increased risk for metastatic disease. There are excellent pictures on page 999 and also on the cover of May 2013, volume 120, number 5.

Retinoblastoma frontiers with intravenous, intra-arterial, periocular, and intravitreal chemotherapy

C L Shields; E M Fulco; J D Arias; C Alarcon; et al.*Eye*. February 2013;27(2):253-64.

The authors explore retinoblastoma diagnostic accuracy in this retrospective review from the Shields' oncology practice at Wills Eye Hospital. Among 2775 patients referred for management of retinoblastoma, 78% had confirmed retinoblastoma and 22% had simulating lesions. Simulating lesions among children ≤2 years old were persistent fetal vasculature, Coats disease, and vitreous hemorrhage, whereas those >5 years showed simulators of Coats, toxocariasis, and familial exudative vitreoretinopathy.

They also review retinoblastoma treatment. The treatment strategy depends on tumor laterality and stage of disease. In bilateral retinoblastoma, intravenous chemotherapy is important as first-line therapy for control of intraocular disease, prevention of metastasis, and reduction in pinealoblastoma and long-term secondary malignancies. Bilateral groups D and E retinoblastoma receive additional subtenon's carboplatin boost for improved local control. If unilateral disease is present, then intra-arterial chemotherapy is often considered. Intra-arterial chemotherapy can also be salvage therapy following chemoreduction failure. Unilateral retinoblastoma of groups D and E are managed with enucleation or globe-conserving intravenous chemotherapy and/or intra-arterial chemotherapy. Intravitreal chemotherapy is cautiously reserved for recurrent vitreous seeds following other therapies.

Hand-held high-resolution spectral domain optical coherence tomography in retinoblastoma: clinical and morphologic considerations.

Rootman DB, Gonzalez E, Mallipatna A, Vandenhoven C, et al
Br J Ophthalmol. 2013 Jan;97(1):59-65.

The authors report on use of both handheld OCT and conventional upright OCT in following retinoblastomas in 16 patients. 22 lesions were imaged. Small lesions were imaged in five cases, all of which were localised to the middle retinal layers. Clinical uses for handheld OCT imaging identified included: diagnosis of new lesions, monitoring response to laser therapy and the identification of edge recurrences. While OCT does not replace indirect ophthalmoscopy, it is an important addition to the armamentarium and offers advantages for detecting very early lesions and recurrence at the edge of lesions.

Intravitreal Melphalan for Refractory or Recurrent Vitreous Seeding From Retinoblastoma

Fariba Ghassemi, Carol L. Shields, Arch Ophthalmol. 2012;130(10):1268-1271.

Chemoreduction, with vincristine, etoposide, and cisplatin, has improved the globe salvage rates, especially in ICRB class D and E. When there is recurrent disease in the form of viable tumor cells collecting in the form of vitreous seeds, there are fewer options. The risk for secondary tumors outside the field of radiation is well known with external beam treatment. The authors propose to evaluate the efficacy and complications of intravitreal chemotherapy, specifically melphalan, for viable vitreous seeding from retinoblastoma. Enucleation would be the only other option in these 12 patients. Dose of melphalan appears to be important with regard to incidence and severity of complications. No serious complications occurred in the group which received 8-10 micrograms, but 4/7 patients had recurrence of vitreous seeds at greater than 6 month follow-up. In the group which received 50 micrograms, 100% achieved long term eradication of vitreous seeds, but complications included cataract, vitreous hemorrhage, subretinal hemorrhage, severe hypotonia, and phthisis leading to enucleation in 2 cases.

Thrombophilia in Patients With Retinoblastoma Receiving Ophthalmic Artery Chemosurgery

Jasmine H. Francis, Y. Pierre Gobin, Aaron Nagiel, Ira J. Dunkel, et.al. Arch Ophthalmol. 2012;130(12):1605-1608.

To prevent thrombus formation with subsequent embolus in patients with retinoblastoma who are treated with ophthalmic artery chemosurgery (OAC), the authors recommend iv heparin at the onset of the procedure and titrated to achieve an activated coagulation time (ACT) of 200-300 seconds (2-3x baseline). The femoral artery sheath is also flushed with heparinized saline.Three patients are described, one with a known predisposition to thrombophilia identified on review of family medial history and two additional patients were discovered after an adverse event incurred after OAC. Risk factors for thrombus formation in retinoblastoma patients who are to undergo OAC include: 15% of the population carries a heritable factor predisposing to thrombophilia or antiphospholipid syndrome,thrombosis formation is more common in patients with malignancy, the procedure itself increase risk of thrombus formation and the chemotherapeutic agent can incite thrombus.

Blue Toe Syndrome as a Complication of Intra-arterial Chemotherapy for Retinoblastoma

Ahmet Sarici, Osman Kizilkilic, Tiraje Celkan, Safa Gode, JAMA Ophthalmol. 2013;131(6):801-802.

Intraarterial chemotherapy has become more popular to treat advanced retinoblastoma. However, severe ocular vascular adverse effects, including ophthalmic artery stenosis and retinal arteriolar embolization have been reported. The authors report a case of extraocular vascular occlusion in a 7 month old infant with retinoblastoma who was treated with intraarterial melphalan. Blue toe syndrome is defined as the sudden onset of acute pain and cyanosis in one or more toes with evidence of a proximal source of emboli, primarily in the femoral or popliteal arteries. The patient was given both oral aspirin and parenteral low-molecular-weight heparin. After 1 week, the blue color and tenderness disappeared.

Superselective Intraophthalmic Artery Chemotherapy in a Nonhuman Primate Model Histopathologic Findings

Brian C. Tse, Jena J. Steinle, Dianna Johnson, Barrett G. Haik, et.al. JAMA Ophthalmol. 2013;131(7):903-911.

Superselective intraarterial chemotherapy(SSIAC) is a treatment for certain patients with retinoblastoma. No preclinical models have been studied to assess variables of flow and drug concentration in an attempt to mitigate ocular and orbital vascular toxicity. The authors studied 6 Macaque monkeys who underwent SSIAC with melphalan or carboplatin. Histopathology of the eye and orbit revealed widespread vascular damage. All contralateral eyes, which were not treated, were normal.

Retinal Vascular Precipitates During Administration

of Melphalan Into the Ophthalmic Artery

Rosanne Superstein, David Lederer, Josee Dubois, Patrick Hamel, et.al. JAMA Ophthalmol. 2013;131(7):963-965.

The authors describe real-time funduscopic findings during superselective intraarterial chemotherapy with melphalan in a 5 month old baby with bilateral retinoblastoma. Vaso-occlusive disease has been described a potentiallysight-threatening complication. The authors hypothesize that intraprocedural vaso-occlusive findings are the cause of vaso-occlusive disease reported by others as late findings. Real-time observation combined with titration of chemotherapy administration may prevent some of these late vaso-occlusive complications. Once intraarterial retinal precipitates or impaired retinal blood flow was detected, the infusion was aborted and findings normalized with time. Electroretinography and examination of retinal and choroidal blood flow performed two months after the procedure were normal and the tumors had regressed

20. ORBIT

The use of self-inflating hydrogel expanders in pediatric patients with congenital microphthalmia in China.

Hou Z, Yang Q, Chen T, et al. J AAPOS 2012;16:458-463

Hydrogel expanders are used in the treatment of congenital anophthalmia. The insertion is minimally invasive and the expander is biocompatible. They attract water from the surrounding tissue and are therefore self-expanding. The authors report their experience using these expanders over a two-year period in a retrospective fashion. There were spherical implants, hemispherical implants, and injectable pellets. There were different indications for which expander to implant (ex. Bony asymmetry, small conjunctival sacs, age of patient). Color Doppler sonograms were obtained 1 day, 3 days, 1 week and 1 month after insertion to assess volume and position of the expander.
There were 17 consecutive patients with unilateral microphthalmia. The average age was 48.4 months with an average follow-up of 21.8 months. There were no cases of infection. Inferior migration occurred in 1 patient, which required reoperation. Another patient removed the expander, which had to be replaced. All patients had subjective satisfactory appearance and were eventually able to wear prostheses. Horizontal palpebral fissure measurements increased from 71.3% to 85.4% that of the contralateral eye. Orbit size increased from 74.7% to 83.5% that of the contralateral eye. The authors feel that these expanders have many qualities that make them the ideal expander in cases of congenital microphthalmia. The fact that they are small when dry and expand after insertion, makes them easier to insert. Also the slow speed of their expansion limits the risk of tissue atrophy and migration or extrusion of the implant.

Management of persistent diplopia after surgical repair of orbital fractures

Loba P, Kozakiewicz M, Nowakowska O, et al. J AAPOS 2012;16:548-553

The authors describe the management of patients with persistent diplopia after reconstructive surgery for orbital fractures with respect to the type of ocular motility impairment. Reconstructive surgery was performed with a mean delay of 29.36 days. 83% of patients had soft tissue or muscle herniation. A mean vertical deviation of 14.8 PD was found in the worst position of gaze. Patients were separated into 4 groups based on orthoptic measurements. 1.IR paresis 2.IR restriction 3.IO involvement 4.IR paresis + restriction group. A few patients did not fit into any of these groups (medial rectus restriction only (n=4) and associated cranial nerve palsy (n=2). Orthoptic reassessment 6-12 months postoperatively showed resolved diplopia in 15%; residual diplopia in extreme gaze in 43.4%, persistent diplopia in 28.3%, and 13.3% were lost to follow-up. Of the persistent diplopia patients 14/17 underwent strabismus surgery. This study from Poland was associated with a greater delay until fracture repair than would typically be seen in the United States and thus this would change the post fracture repair results.

Digital Evaluation of Orbital Development in Chinese Children With Congenital Microphthalmia

Guang Yang, Jing Wang, Qunglin Chang, Zhenchang Wang et al. Am J Opthal September 2012: 154(3): pages 601-609.

This article evaluated the asymmetry of bilateral orbital development in Chinese children with congenital microphthalmia to determine the best timing for treatment and therapy. Thirty-eight pediatric patients of Chinese descent were enrolled with unilateral congenital microphthalmia. Seventy control patients were enrolled. Each patient was evaluated with a Brilliance 64-slice spiral CT scanner. Orbital volume, global volume, orbital widths were all measured. Results demonstrated significant differences between affected and unaffected orbits. In the control group, 69% of adult orbital volume is obtained by age 3 and the proportion rises to 75% by age 5. The affected orbits were smaller and the growth actually slows down or even stops by 3 years of age. The authors speculate that retardation of orbital development affects mainly the inferior and lateral rims. In this study, children with congenital microphthalmia, orbits of both sides grew equally until 3 years of age and then the abnormal side either slowed or stopped growing. Therefore, treatment for maximal orbital growth should occur shortly after birth and continued until 3 years of age

Improving ophthalmic outcomes in children with unilateral coronal synostosis by treatment with endoscopic strip

craniectomy and helmet therapy rather than fronto-orbital advancement

MacKinnon S, Proctor MR, Rogers GF, et al J AAPOS 17;3:259-265

This study compares long-term ophthalmic outcomes of two different surgical techniques for the treatment of children with unilateral coronal synostosis (UCS). A 7-year retrospective record review was performed of consecutive UCS patients, who had undergone endoscopic strip craniectomy (ESC), or fronto-orbital advancement (FOA). Both groups had a mean follow-up of over 21 months. Aniso-astigmatism mean was equivalent in the two groups but the FOA group had a greater standard deviation. FOA patients were more likely to have torticollis, amblyopia, more severe V-pattern and overelevation in adduction, and undergo surgical correction. Parents of children who presented before 4 months of age were given the option of either procedure. Those presenting after 4 months of age underwent FOA because the effectiveness of ESC after 4 months of age is questionable. Twenty-two patients underwent FOA and 21 underwent ESC and helmet therapy. The mean age at first visit was significantly older in the FOA group (9.3 months vs 4.0 months).
The authors correctly identify the fact that the disparate timing of surgical intervention in these two groups may have been a significant factor. In the FOA group, strabismus was more likely to have already been present. FOA is not generally recommended as a surgical procedure at a very young age because of greater operative morbidity and increased likelihood of recurrence. Also UCS may have produced greater orbital symmetry in the FOA group because it had more time to progress. FOA also separates the trochlea from the orbital wall, which could affect pattern strabismus as well. This study was retrospective and the two treatment groups were not randomized which limits comparison. The later intervention, rather than the surgical procedure, could explain the greater ocular morbidity seen in patients who underwent FOA. Early determination of the presence of UCS, as opposed to simple positional plagiocephaly allows earlier diagnosis and potential treatment. This alone, may improve ophthalmic outcomes.

A Quantitative Method for Assessing the Degree of Axial Proptosis in Relation to Orbital Tissue Involvement in Graves' Orbitopathy

Irene Campi, Guia M. Vannucchi, Andrea M. Minetti, Mario Salvi, *Ophthalmology* May 2013;120:1092-1098

This is a retrospective case study that included 50 patients and 29 control subjects who underwent orbital computed tomography (CT). The purpose of the study was to define a method of quantifying axial proptosis in patients with Graves' orbitopathy (GO) and to validate a score that correlates with the orbital

involvement and helps determine the degree of proptosis correction for elective orbital decompression.
The main outcome measures included diagrammatic triangulation of orbital CT. This is best explained on page 1094 of the article.
Conclusions: By measuring the ratio of intraorbital dimensions, the authors were able to quantify the degree of axial proptosis in patients with Graves' orbitopathy. Significant correlation of these measurements with some orbital parameters confirms that this parameter also may be used to measure orbital involvement (GO).
Comments: This is a technical and geometric construction to be used as an alternative to clinical exophthalmometer. This also allows you to look carefully at the size and structure of the extraocular muscles.

Pediatric Anophthalmic Sockets and Orbital Implants Outcomes with Polymer-Coated Implants

Maria Kirzhner, Yevgeniy Shildkrot, Barrett G. Haik, Ibrahim Qaddoumi, *Ophthalmology* June 2013; 120:1300-1304

This is a retrospective, interventional, cohort study designed to compare wrapped and
polymer-coated hydroxyapatite implants in children undergoing primary enucleation with no adjuvant therapies. Outcome measures including implant exposure, extrusion and migration, socket contracture, and formation of pyogenic granuloma.
Conclusions: The use of polymer-coated hydroxyapatite implants is associated with favorable outcomes in the pediatric population. Long-term implant retention is possible in most children.

Orbital Bands in Gomez-Lopez-Hernandez Syndrome

Michel J. Belliveau, Brian W. Arthur, Arch Ophthalmol. 2012;130(11):1496-1497.

Orbital bands are anomalous bands of muscle tissue found between recti muscles or between a rectus muscle and globe. MRI has identified such supernumerary extraocular muscles in 0.8% of orthotropic patients and 2.8% of strabismic patients. The authors present a case report of a child with Gomez-Lopez-Hernandez syndrome, also known as cerebellotrigeminal dermal dyslplasia (OMIM 601853) with orbital bands and A-pattern ET. Given the posterior location of these bands, which may cause restrictive strabismus and be difficult to access surgically, the authors emphasize that stable post-op alignment may be difficult to achieve.

Multiply Recurrent Solitary Fibrous Tumor of the Orbit Without Malignant Degeneration: A 45-Year Clinicopathologic Case

Study

Gregory J. Griepentrog, Gerald J. Harris, Eduardo V. Zambrano, JAMA Ophthalmol. 2013;131(2):265-267

Solitary fibrous tumor (SFT) is a rare mesenchymal spindle cell neoplasm originally described in the pleura and subsequently identified in a number of extrathoracic sites. More than 100 cases of solitary fibrous tumor of the orbit have been reported or reclassified with that diagnosis. Only 4 patients were younger than 10 years when first diagnosed as having orbital SFT. While most reports describe a benign clinical course, there are rare cases of primary malignant orbital SFT. Complete initial resection is a critical prognostic factor in preventing malignant degeneration. The authors describe a patient with orbital SFT whose proptosis was first recognized at age 9 years and who underwent surgical excision at various institutions at ages 12, 22, and 52 years. To their knowledge, this represents the longest histopathologically documented follow-up of a patient with orbital SFT.

Prenatal Presentation of Fronto-orbital Congenital Infantile Fibrosarcoma: A Clinicopathologic Report

Hughie H. Tsang, Peter J. Dolman, Douglas J. Courtemanche, Shahrad Rod Rassekh, et.al. JAMA Ophthalmol. 2013;131(7):965-967.

The authors present a patient who was diagnosed prenatally with congenital infantile fibrosarcoma (CIFS), a mesenchymal spindle tumor which typically occurs in the first year of life. Forty percent of cases are diagnosed at birth or in utero, mainly affecting the extremities. The author's patient presented with the tumor involving the orbit and frontal bone. The tumor was excised with vision sparing surgery. The local recurrence rate is 40% to 50%. The differential diagnosis of a mass in this location include teratoma, rhabdomyosarcoma, neuroblastoma, and granulocytic sarcoma, a manifestation of leukemia.

21. OCULOPLASTICS

Computed tomographic dacryocystography in children undergoing balloon dacryoplasty.

Limongi RM, Magacho L, Matayoshi S, et al. J AAPOS 2012;16:464-467

This study evaluated the volume and circumference of the lacrimal sac and the nasolacrimal duct in children with congenital nasolacrimal duct obstructions (CNLDO), using CT scanning with multidetectors (CT-MD), and digital subtraction dacryocystography (DCG). This was performed before and after balloon dilation. This was a prospective, interventional, case series, of enrolled children between ages 2 and 6 followed for at least 1 year post-op. 18

nasolacrimal ducts of 13 children were included. There was only 1 unsuccessful procedure. The difference in contrast volume before (0.12 +/- 0.08 cm^3) and after (0.07 +/- 0.06 cm^3) the procedure was correlated with success. The size of the most dilated portion of the lacrimal sac was also correlated with success. Lacrimal sac expression, age in months, and difference in contrast volume in percentage, were not found to be correlated with success. The data shows that untreated CNLDO may lead to dilation of the nasolacrimal duct and sac over time. The reduction in contrast volume after the procedure may reflect the release of the 'bottleneck' inside the duct postoperatively. This study was unmasked, small, and had no control group. Also children were exposed to radiation. The amount of radiation was less than that of a routine CT scan of cranial bones, but based on recent literature on the effects of CT scans in children, the authors contention that this procedure (CT-MD and DCG) is useful for complicated cases of CNLDO must be evaluated in terms of risk versus benefit carefully.

Efficiency Of Endoscopic Imaging In Repetitive Probing Following Unsuccessful Probing

Seydi Okumus, Bülent Gürler, Erol Coskun, Cengiz Durucu, Eur J Ophthalmol November – December 2012; 22(6): 882 - 889

Analysis of previous probing failures of 84 eyes with congenital nasolacrimal duct obstruction was done with intranasal endoscopic imaging. The eyes were divided into three groups based on the numbers of unsuccessful attempts with probing (group 1 = 1 attempt, group 2 = 2 attempts and group 3 = 3 attempts). All cases underwent nasal endoscopic-assisted probing and were followed up on the first week, first month, and third month following the procedure. The most common causes of failure were probe progression on the nasal wall in 33.3%, thick membrane and fibrosis in 21.4% different exit location in the nose in 20.2%. The rarest were nasal polyposis in 1.2% and mucocele in 2.4%. The success rate of endoscopic imaging was higher in the group with the fewest previous probings (group 1 = 86%, group 2 = 69.5% and group 3 = 54.5%). Nasal endoscopy not only provides information about the cause of probing failure, it also seem to decrease the failure rate of repeated probings

Treatment of Infantile Capillary Hemangioma of the Eyelid with Systemic Propanolol

Patrizia Vassall, Raimondo Forte, Antonio Di Mezza, Adriano Magli Am J Ophthal January 2013; 155(1): pg 165-170.

This prospective study evaluated the efficacy of systemic propanolol in 17 patients with eyelid infantile capillary hemangiomas. The main inclusion criteria was presence of a hemangioma that could obstruct vision or induce amblyopia. Children with asthma, obstructive pulmonary disease, cardiac disease or progressive intraconal lesion. All children received a dose of 2 mg/kg body weight per day. Size was monitored by B scan, pictures, and clinical exams. This study was blinded for evaluation of response to therapy by two different

ophthalmologists. Mean treatment duration was 2.5 +/- 1.3 months. Complete regression was seen in 9 of the 10 patients which were younger than than 1 year. Children over 1 year of age appeared to respond to treatment but had less of an affect. Age at the onset of treatment may influence final outcome. It is felt proliferative lesions early on are more susceptible to propranolol.

Amblyopia in Childhood Eyelid Ptosis Gregory J. Griepentrog, Nancy Diehl, Brian Mohney. Am J Ophthal June 2013;155(6); pg. 1125-1128.

This retrospective study evaluated the prevelance of amblyopia among children with ptosis over a 40 year period. Sixteen (14.9%) of the 107 patients were diagnosed with amblyopia. All cases of amblyopia occurred in patients with unilateral ptosis with the exception of one. Twelve (14.8%) of the 81 patients with simple congenital ptosis were diagnosed with amblyopia of which 7 (8.6%) cases were the result of occlusion of the visual axis from the ptotic lid. In conclusion, amblyopia affects 1 in 7 patients diagnosed with childhood ptosis.

Intraoperative Prognostic Factors For Probing Outcome In Children With Congenital Nasolacrimal Duct Obstruction Dima Andalib, Reza Nabei Eur J Ophthalmol May June 2013; 23(3): 329 – 332

The authors evaluated the intraoperative prognostic factors for probing outcome in children with congenital nasolacrimal duct obstruction (CNLDO). Prospectively, 69 eyes of 60 children aged 12 to 24 months underwent probing and irrigation by a single oculoplastic surgeon. The surgeon classified the obstruction (simple or complex) and the fluency of irrigation after probing (easy or difficult). There was a significant difference in success rate of probing in eyes with simple obstruction (87.8%) compared with complex obstruction (65%) ($p=0.02$). Also, there was a significant difference in success rate of probing in eyes with easy irrigation (100%) compared with difficult irrigation (53.6%) ($p<0.001$). Probing was more successful in eyes with simple obstruction or easy irrigation in children aged 12 to 24 months. The fluency of irrigation as an objective finding was a reliable intraoperative prognostic factor for probing success.

Clinical And Demographic Characteristics Of Ptosis In Children: A National Tertiary Hospital Study
Rania El Essawy, Mohamed A. Elsada Eur J Ophthalmol May June 2013; 23(3): 356 – 360

A retrospective review of 408 eyes in 336 children with blepharoptosis was conducted in Cairo, Egypt. The mean age at presentation was 3.2 years. The ptosis was unilateral in 65% of the cases, 74% of which involved the left eye. A positive family history was elicited in 19% of the cases. The commonest type was congenital (69%) followed by blepharophimosis syndrome (17%). Frontalis

suspension was the most common surgery (58%) followed by anterior approach levator muscle resection (29%) and Whitnall sling procedure (13%). The mean number of operations performed was 1.5 (range 1-4). Associated strabismus, refractive errors, and amblyopia should be considered for proper management of these patients.

Anisometropia and amblyopia in nasolacrimal duct obstruction

Kipp MA, Kipp MA and Struthers W J AAPOS 17;3:235-238

This paper was a 10-year retrospective review of children age 0-6 years with a diagnosis of nasolacrimal duct obstruction (NLDO). 887/1218 (72.8%) of identified cases were unilateral. Almost 300 patients were excluded for unsatisfactory clinical data. The rate of aniosmetropia was more than double (7.6% versus 3.6%) when comparing the unilateral NLDO cases with the bilateral cases. This difference was statistically significant. The greater hyperopia was found on the side with the NLDO in a statistically significant percentage of the cases (85%). Also the NLDO cohort as a whole showed a greater rate of anisometropia than the general pediatric population.
This study supports complete eye examinations, including cycloplegic refraction for all pediatric patients who present with a NLDO. These children have a higher risk of anisometropia and amblyopia. This does not appear to be a coincident finding alone; rather the NLDO appears to contribute to anisometropia.

Nasolacrimal Duct Obstruction Resolution

Caroline J. MacEwen Arch Ophthalmol. 2012;130(12):1623-1624.

Dr. MacEwen comments on the PEDIG study, which notes that 66% of congenital NLDO diagnosed between 6 – 10 months of age, resolve without surgery within 6 months. She cites 2 additional studies, which corroborate their findings. She adds that the site and complexity of the obstruction contribute more to the success or failure of treatment than age. She concludes that nasal endoscopy is useful in determining such anatomic anomalies.

A Randomized Trial Comparing the Cost-effectiveness of 2 Approaches for Treating Unilateral Nasolacrimal Duct Obstruction

Pediatric Eye Disease Investigator Group* Arch Ophthalmol. 2012;130(12):1525-1533.

To compare the cost-effectiveness of 2 approaches of treating unilateral nasolacrimal duct obstruction. One hundred sixty three infants 6 months to less than 10 months of age with unilateral congenital NLDO were randomized to

immediate office based probing or observation/non-surgical management for 6 months, followed by probing in the operating room for persistent symptoms. Treatment success was defined as absence of clinical signs of NLDO (epiphora, full tear lake, mucous discharge) as assessed by masked exam at age 18 months. Cost was determined from time of randomization to age 18 months, and included cost of surgical procedures and medicines. In the observation group, NLDO signs resolved in 66% within 6 months of enrollment. 92% of the immediate probing group were considered successful at 18 months of age, compared with 82% of the observation/deferred probing group. Immediate office probing was likely more cost effective than the deferred surgery group with the mean cost at $562 in the immediate office probing group compared with $701 in the observation/deferred probing group. However the mean cost is influenced by choice of antibiotic, which varies widely in cost, (e.g. Polytrim vs Vigamox) and whether surgery is performed in an ambulatory care center versus a hospital based operating room. The authors also emphasized that early probing in the office would potentially treat two thirds of infants who would have resolved their signs of NLDO spontaneously.

Positional Change of Lower Eyelid After Surgical Correction of Congenital Ptosis in the Korean Population

Chang Yeom Kim, Su Yan Zhao, Cheng Zhe Wu, Jin Sook Yoon, et.al. JAMA Ophthalmol. 2013;131(4):540-542.

The authors have found that some patients with congenital ptosis have preoperative lower scleral show that is diminished after ptosis surgery. This quantitative study investigates the positional change in the lower eyelid after surgery to correct congenital ptosis. Medical records and clinical photographs of 55 Korean patients with congenital ptosis who underwent corrective surgery were reviewed. Lower scleral show was found in 7 ptotic eyes (8.9%) preoperatively and disappeared in all but 1 eye postoperatively. This study shows lower eyelid elevation after surgical correction of congenital ptosis, especially after frontalis suspension or in bilateral ptosis. Surgeons should inform patients that lower eyelids can displace upward after ptosis surgery and that preoperative lower scleral show can be diminished postoperatively.

Failure of Systemic Propranolol Therapy for Choroidal Hemangioma of Sturge-Weber Syndrome: A Report of 2 Cases

Hatem Krema, Yacoub A. Yousef, Priya Durairaj, Ronaldo Santiago, JAMA Ophthalmol. 2013;131(5) 681-683.

Propranolol was incidentally discovered to induce accelerated involution of infantile cutaneous hemangioma Since that observation, propranolol has successfully been used in the treatment of cutaneous, orbital and ocular

hemangioma. The authors report the outcome of systemic propranolol in 2 patients with choroidal hemangioma and Sturge Weber syndrome. The first patient is a 14 year old girl with nevus flammeus of the left side of the face. She has 20/20 vision OD and HM vision OS. Ultrasonography OS revealed a diffuse choroidal mass, of medium to high internal reflectivity and thickness of 6.3mm causing an exudative retinal detachment. The second patient, a 22 year old man, had bilateral nevus flammeus and choroidal hemangioma with NLP vision OD and 20/300 vision OS. Ultrasound demonstrated a diffuse choroidal hemangioma OS with 2.9 mm thickness. Both patients were treated for 6 months with oral propranolol at a dose of 2 mg/kg/d. No change in the facial or choroidal hemangiomas was noted after 6 months of treatment. The authors postulate that failure of their patients to respond to propranolol may reflect the fact that choroidal hemangiomas have both cavernous and capillary components, which may affect treatment response.

22. INFECTIONS

External ophthalmoplegia in human immunodeficiency virus-infected patients receiving antiretroviral therapy

Pineles SL, Demer JL, Holland GN, et al. J AAPOS 2012;16:529-533

Highly active antiretroviral therapy (HAART) has decreased HIV mortality but increased iatrogenic syndromes, including skeletal muscle dysfunction. The authors discuss a series of HIV-infected patients who presented with bilateral external ophthalmoplegia and blepharoptosis. Medical records were retrospectively reviewed. Five patients aged 44 to 62 who had HIV infection for over a decade were included. The patients had bilateral blepharoptosis and symptoms of diplopia or abnormal pursuit, eccentric gaze, or strabismus. All patients had abnormally slow saccades but pursuits were normal. All 5 patients had abnormal T1-weighted high-resolution MRI of the orbits, with bright patchy signal in all four rectus muscles bilaterally. Extraocular muscle volume were conserved. These findings had only been seen previously in patients with CPEO. Patients were worked up for Kearns-Sayre or other mitochondrial pathology, and also for myasthenia gravis., with negative results. HAART therapy includes nucleoside reverse-transcriptase inhibitors and protease inhibitors, both of which can cause skeletal myopathy. This study was retrospective and subject to selection and referral bias. None of the patients underwent muscle biopsy.

Cat Scratch Neuroretinitis: The Role of Acute and Convalescent Titers for Diagnosis

Archit Gulati; Sushma Yalamanchili, Karl C Golnik, and Andrew G Lee, *J Neuro-Ophthalmol 2012; 32:243-245.*

Two cases of cat scratch neuroretinitis are presented in which the initial acute serological titres were negative or equivocal for Bartonella Henselae. The clinical setting supported treatment for cat scratch disease, and ultimately the convalescent titres confirmed the diagnosis. Although serologic testing is usually seen as the gold standard for initial diagnosis and treatment, the short time frame in which one can reliably expect confirmatory IgM or IgG titres, as well as the high incidence of high seropositivity for IgG titres in the population at large, makes reliance on one set of serological tests inadequate. The authors recommend obtaining convalescent titres two weeks after initial titres to better appraise the likelihood of the diagnosis, and initiating treatment regardless of the outcome of the initial titres when the index of suspicion is high. Indirect fluorescence assay (IFA) and enzyme immune assay (EIA) have low sensitivity profiles; laboratory testing with polymerase chain reaction (PCR) can be helpful in patients with altogether negative serological testing. In short, initiating treatment should be based on clinical suspicion.

Trends in ophthalmic manifestations of methicillin-resistant Staphylococcus aureus (MRSA) in a northern California pediatric population

Amato M, Pershing S, Walvick M and Tanaka S J AAPOS 17;3:243-247

Over an 8-year period, all pediatric cases of culture-positive ophthalmic MRSA were identified in a retrospective cross-sectional review of the largest managed-care healthcare system in Northern California. Over half (58%) of the 137 pediatric cases were community acquired. There was a trend of more new cases towards the end of the study period. The most common presentations were conjunctivitis (40%), stye/chalazion (25%), orbital cellulitis/abscess (19%), dacryocysitis (11%) and brow abscess (3%). Risk of ocular infection increased with male sex, neonates, and multiple infection sites on the body. There was high resistance to bacitracin and ofloxacin. Oral Trimethoprim.sulfamethoxazole showed low resistance, and was the predominant form of therapy, along with topical gentamicin. Resistance to antibiotics increased during the study period. Topical therapy was effective in 29%, oral antibiotics were effective in 47%. IV therapy was required in 12% and 19% required incision/drainage. There were no cases of permanent visual impairment. Unlike prior studies, diabetes was not found to be associated with an increased risk of MRSA infection. The authors recommend early recognition, proper antibiotic selection, and obtaining cultures & sensitivities when resistant or severe ocular infections are present.

Risk Factors for Cytomegalovirus Retinitis in Patients with Cytomegalovirus Viremia after Hematopoietic Stem Cell Transplantation

Sohee Jeon , Won Ki Lee , Yongeun Lee , et al *Ophthalmology* September 2012;119:1892-1898

This study comes from the Department of Ophthalmology, St. Mary's Hospital, Catholic University of Korea/College of Medicine, Seoul-South Korea.
This is a retrospective cohort study designed to evaluate the risk factors for cytomegalovirus retinitis (CMV) in patients with CMV Viremia after hematopoietic stem cell transplantation (ASCT). This study involved 708 patients who underwent HSCT. 363 (51%) developed CMV viremia after HSCT. Of the 363 patients with CMV viremia, 270 underwent retinal examination for CMV retinitis. CMV retinitis was detected in 15 of 270 patients with CMV viremia.
Conclusions: The development of CMV retinitis should be carefully monitored in patients with significant viral load, which is represented by a peak cytomegalovirus DNA level greater than 7.64×10^4 copies/ml or a long duration of cytomegalovirus viremia. This is particularly common in patients who received hematopoietic stem cell transplant from an unrelated or HLA-mismatched donor and showed delayed lymphocyte engraftment.

23. PEDIATRICS/INFANTILE DISEASE

Attention to eyes is present but in decline in 2–6-month-old infants later diagnosed with autism

Warren Jones and Ami Klin Nature November 6, 2013 [epub ahead of print]

This prospective longitudinal observational study evaluated eye fixation from birth to 36 months in 59 subjects at high risk for autism based on family history and in 51 at low risk for based on negative family history. Eye fixation was assessed in all subjects while viewing a video recording of a caregiver looking directly into the camera attempting to visually engage the subjects at months 2, 3, 4, 5, 6, 9, 12, 15, 18 and 24. Of the high-risk infants, 12 (20.3%) were found to have autism at age 36 months. One child from the low-risk cohort was also diagnosed with autism. When compared to those without autism, the subjects with autism demonstrated a statistically significant decline in eye fixation ($p=0.100$) between month 2 to 9. While these data may provide interesting insights and stimulate future research, no validated clinical test was presented or recommended for use by developmental pediatric specialists or ophthalmologists in order to facilitate making the diagnosis of autism in infancy.

24.SYSTEMIC/UVEITIS

Ocular Features in Joint Hypermobility Syndrome/Ehlers-Danlos Syndrome Hypermobility Type: A Clinical and In Vivo Confocal Microscopy Study

Magda Gharbiya, Antonietta Moramarco, Marco Castori, Francesco Parisi et al. Am J Ophthal Septmenber 2012; 154(3): pg. 593-600.

This paper investigates the ocular anomalies associated with Joint Hypermobility Syndrome. Twenty-two patients and 44 eyes were included in this study. Most commons symptom of these patients is dry eye with alterations in tear film stability and also deficiency. In addition, these patients seem to have steeper corneal curvature and higher best-fit sphere index compared to controls. Increased amount of myopia was not seen in this group of patients but a higher percentage of pathologic myopia was present. When pathologic myopia occurred, the vitreous showed fibrillar and beaded appearance. Also there was a decreased cell density in the superficial epithelium and increased stromal keratocytes. This may be due to the dry eye associated with this condition. Asymptomatic lenticular changes were noted in some patients as well including subcapsular lens opacities and fetal nuclei opacities. Ophthalmic exam may help differentiate these patients with milder phenotypes of this condition.

Do Children And Adolescents With Attention Deficit Hyperactivity Disorder Have Ocular Abnormalities?

Eedy Mezer, Tamara Wygnanski-Jaffe Eur J Ophthalmol November – December 2012; 22(6): 931 – 935

A group of 51 children diagnosed with ADHD (32) and ADD (19) were evaluated for visual function and ocular features. The average age was 9.9 years, (± 3.1). Amblyopia was found in 18%, of which the majority was due to ametropia. Strabismus was found in 10%, and absent stereoacuity was found in 6%. Convergence insufficiency was seen in only 4% in contrast to other reports. The refractive errors showed 43% had myopia (< - .50 or higher), hyperopia in 20% (> +3.50), and astigmatism in 20% >= to 1.0 D) Significant ametropia was detected in 42 (83%) of the patients.

Analysis of a Novel Protocol of Pulsed Intravenous Cyclophosphamide for Recalcitrant or Severe Ocular Inflammatory Disease

Ana M. Suelves, Cheryl A. Arcinue, Jesus Maria Gonzalez-Martin, Jonathan N. Kruh, *Ophthalmology* June 2013;120:1201-1209

This is a retrospective, interventional, noncomparative cohort study involving 110 eyes of 65 patients. The purpose of the study was to analyze the success rate of pulsed intravenous cyclophosphamide for noninfectious ocular inflammatory disease and to identify risk factors for failure of therapy. The study occurred in Cambridge, Massachusetts.
Conclusions: Pulsed intravenous cyclophosphamide, when applied according to protocol used by the researchers, results in an extremely high rate of sustained complete remission in patients with recalcitrant and fulminant, vision-threatening ocular inflammatory disease.

The authors describe an excellent safety profile in appropriately trained physicians. This also allows tapering and discontinuing corticosteroids in most patients. Early initiation of therapy may decrease the risk of relapses.

Ocular Manifestations of Xeroderma Pigmentosum Long-Term Follow-up Highlights the Role of DNA Repair in Protection from Sun Damage

Brian P. Brooks, Amy H. Thompson, Rachel J. Bishop, Kenneth H. Kraemer, *Ophthalmology* July 2013;120:1324-1336

This is a retrospective, observational, case series from the clinical center at the National Institute of Health, USA. The study involved 87 participants age 1 to 63 years of age from 1964 to 2011. Eighty-three patients had XP, 3 patients had XP/Cockayne syndrome complex, and 1 patient had XP/trichothiodystrophy complex.
All patients had appropriate ophthalmic examinations consistent with age and developmental stage. Ninety-one percent had at least 1 ocular abnormality. The most common abnormalities were conjunctivitis 51%, corneal neovascularization 44%, dry eye 38%, corneal scarring 26%, ectropion 25%, blepharitis 23%, conjunctival melanosis 20%, and cataracts 14%. Thirteen percent of patients had some degree of visual axis impingement. Five percent of patients had no light perception in one or both eyes. OCULAR SURFACE CANCER OR A HISTORY OF OCULAR SURFACE CANCER WAS PRESENT IN 10% OF PATIENTS. Patients with acute sun burning phenotype were less likely to develop conjunctival melanosis and ectropion but more likely to develop neoplastic ocular surface lesions than nonburning patients. Some patients showed signs of limbal stem cell deficiency.
Conclusions: This longitudinal study reported the ocular status of the largest group of patients systematically examined at 1 facility over an extended period of time. There was a high incidence of structural lid deformities, neoplasms of the ocular surface and eyelids, and inflammatory ocular surface disease. Corneal abnormalities were present in this population.
Burning and nonburning patients with XP exhibit different rates of important ophthalmologic findings including neoplasia. Ophthalmologic characteristics can help refine diagnoses in the case of XP complex phenotypes. DNA repair plays a major role in protection of the eye from sunlight-induced damage.
Comments: This is a must read landmark article for all ophthalmologists. Please see the impressive photographs on pages 1328 and 1329.

Tubulointerstitial Nephritis and Uveitis Syndrome in Children: A Prospective Multicenter Study

Ville Saarel, Matti Nuutinen, Marja Ala-Houhala, Pekka Arikoski, *Ophthalmology* July 2013;120:1476-1481

Tubulointerstitial Nephritis (TIN) is an inflammatory disease of the kidneys. It may be triggered by infectious diseases and numerous medications including non-steroidal anti-inflammatory drugs and antibiotics. However, in these children the cause remains unknown in the majority of cases. The condition may be accompanied by uveitis in which case it is referred to as tubulointerstitial nephritis and uveitis (TINU syndrome). Uveitis is typically anterior and bilateral.
This is a prospective, observational, multicenter, partly placebo-controlled treatment trial performed in Finland. Nineteen children with biopsy proven tubulointerstitial nephritis were evaluated. Clinical features and outcome of uveitis were the main outcome measures.
Conclusions: There is no statistically significant difference in the occurrence of uveitis in patients with tubulointerstitial nephritis in the prednisone and nontreatment groups. In this study the occurrence of uveitis associated with tubulointerstitial nephritis was considered higher than previously reported. UVEITIS RELATED TO TUBULOINTERSTITIAL NEPHRITIS MAY DEVELOP LATE AND IS OFTEN ASYMPTOMATIC . **The ophthalmologic follow-up of all patients with tubulointerstitial nephritis is warranted for at least 12 months starting with 3-month intervals.**

Prevalence of Oculo-auriculo-vertebral Spectrum in Dermolipoma

Jwu Jin Khong,Thomas G. Hardy, Alan A. McNab, *Ophthalmology* August 2013;120:1529-1532

This is a retrospective case series involving patients with primary presentation of ocular dermolipoma. The purpose of the study is to describe the observed frequency of oculo-auriculo-vertebral spectrum (OAVS) in patients with dermolipoma.
Conclusions: Dermolipoma is an independent ocular association of oculo-auriculo-vertebral spectrum (OAVS) that is more commonly observed than previously reported. It is an ocular feature in both the milder and more complex forms of the spectrum.
Comments: Oculo-auriculo-vertebral spectrum (OAVS) is a term first applied by Cohen to describe a phenotypically heterogenous disorder owing to developmental abnormality of the first and second branchial arches. This rare and complex condition is also known as hemifacial microsomia or Goldenhar's syndrome. The diagnosis is based on the presence of combination of features, including microtia, mandibular hypoplasia, anomalies of the cervical spine, and epibulbar dermoid or ocular dermolipoma, also known as lipodermoids.

Dermal Swellings and Ocular Injury after Exposure to Reindeer

Boris Kan; Domenico Otranto; Kristian Fossen; Kjetil Åsbakk *N Engl J Med*; December 2012; 367:2456-2457

The authors describe five children who had visited reindeer herding areas in Norway or Sweden and developed enlargement of occipital lymph nodes, and migratory 2-to-5-cm dermal swellings, including some involving the eyelids. Two children developed ophthalmomyiasis, and one eye lost vision. They were treated with ivermectin once the diagnosis was made of myiasis due to *Hypoderma tarandi,* a bumblebee-like fly that is common in subarctic regions and attaches eggs onto the hair of reindeer

Infantile glaucoma in Rubinstein–Taybi syndrome

J DaCosta; J Brookes *Eye.* September 2012;26(9):1270-1

Rubinstein-Taybi syndrome is characterized by a beaked nose, broad thumb, angulation deformity of the great toe, mental retardation and several ocular findings. The authors present three cases of infantile glaucoma in patients with Rubinstein-Taybi syndrome and review the literature. They stress the importance of measuring intraocular pressure in these patients, to help differentiate corneal clouding and glaucomatous optic atrophy from other types of corneal opacities and optic nerve anomalies found in these patients.

Anterior chamber infiltrate in 3-month-old: rare presentation of myeloid sarcoma

K E Dunbar; G B Hubbard III; J R Wells *Eye.* September 2012;26(9):1274-5

This is a single case report of a baby with bilateral fluffy white anterior chamber infiltrates who subsequently developed bilateral proptosis and lacrimal gland swelling. Lacrimal gland biopsy confirmed myeloid sarcoma. The initial differential diagnosis of anterior chamber infiltrates included retinoblastoma and hypopyon due to endophthalmitis

TYPE 1 NEUROFIBROMATOSIS: A GENO-OCULO-DERMATOLOGIC UPDATE

Rhonda E. SchnurCurrent Opinion in Ophthalmology Sept. 2012 23(5) p. 364-372

Type 1 neurofibromatosis (NF1) is an autosomal dominant geno-oculo-dermatosis caused by mutations or deletions in the neurofibromin gene on chromosome 17p11.2. The NF1 gene encodes neurofibromin. When neurofibromin is deficient, RAS-GTP remains more active and thereby promotes excess cell growth. The NF1 phenotype is caused by dysregulation of the mTOR (mammalian target of rapamycin) signal transduction pathway. The ophthalmologic exam is highly sensitive and specific in screening for NF1 and can distinguish NF1 from other disorders with overlapping clinical features. Optic pathway gliomas (OPGs) are a marker for cerebral arteriopathy and are

associated with decreased retinal nerve fiber thickness. Ciliary body cysts and abnormal retinal pigmentation are associated with NF1. Lisch nodules remain an important diagnostic marker for NF1 in older children and adults and suggest a correlation with underlying iris pigmentation and UV exposure. Effective therapy for NF1 will likely require combined targeting of the NF1 RAS/RHEB/mTOR signal transduction pathway as well as suppressing tumorigenic factors in the stromal microenvironment.

OPHTHALMOLOGY MANIFESTATIONS OF PEDIATRIC CANCER TREATMENT

Jonathon H. Salvin, Dorothy Hendriks Current Opinion in Ophthalmology Sept. 2012 23(5) p. 394-399

Advances in pediatric oncology care have increased survival rates for children with malignancy. As a result, ophthalmologists are seeing more short-term and long-term complications associated with treatment of these conditions. Ophthalmologists need to be aware of cancer treatment-related eye disorders. Multiple eye findings are associated with cancer treatment, including chemotherapy, radiation, bone marrow transplantation, and newer modalities such as intra-arterial chemotherapy. Malignancy and treatment cause immunodeficiency that can lead to infectious disease manifestations, including eye involvement. Paraneoplastic conditions may manifest with eye findings either before the diagnosis of the primary malignancy or as a late finding. The evolution of IVF has raised concerns of increased cancer risks, including ocular tumors. Ophthalmologists who are involved with the care of children undergoing cancer treatment need to be aware of these ocular sequelae. This article reviews the most common eye findings with each type oncologic treatment.

OPHTHALMIC MANIFESTATIONS OF SICKLE CELL DISEASE: UPDATE OF LATEST FINDINGS

Jennifer I. LimCurrent Opinion in Ophthalmology November 2012 23(6) p. 533-536

Recent developments in the diagnosis and management of sickle cell ocular manifestations are reviewed to enable the clinician to better manage the ophthalmic care of these patients. In addition, newer imaging modalities, such as spectral domain OCT and wide-field imaging, have resulted in the detection of subclinical retinopathy related to sickle cell disease. Decreased retinal function (via microperimetry testing) has also been detected in association with areas of retinal thinning. Identification of ocular and systemic factors that are associated with sickle cell retinopathy will help identify those patients who most need to be screened for sickle cell retinopathy. The awareness of subclinical disease as well as the identification of systemic factors associated with higher prevalence of sickle cell retinopathy will aid the clinician in identifying those patients who are at higher risk of retinopathy.

Bilateral Simultaneous-Onset Nongranulomatous Acute Anterior Uveitis Clinical Presentation and Etiology

Andrea D. Birnbaum, Yi Jiang, Roshni Vasaiwala, Howard H. Tessler, et.al. Arch Ophthalmol. 2012;130(11):1389-1394.

Chronic bilateral nongranulomatous anterior uveitis is often associated with juvenile idiopathic arthritis (JIA). In this article, the authors describe the etiology and outcome of patients with simultaneous-onset nongranulomatous bilateral acute anterior uveitis. Their cohort of patients comprised 4288 patients with uveitis diagnosed and treated at the uveitis service of a large tertiary referral center over a 20 year period. One percent or 44 patients met the inclusion criterion of acute (less than 3 months in duration) bilateral nongranulomatous anterior uveitis. Pediatric patients comprised 25% (11/44) of their study cohort. Sixty four percent (7/11) were diagnosed with post-infectious or drug-induced uveitis, 18% (2/11) were idiopathic, 9% (1/11) had tubulointerstitial nephritis (TINU) and 9% (1/11) had Kawasaki disease. Drugs implicated in acute uveitis included Bactrim, Levaquin and penicillin.

Bilateral Ophthalmic Artery Occlusions Due to Probable Varicella-Zoster Virus Vasculopathy

Hari Jayaram, Dinu Stanescu-Segal, Graham E. Holder, Elizabeth M. Graham, Arch Ophthalmol.2012;130(11):1492-1494.

Varicella-zoster virus (VZV) vasculopathies account for almost one-third of arterial ischemic strokes in children. Visual complications are rare, with previous reports occurring secondary to unilateral central retinal artery or posterior ciliary artery occlusion. The first case of an immunocompetent child who became blind due to bilateral ophthalmic artery occlusions secondary to probable VZV vasculopathy is described. Vasculitis associated with VZV either primarily or with reactivation can be difficult to determine diagnostically. In this patient, cerebral angiography showed non-perfusion of both ophthalmic arteries, which correlated with a rapid demise in vision of this child who had the chickenpox 8 weeks earlier. He presented with a panuveitis and retinal vasculitis. MRI was normal and vitreous biopsy revelaed no VZV by PCR. Lumbar puncture was positive for VZV IgG. Treatment with Acyclovir may be indicated to prevent systemic involvement but no improvement in vision was noted in their patient.

Novel Etiological Agent: Molecular Evidence for Trematode-Induced Anterior Uveitis in Children

Sivakumar R. Rathinam, Lalan Kumar Arya, Kim R. Usha, Lalitha Prajna, et.al. Arch Ophthalmol. 2012;130(11):1481-1484

Southeast Asian populations are exposed to at least 70 species of foodborne and waterborne trematodes. Histopathological analysis has provided support

that trematode infections can cause a characteristic granulomatous anterior uveitis in children from South India. The authors present a case report of a 9 year old boy who swam in a local village pond in South India who experienced 2 weeks history of eye redness. Polymerase chain reaction testing of an aqueous tap targeted a whitish granuloma filling the inferior part of the anterior chamber and confirmed the trematode, Procerovum varium. No treatment was described. The authors highlighted the usefulness of PCR testing to identify trematodal infection. Serologic testing often is nonspecific with regard to such infections.

Mycoplasma pneumoniae: The Other Masquerader

Enchun M. Liu, Robert H. Janigian Jr, JAMA Ophthalmol. 2013;131(2):251-252.

The authors present 1 case of bilateral optic papillitis and 2 cases of uveitis secondary to Mycoplasma pneumoniae infection. One patient was 14 and the others were in their 20's. Mycoplasma pneumoniae is a bacterium in the class of Mollicutes and is a common cause of atypical pneumonia, particularly in children and young adults. A Mycoplasma infection primarily manifests as a respiratory tract disease, but an extrapulmonary manifestation has occurred. Ocular disease has been reported, with conjunctivitis being the most frequent finding and less common ophthalmologic manifestations include cranial neuropathies, optic papillitis, and anterior uveitis.

Ocular Complications in Children Within 1 Year After Hematopoietic Stem Cell Transplantation

Viera Kalinina Ayuso, Ymkje Hettinga, Patricia van der Does; Jaap J. Boelens, et.al. JAMA Ophthalmol. 2013;131(4):470-475.

The authors stress the importance of understanding the risk of ocular involvement after hematopoietic stem cell transplantation (HSCT) in the pediatric population because young and severely ill children are unaware of their ocular problems. Forty nine consecutive patients underwent ophthalmologic examination before HSCT, during HSCT, and 3, 6, and 12 months after HSCT. Thirteen (27%) developed an ocular complication during HSCT. These complications included dry eye syndrome in 7 (14%), subretinal hemorrhage in 6 (12%), optic disc edema in 3 (6%), chorioretinal lesions in 2 (4%), vitritis in 1 (2%), and increased intraocular pressure in 1 (2%). Median time to the development of dry eye syndrome was 5 months after HSCT, whereas all other ocular complications were detected within the first 3months. Children with malignant disease had a higher risk of the development of ocular complications compared with children with nonmalignant disease.

Interval Spectral-Domain Optical Coherence Tomography

and Electrophysiology Findings in Neonatal Adrenoleukodystrophy

Robert J. Courtney, Mark E. Pennesi, JAMA Ophthalmol. 2013;131(6): 807-810.

The authors present the ocular findings in a 20 month old toddler with neonatal adrenoleukodystrophy(NALD), confirmed by elevated serum very long-chain fatty acid and phytanic acid levels and cultured skin fibroblast analysis. Vision was noted as fix and follow with each eye. Examination revealed an absent foveal light reflex, and there was chorioretinal atrophy with pigmentary changes in a leopard-spot pattern throughout the midperiphery. Spectral domain OCT through the fovea revealed outer retinal atrophy with loss of the external limiting membrane and the inner-segment ellipsoid line; additionally, hyperreflective opacities were suspended in the vitreous. A linear scan through an area of pigmentary change in the midperiphery showed severe atrophy of outer retinal structures and pigment epithelium atrophy with nodules of hyperreflective material on top of the Bruch membrane. The ERG demonstrated severe loss of bothrod- and cone-driven responses.

Distinct Ocular Expression in Infants and Children With Down Syndrome in Cairo, Egypt :Myopia and Heart Disease

Hanan H. Afifi, Amira A. Abdel Azeem, Hala T. El-Bassyouni, Moataz E. Gheith, , et.al. JAMA Ophthalmol. 2013;131(8):1057-1066.

The authors believe there is a distinct Down syndrome phenotype in Cairo, a historically isolated area. The study compares Down syndrome patients in Cairo with Down syndrome patients in other geographic areas. In this population of patients with Down syndrome, 90 infants and children with Down syndrome were examined and followed over a 3 year period. At initial exam 58% had ocular finding with significant refractive error being most common (41%). Nasolacrimal duct obstruction, blepharoconjuctivitis, or conjunctivitis was found in 18 (20%), strabismus in 13 (14%), cataract in 5 (6%), nystagmus in 3 (3%), and optic nerve dysplasia in 2 (2%). Brushfield spots were not found. 40% had cardiac anomalies. An association with myopia was established.

Congenital Cystic Eye In Utero: Novel Prenatal Magnetic Resonance Imaging Findings

James R. Singer, Patrick J. Droste, Adam S. Hassan, JAMA Ophthalmol. 2013;131(8):1092-1095.

Congenital cystic eye is a rare orbital malformation due to failure of optic vesicle invagination during embryogenesis The malformation consists of anophthalmic orbit containing a fluid-filled cyst and, frequently, rudimentary ocular structures. Associated nonocular malformations include intracranial anomalies and systemic malformations. The authors present a case of congenital cystic eye identified prenatally on ultrasound. After birth, MRI of the brain revealed absence

of the corpus callosum and septum pellucidum, in addition to frontal lobe dysplasia, colpocephaly and contralateral dacryocele. Repeated aspiration is often insufficient and their patient ultimately required surgical excision with silicone implant. The authors suggest that congenital cystic eye in association with midline intracranial defects may represent a novel presentation in the septo-optic dysplasia sequence.