What's New and Important in Pediatric Ophthalmology and Strabismus for 2012

Supplement
March 2012-August 2012

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AMBLYOPIA

A Prospective Pilot Study of Treatment Outcomes for Amblyopia Associated With Myopic Anisometropia

Yi Pang, MD, OD, PhD; Christine Allison, OD, et al.. Arch Ophthalmol. 2012;130(5):579-584 (f12)

The purpose of this study was to determine the efficacy of refractive correction alone and patching treatment with near activities on amblyopia associated with myopic anisometropia in children aged 4 to less than 14 years. The associations of visual acuity (VA) improvement with age, degree of anisometropia, patching compliance, presence of strabismus, and presence of eccentric fixation were also investigated. The authors looked at seventeen amblyopic with range of VA in the amblyopic eye 20/80 to 20/400. The primary outcome was VA after 16 weeks of refractive correction alone and final VA after 16 weeks of patching. The mean (SD) baseline VA in the amblyopic eye was 0.96 (0.27) logMAR, which improved to a mean (SD) of 0.84 (0.24) logMAR with refractive correction and to a mean (SD) of 0.71 (0.30) logMAR after the addition of patching ($P < .001$). Comparing the final VA with the baseline VA, we found that VA improvement averaged 2.59 lines. The final VA in the amblyopic eye was associated with the baseline VA in the amblyopic eye ($P < .001$), the magnitude of anisometropia ($P < .001$), and the level of patching compliance ($P = .04$). The improvement in VA with patching was inversely associated with participants' age ($P = .03$) and presence of eccentric fixation ($P = .02$). In summary, both refractive correction and patching significantly improved the VA of the amblyopic eye associated with myopic anisometropia, with 88% of participants' eyes improving 2 lines or more. Further improvement in VA was observed when patching plus near activities was added to refractive correction and patients were followed for 16 more weeks. Consideration should be given to treat myopic anisometropic amblyopia with refractive correction and patching plus near activities

Lessons from the Amblyopia Treatment Studies-Editorial

Michael X Repka, MD, MBA, Jonathan M Holmes, BM, BCh
Ophthalmology April, 2012; 119:657-658 (f12)

The Pediatric Eye Disease Investigator Group (PEDIG) was established in 1997 to provide a structure for conducting clinical trials in pediatric eye disease as part of the clinical practice.

The network has included participants from the United States and Canada as well as funding for the National Eye Institute. A major area of focus has been the management of children and adolescents with amblyopia caused by strabismus, anisometropia or both. The studies were randomized clinical trials as opposed to retrospective and nonrandomized studies, which were prevalent before the year 2000.

The PEDIG began recruitment of patients with amblyopia in 1999. To date, they recruited about 4,000 subjects for 19 different Amblyopia Treatment Studies (ATS). The purpose of the Pediatric Eye Disease Investigator Group (PEDIG) was to describe clinically important, unexpected findings from these published studies and suggest lessons for future investigations to treat amblyopia.
PEDIG clinical trials have shown that initial spectacle correction is the first step of treatment, which, on its own, will effectively treat many cases of amblyopia. Patching, atropine and Bangerter filters are all effective in active treatments even in dosages that once were considered ineffective. Residual amblyopia continues to be a problem for patients. PEDIG is currently exploring the value of levodopa for patients with residual or refractory amblyopia (that has not responded to traditional treatment modalities).
To date, PEDIG results have not addressed the problem of deprivation amblyopia.
However, for anisometropic and strabismic amblyopia treatment can now be more evidence based than in the 1900s, but many questions remain unanswered. PEDIG continues to be resolute and committed to studying the nuances of amblyopia treatment. PEDIG studies are supported in part by the National Institute of Health, Department of Health and Human Services (EY011751 and EY018810 and Research to Prevent Blindness, Inc. New York).

COMMENT: PEDIG is an investigative group composed of optometrists and ophthalmologists throughout the United States and Canada. They meet at least 4 times a year to discuss treatment results and future protocols.

Comfort of Wear and Material Properties of Eye Patches for Amblyopia Treatment and the Influence on Compliance

Anna M.J. Roefs, Angela M. Tjiam, Caspar W.N. Looman, et al
Strabismus March 2012; 20(1), 3-10 (f12)

This is a proactive, non-randomized study designed to evaluate the mechanical properties, particularly the “breathing ability” of the eye patch comparing 4 main brands of patches for 2 consecutive days in randomized fashion.

Twenty-four children participated in this study. For 8 consecutive days, parents used each of 4 main brands of patches for 2 consecutive days in randomized fashion. After 2 days, a 21 item questionnaire was completed to evaluate comfort of wear for each patch. Compliance was measured electronically using the Occlusion Dose Monitor (ODM). In addition, breathing capacity, resistance to water penetration, opacity, and strength of adhesion were measured.

The study concluded that there were large differences in comfort of wear and mechanical properties between the patches. Breathing capability was

felt to be a major requirement affecting tolerance in children. Further study into the property of the patches was recommended.

COMMENT: This study compared the 3M Opticlude, Master-Aid: Orthopad, Lohmann-Rauscher, Pro-ophta, and BSN Medical Coverlet-S patches. This study includes the “Occlusion Patch Comfort Questionnaire” (OCQ), which was used in the study to quantify and qualify patching characteristics. This study was supported and carried out in the Netherlands, Germany, and Dallas, Texas.

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

Toor SS, Horwood AM, Riddell PM. Br Ir Orthopt J 2012; (9): 9-16. (f12)

This literature-based review addresses the factors that are considered in the outcome of therapy for anisometropic amblyopia. The common ones are patient age, degree of anisometropia, depth of amblyopia, type of treatment and compliance. They note that some studies fail to separate patients with strabismus from those with pure anisometropia, or distinguish between patients with myopia versus hypermetropia. Some go further and highlight the importance of further separating anisohypermetropia from anisomyopia as the former are more likely to become myopic. The premise is that in anisomyopia, the more myopic eye can be used for near, whereas the more ametropic eye in hypermetropia remains blurred at all times. The authors discuss other factors such as aniseikonia, astigmatism and how the accommodation response influences anisometropic amblyopia.

Twice weekly atropine as a primary treatment for amblyopia: How does this compare with daily atropine?

Taylor K, Bryant S. Br Ir Orthopt J 2012; (9): 30-35. (f12)

The orthoptists in this clinic learned from their previous trial in 2009 that for moderate degrees of amblyopia, similar results were obtained whether atropine was instilled daily or twice-weekly. They compared the visual acuity outcomes of each treated group. Results showed that 49 percent of patients treated with twice-weekly atropine achieved a cure (defined as 0.250 or better) and a further 30 percent showed significant improvement (0.275-0.500.) These findings were compared with the previous study using daily atropine. No statistically or clinically significant difference was found in visual acuity outcome in the two treated groups.

They looked at the cost of atropine and noted that a 4-month treatment period of daily atropine cost 4 times more than a twice-weekly regimen. Since the cycloplegic effect of atropine lasts 4-14 days, a trial of once-weekly atropine is being considered. [10 refs]

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VISION SCREENING

Validation of printed and computerised crowded Kay picture logMAR tests against gold standard ETDRS acuity test chart measurements in adult and amblyopic paediatric subjects

N Shah, D A H Laidlaw, S Rashid and P Hysi

Eye. April 2012;26:593-600 (f12)

The authors investigated whether visual acuity measured with pictures overestimates visual acuity measured with letters. They compared visual acuity measured with printed and computerized crowded Kay pictures and against gold standard ETDRS letter charts in 30 adult subjects with various ophthalmic diagnoses and in 40 amblyopic children. Each of these tests was repeated once, to determine test retest variability. The six visual acuity tests were performed in a random order.

The authors found that measured visual acuity with Kay pictures was consistently better than that measured by ETDRS letters by approximately one line for the adult subjects, and by two lines better for the pediatric amblyopic subjects. The printed and computerized Kay pictures were similar to one another. Test retest variability was similar for all testing types, and was approximately 1.5 lines.

Importance of vision screening in children regardless of socioeconomic status

S Abdolrahimzadeh

Eye. March 2012;26:478 (f12)

The author reports on vision screening exams on 380 children ages 6 years and 8 years at a private school in Rome. Higher education was recorded for both parents in 334 families (87.9%) and for one parent in 30 families (7.9%). Refractive errors were considered significant with hyperopia greater than 3 diopters or myopia more than 0.5 diopters. 14.5% of children had significant refractive errors but only 6.3% had already
received a spectacle correction, notwithstanding the high socioeconomic status of the families. The refractive errors deemed significant in this study do not match those considered potentially amblyogenic by AAPOS/AAO. The author comments that even children of higher socioeconomic status require vision screening examinations.

The prevalence of ocular structural disorders and nystagmus among preschool-aged children. Repka MX, Friedman DS, Katz J, et al. J AAPOS 2012;16:182-184. (f12)
The Baltimore Pediatric Eye Disease Survey (BPEDS) was conducted to determine the prevalence of decreased visual acuity, strabismus, amblyopia, refractive error and other ocular disorders. This study describes the prevalence of the 'other ocular disorders'. Of 4132 eligible children, 3990 were enrolled and 2546 were examined. Of these, 159 children (6.2%) had abnormalities of alignment, vision, nystagmus, or structural abnormalities. Structural ocular disorders were found in 41 children. Posterior disorders were more likely to be associated with vision loss. Abnormalities significant enough to be associated with vision loss were very uncommon. The authors opined that development of screening methods targeting these problems is not warranted as the majority would be detected by current screening strategies and treatment options for the detected conditions are largely not available.

Testability of refraction, stereopsis, and other ocular measures in preschool children: The Sydney Paediatric Eye Disease Study. Shih-I Pai A, Rose KA, Samarawickrama C, et al. J AAPOS 2012;16:185-192. (f12)
The authors attempted to document the testability for refraction, ocular biometry, stereoacuity, and retinal imaging in a preschool population-based study of children from various ethnic backgrounds and to determine the lower age limits for applying these tests. The Sydney Paediatric Eye Disease Study identified 3,333 eligible children from various socioeconomic strata. 2,461 children were examined and 2,189 children were included in this report (exclusions were for missing data and lack of cycloplegic refraction). The mean age of included children was 36.9 months. Caucasian, East Asian, and South Asian were the dominant ethnic groups in that order. Retinomax autorefractors yielded better testability than Canon for all age groups. Canon autorefractors require a longer sustained attention span to complete testing. However, Retinomax has been reported to obtain results less consistent with streak retinoscopy. The testability of both autorefractors increased sharply with patient age. Ethnicity was not a factor and testability was greater in boys than girls with the Retinomax autorefractor. Axial length was the most testable of ocular biometry measurements, followed by corneal curvature and anterior chamber depth. Testability of all 3 measurements increased as a function of age. IOLmaster testing feasibility improved substantially after 48 months of age. Lang II was the most testable of the stereoacuity tests, followed by Randot and stereosmile. Lang testing does not require special glasses. Testability also rose in relation to patient age. Stereotest results were difficult to obtain in children younger than 3 years of age. Retinal photographs were difficult to obtain in children younger than 42 months and age was a significant factor for testability.

Visual acuity testability and comparability in Australian preschool children: The Sydney Paediatric Eye Disease Study

Leone JF, Gole GA, Mitchell P, Kifley A, Pai AS, Rose KA.

Eye. Jul 2012;26:925-32. (f12)

Purpose: To establish standardised protocols for vision screening, testability and comparability of three different vision tests were examined in a population-based, cross-sectional sample of Australian preschool children.
1774 children age 2 years and older attempted the Amblyopia Treatment Study HOTV protocol, in which a single HOTV letter with crowding bars is presented to the patient for matching. In addition, in 576 children aged 5 years and older, visual acuity was also tested using the logMAR retro-illuminated HOTV or Early Treatment Diabetic Retinopathy Study (ETDRS) linear charts.
The authors found that testability significantly increased with age for all VA tests. The ATS HOTV with an overall testability of 80% was the most testable of the VA tests ($P<0.0001$). In children aged <3 years testability was low (≤47%). In children aged 5 years and older, testability was higher for the HOTV (94%) than the ETDRS (59%) chart. In those that could complete both types of VA tests, the ATS HOTV visual acuity was 1 line higher than the HOTV linear acuity, and 1.2 lines better than the ETDRS linear acuity ($P<0.0001$).
The implication of this study is that most the ATS HOTV test is suitable for most children ages 3 to 5, and more children ages 5 years and older are testable with the linear HOTV test than the linear ETDRS test. children aged <3 years have poor VA testability. When comparing VA measures using these tests, the higher VA attained using the ATS HOTV needs to be taken into account.

Crowding Ratio in Young Normally Sighted Children

Marjoke J. Dekker, Florine Pilon, Mieke M. C. Bijveld, Gerard C. de Wit, Maria M. van Genderen Strabismus June 2012, Vol 20, No 2: 49-54 (f12)

The purpose of this study is to determine the crowding ration (CR) of children with normal vision. Sixty-two normally sighted children were evaluated and their CR was determined. For children older than 6 years of age, a CR of greater than 2.0 is suspicious and warrants further investigation

Dynamic distance direct ophthalmoscopy, a novel technique to assess accommodation in children

Kothari M, Balankhe S, Paralkar S, Nar D, et al.. (f12)

Indian J Ophthalmol 2012;60:109-14 (Mar-Apr)
This study aims to compare dynamic distance direct ophthalmoscopy (DDDO) with dynamic retinoscopy (DR) in assessment of accommodation in children. There were four components: Component 1: to understand the characteristic digital images of DDDO. Component 2: to compare DDDO with DR for detection of accommodative defects in children (1-16 years). Component 3: to compare DDDO with DR for the detection of completeness of pharmacologically induced cycloplegia in children (5-16 years) and Component 4: to assess which one of the two techniques was more sensitive to detect onset of cycloplegia after instillation of 1% cyclopentolate eye drops. The authors conclude that DDDO is a reliable method to assess accommodation in young children and neurologically impaired patients. This test can assess the accommodative response of both eyes simultaneously and with photography it can be more objective than dynamic retinoscopy.

Screening for visual impairment: Outcome among schoolchildren in a rural area of Delhi

Rustagi N, Uppal Y, Taneja DK. Indian J Ophthalmol 2012;60:203-6 (May-Jun) (f12)
The study aims to assess the magnitude of visual impairment among children and the compliance of students with refraction testing, procurement and use of spectacles in schools of the north- west district of Delhi. Low vision (visual acuity < 20/60) in the better eye was observed in 31 (2.9%) children and blindness (visual acuity <20/200) in 10 (0.9%) children. Compliance with referral for refraction was very low as only 51 (41.5%) out of 123 students could be tested for refraction. Out of 48 students, 34 (70.8%) procured spectacles from family resources but its regular use was found among only 10 (29.4%) students. The poor compliance among students stems out of various myths and perceptions regarding use of spectacles prevalent in the community. The authors conclude that behavior change communication among rural masses about eye health and involving parent-teacher associations and senior students to motivate students for use of spectacles may improve utilization of existing eye health services in rural areas.

Crowding Ratio in Young Normally Sighted Children

Marjoke J. Dekker, BH, Florine Pilon, BH, Mieke M.C. Bijveld, MSc, et al
Strabismus June 2012; 20(2): 49-54 (f12)

This study comes from the Bartimeus Institute for the Visually Impaired, Ophthalmology, Zeist, Netherlands.
The purpose of this study is to determine normal values of the crowding ratio (CR) in children in normally sighted children.

Sixty-two normally sighted children between 4 and 12 years of age were evaluated. The crowding ratio was determined both for distance and near vision. The examinations were performed using commonly available test charts based on the LEA symbols.

At near, the crowding ratio was significantly better than at distance for all ages (crowding ratio less than 2.0). The upper limit of the crowding ratio at distance was less than 2.0 for ages 6 and beyond. The authors concluded that with commonly available tests the crowding ratio could easily be determined in school-aged children. The results show that the crowding ratio greater than 2 is likely to be abnormal at any age and at distance a crowding ratio greater than 2 is likely to be abnormal from ages 6 and beyond. A disparity of more than 2 lines between linear acuity and single optotype acuity should be reason for further investigation. The authors feel that a high crowding ratio may be an indication of cerebral visual impairment.

COMMENT: According to the authors, crowding refers to “impaired target recognition caused by surrounding contours”. Crowding can be expressed by the crowding ratio, which is defined as single optotype acuity divided by linear acuity. According to the authors, crowding is a characteristic feature of strabismic amblyopia but a certain amount of crowding is also present in normal eyes. Young preschool children may also exhibit crowding but it gradually decreases as perceptual maturation proceeds.

Crowding in Central Vision in Normally Sighted and Visually Impaired Children Aged 4 to 8 Years: The Influence of Age and Test Design

Bianca Huurneman, MSc, F. Nienke Boonstra, MD, PhD, Antonius H.N. Cillesse, PhD, et al
Strabismus June 2012; 20(2): 55-62 (f12)

This study comes from various universities in The Netherlands including the Bartimeus Institute for the Visually Impaired, Zeist, The Netherlands.
This article supplements the article “Crowding Ratio in Young Normally Sighted Children”. The purpose of the study is to investigate crowding ratios in children with a visual impairment due to ocular disease in normally sighted children.

All children studied were 4 to 8 years of age. Fifty-eight visually impaired children and 75 normally sighted children were studied. The crowding ratio is defined as single optotype acuity divided by linear acuity (Atkinson, et al, in 1987). “Crowding is generally defined as the deleterious influence of nearby contours on object recognition, a bottleneck in perception or separation difficulty.” (Abraham SB, 1931, and Amos JF, 1987). Crowding can be seen as a development phenomenon, as crowding effects are larger in children than in adults. Crowding also occurs in normally sighted young children, children and adults with

strabismus, adults with congenital nystagmus, and visually impaired adults with ocular disease. Contrast, fixation stability, nystagmus, central scotomas, configuration of stimuli, amblyopia, maturation of retinal receptors, and maturation of selective attention mechanisms have all been implicated in an attempt to explain this phenomenon.

In this study crowding ratios are calculated by dividing single acuity by the linear acuity that was measured binocularly with the Landolt C-test and the LEA figures (LH figures). This study concluded that visually impaired children showed higher crowding ratios than normally sighted children when measured with the standard eye charts. This study illustrates that test design is very important to keep in mind when assessing crowding ratios in visually impaired and normally sighted children.

This study confirmed that there would higher crowding ratios for visually impaired children than normally sighted children. This hypothesis was confirmed when using charts with a fixed inner symbol spacing (ISS). Visually impaired children with nystagmus showed higher crowding ratios in visually impaired children without nystagmus. The chart with proportional inner symbol spacing (ISS) did not reveal differences between normally sighted and visually impaired children nor did it show group, age or nystagmus effects.

COMMENT: This study is the first to compare crowding ratios calculated from different charts (Landolt C and LH charts) in normally sighted children and visually impaired children aged between 4 and 8 years. The results show that inter symbol spacing and fixed or proportional test design have a substantial influence on crowding ratios. THE CHARTS WITH FIXED INNER SYMBOL SPACING GENERALLY MEASURE HIGHER CROWDING RATIOS AND SEEM TO BE MORE SENSITIVE IN MEASURING AGE-RELATED CHANGES IN THE SIZE OF THE CROWDING RATIO. Because of the different outcomes measured with different charts, careful documentation and reporting of the testing conditions including the inner symbol spacing distance was recommended to improve interpretation.

This study shows that when measured with a chart with fixed inner symbol spacing, visually impaired children show higher crowding ratios than normally sighted children. Normally sighted children demonstrate a reduction in the crowding ratio with increasing age. This reduction in crowding ratio is not seen in visually impaired children when they were tested with charts with fixed spacing.

Children with nystagmus showed higher crowding ratios than children without nystagmus. This outcome is considered of great importance because the higher crowding ratios may affect daily activities such as reading. Further investigation is recommended.

Normative data for the crowded logMAR Kay's pictures vision test in children. Saul T, Taylor K. BR IR Orthoptic J 2012; (9): 36-43. (f12)

LogMAR vision tests for children can be divided into two categories: crowded or uncrowded. Most uncrowded tests can overestimate visual acuity. For this reason, crowded logMAR vision tests are thought to be a more precise method of quantifying vision and are recommended in the measurement of visual acuity in children.
In this prospective study, 100 visually-normal children between 3-4 years of age were tested. Normative values were collected using the crowded logMAR Kay picture test. This test, using pictures instead of letter optotypes, has been shown to be comparable to crowded logMAR letter testing. It is important for clinicians to know the age-appropriate normative visual acuity data in children when assessing clinically significant visual acuity abnormalities. [15 refs.]

[Home]

REFRACTIVE ERROR

Accommodative lag and refractive error in infants and toddlers.
Tarczy-Hornoch K. J AAPOS 2012;16:112-117. (f12)
This paper characterized accommodative performance in infants and toddlers as a function of spherical equivalent (SE) and astigmatism. Enrolled children were 5 to 24 months of age. Accommodative lag was measured by a dynamic retinoscopy technique known as modified bell retinoscopy. Refraction was assessed by cycloplegic retinoscopy. Data from 189 participants was analyzed. Of note, 60% were Hispanic. 161/189 had with the rule (WTR) astigmatism and 12/189 displayed against-the-rule astigmatism (ATR). The mean accommodative lag was 0.37 diopters (D). Younger children had greater lags. The younger children also showed lower accommodative demand –response function. Larger degrees of hyperopia, and/or astigmatism were related to increased accommodative lag. Myopes and those with mixed astigmatism showed no significant correlation between lag and astigmatism.
This paper suggests maturation of the accommodation process over the age range studied. Children with larger degrees of hyperopia or astigmatism showed greater accommodative lag. Refractive error in this study was not assessed until after measurements of the accommodative response to avoid a potential bias. Limitations of the study include difficulty in quantifying leads or very large lags of accommodation. There were few enrolled children with large refractive errors, limiting our ability to draw conclusions from this group of patients. Most of the patients were Hispanic so results may not be generalized to other populations.

Finally, this study does not give any information on how this information might change management of various refractive errors.

Annual Changes in Refractive Errors and Ocular Components before and after the Onset of Myopia in Chinese Children

Fan Xiang, MD, Mingguang He, MD, PhD, Ian G Morgan, PhD
Ophthalmology July 2012; 119:1478-1484 (f12)

This is a longitudinal study designed to examine annual changes in refraction and ocular components around the onset of myopia in Chinese children. The participants were twins age 7 to 15 years from the Guangzhou Twin Registry. Participants underwent eye examinations annually from 2006 to 2010. Before the onset of myopia, axial elongation and progression accelerate. After myopic refraction is established, axial elongation and progression decrease.
The authors suggest that the increase before myopia may be due to increased intensity of the study and decreased time outdoors. In contrast, the rapid slowing after the onset of myopia may represent an inhibitory effect of myopic defocus on eye growth.
This study emphasizes that children who are not myopic at the first examination and develop myopia on at least 1 subsequent examination from 2006 to 2010 were included for analysis. Annual change in axial length showed a similar but inverse shape to the spherical equivalent refraction (SER). Annual change in lens power did not change significantly around the onset of myopia.

This study was performed at the State Key Laborator of Ophthalmology, Zhongshan Ophthalmic Center, Sun
Yat-sen University, Guangzhou, China, and the Austrailian National University, Canberra, Australia.

Atropine Dose to Treat Myopia

Virgilio Galvis, MD, Alejandro Tello, MD, Carlos Rodriguez, OD, et al, from Bucaramanga, Colombia.
Ophthalmology August 2012; 119: 1718-1719 (f12)

The Letter to the Editor was in response to an article by Chia A and Chua WH, et al from *Ophthalmology* 2012; 119: 347-354, describing Atropine for the treatment of childhood myopia: safety and efficacy of 0.5%, 0.1% and 0.01% doses (Atropine for the treatment of myopia). The authors of the original article concluded that using Atropine 0.01% had comparable efficacy compared to higher doses of Atropine with fewer side effects for the purpose of controlling myopic progression.

The authors of the Letter felt that with these preliminary results we think that reducing the frequency of administration of 1% Atropine could be an alternative to reducing the concentration of Atropine in control of myopic progression.

The authors of the original article (Chia, Chua, Cheung, et al) responded to the letter and recommended the use of randomized trial to compare reduced dosage Atropine with frequency of administration of 1% Atropine in reducing the progression of myopia.

Comparison of measurements of time outdoors and light levels as risk factors for myopia in young Singapore children

Dharani R, Lee CF, Theng ZX, Drury VB, Ngo C, Sandar M, Wong TY, Finkelstein EA, Saw SM.

Eye. Jul 2012;26:911-8. (f12)

Exposure to outdoor light may be protective against the development of myopia. The authors compared two methods of measuring time outdoor: a parental diary and a light meter worn attached to clothing. In addition, the light meter measured the exposure to light in lux in 5 minute increments throughout the day. These methods were applied to 117 Singapore children aged 6–12 years with and without myopia for one week.
Mean outdoor time according to the diary entries was and time with light levels was 5.44 hours per week during the school week and 7.91 hours per week during school holidays. Time spent with light levels of >1000 Lux according to the light meter was 7.08 h per week during the school week and 9.81 h per week during school holidays. The agreement between the diary and light meter was poor to fair. These methods measure different parameters, time spent outdoors and light intensity, and could therefore capture different aspects of risk in future myopia studies.

Myopic Optic Disc Changes in Adolescents

Young Hoon Hwang, MD, Yong Yeon Kim, MD
Ophthalmology April, 2012: 119:885-886 (f12)

This is a letter to the editor. The authors discuss a population-based cross-sectional study of the adolescent children from Singapore Cohort Study of Risk Factors for Myopia (SCORM) reported that the prevalence of tilted optic disc was 37% and eyes with tilted optic discs were more nearsighted/myopic, astigmatic and longer axial lengths than eyes without tilted discs.

The authors suggest a hypothesis for the optic tilt, namely, a congenital anomaly associated with malclosure of the embryonic optic fissure/fetal cleft and acquired progressive changes in the optic disc according to myopic elongation of the eyeball. They suggest that an investigation with association between longitudinal

changes and the optic disc tilt and other factors may provide important clues to the pathogenesis of myopic optic disc tilt.

Refractive profile in oculocutaneous albinism and its correlation with final visual outcome Claudia Yahalom, Veronica Tzur, Anat Blumenfeld, Gabriel Greifner, Dalia Eli. Ada Rosenmann, Sherry Glanzer, Irene Anteby Br J Ophthalmol 2012;96:537-539 (f12)

Purpose: To evaluate the prevalence of refractive errors in different subtypes of oculocutaneous albinism, and to see if there is any correlation between refractive errors
and final visual outcome in this population.

Results: Refractive errors were mainly astigmatism and hypermetropia. The OCA1A group showed high hypermetropia ($5 dioptres) in 43.4% of patients,reaching significantly higher levels than in other subgroups (p.0.007). Mean visual acuity in logMAR was: OCA1A.0.81, OCA1B.0.64, OCA1C.0.61 and OCA2.0.48. Astigmatism averaged 2.1 dioptres (consistently with-the-rule), and it was homogeneously distributed between all subgroups (53%).
Conclusions: The poorest visual acuity was found in those with OCA1A, which was associated with the highest rate of high hypermetropia (statistically significant different from other subgroups). Astigmatism was the most common visually significant refractive error across all subtypes of albinism.

Comments: These results may help to clarify the prevalence of refractive errors in albino patients and aid the prediction of visual outcome in this heterogeneous population.

Myopia Progression in Children Is Linked with Reduced Foveal mfERG Response. Wing-cheung Ho, Chea-su Kee and Henry Ho-lung Chan IOVS August 2012 53:5320-5325; published ahead of print July 3, 2012, doi: 10.1167/iovs.12-10185. (f12)

The purpose of this study was to assess changes in retinal electrophysiology in children during myopia progression during a 1-year period. Twenty-six children aged from 9 to 13 years were recruited for the global flash multifocal electroretinogram measured at 49% and 96% contrast, in two visits 1 year apart. The amplitudes and implicit times of both direct component (DC) and induced component (IC) measured at these two visits were analyzed and compared. Myopia increased by −0.48 ± 0.32 diopter (D) ($P < 0.001$) during the year, with 24 of 26 children becoming more myopic (range = 0.00 to ∼ −1.38 D); axial length increased by 0.25 ± 0.11 mm ($P < 0.001$) during the year. The increased myopia

was highly correlated with increase in axial length ($r = -0.70$; $P < 0.001$). The central DC and IC amplitudes at 49% contrast reduced significantly as myopia progressed and the paracentral implicit times of these two components were reduced considerably. However, the high-contrast responses were virtually unaffected.

The author's findings suggested that the inner retinal functions in the central retina, with some involvement of the paracentral region, were decreased as myopia progressed in children.

Changes in Lens Power in Singapore Chinese Children during Refractive Development Rafael Iribarren, Ian G. Morgan, Yiong Huak Chan, Xiaoyu Lin, and Seang-Mei Saw. IOVS August 2012 53:5124-5130; published ahead of print July 10, 2012, doi:10.1167/iovs.12-9637. (f12)

The purpose of this study was to examine changes in lens power during refractive development in Singapore Chinese children. Children aged six to nine years from three Singapore schools were invited to participate in the Singapore Cohort study Of the Risk factors for Myopia (SCORM) study. Cycloplegic refractions and biometry were measured annually in the schools over a five year period from 1999. Children were classified into five refractive error groups: persistent hyperopia, emmetropizing hyperopia, persistent emmetropia, newly developed myopia, or persistent myopia. Crystalline lens power was calculated using Bennett's formula. The rate of change per year across the refractive groups was adjusted for age and sex using General Linear Models. There were 1747 children with at least three sets of measurements for lens power calculations. The mean age at baseline was 7.94 ± 0.84 years and the mean spherical equivalent refraction was −0.41 ± 1.71 diopters (D). Lower lens power and lower lens thickness were associated with persistent myopia. As expected, the newly developed myopes and the persistent myopes showed the largest changes in axial length (AL). Changes in lens power and thickness at follow-up were similar in all refractive groups, except for the newly developed myopes, who showed significantly greater decreases in lens power (0.36 vs. 0.29 D/year; $P < 0.001$) and lens thickness (0.015 vs. 0.0003 mm/year; $P < 0.001$) than the persistently emmetropic group. Newly developed myopes showed a significantly greater decrease in lens power than other refractive groups, which may be linked to rapid changes in AL and refraction that occur around the onset of myopia.

Right and Left Correlation of Retinal Vessel Caliber Measurements in Anisometropic Children: Effect of Refractive Error Nichole Joachim, Elena Rochtchina, Ava Grace Tan, Thomas Hong[1], Paul Mitchell, and Jie Jin Wang. Invest Ophthalmol Vis Sci. 2012 Aug 7;53(9): 5227-30. (f12)

Previous studies have reported high right–left eye correlation in retinal vessel caliber. The authors tested the hypothesis that right–left correlation in retinal vessel caliber would be reduced in anisometropic compared with emmetropic children. Retinal arteriolar and venular calibers were measured in 12-year-old children. Three groups were selected: group 1, both eyes emmetropic (*n* = 214); group 2, right–left spherical equivalent refraction (SER) difference ≥1.00 but <2.00 diopter (D) (*n* = 35); and group 3, right–left SER difference ≥2.00 D (*n* = 32). Pearson's correlations between the two eyes were compared between group 1 and group 2 or 3. Associations between right–left difference in refractive error and right–left difference in caliber measurements were assessed using linear regression models. Refractive error ≥2.00 D may contribute to variation in measurements of retinal vessel caliber. In summary, right and left eye correlation of retinal arteriolar and venular caliber estimates tended to be lower in anisometropic compared with emmetropic children, and increasing right–left differences in refractive error or in axial length were weakly associated with increasing right–left differences in retinal vessel caliber estimates. Correction for refractive error is needed in eyes with SER differences ≥2.00 D when examining associations with retinal vessel caliber in study samples with a wide range of refractive powers.

The Association between Time Spent Outdoors and Myopia Using a Novel Biomarker of Outdoor Light Exposure Justin C. Sherwin, Alex W. Hewitt, Minas T. Coroneo, Lisa S. Kearns, Lyn R. Griffiths and David A. Mackey. Invest Ophthalmol Vis Sci. 2012 Jul 1;53(8):4363-70. (f12)

The authors sought to determine whether conjunctival ultraviolet autofluorescence (UVAF), a biomarker of outdoor light exposure, is associated with myopia. They performed a cross-sectional study on Norfolk Island and recruited individuals aged ≥15 years. Participants completed a sun-exposure questionnaire and underwent noncycloplegic autorefraction. Conjunctival UVAF used a specially adapted electronic flash system fitted with UV-transmission filters (transmittance range 300–400 nm, peak 365 nm) as the excitation source. Temporal and nasal conjunctival UVAF was measured in both eyes using computerized photographic analysis with the sum referred to as “total UVAF.” In 636 participants, prevalence of myopia decreased with an increasing quartile of total UVAF (P_{trend} = 0.002). Median total UVAF was lower in subjects with myopia (spherical equivalent [SE] ≤ −1.0 diopter [D]) than participants without myopia: 16.6 mm^2 versus 28.6 mm^2, P = 0.001. In the multivariable model that adjusted for age, sex, smoking, cataract, height and weight, UVAF was independently associated with myopia (SE ≤ −1.0 D): odds ratio (OR) for total UVAF (per 10 mm^2) was 0.81, 95% confidence interval (CI) 0.69 to 0.94, P = 0.007. UVAF was also significantly associated with myopia when analysis was restricted to subjects <50 years, and in moderate-severe myopia (SE ≤ −3.0 D). Prevalence of myopia

decreased with increasing time outdoors (P_{trend} = 0.03), but time outdoors was not associated with myopia on multivariable analysis.

The study authors identified a protective association between increasing UVAF and myopia. The protective association of higher UVAF against myopia was stronger than that of increased levels of time spent outdoors as measured by the study's questionnaire. Further evidence is required from prospective studies to further characterize this relationship between UVAF and incident myopia, and to assess the role of UVAF in myopic progression.

Time Outdoors and Physical Activity as Predictors of Incident Myopia in Childhood: A Prospective Cohort Study Jeremy A. Guggenheim, Kate Northstone, George McMahon, Andy R. Ness, Kevin Deere, Calum Mattocks, Beate St Pourcain and Cathy Williams. Invest Ophthalmol Vis Sci. 2012 May 14;53(6):2856-65. (f12)

Time spent in “sports/outdoor activity” has shown a negative association with incident myopia during childhood. The authors investigated the association of incident myopia with time spent outdoors and physical activity separately. Participants in the Avon Longitudinal Study of Parents and Children (ALSPAC) were assessed by non-cycloplegic autorefraction at ages 7, 10, 11, 12, and 15 years, and classified as myopic (≤−1 diopters) or as emmetropic/hyperopic (≥ −0.25 diopters) at each visit (*N* = 4,837–7,747). Physical activity at age 11 years was measured objectively using an accelerometer, worn for 1 week. Time spent outdoors was assessed via a parental questionnaire administered when children were aged 8–9 years. Variables associated with incident myopia were examined using Cox regression. In analyses using all available data, both time spent outdoors and physical activity were associated with incident myopia, with time outdoors having the larger effect. The results were similar for analyses restricted to children classified as either non-myopic or emmetropic/hyperopic at age 11 years. Thus, for children non-myopic at age 11, the hazard ratio (95% confidence interval, CI) for incident myopia was 0.66 (0.47–0.93) for a high versus low amount of time spent outdoors, and 0.87 (0.76–0.99) per unit standard deviation above average increase in moderate/vigorous physical activity.

Time spent outdoors was predictive of incident myopia independent of physical activity level. The greater association observed for time outdoors suggests that the previously reported link between “sports/outdoor activity” and incident myopia is due mainly to its capture of information relating to time outdoors rather than physical activity.

Myopia Progression in Children Is Linked with Reduced Foveal mfERG Response. Wing-cheung Ho, Chea-su Kee and Henry Ho-lung

Chan. IOVS August 2012 53:5320-5325; published ahead of print July 3, 2012, doi:10.1167/iovs.12-10185. (f12)

The purpose of this study was to assess changes in retinal electrophysiology in children during myopia progression during a 1-year period. Twenty-six children aged from 9 to 13 years were recruited for the global flash multifocal electroretinogram measured at 49% and 96% contrast, in two visits 1 year apart. The amplitudes and implicit times of both direct component (DC) and induced component (IC) measured at these two visits were analyzed and compared. Myopia increased by −0.48 ± 0.32 diopter (D) ($P < 0.001$) during the year, with 24 of 26 children becoming more myopic (range = 0.00 to ∼ −1.38 D); axial length increased by 0.25 ± 0.11 mm ($P < 0.001$) during the year. The increased myopia was highly correlated with increase in axial length ($r = −0.70$; $P < 0.001$). The central DC and IC amplitudes at 49% contrast reduced significantly as myopia progressed and the paracentral implicit times of these two components were reduced considerably. However, the high-contrast responses were virtually unaffected.

The author's findings suggested that the inner retinal functions in the central retina, with some involvement of the paracentral region, were decreased as myopia progressed in children.

Anisometropia in Children from Infancy to 15 Years. Li Deng and Jane E. Gwiazda. Invest Ophthalmol Vis Sci. 2012 Jun 20;53(7):3782-7. (f12)

The purpose of the study was to investigate anisometropia in children from age 6 months to 15 years. Children with refractions at 6 months (n = 1120), 5 years (n = 395), and 12 to 15 years (n = 312) were included in this study. All children were refracted in the laboratory by noncycloplegic retinoscopy. Myopes had spherical equivalent refraction (SER) of the less ametropic eye of less than −0.50 D, hyperopes had SER of the less ametropic eye greater than or equal to 1.00 D, and emmetropes had SER of the less ametropic eye from −0.50 to +1.00 D. The mean difference in refraction between the two eyes was similar at 6 months (0.11 D) and 5 years (0.15 D), increasing to 0.28 D at 12 to 15 years. Using a cutoff of 1.00 D SER for anisometropia, the prevalence was 1.96%, 1.27%, and 5.77% at 6 months, 5 years, and 12 to 15 years, respectively. At 12 to 15 years, the prevalence of anisometropia in the myopes was 9.64% and in the hyperopes was 13.64%, both significantly higher than that in the emmetropes (3.38%, $P < 0.05$). The degree of anisometropia at 12 to 15 years was significantly associated with the refractive error of the less ametropic eye at 12 to 15 years, with and without adjustment for relevant covariates ($P < 0.05$). Infants with significant astigmatism (cylinder power ≥ 1.00 D in one or both eyes) have an increased risk of anisometropia ($P < 0.05$).

The prevalence of anisometropia increases between 5 and 15 years, when some children's eyes grow longer and become myopic. However, anisometropia was found to accompany both myopia and hyperopia, suggesting that other mechanisms in addition to excessive eye growth may exist for anisometropia development, especially in hyperopia.

Association between Ocular Dominance and Spherical/ Astigmatic Anisometropia, Age, and Sex: Analysis of 1274 Hyperopic Individuals Stephan J. Linke, Julio Baviera, Gisbert Richard, and Toam Katz.. Invest Ophthalmol Vis Sci. 2012 Aug 9;53 (9):5362-9. (f12)

The purpose of this study was to determine the association between ocular dominance and spherical/astigmatic anisometropia, age, and sex in hyperopic subjects. The medical records of 1274 hyperopic refractive surgery candidates were filtered. Ocular dominance was assessed with the hole-in-the-card test. Refractive error (manifest and cycloplegic) was measured in each subject and correlated to ocular dominance. Only subjects with corrected distance visual acuity of >20/22 in each eye were enrolled, to exclude amblyopia. Associations between ocular dominance and refractive state were analyzed by means of *t*-test, χ^2 test, Spearman correlation, and multivariate logistic regression analysis. Right and left eye ocular dominance was noted in 57.4 and 40.5% of the individuals. Non-dominant eyes were more hyperopic (2.6 ± 1.27 diopters [D] vs. 2.35 ± 1.16 D; $P < 0.001$) and more astigmatic (−1.3 ± 1.3 D vs. −1.2 ± 1.2 D; $P = 0.003$) compared to dominant eyes. For spherical equivalent (SE) anisometropia of >2.5 D ($n = 21$), the non-dominant eye was more hyperopic in 95.2% (SE 4.7 ± 1.4 D) compared to 4.8% (1.8 ± 0.94 D; $P < 0.001$) for the dominant eye being more hyperopic. For astigmatic anisometropia of >2.5 D ($n = 27$), the non-dominant eye was more astigmatic in 89% (mean astigmatism −3.8 ± 1.1 D) compared to 11.1% (−1.4 ± 1.4 D; $P < 0.001$) for the dominant eye being more astigmatic. The present study is the first to show that the non-dominant eye has a greater degree of hyperopia and astigmatism than the dominant eye in hyperopic subjects. The prevalence of the non-dominant eye being more hyperopic and more astigmatic increased with increasing anisometropia. The authors demonstrated that non-dominant eyes exhibit a greater hyperopic SE power and a higher amount of astigmatism in subjects with anisometropia of >0.5 D. Supplementary information about the complexity and the influence of determinative factors on ocular dominance and the mutual impact of sensory and motor ocular dominance might be gained by implementing functional imaging techniques in characterizing ocular dominance.

[Home]

VISUAL IMPAIRMENT

A survey of severe visual impairment in children attending schools for the blind in a coastal district of Andhra Pradesh in South India.

Krishnaiah S, Subba Rao B, Lakshmi Narasamma K, Amit G.

Eye. Aug 2012;26:1065-70. (f12)

The authors examined 113 children attending Schools for the Blind in Andhra Pradesh in South India. They classified the causes of blindness according to World Health Organization Criteria. The major causes of blindness were congenital eye anomalies, such as anophthalmos and microphthalmos, in 46 children (41.4; 95 confidence interval (CI): 32.3–50.6), followed by retinal disorders (primarily retinal dystrophies) in 21 children (18.9; 95 CI: 11.6–26.2), cataract in 9 children (9.7; 95 CI: 2.9–12.9), and corneal conditions (scar and staphyloma) in 8 children (7.1; 95 CI: 2.4–11.8). More than half the children (56.6) were blind due to conditions that could have been treated or prevented. Preventable causes were ophthalmia neonatorum and trauma, whereas treatable causes were primarily cataract and glaucoma. Approximately half the children were the products of consanguineous marriages. The authors discuss why data from school for the blind is subject to bias. Their data differs from reports from other parts of India in that there was no blindness due to retinopathy of prematurity, and a low incidence of corneal blindness due to vitamin A deficiency, and little rubella. The authors believe that education about the risks of consanguineous marriage may help decrease blindness due to heritable conditions, such as microphthalmos/anophthalmos and retinal dystrophies.

Reading Skills in Children and Adults With Albinsim: The Role of Visual Impairment

John T. MacDonald, MD; Beth R. Kutzbach, MD; Ann M. Holleschau, BA; Suzanne Wyckoff, MD; C. Gail Summers, MD J Pediatr Opthalmol Strabismus 2012; 49:184-188 May/June (f12)

Albinism is a condition closely associated with poor visual acuity; however, are reading skills necessarily poor because of the poor visual acuity? This article found that adults and children with albinism have normal reading skills on standardized tests, however, they have decreased fluency which may require extra rime during tasks that require reading skills in the school system or in the workplace.

Clinical Research on the Ophthalmic Factors Affecting 3D Asthenopia

Seung-Hyun Kim, MD, PhD; Young-Woo Suh, MD, PhD; Jong-Suk Song, MD, PhD; Ji-Hye Park, MD; Yong Yeon Kim, MD, PhD; Kuhl Huh, MD, PhD; Jaebum

Son, PhD; Keetaek Kham, PhD; Taeuk Jeong, PhD; Kyung Soo Pyo, PhD J Pediatr Opthalmol Strabismus 2012; 49:248-253 July/August (f12)

Watching 3D images can sometimes cause 3D asthenopia which refers to eye discomfort, fatigue, headache, and nausea. A survey of 115 volunteers was performed to determine which factors make a person more likely to have 3D asthenopia. Ophthalmic factors such as vision, refractive errors, interpupillary distance, intraocular pressure, tear break-up time, near point of accommodation, strabismus, and retinal abnormalities were evaluated. The only risk factor identified based on the survey to increase likelihood of 3D asthenopia was the presence of exophoria.

Diagnosing Cerebral Visual Impairment in Children with Good Visual Acuity

Maria van Genderen, Marjoke Dekker, Florine Pilon, Irmgard Bals Strabismus Jun 2012, Vol. 20, No. 2: 78–83 (f12)

This purpose of this study was to evaluate various elements that can be used to facilitate diagnosis of cerebral visual impairment (CVI) in children who have good visual acuity and identify the individuals that necessitate further multidisciplinary evaluation. Medical history, MRI findings, visual acuity, crowding ratio, visual field assessment, and the results of ophthalmologic and orthoptic examination were evaluated. A short CVI questionnaire was also given. The most important risk for CVI in children is an abnormal pre- or perinatal medical history. Additional symptoms of cerebral damage include cerebral palsy, visual field defects, partial optic atrophy and a crowding ratio of greater than or equal to 2. The CVI survey questions proved to have little value yielding too many false positives.

Effects of Magnifier Training: Evidence from a Camera Built in the Magnifier

F. Nienke Boonstra, PhD, Ralf F.A. Cox, PhD, Annemieke M. Reimer, MSc, et al *Strabismus* June 2012; 20(2): 44-48 (f12)

This study comes from the Bartimeus Institute for the Visually Impaired, Zeist, The Netherlands, and other institutions in The Netherlands.
This study is an evaluation of 11 visually impaired children to determine the effect of an evidence-based magnifier training on viewing behavior in visually impaired children aged 3 to 6 ½ years of age.
This study supplements 3 previously produced studies at the effects of training on magnifier use in visually impaired children. Camera recordings were used to modify the visual behavior and verify the clinical observations of the examiner.

The study concluded that the magnifier training has a positive effect on viewing behavior in the development of dominance and camera observations provide valuable data on children's viewing behavior.

Profile of Visual Functioning as a Bridge between Education and Medicine in the Assessment of Impaired Vision

Lea Hyvarinen, R. Walthes, C. Freitag, and V. Petz
Strabismus June 2012; 20(2): 63-68 (f12)

This study is from the Faculty of Rehabilitation Sciences, Technical University of Dortmund, Dortmund, Germany.
The purpose of this paper is to report the development of the "Profile of Visual Functioning as a framework for information exchange between schools and medical specialists involved in the education or care of children with impaired vision due to brain damage."

The profile of visual functioning for early intervention and special education should follow the principles of the "International Classification of Functioning, Disability and Health, Children and Youth Version" (ICF-CY), a document by the World Health Organization that covers all areas of functioning for the assessment, early intervention, and education of children with special needs. "Profile of Visual Functioning" gives us the framework for working in collaboration between medicine and education. The "Profile of Visual Functioning" enhances cooperation between education and medicine requiring development of a common language and agreement on how a large amount of information is collected. A basic list of measurements and observations covers functions of most children and thus gives a firm structure to the information exchange.

The authors conclude that the profile of "visual functioning", if used by all stakeholders, may improve the quality of assessments for early intervention and special education. "At the same time, schools and hospitals need to jointly arrange further education so that the names of activities start to make sense to doctors and the medical terms to the teachers."

The Dynamic Landscape of Exceptional Language Development

Annemarie Peltzer-Karpf
Strabismus June 2012; 20(2): 69-73 (f12)

This study was performed at Language Development & Cognitive Science Unit, Graz University, Austria
The purpose of this study is to determine what extent sensory deprivation (decreased vision or decreased hearing) influence the dynamics of language development (particularly in children with decreased vision or decreased hearing.

The findings reported in this study are grounded in studies with vision-impaired children compared with sighted peers as controls (age range 18 months and 3 years). Twelve blind children age 6 to 10 years and 20 hearing impaired/deaf children age 5 to 11 years were compared to children with normal sight and normal hearing.

The findings of the study give evidence that language acquisition and sensory impaired children (decreased vision or decreased hearing) follows the same overall developmental path (as seen in non-sensory impaired children). System-specific temporal discrepancies expressed in protracted phase-shifts and delayed increases of variability are most evident in the early phases of development of sensory impaired children.

Self-organizing maps (SOMs) help to visualize individual and group-specific variation. Self-organizing maps (SOMs) form a dynamic framework that is particularly helpful in assessing children with visual and hearing impairment (sensory impaired children) that helps facilitate reliable information considering that a child's potential for developing cognitive and linguistic skills.

COMMENT: **This is a very difficult article that is well outside of the regular structure and framework for ophthalmologists. Nevertheless, it is very helpful in describing a new "dynamic landscape" for the development of language skills in sensory deprived children (both vision and hearing impaired).**

Diagnosing Cerebral Visual Impairment in Children with Good Visual Acuity

Maria van Genderen, Marjoke Dekker, Florine Pilon, and Irmgard Bals
Strabismus June 2012; 20(2): 78-83 (f12)

This is a retrospective study from the Department of Ophthalmology, Bartimeus Institute for the Visually Impaired and Department of Psychology, Bartimeus Institute for the Visually Impaired.
The purpose of this study is to identify elements that could facilitate the diagnosis of cerebral visual impairment in children with good visual acuity and the general ophthalmology clinic.

Clinical characteristics of 30 children with good visual acuity and cerebral visual impairment (CVI) and compared them with 23 children who were referred with a

suspicion of cerebral visual impairment, but proved to have a different diagnosis. Clinical characteristics included medical history, MRI findings, visual acuity, crowding ratio, visual field assessment, and the results of ophthalmologic and orthoptic examination. All patients were given a short cerebral visual impairment questionnaire.

The results of the study showed that 83% of the children with an abnormal medical history (mainly prematurity and perinatal hypoxia) had cerebral visual impairment, in contrast with none of the children with a normal medical history. Cerebral palsy, visual field defects, and partial optic atrophy only occurred in children with cerebral visual impairment. Forty-one percent of the children with cerebral visual impairment had a crowding ratio greater than 2. The authors suggest that this “may be related to a dorsal stream dysfunction”.

All children with cerebral visual impairment and 91% of the children without cerebral visual impairment gave affirmative answers on the cerebral visual impairment questionnaire. In children with a history of prematurity or perinatal hypoxemia is the most important risk factor for cerebral visual impairment. Additional symptoms or risk factors for cerebral visual impairment include documented/neuro-imaging supported cerebral damage, cerebral palsy, visual field defects, partial optic atrophy, and a crowding ratio greater than 2 may also be suggestive of cortical visual impairment. **CORTICAL VISUAL IMPAIRMENT QUESITONNAIRES HAVE NOT PROVED HELPFUL IN SCREENING FOR TRUE CORTICAL VISUAL IMPAIRMENT AS IT YIELDS “TOO MANY FALSE POSITIVES”.**

Can improving prism fusion range with training also improve stereo-acuity?

Watson M, Davis H, Buckley D. BR IR Orthoptic J 2012; (9): 44-48. (f12)

This study tested whether orthoptic exercises to train the prism fusion range in a group of normal subjects would show improvement in the fusion range and whether this treatment also led to improved stereoacuity. A group of 15 students was recruited. They were randomly divided into the treatment group or the control group. The control group was not treated and asked to return in one week and five weeks. The experimental group was given detailed instructions of performing prism fusion range exercises. All were told to practice at 0.33m only. Half the group did base out (convergence range) first and the other did base in (divergence range.) The stereoacuity and fusional ranges were retested after a week. The treated group ceased training and all participants returned 4 weeks later. Results in the experimental group showed that treating at near only also improved distance prism fusion ranges. Stereoacuity also improved. However, both groups performed similarly at week 5. [10 refs]

[Home]

NEURO-OPHTHALMOLOGY

Case Report: Excessive Blinking and Ataxia in a Child With Occult Neuroblastoma and Voltage-Gated Potassium Channel Antibodies

Allen NM, McKeon A, O'Rourke DJ, O'Meara A, King MD. Pediatrics 2012; 129(5):e1348-52. (f12)

This was a case report of a previously healthy 9-year-old girl who presented with a 10-day history of slowly progressive unsteadiness, slurred speech, and behavior change. On examination there was cerebellar ataxia and dysarthria, excessive blinking, subtle perioral myoclonus, and labile mood. The finding of oligoclonal bands in the cerebrospinal fluid prompted paraneoplastic serological evaluation and search for an occult neural crest tumor. Antineuronal nuclear autoantibody type 1 (anti-Hu) and voltage-gated potassium channel complex antibodies were detected in serum. Metaiodobenzylguanidine scan and computed tomography scan of the abdomen showed a localized abdominal mass in the region of the porta hepatis. A diagnosis of occult neuroblastoma was made. Resection of the stage 1 neuroblastoma and treatment with pulsed corticosteroids resulted in resolution of all symptoms and signs. Excessive blinking has rarely been described with neuroblastoma, and, when it is not an isolated finding, it may be a useful clue to this paraneoplastic syndrome. Although voltage-gated potassium channel complex autoimmunity has not been described previously in the setting of neuroblastoma, it is associated with a spectrum of paraneoplastic neurologic manifestations in adults, including peripheral nerve hyperexcitability disorders.

A case of lorazepam (Ativan)-induced accommodation paresis

J J Jung, S-H Baek and U S Kim

Eye. April 2012;26:614. (f12)

The authors report one case of a 23 year-old man who developed blurred vision at near due to sudden onset of accommodation paresis. The symptoms were temporally correlated with starting lorazepam, and subsided over weeks after discontinuing lorazepam. There was no rechallenge with the drug in this patient. Sudden loss of accommodation has been reported with head trauma, encephalitis, oculomotor nerve palsy, uveitis, and viral diseases and drugs such as tricyclic antidepressants and phenothiazines.

A new perimeter using the preferential looking response to assess peripheral visual fields in young and developmentally delayed children.

Allen LE, Slater ME, Proffitt RV, et al. J AAPOS 2012;16:261-265. (f12)

The authors developed a preferential looking perimeter (PLP) to assess visual fields. They compare this method with confrontation testing on a group of young children, both with and without visual fields defects. These children were unlikely to be able to perform automated static perimetry because of their age, or behavioral and learning difficulties. Enrolled children were between ages 3 and 10. There were 32 controls and 42 children with neurological or ocular pathology that could cause visual field loss. For the controls, PLP was normal in all 32 and 30/32 had normal confrontation visual fields (2 were uninterpretable). In the group with pathology, 18 had field defects confirmed by confrontation, 5 showed no field defects, and 19 had uninterpretable results. PLP demonstrated the same pattern of visual field loss in the 18 with field defects and normal visual fields in the 5 with no field defects on confrontation. Of the 19 with uniterpretable results on confrontation, PLP showed 7 with normal visual fields, 6 with visual field defects, and 6 with uninterpretable results. Interobserver agreement was present in 138 of 140 PLP tests. Based on confrontation as the gold standard, PLP showed a sensitivity and specificity of 100% to detect similar visual field defects. PLP was also interpretable in 31% more patients than confrontation. It was not always possible in this study to mask the child's pathology on confrontation testing. This test may not pick up smaller or milder visual field loss.

Positron Emission Tomography – Letter to the Editor-Case Report

Angelina Cistaro, MD, Natale Quartuccio, MD, Sara Vesco, MD, Marco Pagani, PhD, et al.*Ophthalmology* July 2012; 119:1496-1497 (f12)

This letter reports on the correlation of neuro-ophthalmological findings with a F-fluorodeoxyglucose positron emission tomography-computed tomography study (F-FDG-PET/CT) in a pediatric patient affected by a pure right homonymous hemianopia manifested after a sports related head injury. The patient had no evidence of any neuro-radiologic abnormalities or abnormal visual evoke response testing.

The patient was 14 years of age, right-handed and had a history of nearsightedness. Past medical history is negative for smoking, alcohol, substance abuse, psychiatric disease and previous major traumas. In January 2009 the patient underwent a traumatic brain injury secondary to "uncontrolled ascent" during a volleyball game. Subsequent computed tomography (CT) and magnetic resonance imaging (MRI) showed a "cystic hypodense mass between the lateral ventricles." There was no apparent involvement of the cerebral cortex. Inconsistencies occurred between objective and subjective findings. The patient

also reported transient improvements in his right homonymous hemianopia. There was consideration of the possibility of nonorganic/psychogenic visual loss as well as atypical migraine reviewed and was subsequently discarded.
The patient was referred to the PET Center in November 2009 and underwent a 3D 18F-FDG-PET/CT study. Statistical parametric mapping analysis showed areas of hypometabolism on the left primary visual cortex, left associative visual cortex, left parahippocampal gyrus and thalamus. The PET/CT study demonstrated hypometabolic areas congruent with the symptomatology according to the visual field defect.

The purpose of the paper is to demonstrate that in the absence of any demonstrable structural lesion causing the right homonymous hemianopia in this patient, subtle posttraumatic occipital dysfunction that is best discerned by PET/CT scan, which is felt to be superior to computed tomography and magnetic resonance imaging (including functional MRI-fMRI).
This study is supportive of previous reports by Itoh et al, in the Japanese *Journal of Ophthalmology* 1987. The patient had a full recovery of the right homonymous hemianopia. He returned to school to participate in academic and athletic activities.

Cat Scratch Disease – Letter to the Editor-Correspondence

Clare L Fraser, MBBS, Susan Sanchez, MS, PhD, Nancy J. Newman, MD, Valerie Biousse, MD
Ophthalmology July 2012; 119:1502-1503 (f12)

This letter refers to the previously written article “Clinical Characteristics in 53 patients with cat scratch optic neuropathy” by Chi et al (*Ophthalmology* 2012: 119:183-187). This letter articulates the substantial advancements in our understanding of *Bartonella henselae* (*B. henselae)* in humans.
In North America, the prevalence of *B. henselae* antibodies in cats ranges from 4-7% in the Midwest and 60% in the Southeast with the rates being higher in feral cats. The recommendation from the Infectious Department of the College of Veterinary Medicine, University of Georgia, is that if a cat scratches a person, the wound should be washed immediately with warm soapy water. In addition, cats should not be allowed to lick any open wound on a human.

The transmission of *Bartonella* organisms to humans occurs primarily through cutaneous trauma caused mainly by a scratch from a cat and less likely from a cat bite. Shedding of *B. henselae* in cat saliva has not been clearly documented. *B. henselae* is found in the digestive tract of cat fleas and flea feces. Therefore, the main source of infection appears to be the feces inoculated by contaminated cat claws. The cat itself may not be infected.
The diagnosis of *Bartonella* infection can be determined by polymerase chain reaction, direct culture and blood serum antibodies for *B. henselae*.

The main recommendation of the article is that infection can occur, particularly in cats with infected fleas. Antibody therapy does depress bacteremia in cats, but does not alter the duration of infection. Recurrent bacteremia is common.
Most cats that carry the *B. henselae* bacteria have subclinical infections. Even a cat without apparent bartonellosis must be regarded as a possible reservoir of infection for a minimum of 2-3 months. The main focus of attention should be flea control. Fleas are usually involved in the transmission among cats and the use of acaracide products to eliminate fleas from the environment is critical to also decrease the risk of transmission from cats to humans.

Clinicopathologic and Molecular Analysis of Choroidal Pigmented Schwannoma in the Contest of a PTEN Hamartoma Tumor Syndrome

Giulia Venturini, MSc, Alexandre P Moulin, MD, Manuel Deprez, MD, PhD, et. al.
Ophthalmology April, 2012; 119:857-864 (f12)

The purpose of the study is to report the first case of a choroidal schwannoma in a patient affected by PTEN gene mutation with associated hamartomatous tumor.
The PTEN hamartoma tumor syndrome (PHTS) is an autosomal dominant disorder caused by various mutations in the tumor-supressor gene phosphatase and tensin homolog (PTEN), which is located on the long arm of the 10^{th} chromosome.
The major diagnostic criteria for this syndrome are macrocephaly and breast and thyroid cancer. The minor criteria for this syndrome include lipomas, fibrocystic disease of the breast, hamartomas, intestinal polyps and fibromas. The clinical spectrum of PHTS encompasses several syndromes with variable ocular involvement including Cowden syndrome, Bannayan-Riley-Ruvalcaba syndrome and possibly Proteus syndrome.
The study reports a 10-year-old female patient affected by PHTS who developed unilateral choroidal schwannoma that led to the enucleation of her eye. The molecular involvement of PTEN in this rare intraocular tumor (described for the first time in the context of PHTS) and possible contribution of the NF tumor-suppressor gene in this condition are investigated. This rare tumor showed a unique combination of reduction of PTEN and the absence of NF2 expression.

This study was carried out by the Department of Medical Genetics, Ophthalmology and Pathology at the University of Lausanne, Lausanne, Switzerland.
COMMENT: Comprehensive article describing the PTEN hamartoma and the PHTS syndrome. There is also an excellent discussion of similar syndromes as mentioned above.

Longitudinal Measures of Visual Function, Tumor Volume, and Prediction of Visual Outcomes after Treatment of Optic Pathway Gliomas

John P. Kelly, PhD, Sarah Leary, MD, Paritosh Khanna, MD, Avery H. Weiss, MD
Ophthalmology June, 2012; 119:1231-1237 (f12)

This is a retrospective cohort study designed to examine the longitudinal changes in visual acuity, tumor volume, and visual evoke potentials before and after treatment in children with optic pathway gliomas.
21 patients 0.7 to 9 years of age were initially treated either by chemotherapy (18 patients) or with radiotherapy (3 patients). The patients were followed up with serial magnetic resonance imaging, age corrected visual acuity measurements and visual evoke potential. Longitudinal visual outcome data were obtained on average for 9 years (range 4-16 years). Tumor volumes before and after treatment were estimated in 15 patients. Multivariate regression was use d to predict visual outcomes.

Before treatment, 81% of patients had reduced visual acuity and 81% had optic nerve pallor/atrophy. All patients in this study had reduced visual evoke potential in 1 or both eyes.

After initial treatment tumor volume decreased in 53%, stabilized in 27% and progressively increased in 20%. Treatment arrested the rapid decline of visual acuity loss and stabilized visual acuity for 4-5 years. The rate of visual acuity decline was correlated with tumor shrinkage. 62% of patients required additional treatment with either chemotherapy or radiation because of tumor growth or progressive loss of visual function.
Systemic chemotherapy arrested the decline in visual acuity and stabilized vision on average for 5 years. At presentation, visual evoke potentials were a more sensitive indicator of optic pathway damage than visual acuity or optic nerve appearance. Although tumor reduction or stabilization was achieved in 80% of patients, pre-existing visual damage (documented by objective measurements of tumor volume and visual acuity) limited visual outcomes.

This study was performed at the University of Washington Medical Center and the Seattle Children’s Hospital in Seattle, Washington. The Department of Hematology/Oncology and Radiology and Ophthalmology participated in this study.
COMMENT: The conclusions of the article regarding the clinical value of visual evoke potentials being a “more sensitive indicator of optic pathway damage and visual acuity or optic nerve appearance”, is not a common clinical sentiment. This may indicate potential changes in using standard work-up of children prior to receiving chemotherapy.

Optic disc measurements in full term infants Yogavijayan Kandasamy, Roger Smith, Ian M R Wright, Leo Hartley Br J Ophthalmol 2012;96:662-664. (f12)

Background: The objectives of this study were to measure optic disc size in full term infants and to determine whether this value is influenced by sex or birth weight.
Methods Retinal images from a cohort of full term infants admitted to a tertiary perinatal centre were obtained using a retinal camera.
Results: 35 images of left and right eyes from 35 infants were assessed

Conclusion: Birth weight and sex did not influence the size of the optic disc in term infants. There were no differences in optic disc measurements between male
and female infants and between low birth weight and normal birth weight infants.

Comments: Small sample size, refractive errors are not taken into consideration. But this is a new information.

Maternal Autoantibodies in Autism. Braunschweig D, Van de Water J. Arch Neurol 2012; (June) 69: (6) 693-699. (f12)

Epidemiological studies continue to document a rising increase in rates of ASD in all parts of the world. The underlying mechanism and causative factors are still not fully understood. Recent studies of the immune system in ASD are increasing with several groups identifying fetal protein reactive IgG antibodies in maternal plasma of ASD children. These authors review the current literature in the area of maternal fetal brain-reactive antibodies and other utero immune–related exposures and propose several directions for future research. These include investigating whether subsequent children born to mothers with brain-reactive antibodies have a higher risk of developing autism than the current published familial recurrence rates of 19 percent. Another research approach would identify the target antigens and the affected pathways leading to the behavioral outcome of autism. [50 refs]

Organization in Autism. Kana R, Percy A. Arch Neurol 2012; (July) 69: (7) 906-907. (f12)
This commentary summarizes some of the current research on the causes of autism and should be of interest to those in ophthalmology practice who see ASD patients. The authors address a study recently published on neuron number and size in the prefrontal cortex of children with autism. (Courchesne et al, JAMA 2011 [306(18):2001-2010.) The journal includes the original abstract with this commentary for some interesting reading. The article concluded that brain overgrowth in males with autism involved an abnormal excess number of neurons in the pre-frontal cortex. These authors ask an important question about

the impact of these excess neurons on the functional and anatomical connections in autistic brains. They propose that increased neuronal count may result in altered patterns of connectivity. They suggest that the neuron count be examined in the temporal and parietal lobes and that research be directed to molecular associations to see if they correlate with neuroanatomic findings. [13 refs]

Fractionation of social brain circuits in autism spectrum disorders.

Gotts SJ, Simmons WK, Milbury LA, Wallace GL, Cox RW, Martin A. Brain 2012; (Sept) 135(9): 2711-2725. (f12)

Autism spectrum disorders are developmental disorders characterized by impairment in social and communication abilities along with restricted interests and repetitive behaviors. Converging neuroscientific evidence has shown that ASD-related neuropathology is not localized to one brain region but involves connectivity at the level of whole brain networks. These areas include frontal, temporal and parietal circuits. The authors define the “social brain” as a set of brain regions commonly co-activated across a range of social tasks.
Taking a cohort of 31 high-functioning ASD participants between 12 and 23 years of age with a matched group of 29 typically-developing participants, the authors used fMRI to study the brain circuits during various social activities. The figures in this article are in glorious color and well-captioned for an interesting and thorough review on the topic of ASD. [Multiple references.]

The changing landscape of traumatic brain injury research.

(Editorial) The Lancet Neurology 2012; (Aug) 11: (8). (f12)
This editorial describes a coordinated effort between major funding agencies of neurological research in North America and Europe to address the increasing incidence of traumatic brain injury. TBI is an enormous public health problem and affects all age-groups in all populations. Even in poorer regions, the incidence has been estimated at 150 to 300 per 100,000 individuals per year. The incidence is increasing in low-income and middle-income countries, mostly due to road traffic accidents. In Europe, the case-fatality rate is 3 percent, but is higher in the USA at 6.2 percent. 1-2 percent of TBI survivors live with permanent disabilities. However, few improvements in clinical therapy for TBI have been approved by regulatory agencies.
TBI is predicted to become the third leading cause of global mortality and disability by 2020. Evidence suggests that TBI is a risk factor for dementia, substance abuse and other psychiatric disorders. The number of patients requiring care for eye movement disorders caused by TBI will also increase.

Cognition and Eye Movement: Assessment of Cerebral Dysfunction. White OB, Fielding. J. J Neuro-ophthalmol. (Sept) 2012; 32: 226-273. (f12)
In this state-of-the-art review the authors explore the relationship between cognitive sensory processing and ocular motor abnormalities in neurological disease. Many neurological disorders show deficits in ocular motor function. Most of these are caused by lesions in the brainstem and cerebellum, but disorders that affect the cerebral hemispheres also disrupt cognitive processes. The cognitive control of eye movements requires synchronized circuits between the frontal and parietal lobes as well as interacting with subcortical nuclei. The authors compare cognitive sensory processing with the execution of planned ocular motor tests in Parkinson disease, Huntington disease, and multiple sclerosis. Assessing cognitive function as part of a directed examination of eye movements may provide insight and diagnostic information into cerebral dysfunction. [81 Refs]

The interface of infectious disease and neuro-ophthalmology. Berger JR. J Neuro-Ophthalmol (Sept) 2012; 32: 195-196. (f12)
Although aimed more at neuro-ophthalmology practices, this short two-page article summarizes five articles in the same issue of the journal that have in common an infectious cause for a neuro-ophthalmic disorder. It is valuable because it mentions diseases that do not commonly come to mind in routine differential diagnosis, the most fascinating being the increase in certain fungal infections (invasive fungal sinusitis) as a result of hurricane exposure. One article describes a 90-year-old woman with ptosis as a result of Lyme disease. Serology should be helpful, but is not always positive, as in this patient, and in another article on neuroretinitis from cat scratch disease. A fourth article eventually identified leprosy in a 69 year-old Caucasian woman as the cause of trigeminal and facial palsy. The final case report was on a patient with epileptic nystagmus caused by herpes simplex virus. Every clinician should recognize that nystagmus coupled with an altered level of consciousness may indicate an underlying seizure disorder. [15 Refs]

Auditory-olfactory synesthesia coexisting with auditory-visual synesthesia. Jackson TE, Sandramouli S. J Neuro-Ophthalmol (Sept) 2012; 32: 221-223. (f12)
This is a fascinating account of a patient who reported “seeing sounds” from the age of five. Certain sounds also elicited smells. He thought that this was a normal sensation but later learned that it was unique to him. He was not looking for treatment, but sought a formal diagnosis to help him understand his condition and allow him to explain it to others. He joined other famous individuals who reportedly had auditory-visual synesthesia including Rimsky-Korsakov, Franz Liszt, Duke Ellington and David Hockney.
The short article includes detailed discussion of this individual case and the entity itself. Synesthesia is a condition rarely encountered by ophthalmologists because

of its rare presentation. However because of its visual symptoms, patients may seek advice in an ophthalmology setting. [12 Refs]

Relapsing dorsal midbrain syndrome following interventions for hydrocephalus in aqueductal stenosis. Apkarian AO, Garton HJL, Wesolowski J, Trobe J. J Neuro-Ophthalmol (June) 2012; 32: 124-127. (f12)
This is a detailed case report of a 9-year-old girl who developed transient loss of consciousness after a 6-month history of headaches, unsteady gait, urinary incontinence and poor performance in school. She was diagnosed with hydrocephalus and noted to have optic nerve damage from undiagnosed papilledema, and a relative afferent pupillary defect OS. Seventeen days after shunting, most of her neurological symptoms were resolving and her eye movements were normal. At day 56 there was no change. At day 64, 12 days after a shunt revision, she reported diplopia. Convergence-retraction eye movements were present on upgaze.
The article describes relapsing episodes of DMS with shunt interventions. It is helpful in supporting the theory that rapid enlargement of the ventricles with deformation of the dorsal midbrain, not an alteration in ICP, is the mechanism of DMS. [6 Refs]

Unilateral straight hair and congenital Horner Syndrome. Wang FM, Wertenbaker C, Cho H, Marmor, MA, Ahn-Lee SS, Bernard BA. J Neuro-Ophthalmol (June) 2012; 32: 132-134. (f12)
The authors present three cases of congenital Horner syndrome in which the patients had straight hair ipsilateral to the affected eye. They give detailed discussion on the possible mechanisms for this phenomenon. It is unknown at what stage of development a sympathetic pathway lesion would lead to hair curliness, but as one child developed the Horner syndrome from a birth injury, it is less likely that it occurs in utero. Human hair shape is influenced by a number of factors aside from genetic background. With loss of sympathetic innervation on the affected side, the diminished adrenergic signal to the hair follicles might lead to straighter hair. One would expect that asymmetry of curliness would occur only in preganglionic lesions, as in the three reported cases. [Good photos; 14 Refs.]

Pursuit-paretic and epileptic nystagmus in MELAS. Choi SY, Kin Y, Oh SW, Jeong S-H, Kim JS. J Neuro-Ophthalmol (June) 2012; 32: 135-138. (f12)
The presence of acquired nystagmus usually indicates a brainstem or cerebellar lesion. Cerebral hemispheric lesions rarely give rise to nystagmus. The authors report a 27-year-old man with known MELAS mutation who developed dizziness and blurred vision after heavy drinking four years before. At that time, he was noted to have a left homonymous hemianopia, visual blurring and headache. The visual field defect resolved. Two years later he suffered a seizure after drinking alcohol. At the recent examination he had a left homonymous hemianopia and

spontaneous left-beating nystagmus in primary gaze. Intermittently, the patient exhibited altered mental status, the head turned to the right and left beating nystagmus could be observed. Each episode lasted for several minutes and recurred several times. Epileptic nystagmus has been attributed to irritation of the cortical areas involved in generating eye movements. [Good videos; 12 Refs.]

Paralysis of accommodation with preserved pupillary function as the initial manifestation of Guillain-Barré Syndrome. Fausett BV, Trobe JD. J Neuro-Ophthalmol (June) 2012; 32: 148-149. (f12)
This is a case report of a 7-year-old boy with accommodative loss as the first clinical sign of GBS. His pupillary light reflexes were normal. Three days later he developed dysarthria and ataxia and was admitted to hospital. He was treated with intravenous immunoglobulin and underwent physical therapy. His ophthalmologic and neurologic symptoms resolved, except for reduced deep tendon reflexes. Accommodative paralysis has been reported to occur as the first sign in diphtheria. Adie syndrome includes accommodative dysfunction but the iris sphincter signs are more prominent. The article describes the clinical features of GBS and discusses the pathways involved in sparing the iris sphincter. [Refs 11]

Orbital radiation therapy for thyroid eye disease. Kazim M, Garrity JA. J Neuro-Ophthalmol (June) 2012; 32: 172-176. (f12)
This article is a “point counterpoint” format with discussion by two experts, one for and one against orbital radiotherapy in TED. Much progress has been made in understanding the pathophysiology of TED but therapeutic methods have not advanced at the same rate. Treatment to slow or reverse the active phase is still challenging. ORT is a successful alternative to steroids according to Dr Kazim. He emphasizes case selection because there is only a small window in which the condition is still amenable to therapy. He recommends ORT in cases of progressive limitation of eye movement. He combines ORT with corticosteroids in optic neuropathy to try to avoid surgical decompression. He believes that ORT should not be used on mild TED or in the stable or near-stable phase.

Dr Garrity argues against ORT in TED. The value of medical care is equal to outcomes divided by costs, and he states that by many accounts there is limited value for ORT as a form of therapy. He reviews many studies and concludes that in non-sight threatening TED, ORT has a negligible effect on proptosis or lid position. Although motility may improve in some patients, the changes may not translate into improved health-related quality of life. In weighing the costs, outcomes, risk of complications, and frequent need for additional therapy he states that ORT should be used only in rare circumstances. Further studies are needed to establish more appropriate anti-inflammatory and immunomodulatory therapy. Both authors contributed a rebuttal. The combined opinions provide a current update and good reference list on the topic. [22 Refs.]

Periodic unilateral eyelid retraction in a pediatric patient: Some considerations on botulinum toxin as a temporary treatment option. Denadai R, Bastos EM. J Neuro-Ophthalmol (June) 2012; 32: 189-193. (f12)
In a couple of succinct paragraphs, the authors comment on the paper in a previous issue of this journal (Ghandi NG et al. 2011; 31: 35-352) in which a pediatric patient with unilateral periodic (cyclic) lid retraction underwent surgical repair. Drs Denadai and Bastos suggest the use of botulinum toxin as an alternate treatment method. There followed a formal comment and response from Drs Ghandi and Allen, who also discuss the relative costs and long term outcome. Both include references which would provide further insight into this unusual entity as well as offering other management ideas for the interested reader. [5 Refs; 12 Refs]

Eight-and-a-Half Syndrome. Skaat A, Huna-Baron R. Arch Neurol 2012; (July) 69: (7) 934-935. (f12)
This eye-catching title refers to a rare condition in which a patient with a one-and-a-half syndrome has an associated facial, or VIIth nerve palsy. The term was coined by Eggenberger in 1998. This manifests as a conjugate gaze palsy to one side, an internuclear ophthalmopegia to the other, and a facial palsy ipsilateral to the gaze palsy. The lesion affects the paramedian pontine reticular formation in the region of the abducens nucleus with interruption of the medial longitudinal fasciculus to the other side, but also involves the facial colliculus.

[Home]

NYSTAGMUS

Extraocular Muscles in Patients With Infantile Nystagmus Adaptations at the Effector Level Kathleen T. Berg, BS; David G. Hunter, MD, PhD; Erick D. Bothun, MD; Rosalia Antunes-Foschini, MD; Linda K. McLoon, PhD. Arch Ophthalmol. 2012;130(3):343-349 (f12)

The purpose of this study was to test the hypothesis that the extraocular muscles (EOMs) of patients with infantile nystagmus have muscular and innervational adaptations that may have a role in the involuntary oscillations of the eyes. Specimens of EOMs from 10 patients with infantile nystagmus and postmortem specimens from 10 control subjects were prepared for histologic examination. The following variables were quantified: mean myofiber cross-sectional area, myofiber central nucleation, myelinated nerve density, nerve fiber density, and neuromuscular junction density. The EOMs of patients with infantile nystagmus

displayed a distinct hypoinnervated phenotype including more centrally nucleated myofibers and a greater degree of heterogeneity in myofiber size than did controls. This represents the first quantification of changes in central nucleation and myofiber size heterogeneity, as well as decreased myelinated nerve, nerve fiber, and neuromuscular junction density. These results suggest that deficits in motor innervation are a potential basis for the primary loss of motor control.

Living with nystagmus: a qualitative study

Rebecca Jane McLean, Kate C Windridge, Irene Gottlob Br J Ophthalmol 2012;96:981-986 (f12)

Background/aims: To identify aspects of daily livingaffected by nystagmus. This was done by semistructured interviews.

Results: Analysis identified six domains that were adversely affected by nystagmus; visual function, restriction of movement, standing out/not fitting in, feelings about the inner self, negativity about the future and relationships. Cosmetic appearance of nystagmus, including others' avoidant response to this, was described (n.18), as was others' failure to recognize what it is like to have nystagmus (n.18). Driving issues were frequently raised (n.19) and restrictions in occupation choice/opportunities (n.17) were highlighted. Reliance on others (n. 16) also emerged.

Conclusion: Interviews revealed universally negative experiences of living with nystagmus that are previously unreported. Findings are similar to studies conducted for strabismus.

Comments: The six domains that emerged provide the necessary evidence to develop a patient-derived, disease-specific, QOL tool for nystagmus, allowing questionnaire items to be written that are grounded in the perspectives of individuals with nystagmus.

Intracranial Pathology in Young Children With Apparently Isolated Nystagmus

Mansoor A Shammari, MD; Sahar M. Elkhamary, MD; Arif O. Khan, MD J Pediatr Opthalmol Strabismus 2012; 49:242-246 July/August(f12)

The purpose of this article was to assess the incidence of intracranial pathology in children with nystagmus noted in the first few years of life using brain magnetic resolution imaging (MRI). A retrospective review of children up to 5 years old was conducted and those who had a diagnosis of nystagmus and had MRI of the brain were reviewed. The study found that brain MRI conducted with a normal ocular examination did not lead to significant MRI findings. The only ocular finding that seems to lead to MRI findings in patients with nystagmus was optic

nerve pallor. Clinical description of nystagmus also did not predict intracranial pathology in this study.

Albino Mice as an Animal Model for Infantile Nystagmus Syndrome Ghislaine L. Traber, Chien-Cheng Chen, Ying-Yu Huang, Marcella Spoor, Jeanine Roos, Maarten A. Frens, Dominik Straumann and Christian Grimm. IOVS August 2012 53:5737-5747; published ahead of print July 12, 2012, doi:10.1167/iovs.12-10137 (f12)

Individuals with oculocutaneous albinism are predisposed to visual system abnormalities affecting the retina and retinofugal projections, which may lead to reduced visual acuity and Infantile Nystagmus Syndrome (INS). Due to absence of an established mammalian animal model, mechanisms underlying INS remain elusive. In this study, we screened wild-type mice of varying pigmentation for ocular motor abnormalities in order to identify a possible mouse model for INS. Three albino mouse strains (CD1, BALB/c, DBA/1), and two normally pigmented strains (129S6, C57BL/6) were screened using infrared oculography. Varying visual stimuli (black or white background, stationary pattern, optokinetic, i.e., horizontally rotating pattern) were displayed to the full (fVF) or anterior visual field (aVF) of the restrained mouse. The authors found spontaneous nystagmus, specifically jerks and oscillations, in albino mice under all experimental conditions. Median eye velocity was between 0.8 and 3.4 deg/s, depending on the strain. In contrast, the eyes in pigmented mice were nearly stable with a median absolute eye velocity of below 0.4 deg/s. In albino mice, fVF optokinetic stimuli elicited an optokinetic response (OKR) in the correct direction, albeit with superimposed oscillations. However, aVF optokinetic stimuli evoked reversed OKR in these strains, a well-known feature of INS.

This work presents a promising new mammalian model to study mechanisms of Infantile Nystagmus Syndrome (INS).

Oblique muscle surgery for treatment of nystagmus with head tilt. Lueder GT, and Galli M. J AAPOS 2012;16:322-326. (f12)

The authors report a surgery on the oblique muscles to treat nystagmus patients with torticollis. This was a retrospective surgery of six patients who underwent a superior oblique tendon anterior 50% tenectomy and a contralateral inferior oblique muscle recession. The patients were aged 2 to 13 years. Mean follow-up was 2.2 years. Head tilt improved by an average of 28’. Half of the patients had some improvement in acuity postoperatively as well. This was a small, retrospective study with no masked observers. The authors seem to be somewhat confused in their discussion of other nystagmus surgical techniques.

They confuse improvement in head tilt with improvement in nystagmus waveform and visual function when discussing the surgical procedure performed by Hertle and colleagues.

[Home]

ROP

Prediction of Retinopathy of Prematurity Using the Screening Algorithm WINROP in a Mexican Population of Preterm Infants

Luz Consuelo Zepeda-Romero, MD, MSc; Anna-Lena Hård, MD, PhD; Larissa Maria Gomez-Ruiz, MD, et al. Arch Ophthalmol. 2012;130(6):720-723 (f12)

The purpose of this paper is to retrospectively validate the WINROP (weight, insulin-like growth factor I, neonatal, retinopathy of prematurity [ROP]) algorithm in identification of type 1 ROP in a Mexican population of preterm infants. The study evaluated infants admitted to the neonatal intensive care unit at Hospital Civil de Guadalajara from 2005 to 2010. In this Mexican population of preterm infants, WINROP detected type 1 ROP early in 84.7% of very preterm infants and correctly identified 26.6% of infants who did not develop type 1 ROP. The authors felt that uncertainties in dating of pregnancies and differences in postnatal conditions may be factors explaining the different outcomes of WINROP in this population.

Spectral-Domain Optical Coherence Tomographic Assessment of Severity of Cystoid Macular Edema in Retinopathy of Prematurity

Ramiro S. Maldonado, MD; Rachelle O'Connell, BS; Simon B. Ascher, BS, et al. Arch Ophthalmol. 2012;130(5):569-578. (f12)

This study was performed to investigate whether the severity of cystoid macular edema (CME) in neonates who were 31 to 36 weeks' postmenstrual age, as viewed by spectral-domain optical coherence tomography (SD-OCT) imaging, predicts the severity of retinopathy of prematurity (ROP) or is related to systemic health. 42 neonates were studied had at least 1 SD-OCT imaging session prior to 37 weeks' postmenstrual age and prior to ROP laser treatment, if a laser treatment was performed, and an oph thalmic ROP examination at or after 41 weeks' postmenstrual age, evidence of complete retinal vascularization in zone III, or documentation through telephone report of such information after transfer of care. Cystoid macular edema is common in premature infants (50% of neonates in this study) screened for ROP before 37 weeks' postmenstrual age, with the most common SD-OCT phenotype of a bulging fovea from multiple

elongated cystoid spaces. Detection of CME is not associated with ROP severity; however, tomographic thickness measurements could potentially predict a higher risk of requiring laser treatment or developing plus disease or ROP stage 3. Systemic health factors are probably not related to the development of CME.

A predictive score for retinopathy of prematurity in very low birth weight preterm infants

G U Eckert, J B Fortes Filho, M Maia and R S Procianoy
Eye. March 2012;26:400-406 (f12)

The authors developed a scoring system (ROPScore) applied at the age of six weeks and hypothesized that it might serve as better predictor of any stage ROP or ROP severe enough to necessitate treatment than birth weight (BW) and gestational age (GA). A prospective cohort of preterm infants with BW < 1500 g and/or GA <32 weeks was studied. The score was developed based on BW, GA, proportional weight gain from birth to the 6th week of life, use of oxygen in mechanical ventilation, and need for blood transfusions from birth to the 6th week of life. The score was established after linear regression, considering the impact of each variable on the occurrences of any stage and severe ROP. Receiver operating characteristic (ROC) curves were used to determine the best sensitivity and specificity values for the score.

The sample included 474 patients from a tertiary level hospital in Brazil. The area under the ROC curve for the score was 0.77 and 0.88 to predict any stage and severe ROP, respectively. These values were significantly higher for the score than for BW (0.71) and GA (0.69) alone. The scoring system was simple enough to be routinely used by ophthalmologists during screening examination for detection of ROP. It corroborates other studies that have found poor postnatal weight gain to be predictive of severe ROP. The authors plan to prospectively validate the ROPScore both within and outside their institution.

Training fellows for retinopathy of prematurity care: A Web-based survey

. Wong RK, Ventura CV, Espiritu MJ, et al. J AAPOS 2012;16:177-181. (f12)
A web-based survey, characterized the training received by fellows in Pediatric Ophthalmology and Retina in ROP examination and management. The survey was completed by fellows and their supervising attendings. 40% of programs responded to the inquiry and 100/123 surveys that were e-mailed were completed. This represents an actual survey response of 30% (43% Pediatric Ophthalmology and 24% Retina). Number of ROP examinations performed, was independent of specialty and of responder type (attending vs. fellow). Pediatric fellows tended to be supervised by an attending more frequently for examinations. Retina fellows tended to perform more laser treatments. Reported

self-confidence in ROP management increased dramatically by the end of their fellowship training. Most programs did not report formal evaluations of ROP examinations. Some of the comparisons between Pediatric Ophthalmology fellows and Retina fellows may be biased by the discrepancy in length of fellowship. Many programs were not included in the data since they chose not to participate.

Digital image analysis in retinopathy of prematurity: A comparison of vessel selection methods

Wilson CM, Wong K, Ng J, et al. J AAPOS 2012;16:223-228. (f12)

The authors attempt to determine the optimal method of vessel selection for distinguishing eyes with and without ROP and between ROP stages when semiautomatically quantifying the associated vessel changes from digital retinal images. Color digital images were acquired over a four-year period. 75 images from 75 infants were used. Tortuosity of retinal arterioles and venules increases significantly with ROP presence and severity, and the authors were able to measure this. The increase in tortuosity was greater for arterioles. Vessel width did not increase with either the presence of ROP or with increasing severity of ROP. The lack of vessel width increase contradicts clinical observation, and may represent a lack of resolution ability of the camera images, or the inability of the computer program to detect these differences. Only the results of the 4 most tortuous vessels need to be analyzed. This paper has implications for telemedicine ROP screening possibilities.

Telemedicine detection of type 1 ROP in a distant neonatal intensive care unit.

Weaver DT and Murdock TJ. J AAPOS 2012;16:229-233. (f12)

The authors attempted to demonstrate the feasibility of telemedicine screening for ROP, using the RetCam imaging system. Over a 4.5 year period, premature infants were remotely screened for ROP with a RetCam II by a RN and a neonatal NP. The images were evaluated by one of two Pediatric Ophthalmologists. 582 telemedicine examinations were performed on 137 infants. 13 of these infants (9.5%) were transferred to another hospital based on referral-warranted ROP. The birth weight of the transferred babies was 838grams and the mean gestational age was 26 weeks. This is in comparison to 1239 grams and 29 weeks respectively, for nontransferred infants. These differences were statistically significant. Of the 13 transferred infants, 5 were not treated initially (one was treated much later as an outpatient). All 17 eyes that were treated had a good outcome with no progression to stage 4 or 5 ROP. None of the nontransferred infants required treatment. Telemedicine screening on a weekly basis as utilized in this study had a sensitivity of 100% and a specificity of 96.3% for detecting type 1 ROP. Potential monetary savings are dramatic. The study is limited somewhat by its retrospective nature and small sample size.

Fluorescein angiography to estimate normal peripheral retinal nonperfusion in children.

Blair MP, Shapiro MJ, and Hartnett ME. J AAPOS 2012;16:234-237. (f12)

In children without acute ROP< there is no systematic description of peripheral nonperfusion. Knowledge of normal nonperfusion is important, when evaluating children and their relatives for diseases which affect the peripheral retina. The authors performed fluorescein angiography in conjunction with scleral indentation to image the ora serrata region using the RetCam system during examinations under anesthesia. Most evaluations were of the fellow eye of patients with unilateral disease, or posterior pole disease. 33 eyes of 31 consecutive patients were included. The mean age at angiography was 3.8 years. The mean average of nonperfusion was 0.9 disc diamters (DD) temporally and 0.6 DD nasally. No eye had a measurement >1.5 DD. There was no correlation between age and width of nonperfusion. The authors feel that this data is applicable to the general pediatric population despite the presence of ocular disease. Based on there data, they feel that 2 DD of nonperfusion should be considered abnormal. There was a slight imbalance of female patients to male (18 to 13), but this should not have affected the data significantly.

Delayed onset atypical vitreoretinal traction band formation after an intravitreal injection of bevacizumab in stage 3 retinopathy of prematurity

Lee BJ, Kim JH, Heo H, Yu YS.

Eye. Jul 2012;26:903-10. (f12)

This is a case series of patients who presented with an atypical retinal traction band after intravitreal injection of bevacizumab for stage 3 retinopathy of prematurity (ROP). Three cases (five eyes) are described. The three patients were ex 28, 30 and 32 weeks gestational age who were treated with both laser and bevacizumab for zone 1 with plus or posterior zone 2 with plus stage 3 ROP. They developed an atypical fibrous traction band arising from the major vascularized arcades, not from the border between vascular and avascular retina. Another unusual feature was that the fibrous traction band developed 2.5 to 4 months after the retinas showed regressed ROP. In three eyes, a tractional retinal detachment developed, which could not be successfully treated.
To detect this complication, ROP patients who receive bevacizumab treatment should be closely followed for more than 4 months after the treatment, even if the disease seems to have regressed.

The association between respiratory tract Ureaplasma urealyticum colonization and severe retinopathy of prematurity in preterm infants ≤1250 g

Ozdemır R, Sarı FN, Tunay ZO, Erdeve O, Canpolat FE, Oguz SS, Uras N, Dılmen U.

Eye. Jul 2012;26:992-6. (f12)

Aim: To evaluate the association between respiratory tract Ureaplasma urealyticum (Uu) colonization and development of retinopathy of prematurity (ROP) requiring treatment.
206 infants with birthweight (BW) ≤1250 g born in a third-level neonatal intensive care unit were prospectively identified and underwent ROP screening. Nasopharyngeal swabs for Uu colonization were taken within the first 3 postnatal days. Those with a positive culture within the first 3 days were recultured on day 12. Culture-positive patients were reevaluated on the twelfth day by nasopharyngeal swabs for Uu. The primary outcome was to define whether there was an association between respiratory tract Uu colonization and severe ROP requiring treatment.
A total of 25 (12.1%) infants developed severe ROP requiring treatment. Multivariate analysis demonstrated that BW (OR: 0.64 (95% CI 0.47–0.88); P=0.006), duration of mechanical ventilation (OR: 1.17 (95% CI 1.06–1.28); P=0.001), premature rupture of membrane >18 h (OR: 3.83 (95% CI 1.2–12.2); P=0.02), and Uu positivity in both the 3 and 12 day cultures (OR: 5.02 (95% CI 1.8–13.9); P=0.002) were independent risk factors for the development of severe ROP requiring treatment. Thus, respiratory tract colonization with Uu was independently associated with severe ROP requiring treatment. The authors speculate why colonization with this organism, which predisposes to respiratory complications and death, and which may release factors that influence retinal vascular development, might contribute to risk of severe ROP.

Importance of Early Postnatal Weight Gain for Normal Retinal Angiogenesis in Very Preterm InfantsA Multicenter Study Analyzing Weight Velocity Deviations for the Prediction of Retinopathy of Prematurity

Carolyn Wu, MD; Chatarina Löfqvist, PhD; Lois E. H. Smith, MD, PhD; Deborah K. VanderVeen, MD; Ann Hellström, MD, PhD; for the WINROP Consortium

Arch Ophthalmol. 2012;130(8):992-999. (f12)
This paper's objective was to assess WINROP (*https://winrop.com*), an algorithm using postnatal weight measurements, as a tool for the prediction of retinopathy of prematurity (ROP) in a large geographically and racially diverse study population. The authors performed WINROP analysis retrospectively on

conventionally at-risk infants from 10 neonatal intensive care units. Weight measurements were entered into WINROP, which signals an alarm for an abnormal weight gain rate. Infants were classified into categories of no alarm (unlikely to develop type 1 ROP) and alarm (at risk for developing type 1 ROP). Use of WINROP requires that an infant has (1) gestational age less than 32 weeks at birth, (2) weekly weight measurements, (3) physiologic weight gain, and (4) absence of other pathologic retinal vascular disease. A total of 1706 infants with a median gestational age of 28 weeks (range, 22-31 weeks) and median birth weight of 1016 g (range, 378-2240 g) were included in the study analysis. An alarm occurred in 1101 infants (64.5%), with a median time from birth to alarm of 3 weeks (range, 0-12 weeks) and from alarm to treatment of 8 weeks (range, 1 day to 22 weeks). The sensitivity of WINROP was 98.6% and the negative predictive value was 99.7%. Two infants with type 1 ROP requiring treatment after 40 weeks' postmenstrual age did not receive an alarm. In summary, the authors concluded that WINROP system is a useful adjunct for ROP screening that identifies high-risk infants early to optimize care and potentially reduce the overall number of diagnostic ROP examinations.

Reactivation of Retinopathy of Prematurity After Bevacizumab Injection

Jennifer Hu, MD; Michael P. Blair, MD; Michael J. Shapiro, MD; Steven J. Lichtenstein, MD; John M. Galasso, MD, PhD; Rashmi Kapur, MD

Arch Ophthalmol. 2012;130(8):1000-1006 (f12)

The purpose of this study was to report late reactivation and progression of retinopathy of prematurity (ROP) after intravitreal bevacizumab monotherapy. The study involved retrospective review of 9 patients (17 eyes) with recurrence of ROP after initial treatment with intravitreal bevacizumab monotherapy. Data collected included (1) location and stage of ROP activity, (2) number and timing of treatments, and (3) structural outcomes. Mean age at treatment-requiring recurrence was 49.3 weeks (SD, 9.1 weeks; minimum, 37 weeks; maximum, 69 weeks) postmenstrual age (PMA). The mean time between initial treatment and treatment-requiring recurrence was 14.4 weeks. Age at retinal detachment ranged from 49 to 69 weeks PMA with a median of 55 weeks PMA and mean of 58.4 weeks PMA. The authors concluded that, although intravitreal bevacizumab treatment is effective in inducing regression of ROP, the effect may be transient. Recurrence can occur later in the course than with conventional laser therapy. Late retinal detachment can occur despite early regression. Long-term favorable structural outcome may require extended observation and retreatment. Laser may be a useful treatment for recurrences.

Current Role of Cryotherapy in Retinopathy of Prematurity

A Report by the American Academy of Ophthalmology
Jennifer L Simpson, MD, Michele Melia, ScM, Michael B Yang, MD, Angela N Buffenn, MD, MPH, Michael F Chiang, MD, Scott R Lamberg, MD
Ophthalmology April, 2012: 119:873-877 (f12)

The objective of this study is to evaluate the role of cryotherapy in the current treatment of retinopathy of prematurity.
Literature searches of Pub-Med and the Cochrane Library were conducted on December 2, 2009, for articles published after 1984. The searches included all languages and retrieved 187 relevant citations. Thirteen articles were deemed relevant to the assessment question and were rated according to the strength of evidence. Four articles reported the results from 2 large multicenter randomized clinical trials and the remaining 9 articles reported results of 3 small randomized trials that directly compared cryotherapy and laser.
Neither of the multicenter randomized clinical trials was a direct comparison of cryotherapy with laser. These studies were used to evaluate the comparative trials based on treatment criteria, study population and clinical results.
There is no clear cut undisputable evidence directly comparing cryotherapy and laser treatment for threshold retinopathy of prematurity. However, “literature suggests that neonatal facilities should gain access to laser technology and laser-trained ophthalmic staff to achieve better outcomes for treatment of the disease.” This is an ophthalmic technology assessment by the American Academy of Ophthalmology, San Francisco, California.

Bevacizumab for Retinopathy of Prematurity-Letter to the Editor

Michael W. Stewart, MD
Ophthalmology May, 2012; 119:1091-1092 (f12)

The authors of the Letter bring to light some of the potential systemic interactions of intravitreal bevacizumab (Avastin) for the treatment of Stage 3 ROP. The letter refers to an earlier article by Mosfhfeghi and Berrocal, “Retinopathy of prematurity in the time of bevacizumab: incorporating the BEAT-ROP results into clinical practice” (Ophthalmology 2011; 118:1227-8)
Some of the points of interest are: 1. In animals, bevacizumab readily transfers to the contralateral eye and can be measured in the serum. 2. The BEAT-ROP trial was underpowered to evaluate the safety of intravitreal bevacizumab in premature infants 3. VEGF contributes to the development of pulmonary vasculature and controls the growth and differentiation of the alveolar epithelium, the cells responsible for the production of pulmonary surfactant. Bevacizumab may inhibit the production of surfactant and contribute to the development of

Respiratory Distress Syndrome (RDS) 4. The pharmacokinetics of bevacizumab is unclear in premature children and may explain why the average time of recurrence of ROP in bevacizumab treated patients is 16 weeks post injection.

Both articles were formulated and written in the United States
COMMENT: It is clear that we do not know everything about bevacizumab and how it affects premature children.

A hybrid form of retinopathy of prematurity

Gaurav Sanghi, Mangat R Dogra, Mohit Dogra, Deeksha Katoch, Amod Gupta
Br J Ophthalmol 2012;96:519-522. (f12)
Aims: To study a hybrid pattern of retinopathy of prematurity (ROP) demonstrating both ridge tissue (simulating staged ROP) and flat neovascularisation (simulating aggressive posterior retinopathy of prematurity (APROP)) in the same eye.

Methods Retrospective chart review from January 2006 to June 2010. We reviewed the retinal drawings and Retcam images for a hybrid pattern of ROP, that is, presence of ridge tissue (characteristic of staged ROP) along with flat neovascular syncytium (characteristic of APROP) in the same eye.

Results: 28 eyes of 18 infants had hybrid characteristics. All eyes had severe plus disease, flat new vessels at the junction of the vascular and avascular retina and ridge tissue at variable locations. Three patterns were noted: I Ridge at the junction of vascular and avascular retina (14 (50%) eyes); II Ridge in the vascularised posterior retina (10 (35.71%) eyes); III Ill-defined ridge close to the optic disc, with mat-like fibrous proliferation into the vitreous (4 (14.29%) eyes). After confluent laser photocoagulation, we observed favourable outcome in 92.3% eyes with pattern I, 100% eyes with pattern II and 25% eyes with pattern III disease.

Conclusion: Some eyes with ROP may have abnormal neovascularisation resembling both APROP and classical staged ROP. It is difficult to characterise these eyes according to the international classification of ROP. However, the presence of plus disease should serve as guide to treatment.
Comments: This is an intresting opservation, but the limitation of the present study, is the non-availability of fluorescein angiography, which could have elucidated the characteristics of the atypical neovascularisation and the underlying retina.

IGF-I and Visfatin Levels in Retinopathy of Prematurity

Farhat Cekmez, MD; Fuat Emre Canpolat, MD, Merih Cetinkaya, MD; Ozgur Pirgon, MD; Secil Aydinoz, MD; O. Melih Ceylan, MD; O. Metin Ipcioglu, MD;

Serdar Umit Sarici, MD J Pediatr Opthalmol Strabismus 2012; 49:120-124 March/April(f12)

Retinopathy of prematurity, first described in 1942, has been studied a great deal. Specifically, looking at ways to predict with a certain amount of accuracy the premature infants who will go on to develop ROP. IGF-I levels have been found to be a good predictive marker for ROP. The authors in this study hypothesized that visfatin levels found in adipose tissue may be linked to ROP. They found that although more studies are needed, visfatin levels can be a predictive factor for ROP. This article describes another way one can predict if a premature infant will develop ROP. If more studies corroborate these results, it would be another exciting discovery in a disease that many years ago caused infants to have devastating visual outcomes.

Effect of Resveratrol on Bcl-2 and VEGF Expression in Oxygen-Induced Retinopathy of Prematurity

Wenlin Li, MD; Deyong Jiang, MD J Pediatr Opthalmol Strabismus 2012; 49:230-235 July/August (f12)

The study was conducted to explore the effect of resveratrol on B-cell leukemia/lymphoma-2(Bcl-2) and vascular endothelial growth factor (VEGF) using rats with retinopathy of prematurity (ROP). By inducing ROP in rats and looking at the effects of no resveratrol and various doses of resveratrol, the study concluded that resveratrol can significantly inhibit expression of Bcl-2 and VEGF in the retina of rats with oxygen induced ROP.

Preeclampsia and Retinopathy of Prematurity in Preterm Births.

Yu XD, Branch DW, Karumanchi SA, Zhang JPediatrics 2012; 130(1):101-7. (f12)

This retrospective cohort study of 25,473 preterm neonates examined the influence of maternal gestational hypertension and preeclampsia on the occurrence of ROP in preterm infants. The authors examined the association between gestational hypertension, preeclampsia, and ROP while controlling for potential confounders by multiple logistic regression analysis. Of the 8758 early preterm infants (gestational age <34 weeks), 1024 (11.69%) had ROP, while of the 16 715 late preterm infants, only 29 (0.17%) had ROP. After adjusting for confounders, preeclampsia was associated with a significantly reduced risk of ROP (adjusted odds ratio [aOR], 0.65; 95% confidence interval [CI], 0.49–0.86 for early preterm birth; aOR, 0.10; 95% CI, 0.01–0.93 for late preterm birth; aOR, 0.66; 95% CI, 0.50–0.87 for all preterm births). Gestational hypertension was not significantly associated with ROP at early or late preterm births. The authors concluded that preeclampsia, but not gestational hypertension, was associated with a reduced risk of ROP in preterm births.

PLUS DISEASE IN RETINOPATHY OF PREMATURITY: Diagnostic Impact of Field of View

Rao, Rohini BA[*]; Jonsson, Nina J. MD[*]; Ventura, Camila MD[*,¶]; Gelman, Rony MD[*]; Lindquist, Martin A. PhD[†]; Casper, Daniel S. MD, PhD[*]; Chiang, Michael F. MD

Retina. June 2012; 32(6):1148-1155

Retinopathy of prematurity (ROP) is a leading cause of childhood blindness worldwide. According to the Cryotherapy for Retinopathy of Prematurity (CRYO-ROP) and Early Treatment for Retinopathy of Prematurity (ETROP) trials, treatment of severe disease with laser photocoagulation or cryotherapy can improve structural and visual outcomes.

A key component of the international classification system for ROP is “plus disease,” which is defined as arterial tortuosity and venous dilation in the posterior pole greater than or equal to that of a standard published photograph.

More recently, the revised international ROP classification system defined a new “preplus” category. Presence of plus disease has been shown by the CRYO-ROP and ETROP studies to be the most critical factor in determining treatment for ROP. Therefore, it is essential for plus disease to be diagnosed accurately and consistently. However, plus disease diagnosis has been found to be inconsistent, even among experts. The standard published photograph shows numerous vessels of varying degrees of dilation and tortuosity, leaving the true diagnostic cutoff unclear. Furthermore, the standard photograph has a narrower field of view and larger magnification than clinical diagnostic methods, such as indirect ophthalmoscopy and wide-angle retinal imaging.

The purpose of this study was to examine the impact of retinal field of view and magnification on interexpert reliability of plus disease diagnosis in ROP. This is the first study to investigate plus disease diagnosis as a function of field of view. The key findings are that 1) agreement in plus disease diagnosis from wide-angle images is higher than from medium-angle and narrow-angle images, suggesting that peripheral retinal findings contribute diagnostic information and 2) interexpert agreement in plus disease diagnosis is imperfect.

Aldose Reductase Deficiency Reduced Vascular Changes in Neonatal Mouse Retina in Oxygen-Induced Retinopathy

Zhong Jie Fu, Suk-Yee Li, Norbert Kociok, David Wong, Sookja K. Chung, and Amy C. Y. Lo. IOVS August 2012 53:5698-5712; published ahead of print July 26, 2012, doi:10.1167/iovs.12-10122. (f12)

Retinal neovascularization is the major pathologic process in many ocular diseases and is associated with oxidative stress. Deficiency of aldose reductase

(AR), the first enzyme in the polyol pathway for glucose metabolism, has been shown to reduce oxidative stress and blood vessel leakage. The present study aimed to investigate the effect of AR deficiency on retinal neovascularization in a murine oxygen-induced retinopathy (OIR) model. Seven-day-old wild-type (WT) and AR-deficient ($AR^{-/-}$) mice were exposed to 75% oxygen for 5 days and then returned to room air. Vascular obliteration, neovascularization, and blood vessel leakage were analyzed and compared, as were immuno-histochemical markers for angiogenesis. Compared with WT OIR retinae, $AR^{-/-}$ OIR retinae displayed significantly smaller central retinal vaso-obliterated area, less neovascularization, and reduced blood vessel leakage. Significantly reduced oxidative stress and glial responses were also observed in $AR^{-/-}$ OIR retinae. Moreover, reduced microglial response in the avascular area but increased microglial responses in the neovascular area were found with AR deficiency. Furthermore, expression levels of VEGF, p-Erk, p-Akt, and p-IκB were significantly reduced in $AR^{-/-}$ OIR retinae. The study demonstrated that AR deficiency reduced retinal vascular changes in the mouse OIR model, indicating that AR can be a potential therapeutic target in ischemia-induced retinopathy.

[Home]

PREMATURITY

Characteristics of peripapillary retinal nerve fiber layer in preterm children

Wang J, Spencer R, Leffler JN, Birch EE. Am J Ophthalmol. 2012;153(5):850-855 (f12)

This prospective cross-sectional study examined quantitatively characteristics of the peripapillary retinal nerve fiber layer (RNFL) in preterm children using Fourier-domain optical coherence tomography (FD-OCT). A 3-mm high-resolution FD-OCT peripapillary RNFL circular scan centered on the optic disc was obtained from right eyes of 25 preterm children (10.6 ± 3.7 years old, 8 preterm and 17 with regressed retinopathy of prematurity with normal-appearing posterior poles) and 54 full-term controls (9.8 ± 3.2 years old). Images were analyzed using Spectralis FD-OCT software to obtain average thickness measurements for 6 sectors (temporal superior, temporal, temporal inferior, nasal inferior, nasal, nasal superior), and the global average. The RNFL global average for preterm children was 8% thinner than for full-term controls. In the preterm group, peripapillary RNFL thickness on the temporal side of the disc was 6% thicker than in full-term controls, while all other peripapillary RNFL sectors were 9% to 13% thinner. In the preterm group, temporal sector peripapillary RNFL thickness was correlated with gestational age ($r = -0.47$, $P < .001$), with foveal center total

thickness (r = 0.48, P = .008, 1-tailed), and with visual acuity (r = 0.42; P = .026, 1-tailed). The authors concluded that the significantly thinner RNFL global average for preterm children suggests that prematurity is associated with subclinical optic nerve hypoplasia. Significant correlations between temporal sector RNFL thickness and both the foveal thickness and visual acuity suggest that the peripapillary RNFL is related to abnormalities in macular development as a result of preterm birth.

An Assessment of Ocular Morbidities of Children Born Prematurely in Early Childhood

Altan Goktas, MD; Emin Cumhur Sener, MD; Ali Sefik Sanac, MD J Pediatr Opthalmol Strabismus 2012; 49:236-241 July/August (f12)
This article addresses the various ocular morbidities that may occur in children born prematurely. One hundred and seventeen children with a history of gestational age less than 37 weeks at birth were included in the study. Each child underwent ophthalmologic examination including visual acuity, anterior and posterior examination, refraction, strabismus evaluation and ocular biometry. The study confirms that children born prematurely are at risk for decrease visual acuity, myopia and strabismus.

[Home]

STRABISMUS

Incidence of strabismus and amblyopia in preverbal children previously diagnosed with pseudoesotropia. Silbert AL, Matta NS, and Silbert D. J AAPOS 2012;16:118-119. (f12)

The charts of all children diagnosed with pseudoesotropia over a 10-year period by one Pediatric Ophthalmologist were reviewed. Of the 306 charts which met inclusion criteria, 201 (66%) returned for follow-up. Twenty children (10%) were later found to have strabismus. Another five children were later found to have significant refractive error. The prevalence of strabismus found in this study is higher than that of the general population. This study cannot separate children who acquired strabismus after their initial pseudoesotropia diagnosis versus those who had strabismus at their initial encounter and were missed. Therefore the true percentage of ‘missed diagnosis’ cannot be adequately determined.

Validity of angle of deviation measurements in children with intermittent exotropia. Hatt SR, Leske DA, Liebermann L, et al. J AAPOS 2012;16:120-124 (f12)

The authors attempt to provide thresholds for assessing clinically significant long-term change in angle of intermittent exotropia by analyzing variability associated

with test-retest differences and short-term variability. Twenty-six children were recruited. There was greater variability with larger versus smaller deviations at near but not at distance. Clinically significant change at distance was 4 prism diopters (PD) if the angle of deviation was <=20 PD and 8 PD if the angle of deviation was >20 PD. At near significant change was found to be at least 7 PD and 13 PD, respectively. The authors feel these numbers may be used as a guide to determine clinically significant change. No comment can be made on data from specific age brackets as too few subjects were recruited. Also not everyone would agree that the size of the deviation is the key factor in evaluating the severity of intermittent exotropia. Other factors, such as level of control and stereoacuity are often used as guides to the patients status.

Development of refractive accommodative esotropia in children initially diagnosed with pseudoesotropia.

Mohan K and Sharma A. J AAPOS 2012;16:266-268. (f12)

The authors sought to determine the clinical characteristics of children with pseudoesotropia who later develop a refractive accommodative esotropia. They retrospectively reviewed the records of all consecutive patients diagnosed with pseudoesotropia <=age 3 years, over an 8-year period. A total of 51 children met inclusion criteria. 8 of the 51 children developed refractive accommodative esotropia during follow-up. 1/38 (2.6%) of those with hypermetropia <=1.50 D developed refractive accommodative esotropia compared to 7/13 (54%) who had hypermetropia >1.5 D. Refractive accommodative esotropia developed at an average age of 2.78 years and had a mean hypertropia of 4.28 D. The authors recommend continued follow-up of children with pseudoesotropia who have a hyperopia >1.5 D.

Identifying masked superior oblique involvement in thyroid eye disease to avoid postoperative A-pattern exotropia and intorsion.

Holmes JM, Hatt SR, and Bradley EA. J AAPOS 2012;16:280-285. (f12)

The authors challenge the assumption that patients who receive inferior rectus recession(s) in the context of thyroid eye disease develop A-pattern exotropia because of the inferior rectus weakening. They argue that A-pattern exotropia and intorsion develop due to masked superior oblique muscle tightness in some cases. They present thyroid disease patients who underwent inferior rectus recession and review the results of preoperative and intraoperative testing to detect masked superior oblique muscle tightness. The authors contend that identification of the masked tight superior oblique muscle and addressing this at the time of surgery prevented consecutive intorsion. They discuss the clues for

this condition- minimal preoperative extorsion or frank preoperative intorsion, and superior oblique muscle tightness determined intraoperatively with exaggerated traction testing before and after disinsertion of tight rectus muscles., The authors present 4 cases. Two of the cases already had orbital decompressions and strabismus surgery prior to evaluation of the superior obliques. One of these patients did not have superior oblique tightness and is presented as a contrasting management case. The authors hypothesis would be better supported with more 'clean' cases and with preoperative imaging of the superior oblique muscles.

Intentional extreme anisometropic pseudophakic monovision: New approach to the cataract patient with longstanding diplopia.

Osher RH, Golnik KC, Barrett G, Shimizu K.

J Cataract Refract Surg. Aug 2012;38:1346-51. (f12)

The authors present a case series of twelve patients ages 62 and older with longstanding and stable strabismus who were diplopic. They underwent cataract surgery with intentional targeting of one eye for plano and the other eye for -3.00 to create severe anisometropia. This strategy was successful in nine of twelve patients, eliminating their diplopia. In the other three, diplopia was improved but not eliminated, and these were patients who ended up with a refraction of -2.00 to -3.00 in the eye that was targeted for myopia.
The patients did not receive a preoperative evaluation from an adult strabismus surgeon, and were not counseled regarding various strategies (including strabismus surgery) that the strabismologist could employ to relieve their diplopia and restore fusion. The strategy of intentional extreme monovision, however, may prove useful for patients with intractable diplopia

Accommodative Esotropia-General Correspondence-Letter to the Editor

Sumit Monga, MS, Ramesh Kekunnya, MD
Ophthalmology March, 2012; 119:655-656 (f12)

The authors of the letter refer to an article entitled "The long term follow up of accommodative esotropia in a population based cohort of children" by Mohney et al (Ophthalmology 2011: 118:581-85).
The authors of the Letter bring up the fact that: 1) According to the literature, (Haynes et al, Science: 1965: 148:528-30), accommodation, in an infant, matures by the age of 3-4
months. 2) There are no reports of accommodative esotropia at birth 3) According to the PEDIG- Congenital Esotropia Observational Study (CEOS),

27% of all esotropias referred before 20 weeks (5 months of age) resolve, especially if the esotropia is intermittent at 10 weeks of age. The authors suggest that some of the infants enrolled in the study cohort were truly esophoric and not esotropic, as reported in the paper.
Mohney et al replied: 1) All the children in the study had an esotropia of at least 10 prism diopters 2) The ten prism diopter esodeviation was not always present on the initial exam but did develop an esotropia greater than 10 prism diopters later in the study 3) Family reporting of the age of onset was used as used as the entry exam data. If the parents stated that the eyes were crossed “from birth” this was recorded in the study as “1” day of age. (for the “date of onset” of the deviation)
The authors of the Letter were from Hyderabad and Visakhapatnum, India. Mohney et al are from the Mayo Clinic in Rochester, Minnesota.
COMMENT: Haynes article in Science deserves scrutiny and corroboration. Most infants in my experience don’t have identifiable accommodation until around one year of age, premature children several months later.

Head Posture in Down Syndrome-Letter to the Editor, General Correspondence

Gregory T. Lueder, MD, Brian Arthur, MD, Sylvia R. Codsi, MD, Burton Kushner, MD, Richard Saunders,MD
Ophthalmology May, 2012; 119:1089-1090 (f12)
This Letter to the Editor brings to light Dr Lueder et all’s 2004 article “Head tilt – dependent esotropia associated with trisomy 21”. (Ophthalmology, 2004, 111:596-9). In this article the authors point out that in6/7 of their trisomy 21 patients, their head tilt was related to a V pattern esotropia, which they felt was an example of “comitant” esotropia. The head tilt was effectively treated with horizontal muscle surgery.
In the more recent article, (Ophthalmology**,** 2011:118:1859-64) by Dumitrescu et al, the authors reported that the most common cause of head tilt in Down syndrome patients was “incomitant strabismus” which, according to their criteria, would include V pattern esotropia or any other type of pattern strabismus.
The bottom line of these two articles is 1. Look for pattern or non -pattern strabismus as a cause of head tilt, especially V pattern esotropia 2. Look carefully for incomitant strabismus in these children who are naturally inclined to have atlanto-axial dysplasia and resistance to putting their head in unusual positions.
Both of the above cited articles were produced in the United States

Decreased accommodation during decompensation of distance exotropia

Anna M Horwood, Patricia M Riddell Br J Ophthalmol 2012;96:508-513. (f12)

Objective: Disparity cues can be a major drive to accommodation via the convergence accommodation to convergence (CA/C) linkage, but, on decompensation of exotropia, disparity cues are extinguished by suppression so this drive is lost. This study investigated accommodation and vergence responses to disparity, blur and proximal cues in a group of distance exotropes aged between 4 and 11 years both during decompensation and when exotropic.

Methods: 19 participants with distance exotropia were tested using a PlusoptiXSO4 photo refractor set in a remote haploscopic device that assessed simultaneous vergence and accommodation to a range of targets incorporating different combinations of blur, disparity and proximal cues at four fixation distances between 2 m and 33 cm. Responses on decompensation were compared with those from the same children when their deviation was controlled.

Results:Manifest exotropia was more common in the more impoverished cue conditions. When decompensated for near, mean accommodation gain for the all-cue (naturalistic) target was significantly reduced ($p<0.0001$), with resultant mean under-accommodation of 2.33 D at 33 cm. The profile of near cues usage changed after decompensation, with blur and proximity driving residual responses, but these remaining cues did not compensate for loss of accommodation caused by the removal of disparity.

Conclusions:Accommodation often reduces ondecompensation of distance exotropia as the drive from convergence is extinguished, providing a further reason to try to prevent decompensation for near.

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 Ophthalmol 2012;96:679-682. (f12)

Background/aims: To compare the insertion locations of extraocular muscles between Taiwanese (Han Chinese) and Western populations and to determine whether anatomical differences warrant different surgical guidelines.

Methods: Insertion locations were compared between a Taiwanese population of subjects who had received surgical treatment for strabismus and a control group who had not. Insertion locations and surgical outcomes in the strabismus group were also compared with those reported in other countries.

Results: In Taiwanese subjects, extraocular muscle insertion locations were not significantly different between strabismus subjects and controls. However, the distances
from the insertion location to the limbus of the inferior rectus, lateral rectus and superior rectus were significantly shorter in the Taiwanese subjects than in Western populations.

Conclusion: Extraocular muscle insertion locations for the Taiwanese population in this study significantly differed from those reported in studies of Western populations.
Therefore, surgical guidelines for performing lateral rectus recession to treat exotropia in Western populations may be inappropriate for Taiwanese and other Asian populations.

Comments: Since the data in this retrospective study were only compared with those reported by Apt, prospective studies are needed for further confirmation.

Localising rectus muscle insertions using high frequency wide-field ultrasound biomicroscopy Hayat Ahmad Khan, David R Smith, Stephen P Kraft

Br J Ophthalmol 2012;96:683-687. (f12)
The authors compared the accuracy of the Sonomed UBM(SUBM), a new 'wide-field ultrasound biomicroscope', with the older model Humphrey UBM (HUBM) in localising EOM insertions and compared their ranges of detection of muscle insertions.

Methods: Prospective, double-masked, observational study of 27 patients undergoing primary (n.40 muscles) or repeat (n.10 muscles) horizontal or vertical rectus
muscle surgery. EOM insertional distances were measured with SUBM, and then intraoperatively with callipers.

Results: For all muscles, the differences between SUBM and surgery measurements were less than 1.0 mm. The mean of the SUBM insertion distances was 6.67 mm (SD 1.65 mm) versus 6.7 mm (SD 1.6 mm) at surgery. The intraclass correlation coefficient showed 'excellent' correlation between the two sets of data and was higher
than that reported with HUBM.
Conclusion: The SUBM with its smaller, more manoeuvrable probe handpiece and a wider scanning field was more accurate in detecting muscle insertions compared with HUBM.

Duane Retraction Syndrome: Series of 441 Cases

Ramesh Kekunnaya, FRCS; Amit Gupta, MS; Virender Sachdeva, MS; Sannapaneni, MPH; B. Venkateshwar Rao, MD; Urvish Vashist, MS; Debajit Ray, MS J Pediatr Opthalmol Strabismus 2012; 49:164-169 May/June (f12)

A series of 441 cases is presented that highlights the demographic and clinical profile of various subtypes of Duanes retraction syndrome. The article discusses several differences between type I, II and III as well as unilateral versus bilateral disease. It confirms several factors that are well known in Duanes retraction syndrome and serves as a good review of the disease.

Effectiveness of Prisms in the Management of Diplopia in Patients Due to Diverse Etiologies

Madhura A. Tamhankar, MD; Gui-shang Ying, PhD; Nicholas J. Volpe J Pediatr Opthalmol Strabismus 2012; 49:222-228 July/August (f12)
In a retrospective cohort study, this article discusses the use of prisms in the management of diplopia over a broad range of ocular misalignments and various etiologies. Most of the patients (88%) reported satisfactory resolution of their diplopia with skew deviation and divergence insufficiency showing the highest rate of satisfaction. Prisms were found to be effective with larger ocular misalignments than traditionally thought.

Vertical Heterophoria and Susceptibility to Visually Induced Motion Sickness

Danielle N. Jackson, Harold E.Bedell Strabismus Mar 2012, Vol. 20, No. 1: 17–23 (f12)

The article discusses the relationship between vertical phoria and motion sickness. They tested 43 subjects with vertical phoria and placed them in a situation to induce motion sickness. They found a positive correlation between patients with vertical phorias and motion sickness, more so if the vertical phoria was greater than 0.75 prism diopters. They also attempted to increase and decrease the phorias to see if this would change the amount of motion sickness the subjects experienced, the results showed minimal correlation in this aspect. They suggested that there is a relationship although this relationship may be casual.

Delayed-onset bilateral abducens paresis after head trauma

Salunke P, Savardekar A, Sura S. Indian J Ophthalmol 2012;60:149-50 (Mar-Apr) (f12)

The authors present two cases. The first patient had onset of bilateral abducens paresis 2 weeks after closed head injury and the second patient after 3 days. The cause in the former was detected to be chronic subdural hematoma and in the latter is speculated to be edema/ischemia due to injury to soft tissue structures housing these nerves. The authors propose that another mechanism of delayed onset of abducens nerve palsy may be due to vasospasm or nerve ischemia due to injury to branches of meningodorsal artery. The authors conclude that though rare, delayed abducens palsy needs to be evaluated and may have a treatable cause like elevated intracranial pressure.

Monovision Correction for Small-Angle Diplopia Bujak MC, Leung AK, Kisilevsky M, Margolin E. Am J Ophthalmol 2012; 154(3):586-92. (f12)

This prospective, interventional case series was designed to assess quantitatively the efficacy of monovision correction in the treatment of acquired small-angle binocular diplopia in adult patients. Twenty patients with symptomatic diplopia were enrolled in a prospective treatment trial at a tertiary university neuro-ophthalmology practice. All had stable deviations of 10 prism diopters or less for more than 3 months. Each received monovision spectacles, contact lenses, or both with distance correction in the dominant eye. Half received a +3.00-diopter add and the others received +2.50 diopters. The validated and standardized Diplopia Questionnaire and Amblyopia and Strabismus Questionnaire were used to quantify the efficacy of monovision correction for diplopia by measuring the functional impact on vision-specific quality of life. Based on the results of the Diplopia Questionnaire, 85% of patients experienced significant improvement in diplopia symptoms after monovision correction. There was a statistically significant 58.6% improvement in the Diplopia Questionnaire score in the patients ($P < .0001$). The Amblyopia and Strabismus Questionnaire scores demonstrated improved quality of life and daily function after monovision correction ($P = .03$), especially in the areas of double vision($P = .0003$) and social contact and appearance ($P = .0002$). The authors concluded that monovision decreased the frequency of diplopia and improved subjects' quality of life. They asserted that monovision may be a feasible alternative for presbyopic diplopic patients who are dissatisfied with other conservative treatment options.

Assessing Divergence in Children With Intermittent Exotropia
Laura Liebermann, CO, Sarah R. Hatt, DBO, David A. Leske, MS, et al *Strabismus* March 2012; 20(1), 11-16 (f12)

This study comes from the Department of Ophthalmology, Mayo Clinic, Rochester, Minnesota.

This is a retrospective study that studied 32 children with intermittent exotropia ages 4-13 years in 38 visually normal non-strabismic children. It was designed to report fusional divergence in children with intermittent exotropia.

Fusional divergence break points were measured using a prism bar, staring from a naturally fused state. Distribution of divergence break points was evaluated. Subnormal fusional divergence was defined as below the fifth percentile in visually normal children. Fusional divergence break points for intermittent exotropia were normal at near but bimodal at distance. There is a moderate correlation between divergence and convergence break points at near but only a weak correlation between divergence break points and control groups and divergence break points and the angle of deviation (both were measured at near).

The conclusion of the study is that most children with intermittent exotropia have normal near fusional divergence but nearly half have reduced distance fusional divergence. The study found that the magnitude of near divergence break points correlates with the magnitude of near convergence break point. The main conclusion of the study was that measuring divergence might provide useful information about the “fragility” of fusion status in patients with intermittent exotropia.

Post-vitreoretinal Surgery Strabismus---A Review

N.L. Chaudhry and J.M. Durnian
Strabismus March 2012; 20(1): 26-30 (f12)

This article comes from the St. Paul’s Eye Unit, Royal Liverpool University Hospital, Liverpool, United Kingdom. This is a comprehensive article reviewing the pathogenesis and management of complicated strabismus patients who underwent previous retinal surgery.

The review article includes patients that have undergone vitrectomy procedures, scleral buckling procedures, a combination of intraocular and extraocular procedures. According to the authors, many factors play a role in the development of strabismus including mechanical extraocular muscle changes, local anesthetic administration, and sensory alteration.

The authors reviewed treatment options including occlusion, prisms, botulinum toxin, and surgery. The authors emphasized the importance of prevention of strabismus with the original retinal procedure.

The authors have the following recommendations to prevent strabismus after extraocular muscle surgery: 1) Meticulous dissection during surgery to prevent violation of the orbital fat pad, 2) use of localized tissue-friendly retinal explants, 3) avoiding extraocular muscle manipulation as much as possible, 4) special care when operating around the oblique muscles, and 5) use of sub-tenon anesthetic.

The authors also stress the importance of preparing patients for the possibility of postoperative strabismus and double vision associated with retinal reattachment surgery.

The Role of Cortical Alterations in Infantile Strabismus

Michael C. Brodsky, MD
Strabismus March 2012; 20(1): 35-36 (f12)

This article was written by the author in response to a letter by Dr. Gallegos-Duarte regarding the origin of dissociated vertical deviation. Dr. Brodsky published a paper “Dissociated Vertical Divergence: Cortical or Sub-Cortical in Origin?”

Dr. Gallegos-Duarte published an article “Participation and Neuro-Modulation of the Extrastriate Cortex in Strabismus” in the *Arch Chil Ophthal* 2006; 63:199-209. (Dr. Gallegos-Duarte also has additional publications on the subject in 2006, 2010, and 2 publications in 2007.

Dr. Brodsky, in this particular letter affirms his view that infantile esotropia and other dissociated deviations of eyes from resurgence of subcortical visual reflexes. The cortical visual system injury may potentiate their expression but is not necessary to unlock the physiologic process.

Dr. Brodsky agrees with Dr. Gallegos-Duarte in an “unbalanced input is commonly corrected by cerebral cortex, but when the cortical integrator fails it is feasible that some relatively reflexes may get out of control”. Dr. Brodsky believes that “cortical expression” provides the permissive cause for dissociated deviations even those these conditions are generated by subcortical visual reflexes.

Differences in Gene Expression between Strabismic and Normal Human Extraocular Muscles

Amy L. Altick, Cheng-Yuan Feng, Karen Schlauch, L. Alan Johnson, and Christopher S. von Bartheld. IOVS August 2012 53:5168-5177; published ahead of print July 10, 2012, doi:10.1167/iovs.12-9785. (f12)

Strabismic extraocular muscles (EOMs) differ from normal EOMs in structural and functional properties, but the gene expression profile of these two types of EOM has not been examined. EOM samples were obtained during corrective surgery from patients with horizontal strabismus and from deceased organ donors with normal EOMs. Microarrays and quantitative PCR identified significantly up- and down-regulated genes in EOM samples. Analysis was

performed on probe sets with more than 3-fold differential expression between normal and strabismic samples, with an adjusted *P* value of ≤ 0.05. Strabismic human extraocular muscles show significant changes in gene expression with up-regulation of genes related to extracellular matrix and down-regulation of myosin genes. Expression of genes associated with signaling, calcium handling, mitochondria function and biogenesis, and energy homeostasis also was significantly different between normal and strabismic EOM. Skeletal muscle PCR array identified 22 (25%) of 87 muscle-specific genes that were significantly down-regulated in strabismic EOMs; none was significantly up-regulated. The strabismic transcriptome is a major contribution of the muscle to strabismus, as well as new therapeutic targets. Gene regulation of proteins fundamental to contractile mechanics and extracellular matrix structure appears to be involved in pathogenesis and/or consequences of strabismus, suggesting potential novel therapeutic targets.

Evaluation of the Adult Strabismus-20 (AS-20) Questionnaire Using Rasch Analysis

David A. Leske, Sarah R. Hatt, Laura Liebermann and Jonathan M. Holmes. Invest Ophthalmol Vis Sci. 2012 May 4;53(6):2630-9. (f12)

The Adult Strabismus-20 (AS-20) questionnaire is a 20-item patient-derived strabismus-specific instrument designed to evaluate health-related quality of life (HRQOL) in adults with strabismus. The purpose of this study was to refine the Adult Strabismus 20 (AS-20) health-related quality of life (HRQOL) questionnaire using Rasch analysis. Rasch analysis was performed independently on the original AS-20 using the following steps: dimensionality, response ordering, local dependence, infit and outfit analyses, differential item functioning, subject targeting, and confirmatory dimensionality. Two subscales were present in each of the original AS-20 subscales, for a total of 4 subscales, which were labeled “self-perception,” “interaction,” “reading function,” and “general function.” Response ordering was appropriate for 3 of the subscales but required reduction to 4 response options for the fourth subscale. No notable local dependence was found for any subscale. As a result of fit analysis, 2 items were removed, 1 each from 2 subscales. No significant differential item functioning was seen for sex or age. The resulting 5-item self-perception subscale and 4-item reading function subscale are reliable and target the adult strabismus patient cohort appropriately. The resulting 5-item interaction subscale and 4-item general function subscale have less than optimal reliability.

The AS-20 benefits from reduction to 4 subscales (self-perception, interaction, reading function, and general function) and reducing the response options in the general function subscale from 5 to 4 options. The refined AS-20 may prove to be even more responsive to HRQOL changes in adult strabismus following treatment or changes over time.

Eye Position Stability in Amblyopia and in Normal Binocular Vision.

Esther G. González, Agnes M. F. Wong, Ewa Niechwiej-Szwedo, Luminita Tarita-Nistor and Martin J. Steinbach. IOVS August 2012 53:5386-5394; published ahead of print July 12, 2012, doi:10.1167/iovs.12-9941 (f12)

The authors investigated whether the sensory impairments of amblyopia are associated with a decrease in eye position stability (PS). The positions of both eyes were recorded simultaneously in three viewing conditions: binocular, monocular fellow eye viewing (right eye for controls), and monocular amblyopic eye viewing (left eye for controls). For monocular conditions, movements of the covered eye were also recorded (open-loop testing). Bivariate contour ellipses (BCEAs), representing the region over which eye positions were found 68.2% of the time, were calculated and normalized by log transformation. For controls, there were no differences between eyes. Binocular PS ($\log_{10}$BCEA = −0.88) was better than monocular PS ($\log_{10}$BCEA = −0.59) indicating binocular summation, and the PS of the viewing eye was better than that of the covered eye ($\log_{10}$BCEA = −0.33). For patients, the amblyopic eye exhibited a significant decrease in PS during amblyopic eye ($\log_{10}$BCEA = −0.20), fellow eye ($\log_{10}$BCEA = 0.0004), and binocular ($\log_{10}$BCEA = −0.44) viewing. The PS of the fellow eye depended on viewing condition: it was comparable to controls during binocular ($\log_{10}$BCEA = −0.77) and fellow eye viewing ($\log_{10}$BCEA = −0.52), but it decreased during amblyopic eye viewing ($\log_{10}$BCEA = 0.08). Patients exhibited binocular summation during fellow eye viewing, but not during amblyopic eye viewing. Decrease in PS in patients was mainly due to slow eye drifts.

Deficits in spatiotemporal vision in amblyopia are associated with poor PS. PS of amblyopic and fellow eyes is differentially affected depending on viewing condition. There is decreased fixation stability in the amblyopic eye during binocular and monocular viewing. The fellow eye's fixation stability is comparable to controls during binocular and monocular viewing, but comparable to amblyopic eye in open-loop testing. Only the fellow eye exhibits binocular summation.

Differential Lateral Rectus Compartmental Contraction during Ocular Counter-Rolling.

Robert A. Clark and Joseph L. Demer.. Invest Ophthalmol Vis Sci. 2012 May 14;53(6):2887-96. (f12)

Since the MRI can provide a functional indication of EOM contractility through changes in EOM cross section and volume, the authors sought evidence of selective compartmental activation of the rectus EOMs during head tilt in normal subjects. The lateral rectus (LR) and medial rectus (MR) extraocular muscles (EOMs) have largely non-overlapping superior and inferior innervation territories.

They used magnetic resonance imaging (MRI) in humans to investigate differential compartmental activity in the rectus EOMs during head tilt, which evokes ocular counter-rolling, a torsional vestibulo-ocular reflex (VOR). MRI in quasi-coronal planes was analyzed during target-controlled central gaze in 90°

right and left head tilts in 12 normal adults. Cross sections and posterior partial volumes of the transverse portions of the four rectus EOMs were compared in contiguous image planes 2 mm thick spanning the orbit from origins to globe equator, and used as indicators of contractility.

The horizontal rectus EOMs had significantly greater posterior volumes and maximum cross sections in their inferior compartments ($P < 10^{-8}$). In orbit tilt up (extorted) compared with orbit tilt down (intorted) head tilts, contractile changes in LR maximum cross section ($P < 0.0001$) and posterior partial volume ($P < 0.05$) were significantly greater in the inferior but not in the superior compartment. These changes were not explainable by horizontal or vertical eye position changes. The vertical rectus EOMs did not exhibit significant compartmental contractile changes during head tilt.

Mechanical modeling suggests that differential LR contraction may contribute to physiological cyclovertical effects. Selective activation of the two LR, and possibly MR, compartments correlates with newly recognized segregation of intramuscular innervation into distinct compartments, and probably contributes to non-commutative torsion during the VOR. The present paper also considers the possibility that horizontal rectus EOMs may be capable of generating cyclovertical movements, including sensory torsion, which would open the possibility that horizontal rectus EOMs might also be capable of generating non-commutative torsion.

Effect of sequential injections of hepatocyte growth factor and insulin-like growth factor-I on adult rabbit extraocular muscle.

Willoughby CL, Ralles S, Christiansen SP, and McLoon LK. J AAPOS 2012;16:354-360. (f12)

Growth factors can be used to strengthen extraocular muscles, unlike Botulinum toxin which weakens them. Insulin-like growth factor (IGF) has been shown to increase muscle force and specific tension when injected directly into rabbit extraocular muscles. There is some concern that IGF-I receptor in orbital fibroblasts may be linked to fat expansion in thyroid eye disease. Thus the authors tested a strategy to lower the dosage of IGF-1 needed while maintaining the same effect on muscle force augmentation. Hepatocyte growth factor (HGF) was used as a preinjection before IGF-I to see if it would augment the effect of IGF-I .

Single HGF injection alone did not increase force generation 1 week after treatment. However, sequential HGF injection followed 1 week later by IGF-I did significantly increase force and specific tension. This effect was stronger than a single IGF-I injection alone, regardless of dose. The sequential treatment also increased mean myofiber cross-sectional areas significantly. This treatment

required further study as it may eventually offer another treatment option for strabismus patients.

[Home]

STRABISMUS SURGERY

The effect of general anesthesia and strabismus surgery on the intellectual abilities of children: a pilot study.

Yang HK, Chungh DS, Hwang JM Am J Ophthalmol. 2012;153(4):609-13. (f12)

This prospective observational study sought to determine the influence of general anesthesia and strabismus surgery on children's intellectual abilities. Children 5 to 10 years of age receiving general anesthesia with sevoflurane and undergoing strabismus surgery were included. Intellectual abilities were examined before and 4 weeks after surgery using the Kaufman Assessment Battery for Children. Four subtests representing intellectual abilities related to complex cortical function were examined: identification of objects in a partially completed picture, reproduction of a presented design by using rubber triangles, selecting a picture that completes or is similar to another picture, and memory for location of pictures presented on a page. The study group consisted of 21 children who underwent strabismus surgery under general anesthesia (mean duration, 51.3 minutes). The mean preoperative total score of the 4 subtests was 49.4±6.2. The mean postoperative total score adjusted for potential learning effects and test-retest reliability was 48.1±7.7. There was no significant postoperative change in the total score (P=.108). However, the triangle test score decreased significantly after operation (P=.019), particularly in patients with decreased stereoacuity after surgery. The authors concluded that general anesthesia with sevoflurane and strabismus surgery generally do not affect the intellectual abilities of complex cortical function in children 5 to 10 years of age at 4 weeks after surgery. Cortical functions related to hand-eye coordination may be affected by transient changes in postoperative stereoacuity.

Changes in health-related quality of life 1 year following strabismus surgery

Hatt SR, Leske DA, Liebermann L, Holmes JM. Am J Ophthalmol. 2012;153(4):614-9. (f12)

This retrospective cohort study reported changes in health-related quality of life (HRQOL) 1 year following strabismus surgery. Seventy-three adults undergoing strabismus surgery in a single clinical practice completed the Adult

Strabismus-20 (AS-20) HRQOL questionnaire preoperatively and at 6 weeks and 1 year postoperatively. All included adults were successfully aligned at 6 weeks postoperatively. Success was defined based on clinical criteria. Change in AS-20 psychosocial and function scores was evaluated as: 1) difference in scores between 6 weeks and 1 year postoperatively and 2) proportions exceeding previously published limits of agreement at 6 weeks and 1 year. For patients successfully aligned at both 6 weeks and 1 year (n=51), further improvement in median scores was seen from 6 weeks to 1 year for both the psychosocial scale (83.8 vs 93.8; P<.0001) and the function scale (72.5 vs 77.5; P=.007). Also, a greater proportion exceeded limits of agreement at 1 year than at 6 weeks (psychosocial: 48% vs 30%; P=.007, function: 67% vs 51%; P=.01). For patients who became partial success (n=18) or failure (n=4) at 1 year there was a numerical decrease in scores at 1 year. The authors concluded that adult strabismus patients who remain successfully aligned show continued improvement in both psychosocial and function scores from 6 weeks to 1 year postoperatively, indicating that improvement in HRQOL is long-lasting. They further reported that evaluation of HRQOL should be considered when reporting strabismus surgery outcomes in clinical trials, and may prove helpful in assessing outcomes in clinical practice.

Prognostic factors for recurrence after bilateral rectus recession procedure in patients with intermittent exotropia

S H Lim, B S Hwang and M M Kim

Eye. June 2012;26: 846-852. (f12)

The purpose of this study was to evaluate prognostic factors, specifically age, at the time of surgery, for recurrence after bilateral lateral rectus recession (BLR) in patients with intermittent exotropia. Medical records of 511 subjects who underwent BLR procedures between the ages of 3 and 10 years with more than 12 months of follow-up were retrospectively reviewed. A single surgeon performed all surgeries. Patients' surgical outcomes with a deviation of less than 10 prism diopters (PD) exotropia and less than 5 PD esotropia were defined as a success. Outcomes with more than 11 PD exotropia were designated as recurrences, and those with esotropia of more than 5 PD after 3 months of surgery were noted as overcorrection. Prognostic factors for recurrence were analyzed by multivariate logistic regression.

Of the 511 subjects, 371 had successful surgical outcomes and 129 had recurrences, whereas 11 were found to be overcorrected. Older age at surgery and immediate postoperative alignment proved to be significant factors influencing a favorable outcome by multivariate logistic regression analysis (P<0.05). Gender, photophobia, age at onset of exotropia, spherical equivalent refractive error, astigmatism, spherical equivalent anisometropia, and preoperative deviation size were not significantly predictive of success (P>0.05).

The authors conclude that in BLR recession for intermittent exotropia, increasing patient age at surgery was associated with lower recurrence rates.

Medial rectus muscle pulley posterior fixation sutures in accommodative and partially accommodative esotropia with convergence excess. Mitchell L, and Kowal L. J AAPOS 2012;16:125-130. (f12)

Medial rectus surgery for convergence excess esotropia can be augmented by posterior fixation. This can be performed with posterior scleral fixation sutures, or by fixating the muscle directly to the pulley. The later technique offers a possible safety advantage by avoiding scleral perforation. This study was a retrospective review of the use of this technique. Thirty-one patients underwent this procedure for convergence excess type esotropia and had the minimum required follow-up. Mean follow-up was 12.7 months, but only 38.5% had follow-up of 12 months. At 1 to 3 month follow-up target alignment was achieved in 76.9% of patients at distance and 61.5% of patients at near. Near-distance disparity decreased from 21.8 prism diopters (PD), to 4.5 PD. The only factor found to hamper this reduction postoperatively was a history of amblyopia. Quantitative measurements of intraoperative restriction to adduction produced by pulley sutures was performed in 12 patients and did not show good correlation with reduction of distance-near disparity. No serious complications occurred with this technique. This study was retrospective and really would be better served as a randomized study comparing this technique to other techniques (ex. Scleral posterior fixation, augmented medial rectus recessions). Other techniques also collapse distance-near disparity postoperatively. Follow-up for many patients was short and surgical success was not significantly better than that reported in the literature with other techniques.

Comparison of superior oblique suture spacers and superior oblique silicone band expanders. Awadein A and Gawdat G. J AAPOS 2012;16:131-135. (f12)

Two surgical techniques to treat patients with superior oblique overaction or Brown's syndrome are superior oblique suture spacers and superior oblique silicone band expanders. The results of these two surgical modalities in improving ductions and versions and changing fundus torsion and pattern-strabismus were evaluated. Also, surgery time and complications were reviewed. Consecutive patients who underwent these procedures for superior oblique overaction or Brown's syndrome over a five-year period were retrospectively analyzed. Twenty-five patients met diagnosis and surgery criteria and completed satisfactory follow-up. Thirteen underwent the spacer procedure and 12 underwent the expander procedure. Mean follow-up was 26.5 months and 28.7 month for spacers and expanders respectively. Fifteen of the patients had Brown's syndrome. Two-thirds had complete restoration of normal elevation in adduction after surgery and surgical technique did not alter success rates.

Improvement occurred gradually over a few months. There were no recurrences of decreased elevation. A-pattern strabismus had normal postoperative superior oblique function in ¾ spacer procedures and 2/3 expander procedures. A-pattern improvement and vertical deviations in primary position and in the affected side gaze were both improved and there was no statistically significant difference in the improvement based on which procedure was performed. Both groups showed equal improvement in fundus torsion. The spacer group surgery time was lower than that of the expander procedure and this was statistically significant. Expander patients also had more postoperative inflammation and there was one case of a granuloma-like mass developing which required removal of the implant. Study groups in this paper were small so any differences in success between groups would have been difficult to detect.

Intraoperative monitoring of torsion to prevent vertical deviations during augmented vertical rectus transposition surgery. Holmes JM, Hatt SR, and Leske DA. J AAPOS 2012;16:136-140. (f12)

Augmented vertical rectus transposition surgery to treat 6th nerve palsies and Duane syndrome can produce undesirable postoperative vertical deviations. The authors describe a method of monitoring torsion intraoperatively. The authors feel torsional changes may be associated with vertical changes when performing this procedure. The technique involves marking the 12 and 6 o'clock positions of the limbus and monitoring torsion as the transposition is being performed. The inferior rectus muscle belly position could be adjusted to counteract any induced torsion. Nine patients were included and postoperatively none complained of torsion. Three of the 9 did have a postoperative hypotropia, but only 1 was clinically significant. The authors feel their technique is helpful in decreasing postoperative vertical strabismus, but the rates of this in this paper are similar to those in the published literature. The study was small with a very large confidence interval

Torsional augmentation for the treatment of lateropulsion and torticollis in partial ocular tilt reaction. Brodsky MC, and Holmes JM. J AAPOS 2012;16:141-144. (f12)

Unilateral injury to the brainstem or utricular pathways involved with central vestibular function can cause a compensatory head tilt due to a tilt of the subjective visual vertical. This head tilt is not compensatory for diplopia so vertical misalignment surgery will not improve the patient. Rarely, patients may report lateropulsion (sensation of being pulled to one side). The authors hypothesized that increasing the torsional position of the eyes to further rotate the eyes in the direction of the tilt would realign the vertical poles of the eyes more closely with the subjective visual vertical and eliminate or reduce the need for a compensatory head tilt. They present one patient who underwent horizontal

transposition of the vertical rectus muscles to treat lateropulsion and persistent torticollis. One year after surgery, her head tilt had reduced from 45’ to 5’ and she reported marked subjective improvement. Interestingly double Maddox Rod testing did not actually improve compared to preoperatively. This is a somewhat novel use of vertical rectus muscle transposition that was used in a paradoxical fashion . Based on the success of the procedure in this paper, the authors suggest an alternative mechanism for vertical rectus muscle transposition success. The existing belief is that torsional rotation of the eyes in the direction of the head tilt induces a contraversive tilt of the visual world, which forces the patient to straighten the head to move it into closer alignment with the tilted visual world. These authors suggest the procedure realigns the vertical meridians with a tilted subjective visual vertical, which counter-rotates the tilted subjective vertical back toward the gravitational vertical, removing the need for the head tilt.

Unilateral rectus muscle recession in the treatment of Duane syndrome

. Natan K and Traboulsi EI. J AAPOS 2012;16:145-149. (f12)
This paper was a retrospective review of single rectus muscle recession in a cohort of Duane syndrome patients. 27 patients were included in the study with an average age at surgery of 13 years. Minimum follow-up was 6 months (except for 4 patients who only received 2-3 months of follow-up). 93% of patients experienced an improved head turn postoperatively to <10’. This data is slightly misleading since only 17 of the 27 patients had a head turn of >15’ preoperatively. 85% of patients had alignment in primary position of <10 PD postoperatively but again the data are somewhat misleading because 33% had this alignment preoperatively. Stereoacuity <100 arcsec improved from 31% of patients to 69% of patients. Two patients developed a limitation of adduction postoperatively. Younger patients in this study did not undergo objective measurements of torticollis which clouds the data somewhat. Data was lacking in some cases on stereoacuity and motility restrictions.

Long-term follow-up of bilateral botulinum toxin injections versus bilateral recessions of the medial rectus muscles for treatment of infantile esotropia.

Gursoy H, Basmak H, Sahin A, et al. J AAPOS 2012;16:269-273. (f12)

Infantile esotropia can be treated with botulinum toxin injection or incisional surgery. The authors retrospectively compared the long-term motor and sensory outcomes in these patients treated over a 3-year period. Forty patients were offered botulinum (4 declined) and 61 patients were offered surgery (3 declined). Of the 94 patients who received intervention, 51 satisfied all inclusion criteria (25 botulinum and 26 surgery). The botulinum group patients were treated at a younger age (10 months vs. 12.5 months) and were followed longer (84 months

vs. 75 months). Of the 25 patients who received botulinum injections, 9 received a second injection and 2 required a third injection. 12 of the 16 single injection patients achieved orthophoria and the remaining 4 were treatment failures and received surgery. Of those who required additional injections, 7 of 9 ultimately achieved a successful motor outcome. Motor success rate was 68% in the botulinum group versus 77% in the surgery group. Accommodative esotropia developed in 7 cases in the botulinum group and in 4 cases in the surgical group. In the surgical group, all of the failures had a preoperative deviation of >40 prism diopters. In the botulinum toxin group, the values were more evenly distributed. This study was retrospective and nonrandomized. Botulinum toxin was only offered to families who were deemed likely to cooperate, and therefore this may have influenced outcomes.

Graded anterior transposition of the inferior oblique muscle for V-pattern strabismus.

Akar S, Gokyigit B, and Yilmaz OF. J AAPOS 2012;16:286-290. (f12)

This paper evaluates the efficacy and complications of bilateral graded anterior transpositions of the inferior oblique muscles (ATIO) for the correction of V-pattern strabismus associated with inferior oblique muscle overaction. Consecutive patients over a seven-year period were included. 69 patients were included (44 V-pattern esotropia and 25 V-pattern exotropia). Two-thirds of patients had associated horizontal rectus muscle surgery. Patients received an average follow-up of 18.7 months. There was a statistically significant difference in postoperative inferior oblique overaction and V-pattern. Postoperatively, there were normal versions in 84% of V-pattern esotropes, and 82% had less than 10 prism diopters (PD) of V-pattern. For V-pattern exotropes these numbers were 80% and 80% respectively. Surgical dosing was not found to be an independent predictor of surgical outcome in either group. Objective extorsion was noted in 90% of patients preoperatively with an indirect ophthalmoscope and 0% postoperatively. No patients had subjective torsion complaints pre- or postoperatively. No patient had a postoperative vertical deviation or limited elevation.

Paediatric adjustable strabismus surgery

A Mokashi A, Stead RE and Stokes J.

Eye. Jul 2012;26:1024-5. (f12)

In this research letter, the authors describe their use of adjustable sutures in children ages 8 to 15 years. They use a hangback technique with a 6/0 vicryl adjustable, slip-knot tie, followed by sub-Tenon's levobupivacaine (5 mg/ml). Within 2–3 h of recovery from general anaesthesia, topical tetracaine hydrochloride 1% is instilled and a prism cover test is performed. The suture is

adjusted as required and the conjunctiva closed with 8-0 vicryl in the treatment room. Ninety-five percent (20/21) of our paediatric patients were able to tolerate this technique, with only one requiring a second general anesthetic.

Utility of Adjustable Sutures in Primary Strabismus Surgery and Reoperations

Kamiar Mireskandari, FRCOphth, PhD, Melissa Cotesta, BSc, OC(C), Jenifer Schofield, BA, OC(C)), COMT, StephenP. Kraft, MD, FRCSCS
Ophthalmology March, 2012; 119:629-633 (f12)

Four hundred four patients, older than 12 years who underwent strabismus surgery over a thirteen year period, were evaluated in a retrospective series designed to compare the success of adjustable suture (AS) and nonadjustable suture (NAS) strabismus surgery in reoperations and primary procedures.
All eyes underwent the same hang- back surgery technique in both the AS and NAS groups. Success was defined as alignment within ten prism diopters (PD) for horizontal muscle surgery and within five prism diopters for vertical strabismus surgery, without diplopia or further surgery.
Patients in the AS group required adjustment in 28.85% o cases. Higher overall success was seen with AS patients (77%) versus the NAS patients (69.1%) although, the results were not clinically significant. However, in exotropia surgery, there was a clinically significant advantage for the AS group versus the NAS group (80.8% vs. 65.9%). There was also a higher success for patients undergoing primary surgery (82.5% (AS) vs. 50% (NAS)). However, in the reoperation group there was no statistically significant difference between the AS and NAS groups.
Primary surgery in adults with exotropia has a more successful outcome with adjustable suture surgery. This advantage was not present with esotropia and vertical strabismus surgery.

Study performed at the Hospital for Sick Children, Toronto Ontario,Canada by the Department of Ophthalmology and Visual Sciences
COMMENT: Adjustable strabismus surgery, as a primary procedure, may be more successful than conventional fixed-placement surgery in children and adults with exotropia.

Strabismus Surgery Hemostasis
Research Correspondence-Letter to the Editor

Amit Gupta, MS, DNB, Ramesh Kekunnaya,MD,FRCS, Virender Sachdeva,MS,DNB , Harsha L Rao

Ophthalmology March, 2012; 119: 649-650 (f12)

The authors give a good review of the literature regarding potential systemic side effects of ophthalmic phenylephrine 10% or brimonidine 0.15%, which are frequently used prior to strabismus surgery to prevent intraoperative bleeding. They report a prospective, non randomized, case controlled study comparing preoperative use of brimonidine and phenylephrine with controls to determine the amount of intraoperative bleeding and postoperative subconjunctival hemorrhage in horizontal muscle surgery in children less than 8 years of age. Hydroxypropylmethycellulose (HPMC) was used in the control group and was compared to phenylephrine 10% and brimonidine 0.15% (Groups A, B, C respectively). One drop was placed in each eye 10 minutes prior to surgery. The surgeon was masked to the drop type. Fornix based strabismus surgery was performed and a rating scale was used in all cases. Patients in the brimonidine group showed the greatest reduction in intraocular bleeding and postoperative subconjunctival followed by the phenylephrine group followed by the control group.
This study affirms similar findings by Hong et al (AJO 2007; 144:468—70) which reported that phenylephrine and brimonidine were equally efficacious in reducing bleeding. There was no mention of adverse systemic effects for either medication in the Letter.
The study was performed in Hyderabad, India
COMMENT: Article cites Dr. Fraunfelder's work in 1980 and 2002 describing the adverse effects of topical ocular pledgets soaked with phenylephrine.

Comparison between graded unilateral and bilateral medial rectus recession for esotropia

Lihua Wang, Xiaoming Wang Br J Ophthalmol 2012;96:540-543. (f12)

Aims: To compare the postoperative surgical outcomes and the changes in deviation achieved per millimetre of recession in patients treated by graded unilateral medial
rectus (UMR) or bilateral medial rectus (BMR) recession for small to large angle esotropia with a minimum followup of 6 months. In this retrospective study 102 patients underwent UMR recession and BMR recession for constant esotropia measuring 15-35 prism diopters (PD) and 30-70 PD, respectively.

Results: No significant difference was observed between: (1) the success rates of the BMR and UMR recession groups at postoperative days 1w3 (p.1.00) or at final follow-up (p.0.421); (2) the variation in the mean change in deviation from postoperative days 1w3
to the final follow-up of the UMR (p.0.58) and BMR (p.0.56) recession groups; and (3) the mean correction in PD per millimetre of muscle recession in the UMR and
BMR (p.0.63) recession groups.

Conclusion:Graded UMR recession for 15e35 PD of esodeviation was as effective as graded BMR recession for 30-70 PD of esodeviation. There was no statistical
difference in changes in deviation per millimetre of recession between equivalent amounts of unilateral and bilateral recession.

Comments: The main limitation of the current study is that it is retrospective, and in a retrospective analysis of the results, the preoperative mean near esodeviation (target angle for surgery) in the two groups was not identical, being 26.7 PD in the UMR recession group compared with 48.5 PD in the BMR recession group ($p<0.05$). In addition, the type of surgical procedure (UMR or BMR) chosen for patients with 30 PD and 35 PD of
esodeviation was based on our own experience rather than randomly, and the number of patients with 7-mm (n.2) recessions in the BMR group was too small to be statistically evaluated. A further limitation is that a follow-up of 6 months is relatively short because over time there may be a further reduction in the surgical success rate.

The effect on quality of life of long-term botulinum toxin A injections to maintain ocular alignment in adult patients with strabismus

Joanne Hancox, Shanel Sharma, Kelly MacKenzie, Gill Adams Br J Ophthalmol 2012;96:838-840. (f12)

Methods: 65 patients who had undergone over 25 injections of botulinum toxin A for long-term control of their deviation were identified and asked to fill in and return the Adult Strabismus questionnaire (AS-20) to assess their QoL.
Results: 46 questionnaires were available for analysis. The mean AS-20 score in our patients compared favourably with that reported for normal controls and was much higher than that reported for patients with strabismus.

Conclusion: Long-term injections with botulinum toxin A is a good treatment for maintaining ocular alignment if squint surgery is not indicated and those patients receiving treatment score near the level of normal controls in QoL terms.
Comments: This study id the first of its kind to look at the QOL aspect of strabismus patients who are treated by Botoulinum.

Inferior Oblique Transposition Onto the Equator: The Role of the Equator in Development of Contralateral Inferior Oblique Overaction

TSeung-Hyun Kim, MD, PhD; Jae-Hoon Na, MD; Yoonae A. Cho, MD, PhD J Pediatr Opthalmol Strabismus 2012; 49:98-102 March/April (f12)

Inferior oblique transposition, although a powerful weakening procedure, can create anti-elevation syndrome. The article discusses the possibility of preventing this complication by placing the inferior oblique in different positions at or far behind the equator. The authors found that either if the above surgeries can still cause variable upgaze restriction and contralateral inferior oblique overaction.

Residual Symptoms After Surgery for Unilateral Congenital Superior Oblique Palsy

Ihsan Caca, MD; Alparslan Sahin, MD; Abdullah Cingu, MD; Seyhmus Ari, MD; Umut Akbas, MD J Pediatr Opthalmol Strabismus 2012; 49:103-108 March/April (f12)

The authors discuss unilateral superior oblique palsy with surgical interventions. Forty eight cases were reviewed with an inferior oblique myectomy/disinsertion of the inferior oblique with or without a superior rectus recession. Residual symptoms after vertical muscle surgery were documented. Although there may be patients requiring a second surgery, the above surgical interventions were found to be effective and successful procedures for the treatment of unilateral superior oblique palsy.

The Psychosocial Effects of Strabismus Before and After Surgical Correction in Chinese Adolescents and Adults

Jingling Xu, MD; Xinping Yu, MD; Ying Huang, ND, Jie Chen, MD, Huanyun Yu, MD; Yuwen Wang, MD; Fang Zhang J Pediatr Opthalmol Strabismus 2012; 49:170-175 May/June (f12)

The study evaluates the psychosocial problems associated with strabismus and the effects that corrective surgery has on Chinese adults and adolescents. Not unlike our population, this study found that Chinese adults and adolescents with strabismus have decreased self confidence and self esteem. They also have difficulty interacting in social situations. This is all improved with correction of the strabismus.

Suture Contamination in Strabismus Surgery

H. Sprague Eustis, MD; Annette Rhodes, MD J Pediatr Opthalmol Strabismus 2012; 49:206-209 July/August (f12)

This study evaluated the rate of suture contamination when no pre-treatment is applied to the suture versus antibiotic-coated sutures versus antibiotic/antiseptic –coated sutures. The rate of suture contamination was much lower when antibiotic/antiseptic coating was applied. The contamination is likely from the sutures sitting on the eyelashes when securing the sutures to the muscles. While

the rate of endophthalmitis is low in strabismus in general, we should still take all precautions to prevent suture contamination. Another method to prevent suture contamination is barrier draping of the lids and lashes.

Botulinum Toxin Injection into the Superior Rectus for Treatment of Strabismus

E. Dawson, N. Ali, and J.P. Lee
Strabismus March 2012; 20(1): 24-25 (f12)

This study comes from Moorfields Eye Hospital in London, United Kingdom. The purpose of the study is to evaluate the efficacy of botulinum toxin injection into the superior rectus muscle, particularly in cases of residual hypertropia status post strabismus surgery.

The authors reviewed 7,575 patients who received botulinum A toxin at the Moorfields Eye Hospital. Only 8 cases of superior rectus toxin were identified all of which had residual hypertropia following previous ocular surgery. The surgery comprised 3 patients retinal surgeries, 2 patients with thyroid eye disease, 2 patients with extraocular muscle surgery for thyroid eye disease, and 2 patients received transposition for sixth cranial nerve palsy.

In 7 out of the 8 cases there is a long-lasting mean reduction of 10 prism diopters from the original deviation. The long-lasting mean reduction occurred after the patient received “between 2 and 3 injections”. Ptosis occurred in all but 1 case but resolved with time.

The authors conclude **that the superior rectus toxin injection has very limited indications, but may be considered in residual hypertropia presumed secondary to a tight or overacting superior rectus where the patient can tolerate the temporary post injection ptosis.**

The Management of Traumatic Isolated Inferior Rectus Rupture

Ruchika Batra, MBBS, MRCOphth, Anna Gao, MBChB, PhD, Georges Adrien Shun-Shin, FRCOphth, MBBS
Strabismus September 2012; 20(3): 105-108 (f12)

This is a case report from the Wolverhampton Eye Infirmary, New Cross Hospital, Wolverhampton, United Kingdom.

The article describes a traumatic inferior rectus rupture following blunt trauma in a 42 year old “Heavy goods vehicle driver”. The authors describe a technique of

repair using a Hummelsheim-type procedure. Good alignment was achieved at 1 month following surgery, with an excellent field of binocular vision.

The patient remained asymptomatic for 15 months after surgery. His only residual symptom was vertical double vision in extreme up gaze. The patient was able to continue his job without any significant visual outcome. The authors stress that the surgical outcome was better than described in previous reports of inferior rectus rupture (articles by Paysse and Saunders, et al, *Journal AAPOS* 2000; 4:164-167, “Surgical Management of Strabismus After Rupture of the Inferior Rectus Muscle” and an article by Olitsky and Notaro, *J Paediatr Ophthalmol Strabismus* 2000; 37:50-51, “Anterior Transposition of the Inferior Oblique for the Treatment of a Lost Inferior Rectus Muscle”)

The authors feel that the “Hummelsheim procedure is a useful option in the management of an inferior rectus rupture”.

The ophthalmology surgical competency assessment rubric for strabismus surgery.

Golnik KC, Motley WW, Atilla H, et al. J AAPOS 2012;16:318-321. (f12)

The authors describe a method of designing and validating an assessment tool for rectus recession surgery. The goal is a standardized, internationally valid tool to guide and assess ophthalmology competence. 17 objective benchmarks were created, 16 of which represent different parts of the surgery and 1 representing communication. This is an attempt to provide a global standard, for teaching, training, and evaluation. Subjectivity in assessment should diminish.

Augmented Hummelsheim procedure to treat complete abducens nerve palsy.

Couser NL, Lenhart PD, and Hutchinson AK. J AAPOS 2-12;16:331-335. (f12)

The authors present a series of patients with complete abducens nerve palsy treated with an augmented Hummelsheim procedure (combined with medial rectus recession). This was a retrospective review. Ten patients were analyzed and followed for a mean follow-up of 28 weeks. The average age of the patients was 29 years. Six of the 10 had isolated unilateral traumatic abducens palsies and the other four were more complex. The mean primary position esodeviation improved from 43 PD to 6 PD with surgery at distance and from 43 PD to 9 PD at near. One patient developed a presumed slipped inferior rectus muscle, and was one of two patients who developed an induced vertical deviation after surgery. Slight abduction improvements were noted. This surgical procedure spares at

least one ciliary vessel in each of the vertical rectus muscles and avoids placing a lateral fixation suture.

Function of transected or avulsed rectus muscles following recovery using an anterior orbitotomy approach. Pineles SL, Laursen J, Goldberg RA, et al. J AAPOS 2012;16:336-341. (f12)

Only about half of muscles that are traumatically disinserted are recovered. The authors use an anterior orbitotomy approach with a recovery rate of 100% based on prior publications. This study evaluated muscle function after retrobulbar recovery via this technique. Eleven patients were included. Mean time from injury to muscle retrieval was 10 months and mean postoperative follow-up averaged 16 months. Postoperatively, there was a significant improvement in ductions in the field of action of the affected muscle, primary position deviation, and deviation in the field of action of the affected muscle. Interestingly, the only statistically significant associations with a successful outcome were a larger preoperative deviation in the primary position and less restriction on forced duction testing of the antagonist muscle. This probably can be explained by less coexisting restriction of the opposing muscle. Another possibility is that the surgeon was more aggressive with muscle advancement in cases with larger preoperative deviations. Patients with injury to the medial rectus muscle or with an injury from sinus surgery were less likely to have a successful subjective surgical outcome. Overall duction improvements were limited. Muscle recovery is advantageous over transpositions because of decreased risk of anterior segment ischemia and a possible larger field of single binocular vision due to active force generation by the recovered muscle.

Clinical findings, orbital imaging, and intraoperative findings in patients with isolated inferior rectus muscle paresis or underaction. Awadein A. J AAPOS 2012;16:345-349. (f12)

This study presents clinical, orbital imaging, and intraoperative findings of patients with inferior oblique underaction from various causes. Cases were culled from a 4-year retrospective review of medical records. Twenty-eight patients with a mean age of 39.8 years were identified. The most common cause was orbital trauma. Half of the patients had an exotropia in addition to the hypertropia and most displayed an A-pattern. All patients had subjective intorsion. Less than half of the patients displayed abnormalities on imaging.

Retinal vessel changes after laser treatment for retinopathy of prematurity. Kwon JY, Ghodasra DH, Karp KA, et al. J AAPOS 2012;16:350-353.

Patients with type 1 ROP had retinal vessel caliber and tortuosity measured. Measurements were repeated after laser treatment and the rate of change was also evaluated. Seventy-nine infants were treated with laser photocoagulation for type 1 ROP and thirty eyes met all inclusion criteria.

[Home]

CATARACT

Clinical characteristics, course, and visual prognosis of partial cataracts that seem to be visually insignificant in children. Choi J, Kim JH, Kim S-J, et al. J AAPOS 2012;16:161-167. (f12)

The authors report data from a retrospective chart review on clinical characteristics, course, intervention, and visual prognosis of partial cataracts that were visually significant over a 15-year period. One hundred and eighty-four eyes of 110 children were included in this study. Posterior subcapsular cataracts were the most common type in this study and the most common etiology was idiopathic followed by steroid induced. The mean age at first diagnosis was 6.2 years and mean follow up was 5.2 years. The most common associated ocular abnormality was cataracts. Slightly more than one-third of patients had systemic or syndromic abnormalities (the most common of which was nephrotic syndrome). Seventy-two of the 184 eyes required refractive correction. In all nonsurgical treatment groups, the final visual acuity showed improvement compared to the initial visual acuity. No progression of the lens opacity was noted in 163/184 eyes. The remaining 21 eyes progressed and received cataract extraction and intraocular lens implantation at a mean age of 10.7 years. The group that showed progression had worse visual acuities at the initial examination. It is important to view the data presented in this article in the context that the cataracts were partial and presented data cannot be applied to all cataracts. This is supported by the late first age at diagnosis.

Clinical and molecular analysis of children with central pulverulent cataract from the Arabian Peninsula Arif O Khan, Mohammed A Aldahmesh, Jawahir Y Mohamed, Fowzan S Alkuraya Br J Ophthalmol 2012;96:650-655. (f12)

Aim: To clinically and genetically characterise central pulverulent cataract in a consecutive cohort of children from the Arabian Peninsula
Methods: Ophthalmic examination, homozygosity mapping in a consanguineous family and candidate gene analysis.
Results: All 16 children (4e16 years old, mean 9 years; seven girls and nine boys from 10 families) had bilateral central nuclear dust-like lenticular opacities. Two patients (one family) had cortical riders and six had associated strabismus. Cycloplegic retinoscopy was usually hyperopic (13/16; right eye spherical equivalent +0.50 to +6.25 dioptres, mean +3.50) but was sometimes myopic (3/16; right eye spherical equivalent

0.50 to _11.75, mean _6.50). In children with amblyopia (5/16), the cause was significant uncorrected ametropias rather than the lens opacities. Three patients had uncomplicated unilateral cataract surgery suggested by an outside second opinion that did not improve bestcorrected visual acuity. Homozygosity mapping for one
consanguineous family suggested the candidate gene CRYBB1. Sequencing of this gene revealed a homozygous c.171del mutation (p.N58Tfs*107) witha shared haplotype in all 16 children.
Conclusions: Central pulverulent cataract in this consanguineous population does not significantly impact visual acuity during early childhood, can be associated with significant ametropias (with amblyopia and/or strabismus) and is specific for a homozygous CRYBB1 founder mutation. Primary management in children is
typically spectacle correction based on cycloplegic retinoscopy to treat significant refractive error rather than paediatric cataract surgery

[Home]

CATARACT SURGERY

Glued intrascleral fixation of posterior chamber intraocular lens in children

Kumar DA, Agarwal A, Prakash D, Prakash G, Jacob S, Agarwal A. Am J Ophthalmol. 2012;153(4):594-601. (f12)

This retrospective observational case series evaluated the short-term results of glued intrascleral fixation of posterior chamber intraocular lens (glued IOL) in children without adequate capsular support. Forty-one eyes of 33 children who underwent glued IOL implantation were retrospectively evaluated. The indications were postsurgical aphakia, subluxated cataract, ectopia lentis, traumatic subluxation, and decentered IOL. Main outcome measures: Visual acuity (VA), endothelial cell changes, intraoperative and postoperative complications. The mean age at the time of glued IOL was 10.7±3.6 years (range 5-15). The mean duration of follow-up after surgery was 17.5±8.5 months (range 12-36). The mean postoperative best spectacle-corrected visual acuity (BCVA in decimal equivalent) was 0.43±0.33 and there was significant change noted (P<0.001). Postoperatively, 20/20 and >20/60 BCVA was obtained in 17.1% and 46.3% of eyes respectively. BCVA improvement more than 1 line was seen in 22 eyes (53.6%). The mean postoperative refraction was myopic (-1.19±0.7 diopters [D]) in 19 eyes and hyperopic (+1.02±0.7 D) in 22 eyes. The mean endothelial loss was 4.13% (range 1.3%-5.94%). The 3 causes of reduced BCVA were the preexisting corneal, retinal pathology, and amblyopia. Postoperative complications included optic capture in 1 eye (2.4%), macular edema in 2 eyes (4.8%), and clinical decentration in 2 eyes (4.8%). There was no postoperative retinal detachment, IOL dislocation, endophthalmitis, or

glaucoma. The authors concluded that short-term results in children after glued IOL were favorable, with a low rate of complications. However, regular follow-ups are required since long-term risks are unknown.

Long-term Outcomes of Undercorrection Versus Full Correction After Unilateral Intraocular Lens Implantation in Children

Lambert SR, Archer SM, Wilson ME, Trivedi RH, del Monte MA, Lynn M. Am J Ophthalmol. 2012; 153(4):602-8. (f12)

This retrospective case-control study evaluated the impact of full correction vs undercorrection on the magnitude of the myopic shift and postoperative visual acuity after unilateral intraocular lens (IOL) implantation in children. The medical records of 24 children who underwent unilateral cataract surgery and IOL implantation at 2 to <6 years of age were reviewed. The patients were divided into 2 groups based on their 1-month-postoperative refraction: Group 1 (full correction) -1.0 to +1.0 diopter (D) and Group 2 (undercorrection) ≥+2.0 D. The main outcome measures included the change in refractive error per year and visual acuity for the pseudophakic eyes at last follow-up visit. The groups were compared using the independent groups t test and Wilcoxon rank sum test. The mean age at surgery (Group 1, 4.2±0.9 years, n=12; Group 2, 4.5±1.0 years, n=12; P=.45) and mean follow-up (Group 1, 5.8±3.7 years; Group 2, 6.1±3.5 years; P=.69) were similar for the 2 groups. The change in refractive error (Group 1, -0.4±0.5 D/y; Group 2, -0.3±0.2 D/y; P=.70) and last median logMAR acuity (Group 1, 0.4; Group 2, 0.4; P=.54) were not significantly different between the 2 groups. The authors concluded that they did not find a significant difference in the myopic shift or the postoperative visual acuity in children aged 2 to <6 years of age following unilateral cataract surgery and IOL implantation if the initial postoperative refractive error was near emmetropia or undercorrected by 2 diopters or more.

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

Kekunnaya R, Gupta A, Sachdeva V, Rao HL, Vaddavali PK, Prakash VO. Am J Ophthalmol 2012; 154(1):13-9. (f12)

This retrospective comparative study assessed the accuracy of IOL power calculation formulae in children less than 2 years of age. 128 eyes of 84 with congenital cataract who underwent primary IOL implantation were analyzed. Data were analyzed for prediction error using the 4 commonly used IOL power calculation formulae. The absolute prediction error was calculated with each of the formulae and the formula that gave least variability was determined. The formula that gave the best prediction error was determined. The mean age at surgery was 11.7 ± 6.2 months. Absolute prediction error was found to be 2.27 ± 1.69 diopters (D) with SRK II, 3.23 ± 2.24 D with SRK T, 3.62 ± 2.42 D with Holladay, and 4.61 ± 3.12 D with Hoffer Q. The number of eyes with absolute

prediction error within 0.5 D was 27 (21.1%) with SRK II, 8 (6.3%) with SRK T, 12 (9.4%) with Holladay, and 5 (3.9%) with Hoffer Q. Comparison between different formulae showed that the absolute prediction error with SRK II formula was significantly better than with other formulae (P < .001). Prediction error with SRK II formula was not affected by any factor such as age (P = .31), keratometry (P = .32), and axial length (P = .27) of the patient. Axial length influenced the absolute prediction error with Holladay (P = .05) and Hoffer Q formulae (P = . 002). Mean keratometry influenced prediction error (P = .03) with SRK T formula. The authors concluded that although absolute prediction error tends to remain high with all present IOL power calculation formulae, SRK II was the most predictable formula in their series.

Predictability of Intraocular Lens Calculation and Early Refractive StatusThe Infant Aphakia Treatment Study

Deborah K. VanderVeen, MD; Azhar Nizam, MS, et al for the Infant Aphakia Treatment Study Group.. Arch Ophthalmol. 2012;130(3):293-299 (f12)

The goal of this study was to report the accuracy of intraocular lens (IOL) power calculations and the early refractive status in pseudophakic eyes of infants in the Infant Aphakia Treatment Study.Eyes randomized to receive primary IOL implantation were targeted for a postoperative refraction of +8.0 diopters (D) for infants 28 to 48 days old at surgery and +6.0 D for those 49 days or older to younger than 7 months at surgery using the Holladay 1 formula. Refraction 1 month after surgery was converted to spherical equivalent, and prediction error (PE; defined as the calculated refraction minus the actual refraction) and absolute PE were calculated. Fifty-six eyes underwent primary IOL implantation; 7 were excluded for lack of postoperative refraction (n = 5) or incorrect technique in refraction (n = 1) or biometry (n = 1). Overall mean (SD) absolute predictive error (PE) was 1.8 (1.3) D and mean (SD) PE was +1.0 (2.0) D. Absolute PE was less than 1 D in 41% of eyes but greater than 2 D in 41% of eyes. Mean IOL power implanted was 29.9 D (range, 11.5-40.0 D); most eyes (88%) implanted with an IOL of 30.0 D or greater had less postoperative hyperopia than planned. Multivariate analysis revealed that only short axial length (<18 mm) was significant for higher PE. Overall, short axial length correlates with higher PE after IOL placement in infants. Less hyperopia than anticipated occurs with axial lengths of less than 18 mm or high-power IOLs. Quality A-scans are essential and higher PE is common, with a tendency for less hyperopia than expected.

Glaucoma-Related Adverse Events in the Infant Aphakia Treatment Study1-Year Results

Allen D. Beck, MD; Sharon F. Freedman, MD; Michael J. Lynn, MS, et al for the Infant Aphakia Treatment Study Group. Arch Ophthalmol. 2012;130(3):300-305 (f12)

The purpose of this study was to report the incidence of glaucoma and glaucoma suspects in the IATS, and to evaluate risk factors for the development of a glaucoma-related adverse event in patients in the IATS in the first year of follow-up.A total of 114 infants between 1 and 6 months of age with a unilateral congenital cataract were assigned to undergo cataract surgery either with or without an intraocular lens implant. Of these 114 patients, 10 (9%) developed glaucoma and 4 (4%) had glaucoma suspect, for a total of 14 patients (12%) with a glaucoma-related adverse event in the treated eye through the first year of follow-up. Of the 57 patients who underwent lensectomy and anterior vitrectomy, 5 (9%) developed a glaucoma-related adverse event; of the 57 patients who underwent an intraocular lens implant, 9 (16%) developed a glaucoma-related adverse event. The odds of developing a glaucoma-related adverse event were 3.1 times higher for a child with persistent fetal vasculature and 1.6 times higher for each month of age younger at cataract surgery. Modern surgical techniques do not eliminate the early development of glaucoma following congenital cataract surgery with or without an intraocular lens implant. Younger patients with or without persistent fetal vasculature seem more likely to develop a glaucoma-related adverse event in the first year of follow-up. Vigilance for the early development of glaucoma is needed following congenital cataract surgery, especially when surgery is performed during early infancy or for a child with persistent fetal vasculature.

Predictors of adherence to occlusion therapy 3 months after cataract extraction in the Infant Aphakia Treatment Study. Drews-Botsch CD, Hartmann EE, and Celano M. J AAPOS 2012;16:150-155. (f12)

Patching compliance in the Infant Aphakia Treatment Study (IATS) was assessed every 3 months by trained interviewers questioning the primary caregiver. Data was obtained on 104 children. Three months after surgery, 60% reported achieving at least 75% of prescribed patching and almost half reported achieving at least 90%. Aphakia and pseudophakic rates were similar. Greater socioeconomic status correlated with better rates of adherence. Lower reported parenting stress and private insurance both correlated with increased patching wear-time in the 3-month postop time period. White children patching rates were reported as higher than in non-white children at the six-month follow-up.

Adherence to Occlusion Therapy in the First Six Months of Follow-Up and Visual Acuity among Participants in the Infant Aphakia Treatment Study (IATS). Carolyn D. Drews-Botsch, Marianne Celano, Stacey Kruger, E. Eugenie Hartmann, for the Infant Aphakia Treatment Study. Invest Ophthalmol Vis Sci. 2012 Jun 5;53(7):3368-75. (f12)

Achieving good vision in infants born with a unilateral cataract is believed to require early surgery and consistent occlusion of the fellow eye. This study examined the relationship between adherence to patching and grating acuity. Data came from the Infant Aphakia Treatment Study, a randomized clinical trial of treatment for unilateral congenital cataract. Infants were either left aphakic (n = 53) or had an intraocular lens implanted (n = 55). Patching was prescribed 1 hour per day per month of age until 8 months of age and 50% of waking hours thereafter. Adherence was measured as the mean percentage of prescribed patching reported in a 7-day diary completed 2 months after surgery, and 48-hour recall interviews conducted 3 and 6 months after surgery. Grating visual acuity was measured within 1 month of the infant's first birthday (n = 108) using Teller Acuity Cards by a tester masked to treatment. Nonparametric correlations were used to examine the relationship with grating acuity.

On average, caregivers reported patching 84.3% (SD = 31.2%) of prescribed time and adherence did not differ by treatment (t = −1.40, df = 106, p = 0.16). Adherence was associated with grating acuity ($r_{Spearman}$ = −0.27, p < 0.01), but more so among pseudophakic ($r_{Spearman}$ = −0.41, p < 0.01) than aphakic infants ($r_{Spearman}$ = −0.10, p = 0.49).

This study empirically has shown that adherence to patching during the first 6 months after surgery is associated with better grating visual acuity at 12 months of age after treatment for unilateral cataract and that implanting an intraocular lens is not associated with adherence.

Fixation control and eye alignment in children treated for dense congenital or developmental cataracts. Birch EE, Wang J, Felius J, et al. J AAPOS 2012;16:156-160. (f12)

The authors evaluate factors that affect ocular motor outcomes following cataract surgery during the first 5 years of life. Eye movement recordings were utilized for nystagmus evaluation. Forty-one children were enrolled, with an even distribution of congenital and developmental cataracts. Twenty-seven of these children developed strabismus. Congenital cataracts and visual deprivation >6 weeks were associated with a significant increased risk for strabismus. Nystagmus developed in 29 children and risk factors for this condition were strabismus, visual deprivation >6 weeks duration, and infantile onset (<= 12 months) of the cataracts. Asymmetric fixation instability was present in 14 children. Risk factors for interocular difference in type or amplitude of nystagmus were unilateral cataract, or congenital onset. The authors propose that time of onset, duration, and severity of the cataract(s) all contribute to the ocular motor outcome in these children.

Vitrectorhexis and lens aspiration with posterior chamber intraocular lens implantation in spherophakia.

Al-Haddad C, Khatib L.
J Cataract Refract Surg. Jul 2012;38:1123-6. (f12)

The authors describe a technique very similar to the pediatric cataract surgery technique pediatric ophthalmologists are already familiar with, but apply this technique to a seven year-old patient with bilateral spherophakia causing chronic angle closure glaucoma. The authors describe their technique and provide a video.

After an anterior chamber maintainer is placed, the 20 Gauge ocutome is introduced through a limbal incision to perform a circular vitrectorhexis to avoid excessive manipulation of the unstable lens, followed by gentle cortex aspiration. A foldable IOL is injected. In the right eye, the authors inject a three piece lens, and one haptic goes into the sulcus and the other in the bag. In the left eye, the authors inject a one piece lens into the capsular bag. Through a pars plana incision, the ocutome is used to perform a posterior capsulotomy to prevent late posterior capsule opacification. The authors follow the patient for four months. The patient has transient phacodonesis in the left eye.

This surgical technique report provides a useful example of how to remove a spherophakic lens with minimal stress on the zonules, preserving the capsular bag. The authors do not have enough follow-up to comment on the safety of IOL implantation. The authors comment that primary posterior capsulotomy is performed in children under age 18 years, but this is open to debate.

Outcomes of pediatric cataract surgery in anterior persistent fetal vasculature.

Vasavada AR, Vasavada SA, Bobrova N, Praveen MR, Shah SK, Vasavada VA, Pardo A JV, Raj SM, Trivedi RH.
J Cataract Refract Surg. May 2012;38(5):849-57. (f12)

The pupose of the study was to report intraoperative techniques and postoperative outcomes for the treatment of pediatric cataracts due to persistent fetal vasculature (PFV). 33 eyes at two centers in India and the Ukraine underwent surgical treatment for unilateral PFV cataracts at the age of 1-24 months. The mean age at surgery was 6.3 (± 5.2) months.

The authors used a limbal approach via two paracenteses. Anterior continuous curvilinear capsulorhexis (CCC) was performed in 23 eyes (70%). Intraoperatively, posterior capsule plaque was seen in 20 eyes (61%). The authors describe cutting the posterior capsular plaques with microscissors. In 10 eyes (30%), the lens was converted into a fibrovascular mass. In 31 eyes (94%), 2-port limbal anterior vitrectomy was performed, and in 2 eyes (6%), pars plana vitrectomy was performed. Intraocular lens implantation was performed in 16 eyes (49%); 17 eyes (51%) were left aphakic.

The authors followed the patients for three years. Visual axis obscuration occurred in 6 eyes (18%), requiring additional intraocular surgery. Two eyes (6%) developed glaucoma necessitating surgery. Comparing preoperative to postoperative vision, visual acuity was reported as stable in 11 eyes (33%) and

improved in 22 eyes (67%). However, these visual acuity comparisons were primarily based on whether the subjects were able to fix and follow light. Optotype visual acuity was available for 16 eyes at the age of 3 years. 8 aphakic eyes had a mean visual acuity of logmar 0.56 (+0.18) (Snellen 20/73) and 8 pseudophakic eyes had a mean visual acuity of logmar 0.54+0.46 (Snellen 20/69).

The authors conclude that good visual outcomes can be obtained in PFV eyes after surgical intervention, with an acceptable rate of serious postoperative complications.

Management and outcomes of cataract in children: The Toronto experience.

Lim Z, Rubab S, Chan YH, and Levin AV. J AAPOS 2012;16:249-254. (f12)

The authors review a large cohort of patients with pediatric cataracts and what factors affect management and outcomes. This was an 11- year retrospective review. Cataract extraction was performed on 828 of 1122 eyes. Patients with total, nuclear, and lamellar cataracts were significantly more likely to undergo surgery. Anterior polar and lamellar cataracts were more likely to be managed without surgery. Mean follow-up was 31 months. 102/828 surgical eyes required additional surgeries. The most common indication for additional surgeries was opacification within the pupil, which occurred in 50/374 (13%) pseudophakic eyes and 20/462 (4%) of aphakic eyes. Glaucoma occurred in 123/1122 eyes (11%). Slightly more than half had glaucoma prior to cataract surgery. Glaucoma was more prevalent in aphakic eyes than in psuedophakic eyes (12% vs 1%). These numbers are probably biased to some degree by the fact that the aphakic patients averaged an earlier age for their cataract surgery and the pseudophakic patients had fewer risk factors for glaucoma. Age at surgery for aphakic glaucoma averaged 24 months versus 108 months for pseudophakic glaucoma. Cataract surgery performed earlier in life has been reported to increase the risk of glaucoma, but this data is also biased by the difference in average age at the time of cataract surgery between the two groups. The prevalence of strabismus was 22%. However only 13% of pseudophakic patients developed strabismus versus 36% of those using contact lenses for visual rehabilitation. Persistence of poor postoperative vision after cataract surgery was significantly correlated with strabismus. Final acuity of 20/30 or better was obtained in 39% of unilateral cataracts, as well as in the right eye of bilateral cases. Unilateral pseudophakic patients had a greater likelihood of 20/30 or better acuity and a lower likelihood of 20/400 or worse acuity. This discrepancy was not found in bilateral cases. Visual outcomes were significantly better for posterior subcapsular, nuclear, lamellar and traumatic cataracts.

Intraocular Lenses in Children – Letter to the Editor

Sumit Monga, FRCS, Muralidhar Ramappa, MD, Ramesh Kekunnaya, FRCS, Sunita Chaurasia, MD, Harsha Rao, MD, DNB.
Ophthalmology July 2012; 119:1503-1504 (f12)

This letter refers to the article “Complications, adverse events, and additional intraocular surgery 1 year after cataract surgery in the infant aphakia treatment study” by Plager et al (*Ophthalmology* 2011; 118:2330-2334). The authors had some questions regarding the methodology of the original article. The authors felt that the psychological trauma that parents and young children undergo with additional surgery to remove reproliferated lens epithelial cells should not be overlooked. The limited data available from the Infant Aphakia Treatment Study as well as other retrospective studies evaluate safety profiles of primary IOL implantation in infantile patients revealed much fewer operation rates. The point of the editorial is that “primary IOL implantation in infants would be more affectively studied in multi-center trials with centers from around the world.” The reply by Dr. Plager et al emphasized that all patients underwent surgery using the same protocol regardless of which of the 12 study centers the patient was operated in. The patients underwent a pars plana approach to the primary posterior capsulotomy and anterior vitrectomy because of a history of greater access it allows and the decreased likelihood of displacing the intraocular lens from the bag during surgical manipulation.

Pediatric IOL Opacification

O. Murat Uyar, MD, FEBO, Vedat Kaya, MD
Ophthalmology August 2012; 119: 1717-1718 (f12)

This Letter to the Editor is in response to the previous article in *Ophthalmology* 2011; 118: 2128-2132, by Pehere NK, Bojja S, Vemuganti GH, et al, titled “Opacification of Intraocular Lenses Implanted During Infancy: a Clinicopathologic Study of 4 explanted Intraocular Lenses”.

The authors of the Letter to the Editor had the following observations: Opacification of intraocular lenses (IOLs) after implantation can be related to various factors including the IOL manufacturing process, IOL storage, adjuvants used during surgery, surgical techniques, patient-associated conditions, and mainly postoperative inflammation.

The authors of the original paper report the “fish-egg” appearance of certain intraocular lenses as a “novel finding”. The authors of the letter state that they have seen similar precipitations in certain types of hydrophilic acrylic lens implants. The authors of the letter stress that in the infant group, intraocular lens implantation has a high complication and reoperation rate. The rate, character, and severity of intraoperative complications, adverse events and additional

intraocular surgeries were numerically higher in the intraocular lens group compared with the contact lens group during the first postoperative year.

The Letter by Uyar and Kaya was responded to by the original authors Niranjan Pehere, Sreedhar Bojja, Geeta Vemuganti, et al, from Hyderabad, India. Dr. Pehere stated that “IOL implantation in infancy is controversial and more experience with intraocular lenses and their placement need to be studied. All pediatric cataract surgeons should meticulously document all adverse events, study them in detail and report the outcomes.”

Early Experience with the Femtosecond Laser for Cataract Surgery

Shveta Jindal Bali, MBBS, MD, Chris Hodge, BAppSc (Orth), Michael Lawless, MBBS, FRANZCO, Timothy V. Roberts, MBBS, MMed, Gerard Sutton, MBBS, MD
Ophthalmology May, 2012; 119:891-899 (f12)

The purpose of this Australian based prospective, consecutive cohort study is to describe the intra-operative complications and the learning curve associated with the intraocular use of femtosconsd (FS) laser for the removal of adult cataracts. The study included 200 eyes, operated by six different surgeons with a mean operative age of 69 years. The cases underwent anterior capsulotomy, lens fragmentation, and corneal incisions with the FS laser. The procedure was then completed with phacoemulsification and insertion of an intraocular lens.
Data regarding patient demographics, preoperative investigations and intraoperative complications were collected and divided into four chronologically stratified groups (looking for learning curve related complications). 10% of eyes had free-floating anterior capsular fragments (tags); 4% had anterior radial tears; 3.5% with a posterior capsular rupture and 2% with a dropped nucleus (4/200 cases).
The surgeons experienced most of their complications in the first hundred cases; surgeons with previous FS laser experience had fewer complications.
The study demonstrated a clear learning curve associated with the procedure that seemed to flatten with surgical experience.

The study was performed at the Vision Eye Institute in Chatswood, Australia and the Save Sight Institute affiliated with the Sydney Medical School/University of Sidney, Sydney, Australia
COMMENT: This article heralds the transition to FS laser surgery for adult and ultimately pediatric cataract surgery, which has already started outside of the United States. Please read Dr. Roger F Steinert’s editorial on page 889 for historical perspective and future challenges in the United States.

Comparing the astigmatic outcome after paediatric cataract surgery with different incisions. Gupta A, Ramappa M, Kekunnaya R, Vaddavalli PK, Ray D, Chaurasia S, Rao HL, Garudadri CS. Br J Ophthalmol. 2012 Mar;96(3):386-9 (f12)

Aims: To compare astigmatism with clear corneal incision versus scleral tunnel incision following paediatric cataract surgery with primary intraocular lens implantation.
Retrospective, comparative case series of 218 eyes of 138 children <12 years of age undergoing cataract extraction with intraocular lens implantation.The study cohorts were grouped into two categories i.e clear corneal versus scleral tunnel The mean postoperative astigmatism in group 1 was 1.2860.97 D, 1.4261.00 D and 1.3860.98 D at 1, 3 and 6 months respectively. The mean astigmatism i group 2 was 1.3461.20 D, 1.1360.88 D and 1.0360.89 D at 1, 3 and 6 months respectively. The amount of astigmatism was comparable between the two groups at 1 month postsurgery (p.0.90), while it was significantly lower in the corneal incision group at 3 (p.0.03) and 6 months (p.0.01).
Conclusions: Postoperative astigmatism after paediatric cataract surgery by clear corneal incision was lower compared with scleral incision.

Comments: Good study which compares the two incisions. No cases of wound infection.
But no preop keratometry reading and multiple optometrists did the refraction.

Artisan iris-claw lenses for the correction of aphakia in children following lensectomy for ectopia lentis Catherine Cleary, Bernadette Lanigan, Michael O'Keeffe Br J Ophthalmol 2012;96:419e421.(f12)

Purpose To describe the results of Artisan iris-claw lens implants in children with aphakia following lensectomy for ectopia lentis.
Method: We measured visual acuity, refractive error and endothelial cell counts and recorded complications in a group of children after insertion of Artisan iris-claw implants.
Results: Artisan implants were implanted in eight eyes of five children. Mean follow-up was 28 months (range: 4e58 months). The mean preoperative LogMAR bestcorrected visual acuity was 0.2160.2, and postoperatively, mean LogMAR uncorrected visual acuity was 0.0460.09 (p.0.04). Mean endothelial cell countwas 33126277 cells preoperatively and 29136268 cells postoperatively, representing a mean cell loss of 14.2% (p<0.001). Mean defocus equivalent was 11.3861.04 preoperatively and 2.3460.66 postoperatively. We encountered no postoperative complications in any of the eyes.

Conclusion: Artisan iris-claw implants are safe and effective in the correction of aphakia in children following lensectomy for ectopia lentis, enabling good spectaclefree distance vision in 75% of eyes with no postoperative complications.

Comments: Even though the sample size is small, Artisan lens may be promising in young children with lack of posterior capsule.

Can virtual reality simulation help to determine the importance of stereopsis in intraocular surgery? Salman Waqar, Olayinka Williams, Jonathan Park, Neil Modi, Thomas Kersey,Tamsin Sleep Br J Ophthalmol 2012;96:742-746. (f12)

Aim: To establish the effect of acute loss of stereopsis on simulated intraocular surgical performance.
Methods: This study was performed using the EYESi ophthalmic surgical simulator. Thirty junior doctors with no previous ophthalmic surgical experience were enrolled
and distance visual acuity (Snellen), near visual acuity and stereoacuity (Frisby) were recorded. All participants completed a standard introductory programme on the
forceps module to eliminate the learning curve. They then undertook four attempts of level 4 forceps module binocularly and another four monocularly to simulate an acute loss of stereopsis. Total score, odometer movement, corneal area injured, lens area injured and total time taken were recorded.
Results: Mean age was 31 years (SD69). None had amblyopia, with all demonstrating distance visual acuity of 6/6 or better and N6 for near. Mean stereopsis was 35 s of arc (SD618). Average total score decreased from 60 while operating binocularly to 47 monocularly ($p<0.05$). Average corneal area injured increased from 0.95 mm2 to 2.30 mm2 ($p<0.05$), average lens area injured increased from 1.76 mm2 to 3.53 mm2 ($p<0.05$)
and average time taken increased from 69.6 s to 77.4 s ($p<0.05$).
Conclusion: The importance of stereopsis for intraocular surgery is difficult to establish in a live theatre setting without compromising patient safety.

Comments: This study demonstrates a statistically significant decrease in simulated intraocular surgical performance with acute loss of stereopsis in potential ophthalmic training applicants. Caution is recommend in using these results to advocate stereopsis testing as a screening tool in interviews because some participants performed well despite an absence of stereopsis.

Simultaneous Bilateral Cataract Surgery With IOL Implantation in Children in Kenya Dan Gradin, MD, ABO; Daniel Mundia, MBChB, MMed J Pediatr Opthalmol Strabismus 2012; 49:139-144 May/June (f12)

There is great pressure in developing countries to decrease costs, increase efficiency in the operating room and to decrease days of admission to the hospital. In an effort to comply with the above criteria, the authors began performing bilateral same day cataract surgery with IOL insertion. The goal was to comply with the criteria imposed by the institution without compromising patient safety. In addition to resulting in a 20% cost reduction and decreasing hospital stay times from 8 days to 5 days. In simultaneous bilateral cataract surgery the late complications, although higher than sequencial surgery, were not sight-threatening. Simultaneous bilateral cataract surgery proved to be a good alternative to sequential surgery in developing countries without compromising patients' needs and satisfaction and most importantly, safety.

Subluxation of the Crystalline Lens: A No-Ring Approach

Antonio Carlos L. Rodrigues, MD; Rupal H. Trivedi, MD, MSCR; M. Edward Wilson, MD J Pediatr Opthalmol Strabismus 2012; 49:157-163 May/June(f12)

The article offers a surgical alternative for subluxed lenses in children and young adults that offers centration of the intraocular lens without using a capsulotension ring or suturing to the sclera. The article details the surgical technique well with figures to assist in explaining the technique. An interesting read for those who perform cataract surgery in this population. The article also discusses post operative complications and treatment as they compare to other techniques

Completion rates of anterior and posterior continuous curvilinear capsulorrhexis in pediatric cataract surgery for surgery performed by trainee surgeons with the use of a low-cost viscoelastic

Muralidhar R, Siddalinga Swamy GS, Vijayalakshmi P. Indian J Ophthalmol 2012;60:144-6 (Mar-Apr) (f12)

The study aims to determine the success rates for anterior and posterior capsulorrhexis and intraocular lens (IOL) implantation in the bag for pediatric cataract surgery performed with the aid of a low-molecular-weight viscoelastic. Children less than 6 years of age who underwent cataract surgery with IOL implantation in the period May 2008-May 2009 were included. The surgeries were done by pediatric ophthalmology fellows. The success rate for completion was 66.7% and 88.2 % for anterior and posterior capsulorrhexis, respectively. IOL implantation in the bag was successful in 87.9%. **Conclusions:** 2% hydroxypropylmethylcellulose is a viable low-cost alternative to more expensive options similar to high-molecular-weight viscoelastics. This is of great relevance to hospitals in developing countries.

Anti-inflammatory Effect of Low-Molecular-Weight Heparin in Pediatric Cataract Surgery: A Randomized Clinical Trial Vasavada VA, Praveen MR, Shah SK, Trivedi RH, Vasavada AR Am J Ophthalmol 2012;154(2):252-8. (f12)

This prospective masked randomized controlled trial sought to determine if intraocular infusion of low-molecular-weight heparin (enoxaparin) reduces postoperative inflammation in pediatric eyes undergoing cataract surgery with IOL implantation. Twenty children (40 eyes) undergoing bilateral cataract surgery with IOL implantation were randomized to receive enoxaparin in the intraocular infusion fluid (BSS) (Group I) or not to receive enoxaparin (Group II). The first eye was randomly assigned to 1 of the 2 groups and the second eye received alternate treatment. Patients were followed up in the first week and 1 and 3 months after surgery. Outcomes included: anterior chamber flare and cells (Hogan's criteria), cell deposits on IOL, posterior synechiae. The authors reported that one week postoperatively, no eyes had >grade 2 flare/cells. Proportion of eyes with grade 2 cells was higher in eyes that did not receive enoxaparin (Group II: 80% vs Group I: 40%, *P* = .009). In the first week >10 small cell deposits were noted in the eyes that received enoxaparin (Group I: 20%, Group II: none, *P* = .005). Large cell deposits first appeared at 1 month in 40% of eyes in Group I and 55% of eyes in Group II (*P* = .34) and increased at 3 months (60% in both groups, *P* > .999). Posterior synechiae were seen in 10% of eyes in Group I at 1 month, which persisted at 3 months; no eyes in Group II showed posterior synechiae (*P* = .14). The authors concluded that their study suggests that there does not seem to be a benefit of using enoxaparin in the infusion fluid with respect to early postoperative inflammation.

[Home]

REFRACTIVE SURGERY

GENETICS

Myotonia congenita with strabismus in a large family with a mutation in the SCN4A gene

Du H, Grob SR, Zhao L, Lee J, El-Sahn M, Hughes G, Luo J, Schaf K , Duan Y, Quach J, Wei X, Shaw P, Granet D, Zhang K.

Eye. Aug 2012;26:1039–1043. (f12)

The authors report on the ocular and genetic findings of five of seven members of a Caucasian family with myotonia congenita. Four of the five patients had eyelid myotonia, or delayed eyelid opening after forceful lid closure. All five patients had prior strabismus surgery and exhibited residual small angle esotropias. The CLCN1 gene coding for a chloride channel and the SCN4A gene coding for a sodium channel were sequenced and analyzed for mutations. These ion channels are important for skeletal muscle contraction. Sequencing results revealed a point mutation in the SCN4A gene in all five affected family members. The proband initially presented to an ophthalmologist at age 5-6 months with eyelid myotonia, and the ophthalmologist may thus be the first person to make the diagnosis. There is an association between myotonia congenita and malignant hyperthermia, which has implications when these patients undergo general anesthesia.

PAX6 Mutations Identified in 4 of 35 Families with Microcornea

Panfeng Wang,Wenmin Sun, Shiqiang Li, Xueshan Xiao, Xiangming Guo, and Qingjiong Zhang**.** IOVS September 2012 53:6338-6342; published ahead of print August 14, 2012, doi:10.1167/iovs.12-10472 (f12)

2) Mutations in paired box gene 6 (PAX6) are the major cause of aniridia that may be associated with several other developmental anomalies of the eye, including microcornea in rare cases. However, systemic evaluation of PAX6 in patients with microcornea as the major sign has not been reported. This study aims to detect PAX6 mutations in patients with microcornea. This study screens PAX6 gene in a microcornea cohort with 35 patients to evaluate the relationship between PAX6 and microcornea. The authors identified two novel and a known mutation of *PAX6* in four probands with microcornea, accounting for 11.4% of microcornea in this cohort. The findings not only expand the spectrum of *PAX6* mutations, but also suggest that *PAX6* mutations may be a common cause of microcornea. The results show that PAX6 may be a common cause of microcornea with/without iris hypoplasia.

Non-syndromic retinal ciliopathies: translating gene discovery into therapy.

Human Molecular Genetics 2012; 1-14. Estrada-Cuzcano A, Roepman R, Cremers FPM, den Hollander AI, Mans DA. (f12)

Synopsis: 158 genes have been discovered to cause retinal degeneration, and about a third of these involve genes related to cilia. The photoreceptor outer segment, connecting cilium and basal body actually form a specialized sensory cilium for vision. The connecting cilium in the photoreceptor is involved in bidirectional transport of proteins needed for

vision, so mutations in genes that regulate the formation, function or maintenance of this structure are likely to cause retinal disease. Many of the ciliopathies are multiorgan syndromes that also cause retinal dysfunction, but at least 21 genes encoding ciliary proteins are known to cause isolated, non-syndromic retinal degenerations. Because of the unique protein interactions in ciliopathies, these retinal degenerations will have special challenges for treatment, which the authors outline in this excellent review article.

Mutations in NMNAT1 cause LCA with early onset severe macular and optic atrophy. Perrault I et al. *Nature Genetics* 2012 July 29 Epub ahead of print. **NMNAT1 mutations cause LCA.** Falk MJ et al. *Nature Genetics* 2012 July 29 Epub ahead of print. **Mutations in NMNAT1 cause LCA and identify a new disease pathway for retinal degeneration.** Koenekoop et al. *Nature Genetics* 2012 July 29 Epub ahead of print. **Exome sequencing identifies NMNAT1 mutations as a cause of LCA.** Chiang et al. **Nature Genetics** 2012 July 29 Epub ahead of print. (f12)

Synopsis: Four papers were published simultaneously reporting the discovery of another gene for LCA. This locus (chromosome location) was previously reported and called LCA9, but the gene was not known. Exome sequencing, in which a person's entire coding DNA is examined, rather than individual genes or locations, allowed several groups to identify it. The inheritance is autosomal recessive. The unique clinical feature of this type of LCA is a macular coloboma-like lesion due to the central retina being devoid of normal tissue. Early onset optic atrophy may also be a feature. Of note, the protein product of the *NMNAT1* gene is a neuro-protective molecule which has been studied in animals with neurodegenerative disorders. This genetic subtype of LCA should be considered in patients who present with a macular coloboma-like lesion.

OCRL localizes to the primary cilium: a new role for cilia in Lowe syndrome. Luo et al. *Human Molecular Genetics* 2012 August 1;21(15): 3333-3344. **The Lowe syndrome protein OCRL1 is involved in primary cilia assembly.** Coon et al. *Human Molecular Genetics* 2012;21:1835-1847. (f12)

Synopsis: Oculocerebral renal syndrome of Lowe, or Lowe syndrome, is an X-linked disorder in which congenital cataracts, congenital glaucoma, mental retardation and kidney dysfunction are present. It is caused by mutations in the OCRL gene. This study used zebrafish to show that OCRL knock-down resulted in defective cilia formation and cilia-related abnormalities. They found that OCRL localizes to the primary cilium of RPE, fibroblasts and kidney tubule cells. Lowe syndrome now joins many other conditions which affect the eye as a ciliopathy.

Real-time ophthalmoscopic findings of intraophthalmic artery chemotherapy in retinoblastoma. Fallaha et al. *Arch Ophthalmol* 2012;130(8):1075-7. (f12)

Synopsis: Superselective intraophthalmic artery chemotherapy is becoming a more common treatment for retinoblastoma, however there can be complications. The authors present real time photographs of a case which shows the effect on the arterioles during the infusion.

Effect of VEGF-targeted antisense gene therapy on retinoblastoma cell line SO-RB50 in vitro and in vivo. Xin et al. *Int J Ophthalmol* 2012;5(4):440-7 . (f12)

Synopsis: Polyamidoamine dendrimers were used to deliver VEGF antisense oligodeoxynucleotides to cultured cells and to a mouse tumor xenograft of retinoblastoma and tumor growth was monitored. The cells in culture had decreased viability and the tumors in the mice decreased in size. Dendrimers are a novel delivery system for genes, and this study shows they can also be used to deliver antisense DNA to negate tumor DNA.

A novel translocation t(11;13)(q21;q14.2) in a child with suprasellar primitive neuroectodermal tumor and retinoblastoma. Huddleston et al. *Ophthalmic Genetics* 2012 Aug 27 Epub ahead of print. (f12)

Synopsis: A child was treated for a suprasellar primitive neuroectodermal (sPNET) tumor at 1 year of age. At age 6 when unilateral retinoblastoma developed a karyotype was performed and was found to affect the retinoblastoma gene on chromosome 13 with a translocation, 46, XX,t(11;13)(q21;q14.2). This translocation and unusual combination has not been reported before and highlights the importance of genetic testing, even in cases of unilateral retinoblastoma.

Parental diet and risk of retinoblastoma resulting from new germline RB1 mutation. Bunin et al. Environ Mol Mutagen 2012;53(6):451-61. (f12)

Synopsis: Parents of 206 bilateral sporadic (new germline mutation) retinoblastoma cases in North America were contacted by phone as well as parents of 233 controls and a food intake survey covering the year

before the child's birth was taken. Fathers' intake of dairy and fruit was associated with decreased risk and intake of cured meats and sweets was associated with increased risk. The odds ratio for fathers eating cured meat was 5.05. There was no association of new germline Rb mutations with maternal food intake, which is in line with the fact that 85% of new germ line RB1 mutations occur on the paternal allele. Further studies would be needed to confirm this result.

Are children born after infertility treatment at increased risk of retinoblastoma? Foix-L'Helias et al. *Hum Reprod* 2012;27(7): 2186-92. (f12)

Synopsis: No! But they found a slightly increased odds ratio with maternal age older than 35 and for women for whom time to pregnancy was greater than 24 months. No data on fathers was presented.

(106) Ruthenium Plaque Therapy (RPT) for Retinoblastoma. Murakami et al. *Int J Radiat Oncol Biol Phys* 2012;84(1):59-65. (f12)

Synopsis: The authors report on 90 eyes of 85 patients treated with RPT. Two patients died of metastatic disease. Radiation complications included retinal detachment in 13.3%, proliferative retinopathy in 6.7%, rubeosis iridis in 2.2%, and PSC cataract in 25.6%. Long term control at 2 years was achieved in 33.7%, and 58.7% retained the eye at 2 years. The authors conclude that "RPT is an effective eye-preserving treatment for retinoblastoma," but these results should be carefully compared to standard therapy.

Long-term medical outcomes in survivors of extra-ocular retinoblastoma: the Memorial Sloan Kettering Cancer Center experience. Friedman et al. *Pediatr Blood Cancer* 2012 August 21 Epub ahead of print. (f12)

Synopsis: 19 survivors of extra-ocular retinoblastoma treated from 1992 to 2009 were studied. Patients received chemotherapy with or without radiotherapy and autologous stem cell transplant. Median follow up was 7.8 years. 79% had hearing loss, 37% had short stature, and 31% had secondary malignancies. Two patients died of secondary malignancies.

Topical ocular sodium 4-phenylbutyrate rescues glaucoma in a myocilin mouse model of primary open-angle glaucoma. Zode et al. *Invest Ophthalmol Vis Sci* 2012;53(3):1557-65. (f12)

Synopsis: In a mouse model of myocilin glaucoma, topical PBA reduced IOP, while IOP was unaffected in normal mice. The mechanism is thought to be a decrease in endoplasmic reticulum stress and less build up of abnormal myocilin in the TM. This could be a new mechanism of treatment for myocilin related glaucoma in humans.

Mutational Analysis of SDCCAG8 in Bardet –Biedl syndrome patients with renal involvement and absent polydactyly. Billingsley et al. *Ophthalmic Genetics* 2012;33(3):150-4. (f12)

Synopsis: BBS is a multi-organ disorder caused by at least 18 different genes. In this paper the authors describe 5 patients from 4 families with BBS caused by mutations in the SDCCAG8 gene. The retinal degeneration in these patients was typical with rod worse than cone disease, however the patients had severe renal disease requiring early transplant, but no polydactyly. This is an important genotype-phenotype study of this heterogeneous disorder.

Exome sequencing identifies mutations in LZTFL1, a BBSome and trafficking regulator, in a family with Bardet-Biedl syndrome with situs inversus and insertional polydactyly. Marion et al. *J Med Genet* 2012 May;49(5):317-21. (f12)

Synopsis: The gene LZTFL1 was recently characterized as a gene important in cilia and functioning of the BBSome. In this study the authors used exome sequencing to identify the gene in a family with a syndrome having features of BBS, and were able to identify LZTFL1 mutations as the cause, making this BBS17. Of note, the family has insertional polydactyly, not post-axial, with the extra digits in between 2 fingers or near the palm of the hand. In addition to retinal degeneration and other features of BBS, these patients also have situs inversus.

In search of triallelism in Bardet-Biedl syndrome. Abu-Safieh et al. Eur J Hum Genet 2012 Apr;20(4):420-7. (f12)

Synopsis: BBS has long been described as an autosomal recessive disorder, but some recent studies have suggested it is triallelic, i.e. that 3, not 2, mutations must be present in some cases or families to cause disease. The authors of the current study examined patients from 29 families with BBS and in every family were able to find 2 mutations in trans in the same BBS gene. They did not find segregation of other disease causing mutations in other BBS genes acting as modifiers of penetrance. Of note, they found a family with BBS9 in which some patients have typical BBS with RP, obesity, polydactyly and other typical features, while other family members have only isolated RP with the same 2 mutations. This well done study is important in that it supports Mendelian autosomal recessive inheritance as the mode of transmission of BBS, but also highlights the extreme heterogeneity of expression, which may be due to modifier genes. It also gives an example of isolated RP caused by a BBS gene.

Ocular manifestations of the autoinflammatory syndromes. Tarabishy et al. *Ophthalmic Genetics* 2012 Aug 27 Epub ahead of print. (f12)

Synopsis: The authors review these rare inherited disorders of immunity characterized by recurrent inflammation which can severely affect the eye. These disorders include cryopyrin-associated periodic syndromes, familial cold autoinflammatory syndrome, Muckle Wells syndrome and periodic fever syndrome. Because these are so rare, they may be overlooked in the differential of inflammatory eye disease, making this an important review.

Tuberous Sclerosis Complex: Genotype/Phenotype Correlation of Retinal Findings. Aronow et al. *Ophthalmology* 2012 May 16 Epub ahead of print. (f12)

Synopsis: Patients with retinal findings in TS are more likely to also have subependymal giant cell astrocytomas, angiolipomas, cognitive impairment, and epilepsy than those without retinal findings. TSC2 mutations are more common in patients with retinal findings.

Genetic variants on chromosome 1q41influence ocular axial length and high myopia. Fan et al. *PLoS Genet* 2012 June; 8(6)e1002753. (f12)

Synopsis: Myopia is an important cause of vision loss especially in Asia. Myopia clearly has genetic underpinnings. This study is a meta-analysis of 3 genome wide association studies of large numbers of Asian patients with high myopia. A locus on 1q41 was identified and found to harbor ZC3H11B. ZC3H11B and 2 neighboring genes are expressed in retina, RPE and sclera, making them excellent candidates for having a role in myopia. This study adds to the search for the genetic causes of this complex ocular disorder.

Gene delivery to mitochondria by targeting modified adenoassociated virus suppresses LHON in a mouse model. Yu et al. *Proc Natl Acad Sci USA* 2012 May;109(20):E1238-47. (f12)

Synopsis: In this exciting study the authors were able to introduce a normal ND4 gene into the mitochondrial genome by gene therapy. They show that in cells with the 11778 mutation it restored ATP synthesis. In a mouse model of LHON it was able to suppress the vision loss and optic atrophy seen in this model. This is very hopeful for the possibility of gene therapy in LHON, a cause of catastrophic vision loss in young adults, and in other mitochondrial disorders as well.

AAV-mediated cone rescue in a naturally occurring mouse model of CNGA3-achromatopsia. Pang et al. *PLoS One* 2012 April;7(4):e35250. (f12)

Synopsis: Achromatopsia is a debilitating disorder in which severe cone dysfunction causes reduced visual acuity, colorblindness and photophobia. About 1 in 4 cases is caused by mutations in CNGA3. In a mouse model of this disorder the authors used AAV5 to deliver a normal copy of the gene to the retina. They saw a rescue of the cone ERG, improved visual acuity, and normal expression and localization of Ma dn S opsins. The results were stable for at least 5 months after treatment. This study is the first step in developing a gene therapy trial for humans with achromatopsia.

Recommendations for Genetic Testing of Inherited Eye Diseases: Report of the American Academy of Ophthalmology Task Force on Genetic Testing.

Stone EM, Aldave AJ, Drack AV, Maccumber MW, Sheffield VC, Traboulsi E, Weleber RG. *Ophthalmology*. 2012 Aug 31. [Epub ahead of print] (f12)

Synopsis/Abstract: Genetic testing can make a very positive impact on individuals and families affected with inherited eye disease in a number of ways. When properly performed, interpreted, and acted on, genetic tests can improve the accuracy of diagnoses and prognoses, can improve the accuracy of genetic counseling, can reduce the risk of disease occurrence or recurrence in families at risk, and can facilitate the development and delivery of mechanism-specific care. However, like all medical interventions, genetic testing has some specific risks that vary from patient to patient. For example, the results of a genetic test can affect a patient's plans to have children, can create a sense of anxiety or guilt, and can even perturb a patient's relationships with other family members. For these reasons, skilled counseling should be provided to all individuals who undergo genetic testing to maximize the benefits and minimize the risks associated with each test.

[Home]

RETINOBLASTOMA

Outcome of Children With Retinoblastoma and Isolated Choroidal Invasion.

Andrea Bosaleh, MD; Claudia Sampor, MD; Verónica Solernou, MD; et al. . Arch Ophthalmol. 2012;130(6):724-729 (f12)

This paper's goal was to evaluate the outcome of children with different degrees of choroidal invasion, to compare different systems for grading the extent of choroidal invasion, and to assess the role of concomitant prelaminar optic nerve and anterior segment invasion as predictors of extraocular relapse. This was a retrospective analysis of children included in 4 prospective protocols (January 1, 1989, through June 31, 2010). Children with postlaminar optic nerve or scleral involvement and overt extraocular disease were excluded. The probability of 5-year event-free survival was 98.1% and the probability of overall survival was 98.7% because 1 patient relapsed. Children with massive invasion had a significantly lower event-free survival probability (94.2%) compared with those with focal invasion (99.2%) (P = .04). However, no significant difference was found in overall survival probability (98.7% vs 99.2%; P = .29). No significant effect of other risk factors was found. The authors concluded that survival was excellent without adjuvant therapy, and no other factors correlated with survival. Children with massive invasion have a higher relapse rate but comparable survival to those with focal invasion provided that aggressive therapy for extraocular relapse is available with adequate safety conditions.

Intravenous Chemoreduction or Intra-arterial Chemotherapy for Cavitary Retinoblastoma Long-term Results

Duangnate Rojanaporn, MD; Swathi Kaliki, MD; Carlos G. Bianciotto, MD, et al Arch Ophthalmol. 2012;130(5):585-590 (f12)

The goal of this study was to assess the long-term results of chemotherapy for cavitary retinoblastoma. This was a retrospective, nonrandomized, interventional case series of 26 cavitary retinoblastomas in 25 eyes of 24 patients. Retinoblastomas were treated with intravenous chemoreduction and/or intra-arterial chemotherapy. Of 24 patients with cavitary retinoblastoma, the mean age at diagnosis was 16 months. The mean number of cavitary tumors per eye was 1 (median, 1; range, 1-2), with a mean tumor basal diameter of 13 (median, 13; range, 7-24) mm and mean tumor thickness of 7 (median, 6; range, 3-17) mm. The mean number of cavities per tumor was 2 (median, 2; range, 1-5), with a mean cavity diameter of 3 (median, 2; range, 1-10) mm. Related features included vitreous seeds in 7 tumors (27%), subretinal seeds in 6 (23%), and subretinal fluid in 13 (50%). Intravenous chemoreduction was used in 23 tumors (88%); intra-arterial chemotherapy, in 2 (8%); and both, in 1 (4%). After treatment, the mean reduction in tumor base was 22% and mean reduction in tumor thickness was 29%. Despite minimal reduction, tumor recurrence was noted in only 1 eye (4%), globe salvage was achieved in 22 (88%), and there were no cases of metastasis or death during 49 (range, 6-189) months of follow-up. Despite minimal visible tumor response to chemotherapy, cavitary retinoblastoma displays relatively stable long-term results.

Outcome, Pathologic Findings, and Compliance in Orbital Retinoblastoma (International Retinoblastoma Staging System Stage III) Treated with Neoadjuvant Chemotherapy
A Prospective Study

Venkatraman Radhakrishnan, MD, Seema Kashyap, MD, Neelam Pushker, MS, et al
Ophthalmology July 2012; 119:1470-1477 (f12)

This is a prospective study and case series involving 28 consecutive international retinoblastoma staging system stage III (IRSS) retinoblastoma patients. The patients were enrolled prospectively. Planned therapy for patients included treatment with a uniform protocol consisting of neoadjuvant chemotherapy followed by enucleation, adjuvant radiotherapy, and chemotherapy.

Main outcome measures were event-free survival (EFS) and overall survival (OS) using Kaplan-Meier survival analysis. This study shows that neoadjuvant chemotherapy was able to avoid exenteration in all operated patients; however, residual viable tumor was present in 95% of enucleated specimens. Bilaterality

and tumor presence in the optic cut end after neoadjuvant chemotherapy were associated with inferior outcome.

This study was performed at the Department of Medical Oncology, Department of Ocular Pathology, Department of Ophthalmology, and Department of Radiodiagnosis, All India Institute of Medical Sciences, New Delhi, India
COMMENT: The authors feel that neoadjuvant chemotherapy prevents orbital exenteration in most cases. Central nervous metastasis was the most common site of relapse and death. The overall rate of compliance with treatment was 67.8%. Adjuvant chemotherapy consisted of Vincristine, carboplatin on day 1 and Etoposide on days 1 and 2 every 4 weeks. This was followed by enucleation or exenteration of the involved eye, external beam radiotherapy to the involved eye, and adjuvant vincristine and carboplatin chemotherapy.

Fluorescein Angiographic Findings after Intra-arterial Chemotherapy for Retinoblastoma

Carlos Bianciotto, MD, Carol L Shields, MD, Juan C Iturralde, MD, Ahmet Sarici, MD, Pascal Jabbour, MD, Jerry A Shields, MD
Ophthalmology April, 2012: 119:843-849 (f12)

This is a retrospective case series of 24 eyes of 24 patients designed to evaluate fluorescein angiography findings after intra-arterial chemotherapy (IAC) for retinoblastoma.
Fifty-five, intra-arterial chemotherapy procedures for delivery of melphalan 5mg and possible carboplatin 30mg were discussed. The main outcome measures were vascular flow of the iris, retina and choroids after intra-arterial chemotherapy (IAC). Fluorescein angiography suggests that vascular perfusion to the retina and choroids can be compromised after IAC for retinoblastoma. “The most common vascular abnormality was choroidal sector or diffuse nonperfusion.”
This study was performed by the Ocular Oncology Service, Wills Eye Institute, Philadelphia, Pennsylvania.
COMMENT: There are excellent photographs on page 846 showing choroidal nonperfusion in patients treated with IAC.

Ultrasound biomicroscopy evaluation of anterior extension in retinoblastoma:
a clinicopathological study

Alexandre P Moulin, Marie-Claire Gaillard, Aubin Balmer, Francis L Munier
Br J Ophthalmol 2012;96:337-340(f12)

Aim: To determine the value of ultrasound biomicroscopy (UBM) in the assessment of posterior chamber involvement in advanced retinoblastoma The sensitivity of UBM in the assessment of posterior chamber invasion by retinoblastoma was 81% and the specificity was 100%.

Conclusion: In selected cases of advanced retinoblastoma, UBM appears to represent a valuable tool in the precise evaluation of anterior extension of disease, with good sensitivity and specificity for the assessment of posterior chamber involvement. UBM
may provide useful criteria governing the indication for enucleation.

Intra-arterial chemotherapy for retinoblastoma in eyes with vitreous and/or subretinal seeding: 2-year results David H Abramson, Brian P Marr, Ira J Dunkel, Scott Brodie, Emily C Zabor, Sarah J Driscoll, Y Pierre Gobin

Br J Ophthalmol 2012;96:499-502. (f12)

Background/aims: To review the effectiveness of intraarterial chemotherapy for advanced intra-ocular retinoblastoma with vitreous and/or subretinal seeds in naive (untreated) and previously treated eyes.

Methods: 76 eyes of 67 patients with retinoblastoma with subretinal and/or vitreous seeding treated with intra-arterial chemotherapy
Results: Despite advanced intraocular disease with seeding, the majority (56/76) of eyes were saved; 20/76 eyes were enucleated. Among treatment-naive eyes, the 2-year probability of ocular salvage was 83% (95% CI 27% to 97%) for eyes with subretinal seeding only, 64% (95% CI 24% to 87%) for eyes with vitreous seeding only, and 80% (95% CI 40% to 95%) for eyes with both. Among eyes that received previous treatment and had progressed, the 2-year probability of ocular salvage was 50% (95% CI 15% to 78%) for eyes with only subretinal seeding, 76% (95% CI 48% to 91%) for eyes with vitreous seeding only, and 54% (95% CI 20% to 79%) for eyes with both. Nine of 29 naive eyes (31%) were cured with intra-arterial (super-selective ophthalmic artery infusion of chemotherapy) chemotherapy alone.
Conclusion: Unlike radiation or systemic chemotherapy, intra-arterial chemotherapy can usually prevent the need for enucleation in naive eyes with advanced intraocular retinoblastoma with seedingdespecially if the seeding is subretinal.

Comments: Vitreous and subretinal seeding remain a challenge for clinicians treating intraocular retinoblastoma, but the success of intra-arterial chemotherapy in salvaging eyes (without the need for external beam irradiation) is encouraging.

Combined intravitreal and subconjunctival carboplatin for retinoblastoma with vitreous seeds

Stephen J Smith, Jose S Pulido, Diva R Saloma, Brian D Smith, Brian MohneyBr J Ophthalmol 2012;96:1073-1077. (f12)

Background: To describe the technique of intravitreal chemotherapy preceded by subconjunctival chemotherapy for the treatment of vitreous seeds in advanced stage retinoblastoma.

Methods: This non-comparative interventional case series retrospectively reviewed the medical records and postenucleation histopathological findings of two patients who presented within weeks of each other with bilateral retinoblastoma, ReeseeEllsworth (R-E) stage Vb in the worse eye. Both patients had failed systemic chemotherapy prior to receiving a single treatment of 0.5 ml (5 mg per 0.5 ml) of subconjunctival carboplatin,
through which 0.05 ml (3 mcg per 0.05 ml) of carboplatin was injected into the vitreous (Case 2 received 0.1 ml of intravitreal carboplatin). The subconjunctival chemotherapy was given to reduce the risk of orbital tumour seeding following intravitreal injection. Following enucleation, ocular toxicity and the presence or absence of viable tumour cells at the intravitreal injection site were recorded.

Results: Histopathological examination did not reveal patency of the pars plana intravitreal penetration site in either case at 6 weeks post-treatment nor was malignant seeding detected in the area of injection.Examination of the two enucleated eyes did not demonstrate structural toxicity to the cornea, anterior segment, iris or retina. Additionally, both cases were followed for over 37 months post-treatment, without the
occurrence of orbital malignancy.
Conclusions: Injecting a bleb of subconjunctival chemotherapy prior to intravitreal drug delivery appeared to mitigate the risk of orbital tumour seeding in two patients with advanced stage retinoblastoma. Incorporating this technique may allow further
investigation of intravitreal chemotherapy for the treatment of vitreous seeds in retinoblastoma.

Comments: While a two patient case series is insufficient to conclusively state that the technique of subconjunctival chemotherapy prior to intravitreal injection eliminates the risk of tumour seeding, the results of past studies indicate that this risk may be less than previously believed, and the addition of subconjunctival treatment as described in this report may enable further investigation of intravitreal treatment.

Intravitreal chemotherapy for vitreous disease in retinoblastoma revisited: from prohibition to conditional indications. Francis L Munier, Marie-Claire Gaillard, Aubin Balmer, Sameh Soliman, Gregory Podilsky, Alexandre P Moulin, Maja Beck-Popovic.Br J Ophthalmol 2012;96:1078-1083 (f12)

Background: Tumour control of vitreous seeds remains challenging owing to their resistance to radiation and systemic chemotherapy. Objective To describe the short-term efficacy of intravitreal melphalan for vitreous disease in retinoblastoma using a new injection technique and dose.

Methods: This study is a retrospective non-comparative review of 23 consecutive heavily pretreated patients(23 eyes) with active vitreous seeding and eligible for intravitreous chemotherapy (IViC). They received a total of 122 intravitreal injections of melphalan (20-30 mg) given every 7-10 days. The ocular status was objectively monitored under anaesthesia with fundus photography.

Results: All patients are alive without evidence of extraocular spread (95% CI 82.19% to 100%). Concomitant treatments, including other chemotherapeutic modalities, were used until complete sterilisation of the retinal seeding source and subretinal seeds. Globe retention was achieved in 87% (20/23) of cases. All retained eyes were in complete remission after a median follow-up period of 22 months (range 9-31 months). The Kaplane Meier estimate of ocular survival rates at 2 years was 84.14% (95% CI 62.48% to 95.28%). A localised peripheral salt-and-pepper retinopathy was noted in 10 eyes (43%) at the site of injection.

Conclusions: This study reports the first clinically documented case series of patients with retinoblastoma treated with IViC. Of note, none of the treated eyes required external beam irradiation to control the vitreous seeding. Further studies are required to assess IViC retinal toxicity and to better delineate its role in the management of retinoblastoma.

Comments: Although IViC appears to offer a safe and efficient salvage option, its validation awaits the results of a prospective phase II clinical trial. Special attention will be paid to retinal toxicity assessed by electroretinogram, fluorescein angiography and optic coherence tomography. If validated, IViC may prove to be useful as salvage treatment for recurrent or resistant vitreous seeds, and also useful as a prophylactic measure in cases of iatrogenic seeding after photocoagulation and plaque surgery, or secondline treatment for group B eyes with ruptured internal limiting membrane (as assessed by fluorescein angiography).

First-Year Experience of Chemotherapy for Advanced Retinoblastoma in Tanzania: Disease Profile, Outcomes, and Challenges in 2008

Zia I. Carrim, MRCOphth; Jane Kajaige, AMO; Richard J. Bowman, FRCOphth; Tim E. Lavy, FRCOphth; Patricia Scanlan, MRCPI (paeds) J Pediatr Opthalmol Strabismus 2012; 49:176-183 May/June (f12)

Mortality associated with retinoblastoma may seem like a rare entity these days in North America (5% in Europe and North America); however, if we were to look at the African continent, you would find this number close to 70%. Until 2007, the only treatment modalities were surgery and external beam radiation treatment. In 2007, chemotherapy was offered as an option due to funding at one center in Tanzania. This study summarizes results of chemotherapy in 2008 for ocular and orbital disease. Although chemotherapy offered some benefit, many other factor proved to contribute to the increased mortality rate other than treatment offered. The article discusses these other factors as well and offers possible solutions.

EFFECT OF INTRAARTERIAL CHEMOTHERAPY ON RETINOBLASTOMA-INDUCED RETINAL DETACHMENT

Shields, Carol L. MD[*]; Kaliki, Swathi MD[*]; Shah, Sanket U. MD[*]; Bianciotto, Carlos G. MD[*]; Jabbour, Pascal MD[†]; Shields, Jerry A. MD Retina. March 2012; 32(4):799-804

Most eyes with total retinal detachment from retinoblastoma (RB) have traditionally been considered to be nonsalvageable and have been managed with enucleation. With advances in chemotherapy, three-drug intravenous chemoreduction has been shown effective for RB control and successful in bringing about resolution of tumor-related retinal detachment. In more recent years, some interest has focused on localized intraarterial chemotherapy (IAC) (ophthalmic artery) in an effort to control the tumor and avoid systemic toxicities. However, the effect of IAC on eyes with total or partial retinal detachment has not been studied in detail and this study looks at that. It is a retrospective, noncomparative, interventional case series including 15 patients, with intraarterial (ophthalmic artery) chemotherapy as the intervention. Resolution of retinal detachment was the main outcome measure.

Of 15 patients with RB induced retinal detachment managed with IAC, 7 (47%) presented with total retinal detachment and 8 (53%) with partial detachment. The eyes were classified as International Classification of Retinoblastoma group C (n = 2, 13%), group D (n = 4, 27%), and group E (n = 5, 33%). There were 4 eyes (27%) that received IAC as secondary treatment after the failure of other treatment methods. After IAC, all tumors showed regression. Of the 7 eyes with

initial total retinal detachment, complete resolution of fluid was found in 3 (43%) and partial resolution in 4 (57%). Of the 8 eyes with initial partial retinal detachment, complete resolution of subretinal fluid was found in all 8 (100%), but 1 eye later developed recurrent subretinal fluid. The mean interval to complete resolution of subretinal fluid was 2 months. Globe salvage was achieved in 5 of 7 eyes (71%) with total retinal detachment and 6 of 8 eyes (75%) with partial retinal detachment. The authors concluded that RB induced retinal detachment showed complete resolution of detachment after IAC in 43% (total RD) and 100% (partial RD).

Intravenous and intra-arterial chemotherapy for retinoblastoma: what have we learned?

Shields, Carol L. Kaliki, Swathi; Rojanaporn, Duangnate; Al-Dahmash, Saad; Bianciotto, Carlos G. Shields, Jerry A.

Current Opinion in Ophthalmology. May 2012; 23(3):202-209

This is a literature review of chemotherapy for retinoblastoma (RB) and compares the two current delivery methods, intravenous and intra-arterial.

In 1996, intravenous chemotherapy (chemoreduction) for RB was introduced and published studies showed impressive tumor control, without the need for external beam radiotherapy or enucleation. Later reports showed continued impressive long-term control, minimal systemic toxicities, likely prevention of pinealoblastoma (trilateral RB), and reduction in numbers of germline mutation second cancers. There is no reported ophthalmic toxicity and no evidence of reduction in fertility with chemoreduction. In 2011, intra-arterial chemotherapy was introduced as way to deliver targeted therapy and avoid systemic toxicity of chemoreduction. To date there have been several studies and conflicting editorials in the literature. This technique requires a catheterization from the femoral artery into the ophthalmic artery. Outstanding tumor control is achieved with only three cycles. However, ocular ischemic events have been noted. Further improvements in this technique could minimize complications.

Both intravenous and intra-arterial chemotherapy are powerful methods for RB control. Intra-arterial chemotherapy provides excellent tumor control for slightly more advanced eyes with RB and, in addition, can be used to treat eyes that fail other methods. However, local ocular toxicities can be vision-threatening and long-term systemic toxicities are not yet understood.

US and MRI of pediatric ocular masses with histopathological correlation

Rachel C. Brennan & Matthew W. Wilson & Sue Kaste &

Kathleen J. Helton & M. Beth McCarville Pediatric Radiology. June 2012; 42(6): 738-749

Pediatric ocular masses pose a challenging diagnostic dilemma. This is a nice review article of the differential diagnosis of leukocoria with clinical and radiographic illustrations of each. In addition, distinctive clinical and radiographic features of each entity are highlighted to assist in making the correct diagnosis.

The US and MRI appearances of the normal eye are reviewed. In general, US is useful to delineate areas of calcification within intraocular lesions, identify tissue interfaces and
reveal patterns of vascularity using Doppler technology. MRI is less sensitive than US for detecting calcification but more sensitive to tumor extension into the optic nerve and subarachnoid spaces. Therefore, these modalities are complementary and both are used routinely for intraocular tumor diagnosis. CT scans are not indicated for assessment of intraocular tumors due to exposure to ionizing radiation and the association of radiation-induced secondary malignant neoplasms in children with hereditary retinoblastoma. In modern-day practice, the combination of US, MRI and fundoscopy obviates the need for CT in the setting of pediatric ocular tumors.

[Home]

TRAUMA

Ocular Trauma Score as a predictor of final visual outcomes in traumatic cataract cases in pediatric patients.

Shah MA, Shah SM, Applewar A, Patel C, Patel K.
J Cataract Refract Surg. Jun 2012;38(6):959-65. (f12)

The authors studied 354 pediatric traumatic cataracts (children ages 0-18 years) in India. The patients underwent open globe repair, cataract removal, amblyopia and strabismus treatment (when necessary), and had determination of corrected distance visual acuity after rehabilitation. The authors were interested in whether the ocular trauma score (OTS) predicted final visual acuity.

The authors concluded that the OTS predicts final visual acuity in pediatric traumatic cataracts. Eyes with a low OTS (= poor score) are likely to have poor final visual acuity. Understanding the manuscript required reading the original OTS publication.1 The OTS is determined primarily by visual acuity at presentation, with better visual acuity giving a higher OTS. Points are then taken off for rupture, endophthalmitis, perforation, retinal detachment, or afferent pupillary defect. The conclusion that the OTS predicts final visual acuity in

pediatric traumatic cataracts may be similar to concluding that initial visual acuity predicts final visual acuity. However, it's not a tightly correlated relationship, and OTS scores of 3 and 2, which most patients fell into, showed a wide range of final visual acuities, ranging from > 20/40 to LP and NLP.

The manuscript provides descriptive information about pediatric traumatic cataracts. Older boys ages 11-18 years are most commonly affected. In this population, the most common cause was injury by a wooden stick (51%) and occurred most often during play (55%). Open globe injuries (83%) were more common than closed globe injuries (17%), and open globe injuries were associated with better final visual acuity.

Ocular Trauma Score: A Useful Predictor of Visual Outcome at Six Weeks in Patients with Traumatic Cataract

Mehul A Shah, MD, Shreya M Shah, MD, Adway Applewar, MD, et al
Ophthalmology July 2012; 119:1336-1341 (f12)

This is a retrospective cohort study of 787 eyes. This study was designed to validate predictive value of the Ocular Trauma Score (OTS) in injury cases with traumatic cataracts. This was performed in January 2003 to December 2009. Cataracts were classified as corneal lenticular opacity. A total of 787 eyes of 787 subjects with traumatic cataracts were enrolled using specific inclusion criteria. The eyes were examined to review comorbidities, such as trauma. Surgery was performed for traumatic cataracts, lenses were implanted, and patients were treated for amblyopia as applicable. Patients were evaluated 6 weeks postoperatively. They were evaluated based on the Birmingham Eye Trauma Terminology System (BETTS). The patients were divided into open globe injury and closed globe injury classification.
At 6 weeks postoperatively 245 eyes (31%) had a visual acuity greater than 20/140 and 480 eyes (61%) had a visual acuity greater than 20/200. The Ocular Trauma Score prediction was not significantly different when compared with actual visual outcome at 6 weeks postoperatively in all OTS categories. The Ocular Trauma Score was found to be a reliable tool to predict visual outcome in cases of traumatic cataract 6 weeks postoperatively.

This study was performed in Drashti Netralaya, Dahod, Gujarat, India.

(1) Kuhn, F, Maisiak, R, Mann, L, et al. (2002). The Ocular Trauma Score (OTS). Ophthalmology clinics of North America, 15(2), 163-5, vi.

Periorbital infections after Dermabond closure of traumatic lacerations in three children

. Yeilding RH, O'Day DM, Li C, et al. J AAPOS 2012;16:168-172. (f12)

The authors describe cases of periorbital cellulitis after closure of traumatic periorbital lacerations with 2-octyl cyanoacrylate (Dermabond). Three patients presented over a four week period to three different emergency rooms. A literature review shows a 1.8% rate of infection with tissue adhesive wound closure compared with a 0.3% infection rate with standard wound closure (not a statistically significant difference). Cultures grew Streptococcus pyogenes in 2 of the 3 cases and 2 cases required surgical intervention. The authors comment that adequate skin preparation of the wounds may not have occurred, due to poor patient cooperation because of age, or because the clinician was unaware this was necessary. Dermabond use may shorten the time physicians spend sterilizing a wound versus traditional closure. Wounds also may have become contaminated after closure. One of the three cases involved necrotizing fasciitis, which has not previously been reported with Dermabond use.

Twelve-Year Review of Pediatric Traumatic Open Globe Injuries in an Urban U.S. Population

Sebastian P. Lesniak, MD; Alain Bauza, BS; Jung H. Son, BS; Marco A Zarbin, MD, PhD; Paul Langer, MD; Suquin Guo, MD; Rudolph S. Wagner, MD; Neelakshi Bhagat, MD J Pediatr Opthalmol Strabismus 2012; 49:73-79 March/ April

The purpose of the article is to characterize the demographics and clinical features of pediatric open globe injuries in an urban population in the United States. In addition, it compares final vision to the vision predicted by the ocular trauma score, an eye trauma classification system. Results show the various types of injuries and complications even after repair of the initial injury. The final and predicted vision were found to be comparable. The article serves as a good reference for the types of injuries that cause open globe trauma in the pediatric population.

[Home]

ANTERIOR SEGMENT

Scalability and Severity of Keratoconus in Children

Leoni-Mesplie S, Mortemousque B, Touboul D, Malet F, Praud D, Mesplie N, Colin J. Am J Ophthalmol 2012; 154(1):56-62. (f12)

This retrospective study was performed to assess the severity of keratoconus at diagnosis and its scalability over a period of 2 years in children compared to adults. In total, 216 patients were studied, comprising 49 patients (22.7%) aged ≤15 and 167 patients (77.3%) aged ≥27 years at diagnosis, who were seen

within 2 years of diagnosis. Severity at diagnosis was assessed using Krumeich's classification, and the scalability criteria of the US Food and Drug Administration (2010) were used. Student t tests and $\chi 2$ tests were performed to compare the 2 groups. Keratoconus in children was significantly more severe at diagnosis, with 27.8% being stage 4 vs 7.8% of adults ($P < .0001$). In addition, ophthalmoscopic signs were more frequent in children (42.9% vs 29.5%, $P = .05$), while mean values of maximum, average, and minimum keratometry as well as simulated keratometric astigmatism were higher ($P < .0001$, $P = .0002$, $P = .0005$, and $P = .001$, respectively). After diagnosis, keratoconus did not evolve more frequently in children. However, in the case of progression, keratoconus evolved faster in children, with significant differences in the spherical equivalent and maximum and minimum keratometry ($P = .03$, $P = .02$, $P = .04$, respectively). The authors concluded that at the time of diagnosis, keratoconus is often more advanced in children than in adults, with faster disease progression. Early detection and close monitoring are therefore crucial in young patients.

Fixation and its role in the causation, laterality and location of pterygium: a study in amblyopes and non-amblyopes

A Sudhalkar

Eye. March 2012;26:438-443 (f12)

This study evaluated the laterality (right versus left versus both eyes) and location (nasal versus temporal) of pterygia in relation to fixation. This was a prospective, observational, case–control study. Cases were defined as patients with primary pterygium who had unilateral amblyopia with eccentric fixation. Controls were age-matched patients with primary pterygium, but without amblyopia and without eccentric fixation. All patients underwent complete orthoptic evaluation. The mean age of subjects was 47.1±5.25 years in cases (n=107) and 48.2±4.75 years in controls (n=310; P=0.78). The groups came from the same geographic area and were similar in terms of ultraviolet light exposure.

All pterygia were nasal. Among the cases, 88 (82.2%) patients demonstrated suppression of the amblyopic eye and all of these patients had pterygium only in the fixing eye. 19 (17.8%) patients had abnormal retinal correspondence (ARC) and all of these patients had bilateral pterygia.

Among the controls, 192 (61.9%) eyes had bilateral pterygia and 118 (38.1%) eyes had unilateral pterygium. In controls, pterygium was strongly associated with eye dominance, with the dominant eye being affected first in 85% of bilateral pterygia, and the dominant eye affected in 83% of unilateral pterygia.

The author concludes that fixation appears to be important role in causing pterygium and determining its location and laterality. The author is not describing pterygium to be associated with strabismus.

I have trouble understanding and believing these study results. I don't understand why pathophysiologically there would be any relationship between pterygia and foveal versus nonfoveal fixation. Equally puzzling would be a relationship between pterygium laterality and eye dominance in a nonstrabismic patient. Why would pterygium develop in both eyes if there is ARC, but not develop in amblyopic eyes with eccentric fixation, and would this relationship hold true for **all** patients? Perhaps there is a confounder at play in this study, such as angle of strabismus (which could influence ultraviolet light exposure or tear film / lid interactions). Perhaps the study was biased: the study examiner was masked to the objective of the study, but I wonder how rigorously.

Tofacitinim (CP-690,550), a Janus Kinase Inhibitor for Dry Eye Disease

Results from a Phase ½ Trial
Shiao Hui (Melissa) Liew, MBBS, MRCOphth, Kelly K Nichols, OD, PhD, Karen J Klamerus, PharmD
Ophthalmology July 2012; 119:1328-1335 (f12)

This is a study designed to evaluate the safety and efficacy of topical ophthalmic Tofacitinib (CP-690,550), a novel Janus kinase inhibitor, in treating dry eye disease (DED). This is a randomized, double-masked, multicenter study. Three hundred and twenty-seven patients 18 years of age and older with dry eye disease was diagnosed for 6 months or more. In the United States topical cyclosporin ophthalmic emulsion (0.05%, Restasis) and a delayed-release lubricant (Lacrisert) is available. Tofacitinib (CP-690, 550) is a potent, selective inhibitor of the Janus kinase (JAK) family. Signaling is critical for immune cell activation, proinflammatory cytokine production, and cytokine signaling. Tofacitinib inhibits JAK1, JAK2, and JAK3 invitro with functional cellular selectivity for JAK1 and JAK3 greater than JAK2. Inhibition of JAK1 and JAK3 by Tofacitinib blocks signaling to the common gamma chain containing receptors for several cytokines. Tofacitinib modulates adaptive immunity with limited affects on hematopoiesis. It has also demonstrated efficacy as oral and topical nonophthalmic formulations in the treatment of other inflammatory autoimmune diseases, such as rheumatoid arthritis, psoriasis, and transplant rejection.

This was a 2 stage; phase 1/2 prospective randomized double-masked, multicenter study in patients with dry eye disease. This study demonstrated a trend for improving both signs and symptoms of dry eye. All doses of Tofacitinib exhibit a reasonably safe profile and were well tolerated by patients with dry eye disease.

This study was performed at the University of Houston College of Optometry, Houston, Texas, under the auspice of Pfizer Inc., Collegeville, Pennsylvania, and Department of Ophthalmology and Visual Sciences, University of Louisville School of Medicine, Louisville, Kentucky.

Association of Single Nucleotide Polymorphisms of Interleukins-1B, -6, and –12B with Contact Lens Keratisi Susceptibility and Severity

Nicole A carnt, B. Optom, Mark D.P. Willcox, PhD, Scott Hau, MSc, MCOptom, et al
Ophthalmology July 2012; 119:1320-1327 (f12)

This is a retrospective case control study presented at Association for Research and Vision Ophthalmology, Fort Lauderdale, May 2011. One hundred and twelve cases with keratitis and 205 controls were recruited for the studies conducted at Moorfileds Eye Hospital and in Australia during 2003 through 2005. Buccal swab samples were collected on Whatman FTA cards and mailed by post for analysis of interleukin genotypes.
Relative risk of developing contact lens-related keratitis and more severe forms of the disease based on allele, genotype, and haplotype associations. Single nucleotide polymorphisms of interleukin IL-6 are known to reduce protein expression of the cytokine and thus ocular immune defense systems and carriers of these single nucleotide polymorphisms (SNP) were more likely to experience more severe and microbial keratitis. This suggests that IL-6 decreases in severity and susceptibility of contact lens related keratitis.

This study was performed at School of Optometry and Vision Science, University of New South Wales, Sydney, Australia, and Moorfields Eye Hospital NHS Foundation Trust, London, UK, and the Institute of Ophthalmology, University College, London, UK.
COMMENT: Single nucleotide polymorphisms of interleukin (IL-6) were involved in pro-inflammatory pathway of inflammatory keratitis. Regulation of these cytokines may lead to new therapeutic modalities and treatment of inflammatory microbial keratitis.

Congenital Fibrovascular Pupillary Membranes: Clinical and Histopathologic Findings

Scott R Lambert, MD, Edward Buckley,MD, Phoebe Lenhart,MD, Qing Zhang,MD, Hans E. Grossiniklaus, MD
Ophthalmology March, 2012; 119:634-641 (f12)

This study reports a case series describing the clinical and histopathologic findings associated with congenital fibrovascular pupillary membranes.
Seven infants were reported, six with a unilateral congenital pupillary membrane and one with “classic” persistent fetal vasculature (PFV)
All patients underwent anterior segment surgery (lensectomy, pupilloplasty, or membranectomy) and histopathologic examination was performed on the excised membranes.

Sixty six percent (4/6 patients),of the operated eyes, required repeat surgery for recurrent membranes. The vision was described as “excellent” in 4/6 patients. The two patients that did not require repeat surgery had multiple spincterotomies performed at the time of the original surgery.
Histopathologic evaluation showed the presence of smooth muscle actin only in the blood vessel walls of pupillary membranes. In the single eye with persistent fetal vasculature and in reoperated eyes, collagenized fibrovascular tissue that was immunoreactive for smooth muscle actin was present.
The authors feel that congenital fibrovascular pupillary membranes in infants are likely to be a variant of PFV that may recur if incompletely excised. Iris spincterotomies and broad excision of the pupillary membrane, performed at the time of initial surgery, may prevent the need for repeat surgery.

The study was carried out at the Departments of Ophthalmology at Emory and Duke Universities.
COMMENT: Excellent anterior segment color photos and histopathologic sections.

Use of the Boston Ocular Surface Prosthesis (BOSP) in the Management of Severe Periorbital Thermal Injuries: A Case Series of 10 Patients

Kevin Kalweriskey, MD, Bret Davies, MD, Lisa Mihora, MD, Craig N. Czyz, DO, Jill A. Foster, MD, Sheri DeMartelaere, MD
Ophthalmology March, 2012; 119: 516-521 (f12)

This article reports 10 cases of severe ocular and periorbital burns that required use of the Boston Ocular Surface Prosthesis.
The article has an excellent description and photographs of the BOSP and how it fits over the eye like a large, rigid, scleral contact lens. The prosthesis has an optical zone, transition zone and haptic zone that helps kept the prosthesis in place above the surface of the cornea. There is a liquid reservoir between the prosthesis and the injured cornea. This lens would have value in preventing symblepharon formation in Stevens-Johnson and burn patients.
Study performed at the San Antonio Uniformed Services Health Education Consortium in San Antonio Texas
COMMENT: Excellent pictures. Most cases involved third degree burns to all eyelids with associated exposure keratopathy.

Comparison between Aniridia with and without PAX6 Mutations

Clinical and Molecular Analysis in 14 Korean Patients with Aniridia
Hyun Taek Lim, MD, PhD, Eul-Ju Seo, MD, PhD, Gu-Hwan Kim, PhD, Hyosook Ahn, MD, PhD, et. al.

Ophthalmology June, 2012; 119:1258-1264 (f12)

This is a comparative case series of 14 Korean patients from 10 families with aniridia.

The purpose of this study was to describe clinical molecular characteristics of Korean patients with aniridia and to compare the clinical phenotype between those having an identifiable PAX6 mutation and those not. The main outcome measures were severity of ocular abnormalities and genetic findings. This study has 2 novel PAX6 mutations to those previously reported, providing further evidence for genetic and phenotypic heterogeneity in aniridic ocular malformation. There is no difference in the clinical phenotype between patients with and without detectable mutations in the PAX6 gene. The wide variability of ocular phenotype regardless of the presence or absence of PAX6 mutations calls for a further appreciation of the complexity of molecular diagnosis of aniridia and suggests that this ocular malformation may be better regarded as a group of heterogeneous disorders rather than a single disease entity. There may be associated mutations in PAX6 and/or other genes located elsewhere in the human genome.

This study was performed in the Department of Ophthalmology, Department of Laboratory Medicine and Department of Pediatrics at the Asan Medical Center Children's Hospital, University of Ulsan College of Medicine, Seoul, Korea.
COMMENT: This article has an excellent analysis of genomic molecular karyotyping and candidate gene sequencing. The PITX3 and FOXE3 genes were sequenced as a candidate gene analysis for the PAX6 negative cases. This is an excellent article summarizing state of the art genomics for aniridia.

Mooren's ulcer in children

Anurag Mathur, Jatin Ashar, Virender Sangwan
Br J Ophthalmol 2012;96:796-800. (f12)
Purpose: To describe the epidemiology, clinical features, management and outcomes of paediatric Mooren's peripheral ulcerative keratitis.

Methods: All patients with Mooren's ulcer aged <18 years presenting at a single centre from 1987 to 2010 were enrolled.

Results: 14 eyes of 11 children (seven males and four females with an average age of 12.4562.25 years at presentation) with Mooren's ulcer were included. Eight cases were unilateral and three bilateral. Symptoms at presentation were more severe than in adults. Trauma was the commonest predisposing factor. Eight eyes had severe corneal involvement. Medical management included intensive topical steroid therapy, oral steroid
therapy and immunosuppressant agents.Surgical therapy included tissue adhesive and bandage contact lens application, amniotic membrane

transplantation, optical penetrating keratoplasty and limbal stem cell transplantation and was performed in most eyes as partof primary management or later during the disease course. Patients were followed up for a mean of 69.1 weeks. Ten eyes healed successfully and one developed descemetocele. Three eyes developed secondary infections, one of which ultimately became phthisical. In most eyes, final vision either remained stable or was better than at presentation.
Conclusions: Our data suggest demographic and clinical features of Mooren's ulcer in children differ from those in adults. Good anatomical results and stable visual results
can be achieved with appropriate medical and surgical therapies.

Comments: This is a good series which discusses the rare entity

Childhood blepharokeratoconjunctivitis: characterising a severe phenotype in white adolescents

Samer Hamada, Imran Khan, Alastair K Denniston, Saaeha Rauz1,Br J Ophthalmol 2012;96:949-955. (f12)

Methods: A cohort of 10 white patients (20 eyes), median age 15.2 (range 6e27) years were identified among 62 patients with BKC attending a tertiary referral centre. Clinical features were graded and lid/conjunctiva swabs were performed, before instituting a hierarchical therapeutic protocol comprising lid hygiene, topical/ systemic antibiotics, intensive topical glucocorticoids and systemic immunosuppression.

Results: The median duration of symptoms prior to presentation was 4.3 (range 1.2e16.3) years, with 14 eyes (nine patients) demonstrating 3608 peripheral corneal vascularisation associated with encroachment/ involvement of the visual axis in 10 eyes (six patients).
Corneal perforation(s) occurred in three eyes (two patients). Intensive topical glucocorticoids enabled disease control in 10 eyes (seven patients). In six eyes (three patients), persistent active disease necessitated systemic immunosuppression (azathioprine (2), mycophenolate mofetil (1), prednisolone (1)) achieving
disease remission within three months with no adverse events reported.

Conclusions: Suboptimal treatment of BKC in white children may permit a progressively destructive sightthreatening phenotype, which may last into adulthood and require immunosuppression. Appropriate aggressive steroid-based and steroid-sparing strategies are vital fordisease remission.

Surgical Management of Pediatric Limbal Dermoids With Sutureless Amniotic Membranes Transplantation and Augmentation

Amir Pirouzian, MD; Huck Holz, MD; Kevin Merrill, MD; Rattehalli Sudesh, MD; Kris KArlen, MD J Pediatr Opthalmol Strabismus 2012; 49:114-119 March/April (f12)

This article examines a new technique using amniotic membrane to reconstruct the ocular surface after removal of a limbal dermoid. This is a case series of 3 patients all with corneal limbal dermoids treated with excision and placement of an amniotic membrane. The authors introduce an interesting idea for treatment of limbal dermoids especially for those who have an interest in ocular surface disease.

Oral cyclosporine therapy for refractory severe vernal keratoconjunctivitis

Gokhale NS, Samant R, Sharma V. Indian J Ophthalmol 2012;60:220-3 (May-Jun) (f12)

The authors report the success of oral cyclosporine therapy in a patient with severe vision-threatening vernal keratoconjunctivitis. A child presented with severe allergy which was not controlled with topical steroids, cyclosporine and mast cell stabilizers. Oral steroids were required repeatedly to suppress inflammation. The child showed a dramatic improvement and stabilization with oral cyclosporine therapy. The authors were able to discontinue his oral steroids totally and were also able to shift him onto milder steroid drops. The patient tolerated cyclosporine therapy very well with no major side effects. His ocular surface stabilized though he still has seasonal fluctuation of the surface inflammation. The authors conclude that in this case oral cyclosporine was used with success and no significant side effects in this child with recalcitrant VKC.

Boston keratoprosthesis and Ahmed glaucoma valve for visual rehabilitation in congenital anterior staphyloma

Srinivasan B, Choudhari NS, Neog A, Latka S, Iyer GK. Indian J Ophthalmol 2012;60:232-3 (May-Jun) (f12)

The authors report on a case of congenital anterior staphyloma with a promising result following staphylectomy with implantation of a keratoprosthesis and a glaucoma drainage device in a seven-month-old child. Six months after the surgery, the child was fixing and following light with the right eye. Retinoscopy value was +2.50 diopters. The keratoprosthesis was well-integrated. Ahmed glaucoma valve was *in situ*. An elevated conjunctival bleb was noted. Digital tension was normal.

Trachoma: an update on prevention, diagnosis, and treatment

Bhosai, Satasuk Joy Bailey, Robin L.; Gaynor, Bruce D.; Lietman, Thomas M.

Current Opinion in Ophthalmology. July 2012; 23(4):288-295

Trachoma is the leading infectious cause of blindness worldwide, resulting from recurrent infection with ocular strains of Chlamydia trachomatis It occurs primarily in areas with poverty and poor hygiene. Recurrent inflammation of the eyelids leads to a cascade of conjunctival scarring, entropion, and trichiasis. These processes then lead to secondary corneal ulcers and eventually blindness. This is a review of recent clinical and epidemiological studies regarding the prevention, diagnosis, and treatment of trachoma.

Newer studies propose novel diagnostic tests that appear sensitive for the detection of ocular chlamydial infection. For example, recent studies with ribosomal RNA-based nucleic acid amplification tests (NAATs) have demonstrated improved sensitivities compared to DNA-based NAATs. The progression of scarring has now been characterized with confocal microscopy. Immunologic studies have further explored the etiology of clinical sequelae, suggesting that chronic inflammation can lead to progressive scarring even in the absence of active infection with Chlamydia. Mass oral azithromycin distribution remains a mainstay of treatment and studies have assessed the appropriate frequency and duration of treatment programs. Current studies have also explored ancillary effects of azithromycin distribution on mortality and bacterial infections.

In summary, trachoma programs have had remarkable success at reducing chlamydial infection and clinical signs of trachoma. Recent work suggests improved methods to monitor infection and scarring, and better ways to distribute treatment. Whereas studies continue to demonstrate reduction in infection in hyperendemic areas, more work is necessary to achieve elimination of this blinding disease.

Efficacy and potential complications of difluprednate use for pediatric uveitis

Slabaugh MA, Herlihy E, Ongchin S, van Gelder RN. Am J Ophthalmol. 2012;153(5):932-8. (f12)

This retrospective observational case series evaluated the clinical effect of topical difluprednate in pediatric patients for treatment of noninfectious uveitis. Twenty-six eyes of 14 pediatric patients with noninfectious uveitis who were treated with topical difluprednate were evaluated. Anterior and posterior cell grade, visual acuity, intraocular pressure (IOP), and cystoid macular edema (CME) were recorded at each visit. Main outcome measures were changes in anterior segment cell, CME, visual acuity, and IOP and development of a visually significant cataract. A significant (≥ 2-grade decrease or decrease to 0 in anterior

segment cell) reduction in anterior segment inflammation was observed during treatment with topical difluprednate in 88% of eyes (22/25) when used as an adjuvant to systemic immunomodulatory therapy. In addition, improvement in CME associated with uveitis was seen in response to topical therapy with difluprednate in 78% of eyes with CME (7/9). A significant IOP response (IOP increase of ≥ 10 mm Hg from baseline and IOP ≥ 24 mm Hg) was seen in 50% of eyes (13/26) and in 50% of patients (7/14); 3 eyes of 2 patients required glaucoma surgery. Cataract formation or progression was observed in 39% of eyes (10/26) and in 43% of patients (6/14); 5 eyes of 3 patients required cataract surgery. The authors concluded that difluprednate is an effective agent for both control of anterior segment inflammation and reduction of CME in pediatric patients with uveitis when used as an adjuvant to systemic immunomodulatory therapy. A high rate of steroid-induced IOP elevation and cataract formation is seen in this population. Close monitoring of pediatric patients receiving difluprednate was recommended.

Clinical Features and Presentation of Infectious Scleritis from Herpes Viruses-
A Report of 35 Cases

Luis Alonso Gonzalez-Gonzalez, MD, Nicolas Molina-Prat, MD, Priyanka Doctor, MS, et al
Ophthalmology July 2012; 119:1460-1464 (f12)

This is a retrospective case series of 35 patients with infectious scleritis from herpes virus to describe the clinical features and presentations of infectious scleritis resulting in herpes viruses. Correlation between classification, severity in symptoms, and diagnosis of herpetic-associated scleritis. Vision loss, presence of associated uveitis, keratitis, glaucoma or systemic disease were documented for follow-up.
The association between scleritis and infectious disease may be higher than previously reported by other series. Herpes viruses account for 7% of all scleritis cases and its diagnosis may be challenging where there is not a classically diagnostic picture. The reported prevalence of herpetic scleritis varies from study to study ranging from 4.2% to 7.5%. The herpes virus is the most common etiology followed by syphilis and pseudomonas. Scleral biopsy was not possible. Herpes simplex virus/herpes zoster virus titers (IgG and IgM) as well as clinical responses to oral Acyclovir established at the present of diagnosis.
The authors suggest considering biopsy and analysis of scleral tissue by specific anti-herpes simplex virus and anti-herpes zoster virus immunofluorescence to establish a definitive diagnosis, especially in cases with clinical findings or equivocal.

This study was performed at the Massachusetts Eye Research and Surgery Institution, Cambridge, Massachusetts, Institute Clinic of Ophthalmology, Hospital Clinic of Barcelona, Spain, and Bay View Clinic, Mumbai, India.

COMMENT: Excellent pictures of clinical features of infectious scleritis from herpes virus page 1462.

Clinical Manifestations of Human Immunodeficiency Virus-Induced Uveitis

Paradee Kunavisarut, MD, Wasna Sirirungsi, PhD, Kessara Pathanapitoon, MD, PhD, Aniki Rothova, MD, PhD
Ophthalmology July 2012; 119:1455-1459 (f12)

The purpose of this prospective cohort study of 6 patients with HIV uveitis described clinical manifestations of patients with Human Immunodeficiency Virus (HIV) induced uveitis in Thailand. The main outcome measures were clinical manifestations and laboratory findings. Six patients (8 eyes) with HIV-induced uveitis who had extremely high intraocular plasma HIV-1 RNA ratio. Human Immunodeficiency Virus induced uveitis should be suspected in HAART-naïve, HIV-positive patients or in those in whom this treatment fails and who have anterior uveitis without any retinal lesions. These patients usually exhibit no response to topical corticosteroids. The concurrent determination of Human Immunodeficiency Virus (HIV) load in the intraocular fluids and plasma may clarify the cause of HIV-associated uveitis.

This study was performed at the Department of Ophthalmology and Division of Clinical Microbiology, Chiang Mai University, Chiang Mai, Thailand, and Department of Ophthalmology, University Medical Center Utrecht, Utrecht, The Netherlands.

Risk of Corticosteroid-Induced Hyperglycemia Requiring Medical Therapy among Patients with Inflammatory Eye Diseases

Joshua D. Udoetuk, MD, Yang Dai, MS, Gui-Shuang Ying, PhD, et al, for the Systemic Immunosuppressive Therapy for Eye Diseases Cohort Study Research Group
Ophthalmology August 2012; 119: 1569-1574 (f12)

This is a retrospective core study designed to identify the incidence and risk factors for corticosteroid-induced hyperglycemia requiring medical therapy among patients with inflammatory eye diseases. Eligible patients who use oral corticosteroids during follow-up were identified and followed longitudinally for initiation of hypoglycemic medications over 1 year after beginning corticosteroids. The remaining eligible patients were followed for 1 year after initial visit. Survival analysis was used to calculate the risk of hyperglycemia requiring medical therapy to identify potential risk factors and the main outcome measures for initiation of hypoglycemic medications.

The results of the study suggest that absolute risk of corticosteroid-induced hyperglycemia that is detected and treated with hypoglycemic therapy in ocular inflammation is low. Older age and African-American races also are considered risk factors for the development of corticosteroids-induced hyperglycemia.

“PHYSICIANS WHO USE SYSTEMIC CORTICOSTEROIDS FOR OCULAR INFLAMMATORY DISEASE SHOULD BE AWARE OF THIS RISK AND SHOULD CONSIDER SURVEILLANCE FOR HYPERGLYCEMIA AMONG HIGH RISK PATIENTS.”

Ocular Toxocariasis: Epidemiologic, Anatomic, and Therapeutic Variations Based on a Survey of Ophthalmic Subspecialists

Dana Woodhall,MD , Michelle C. Star,BA, Susan P. Montgomery, DVM,MPH, Jeffrey L. Jones,MD,MPH, Flora Lum,MD, Russell W. Read,MD,PhD, Ramana S. Murthy,MD
Ophthalmology June, 2012; 119:1211-1217 (f12)

This is a web-based, cross-sectional survey designed to assess the current prevalence, diagnosis and treatment of ocular toxocariasis in the United States. An electronic survey was sent to 3020 ophthalmologic subspecialists belonging to the American Uveitis Society, (AUS), American Association of Pediatric Ophthalmology and Strabismus (AAPOS), American Society of Retinal Specialists (ASRS).
159 patients were reported by 559 respondents. The median patient age was 11.5 years. 45% lived in the southern regions of the United States. 69% owned a dog or cat. 85% of affected patients reported visual loss; 71% with permanent visual loss; 29% had temporary visual loss. The most common cause of total visual loss was subretinal granulomatous mass or scar which along with vitritis and central scotoma was also the most common clinical finding. Ocular toxocariasis continues to occur in the United States and it affects mostly children and causes permanent visual loss in many patients.

This study was performed by the Center for Disease Control in Atlanta, Georgia in conjunction with the American Academy of Ophthalmology, University of Alabama, Birmingham and Indiana University Glick Eye Institute.

TREATMENT OF INTRACTABLE POSTERIOR UVEITIS IN PEDIATRIC PATIENTS WITH THE FLUOCINOLONE ACETONIDE INTRAVITREAL IMPLANT (RETISERT)

Patel, Chirag C MD; Mandava, Naresh MD; Oliver, Scott C N MD; Braverman, Rebecca MD; Quiroz-Mercado, Hugo MD; Olson, Jeffrey L MD

Retina. March 2012; 32(3):537-542

Chronic noninfectious posterior uveitis is a vision-threatening and potentially disabling condition. Onset in childhood can be especially debilitating due to a lifetime of visual impairment. Traditionally, these children have been treated with local or systemic steroids, or steroid-sparing agents. In addition to many side effects associated with steroids, administering these agents can be challenging in children. Eye drops are often difficult for parents to administer and can lead to noncompliance with therapy. Periocular or intravitreal steroid injection usually must be performed in the operating room. If repeated injections are required every 2 months to 3 months, this can be especially cumbersome. Pediatric patients can therefore be an ideal population for the Retisert implant, which is an intravitreal implant containing .59mg of fluocinolone acetonide. The implant is designed to last 3 years and can therefore circumvent many of the problems of traditional therapy.

The purpose of this study was to evaluate the efficacy and safety of the Retisert implant in pediatric patients with intractable noninfectious posterior uveitis. This is a retrospective chart review. The study included 6 eyes in 4 patients younger than 18 years. Mean age at implant placement was 9.2 years (range, 6–13 years). All patients had failed prior conventional therapy. Mean follow-up duration was 698 days (range, 376–1,189 days). Postoperative visual acuity improved by >=3 lines in 3 eyes. Inflammation was well controlled postoperatively leading to successful discontinuation of topical steroids in all six eyes. There were no cases of postoperative infection. There were no postoperative complications of surgical technique. As with any steroid treatment, concerns for the development of cataract and increased IOP remain.

Although this is a small series, the data demonstrate evidence that the Retisert implant may be a viable option for children with chronic noninfectious uveitis.

[Home]

RETINA

Retinal detachment associated with optic disc colobomas and morning glory syndrome

S Chang, E Gregory-Roberts and R Chen

Eye. April 2012;26:494-500 (f12)

A retrospective review of 5 patients with optic nerve coloboma or morning glory syndrome with associated retinal detachment or retinoschisis was conducted. The authors describe these patients' clinical findings, spectral domain optical coherence tomography (OCT) imaging, intraoperative findings, and treatment outcomes. The authors created 3-d OCT reconstructions, showing retinoschisis and retinal detachment. The authors frequently noted a retinal break within the optic disc defect with three-dimensional OCT imaging. Glial tissue was

sometimes observed within the anomalous defect. Vitrectomy and resection of the tractional tissue was performed in some cases, and one case with no traction was found to have spontaneous retinal reattachment. The authors speculated that a strong attachment of the vitreous to the anomalous optic disc may contribute to the pathogenesis of peripapillary retinal detachment associated with colobomatous optic disc anomalies, either directly or by creating a secondary retinal break.

Idiopathic macular hole in a child

J C Park and K N Frimpong-Ansah
Eye. April 2012;26:620-621. (f12)

The authors report that an 8 year-old girl had a unilateral macular hole and no known history of trauma. She was treated with pars plana vitrectomy, internal limiting membrane peeling and C3F8 gas, with closure of the macular hole and some visual improvement.

The lack of known history of trauma does not exclude the possibility that this was a traumatic macular hole. The child had been treated for “amblyopia” in the same eye at the age of 6 years, and it’s possible that the macular hole was missed at that exam and possible trauma was not recalled from > 2 years previously.

Ophthalmologic findings in Aicardi syndrome

Fruhman G, Eble TN, Gambhir N, et al. J AAPOS 2012;16:238-241. (f12)

Ophthalmologic features of 40 girls with Aicardi syndrome are reported. Subjects were evaluated during a family conference of the Aicardi Syndrome Foundation. The median age was 5 years. 39 of 40 had seizures within the first 6 months of life, and all 40 had partial or complete agenesis of the corpus callosum. Chorioretinal lacunae (88% of evaluated eyes), followed by optic nerve abnormalities (81%) were the most common ophthalmic findings. Coloboma was the most observed optic nerve anomaly. This study group was self-selected which may introduce bias in the frequency of various ophthalmic findings.

Pediatric Posterior Scleritis – Letter to the Editor

Genie M Bang, MD, Michael C Brodsky, MD
Ophthalmology July 2012; 119:1505 (f12)

The letter emphasizes that neuroimaging abnormalities can establish a diagnosis of posterior scleritis and obviate the need for ocular ultrasound or optical coherence tomography (as was reported in the original article by Cheung and Chee.

The authors of the letter describe a child with a 1 month history of severe bilateral eye pain and bilateral posterior scleral thickening wit prominent peribulbar enhancement without evidence of myositis, dacryoadenitis, or perineuritis on MRI imaging. The authors emphasized that the spectrum of posterior scleritis in children extends to include isolated posterior scleral inflammation producing severe pain in the absence of any other retinal manifestation. Treatment can be carried out with the use of steroids with adjunct Methotrexate chemotherapy. The authors of the letter emphasize that “enhanced MRI may be particularly useful for identifying this common variant of posterior scleritis in a child with bilateral ocular pain”.

Quality of Nonmydriatic Digital Fundus Photography Obtained by Nurse Practitioners in the Emergency Department: The FOTO-ED Study

Cedric Lamirel,MD, Beau B. Bruce,MD,MS, Daniel W. Wright,MD, Kevin P. Delaney,MPH et al.
Ophthalmology March, 2012; 119:617-624 (f12)

This is a prospective, cross-sectional study designed to evaluate the use of a newly developed rating scale to assess the quality of fundus photography performed in the emergency room by non-ophthalmic-trained nurse practitioners. 350 patients (with complaints of headache, acute focal neurologic deficit, acute visual changes or elevated diastolic blood pressure greater than 120mmHg) were enrolled in the in the Fundus photography versus Ophthalmoscopy Trials Outcomes in the Emergency Department (FOTO-ED) study who were photographed by nurse practitioners after less than 30 minutes of training followed by supervision.

Photographs were graded for quality by two neuro-ophthalmologists on two separate occasions. Photos were statistically compared for image quality of the optic disc, superior and inferior temporal arcades and the macula. A five point rating scale was used.
The authors concluded that the five point rating scale is a reliable measure of nonmydriatic photograph quality and that the current study may have a direct impact in the use of use of nonmydriatic fundus cameras in evaluating patients by non ophthalmic personnel in the Emergency Department.
The study was performed by multiple departments at Emory University in Atlanta, Georgia
COMMENT: There is an excellent description of image compression technology, the use of the KOWA nonmyd-alpha D 5 megapixel fundus camera and analysis of the fundus photographs on page 618.

Retinal Structure, Function and Molecular Pathologic Features in Gyrate Atrophy

Panagiotus I. Sergouniotis,MD, Alice E. Davidson,PhD, Eva Lenassi,MD, Sophie R. Devery, MSc, Anthony T. Moore, MA,FRCOphth, Andrew R. Webster,MD, FRCOphth
Ophthalmology March, 2012; 119: 596-605 (f12)

This is a retrospective case series of seven unrelated patients, ages 10-52 years with clinical and biochemical evidence of gyrate atrophy.
The purpose of the study is to describe phenotypic variability and to report novel mutational data in patients with this condition.
The coding region and intron-exon boundaries of ornithine aminotransferase (OAT) were analyzed. Fundus photographs, spectral domain-optical coherence tomography (SD-OCT), fundus autofluorescence (FAT) and microperimetry testing were performed.

Fundus autofluorescence best reveals the extent of neurosensory dysfunction in gyrate atrophy patients. Macular edema is a uniform finding. The fovea is thickened in the early stages of the disease and retinal tubulation is present in advanced disease. Analysis of leukocyte RNA complements the high sensitivity of conventional sequencing of genomic DNA for mutation detection in this gene.
Study performed at the University College London, Moorfields Eye Hospital Special Trustees, London, United Kingdom
COMMENT: Article has excellent fundus photographs, autofluorescence and OCT images. The images are worthwhile to look at.

Photodynamic Therapy for Best Disease Complicated by Choroidal Neovascularization in Children

Sengul Ozdek, MD; Mehmet Cuneyt Ozmen, MD; Hasan Ali Tufan, MD; Gokhan Gurelik, MD; Berati Hasanreisoglu, MD J Pediatr Opthalmol Strabismus 2012; 49:216-221 July/August (f12)

This was a case series of 5 eyes in 4 children who were diagnosed with Best viteliform macular dystrophy (BVMD) and choroidal neovacularization (CNV). One single session of photodynamic therapy (PDT) was performed in these children and the results were documented based on fluorocein angiography and ocular coherence tomography. The case series found that a single session of PDT had a favorable response on CNV associated with BVMD and may be a treatment to consider when vision is compromised.

MACULAR EPIRETINAL MEMBRANE PEELING TREATMENT OUTCOMES IN YOUNG CHILDREN

Ferrone, Philip J; Chaudhary, Khurram M Retina. March 2012; 32(3):530-536

Epiretinal membranes (ERM) are semitranslucent fibrovascular membranes on the inner surface of the retina along the inner limiting membrane. They are most commonly seen in older adults, typically from posterior vitreous separation, and are rarely seen in the pediatric population. In children they have been estimated to occur with a frequency of about 1 in 21,000. In such cases, an ERM is usually due to an underlying cause such as intraocular inflammation, trauma, or combined hamartoma of the RPE and retina. Other rare causes include retinopathy of prematurity (ROP), familial exudative vitreoretinopathy (FEVR), retinal vascular diseases, Coats disease, Norrie disease, ocular toxocariasis, pars planitis, and other pathologies.

Very few studies have evaluated the treatment of children with ERMs and there are currently no guidelines for surgical removal. ERMs in children are thicker and whiter and more challenging to remove. The purpose of this study is to describe macular ERM peeling treatment outcomes in children 16 years and younger. This study is a retrospective chart review. The medical records of all vitrectomies from 1998 through 2010 were reviewed. Patients 16 years or younger were selected, who had primarily macular or posterior pole disease secondary to an ERM. Patients with ROP, Coats disease, Norrie disease, and incontinentia pigmenti were excluded. Fourteen patients underwent vitrectomy with ERM peeling, and their preoperative and postoperative visual acuities were compared. The mean age at surgery was 8 years, with an average follow-up of 4.2 years. The average presenting visual acuity was 20/258. Average postoperative visual acuity was 20/100, with improvement in 12 patients (86%) and no change of visual acuity in 2 patients (14%). Pediatric ERMs respond well to vitrectomy and membrane peel with a low likelihood of recurrence.

[Home]

ORBIT

Case Report: Watery Eyes During Urination Bulwer C, Hodin M, Mansy S Pediatrics 2012; 129(3):e803-5. (f12)

This was a case report of an unusual case of tear production with painless micturition, only once described in the medical literature in 1932. A 3-year-old girl with painless “watering of her eyes every time she passed urine” present since birth and occasionally associated with a vacant look and dropping of her jaw was reported. With only 1 previously described case in the literature but clear evidence from online fora, the authors state that this phenomenon may be more common than previously thought

Resolution of Congenital Nasolacrimal Duct Obstruction With Nonsurgical Management

Pediatric Eye Disease Investigator Group .. Arch Ophthalmol. 2012;130(6):730-734 (f12)

This was a randomized trial to determine how often nasolacrimal duct obstruction (NLDO) resolves with 6 months of nonsurgical management in infants aged 6 to less than 10 months. 107 infants aged 6 to less than 10 months who had NLDO and no history of nasolacrimal duct surgery were prescribed 6 months of nasolacrimal duct massage and topical antibiotics as needed. Resolution of the NLDO was assessed 6 months after study entry and was defined as the absence of all clinical signs of NLDO (epiphora, increased tear lake, or mucous discharge) and not having undergone NLDO surgery. At the 6-month examination, which was completed for 117 of the 133 eyes (88%), the NLDO had resolved without surgery in 77 eyes (66% [95% CI, 56%-74%]). None of the baseline characteristics evaluated, including age, sex, laterality, and prior treatment were found to be associated with resolution. In summary, in infants 6 to less than 10 months of age, more than half of eyes with NLDO will resolve within 6 months with nonsurgical management. Knowledge of the rate of NLDO resolution in infancy without surgery will help clinicians and parents effectively discuss treatment options.

Orbitofacial neurofibromatosis: clinical characteristics and treatment outcome

I A Chaudhry, J Morales, F A Shamsi, W Al-Rashed, E Elzaridi, Y O Arat, C Jacquemin, D T Oystreck and T M Bosley

Eye. April 2012;26:583-592 (f12)

This is a retrospective case series that includes clinical observations and surgical management of 60 patients (31 females and 29 males; mean age, 14 years) with neurofibromatosis type 1 affecting the orbits or periorbita. The patients were seen at the King Khaled Eye Specialist Hospital in Saudi Arabia over a 23 year time period.

Patients were identified and medical records reviewed for demographic data, ophthalmologic examinations, surgical interventions, and procedure outcome to create a retrospective, non-comparative case series of patients with OFNF seen at one medical center over a 23-year period. The patients were followed for an average of 5.7 years (range 1 month to 23 years). Presenting signs and symptoms included eyelid swelling in all patients, ptosis in 56 (93.3%), proptosis in 34 (56.6%), dystopia or strabismus in 30 (50%), and decreased visual acuity in 50 (83.3%). Surgical intervention included ptosis repair in 54 (90%; mean 1.6 surgical procedures), facial and orbital tumor debulking in 54 (90%; mean 2.3 surgeries), and canthoplasty in 28 (46.6%) patients. Eleven patients required enucleation or exenteration of a blind eye. Patients with OFNF often require multiple procedures to preserve vision, prevent additional disfigurement, and

achieve cosmetic improvement. The approach to each patient must be individualized.

Atypical Presentations of Orbital Cellulitis Caused by Methicillin-Resistant Staphylococcus aureus

Marc T. Mathias, MD, Michael B. Horsley, MD, Louise A Mawn, MD, Stephen J. Laquis, MD, Kenneth V. Cahill, MD, Jill Foster, MD, Malena M. Amato, MD, Vikram D. Durairaj, MD
Ophthalmology June, 2012; 119:1238-1243 (f12)

This is a multicenter retrospective case series designed to evaluate the epidemiologic and clinical features of orbital cellulitis caused by Methicillin-resistant Staphylococcus aureus (MRSA).
15 patients with culture positive MRSA orbital cellulitis were evaluated. All recent cases of orbital cellulitis at several hospitals and surgical centers were reviewed. Cases with culture-positive MRSA from aspirates were identified. The data collected and analyzed retrospectively included patient demographics, medical history, presenting sign, imaging results, surgical procedure performed, surgical culture results, visual acuity at presentation, visual acuity at last follow-up, and duration of antibiotics.

The main outcome measures were presenting sign, radiographic evidence of paranasal sinus disease, the presence or absence of antecedent upper respiratory infection, and final visual acuity. 15 cases were identified. The mean patient age was 31.9 years. Lid swelling was the presenting sign in 14 of 15 patients. No patients had preceding upper respiratory infection and 1 patient had antecedent eyelid trauma. Only 3 of the 15 patients had documented adjacent paranasal sinus disease on imaging. Lacrimal gland abscess or dacryoadenitis was the presenting finding in 5 of 15 patients. Multiple orbital abscesses were identified in 4 of 15 patients by computer tomography or magnetic resonance imaging. 14 of 15 cases required surgical intervention and 4 of 15 cases had loss of visual acuity to light perception or worse. All 4 of these cases had a delay in referral for surgical intervention.

In these 15 patients with MRSA orbital cellulitis, the typical clinical setting of orbital cellulitis was absent. In particular there was no identified antecedent upper respiratory illness nor was there a preceding traumatic injury. Lid swelling in the absence of recent upper respiratory illness, lacrimal gland focus, multiple orbital abscesses, and lack of adjacent paranasal sinus disease may be predictive factors that suggest MRSA as the causative agent in patients with this type of orbital cellulitis.

This study was performed at the Rocky Mountain Lions Eye Institute, University of Colorado, Vanderbilt Eye Institute, Vanderbilt University School of Medicine, Nashville, Tennessee, Havener Eye Institute, Ohio State University, Columbus,

Ohio, Department of Ophthalmology, Haywood/Freemont Medical Centers, Union City, California.
COMMENT: This article is a unique profile of orbital cellulitis in patients with Methicillin-resistant Staphylococcus aureus, namely the paucity or absence of antecedent upper respiratory illness or preceding traumatic orbital injury.

Thyroid associated orbitopathy

Maheshwari R, Weis E.
Indian J Ophthalmol 2012;60:87-93 (Mar-Apr) (f12)
This is a review article that summarizes the pathophysiology, clinical presentation, and management options for the different stages of disease. A classification scale for TRO is presented, as well as management flow charts.

[Home]

PLASTICS

Acquired Brown's syndrome following cosmetic blepharoplasty

C Wilde, M Batterbury and J Durnian
Eye. May 2012; 26:757-758 (f12)

The authors report a case of unilateral acquired Brown's syndrome after cosmetic blepharoplasty in a 56-year-old male. The patient had diplopia in right gaze approaching primary position. This case may be the fourth reported one.

Success rates of primary probing for congenital nasolacrimal obstruction in children.

Arora S, Koushan K, and Harvey JT. J AAPOS 2012;16:173-176. (f12)

This retrospective comparative case series evaluated all patients who underwent a primary probing and irrigation for NLDO by one of the authors, over a 56 month period. Primary outcome was treatment success or failure at 3 months. Overall success was 72%, with no appreciable difference between boys and girls. The best success rates were in children <3 years of age. Age was a significant risk factor for a failed probing. The older age group was a smaller sample size which could skew results, and the data in this study is consistent with prior publications

Associated signs, demographic characteristics, and management of dacryocystocele in 64 infants.

Dagi LR, Bhargava A, and Melvin P. J AAPOS 2012;16:255-260. (f12)

This study was designed to capture a significant sample size to assess the influence of age at presentation, sex, associated infection, laterality and presence of intranasal cyst on the choice of intervention to achieve successful resolution of dacryocystocele. All patients diagnosed with this condition over a 15-year period at one institution were included. 79 eyes were included (64 patients). Almost 70% of patients presented before 30 days of life. Half of the patients were treated successfully without general anesthesia (digital massage or in-office probing). Of the remaining 31 patients, 19 were successfully treated with a simple probing under anesthesia (2 with stent placement and 1 with balloon dilation). 12 patients received a more complex procedure (endoscopic procedure +/- excision of an associated intranasal cyst). Bilateral cases had a 6.4x increased odds of an endoscopic procedure. All patients were successfully treated. Female infants constituted 63% of the patients. The authors comment that since neonates are obligate nasal breathers, intranasal cysts can cause respiratory distress, especially when the infant is nursing or feeding. CT imaging was not performed in all patients, so the prevalence of intranasal cysts may have been underestimated.

Entropion in children with isolated peripheral facial nerve paresis

Alsuhaibani AH, Bosley TM, Goldberg RA, Al-Faky YH.

Eye. Aug 2012;26:1095-8. (f12)

Adults with facial nerve paresis generally develop lower eyelid ectropion. However, in this consecutive case series of pediatric patients with facial nerve paralysis due to congenital, traumatic, and middle ear infection causes, all patients developed lower eyelid entropion rather than ectropion. The 10 patients (4 males and 6 females; mean age at presentation, 4 years) had nonsyndromic unilateral facial nerve paralysis. All patients also had lower eyelid retraction (mean 2.3 mm) and lagophthalmos (mean 2.9 mm). None had enophthalmos, lower eyelid ectropion, or brow ptosis. Entropion formation has also been reported in five children with syndromic facial nerve paralysis. Thus, entropion after facial nerve paralysis may be relatively common in pediatric facial nerve paralysis from various causes.

Congenital Nasolacrimal Duct ObstructionDelineation of Anatomic Abnormalities With 3-Dimensional Reconstruction

Avery H. Weiss, MD; Francine Baran, MD; John Kelly, PhD

Arch Ophthalmol. 2012;130(7):842-848. (f12)
The purpose of this paper was to characterize anatomical abnormalities of the distal nasolacrimal duct (NLD) in children with congenital obstruction using high-resolution computed tomographic imaging. The anatomy of the bone and soft tissue of the NLD and the postductal anatomy of the inferior meatus in 6 children with a wide spectrum of NLD obstructions were imaged by computed tomographic scans and then reconstructed in a 3-dimensional viewer. The axial computed tomographic images and 3-dimensional reconstructions showed bony obstructions of the distal NLD in 3 children, membranous obstructions of the distal NLD in 1 child, and a postductal obstruction in the inferior meatus in 1 child. One child had a combined soft tissue obstruction of the NLD and post-NLD obstruction. In summary, this study provides anatomic evidence of a bony or membranous obstruction at the distal portion of the NLD or of a postductal obstruction at the inferior meatus in children with congenital NLD obstruction.

Efficacy of systemic propranolol for severe infantile haemangioma of the orbit and eyelid: a case study of eight patients

F Thoumazet, C Le´aute´-Labre`ze, J Colin,1 B Mortemousque Br J Ophthalmol 2012;96:370 -374.(f12)

Aim: To assess the efficacy of systemic propranolol for severe capillary haemangiomas involving eyelid and orbit.
Method: This was a longitudinal retrospective study that began in November 2007, involving eight children with disfiguring orbit and eyelid capillary haemangioma who received oral propranolol therapy.
In all kids there was 100% regression. But the there is no New information in the article.

Management of Unilateral Versus Bilateral Lacrimal Drainage System Dysfunction in Down Syndrome

Yasser H. FAky, MD, FRCS; Ahmad Mousa, MSc; Hattan T. Alkhiary, MD; Abdul Rahman Al-Mosallam, MD J Pediatr Opthalmol Strabismus 2012; 49:109-113 March/April (f12)
Children with Down's syndrome have systemic features that can alter ormal functioning of the lacrimal drainage system including a hypoplastic nasal bone and a flat nasal bridge. Theses can change the anatomical features of the medial canthus area. Lacrimal drainage system dysfunction is known to be more common in patients with Down's syndrome. The aim of the study is to evaluate the treatment outcomes of unilateral versus bilateral congenital nasolacrimal duct obstruction in patients with Down's syndrome and to highlight the effect of other features that may result in poor outcomes. The study found that unilateral disease and lower-end NLDO are good prognostic factors. For bilateral disease and mild tearing, it is advisable to wait until 5 years or older.

Community-acquired Methicillin-Resistant *Staphylococcus aureus* Bilateral Acute Dacryocystitis in a Neonate

Chandravanshi SL, Sutrakar SK, Bajaj N. Indian J Ophthalmol 2012;60:155-6 (Mar-Apr) (f12)

The authors present a case of bilateral acute dacryocystitis secondary to congenital nasolacrimal duct obstruction (CNLDO), caused by community-acquired methicillin-resistant *Staphylococcus aureus* (CA-MRSA). The mother had taken antibiotics for bilateral MRSA mastitis during pregnancy. Bilateral incision and drainage of the abscess was performed under general anesthesia. Swab cultures demonstrated *Staphylococcus aureus*, resistant to methicillin. The patient was given IV vancomycin and topical 5% vancomycin solution was given QID. The lacrimal swelling and erythema resolved completely over the course of two days. Probing and irrigation of the nasolacrimal duct was performed in both eyes two weeks after presentation which resolved CNLDO. Patient was free from lacrimal symptoms at six month's follow-up.The probable source of infection was maternal breast skin, which reached the nasolacrimal system via skin-to-skin contact during breast feeding.

Single stage surgery for Blepharophimosis syndrome

Bhattacharjee K, Bhattacharjee H, Kuri G, Shah ZT, Deori N. Indian J Ophthalmol 2012;60:195-201 (May-Jun) (f12)

The authors report the functional and cosmetic outcome of single stage surgical procedure for correction of the classic components of Blepharophimosis syndrome. So far, most of the reported surgical techniques for BPS involve multistage procedures. 11 patients with Blepharophimosis syndrome were operated between July 2004 and April 2008. Each patient had undergone the correction of epicanthus inversus, telecanthus, palpebral phimosis, and bilateral ptosis as a single-stage surgical procedure. The surgical outcome was assessed both functionally and cosmetically.Patients were examined and photographed before and after surgery. The mean follow-up was 3 years.There was postoperative improvement in visual acuity, astigmatism, telecanthus, and superior visual field. All the patients had a stable functional and cosmetic result after a mean follow-up period of 3 years. The authors conclude that single stage surgery for BPS can be successful.

Intralesional bleomycin for the treatment of periocular capillary hemangiomas

Smit DP, Meyer D. Indian J Ophthalmol 2012;60:326-8 (Jul-Aug) (f12)

The authors describe the use of intralesional bleomycin injections (IBIs) to treat

potentially amblyogenic lesions in two cases where other modalities have failed. In both cases monthly IBIs successfully cleared the visual axis of the affected eye before the age of 1 year thus preventing permanent sensory deprivation amblyopia. A total of five and nine injections, respectively, were used and no significant side effects were noted. The authors conclude IBI appears to be a useful alternative in the treatment of periocular capillary hemangiomas refractory to more conventional modalities

[Home]

GLAUCOMA

Influence of Parental Health Literacy and Dosing Responsibility on Pediatric Glaucoma Medication Adherence

Rebecca B. Freedman; Sarah K. Jones, BS; Alice Lin, MD; Alan L. Robin, MD; Kelly W. Muir, MD, MHS.
Arch Ophthalmol. 2012;130(3):306-311. (f12)

The goal of this study was to assess glaucoma medication adherence in children, hypothesizing that poor parental health literacy and eye drop instillation by the child are associated with worse adherence. This was a prospective, observational study which enrolled pediatric patients with glaucoma who were prescribed eye drops. The study included 46 of the 50 enrolled children who used the Medication Event Monitoring System for 30 days or more. Adherence ranged from 43% to 107% (93% [12%]) and was not associated with age (slope, 0.09 [0.52]; P = .86) but decreased with the parent's lower health literacy (slope, 0.62 [0.24]; P = .01).The mean number of dosing errors for medications prescribed daily vs twice daily was similar (3.3 vs 2.9; P = .66). The proportion of doses taken on schedule (within 2 hours of prescribed dosing interval) ranged from 3% to 97% (median, 34%; mean, 41% [24%]) and was better when the parent vs the child instilled eye drops (46% [26%] vs 23% [19%]; P < .001). In summary, time-dependent glaucoma medication adherence was better when the parent was responsible for eye drop instillation. Overall decreased adherence was associated with decreased parental health literacy. Children of parents with poor health literacy are vulnerable to poor medication adherence; efforts to address poor health literacy may improve outcomes.

Age variations in intraocular pressure in a cohort of healthy Austrian school children

W A Dusek, B K Pierscionek and J F McClelland
Eye. June 2012;26: 841-844 (f12)

This study provides normative data on intraocular pressure (IOP) in 211 normal European children ages 6 to 15 years. The Icare tonomoter was used. Mean IOP was higher in males (=15.0 mm Hg) than in females (=14.4 mm Hg; P = 0.04). Mean IOP increased from approximately 12 mm Hg to 17 mm Hg for both genders from the ages of 6 to 9 years. Thereafter, there was no clear relationship between IOP and age.

Comparison of Tono-Pen and Goldmann applanation tonometers for measurment of intraocular pressure in healthy children.

Bradfield YS, Kaminski BM, Repka MX, et al. J AAPOS 2012;16:242-248. (f12)

The primary objective of this study was to assess the agreement between Goldman applanation tonometry (GAT) and Tono-pen measurements in children. Other objectives included: identifying factors that influence the agreement between GAT and Tono-Pen, to assess the correlation between IOP measured with the GAT and central corneal thickness (CCT), and to estimate the precision of Tono-Pen IOP measurements in children in an office setting and under general anesthesia (GA). Most GAT measurements were slit-lamp mounted in an office setting, but a few were performed with a handheld GAT (in the office setting or under GA). Tono-Pen measurements were similar to or slightly higher than the GAT measurements when intraocular pressure (IOP) was between 10 and 20mm Hg. Below 11 mm Hg. Tono-Pen measurements trended slightly lower than GAT when estimated IOP was less than 11 mm Hg. Tono-pen measurements were slightly higher than hand-held GAT under GA. The supine position of the patients under GA may have influenced this difference. Larger differences between instruments were found with younger age. With IOP ranging from 6mm Hg to 23mm Hg, GAT measurements were 1.9 mm Hg higher for every 100 um increase in CCT. IOP measurement error increased with increasing office IOP and under GA. In this study, investigators obtaining retest measurements were not masked which may induce a bias. A small sample size for handheld GAT under GA may have affected the data. Also, only normal subjects were enrolled, so the data may not be applicable to children with glaucoma or ocular hypertension. The authors advise that when using the Tono-Pen, multiple measurements be obtained.

Congenital Ectropion Uvea and Mechanisms of Glaucoma in Neurofibromatosis Type 1: New Insights

Deepak P Edward, MD, Jose Morales, MD, Rachida A Bouhenni, PhD, et al *Ophthalmology* July 2012; 119:1485-1494 (f12)

This is a retrospective case series involving enucleated eyes of 5 cases of NF-1 associated glaucoma. The purpose of the study is to describe the clinical and pathologic features of congenital ectropion uvea associated with glaucoma and neurofibromatosis 1 (NF-1). Ectropion uvea and NF-1 glaucoma is secondary to

endothelialization of the anterior chamber angle and is associated commonly with severe pediatric glaucoma in NF-1 patients. Endothelial cell proliferation may be related to overexpression of Ras (Rat sarcoma)-MAPK genes in these eyes.

This study was performed at the Department of Ophthalmology, Summa Health System, Akron, Ohio, and King Khaled Eye Specialist Hospital, Riyadh, Saudi Arabia, and Wilmer Eye Institute, Johns Hopkins University, Baltimore, Maryland, and the Department of Pathology, Duke University Medical Center, Raleigh, North Carolina.
COMMENT: Color photographs on page 1487 and histopathological photomicrographs on pages 1489 were very illustrative of the clinical and histopathological findings associated with this disorder.

Systematic Review of the Agreement of Tonometers with Goldmann Applanation Tonometry

Jonathan Alistair Cook, PhD, BSc, Adriana Paola Botello, MSc, Andrew Elders, MSc, BSc, et al, for the Surveillance of Ocular Hypertension Study Group
Ophthalmology August 2012; 119: 1552-1557 (f12)

This was a systematic review and meta-analysis of directly comparative studies assessing the agreement of 1 or more tonometers with the reference tonometer, Goldmann applanation tonometer (GAT). A total of 11,582 participants (15,525 eyes) were included in this study.

Noncontact tonometer and handheld applanation tonometer seemed to achieve a measurement closest to the Goldmann applanation tonometer. However, there was substantial variability in the measurements both within and between the studies.

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CONGENITAL INFECTIONS

PEDIATRICS

Childhood outcomes after hypothermia for neonatal encephalopathy

Gustafson KE, Leach TM, Green C, Bara R, Petrie Huitema CM, Ehrenkranz RA, Tyson JE Shankaran S, Pappas A, McDonald SA, Vohr BR, Hintz SR, Yolton K,, Das A, Hammond J, Peralta-Carcelen M, Evans PW, Heyne RJ, Wilson-Costello DE, Vaucher YE, Bauer CR, Dusick AM, Adams-Chapman I, Goldstein RF, Guillet

R, Papile LA, Higgins RD; Eunice Kennedy Shriver NICHD Neonatal Research Network
N Engl J Med. May 2012;366:2085-92 (f12)

The authors previously reported early results of a randomized trial of whole-body hypothermia for neonatal hypoxic–ischemic encephalopathy, showing a significant reduction in the rate of death or moderate or severe disability at 18 to 22 months of age. This publication reported on long-term results at age 6-7 years.

Babies suffering from hypoxia/ischemia at birth were randomized to usual care or to whole body cooling to 33.5 degrees Celsius, followed by slow rewarming, for the first 72 hours of life. The primary outcome was death or IQ<70 at age 6-7 years. The study involved 208 babies, and the primary outcome data were available for 190. There was a trend toward a better outcome in the hypothermia compared to the control group: death or an IQ score below 70 occurred in 46 (47%) of the hypothermia babies and 58 (62%) of the control babies (P = 0.06). The borderline significance of the primary outcome was largely driven by deaths, which were lower in the hypothermia group compared to the control group (P=0.04), and occurred primarily before age 18 months. There is concern that reduction in mortality might come at the price of increased survival of severely disabled children. However, there was no evidence of an increased rate of low IQ, severe disability, or cerebral palsy at age 6-7 years among surviving children treated with hypothermia compared to controls. Bilateral blindness was rare in both groups: 1 /67 in the hypothermia group and 2/50 in the control group (P = 0.42).

The study confirms that neonatal cooling within the first hours of life for babies with birth asphyxia decreases death and does not result in an increase in severe disability.

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INFANTILE DISEASE

Povidone Iodine in Ophthalmia Neonatorum-General Correspondence-Letter to the Editor

Wendell J. Scott, MD, Craig D Eck, BS
Ophthalmology March, 2012; 119:653 (f12)
The authors refer to David et al.’s article describing the “Efficacy comparison between povidone iodine -2.5% and tetracycline 1% in prevention of ophthalmia neonatorum (Ophthalmology 2011:118:1454-8).

They point out some overlooked aspects of povidone iodine formulation for ophthalmic use in the prevention of ophthalmia neonatorum.
Free iodine is the active anti-bacterial component of povidone-iodine preparations and 1% povidone-iodine (PI) has a faster kill rate of bacteria than 10%. This is because free iodine parts per million (ppm) is inversely related to the concentration. Therefore, lower concentrations of PI, such as 2.5% will be more effective in killing bacteria than 10% concentrations.
Furthermore, a low pH (3.4-4.5-for PI) is necessary for the bioactivity of the product. This low of a pH also causes a pain and burning sensation upon instillation as well as direct chemical irritation to the eye. This effect can be reduced but not eliminated by further dilutions of the medication such as 0.25 % - Shimeda et al (AJO-2011;151; 11-17) – as well as by combination with dexamethasone (0.15%) which raises the pH to less irritating levels and allows it to be better tolerated by patients without altering the kill rate. It seems to be very effective in treating adenoviral conjunctivitis (but was not evaluated in its effectiveness in preventing ophthalmia neonatorum or “other” infections) (David et al-2012-reply)
The specific formulation and preparation of povidone- iodine need s to be taken into account (parts per million, pH and osmolarity) in addition to the concentration when analyzing comparative studies. “All brands and formulations of povidone-iodine are not created equal.”
The authors are actively engaged in developing such a product.
The Letter was authored in Springfield and Columbia, Missouri.
COMMENT: Very good description on the pharmacokinetics of povidone- iodine

[Home]

SYSTEMIC

Ocular Whipple’s Disease: Therapeutic Strategy and Long-Term Follow-up

Valerie Touitou, MD, PhD, Florence Fenollar, MD, PhD, Nathalie Cassoux, MD, PhD, Helene Merle-Beral, MD, PhD, et al
Ophthalmology July 2012; 119:1465-1469 (f12)

This is a retrospective case series designed to characterize clinical features of ocular Whipple’s disease (WD) and determine the long-term prognosis after antibiotic treatment. Whipple’s disease (WD) initially known as intestinal lipodystrophy was first reported in 1907 by George H. Whipple. This disease was primarily fatal in all cases until the first patient with Whipple’s disease was accidentally cured using chloramphenicol in 1952. It was not until 1991 that a bacillus responsible for the condition of Whipple’s disease was determined. The bacillus is *Tropheryma whipplei*. The bacillus was first cultured in 2000 and is still not routinely used for diagnostic purposes.

Whipple's disease is a multisystemic disease, which include gastrointestinal, central nervous system, joints and lymph nodes. Ocular manifestations are rare in patients with Whipple's disease and usually occur late in the course of this disease. Most of the time patients' ocular manifestations also have a history of gastrointestinal symptoms. A broad spectrum of manifestations includes uveitis, retinitis, retinal hemorrhages, choroiditis, and keratitis. Various neuro-ophthalmologic manifestations have also been reported including ophthalmoplegia, supranuclear gaze palsy, oculo-masticatory myorhythmia, and pendular nystagmus associated with tongue and mandibular myoclonus. Additional findings include myoclonus, ptosis, papilledema, and optic nerve atrophy.

The medical records of patients between January 1993 and December 2010 were evaluated. The initial complaint in most cases was chronic steroid resistant uveitis. This diagnosis was based on cytologic examination and molecular analysis of samples (cerebral spinal fluid, vitreous, duodenum, or any other involved lymph node).
Main outcome measures demographic and clinical characteristics of patients with positive PCR for *Tropheryma whipplei* or periodic acid-Schiff-positive macrophages in the vitreous. Ocular Whipple's disease seems to be a neurologic manifestation of Whipple's disease. Trimethoprim-sulfamethoxazole and Rifampin is an efficient treatment. Treatment needs to be considered for at least 1 year. Long-term low-dose antibiotic therapy may reduce the rate of relapse, neurologic involvement, and death.

This study was performed at the Department of Ophthalmology, Laboratory of Microbiology, Department of Biological Hematology, at the Pitie-Salpetriere Hospital, Paris, France.
COMMENT: Whipple's disease seldom affects children but is also a rare condition that pediatric ophthalmologists should be aware of.

Epidemiology, risk factors and management of paediatric diabetic retinopathy

Marla B Sultan, Carla Starita Kui Huang Br J Ophthalmol 2012;96:312-317 (f12)

This is a good review article looking at the issue of childhood diabetic retinopathy. Issue of screening and management protocols are explained in detail.. Duration of diabetes and glycemic control are the most important determinants of the retinopathy.

Topical Ophthalmic Moxifloxacin Elicits Minimal or No Selection of Fluoroquinolone Resistance Among Bacterial Isolated from the Skin, Nose and Throat

Steven J. Lichtenstein, MD; Liberation DeLeon, MD; Warren Heller, MD; Bonnie Marshall, MT; Gale Cupp, MS; Kimberly Foster, MT; Celeste McLEean, BS; Stuart Levy, MD; David W. Stroman, PhD J Pediatr Ophthalmol Strabismus 2012; 49:88-97 March/April (f12)

The authors investigated children between the ages 1-12 who were treated with moxifloxacin for bacterial conjunctivitis and resolved to answer the question of whether the use of moxifloxacin changes the susceptibility of bacterial isolates in the eyes, cheeks below the eyes and nares. The main concern was that this would cause an emergence of moxifloxacin resistance bacterias (i.e. H. influenza, S. pneumonia and/or S. aureus) in the eyes or other body sites – a concern of pediatricians when prescribing fluoroquinolones. The article found no selection of resistant strains after the use of moxifloxacin topically for 7 days.

Ocular Abnormalities and Systemic Disease in Down Syndrome

Branka Stirn Kranjc Strabismus Jun 2012, Vol. 20, No. 2: 74–77 (f12)

Ocular abnormalities in children with Down syndrome were investigated. The most common abnormalities found in 65 children from 2008-2010 in Slovenia were nystagmus, esotropia, epiphora, Brushfield spots, lens opacities, abnormalities of retinal vessels, foveal hypoplasia or retinal pigment epithelium hyperplasia. Refractive errors in children with Down syndrome were more commonly hyperopia followed by astigmatism, followed by myopia. Of the many ocular abnormalities found in this population, only nystagmus was related to heart problems and heart operations.

Nanophthalmos and situs inversus totalis

Sahin A, Oltulu R, Kivrak AS, Özkagnici A. Indian J Ophthalmol 2012;60:319-21 (Jul-Aug) (f12)

The authors report a unique case of a nanophthalmic patient with situs inversus totalis. The report underlines the importance of the systemic examination of the nanophthalmic patients to detect systemic malformations and visceral transpositions.

Ocular Abnormalities and Systemic Disease in Down Syndrome Retrospective clinical study, University Eye Hospital, Ljubljana, Slovenia

Branka Stirn Kranjc, MD, PhD
Strabismus June 2012; 20(2), 74-77 (f12)

This is a retrospective clinical study from the University Eye Hospital, Ljubljana, Slovenia.
This study correlates eye problems with systemic disease, such as heart disease, hypotony, hypothyroidism, hearing loss, and others. The ocular findings include nystagmus 29%, esotropia 26%, epiphora 21.5%, Brushfield spots 16.9%, lens opacity 12.3%, abnormalities of the retinal vessels, foveal hypoplasia or retinal pigment epithelial hyperplasia 32% and optic disc pallor 7.6%. Hyperopia was present in 36.9% and was the most frequent refractive error in the group followed by astigmatism 29.2% and myopia 24.6%. No diagnosed systemic abnormalities were found in 18.3% of the children, while 30.7% had congenital heart defects.

Hypothyroidism, hypotony, hearing loss, gastrointestinal tract malformations, and leukemia were less common. Nystagmus was related to myopia and esotropia, and to heart disease and heart operations.

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