Purpose: To delineate the role of impaired central processing in supra-nuclear motion circuits versus abnormal static torsion resulting in altered direction of recti muscle pull.

Methods: Eye movements were measured using high-precision video-oculography (VOG) (Eyelink 1000, SR research) in subjects with infantile pattern strabismus (IPS n=3), late onset pattern strabismus (LPS n=2), infantile comitant strabismus (IS n=1), late onset comitant strabismus (LS n=4) and from healthy controls (C n=15). Horizontal and vertical eye positions were simultaneously recorded during fixation, saccades, step-ramp pursuit and vestibulo-ocular reflex (VOR). Static torsion was measured with fundus photography.

Results: Oculomotor responses in the plane orthogonal to desired movements (cross-coupled responses) were seen in patients with pattern strabismus. The cross-coupled response was larger in IPS than LPS. The cross-coupled responses were evident during saccades more than smooth pursuit but not during VOR. There was a greater amount of static torsion as measured by fundus photos in IPS compared to LPS. The amount of cross coupled responses in human pattern strabismics was smaller compared to non-human primates reared by alternate monocular occlusion.

Conclusions: Cross coupled responses were seen in pattern strabismics and its amplitude varied with different class of eye movements. Infantile onset pattern strabismus could be due to abnormal neural connections whereas late onset pattern strabismus or pattern strabismus in patients with craniofacial anomalies could be due to orbital pulley instability.

Commercial Relationships: Fatema Ghasia, None; Aasef Shaikh, None; Mark Walker, None

Support: Knights Templar Pediatric Eye Research Award, Fight for Sight Grant in Aid

Program Number: 2572 Poster Board Number: C0276
Presentation Time: 3:45 PM–5:30 PM

Abducens Neuron Responses in Monkeys with Strabismus
Mehmet Agaoglu1, 2, Anand C. Joshi1, Sevda Agaoglu1, 2, Vallabh E. Das1. 1College of Optometry, University of Houston, Houston, TX; 2Electrical and Computer Engineering, University of Houston, Houston, TX.

Purpose: Disrupting binocular vision during the first few months of life in a monkey results in strabismus. The objective of this study was to investigate response properties of abducens motoneurons (ABN) in relation to horizontal misalignment in monkeys with strabismus.

Methods: Burst- tonic (BT) activity of 49 neurons in the abducens nucleus (17-Left Abducens LTB; 32-Right Abducens RTBT) was recorded from one strabismic monkey (OD: ~30° XT; OS: ~15° XT) during horizontal smooth pursuit (0.2 Hz, ±15°) under each monocular viewing condition. Neuronal firing rates (FR) and horizontal component of eye position and velocity (Epos, Eneg) were used to identify regression coefficients (K-position, R-velocity, C-constant) in a first-order model (FR = K*Epos + R*Eneg + C) for each tracking condition.

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Fixation Stability in Monkeys with Strabismus
Onkar H. Pirdankar, Harold E. Bedell, Vallabh E. Das. College of Optometry, University of Houston, Houston, TX.

Purpose: To assess the effect of fixation target parameters on fixation stability of the viewing and fellow eyes in normal and strabismic monkeys.

Methods: One normal and three exotropic monkeys were presented with four differently shaped fixation targets (x, disk, disk + cross, and %), with diameters of 0.5°, 1° or 2°, during monocular or binocular viewing. Fixation targets were white on a black background or vice versa. Eye movements were recorded using the magnetic search coil technique and fixation stability quantified by calculating the Bivariate Contour Ellipse Area (BCEA). Multi-factorial ANOVA was performed to analyze the effects of target shape, size, background and viewing condition on BCEA.

Results: BCEA was greater (more fixation instability) in the three strabismic monkeys compared to the normal monkey. The BCEAs of the covered (deviated) eye of the strabismic monkeys was significantly greater than the BCEA of the fixating eye (paired t-test p<0.001), but not in the normal monkey. Target shape and size significantly affected fixation stability in all monkeys. A disk-shaped target resulted in significantly higher BCEAs than other target shapes in the viewing eyes of the normal monkey and 2/3 strabismic monkeys (p<0.001). A similar target-shape effect also was observed in the covered eye (p<0.03). Best fixation was elicited with a 0.5° target in the normal monkey and a 1.0° target in the strabismic monkeys, both in the viewing (p<0.005) and the covered eye. Background effects were idiosyncratic among the monkeys. Fixation stability was similar during monocular and binocular viewing in all monkeys. Analysis of the BCEA ratios for the viewing and covered eyes revealed no significant effect of target shape in any monkey and no significant effect of target size except in one monkey, suggesting proportional changes in the stability of both eyes due to shape and size.

Conclusions: The difference in fixation stability between normal and strabismic monkeys is likely due to underlying amblyopia, abnormal drifts and nystagmus eye movements. Target parameters (shape and size) that influence fixation stability in a normal animal also affect fixation stability in strabismus. The influence of target parameters on fixation likely functions via conjugate mechanisms, as proportional effects were observed in both eyes of all monkeys.

Commercial Relationships: Onkar H. Pirdankar, None; Harold E. Bedell, None; Vallabh E. Das, None
Support: NIH Grant R01 EY015312; NIH Grant R01 EY022723; NIH Grant P30 EY07551

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Presentation Time: 3:45 PM–5:30 PM

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Program Number: 2575 Poster Board Number: C0279

Postural control in children and young adults with vergence insufficiency
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**Purpose:** To study the postural control in children and young adults with vergence insufficiency. Deficits of vergence anomalies and ocular instability or misalignment can influence postural control. Vergence deficits that deteriorate gaze stabilization during body displacement had been described to cause blurry vision and vertigo.

**Methods:** A posturography system (Dynaport) was used to examine posture in quiet stance. The child was asked to fixate a target at 40 cm or at 200 cm, either with eyes open (vision condition) or with eyes covered by a monocular (monocular vision condition) or a binocular cover (no vision condition). During 30 sec data were recorded by Mira 2 software. Eighteen children and young adults (range 6 to 35 year-old) were selected and tested, 8 presenting vergence insufficiency and 10 normal subjects. The patients had complete ocular and orthoptic evaluation examination initially and repeated at 3 months after treatment including orthoptic reeducation.

**Results:** Ocular examination assessed the vergence insufficiency of the 8 subjects and hyperopia was present in 7 of them. Before treatment, amplitude of postural movements in subjects with vergence insufficiency was significantly larger than in normal subjects. Of the 8 subjects with vergence insufficiency, 6 presented a significant data improvement of amplitude of postural movements. Data with different binocular, monocular and no vision conditions are detailed and analysed.

**Conclusions:** Binocular visual information, such as vergence disparity, is essential in stabilizing posture at far and near distance. Postural instability reported in children with vergence abnormalities could be due to poor vergence inputs and/or to immature compensatory mechanisms controlling postural stability as vestibular, somatosensory inputs and/or cerebellar processes. This simple technique can be useful to explore and evaluate abnormal binocular conditions before and after treatment. Further studies are needed to evaluate binocular pathologies including vergence anomalies and strabismus.

**Commercial Relationships:** Dominique Bremond-Gignac, None; Julien Ricard, None; David Rivalan, None; Clement Dhainaut, None; Pascal Louage, None; Zoi Kapoula, None

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Program Number: 2576 Poster Board Number: C0280

Presentation Time: 3:45 PM–5:30 PM

Fixational eye movements when viewing focused and defocused images
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**Purpose:** To determine whether the patterns of fixational eye movements are altered when images are defocused (low pass-filtered).

**Methods:** Six subjects (3 male, 3 female, 22 - 32 years of age, Caucasian origin) were asked to inspect an image on the computer screen which was either “in focus” or low-pass filtered by convolution with a point spread function that was equivalent to 5D of myopic defocus. The screen was at 79 cm distance and one pixel on the screen subtended a visual angle of 1.45 arcmin. The movements of the right eye were tracked at 166 Hz sampling rate in a highly magnified video image (39.9 pixels/mm) by custom-developed software (Visual C++ 8.0). A commercial IR camera (DMK 22AU03, www.theimagingsource.com) was used and a single IR LED that generated a bright first Purkinje image. The angular resolution in natural eyes was determined to be 1-2 arcmin. Differences in eye movements variables were determined in the pooled data of all subjects using t-tests, analyzing the horizontal and vertical components of the eye movements separately.

**Results:** The frequency of horizontal microsaccades (defined as saccades with an amplitude between 20 and 120 arcmin) increased from 3.3±2.9 to 6.3±4.4 in low-pass filtered images recorded over 4 secs (p<0.05). The frequency of vertical microsaccades increased with the low pass-filtered image from 1.1±1.7 to 3.1±3.6 in 4 secs (p<0.05). Vertical saccades were generally smaller than horizontal ones (p<0.01). The amplitudes of horizontal microsaccades increased from 48.1±17.5 arcmin to 68.4±29.4 arcmin (p<0.01). Amplitudes of vertical microsaccades did not change (34.8±13.4 to 39.8±16.8 arcmin). The horizontal “diameter of the distributions of gaze during fixation” (described by Cherici et al, J Vis 2012, who found about 18 arcmin) changed from 15.3±6.0 arcmin to 17.6±13.1 arcmin (p=0.34; one subject p<0.01, one subject p<0.05) and vertically from 11.7±5.1 to 12.3±6.8 arcmin (p=0.69; one subject p<0.01).

**Conclusions:** The spatial frequency content of an image has impact on the pattern of fixational eye movements in humans. Accordingly, we expect that uncorrected myopic subjects have different patterns of fixational eye movements.

**Commercial Relationships:** Ulrich Wildenmann, None; Frank Schaeffel, None; William R. Bobier, None

**Support:** PITN-GA-2010-264605

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Program Number: 2577 Poster Board Number: C0281

Presentation Time: 3:45 PM–5:30 PM

Visual perception and oculomotor control are robust to acute hypoxia
Charlotte J. Connell1, Jhordaine B. Charlton1, Clare E. Turner1, Joanna M. Black2, Gustav Kuhn2, Michael P. Claffey2, Benjamín Thompson2, Nicholas Gam1. 1Exerc. Neurometabolism Lab., Univ. Auckland, Auckland, New Zealand; 2Dep. Optometry & Vision Sci., Univ. of Auckland, Auckland, New Zealand.

**Purpose:** Hypoxia impairs cerebral metabolism and represents a significant challenge to brain homeostasis and cognitive function. Therefore, experimentally-induced hypoxia provides a model for brain injuries that impair cellular energy provision. The aims of this study were 1) to investigate the effect of acute, severe hypoxia on visual perception and saccadic eye movements, and 2) compare the effects of hypoxia on visual and cognitive function.

**Methods:** Within a single-blind, randomized, matched-pairs design, 26 healthy participants (10 male) inspired either a hypoxic (FiO2 = 0.10) or sham (FiO2 = 0.21, atmospheric air) gas mixture for 90 min. Saccadic eye movements, assessed within the context of an established social attention paradigm, were measured at baseline and intervention. Global motion perception and cognitive performance were assessed using random dot kinematograms and the CNS Vital Signs test battery respectively. In addition, chromatic discrimination (Farnsworth-Munsell 100-hue test), perceived symptoms of hypoxia (environmental symptoms questionnaire), cardiovascular responses and oxygen saturation were recorded at regular intervals throughout the protocol.

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Results: Hypoxia significantly reduced arterial oxygen saturation (SaO2 = 79 ± 6%, p < 0.001) and severely impaired a wide range of cognitive processes, compromising neurocognitive index scores by -20 ± 13% (p = 0.025). Saccadic eye movements (peak velocity, latency and accuracy), global motion perception, and chromatic discrimination were not affected by hypoxia. However, visual disturbances including blur and dimmed vision were reported on the environmental symptoms questionnaire during the hypoxic condition. Conclusions: Oculomotor control and visual perception are robust to a severe hypoxic insult that significantly impairs higher order cognition. Visual disturbances reported by participants during hypoxia did not influence objective measures of visual function. Energy provision to networks responsible for controlling saccadic eye movements, global motion perception and color perception appear to be protected during a severe, acute oxygen deficit.

Commercial Relationships: Charlotte J. Connell, None; Jhordaine B. Charlton, None; Clare E. Turner, None; Joanna M. Black, None; Gustav Kuhn, None; Michael P. Claffey, None; Benjamin Thompson, US12528934 (P), US8006372B2 (P); Nicholas Gant, None

Program Number: 2578 Poster Board Number: C0282
Presentation Time: 3:45 PM–5:30 PM

Gaze tracking of pitched balls through apertures

Nicholas Fogt1,2, Marc A. Burcham3. 1College of Optometry, Ohio State University, Columbus, OH; 3Ophthalmology, Nationwide Children’s Hospital, Columbus, OH.

Purpose: It has been shown that experienced baseball players tend to move the head significantly and the eyes very little when tracking pitched balls. Attempting to track pitched balls through apertures in front of the eyes might force individuals to adopt the pattern of head-eye coordination used by experienced players. The purpose of this experiment was to determine whether individuals would adopt the appropriate pattern of head and eye coordination (head movement with rotational vestibulo-ocular reflex (RVOR) suppression) to view pitched balls through small apertures.

Methods: Data were gathered from 10 subjects with variable baseball experience. Subjects viewed tennis balls (76mph) thrown by a pneumatic pitching machine 43.6ft away. Head movements (inertial sensor) and (left) eye movements (video tracker) were recorded. Head, eye, and ball positions were synchronized by software. Two tracking trials (50 pitches each) were utilized. In the first trial (no-aperture/NA condition), subjects attempted to keep their eyes on the ball as long as possible. In the second trial (aperture/A condition), subjects attempted to keep the eyes pointed at the ball while wearing small (3.3deg) apertures centered over both eyes.

Results: Blink-related data were discarded. Data were analyzed at 300ms after pitch release (ball 8 ft from the subject). Head rotation, eye rotation, and gaze (head+eye) errors were calculated. Mean eye rotations (mean NA = 19.4±9.8deg) were always in the direction of the ball and always much larger than mean eye rotations in both conditions. Depending on the subject, mean eye movements could be in the direction of the ball, opposite the ball, or near zero. Four of 10 subjects made mean eye movements less than 1.7deg (half-angle of apertures) with the apertures. There were no statistically significant (p<0.05) differences in the NA and A conditions for mean head movements, mean eye movements, or mean gaze errors (paired t-test).

Conclusions: Four of 10 subjects suppressed the RVOR sufficiently to maintain the visual axis through the aperture. However, there were no statistically significant differences between the mean head movements, mean eye movements, or mean gaze errors in the NA and A conditions. Taken together, these results suggest that the apertures resulted in no consistent changes in head and eye movements. Subjects generally showed similar head and eye tracking behaviors with and without the apertures.

Commercial Relationships: Nick F. Fogt, Patent #8553936 (P); Marc A. Burcham, None

Support: Optometric Educator’s Incorporated
Plasticity of the Vergence Response

Lynn D. Greenspan. Foundations Of Optometric Medicine, Salus University, Elkins Park, PA.

**Purpose:** The purpose of this study is to assess the correlation between findings of autonomic dysregulation and post-concussion vision symptoms. Traumatic brain injury is associated with autonomic dysregulation (dysautonomia) causing storms of excess sympathetic tone also known as paroxysmal sympathetic hyperactivity (PSH). A high baseline state of hyper-arousal and decreased parasympathetic activity can be life-threatening in severe TBI. In this study we propose that a mild form of PSH or dysautonomia found in mild traumatic brain injury is associated with visual symptoms in post-concussion syndrome (PCS). Exaggerated sympathetic tone creates a fight or flight response which opposes the near triad reflex. Increased effort to counteract the excess sympathetic tone results in asthenopia from accommodative, convergence and ocular motor dysfunction in PCS.

**Methods:** A retrospective cohort study was performed on the vision clinic records from 2010-2013 in a rehabilitation hospital. Inclusion criteria: 1) records of diagnosed post-concussion syndrome 2) visual findings of large or minimally reactive pupils, decreased accommodation and convergence, photophobia, peripheral visual field constriction or motion sensitivity 3) symptoms of autonomic dysregulation, including dizziness, orthostasis, erratic heart rate or pulse, exercise intolerance, pause, anxiety, pain syndromes, or temperature dysregulation. Three symptom lists were generated based on established symptom surveys: (1) Post Concussion Symptom Survey, PCSS (2) Brain Injury Vision Symptom Survey, BIVSS and (3) a dysautonomia symptom survey. Key data points included (1) the number of subjects with both dysautonomic symptoms and post-concussion vision findings (2) the correlation between symptoms in each condition (3) the predictive value of key vision findings most highly correlated to autonomic dysregulation.

**Results:** Of 712 records, 309 were post-concussive and 207 of those had at least 8 symptoms or findings related to acquired dysautonomia (67%). 190 of the 207 (91.7%) had all 5 key vision findings that most highly correlated to autonomic dysregulation.

**Conclusions:** Acquired dysautonomia explains post-concussion vision findings and in mild traumatic brain injury. Future studies will assess the benefits of treatment modalities effective in vision symptoms and findings in mild traumatic brain injury. Future studies will assess the benefits of treatment modalities effective in vision symptoms and findings in mild traumatic brain injury. Future studies will assess the benefits of treatment modalities effective in vision symptoms and findings in mild traumatic brain injury. Future studies will assess the benefits of treatment modalities effective in vision symptoms and findings in mild traumatic brain injury.

**Commercial Relationships:** Lynn D. Greenspan, None

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Plasticity of the Vergence Response

Muriel Dysli1, 2, Mathias Abegg1.

**Purpose:** Vergence movements are slow disconjugate eye movements which may be triggered by image disparity or accommodation. There exist numerous clinical contexts where image disparity may vary with the direction of gaze. A common example is a sixth cranial nerve palsy with increasing image disparity in gaze toward the affected muscle. Adaptive changes to such incomitant image disparity have been poorly investigated and are the scope of this study.

**Methods:** Vergence stimuli of gaze dependent magnitude were used to mimic the image disparity of an incomitant strabismus. In a first experiment prisms were placed such that stimuli were viewed through the prisms in one gaze direction but not in the other gaze directions. In a second experiment we used a haploscope to modify image disparity according to gaze. We measured vergence responses that were made after a saccade shifting gaze from left to right, with increased image disparity in right gaze. We analysed changes of rise time or slope, latency, and amplitude over time.

**Results:** Increased image disparity in right gaze led to a decrease of vergence rise time (p=0.055) and latency (p=0.048) within minutes. Using the haploscope to deliver vergence stimuli, we again found a significant increase in slope (p=0.001), but not in latency (p=0.336).

**Conclusions:** In this study we show that repetitive increase of the vergence demand leads to rapid improvement of the vergence response kinetics with a moderate effect on the latency. This novel type of vergence plasticity helps to rapidly restore stereovision after a saccade is made into a field of gaze with increased image disparity.

**Commercial Relationships:** Muriel Dysli, None; Mathias Abegg, None

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Head oscillations in infantile nystagmus during tasks of varying visual demand

Frank A. Proudlock, Vijay Patel, Rebecca J. McLean, Irene Gottlob. Ophthalmology, University of Leicester, Leicester, United Kingdom.

**Purpose:** Involuntary head oscillations are often associated with infantile nystagmus (IN) although the nature and cause of the oscillations is unclear. Head oscillations in IN increase during tasks with higher visual demand, however it is unclear if this simply reflects normal patterns of head tremor or whether the oscillations provide any benefit in reducing nystagmus. We compare head and eye oscillations in IN volunteers with age-matched controls during tasks of varying visual demand.

**Methods:** Horizontal and vertical head and eye oscillations were measured at 1000Hz using a head and pupil tracker in 17 IN participants (10 with albinism, 6 idiopathic, 1 acomitant) and 14 controls when performing two tasks of high visual demand (visual acuity and reading tasks where letters become progressively smaller) and three tasks of low visual demand (eyes closed, fixation task, fixation with mental maths task). The amount of head oscillations were quantified by summing the area under the curve of power spectral density functions between 1-10Hz. Nystagmus characteristics were evaluated during the visual acuity and fixation tasks and were compared when the head was fixed using a bite bar.

**Results:** A larger degree of head oscillations were observed in both the IN and control group when performing the two tasks of higher visual demand compared to the fixation and eyes closed task. The differences were highly significant in the IN group (p<0.005 for all comparison) but only some comparisons reached significance in controls as larger head oscillations were observed in the IN group overall. Nystagmus intensity and foveation characteristics were not significantly different between tasks although the nystagmus frequency was slower during the fixation tasks compared to the visual acuity tasks (p<0.05). Nystagmus oscillations were even slower during the mental maths task. Fixing the head did not significantly change the nystagmus.

**Conclusions:** Similar patterns of head oscillations were observed in both the IN and control group with some differences in controls as larger head oscillations were observed in the IN group overall. Nystagmus intensity and foveation characteristics were not significantly different between tasks although the nystagmus frequency was slower during the fixation tasks compared to the visual acuity tasks (p<0.05). Nystagmus oscillations were even slower during the mental maths task. Fixing the head did not significantly change the nystagmus.

**Commercial Relationships:** No
Commercial Relationships: Frank A. Proudlock, None; Vijay Patel, None; Rebecca J. McLean, None; Irene Gottlob, None

Support: Ulverscroft Foundation

Program Number: 2583 Poster Board Number: C0287

Presentation Time: 3:45 PM–5:30 PM

Prevalence of Gaze Apraxia in Children with Cerebral Palsy

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Purpose: Gaze apraxia is difficulty in initiating and executing saccades, evident as abnormally long latencies and/or subnormal saccade amplitude. The purpose of the current study was to determine the prevalence and severity of gaze apraxia in children with different severities of cerebral palsy (CP).

Methods: Horizontal and vertical visually evoked saccade testing was conducted by clinical observation in 131 children with CP. Stimuli were positioned ± 20° from central gaze as the child viewed binocularly. OKN fast-phases were quantified using large-field stripe motion and laboratory eye movement recordings in an additional 19 CP children. Responses were compared to a control group of 32 normal children.

Results: Gaze apraxia prevalence for CP children was 55%. Prevalence was linked to severity of CP (Gross Motor Function Classification Systems [GMFCS] level 1 = mildest to 5 = most severe). Gaze apraxia was detected in 23% of GMFCS 1 children. Prevalence increased systematically with increasing GMFCS levels (chi square, p < .01) to 87% at GMFCS 5. OKN fast-phase frequency and amplitude in CP children across all GMFCS levels was 36% of control values.

Conclusions: Gaze apraxia, evident as saccadic or OKN fast-phase dysfacility, is evident in over one-half of children with CP. The prevalence of the apraxia increases with severity of CP. The apraxia degrades visual function, which is dependent upon prompt and accurate foveation of targets at different locations.

Commercial Relationships: Daniel Greenstein, None; Lawrence Tychsen, None

Program Number: 2584 Poster Board Number: C0288

Presentation Time: 3:45 PM–5:30 PM

The long term progression of eye-movements in relationship to birth order in children

Christine L. Allison, Darrell G. Schlange. Pediatric Optometry & Binocular Vision, Illinois College of Optometry, Chicago, IL.

Purpose: The purpose of this longitudinal study was to analyze eye movement data related to birth order in a group of children upon entering Kindergarten, and then again upon entering 3rd grade. We have previously reported on a significant difference in eye movements among the pre-Kindergarten groups of children with no siblings, first born with siblings, and children who are not first born and have siblings. The findings have led to the theory that first born children and children with no siblings may exhibit better saccades and fixation control prior to entering Kindergarten due to differences in the type of activities they pursue. Once children are in school, their activities are more regulated, thus we theorized that the same children prior to entering 3rd grade would not show a significant difference in eye movements.

Methods: 112 children with similar academic/socioeconomic backgrounds were examined the summer prior to Kindergarten. 33 of the same children have been examined again upon entering 3rd grade. The children were given comprehensive exams including tests of accommodation and vergence. The children also received eye-movement analysis with the Visagraph visual skills protocol. The caregivers completed a survey regarding the number/ages of siblings, prior school history, and amount of time spent on tasks like reading, using computers, and playing outdoors.

Results: Children in the pre-Kindergarten group who were first born exhibited better fixation control with fewer off-target drifts (F 6.09, p = <0.05) and more efficient horizontal saccades (F 5.96, p<0.05). The Bland-Altman analysis of the 3rd grade group indicated good agreement among all groups for saccadic accuracy, saccadic speed, fluency, and fixation accuracy.

Conclusions: The activities that first born children are encouraged to perform regularly prior to Kindergarten may lead to better eye movement skills at that age. However, by 3rd grade, a child is using eye-movements to learn, and uses visual tracking, saccadic sequencing and reading fluency to read at a higher level. Our data suggests that the first born group continues to have moderately improved eye-movement performance compared with the others, but not significantly better. The children in this study will be re-evaluated prior to 6th grade to determine if any new trends with eye movements or binocular stability develop.

Commercial Relationships: Christine L. Allison, None; Darrell G. Schlange, None

Program Number: 2585 Poster Board Number: C0289

Presentation Time: 3:45 PM–5:30 PM

Ocular Motility Disturbances in Orbitofacial Neurofibromatosis

Type 1

Thomas M. Bosley1, Ibrahim A. Aloraonen, Jose Morales1, Imtiaz A. Chaudhry1, Sahar M. Elkhamey1, Darren T. Oystre1.

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Purpose: To review ocular motility disturbances in patients with orbitofacial neurofibromatosis type 1 (OFNF) who have neurofibromas on the lid, brow, or face during infancy or early childhood. OFNF is commonly associated with enlargement of the ipsilateral globe, sphenoid and orbital dysplasia, and neurofibromas involving the orbit and cavernous sinus, all of which can affect ocular motility.

Methods: Retrospective medical record review and re-examination of selected patients from one institution.

Results: Forty-nine patients met National Institutes of Health criteria for OFNF and had adequate clinical and neuroimaging information to assess ocular motility and factors affecting eye movements. Fourteen patients had no strabismus, all of whom had good vision with no ducational abnormalities on either side despite large globes, sphenoid dysplasia, and neurofibromas in the orbit and/or cavernous sinus in many. Eight patients with comitant strabismus also had no ducational abnormalities with a similar constellation of anatomic abnormalities, but these patients all had poor vision in at least one eye. Twenty-seven patients had incomitant strabismus, all with downward displacement of the globe and ducational limitations (Figure).

Conclusions: Pathologic anatomic changes associated with OFNF do not always cause ocular motility abnormalities. Strabismus generally was not present when ocular motility was full and visual acuity was good despite an enlarged globe, substantial distortion of orbit and sphenoid anatomy, and tumor in orbit and/or cavernous sinus. Comitant strabismus occurred with full ocular motility and reduced vision in at least one eye. Incomitant strabismus was always accompanied by reduced vision and a ducational abnormality in one or both eyes that in general seemed due to anatomic abnormalities of the orbit and skull rather than to partial or complete cranial nerve palsy.

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Unfortunately, the causes of ductional abnormalities in OFNF are currently difficult or impossible to treat.

External photo in (A) primary gaze, (B) right gaze, (C) left gaze, (D) up gaze, and (E) down gaze. (F) Axial T2-weighted MR image showing left sphenoid dysplasia causing mass effect on the extraocular muscles and optic nerve (open arrow). (G) Enhanced axial T1-weighted image showing plexiform neurofibroma involving the left orbit and eye lid (open arrow). (H) Coronal T1-weighted image showing left sphenoid dysplasia (open arrow).

Commercial Relationships: Thomas M. Bosley, None; Ibrahim A. Alorainy, None; Jose Morales, None; Imtiaz A. Chaudhry, None; Sahar M. Elkhamary, None; Darren T. Oystreck, None

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