Mastery and self-esteem mediate the association between visual impairment and mental health: outcomes of a population-based longitudinal cohort study

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Purpose: The high prevalence of mental health problems in older adults with visual impairment is well known. However, to determine the optimal approach of treating these problems, it is necessary to gain more insight into the association between visual impairment and mental health. We provide novel insights into this association by using longitudinal data, studying the association for reversible and irreversible vision loss separately, and investigating the potential mediating roles of mastery and self-esteem.

Methods: A longitudinal population-based cohort study was performed, using data from the Longitudinal Aging Study Amsterdam, collected from 2001 until 2012. Participants (n=2599, mean age 72 years, 56% female, 1.2% irreversible- and 0.3% reversible vision loss) participated in measurements four times over a period of 10 years. Factor analysis and Item Response Theory were used to determine psychometric properties of the main outcome measures. Associations between visual acuity, depression, anxiety, mastery, self-esteem and mediation analyses were performed using linear mixed models.

Results: Better visual acuity was associated with significantly less depression (β=0.24, p<0.001), greater mastery (β=0.48, p<0.001) and higher self-esteem (β=0.17, p<0.01). No significant association with anxiety was found (β=0.03, p=0.58). When self-esteem was included as a mediator, the association between visual acuity and depression was suppressed but still significant (β=0.16, p<0.01), indicating partial mediation with a mediated proportion of 25%. When mastery was included as a mediator, the association was no longer significant (β=0.05, p=0.43), indicating full mediation with a mediated proportion of 75%. Compared to irreversible vision loss, reversible vision loss was associated with significantly less depression and anxiety.

Conclusions: Based on these results we can conclude that especially people with irreversible vision loss tend to experience mental health problems. Moreover, self-esteem and mastery play a significant role in the association between visual impairment and mental health problems. People with lower mastery and self-esteem could be identified as those at risk of developing mental health problems and treatment could be targeted at increasing mastery and self-esteem to prevent and reduce these problems in this population.

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Ivar Maaswinkel, None; Ger van Rens, None; Ruth M. Van Nispen, None

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Predictive value of self-report psychological status in comparison to Geriatric Depression Scores in visually impaired patients

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Purpose: Depression is a common co-morbidity among patients with visual impairment, yet detecting patients at risk can be time-consuming and cumbersome with administering formal questionnaires. Our objective is to determine the predictive value of self-report psychological status obtained through a single intake question as compared with the Geriatric Depression Scale (GDS).

Methods: A total of 764 patients were recruited from 28 low vision rehabilitation (LVR) centers in the United States. Eligibility criteria required participants to be new patients to the LVR center. Among the series of questionnaires that were administered via telephone before the initial clinical examination was a standardized “check-box” intake survey that included a single question asking patients to “describe their current emotional state” and the 15-item GDS. The GDS was used as the gold standard to assess depression severity and outcome variables were estimated using Rasch analysis.

Results: Distributions of GDS severity measures of individual patients who reported being depressed, sad, anxious, and/or well-adjusted on the intake were greater than those who did not report the same symptom (p<0.005). A General Linear Model (GLM) was used to estimate the predictive weight of each of the self-report intake indicators. The binary indicator weights are: depressed (b=0.95, p=0.002; OR=2.58); sad (b=0.88, p=0.005; OR=2.42); anxious (b=0.82, p=0.005; OR=2.27); not well-adjusted (b=0.79, p=0.012; OR=2.21). When analyzing the within patient agreement between the depression indicator model and clinical depression diagnoses based on the GDS, a weighted kappa of 0.385 was estimated.

Conclusions: Patients who report that they are depressed, sad, anxious, and/or not well-adjusted on an intake questionnaire have greater level depression severity based on GDS measures than patients who do not report these symptoms. On a population basis, the GLM shows good separation of depression severity distributions between patients who report these psychological indicators vs those who do not. However, at the level of the individual patient, although kappa is significantly greater than zero, concordance between these indicators and a depression diagnosis based on GDS criterion is weak.

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Blind registration in Trinidad

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Purpose: Little is known about the causes and impact of blindness in the Caribbean. This study of the Trinidad Blind Welfare Association register explored historic causes of blind registration, and characteristics and experiences of registered clients.

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Purpose: Patients with low vision due to glaucoma have more difficulty performing daily activities, are at greater risk for injury, and suffer higher rates of depression, but these issues have not been adequately characterized. We performed a retrospective analysis of patients with a primary diagnosis of glaucoma presenting to a low vision clinic in order to describe their characteristics.

Methods: All patients with a primary diagnosis of glaucoma who presented to Hoover Low Vision Rehabilitation Services from August 2012 through September 2016 were included. Data collected included patient’s age, visual acuity, Pelli-Robson contrast sensitivity, visual field measures, and difficulty of various activities, including reading, watching TV, driving, ambulating, recognizing faces, and using a computer. Patients were also asked about depression and visual hallucinations. Descriptive statistical analysis was performed.

Results: Patient ages ranged from 22 to 103, with a mean of 77 years; only 13% were under 60 years of age. Visual acuity in the better-seeing eye ranged from 20/20 to worse than 20/400, with a median of 20/40. 54% of patients had visual acuity of 20/40 or better. However, 61% needed 4 times as much contrast as normal on the Pelli-Robson contrast sensitivity chart. Humphrey Visual Field (HVF) 24-2 available in 68 patients gave a median value of the mean deviation of -14.36 dB. Of 83 patients queried, 36% reported depression and 24% reported visual hallucinations. Of 85 patients who were drivers, 78% had stopped driving due to their vision. Of the 41 patients who used computers, 56% reported difficulty. Percentages of patients who reported difficulty with other tasks included 60% recognizing faces, 85% reading, 44% watching TV, and 66% mobility. Correlation between logMAR visual acuity and Pelli-Robson contrast sensitivity was -0.64, contrast sensitivity and HVF mean deviation was -0.24, and logMAR visual acuity and HVF mean deviation was -0.11.

Conclusions: This study quantifies the visual difficulties of glaucoma patients in a low vision setting. This information will give low vision providers a better understanding of this population’s unique needs and thus help patients achieve more independence in their daily life.

Commercial Relationships: Jennifer Wall, None; Michelle Bianchi, None; Janet S. Sunness, None

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Presentation Time: 11:00 AM–12:45 PM

Binocular contrast suppression in patients with glaucoma
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Purpose: Prior research suggests that binocular measures of vision equal or exceed measures obtained from each eye individually. Anecdotally, however, glaucoma patients sometimes express that a poorly-seeing eye can depress their binocular vision below the level of their better-seeing eye, a phenomenon referred to as suppression. We performed a cross-sectional, clinical study to look for evidence of suppression in a glaucoma population.

Methods: The contrast sensitivity function (CSF), measuring contrast sensitivity at varying letter sizes to model the area under the curve log CSF (AULCSF), was evaluated in 29 primary glaucoma patients with varied disease severity using the qCSF testing device (Adaptive Sensory Technology). Testing was performed in monocular and binocular conditions. Regression models were constructed in which the binocular - better-eye AULCSF difference was the dependent variable, inter-eye AULCSF difference was the independent variable, and age was a covariate. Patients were said to have a small inter-eye difference if right and left eye AULCSF values were within 0.3 and a large inter-eye difference if the values differed by more than 0.3.

Results: Subjects had a mean age of 69.6 (1.7) years and an average visual field mean deviation of -5.9 (1.4) and -13.1 (1.5) in the better and worse eyes, respectively. For all subjects, binocular AULCSF was 0.10 (0.12) greater than better-eye AULCSF. In the 15 patients with a small inter-eye difference, binocular AULCSF was 0.16 (0.13) greater than better-eye AULCSF, while in the 14 patients with a large inter-eye difference, binocular AULCSF was 0.03 (0.11) greater than better-eye AULCSF (p=0.003). No patient with a small inter-eye difference had worse binocular than better-eye AULCSF, while 7 of 14 patients with a large inter-eye difference had worse binocular than better-eye AULCSF (p=0.001). In regression models, each 0.1 increment in inter-eye AULCSF difference was associated with a 0.02 decrement in binocular - better-eye AULCSF difference (95% CI -0.03 to -0.002, p=0.024) and 1.43 higher odds of a worse binocular than better-eye AULCSF (95% CI 1.06 to 1.93, p=0.018).

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Conclusions: Glaucma patients with large visual differences from a poorly-seeing eye may experience suppression of vision in their better eye and lose binocular advantage when testing CSF. Better-eye monocular visual measures may not be an accurate representation of binocular vision in glaucoma patients.

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Presentation Time: 11:00 AM–12:45 PM
Point prevalence and incidence of visual impairment following stroke
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Purpose: To report annual point prevalence and incidence of visual impairment in an acute adult stroke population.

Methods: A prospective, epidemiology study was conducted from 1-6.14 to 30.6.15 across 3 stroke units. All stroke admissions were identified by the stroke research nurses. Deaths were coded. The orthoptic research team assessed all remaining patients on the stroke unit. Patients who could not be assessed were coded for reasons why. The remaining patients had standard clinical assessment of visual acuity, visual fields, ocular alignment, ocular motility, visual inattention and visual perception.

Results: There were 51% males and 49% females with mean age of 73.4 (SD 13.8) years. 1291 patients were recruited: 99 died before full assessment and 169 could never be assessed. Overall 1023 patients were assessed with diagnosis. Over half were assessed at baseline. 622 could not be assessed at baseline and were subsequently reviewed. 959 underwent full visual assessment at a mean of 19.4 days. 279/1023 (28%) had normal eye exams. 744/1023 (72%) had visual impairment: 55.8% with impaired central vision, 41.7% with eye movement abnormalities, 28% with visual field loss, 27.2% with visual inattention and 4.3% with visual perceptual disorders. 75/1023 (7.3%) had visual impairment due to pre-existent causes.

Conclusions: The point prevalence of post-stroke visual impairment in acute adult stroke survivors undertaking visual assessment is 73% with incidence of stroke-related visual impairment being 65.4%. This is higher than previous reports, and highlights the need for integration of visual assessment as a core post-stroke assessment. Full visual assessment possible for most stroke survivors by 4 days. Thus early visual assessment is feasible and important in that information can be provided on visual status and the functional significance of this to the stroke team, patients and carers.

Commercial Relationships: Fiona J. Rowe, Haag Streit AG (C); Lauren R. Hepworth, None; Kerry Hanna, None; Claire Howard, None
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Presentation Time: 11:00 AM–12:45 PM
Characteristics of a low vision population with geographic atrophy (GA) from age-related macular degeneration
Carol A. Applegate, Emuna Singman, Michelle Bianchi, Janet S. Sunness. Hoover Low Vision & Rehabilitation, Greater Baltimore Medical Center, Baltimore, MD; Ophthalmology & Visual Science, University of Maryland, Baltimore, MD; Stevenson University, Stevenson, MD.

Purpose: To describe clinical and visual function characteristics from a retrospective study of a GA low vision population.

Methods: New patients with GA without CNV in both eyes, and without other significant ocular disease, seen at our low vision clinic between July 2012 and September 2016 are included. Visual function measures were analyzed, including best-corretcted ETDRS visual acuity, Pelli-Robson contrast sensitivity, and MNRead testing. Optos SLO/TOC/micrometer testing was used to measure GA area and pattern.

Results: 91 patients met these criteria. 78% were women. Median age 86, with 17% younger than 80, 48% between 80 and 89, and 34% 90 or older. The median logMAR VA in the better-seeing eye was 0.47 (20/60), with 37% 20/40 or better, 47% between 20/50 and 20/100, and 15% worse than 20/100. The median difference in visual acuity between eyes of each patient was 2.6 lines, with 21% having between 3 and 5 lines difference, and 26% with 6 or more lines difference. Pelli-Robson log contrast sensitivity had a median of 1.13, with 45% requiring >=4 times more contrast than normal. For the 66 patients whose GA area could be measured, the area of GA in the less-involved eye had a median of 4.7 sq mm, with 32% having less than 2.5 sq mm, and 27% having greater than 7.5 sq mm. GA patterns for all eyes tested were multifocal in 27%, horseshoe in 15%, ring in 10% and solid in 47%. Median maximum MNRead reading speed in the 61 patients tested was 158 wpm, with 21% reading less than 100 wpm. 20% of patients had better reading rates at smaller character sizes than at the larger character sizes.

Conclusions: These data characterize a low vision GA population, and can serve as a basis for understanding the low vision needs of these patients and for comparing this group to treatment groups in clinical trials.

Commercial Relationships: Carol A. Applegate, Emuna Singman, Michelle Bianchi, Janet S. Sunness, Cell Cure (C), The Eye Machine (C), Acucela (C), Genentech (Roche) (C)

Program Number: 4665 Poster Board Number: B0572
Presentation Time: 11:00 AM–12:45 PM
Potential Mediators of the Relationship between Socioeconomic Status and Vision in People with Age-related Macular Degeneration
Bradley E. Dougherty, San-San L. Cooley, Ellen Segerstrom, Frederick H. Davidorf. College of Optometry, The Ohio State University, Columbus, OH; Ophthalmology, The Ohio State University, Columbus, OH.

Purpose: There is some evidence that lower socioeconomic status (SES) is related to increased age-related macular degeneration (AMD) prevalence. We recently found that poorer visual acuity (VA) with habitual correction was related to lower education level in a
sample of people diagnosed with AMD. The purpose of this study was to evaluate the potential roles of social support, smoking history, refractive correction status, and comorbidities in the relationship between SES and VA and self-reported visual functioning in patients from this sample.

**Methods:** SES was represented as highest level of education. The ENRICHD Social Support Inventory, Impact of Vision Impairment Scale, and Charlson Comorbidity Index were administered to patients with AMD. VA with habitual correction was measured using an ETDRS chart with letter-by-letter scoring. Smoking status was assessed (current smoker, former smoker, or never smoker), as was spectacle wear. Multiple regression models and Fisher’s exact tests were used to examine relationships among potential predictors, SES, and vision.

**Results:** 124 people with AMD (54% female, mean±SD age of 82±9 years, 47% receiving anti-VEGF injections at time of study visit) were recruited. The IVI and ESSI were scored using Rasch analysis. There was a significant univariate relationship between education level and both VA (p=0.009) and self-reported visual functioning (p=0.014), with SES remaining a significant predictor of VA after adjusting for age (p=0.04). There was no significant relationship between SES and comorbidities (p=0.429), social support (p=0.265), or history of smoking (p=0.304). Spectacle wear was significantly related to both SES (p=0.018) and visual acuity (p=0.001). SES was significantly related to response on the “accessing information” item of the IVI (p=0.01), with low SES related to lower scores.

**Conclusions:** Lower SES was related to poorer visual acuity in people with AMD. People in low SES categories were less likely to wear spectacles, and this was related to poorer visual acuity. We did not find relationships between SES and several potential mediators of the relationship, including social support, history of smoking, or comorbidities.

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**Support:** NIH K23 EY022940, The Ohio Lions Eye Research Foundation

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**Presentation Time:** 11:00 AM–12:45 PM

### How Many Patients in a Retinal Practice Have Low Vision?

**Donald C. Fletcher¹, ², Taylor D. Fletcher, Laura Walker².

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**Purpose:** To assess what proportion of patients being seen in a retinal practice could possibly benefit from referral to a low vision rehabilitation clinic based on visual acuity alone.

**Methods:** The records of 1017 consecutive unique patients seen over a 2 month period in a Florida retinal practice were reviewed. Patients best corrected visual acuity was recorded for each eye. Patients with visual acuity worse than 20/60 in each eye (ICD 9 category Low Vision) were noted and diagnosis recorded.

**Results:** Patient age median (range) was 75 (12 – 103) years with 55% female. Best eye corrected visual acuity for all patients median (range) was 20/30 (20/12 to HM). 18% were classified as low vision based on acuity, best eye median (range) 20/80 (20/70 to HM). Of these patients 51% had exudative AMD, 16% had atrophic AMD, 14% had diabetic retinopathy and 19% had some other diagnosis.

**Conclusions:** A very significant proportion of patients seen in a retinal practice are potential candidates for referral for low vision services. The majority of these patients with decreased acuity had AMD or diabetic retinopathy. These patients are likely to have significant reading problems which could be helped with low vision rehabilitation. This supports the assertion that retinal practices may be a good location to offer low vision rehabilitation services where these patients are already comfortable with receiving medical care.

**Commercial Relationships:** Donald C. Fletcher; Taylor D. Fletcher, None; Laura Walker, None

**Program Number:** 4667 Poster Board Number: B0574
**Presentation Time:** 11:00 AM–12:45 PM

### Visual acuity in Stargardt disease patients after age 40

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**Purpose:** In order to better define visual loss in Stargardt disease patients later in life, we investigated the most recent visual acuity in a cohort of Stargardt disease patients beyond 40 years of life.

**Methods:** The most recent best corrected visual acuities (BCVA, Snellen or Feinbloom Low Vision Chart) in the better-seeing eye of 225 patients with Stargardt disease over 40 years of age were recorded in this retrospective study. Also included in the analysis were the age of subjective onset for symptoms and duration of symptoms (“duration”=age-onset). The Mann-Whitney U test was used to compare differences between groups (BCVA), while the Spearman rank-order correlation was used to measure associations between continuous variables (BCVA versus age, onset, and duration of symptoms).

**Results:** The median age of the Stargardt disease patients was 53.1 years, with an interquartile range (IQR) of 46.9-61.3 and a range of 41 to 88 years. The median BCVA of all patients showed a bimodal distribution (shown in Figure 1), with a median BCVA of 1.00 logMAR (20/200 Snellen, IQR 0.22 to 1.10 logMAR). Twenty-five patients (11.1%) had worse than 20/400 vision, and ten patients (4.4%) had worse than 20/800 vision. In the subset of 19 patients with subjective onset of age 10 or younger, the median BCVA was 1.30 logMAR (20/400, IQR 1.00 to 1.60). Eleven of these 19 patients had BCVA of 20/400 or better, four were between 20/500 and 20/800, and the remaining four had BCVA worse than 20/800. There was only a modest correlation of BCVA with age (r=0.17, P=0.01), but there were stronger correlations between BCVA and both subjective onset (r=-0.62, P<0.001) and duration of symptoms (r=0.7, P<0.001).

**Conclusions:** Although many Stargardt disease patients lose visual acuity to the 20/200-20/400 range, and some lose visual acuity beyond 20/400, only a small number of our patients were found to have “counting fingers” vision or worse, even with early onset of symptoms. These numbers will be valuable in counseling Stargardt disease patients on visual loss and could have value in planning treatment trials.
Visually Impaired (VI) Paralympic skiers are classified

Frederick T. Collison

Static VA was quantifiable in 55 athletes (1.54 logMAR

A wide range of visual functions can be measured in VI

Patient satisfaction with the outcome of low vision

Support: Agitos Foundation 2015 Grant Support Program grant

Factors Influencing Goal Setting in Low Vision Rehabilitation

Theresa Smith1, Lisa Foret2, Guy Davis3, Robert W. Massof4.

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Purpose: Patient satisfaction with the outcome of low vision rehabilitation is linked to achievement of patient-identified goals. Patient and therapist perspectives frequently differ on whether or not a goal is achievable. Many of the factors affecting the therapist’s decision-making process have face validity or are supported by anecdotal evidence. The aim of this study is to elucidate factors individual therapists consider when deciding whether to include a patient-selected goal in the plan of care (POC).

Methods: An emergent mixed method study design was used. Baseline functional independence ratings (FIM scores) and expected FIM scores as a consequence of therapy were assigned by the therapist to each patient-selected goal for 45 home health care patients having low vision. Qualitative data were obtained using a survey of 11 plausible factors elicited from the therapist that influence goal setting. The 24 goals with higher probability of not being included in the POC were selected from the 50 goals of the Activity Inventory. These goals were then organized into 4 goal categories: ADLS, IADLS, Social Participation, and Leisure. The goal categories were populated with factors likely to affect goal setting. Concurrently, logistic regression analysis was performed on the therapist’s baseline and expected FIM ratings and on measures of mental and physical health for each patient-selected goal to determine how well each predicts that the goal will be included in the POC.

Results: The goal category most likely to have patient-selected goals excluded from the POC was IADLS with Leisure a close second. Logistic regression showed that the FIM score the therapist expected the patient to be able to achieve after rehabilitation is a very strong predictor of whether or not the goal is included in the POC (other measures of patient mental and physical health state were not predictive).

Conclusions: From qualitative analysis, we conclude that therapists give greatest weight to ADL- and social interaction-related goals in constructing the POC and less weight to IADL- and leisure-related goals. Based on quantitative analysis of therapist’s FIM scores, we conclude that estimates of the patient’s rehabilitation potential is the strongest factor for inclusion of the goal in the POC.

Commercial Relationships: Theresa Smith, None; Lisa Foret, None; Guy Davis, None; Robert W. Massof, None

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Presentation Time: 11:00 AM–12:45 PM

Stargardt over 40 years

BCVA in 0.3 logMAR columns

![Figure 1: Histogram of best corrected visual acuity (BCVA) in 225 Stargardt disease patients between 41 and 88 years of age. Note the bimodal distribution of visual acuity, with 38% of patients falling in the 20/15 to 20/50 range and 44% falling in the 20/125 to 20/400 range.](image)
Purpose: To evaluate the demographic, medical, and psychosocial variables associated with awareness of low vision services (LVS) among retina clinic patients.

Methods: An anonymous survey including multiple choice and Likert-style questions was created on Research Electronic Data capture (REDCap) software. Patients with best-corrected visual acuity (BCVA) 20/50 or worse in their better seeing eye were recruited from five retina provider clinics at an academic medical center during November and December 2016. All patients were interviewed by one of two interviewers regarding demographics, health status, awareness of and experiences with LVS. Ocular diagnoses and visual acuity were recorded. Logistic regression models will be used to identify statistically significant variables and patient LVS knowledge (p<0.05).

Results: To date, 48 patients were interviewed, including 30 women and 18 men with a mean age of 73 years (SD=14). Thirty (63%) patients were unaware of LVS. Mean age of the patients aware and unaware of LVS was 80.3 (SD = 10.3) and 68.7 (SD = 15) years, respectively (p=0.005). The majority (89%, n=22) of patients with a primary ocular diagnosis of age-related macular degeneration (AMD) were aware of LVS, while most patients with a primary ocular diagnosis of proliferative diabetic retinopathy (PDR) (77%, n=25) were unaware of LVS. The highest level of education attained was high school in 67% (n=20) of the patients unaware of LVS, compared to higher education in 78% (n=14) of the patients aware of LVS (p<0.05). Patients with more severe vision loss were more likely to be aware of LVS (p<0.001). Of the 30 patients who reported no knowledge of LVS, 27 (90%) had a BCVA < 20/50 but >20/200. Of the 18 patients who reported knowledge of LVS, 10 (56%) had a BCVA of 20/200 or worse.

Conclusions: The majority of patients were unaware of LVS. Higher education level, older age, more severe vision loss, and a primary ocular diagnosis of AMD were positively correlated with awareness of LVS. Further investigation in larger study populations is warranted in order to improve awareness and utilization of LVS.

Commercial Relationships: Ramunas Rolius, None; Mona Lotfi-pour, None; Michael Langue, None; Ingrid U. Scott, None

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Presentation Time: 11:00 AM–12:45 PM

Visual acuity in children with Cerebral Visual Impairment: Comparison of Cardiff cards to Lea single symbols with contour bars at 100% and 50% spacing

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Purpose: Cerebral visual impairment (CVI) is the leading cause of vision loss in children in industrialized nations, and results from an insult to the developing brain. There is still much to be learned about the visual impairments associated with CVI. Preferential looking procedures using gratings and the Cardiff picture cards successfully measure visual acuity in non-verbal patients; but both grating acuity and Cardiff cards tend to underestimate the visual acuity reduction associated with amblyopia. It is anticipated that Cardiff likely would underestimate visual acuity loss in children with CVI due to the lack of contour interaction effects. A comparison of Cardiff cards to Lea symbols with contour bars at 100% and 50% spacing was evaluated through a retrospective, observational clinical study.

Methods: Binocular visual acuity was measured during a comprehensive vision examination of 15 children (ages 4-18 years; mean 8.6 years) presenting with a diagnosis of CVI. Single Lea symbols with contour flanker bars at 50% and 100% spacing were presented in a two-alternative forced choice task using the apple and the house as the test optotypes. Binocular visual acuity was measured with the Cardiff Visual Acuity Test which required detection of a single shape on a grey background. Visual acuity thresholds were determined using the modified acuity card procedure by observation of the child’s eye gaze, pointing, and/or naming of the optotype.

Results: The ranges for visual acuity were: Cardiff logMAR 0 to 0.6; Lea 100% logMAR 0 to 1.18; Lea 50% logMAR 0.23 to 1.38. A Bland Altman analysis (mean vs. difference in visual acuity) comparing Cardiff to 100% Lea and 50% Lea was performed. Cardiff acuity underestimated the vision loss compared to 100% and 50% Lea by -0.35 logMAR and -0.54 logMAR, respectively. The difference in acuity measures increased with worse acuity.

Conclusions: For 93% of the patients with CVI, 50% and 100% Lea symbols yielded a more reduced acuity compared to the Cardiff Acuity Test; the latter only requires detection of the symbols, whereas the Lea symbols require discrimination between an apple and a house optotype. Reporting visual acuity in the presence of contour interaction in children with CVI would be helpful when making educational recommendations to the child’s parents and care team.

Commercial Relationships: Jasmine Junge, None; Deborah A. Orel-Bixler, None

Support: NIH T32 Institutional Training Grant
Conclusions: The functional ERG and Vep exams identify more (70.57%) retinal and visual pathway diseases than the funduscopy (47.05%), i.e. the functional alteration can be detected in advance compared to the morphologic alteration. Fundus examination performed using an indirect ophthalmoscope equipped with a videocamera provides a good and relatively inexpensive retinal phenotype characterization.

![Fundus picture in a 15-month-old child: a large macular dystrophy area is evident in a severe retinal dystrophy](image)

A highly reduced and delayed ERG is present in the same child of the previous figure.

**Commercial Relationships:** Giulio Ruberto, None; Rosanna Guagliano, None; Donatella Barillà, None; Margherita Bensi, None; Paolo E. Bianchi, None

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**Purpose:** Zika virus (ZIKV) has spread to several countries in the Americas, since the first reports from Brazil. Microcephaly, central nervous system, and eye abnormalities have been associated with ZIKV infection as a result of perinatal transmission and can impact visual development. We performed a prospective clinical study to evaluate the role of overplus prescription in improving visual function in infants with severe visual impairment due to congenital ZIKV infection.

**Methods:** Methods: We enrolled infants whose mothers had blood or urine specimens confirmed for ZIKV by reverse-transcriptase-polymerase-chain-reaction (RT-PCR) assays. Genetic diseases and other prenatal infectious diseases (i.e. rubella, cytomegalovirus, toxoplasmosis, and syphilis) were ruled out serologically. Detailed demographic, medical, and prenatal history information, as well as clinical findings were documented. Complete clinical examination, intra-cranial ultrasound, neurologic examination were also performed. External ocular examination, visual behavior, motility evaluation, as well as indirect ophthalmoscopy, were performed in all subjects. Cycloplegic refraction with phylephrine 2.5% and tropicamide 1% was performed at a minimum 3 months of age. Assessment of visual behavior included evaluation of the ability to perceive light, human face, fix and follow objects. Those unable to fix and follow were considered at high risk of low visual development and eligible for overplus of +3.00D prescription after 3 months of age and reassessed 3 months later.

**Results:** Results: Two hundred and six infants with suspected congenital ZIKV born between October 2015 and July 2016 were examined. ZIKV infection during pregnancy was confirmed by mother’s blood or urine RT-PCR in 91 patients. Forty eight (48%) were female. Of the 91, 64 had central nervous system (CNS) disorders; 14 had microcephaly and 18 had eye abnormalities. At 3 months, 7/91 were unable to fix and follow of which 6 had nystagmus and all 7 had severe CNS abnormalities. After 3 months of +3.00D overplus glasses, fix and follow was seen in 6 of the 7 patients.

**Conclusions:** Conclusion: Infants exposed to ZIKV prenatal infection are at risk of developing low vision from a combination of CNS and ocular abnormalities. Overplus correction with +3.00D after 3 months of age may compensate for loss of accommodation in infants with congenital ZIKV.

**Commercial Relationships:** Andrea Zin, None; Julia Rosseto, None; Irena Tsui, None; Olivia Zin, None; Joel Silveira, None; Zilton Vasconcelos, None; Maria Elisabeth Lopes Moreira, None

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**Purpose:** The management of visually impaired infants is difficult, especially concerning visual function and prognosis. We therefore retrospectively evaluated the results and the prognostic value of flash visual evoked potentials (VEP) in children with severe visual impairment or blindness.

**Commercial Relationships:** Andrea Zin, None; Julia Rosseto, None; Irena Tsui, None; Olivia Zin, None; Joel Silveira, None; Zilton Vasconcelos, None; Maria Elisabeth Lopes Moreira, None

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**Purpose:** Usefulness of flash-VEP in childhood visual impairment

**Commercial Relationships:** Klaus Rohrschneider. Department of Ophthalmology, University of Heidelberg, Heidelberg, Germany.
Methods: 105 infants with severe visual impairment or functional blindness were examined and mostly followed for further development. VEP recording was performed unilaterally and bilaterally using the standard ISVEC recommended electrode set-up with the medetec LED goggles and a medetec synergy system. We correlated VEP results with visual function and further visual outcome.

Results: In 15 of 45 patients with normal VEP only pupillary light reaction was detected, however 7 were less than 1 year of age. In contrast only 4 out of 55 children with missing VEP had reliable visual function, two of them measurable visual acuity. During follow-up extinguished VEP changed to normal examinations when visual function markedly improved in 8 patients. During follow-up, kids with initially maintained VEP mostly showed positive visual development.

Conclusions: Although flash-VEP has only limited value in differentiation of severe visual impairment and blindness, it may be useful for counseling those patients concerning further visual prognosis. For confirmation of blindness with the VEP it is necessary to have more than one exam with absent VEP because developmental changes may lead to normal results. Due to missing maturation of the optic nerve examination during the first year of life may be of limited value. However, abnormal VEP does not necessarily demonstrate poor prognosis, even in the first years of life.

Commercial Relationships: Klaus Rohrschneider, None

Program Number: 4675 Poster Board Number: B0582
Presentation Time: 11:00 AM–12:45 PM
Long-term Outcome of Conventional and Half-fluence Photodynamic Therapy for Chronic Central Serous Chorioretinopathy
BoKwon Son, Eung-Suk Kim, Seung-Young Yu. Kyung Hee University Hospital, Seoul, Korea (the Republic of).

Purpose: To evaluate the long-term efficacy and safety of conventional photodynamic therapy (PDT) and half-fluence PDT in chronic central serous chorioretinopathy (CSC)

Methods: Retrospective review of chronic CSC patients treated with conventional PDT or half-fluence PDT and a minimum follow-up of 12 months between October 2007 and June 2015. Best-corrected visual acuity (BCVA), central retinal thickness (CRT), neural retinal thickness (NRT), choroidal thickness (CT), recurrence of CSC after PDT and resolution of subretinal fluid (SRF) at 12, 24 and 36 months were assessed.

Results: The study included 52 eyes completed the 36-month follow-up. 28 eyes received conventional PDT and 24 eyes received half-fluence PDT. The mean follow-up period was 45 months. The mean logMAR BCVA improved significantly (P < .001), both in the conventional group (from 0.36±0.32 to 0.15±0.27) and in the half-fluence group (from 0.31±0.29 to 0.15±0.28) at 36 months, without significant difference between the 2 groups (p=0.711). The mean CRT decreased significantly (P < .001), both in the conventional group (from 395.2±185 to 205.2±45.8) and in the half-fluence group (from 341.8±97.9 to 205.6±50.1), at 36 months, without significant difference between the 2 groups (p=0.326). Both groups showed significant reduction in CT at months 36 after PDT with significant difference (p<0.001). At 36 months, all patients in the both groups had complete absorption of subretinal fluid (SRF). There were no recurrence of SRF during the follow-up.

Conclusions: Both treatments were effective and safe in chronic central serous chorioretinopathy treatment with a significant improvement in the long term, both anatomic and visual, without recurrence of SRF.

Commercial Relationships: BoKwon Son, None; Eung-Suk Kim, None; Seung-Young Yu, Zeiss (F), Norvatis (F), Allergan (F), Bayer (F)

Program Number: 4676 Poster Board Number: B0583
Presentation Time: 11:00 AM–12:45 PM
Anatomo-functional retinal changes after peeling and no peeling of the ILM
Paolo Corazza, Daniele Ferrari, Matteo Badino, Silvio Lai, Carlo E. Traverso. Clinica oculistica DiNOGMI University of Genova IRCCS AOSP UNIV IST GE, Genova, Italy.

Purpose: To analyze functional and anatomical different outcomes in patients affected by idiopatic epiretinal membrane (iERM) undergoing pars plana vitrectomy (PPV) 25 Gauge with and without ILM peeling.

Methods: This is a single center study. Patients affected by iERM were randomized in two groups: PPV associated with ILM peeling (Double Peeling group DP; n=10) and PPV without ILM peeling (Single Peeling group SP; n=10). A complete ophthalmological examination and best-corrected visual acuity (BCVA) evaluation by ETDRS charts were performed. Retinal sensitivity and microperimetric parameters were tested by MP1 microperimetry. Anatomical outcomes were analyzed with swept source optical coherence tomography (ss-OCT). One year after surgery Optical Coherence Tomography Angiography (OCTA) was performed. The scan of the superficial and of the deep capillary plexus was than elaborated by ImageJ software.

Results: After a 6 month follow-up, BCVA improved in all patient with no difference between the two groups (p = 0.671). A significant (p = 0.019) postoperative difference was observed in mean retinal sensitivity in the 4° central area. The distance between fovea and disc margin was decreased in the SP group (p = 0.007). Fixation index was also evaluated by the central dominance percentage. SP group showed an average improvement of 38.3± 20.56%, whereas DP group had a mean improvement of 8.4± 16.87% (p=0.002). OCTA showed mean FAZ of 0.215 mm² ± 0.058 and 0.112 mm² ± 0.013 in the SP and DP group respectively. The difference in FAZ measurement between groups is statistically significant (P < 0.001) in the superficial plexus only. There is no statistically significant difference in the foveal vessel density of deep plexus between groups (P=0.417).

Conclusions: In terms of improvement of visual acuity both surgical practices are effective. Mean retinal sensitivity in 4° central area and central fixation stability significantly improved more in patients treated with the SP technique. OCTA might become a useful tool for preoperative and postoperative evaluation of epiretinal membrane.

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Fig.1: Angio OCT of patients of single peeling group and the ImageJ modified image

Fig.1: Angio OCT of patients of double peeling group and the ImageJ modified image

Commercial Relationships: Paolo Corazza, None; Daniele Ferrari, None; Matteo Badino, None; Silvio Lai, None; Carlo E. Traverso, None
Short term Efficacy of intravitreal dexamethasone implant in patients with macular edema (ME) secondary to vascular retinal diseases

**Purpose:** Purpose: To evaluate prospectively the short term efficacy of intravitreal dexamethasone implant (IDI) in patients with macular edema (ME) measuring central macular thickness (CMT) and visual acuity modification up to one month. Secondary endpoints were the evaluation of choroidal thickness and the safety of the drug in terms of intraocular pressure and other ocular and systemic side effects.

**Methods:** Methods: In this prospective study, the authors observed 15 consecutive patients affected by retinal vascular diseases (diabetes, retinal vein occlusion) and macular edema (central macular thickness more than 300μ) treated by intravitreal injection of long release intravitreal dexamethasone (Ozurdex™). Fifteen eyes of 15 consecutive patients were included in the study. Central macular thickness and choroidal thickness were evaluated by enhanced depth optical coherence tomography (EDI-OCT). EDI-OCT, intraocular pressure (IOP) and best correct visual acuity (BCVA) were reported at baseline and after 3, 6, 24 hours, and then after 1 week and 1 month later.

**Results:** Results: We observed a reduction in CMT since the first measurement, after 3 hours from the injection and progressively continuing along at the next follow up, to one months. Choroidal thicknesses showed no significant changes from the baseline visit to the last follow up 1 month later post-injection (P > 0.05). Visual acuity gain shows a correlation with the reduction of CMT. We found a significant increase of BCVA after 3, 24 hours, 1 and 4 weeks of treatment. One month after IDI, CRT was 296.7±109.9 and was significantly lower compared with pretreatment value 506.7±143.1. No significant change in IOP after the treatment was observed. Secondary endpoint: no systemic neither ocular side effects were noticed.

**Conclusions:** Conclusion: The reduction in central macular thickness is appreciated as early as 24 hours after the injection of intravitreal Ozurdex with progressively reduction during the follow up of the study (up to one month). Choroidal thickness measurement seemed to be unchanged after IDI from baseline to end of study. No differences was observed between the group of DME and RVO. The study shows the early effect of IDI and it confirms the safety profile of the drug.

**Commercial Relationships:** matteo federici, None; VALERIA PAGLIEI, angela lanza, None; MARIA CRISTINA SAVASTANO, Giulia Midena, Benedetto Falsini, ANGELO MARIA MINNELLA, aldo caporossi, None

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Program Number: 4678 Poster Board Number: B0585
Presentation Time: 11:00 AM–12:45 PM

How do patients rate their subjective symptoms after CNGA3 gene therapy: First application of the instrument A3-PRO

Barbara Wilhelm1, Andreas Koegel2, Nadine Kahle2, Tobias Peters1, Stylianos Michalakis2, Martin Biel2, Mathias Seeliger4, Eberhart Zrenner2, Bernd Wissinger2, Susanne Kohl3, Marius Ueffing4, Karl Ulrich Bartz-Schmidt5, Dominik M. Fischer5, Ditta Zobor5, 1Centre for Ophthalmology, STZ ophthalmology, University Hospitals Tübingen, Tübingen, Germany; 2Institute for Ophthalmic Research, Centre for Ophthalmology, University Hospitals Tübingen, Tübingen, Germany; 3Center for Integrated Protein Science Munich (CIPSM) at the Department of Pharmacy - Center for Drug Research, Ludwig-Maximilians-Universität München, München, Germany, Germany.

**Purpose:** There are limitations of wide-spread standard measures for vision-oriented quality of life like the VFQ25. Specific symptoms in low vision caused by hereditary disorders and their short-term variations during an interventional trial may not be sufficiently reflected. This is especially true for Achromatopsia, a hereditary retinal degeneration with severe visual impairment in daily life, its leading symptom being glare in normal daylight conditions. Our aim was to develop a patient reported outcome (PRO) measure as a complement to the VFQ25 in the first gene therapy trial in CNGA3-linked Achromatopsia.

**Methods:** A3-PRO is a short questionnaire to measure the patients’ subjective visual performance and its development during therapy in a clinical trial, using five criteria – each represented by a single item. In order to also consider the possibility of sporadic deteriorations we provided a symmetric response scale, although distinctive declines were not expected. The patients rate the development of all aspects on a scale ranging from “almost always worse” (−4) to “almost always better” (+4) in nine indicated steps, including “unchanged” (0) as middle category. The questions are read by an interviewer, so the questionnaire contains a brief instruction and supplementary explanations to each item. The inventory is completed by a simple five-level scale of the patient’s overall satisfaction with the therapy. The questions were deduced from the key symptoms identified in an observational trial in the target cohort.

**Results:** A3-PRO was easy to assess for both patient and interviewer. The time need was appr. five minutes per test. Interim results of the ongoing trial (n=9; 1 f, 8 m; age range: 24 to 59 y) are reported (NCT02610582). The questionnaire was able to reflect intra- and interindividual changes of subjective symptoms over the time of the study in all three dosage groups. Our preliminary findings need to be confirmed in the final analysis.

**Conclusions:** A3-PRO is an easy applicable tool for the investigation of Achromatopsia. It provides valuable quantitative data on patient perception of symptom development after treatment and is superior to open, non-standardized questions in the clinical trial setting.

**Commercial Relationships:** Barbara Wilhelm, None; Andreas Koegel, None; Nadine Kahle, None; Tobias Peters, None; Stylianos Michalakis, Eyeserv GmbH (P); Martin Biel, Eyeserv GmbH (P); Mathias Seeliger, Eyeserv GmbH (P); Eberhart Zrenner, Eyeserv GmbH (P); Bernd Wissinger, None; Susanne Kohl, None; Marius Ueffing, None; Karl Ulrich Bartz-Schmidt, None; Dominik M. Fischer, None; Ditta Zobor, None

**Support:** Tistou and Charlotte Kerstan Stiftung, Germany, RD-Cure Project

**Clinical Trial:** NCT02610582
ARVO 2017 Annual Meeting Abstracts

Program Number: 4679 Poster Board Number: B0586

Presentation Time: 11:00 AM – 12:45 PM

**Natural History of Retinal function and structure in a French cohort of patients with Usher Syndrome Type 1 due to MYO7A Mutations**

Saddek Mohand-Said, Line Azoulay-Sebban, Isabelle S. Audo, Céline Chaumette, Céline Devisme, Karine Becker, Emmanuel Gutman, Caroline Segaut-Prevost, Ronald BUGGAGE, Jose A. Sahel, CHNO Quinze-Vingts / CIC Inserm, Paris, France; 2Streetlab, Paris, France; 3Department of Genetics, Institut de la Vision/INSERM/UPMC/CNRS/CHNO, Paris, France; 4Sanofi, Chilly-Mazarin, France; 5Inserm, U968; UPMC Univ Paris 06, UMR_S968, Institut de la Vision; CNRS, UMR 7210; CHNO des Quinze-Vingts, INSERM-DHOS CIC 1423, Paris, France, Paris, France.

**Purpose:** Usher Syndrome type 1 is a rare disease (prevalence: 1/50,000 in Europe) characterized by retinitis pigmentosa (RP), a congenital profound deafness and vestibular dysfunction. This study evaluates the natural history and retinal phenomenographic differences in patients with Usher syndrome type 1B (USH1B) due to MYO7A mutations.

**Methods:** Retrospective, chart-review study. The USH1B population included 45 patients from 38 families (age range, 9-61 years; median, 27) followed at the CHNO des Quinze-Vingts, Paris. Recorded visual assessments included visual acuity (VA), visual field (VF) and optical coherence tomography (OCT). Structure-function correlations were performed in comparison with age and phenotypic variability was studied.

**Results:** The median best eye visual acuity (BCVA) for the cohort was 0.5 in decimal (range, 0.05–1.25), and correlated to the age (Spearman’s rank correlation coefficient, r = -0.57; p = 0.0001), with central retina thickness (CRT) and length of the ellipsoid zone (lEZ) on OCT (r = 0.51; p = 0.0025; r = 0.53; p = 0.0016, respectively). Correlation between BCVA and CRT is higher when analyzing data from patients without edema (r = 0.72; p = 0.0005). VA decreases with advancing age but it is preserved until approximately 50 years. VF loss follows a pattern similar to non-syndromic RP, initially showing scotomas deepening in mid-periphery in the first decade, and progressively coalescing and extending. The residual tubular VF is also progressively reduced over time. The median of the diameter of the residual central VF was 35° (range, 3°–170°), and was also correlated with age (r = -0.46; p=0.0014).

OCT shows epiretinal membranes in 8% of cases, and cystoid macular edema (CME) in 39% especially in younger patients. The CME rate was 67% in patients under 20 years old, 29% between 21 and 40 years old and 17% over 40. In the patients without CME, the median CRT was 154.8 (range, 100–279 μm), and is correlated to lEZ (r = 0.75; p = 0.0002).

**Conclusions:** This study provides a better understanding of the natural progression of the ocular disease in patients with USH1B. The additional knowledge on this rare genetic form of RP will facilitate the appropriate selection of USH1B patients for whom treatment with gene therapy would be expected to provide the optimal benefit-risk balance in ongoing clinical trials.

**Commercial Relationships:** Saddek Mohand-Said, None; Line Azoulay-Sebban, None; Isabelle S. Audo, None; Céline Chaumette, None; Céline Devisme, None; Karine Becker, None; Emmanuel Gutman, None; Caroline Segaut-Prevost, Sanofi (E); Ronald BUGGAGE, Sanofi (E); Jose A. Sahel, None

Support: Sanofi

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Program Number: 4680 Poster Board Number: B0587

Presentation Time: 11:00 AM – 12:45 PM

**Effects of Retinal Gene Therapy on Auditory-Visual Cross Modular Plasticity: Does Re-Established Vision Kick Auditory Activity out of the Occipital Lobe?**

Aimee E. Willett, Mani Mahmoudian, Gloria J. Young, Albert M. Maguire, Jean Bennett, Manzar Ashtari

1Center for Advanced Retinal and Ocular Therapeutics (CAROT), University of Pennsylvania, Philadelphia, PA; 2Department of Ophthalmology, University of Pennsylvania, Philadelphia, PA.

**Purpose:** We used functional magnetic resonance imaging (fMRI) to explore the fate of auditory-driven cross modal (CM) activity within the visual cortex in a group of Leber’s Congenital Amaurosis (LCA) patients after sight restoration through retinal gene therapy (GT).

**Methods:** Participants consisted of 8 LCA patients and 8 normal-sighted controls. LCA patients were administered retinal GT in their worse seeing eye as part of a Phase-I clinical trial and subsequently received GT in their contralateral eye in a second follow-on clinical trial. Auditory (tonal interval) and visual (checkerboard) task fMRI activations were recorded an average of 3 months before and 3 years after GT re-administration. fMRI group general linear model analysis was conducted using BrainVoyagerQX. Pearson correlations between auditory and visual task occipital cortex activations were also performed.

**Results:** Group results from the auditory task show significant CM visual cortex activity before and after vision restoration in LCA patients. An increase in visual cortex activation area is observable 3 years after GT, Controls show no visual cortex activation during the auditory task. Results from visual task stimulation of the left and right eyes after GT show significant and widespread occipital lobe activations. Correlations show strong trends and significant relationships between areas of visually vs. auditory stimulated visual cortex activation areas at the 3-year time point. Activity of the right hemisphere has the strongest correlation between both tasks (p<0.03).

**Conclusions:** Our results show persistence of CM auditory activity within the visual cortex in a group of LCA subjects 3 years after successful vision restoration. Significant correlations between the auditory and visual task activations within the primary visual cortex 3 years after vision restoration showed a strong relationship between CM plasticity and response to vision. This may indicate a relationship between the strengthening of the visual pathway as a result of GT and increase in CM activation.

**Commercial Relationships:** Aimee E. Willett, None; Mani Mahmoudian, None; Gloria J. Young, None; Albert M. Maguire, US Patent number 8147823 (P); Jean Bennett, US Patent number 8147823 (P); Gensight Biologics (C), Sanofi-Aventis (C), Avalanche Technologies (C); Manzar Ashtari, None

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Phase II/III Visual Acuity Outcomes 1.5-Years Post-Treatment with rAAV2/2-ND4, an Investigational Gene Therapy for ND4 LHON

Scott Uretsky1, Nizza Thomasson1, Celine BOUQUET2, Anne Galy1, Jean Philippe Combial3, Serge Fitoussi4, Jose A. Sahel1,3, Catherine Vignal4,5,6. 1Clinical, GenSight Biologies, Paris; France; 2UPMC Univ Paris 06, INSERM U968, CNRS UMR 7210, Sorbonne Universités, Paris, France; 3Institut de la Vision, Paris, France; 4Centre Hospitalier National d’Ophthalmologie des Quinze-Vingts, Paris, France; 5Foundation Rothschild, Paris, France.

Purpose: rAAV2/2-ND4 is an experimental gene therapy enabling allotopic transgene expression. We report visual acuity (VA) outcomes 1.5-years post-treatment in a Phase II/IIIa (NCT02064569) open-label, dose-escalation safety study.

Methods: LHON patients with G11778A-ND4 mutation with stable vision loss received a single intravitreal injection of rAAV2/2-ND4 in their worst-seeing eye. Three patients were included in each dose-escalation cohort (9x10^9, 3x10^10, 9x10^10, 1.8x10^11 vg/eye) and the extension cohort (9x10^10 vg/eye) for a total of 15 patients. Post-hoc analysis of patient groups with ≤2-years vs. >2-years of vision loss at treatment and excluding the worst baseline VA (LogMAR>2.79) was performed.

Results: At baseline (N=15) mean/median (range) LogMAR in treated-worst and untreated-best eyes was 2.29/2.79 (1.10-3.01) and 2.03/2.01 (1.00-3.18) respectively; the difference between means is 0.265 (p=0.0342). Mean (range) vision loss duration was 72.3 months (8-271; median 22). All patients completed 48-week follow-up; one patient withdrew consent and 14 patients have completed 1.5-year follow-up. Mean LogMAR changes from baseline to 1.5-years post-treatment are as follows: For all patients (N=14): treated-worst eyes -0.612 vs. untreated-best eyes -0.308; mean difference -0.304. Excluding patients with baseline LogMAR≥2.79: group with ≤2-years vision loss (N=5) treated-worst eyes -0.632 vs. untreated-best eyes -0.234; mean difference -0.398. In the >2-year vision loss group (N=6) treated-worst eyes -0.451 vs. untreated-best eyes +0.071; mean difference -0.523 with VA change of 3/6 patients driving results.

Conclusions: Mean LogMAR change at 1.5-years improved in treated-worst and untreated-best eyes; there is a sustainable, clinically relevant, greater improvement of ≥0.3LogMAR in treated-worst versus untreated-best eyes. For patients with ≤2-years vision loss, mean differences favoring treated-worst eyes are noted at 36-weeks and increase in magnitude at subsequent follow-up. Those affected ≤2-years show a clinically relevant (≥0.3LogMAR) mean difference for the first time at 1.5-years post-treatment.

Commercial Relationships: Scott Uretsky, GenSight Biologies (E); Nizza Thomasson, GenSight Biologies (E); Celine BOUQUET, GenSight Biologies (E); Anne Galy, GenSight Biologies (E); Jean Philippe Combial, GenSight Biologies (E); Serge Fitoussi, GenSight Biologies (E); Jose A. Sahel, ERC Synergy “HELMHOLTZ” (F), Foundation Fighting Blindness (F), Genesignal (C), Pixium Vision (I), Chronocam (I), Pixium Vision (C), GenSight Biologies (I), GenSight Biologies (C), Chronoflame (I), GenSight Biologies (F), LIFESENSES (ANR-10-LABX-65) (F), Banque publique d'Investissement (F), LabEx (F); Catherine Vignal, GenSight Biologies (C)

Clinical Trial: NCT02064569

Program Number: 4682 Poster Board Number: B0589
Presentation Time: 11:00 AM–12:45 PM
Correlation analysis between Argus II Retinal Prosthesis Patients performances and electrophysiological data

Laura Cinelli, stanislao rizzo. Ophthalmology, Azienda Ospedaliero Universitaria Careggi, Florence, Italy.

Purpose: The Argus II Retinal Prosthesis System (Argus II) is currently the first and only retinal prosthesys with both FDA (2013) and CE (2011) approval and has been implanted in more than 210 severely vision impaired patients. Since 2011 we implanted 29 patients with the Argus II in our centers. We followed up closely patients’ progresses using three visual function tests designed to measure their ability to localize objects, discriminate motion, and discriminate gratings’ orientation.

Noticing that the population performed statistically better with the system on versus off in these tasks, but that the results varied from patient to patient, we wanted to verify if electrophysiological factors are affecting the performances in these subjects, therefore an analysis of the retinal health and implant position could predict or correlate with patients performances.

We chose electrode-retina distance (known factor in determining the amount of current necessary to induce a visual percept), retina thickness (associated with the extent of retinal degeneration in advanced stages of RP), and electrical thresholds.

Methods: Starting from OCT images obtained by Heidelberg Engineering or Nidek systems, we measured the electrode-retina distance, retinal thickness and electrodes-fovea distances of 19 Argus II patients. Electric threshold measurements and visual acuity tests were done by the custom developed Argus II software. Correlation analysis between the parameters has been performed in MATLAB and Excel.

Results: We performed correlation analysis between patients’ performances and electrophysiological and geometric data. We found no correlation between retinal thickness and ability to localize objects or discriminate movements. We observed a slight correlation between the mean distance between implant and retina and both the visual function tests. Average threshold values correlate only with the ability of movement detection.

Conclusions: Our results show that electrode–retina distance and thresholds values affect performances. We conclude that a tight juxtaposition of the array to the retina is critical for optimizing the benefit of the patients.

Commercial Relationships: Laura Cinelli, None; stanislao rizzo, None

Clinical Trial: NCT01490827

Program Number: 4683 Poster Board Number: B0590
Presentation Time: 11:00 AM–12:45 PM
Handheld VEP in elderly and/or non-verbal, non-mobile Glaucoma Patients

ivy S. kim1, Gloria Wu1, Mary Gao2, Bonnibel Bautista2, 1Molecular, Cellular, and Developmental Biology, University of California, Santa Barbara, Santa Barbara, CA; 2Psychology, Saint Louis University, Baguio, Philippines; 1Integrative Biology, University of California, Berkeley, Berkeley, CA; 1Ophthalmology, University of California, San Francisco, San Francisco, CA.

Purpose: Visually Evoked Potential (VEP) has been shown to be helpful in examining the integrity of the visual pathways in nonverbal children and adults. The use of a portable, handheld VEP in the setting of elderly, non-mobile and/or non-verbal glaucoma patients has not been well studied.

Clinical Trial: NCT02064569

Program Number: 4681 Poster Board Number: B0588
Presentation Time: 11:00 AM–12:45 PM
Phase I/IIa Visual Acuity Outcomes 1.5-Years Post-Treatment with rAAV2/2-ND4, an Investigational Gene Therapy for ND4 LHON

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Purpose: To evaluate VEPs using the RETeval device in elderly, non-mobile, and/or non-verbal glaucoma patients in a clinical setting.

Methods: The RETeval device served as the ganzfeld; flash VEP was performed on patients, using ISCEV Vi guidelines of electrode placement. The RETeval flash VEP followed ISCEV Vi protocol for flash VEP, 3 cd s-2. All patients were dilated to 6mm. VEPs were performed in 2016. Inclusion criteria: vision 20/20 to 20/60, glaucoma patients diagnosed by previous eye MD or optometrist, previous VF documenting glaucoma. Inability to sit for visual field, inability to operate hand held remote control for automated visual field testing, inability to speak or communicate with staff. Exclusion criteria: macular laser or previous optic neuritis history.

Control patients: no glaucoma, no optic nerve disease, visual acuity 20/20-20/60. Measurements of P2, N2 performed manually (IK and verified by GW).

Results: 8 Glaucoma patients: 5 females, 3 males, avg age=74.7 yrs (sd=12.5, range=59-94 yrs). 8 Controls : 3 females, 5 males, avg age=66.0 yrs (sd=14.9, range=43-83 yrs).

P2 amp: (Glc) P2 (avg OD and OS)=27.5 uV, sd=11.0, range=18-48 uV; Controls P2 (avg OD and OS)=14.7 uV, (sd=9.7, range=7.5-33.5) unpaired t-test: p=0.028

P2 Implicit time (P2IT): Glc v C, t-test not significant.

Conclusions: This small study shows that there may be clinical usefulness in the handheld flash VEP in the setting of poorly mobile, elderly and/or non-verbal patients with glaucoma. This handheld VEP holds promise in the follow up of our most vulnerable patients who need eye care.


Program Number: 4684 Poster Board Number: B0591

Presentation Time: 11:00 AM–12:45 PM

Optic radiation damage relates to reduced V1 and thalamus volume in cortical/cerebral visual impairment

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Purpose: Cortical/cerebral impairment (CVI) is the leading cause of pediatric visual impairment in developed countries. It is caused by pre/perinatal brain damage, which impairs the development of vision-related pathways, leading to visual field loss and deficits in higher order visual processing. The extent to which the primary visual pathway is damaged in CVI is unknown. To this end, the current study examined the volume of the thalamus and V1, as well as tractography of the optic radiations in individuals with CVI compared to typically sighted controls (CTL).

Methods: Subjects were scanned on a 3T Philips Achieva MRI system to acquire two structural T1w and a 64-direction high angular resolution diffusion imaging (HARDI) image (Np=6, Np=6). CVI was caused by either periventricular leukomalacia (PVL N=4) or epilepsy (non-PVL N=2). T1 data were processed in FreeSurfer to measure thalamus volume, V1 volume, and V1 cortical thickness. HARDI data were reconstructed in DSiStudio. Optic radiations were quantified for tract number, tract volume, average fractional anisotropy (FA), and generalized fractional anisotropy (gFA).

Results: CVI was associated with significant reductions in thalamus volume (CVI=4993.8±1824.1 mm3, CTL=7864.8±2099.6 mm3 (mean±SD), p <0.005), V1 volume (CVI=1752.3±258.7 mm3, CTL=2218.8±412.2 mm3, p<0.01), V1 thickness (CVI=1.5±0.1 mm3, CTL=1.7±0.08 mm3, p<0.05), and optic radiations volume (CVI=6059.2±5745.6 mm3, CTL=19187.1±8787 mm3, p<0.005). Decreased fiber number was only observed in CVI due to PVL (CVI=1565.8±2700, CTL=23965±18451, p<0.05; CVI vs. \[10846±14714\], QA was significantly increased in CVI (QA=0.23±0.07, CTL=0.16±0.03, p<0.05), while there were no significant differences in gFA. Significant correlations were observed between thalamus volume and QA (r²=0.59, p<0.005), tract volume (r²=0.52, p<0.05), and number of reconstructed fibers (r²=0.5, p<0.01). Visual inspection of the optic radiations revealed that fewer fibers were reconstructed to the upper banks of the V1 sulcus in individuals with CVI due to PVL.

Conclusions: The results indicate that the optic radiations, thalamus, and V1 are reduced in CVI. This may depend on the aetiology of CVI. For example, the optic radiations representing the lower visual field were decreased only in CVI due to PVL, corresponding to documented visual field loss.

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Ophthalmology - Lions Vision Center, None; We developed a stereo camera pair with 14.0 cm base disparity, ¼" VGA (640x480) CMOS sensors, and 2.84 mm focal length, providing 7 arcmin resolution. The cameras, mounted on a flat rigid frame, are clipped onto the Argus II glasses. A USB video processing unit (VPU) replaces the standard camera input.

**Methods:** Patients underwent ophthalmic examination, Optical coherence tomography (OCT), autofluorescence, functional testing including Full Field ERG and genetic mutational screening. Further tests were performed in some patients, including fundus fluorescein angiography (FFA).

**Results:** R31IQ homozygous mutation was identified in all three patients with additional heterozygous NRL: c.654delC in the child aged 3. Patients were aged 3, 32 and 40 years; all had night blindness from childhood and reducing VA. The 3 year old child had esotropia of the RE, VA 0.1 (RE), 0.6 (LE), physiological hypermetropia and astigmatism, normal appearing anterior segment optic nerves and peripheral retina, large interpapillomacular subretinal fibrosis with multiples yellowish subretinal deposits in the right eye, subretinal yellow deposits temporal to the fovea in the left one. Autofluorescence showed multiples hyperfluorescent dots in the posterior pole and large areas of hypofluorescence where fibrosis has been previously described. Fluorescein angiography showed central retinal atrophy, absence of foveal avascular zone and absence of vascular anomaly, leakage or ischemia. OCT showed full thickness atrophy in both eyes predominant in the right one. ISCEV Standard Full Field electroretinogramm under narcosis showed markedly reduced scotopic responses and slightly reduced photopic waves.

**Conclusions:** The phenotype in ESCS is highly variable, the associated Nrl mutation might play some additional role in the severity of the disease.

**Commercial Relationships:** youssr louati; Veronika Vaelvik, None; Daniel F. Schorderet, None; Imen Habibi, None

**Program Number:** 4685 Poster Board Number: B0592

**Presentation Time:** 11:00 AM–12:45 PM

**Consanguineous Tunisian Family With Enhanced-S-cone Syndrome**

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**Purpose:** To report the clinical, electrophysiological and genetical characteristics of 3 family members with homozygous NR2E3 gene mutation, classically associated with Enhance-S-cone syndrome. The youngest 3 years old patient had an additional heterozygote NRL mutation and a clinically more advanced disease

**Methods:** Patients underwent ophthalmic examination, Optical coherence tomography (OCT), autofluorescence, functional testing including Full Field ERG and genetic mutational screening. Further tests were performed in some patients, including fundus fluorescein angiography (FFA).

**Results:** All 4 Argus II users were 100% correct on the chair presence test with filtering; without filtering 3 performed at chance, while the 4th scored 80% (n.s.) but took 49 s (17 s with filtering). On the near presence test subjects were 90-100% correct with, vs. at chance without, filtering. One subject was able to perform the chair position test both with and without filtering, while another was at chance in both conditions; for the near position test all subjects were above chance (67–89%) with, and at or near chance (22–56%) without filtering. For the depth discrimination at 1.50 m, the minimum depth difference successfully detected was 10–20 cm, while at 40 cm a depth difference of 2.5 cm was reliably detected.

**Conclusions:** Argus II users can benefit from depth-filtered imagery in 2 ways: 1) by detecting the presence of objects within a range of interest; 2) by estimating relative distances of objects to within 10%. A production version of the system is under development.

**Commercial Relationships:** Gislin Dagnelie, Quadra Logic Technologies (F), eSight Corp (C), Second Sight Medical Products (P), Second Sight Medical Products (C), eSight Corp (I); Greg Seifert, Advanced Medical Electronics Corp (E); Paul Gibson, Advanced Medical Electronics Corp (E); Arup Roy, Second Sight Medical Products (E); Michael P. Barry, Second Sight Medical Products (P); Avi Caspi, Second Sight Medical Products (P), Second Sight Medical Products (C)

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**Presentation Time:** 11:00 AM–12:45 PM

**Retinal prosthesis users' shifts in hand-camera coordination correlate with changes in eye orientation**

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**Purpose:** Determine whether previously observed shifts in hand-camera coordination of retinal prosthesis users correspond with long-term changes in eye orientation.

**Methods:** Two Argus II retinal prosthesis users performed 3 pointing tasks on 2 different setups every 1–2 weeks over 8 months. One setup used a chinrest with an eye-tracking camera. The second setup allowed the participant to use the Argus II glasses and camera to visually scan a touchscreen with head movements. On the eye-tracking tower, participants were asked to either point to percepts using a touchscreen when electrodes were activated by computer control, or touch the screen directly in front of the nose, when no stimulation was provided. When using the Argus II glasses, participants touched randomly located 4° circular targets as accurately as possible 100–300 times per session. Eye orientation was measured by tracking the implanted eye’s pupil center and a corneal reflection. Optimal camera alignment positions
(OCAPs) were estimated from pointing errors when using the Argus II camera. Correlated eye-tracking and OCAP data were collected on the same day. Correlation significance was determined using bootstrap resampling of ordinate values. Rates of change were estimated using ordinary least squares regression.

**Results:** Over the measurement period, subject S1’s OCAP shifted leftward 0.02°/day and downward 0.05°/day. S2’s OCAP shifted downward 0.01°/day. Each of these OCAP-time correlations were significant ($p < 10^{-6}$).

Horizontal and vertical shifts in OCAPs correlated with changes in average eye orientation for both subjects. S1’s horizontal and S2’s CAP changes correlated best with stimulation-OFF eye orientation changes (S1 horizontal: Pearson’s correlation coefficient $r = 0.26$, $p < 10^{-4}$; S2 horizontal: $r = 0.07$, $p < 10^{-1}$; S2 vertical: $r = 0.08$, $p < 10^{-4}$).

S1’s vertical CAP changes correlated best with stimulation-ON eye changes ($r = 0.13$, $p < 10^{-4}$). Subtracting average changes in eye orientation from OCAPs for S1 reduced the horizontal rate of change by 47% and vertical by 104%. S2’s OCAP vertical rate of change was corrected by 103%.

**Conclusions:** Changes in hand-camera coordination correlate with long-term changes in average eye orientation. Eye tracking integrated into a visual prosthesis may be used to estimate and at least partially compensate for changes in hand-camera coordination over time.

**Commercial Relationships:** Michael P. Barry, Second Sight Medical Products (F); Sophia Díaz-Aguilar, None; Liancheng Yang, None; Gislin Dagnelie, Quadra Logic Technologies (F), eSight Corporation (C), eSight Corporation (I), Second Sight Medical Products (P), Second Sight Medical Products (C).

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**Presentation Time:** 11:00 AM–12:45 PM

**Prosthetic vision and sensory substitution are comparable to native ultra-low vision in visual performance assessment Olukemi Adeyemo, Duane Geruschat, Gislin Dagnelie. Johns Hopkins Wilmer Eye Institute, Baltimore, MD.**

**Purpose:** To test the assumption that visual performance with ultra-low vision (ULV) is similar regardless of vision modality (i.e. native, retinal implant, sensory substitution) and can be measured using the ULV activities of daily living (ULV-ADL) assessment toolkit.

**Methods:** Thirty-five patients were recruited for this study: 25 with native ULV – i.e., from natural causes, 4 with an Argus II retinal implant and 6 with a Brainport (sensory substitution device using tongue stimulation). The 17-item ULV-ADL toolkit was administered in their home environment. Tasks in the ULV-ADL are administered at 3 difficulty levels and include everyday activities such as detection of a towel on a rack, a moving cursor, a lit candle on a table, etc. Item and person measures were calculated from the ULV-ADL scores using Winsteps®.

**Results:** The item measure distribution of the ULV-ADL items in native ULV participants ranged from -3.94 (less difficult to complete) to +2.24 (more difficult to complete) logits. A narrowed difficulty range of -2.02 to 2.32 logits was obtained when scores from all participants were included. Person measures in our sample ranged from -2.02 to 5.35 logits. The Argus II and Brainport users were within the same range as the native ULV subjects. The Argus II users in our sample had lower ability compared to others while subjects with higher ability had larger standard errors (i.e. cruder estimates) since their ability exceeded the difficulty of the ULV-ADL items. A principal components analysis indicated that 56% of the variance was explained by the model, and showed no evidence of additional dimensions (largest unexplained component, 5.1%).

**Conclusions:** Despite the small sample size, our data set confirms the assumption that native, prosthetic and substitute ULV all exhibit similar behavior and can be measured using the ULV-ADL toolkit. This demonstrates the instrument’s potential across the ULV rehabilitation field, irrespective of the cause of the ultra low vision.

We intend to continue examining this in a larger sample and compare person measures from our self-report questionnaire (ULV-VFQ) and the ULV-ADL.

**Commercial Relationships:** Olukemi Adeyemo, None; Duane Geruschat, Second Sight Medical Products (C); Gislin Dagnelie, Second Sight Medical Products (P), Second Sight Medical Products (C).

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**Presentation Time:** 11:00 AM–12:45 PM

**Comparison between the only Usher Syndrome patient implanted with ARGUS II® and patients affected with Retinitis Pigmentosa utilizing the same retinal prosthesis system. One year follow up Sarah Karam Palos, Maximiliano Olivera, Maria Iglesias Alvarez, Jeroni Nadal Reus. Ophthalmology, Barraquer Ophthalmological Center, Zaragoza, Spain.**

**Purpose:** To analyze the acquired abilities by the only (worldwide known) Usher Syndrome (US) patient who underwent ARGUS II® retinal prosthesis implant after one year follow up compared to the patients affected with Retinitis Pigmentosa (RP) also implanted with the device in our center.

**Methods:** We analyzed the results of rehabilitation in our three patients implanted with ARGUS II, two women (41 and 50 years old) diagnosed of Retinitis Pigmentosa, who were legally blind for more than 15 years, and a 51-years-old man diagnosed of Usher Syndrome, legally blind for 16 years. We analyzed general health condition, surgical time and perioperative complications, time to first device power on, light/shadow perception, high contrast shape identification, movement identification, high contrast letters, signs and general life achievements (standard questionnaires and methodology for all the patients), together with using time of the device including both rehabilitation-pauted time and free-will usage of the device.

**Results:** Surgical time improved from 3:15 hours to 2:20 hours. We relate this time variability due to a logical surgeon’s learning curve. No perioperative complications were observed and time to first device power on were the same (15 days) for all cases. After two months of ARGUS II® device usage both patients affected with Retinitis Pigmentosa (RP) showed a much faster progression on rehabilitation therapy and achievements. At the third month US patient showed an extraordinary improvement on the general life tasks and became much more independent for daily life compared to RP patients, being even capable to discriminate a broader contrast gamma. After one year follow-up results are persistent. Because our study is about the only US Syndrome patient in the world with a fully functional ARGUS II® device implanted, we are not able to analyze statistical data.

**Conclusions:** Although Usher Syndrome is not contemplated as an indication for ARGUS II® implant, mainly because of their learning and communicative impairment, the result observed with our patient should be encouraging to considerate US, and probably other similar conditions, also as retinal prosthesis implants suitable receptors.

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