



ICO

# ILLINOIS COLLEGE OF OPTOMETRY

## 2015 RESEARCH PRESENTATIONS

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# The Association of Schools and Colleges of Optometry to Convene an Interprofessional Education Summit in February 2016

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Goals of the IPE Summit	Objectives
<b>To assemble representatives from ASCO member institutions and other champions of interprofessional collaboration</b>	<b>Create opportunities among attendees for networking and collaboration</b>
	<b>Raise the role optometry plays in interprofessional healthcare teams</b>
	<b>Identify synergies in obtaining grant funding for developing and sustaining IPE and IPP</b>
<b>Share current best practices in IPE and IPP</b>	<b>Communicate techniques, strategies and considerations for IPE and IPP, including engagement of optometry in team-based patient care</b>
	<b>Identify outcome measures and tools to assess the impact of IPE and IPP on student learning</b>
<b>Share ideas on future development of IPE and IPP</b>	<b>Explore future options for enhanced engagement in IPE and IPP</b>
	<b>Develop strategies to assess the impact of IPE and IPP on patient outcomes</b>

## BACKGROUND

Interprofessional practice (IPP), in its higher form, occurs when healthcare professionals from different disciplines and professions work in teams, facilitated by regular face-to-face meetings, to make unified decisions about a patient's care.

Nationally, the Association of Schools and Colleges of Optometry (ASCO) was the first "Supporting Organization" of the Interprofessional Education Collaborative (IPEC); a coalition of health professions education associations promoting interprofessional education (IPE) and collaborative health care.

In 2013, ASCO developed an interprofessional education task force that was, in part, charged with identifying best practices within ASCO institutions to prepare graduates for team-based practice. In 2014 the task force became an ad hoc Committee within ASCO.

To affirm optometry's commitment to interprofessional practice, ASCO continues to seek new ways to be included in models of team-based care.

## METHODS

A Survey of Interprofessional Education Programs in the Schools and Colleges of Optometry was mailed to each President and Chief Academic Officer of the twenty-one U.S. schools and colleges of optometry. The sixteen question survey, originally developed in 2011 by an ASCO Government Affairs staff member was modified to ask about existing IPE activity as well as challenges and future plans relating to IPE at each school. The survey was revised and re-administered in 2015. Each of the ASCO member institutions completed the survey and the results were tabulated by the ASCO office. The ASCO task force in turn analyzed the results and made recommendations to ASCO for strengthening optometry's role in patient care teams.

## RESULTS

ASCO plans to convene a national IPE Summit in February 5 & 6, 2016 at Southern California College of Optometry at Marshall B. Ketchum University. Three main goals of the IPE Summit with corresponding objectives have been defined (see table above).

## CONCLUSION

The IPE Summit is an important next step in ASCO's commitment to supporting optometry's role in IPE, team-based collaborative patient centered care and ultimately improved patient outcomes. Two representatives from each ASCO member institution have been invited to participate in the Summit as well as representatives from important stakeholders in optometric education including the AOA and VA.

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# Linking Student Performance to Timeliness and Accuracy of Patient Encounter Logging: The Carrot and the Stick.

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## INTRODUCTION

A patient encounter log (PEL) is used by most Schools and Colleges of Optometry to document and assess students' clinical experiences. The medical education literature demonstrates that no more than 83% of patient encounters are recorded in medical student logbooks.

Diverse documentation strategies are used to track student's clinical encounters:

- Paper
- Hand-held devices
- Web-based logging systems

Each system has limitations since all rely upon students' willingness to accurately document the information. Students may not report all encounters, may carelessly or erroneously recall information, and may even falsify data.

The author previously demonstrated that linking the student evaluation process to a patient encounter logging system improves the accuracy of student patient encounter logs at the Illinois College of Optometry during the third professional year.

Question: Does evaluating clinical performance of first year students based, in part, a PEL improve logging?

## BACKGROUND

First year students at the Illinois College of Optometry (ICO) are assigned one session of patient care for five weeks in the Primary Care Clinic of the Illinois Eye Institute (IEI) during the winter and spring quarter as part of the Clinical Assistant Program (CAP). The students are assigned to a faculty preceptor who then assigns the students to work with one of their third or fourth year students during the session.

Students use Meditrek ©, a medical education management system to log their patient encounters online. A previous study demonstrated third year students who log their encounters with high frequency (promptly after each patient care session) report a statistically higher average number of patient encounters/session than students who log their encounters less frequently.

First year students started in the CAP program observing patient care or assisting patients navigate through IEI during their assignments. During AY 2013-2014 students began to perform entrance test skills while being observed by third or fourth year students. Each student was required to perform a set of identified skills over the course of their two quarters of patient care.

CAP students were initially evaluated on an 'Unsatisfactory/Satisfactory' grading system based on their attendance and completion of their patient care logs. As with third year students, it became apparent that students could be classified as frequent (logging experiences after each encounter) or infrequent (logging experiences only at the end of rotation) loggers.

A new evaluation system was put in place during AY 2014-2015 using an 'Unsatisfactory/Satisfactory/Honors' grading system. Students may achieve an 'Honors' performance based, in part, the timeliness logging their patient care experiences. A student could also achieve an 'Honors' performance based upon their level of engagement in patient care activity during their assigned sessions.

## METHODS

### Pre-study period: AY 2012-2013

- Students recorded patient encounters on-line via Meditrek ©
- The number of patient encounters for each student was identified
- Students were evaluated on a 'Satisfactory/Unsatisfactory' basis



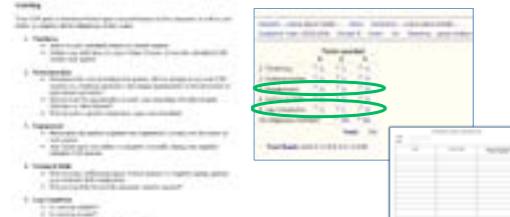
### Pre-study period: AY 2013-2014

- Students recorded patient encounters on-line via Meditrek ©
- Students completed 'Skills Assignment' form and submitted an in-progress form at the conclusion of winter quarter and the completed form at the conclusion of spring quarter.
- Number of patient encounters for each student identified
- Students continued to be evaluated on a 'Satisfactory/Unsatisfactory' basis



### Study Period AY 2014-2015

- Students completed 'Skills Assignment' form and submitted an in-progress form at the conclusion of winter quarter and the completed form at the conclusion of spring quarter
- A 'Supplemental Skills Assignment Log' was initiated to encourage students to perform additional skills beyond the minimum requirements
- New grading system reviewed during a required orientation session prior to the start of winter quarter
- Number of patient encounters for each student identified using the Meditrek © logging system

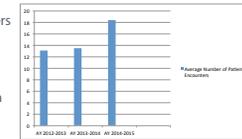


## RESULTS

# No statistical difference was found in the average number of patient encounters reported by first year students when only observing in patient care and when actively engaged in patient care.

\* First year students logged a statistically higher number of average patient encounters when their evaluation was tied to the timeliness of reporting their experiences (18.4) and their engagement during their patient care sessions (p<.0001) than when only observing in patient care (13.1) or when actively engaged in patient care (13.5).

Academic Year	Number of Students	Average Patient Encounters
2012-2013	146	13.1 *
2013-2014	166	13.5 *
2014-2015	156	18.4 *



## CONCLUSIONS

- Actively engaging students in patient care does not impact the average number of patient care experiences logged by first year students when students are graded on a 'Satisfactory/Unsatisfactory' basis.
- Grading first year students on an 'Honors/Satisfactory/Unsatisfactory' basis increases the average number of patient care experiences logged when the evaluation system is tied to the timeliness of students' logging their encounters and their engagement in performing clinical skills during their CAP sessions.
- While it appears evaluating first year students on the timeliness of their logging and engagement in patient care it is not clear that the students' log entries are more accurate. This facet of student logging deserves additional study.

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# The Importance of Comprehensive Eye Care for Children entering Kindergarten in the Chicago Public Schools

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## BACKGROUND

Despite recommendations stressing the importance of eye exams prior to elementary school, the majority of children enter school without comprehensive eye care. Illinois is one of only a couple of states that require a comprehensive eye exam to be completed upon entering school for the first time. Typically, this refers to children entering Kindergarten though it is also required for children moving into the district at any grade.

For the 2013-2014 school year, compliance with this Illinois law was only 62.1% for the entire state according to the Illinois State Board of Education. Compliance for CPS was 14.2% and the rest of the state was 73.4%. This study breaks down the exam findings for children entering Kindergarten or in the year prior to their entrance into Kindergarten to the Chicago Public Schools (CPS) who visited the clinic and were seen from July, 2013 through June, 2014. Results of this study will be shared with Chicago Public Schools Health and Wellness department in order to stress the importance of schools complying with the vision laws of the state.

## METHODS

Between July 1, 2013 and June 30, 2014, 635 children in or entering Kindergarten within one year were seen at the Illinois Eye Institute at Princeton (IEI at Princeton), a school-based clinic serving CPS students, for a comprehensive vision exam. Children seen at the clinic were either brought to the clinic by bus from their schools or brought to the clinic by their parents/guardians. Of the 635 children seen, 22 (3.9%) presented with a history of a previous correction. A complete eye examination was performed, including cycloplegia when permission was given by the parent/guardian. The breakdown by race can be seen in Chart 1.

Chart 1: Racial Distribution

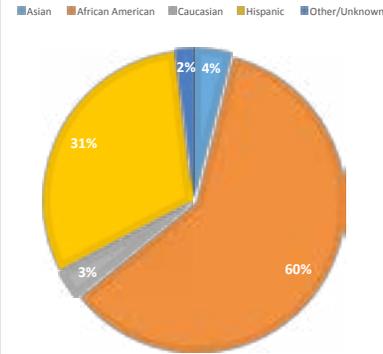


Table 1: Cycloplegic Autorefraction

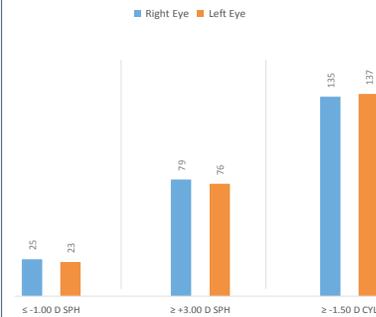
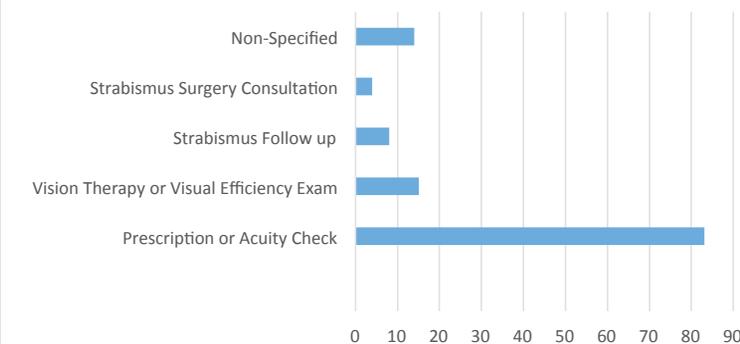


Table 2: Reasons for Follow Up Care



## RESULTS

Entering distance visual acuity in the right eye was 20/25 or better with 13.4% less than 20/40. Cycloplegic autorefraction found 4.1% had at least 1 diopter of myopia, 13.7% had ≥3.00 diopters hyperopia, and 24.6% had ≥1.50 diopters astigmatism. (see Table 1) Strabismus was detected in 6.9% and amblyopia in 9.9% of the children. All intermittent and constant horizontal and vertical strabismus was included in this count. Refractive correction was given to 36.5% of the children. In addition, 164 children (26.0%) required follow up care. The most common reason for further care was being prescribed a new or significant prescription or visual acuity check. (see Table 2)

## CONCLUSIONS

The results of this study demonstrate the importance of eye exams for children entering school. Many of these children had already begun their Kindergarten year, and some may have visited the clinic as a result of a failed vision screening rather than only to fill the Illinois examination requirement. This highlights the fact that the population seen may not be reflective of a general population and this group may have higher than normal percentages of vision problems. The majority of the patients seen at the IEI at Princeton fall below the federal poverty level. The research has demonstrated that this is a population associated with higher than average rates of vision disorders in children. The results of this study will be shared with the CPS Office of Health and Wellness to better educate administrators, teachers, and parents of the importance of eye examinations in young children.

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# Effect of Chicago Public School students Receiving Vision Care at the IEI at Princeton Clinic on Grade Point Average and Math and Reading Scores

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Valeria Michelman, Paul Moore, Julia Gwynne, Consortium on Chicago School Research, University of Chicago, Chicago, IL

## PURPOSE

It is suspected that correcting children's vision will have a positive impact on their academics. This study looked at the effect of attending a school-based vision clinic for an eye exam on grade point average (GPA), math and reading test scores.

## BACKGROUND

The Illinois College of Optometry partnered with the Chicago Public Schools (CPS) to open the Illinois Eye Institute at Princeton Vision Clinic in January 2011. The clinic was opened in response to the lack of follow through for children who failed vision screenings and were referred and who were not seeking appropriate comprehensive vision care. CPS has developed a sophisticated system to ensure vision screenings of children at regular times in their academic lives, more specifically, they are screened at kindergarten, second and eighth grades, as well as, children who are considered at risk for a vision problem. The gap in ensuring that children received needed eye care when referred led to the opening of the IEI at Princeton School-based Vision Clinic.

## METHODS

Demographic data along with the results of the examination have been collected for all children that have attended the clinic. The Consortium on Chicago School Research at the University of Chicago has a memorandum of understanding with the CPS which provides them access to attendance, tests scores, GPA along with other information relating to CPS students academic efforts.

Standardized test scores and grade point average (GPA) for children included in the study were analyzed from the year prior to attending the IEI at Princeton Clinic and minimally the year after the visit. A student fixed effects model was used to estimate within-student change in outcome following the visit. Academic data included longitudinal Chicago Public Schools (CPS) standardized scores and GPAs from 2007 - 2014 and patient eye exam data on 14,663 CPS students seen from Jan. 2011 and Dec. 2013. This phase of the study looked at effect of race and magnitude and type of refractive error on academic outcomes.

The population served were 14,929 students who visit the clinic (14,663 were enrolled in CPS). Children seen were referred due to screening failure, concern about vision, lost/broken glasses, and need for compliance with mandate of a comprehensive eye exam upon entry to school or simply brought in for an eye exam by the parent.

The following describes the make up of the patients that were seen who were students in CPS.

### Demographics of population

	Characteristic	IEI at Princeton	Resident School	CPS (district)
Race	Black	60.6%	57.9%	41.4%
	Latino	35.4%	37%	44.7%
	Other(white, mixed, Asian, middle eastern, unknown)	4.0%	5.1%	13.9%
SES	Free/Reduced Lunch	94.3%	88.7%	83.5%
	Neighborhood Poverty	0.489	0.462	0.205

### Academic Profile

	Characteristic (average)	IEI at Princeton	Resident School	CPS (district)
	GPA (5 point scale)	2.42	2.49	2.58
	Math Score	-0.324	-0.161	0.019
	Reading Score	-0.317	-0.149	0.015
	Attendance	94%	93.4%	93%
	Special Education	21%	12.9%	12.6%

## RESULTS

Academic data was available for 13,144 of the subjects. Subjects were excluded in Grades K-2 since there were no standardized test scores available for them pre and post exam.

The population seen at IEI at Princeton were known to perform lower on testing (math- -0.32, reading - -0.317, GPA 2.42) then the average CPS student (math- +0.2, reading - +0.02, GPA 2.58).

This presentation is reporting on the analysis that looked at estimating change in effect for specific subgroups of subjects, specifically, race differences and those found to have specific refractive errors.

The analysis looked at the Latino/Hispanic versus Black/African American. The students were separated into two groups – Latino and non-Latino. Note that almost all non-Latino students (94%) are Black. Latinos demonstrated improved reading scores but no change in math or GPA.

### Race/ethnicity

	Parameter	Reading Score			Math Score			GPA		
		Estimate	p-value	Sig.	Estimate	p-value	Sig.	Estimate	p-value	Sig.
Non-Latino	Effect	-0.031	0.001	*	0.025	0.004	*	0.049	0.000	*
Latino	Effect	0.036	0.001	*	0.025	0.018		-0.006	0.665	

After the exam, statistical significance in outcomes was seen for subjects with moderate/high hyperopia (improved GPA) and astigmatism >1.25 (improved GPAs). Refractive error data reflected autorefraction findings 30 minutes after the subject was administered 1 drop 2.5 % phenylephrine, 1 drop 1% tropicamide, and 2 drops 1% cyclopentolate.

### Refractive error: measure of type and severity of vision problem

Subpopulation	Parameter	Reading Score			Math Score			GPA		
		Estimate	p-value	Sig.	Estimate	p-value	Sig.	Estimate	p-value	Sig.
Low or no refractive error (wet)	Effect	-0.010	0.270		0.029	0.001	*	0.015	0.218	
	Trend	-0.018	0.004	*	-0.023	0.000	*	-0.023	0.037	
Moderate or high hyperopia - > 2.00 diopters(wet)	Effect	0.007	0.736		0.032	0.085		0.084	0.001	*
	Trend	-0.030	0.032		-0.027	0.048		-0.013	0.598	
Moderate or high myopia - <3.00 diopters (wet)	Effect	-0.014	0.602		0.029	0.271		0.006	0.872	
	Trend	-0.024	0.312		0.018	0.394		-0.002	0.967	

## CONCLUSIONS

These results suggest that attending the clinic improves academic outcomes for students with moderate/high hyperopia and high astigmatism and Latinos. This confirms the fact that an eye exam has a positive impact on academic performance in low income Latino children in the CPS system and children with moderate or high hyperopia.

Several challenges preclude the results from being extrapolated to general populations: subjects were primarily low income, high number special education students and self-selection. The effect seen may be less than expected since there was no mechanism to ensure that the glasses were actually worn as recommended.

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# Development of A Reference Database of OCT Parameters in a Pediatric Population

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## INTRODUCTION

Optical coherence tomography (OCT) is a critical tool in diagnosis and clinical management of patients. The newer Cirrus HD-OCT is a spectral domain (SD) OCT that collects more data in a shorter period of time as compared to time domain (TD) OCT technology, which allows for faster and more precise scanning.

There is currently no normative data for the Cirrus HD-OCT for patients under 18 years of age. This poses a clinical challenge because the practitioner is unable to compare patient OCT results with age, sex, and race matched norms. Numerous authors have demonstrated that race, sex, and age effect OCT parameters. Hence, adult normative data cannot be extended to pediatric populations.

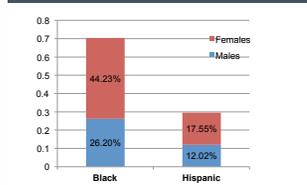
The aim of this study is to establish a reference database of RNFL thickness, macular thickness, and ONH parameters for the patient population at IEL at Princeton, which consists of predominantly Black and Hispanic children. This reference database will facilitate evaluation and management of physiologic abnormalities.

## METHODS

### Data Collection

- Data collected on 9 to 17 year olds with race designations of Black or Hispanic from November 2014 to March 2015 on patients who presented to the IEL at Princeton clinic, as part of their routine dilated exam
- Data collected:** age, sex, race, ocular/medical history, family history, VA, cycloplegic auto refraction (Full Auto Ref R-F10), C/D ratio, IOP (TA011, iCare Finland), fundus photography (Visucam PRO NN)
- Exclusion criteria:** VA < 20/40; IOP > 22 mmHg; > +8.00 D or < -12.00 D refractive error; no history of ocular surgery; no history of ocular, neurologic, or systemic disease; signal strength <8 on OCT
- Cirrus HD-OCT scans** (software ver. 3.0.0.64; Carl Zeiss Meditec, Inc)
  - 200x200 optic disc cube and 512x128 macular cube
  - Ganglion cell analysis, macular thickness analysis, and optic nerve head and retinal nerve fiber layer (RNFL) thickness analysis was performed

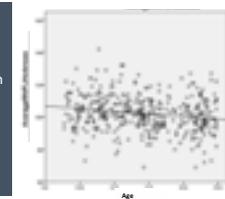
**Figure 1:** Sex distribution for Black and Hispanic patients



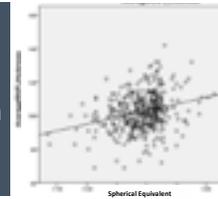
**Figure 2:** Average overall RNFL thickness by quadrant



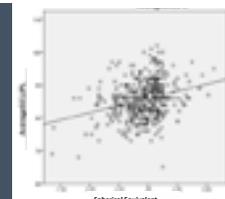
**Figure 3:** Average RNFL thickness in relation to age



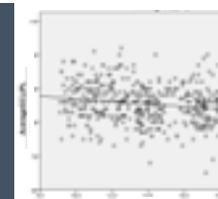
**Figure 4:** Average RNFL thickness in relation to spherical equivalent



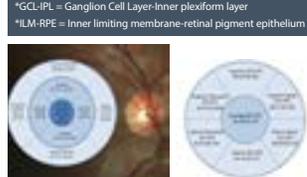
**Figure 5:** Average GCL+IPL thickness in relation to SE



**Figure 6:** Average GCL+IPL thickness in relation to age



**Figure 7:** Overall average macular parameters (central subfield thickness, ILM-RPE, and GCL+IPL)



**Table 1: Summary of current pediatric normative SD-OCT studies evaluating RNFL thickness**

Author	Age Median	n	Race	Ave RNFL thickness	OCT
Yates et al <sup>1</sup>	5.55	83	White (57 of 83)	107.6±1.3	Spectralis
Ella et al <sup>2</sup>	6.53	344	White	98.46±10.79	Cirrus
Barrio Barrio et al <sup>3</sup>	4.17	301	White	97.49±0.0	Cirrus
Taylor et al <sup>4</sup>	10.6±1.1	121	White	99.41±6.5	Cirrus
Taylor et al <sup>5</sup>	10.15	1521	White	99.41±6.5	Cirrus
Totan et al <sup>6</sup>	11.7	296	Turkish	96.91±10.31	Cirrus
Turk et al <sup>7</sup>	10.6±1.1	107	Turkish	106.45±9.47	Spectralis
Age at onset (present study)	9.97	406	Black Hispanic	102.31 ± 10.97 µm	Cirrus

## RESULTS AND DISCUSSION

- N = 406 (70.4% Black; 29.6% Hispanic; 38.2% Male) **Figure 1**.
- Average **RNFL** thickness was 102.31 ± 10.97 µm. This finding is comparable to other pediatric studies of SD-OCTs (**Table 1**). The current Cirrus normative database, which included 284 adults aged 18-84 had RNFL thickness values slightly thinner than this study.<sup>11</sup>
- RNFL** thickness followed ISNT rule (**Figure 2**), decreased with age (**Figure 3**), and increased with hyperopic SE (**Figure 4**), which agrees with the literature. See **Table 2** for average RNFL thickness by sex and race.
- RNFL** thickness decreased with increasing myopic refractive error when controlling for age.

**Table 2: Average RNFL thickness by sex and race**

	Mean RNFL thickness (µm)
Black (n=293)	102.84 ± 11.39
Hispanic (n=123)	101.04 ± 9.85
Male (n=159)	100.94 ± 11.03
Female (n=257)	103.15 ± 10.87
Overall (n=416)	102.31 ± 10.97

**Table 3: Median (range) of optic nerve head parameters**

	Vertical C/D ratio	Average C/D ratio	Cup volume (mm <sup>3</sup> )	Disc area (mm <sup>2</sup> )
Black (n=293)	0.45 (0.05-0.75)	0.48 (0.06-0.79)	0.122* (0-1.123)	3.01* (1.20-4.09)
Hispanic (n=123)	0.43 (0.05-0.72)	0.45 (0.07-0.72)	0.088* (0-0.922)	1.89* (1.04-3.55)
Male (n=159)	0.47* (0.05-0.73)	0.50 (0.07-0.79)	0.094* (0-0.922)	2.00 (1.04-4.09)
Female (n=257)	0.43* (0.05-0.70)	0.45 (0.06-0.74)	0.134* (0-1.123)	1.95 (1.20-3.55)

\*Indicates statistical significance, P<0.05 (2-tailed, spearman correlation)

**Table 4: Mean (range) of optic nerve head parameters**

	Mean central subfield thickness (µm)	Cube average thickness (µm)	Average GCL+IPL thickness (µm)
Black (n=293)	236.52 ± 21.19*	278.87 ± 13.28	86.24 ± 5.09*
Hispanic (n=123)	243.32 ± 20.26*	277.88 ± 11.13	82.85 ± 6.05*
Male (n=159)	243.58 ± 20.75*	278.49 ± 12.70	85.67 ± 6.36
Female (n=257)	239.97 ± 22.40*	276.11 ± 11.46	85.49 ± 6.39
Overall (n=416)	238.52 ± 21.57	278.21 ± 11.94	85.56 ± 5.74

Table 4: Mean (range) of optic nerve head parameters

- Black children had a larger **disc area** and **cup volume** than Hispanic children (P=0.009, P=0.007) (**Table 3 and 4**). **Disc area** is known to be larger in patients of African and Hispanic descent than whites.<sup>5,20,26,28-39</sup>
- Average C/D ratio** and **vertical C/D ratio** were similar for Blacks and Hispanics.
- Central subfield thickness** was increased in males (P=0.001, R2=0.026) and Hispanics (P=0.013, R2=0.015) but was not impacted by age (P=0.540) or SE (P=0.060), which agrees with the literature.
- Average **ganglion cell layer + inner plexiform layer** (GCL+IPL) thickness increased with hyperopic SE (P<0.001, R2=0.058) and age (P=0.014, R2=0.014) as shown in **Figures 5 and 6**. **GCL+IPL** is a sensitive measure vision loss due to glaucoma. Our study is in agreement with Totan et al, who found a mild negative correlation with average **GCL+IPL** thickness and age.<sup>19</sup> See **Table 4** and **Figure 7** for macular parameters.

## CONCLUSION

This is an ongoing study that seeks to establish a pediatric reference database for the Zeiss Cirrus HD-OCT. Data collection will continue until sample sizes are equalized between age and ethnic groups and complete results will be presented at future meetings. This data will allow for a more appropriate assessment of the optic nerve and macula for Hispanic and Black patients aged 9 to 17.

## FINANCIAL SUPPORT

This work was supported by the Illinois Society for the Prevention of Blindness (ISPB) and the Illinois Children's Healthcare Foundation.

## CONTACT INFORMATION

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# Loss of s-Cone and Scotopic Visual Function in Individuals with Retinitis Pigmentosa

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## Introduction

Some retinitis pigmentosa (RP) patients present early in life with normal rod and cone function, while others have loss of both, and some have only cone function at a young age. The progression rate varies across patients<sup>[1]</sup>.

Over 20 years ago, it was reported that some RP patients initially lose short wavelength cones<sup>[2,3]</sup> (i.e., s-cones for blue-violet colors).

At that time, it was hypothesized that a mechanism might affect a shared biochemical pathway vulnerability found in both rods and s-cones selectively, but not other cones.<sup>[2]</sup>

However, no further research on the relationship between loss of s-cone function and rod-mediated scotopic night vision has been published.

Greater loss of s-cone function, compared to l- (long wavelength) and m- (medium wavelength) cones, has also been previously documented in patients with other eye diseases; e.g. diabetic retinopathy and glaucoma<sup>[4]</sup>.

In this current study, we explored whether history of vision loss or current visual function status may predict which RP patients are susceptible to s-cone loss.

## Methods

### Participants

A total of 22 participants with a confirmed diagnosis of RP and VA better than 20/400.

Ages ranging from 25-70 years; Mean of 46 yrs.

11 Caucasians, 6 Hispanic/Latinos, 3 African Americans, and 2 Asians

12 males and 10 females

### Vision Tests

Cone Contrast Test (CCT) by Innova

- Staircase rapid test
- Evaluates red, green & blue cones
- 5-16 letters per cone type
- ~7-12 minutes total for OD & OS



Visual Acuity (VA; ETDRS Chart):

measured at 1 or 3 meters (depending on level of vision) with best correction

Goldmann Visual Field (VF): Manual Kinetic Perimetry; calculated log retinal area seen by the patient for V4e and III4e target sizes

Quick Contrast Sensitivity Function (qCSF) by Adaptive Sensory Technology:

computerized test using a Bayesian adaptive algorithm to determine contrast sensitivity at various spatial frequencies (letter sizes) and calculates the area under the curve (AUC)

AdaptDx by Maculogix:

Dark Adaptation Testing (i.e., rate and sensitivity; also determines if scotopic sensitivity is mediated by cones only, or both cones and rods) at 5 degrees from fixation

### Test Frequency

Two visits within 4 weeks apart, each lasting about 5-6 hours.

At each visit, the CCT was administered twice and all other vision tests were completed once.

All tests were administered to each eye individually and both eyes were tested.

### Self-reported history of vision loss

Age of onset for night vision loss and peripheral vision loss

Ability to see a sky full of stars at night in a rural area as a child

### Data Analyses

For analysis of CCT results, we used the best score from either visit (of 4 tests) for each eye.

According to the test manufacturer, reductions in CCT scores are significant when there is a difference of  $\geq 15$  points. We used this criteria to determine if s-cone (blue) sensitivity loss was significantly greater than the other cone types.

For the analysis of the other vision test data, we used the better of the scores between the two visits, for each eye.

We used multilevel model linear or logistic regressions to account for the correlation between subjects' eyes (STATA 13).

## Hypotheses: Why are cones lost in RP, which is due to a rod-specific mutation?

A long standing question in the field of RP research is why cones depend on rods for survival.

Listed below are a few of the hypotheses that may explain secondary cone loss in a disease that primarily results in widespread rod death:

Toxic agents released with rod death: either directly from rods or from activated retinal microglia that migrate to the photoreceptor layer<sup>[4,5]</sup>.

Rods produce a neurotrophic factor<sup>[4,5]</sup> that is required by cones for survival.

Oxidative damage from rod loss may cause death of cones<sup>[4,5]</sup>.

Reduced activity in the mTOR pathway (affects nutrition, results in starvation of cones)<sup>[6]</sup>.

Loss of structural support (due to rod loss, causes reduced contact between the cones and retinal pigment epithelium (RPE)) may result in a decrease in glucose supply and the starvation of cones<sup>[6]</sup>.

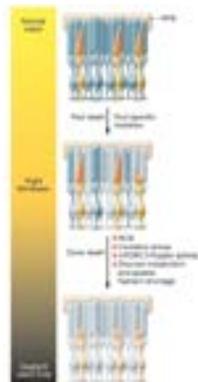


Figure 3: Rod death (represented by blue photoreceptors) is caused by a rod-specific mutation; however, subsequent cone death (represented by orange photoreceptors) can be caused by multiple factors, including increases in reactive oxygen species (ROS), increased oxidative stress, a decrease in the activity of the mTORC1 pathway, and a decrease in the supply of glucose metabolism and uptake, resulting in nutrient shortage<sup>[6]</sup>.

## Hypotheses: Why is there a greater loss of s-cones versus l- and m-cones in RP?

S-cones may be more susceptible to changes occurring at the RPE (retinal pigment epithelium) due to morphologic differences when compared to l- and m-cones.

S-cones (blue) have a longer / larger inner segment and deeper innervation projecting further into the sub-retinal space towards the RPE (i.e., closer to the RPE).

S-cones are the least numerous, representing only represent 10% of all cones, thus a loss of a few cells could have a greater impact on visual function.

S-cones are located parafoveally, outside of central vision, which is where RP patients tend to develop scotomatous and constricted visual field loss.

Figure 4

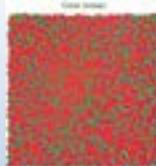


Figure 5

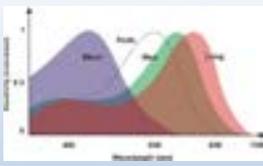


Figure 4: The image displays the location of the cone photoreceptors in the macula. S-cones are represented by the blue dots, m-cones are green dots, and the l-cones are red.

Figure 5: Displays sensitivity in terms of wavelength for each cone type, as well as rods.

## Results

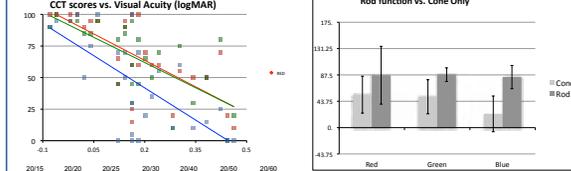
Out of 22 RP patients tested, only 15 (68%) patients had measurable cone function with the CCT.

How much VA loss was noted in patients who had no measurable CCT results? Worse than 20/50

How much VA loss was noted in patients with measurable loss of CCT sensitivity?

Since VA is mediated by cones, it's not surprising that with early VA loss (20/25-20/40) there were measured reductions in CCT scores (<75).

CCT scores according to AdaptDx Scotopic Vision: Rod function vs. Cone Only



For the 15 presumed RP patients with measurable CCT results, all of those who did not have any measurable rod function (i.e., reduced night vision mediated by cones only, n=10), had the greatest loss for s-cone sensitivity (blue) when comparing the 3 cone types, while those with rod function (n=5) had either normal s-cone function or relatively equal reductions for all 3 cones [see above figure to the right].

One of these subjects with measurable rod function in both eyes, had a slower rod response rate (i.e., slightly delayed dark adaptation) and reduced CCT scores for all 3 cone types in one eye, whereas the better eye had a normal dark adaptation rate and normal CCT scores for all 3 cone types. This case represents more advanced visual function loss in one eye due to the progression of RP.

In a subgroup analysis of the 10 patients without rod function, 2 patients (4 eyes) who had normal red and green CCT scores but reduced blue CCT results were compared to 8 patients (13 eyes) who had decreased red, green and blue CCT scores. The 2 patients who maintained normal red and green cone function had statistically significantly larger Goldmann VF log retinal area with the V4e (larger) test target (p<0.01) and significantly shorter duration of VF loss (i.e., mean 12.5 vs. 23.4 years; p=0.003).

How well does current visual function loss and/or history of vision loss predict reduced CCT results?

In multivariate regression analyses, statistically significant predictors of reduced CCT sensitivity of s-cones (blue) were: reduced VA (p<0.01) and absence of current rod function (p<0.001); While reduced l-cone (red) sensitivity was significantly predicted by reduced qCSF and decreased Goldmann VF log retinal area with the III4e test target (both p<0.01).

## Discussion / Conclusions

RP patients with normally functioning rods had normal CCT scores for all 3 cone types; then as rod function was slightly decreased (i.e., delayed dark-adaptation rate), we noted reductions in sensitivity for all 3 cone types.

Greater reduction in s-cone function than l- and m-cone function was found only in RP patients without current measurable rod function; some of these patients never had rod function (i.e., never seen stars), while others lost rod-mediated vision with disease progression.

These various patterns of rod and cone function loss in RP patients represent different stages of disease progression and/or different subtypes of RP,<sup>[1]</sup> which is genetically and phenotypically heterogeneous.

The greater loss of sensitivity for s-cones than l- and m-cones in the absence of functional rods may be likely explained by a combination of multiple mechanisms studied in animal models of RP (e.g., structural, toxic, nutritional, trophic, and/or oxidative factors). While it is not currently possible to measure these factors in the retina of human subjects, we hope that future clinical trials will explore potential therapeutic approaches related to these mechanisms and measure the responsiveness of specific cone types, which may reveal valuable insights about s-cone loss, thus furthering our knowledge of RP pathophysiology.

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## Acknowledgments

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# Acute Retinal Necrosis Secondary to Herpes Simplex Virus Type 2 in an Adolescent Male

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## INTRODUCTION

Acute retinal necrosis (ARN) is a rare, necrotizing retinopathy presenting with discrete, rapidly progressive areas of circumferential retinal necrosis with an associated panuveitis and vasculitis. Visual prognosis is poor, with an incidence of retinal detachment up to 85%, and the rate of bilateral involvement 36-70%. Standard therapy consists of intravenous acyclovir for seven to ten days followed by oral valacyclovir. Although, recent small pilot studies have reported favorable outcomes with oral antivirals only. It has been shown that patients with HSV-2 may show a relatively aggressive clinical course and variable response to treatment.

## CASE REPORT

A 17-year-old African American male presented with a painless, red, right eye for 1 month.

- PMHx: Neonatal HSV-2 encephalitis with recurrent skin rashes
- POChx: Unremarkable
- Current meds: None
- BCVA: 20/400 OD, 20/20 OS
- Anterior segment examination: 4+ cell, 2+ flare, and diffuse granulomatous keratic precipitates OD (see Figures 1).
- IOP via GAT: 43mmHg OD, 12mmHg OS
- Posterior segment evaluation: Peripheral white retinal necrosis superior nasal OD with 3+ vitritis. The retina was attached (see Figures 2, 3).
- There was no involvement OS (see Figure 4).

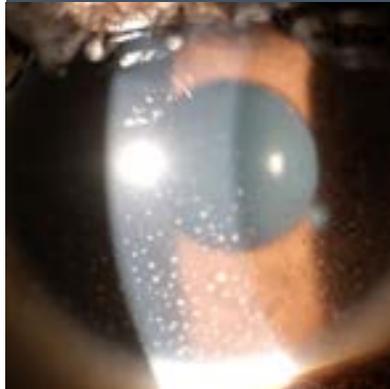
The patient was urgently referred to a retina specialist.

The retina specialist initiated oral bactrim and valacyclovir. Vitreous PCR and blood work revealed positive HSV-2 titers only. Syphilis, HIV, tuberculosis, sarcoidosis and toxoplasmosis tests were negative.

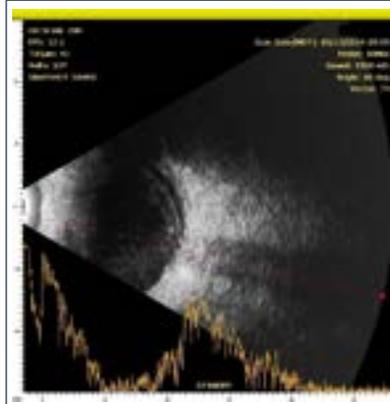
At 1 week follow-up, the vitritis was resolving and a shallow RD was present nasal. Barricade laser was performed (see Figure 5) and oral prednisone 40mg/day was added. Vision improved to 20/125, and remained stable for 7 months.

Late-onset nasal RD extending to the optic nerve (see Figure 6) and optic neuropathy occurred at month 8, and vision worsened to light perception. Pars plana vitrectomy and scleral buckle OD were performed. The left eye remains uninvolved. He will continue oral valacyclovir for prevention of bilateral involvement.

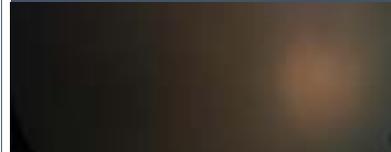
**FIGURE 1**  
Initial presentation of corneal keratic precipitates.



**FIGURE 2**  
B-scan OD demonstrating posterior vitreous cells.



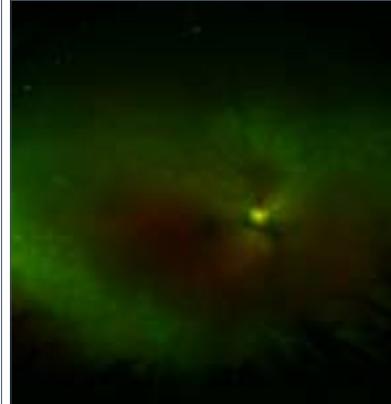
**FIGURE 3**  
Hazy view of posterior pole OD, secondary to vitritis.



**FIGURE 4**  
Unremarkable OS findings.



**FIGURE 5**  
Optomap at 3 month follow-up post-barricade laser treatment and resolution of panuveitis.



## CONCLUSION

This case demonstrates a rare retinopathy, ARN, in an adolescent male due to reactivation of neonatally acquired HSV-2 infection. This case initially responded well to oral valacyclovir and prednisone, however late-onset complications (retinal detachment and optic neuropathy) lead to poor visual outcome. ARN's predilection towards aggressive inflammatory retinal destruction requires urgent diagnosis and referral for best visual prognosis and prevention of bilateral involvement.

**FIGURE 6**  
SD-OCT demonstrating nasal retinal detachment OD extending to optic nerve.



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# Efficacy of Loteprednol Etabonate Ophthalmic Gel 0.5% for the Treatment of Evaporative Dry Eye and Meibomian Gland Dysfunction

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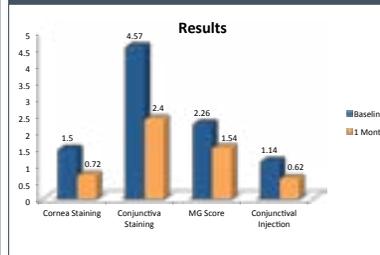
## BACKGROUND

Meibomian gland dysfunction (MGD) is the leading cause of evaporative dry eye (EDE). Evidence suggests that inflammation is associated with the development and perhaps progression of MGD.<sup>1</sup> Lotemax® (loteprednol etabonate ophthalmic gel) 0.5% Gel Drop Formation (LE Gel) has dose uniformity without shaking and fewer preservatives than loteprednol etabonate suspension 0.5%.<sup>2</sup> The mucoadhesive vehicle of LE Gel contains glycerin and propylene glycol which allows for longer contact time with the ocular surface.<sup>2</sup> The mucoadhesive vehicle coats the mucous membranes of the ocular surface similar to artificial tears. We studied the treatment effect of loteprednol etabonate ophthalmic gel 0.5% on the signs and symptoms of EDE and MGD.

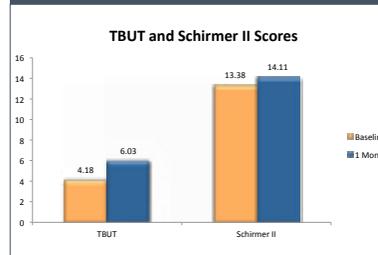
## METHODS

In an open label, prospective, multi-centered study, patients with meibomian gland dysfunction were treated with loteprednol etabonate ophthalmic gel 0.5% twice a day for thirty days bilaterally. Tear break-up time (TBUT), corneal staining, conjunctival staining, Schirmer II scores, meibomian gland lid margin score, and tear osmolality were evaluated at baseline and after 30 days of treatment. Ocular Surface Disease Index (OSDI)<sup>3</sup> and Standard Patient Evaluation Eye Dryness (SPEED)<sup>4</sup> questionnaires were administered at baseline, after two weeks of treatment, and after 30 days of treatment. Intraocular pressure (IOP) and visual acuity were measured pre and post treatment to assess patient safety.

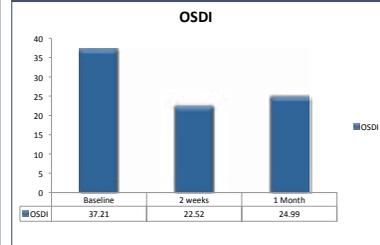
**FIGURE 1**  
Corneal Staining was reduced by 52.0% (p=0.006); Conjunctival staining was reduced by 47.5% (p=0.002); Meibomian Gland (MG) Score was reduced by 31.9% (p<0.001); Conjunctival Injection was reduced by 56.0% (p<0.001).



**FIGURE 2**  
There was a statistically significant increase (p=0.005) in tear break-up time (TBUT) after one month of treatment with an increase of 44.3%. Schirmer II score did not show a significant change from baseline (p=0.93).



**FIGURE 3**  
The Ocular Surface Disease Index (OSDI) was reduced by 32.8% after one month of treatment and patients experienced improved symptoms at 2 weeks. This statistically significant reduction of the mean OSDI (p=0.004) also resulted in an improvement by at least 1 severity level on the OSDI severity scale. OSDI scoring: 13-22 (mild), 23-32 (moderate) and 33-100 (severe).



**FIGURE 4**  
SPEED scores were reduced by 28.4% (p<0.001). Mean SPEED scores were reduced by at least one severity level after treatment. SPEED scoring: 1-5 (mild); 6-9 (moderate); 10+ (severe).



## RESULTS

30 patients (20 females and 10 males) were enrolled and completed the study. Average age was 44.7 years. After 30 days of treatment, all objective parameters evaluated showed a statistically significant improvement except for Schirmer II and Tear Osmolality (Figures 1 & 2). There was also a significant reduction of patient symptoms as measured by the OSDI and SPEED (Figures 3 & 4). IOP and visual acuity were unchanged and no adverse events took place.

## CONCLUSION

- Loteprednol etabonate ophthalmic gel 0.5% used twice a day significantly improved the clinical signs associated with EDE resulting from MGD.
- Loteprednol etabonate ophthalmic gel 0.5% showed significant reduction in severity of symptoms associated with EDE resulting from MGD.
- Results indicate that LE Gel is a safe and effective treatment option for EDE and MGD.

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# A COMPARISON OF TWO CLASSIFICATION METHODS FOR FULL THICKNESS MACULAR HOLES: A RETROSPECTIVE ANALYSIS



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## BACKGROUND

A full-thickness macular hole (FTMH) is defined as a foveal lesion that interrupts all macular layers from the internal limiting membrane to the retinal pigmented epithelium. In the 1980's, Donald M. Gass began classifying FTMH's based on clinical presentation. In 2013, the International Vitreomacular Traction Study (IVTS) started a new classification scheme, meant to eliminate any contradictions within (and modifications to) the Gass scheme and find a consensus using an Optical Coherence Tomography (OCT)-based system that used size rather than anatomy to classify macular holes.

While approximate estimates of visual function have long been noted with the Gass staging of FTMH's, the IVTS method has not yet correlated staging with visual function on a quantifiable basis, and the two methods have yet to be compared.

It is the purpose of this study to directly compare the IVTS and Gass systems for classifying idiopathic FTMH's with regard to visual function.

## METHODS

The charts of 276 patients at the Illinois Eye Institute from 2010 to 2014 were reviewed for idiopathic FTMH's, 64 of which met our criteria of patients with no concurrent macular disease (mean age=70yr).

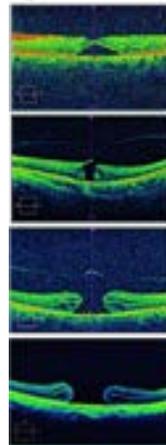
Each FTMH was categorized into Gass stage 1, 2, 3, or 4 based on anatomical presentation and IVTS stage small, medium, or large (see **Tables 1 and 2**) using measurements and anatomical analyses of the minimum hole width from OCT images (see **Figures 1 and 2**). The size of each macular hole was measured at its narrowest opening using the OCT program caliper.

The average best-corrected visual acuity (BCVA) for each stage and method was calculated and then the correlation between the BCVA logMAR and FTMH stage for each method was obtained.

Stage	# of macular holes
Stage 1	4
Stage 2	6
Stage 3	6
Stage 4	48

Stage	# of macular holes
Small	9
Medium	9
Large	46

Figure 1



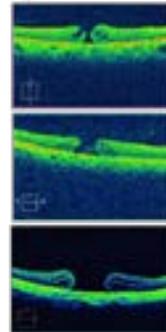
**Gass Stage 1 FTMH**  
(No separation between the vitreous and the fovea)

**Gass Stage 2 FTMH**  
(Foveal dehiscence and formation of pseudo-operculum)

**Gass Stage 3 FTMH**  
(Continued vitreo-macular traction; operculum lies anterior to foveal plane)

**Gass Stage 4 FTMH**  
(Smoothing of retinal tissue edges; complete separation of vitreous from fovea)

Figure 2



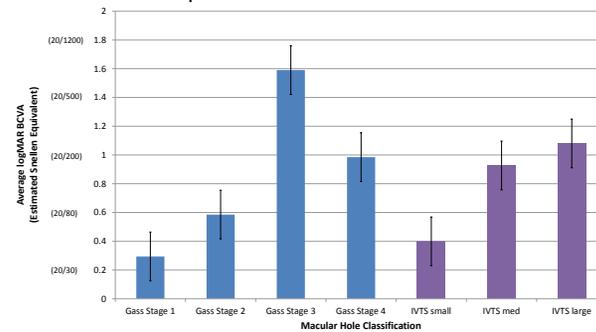
**IVTS Stage Small FTMH**  
(≤ 250µm)

**IVTS Stage Medium FTMH**  
(> 250µm but ≤ 400µm)

**IVTS Stage Large FTMH**  
(> 400µm)

## RESULTS

BCVA Comparisons of Gass vs. IVTS Classification Methods for FTMH's



Stage	# of macular holes
IVTS Small & Gass Stage 1	4
IVTS Small & Gass Stage 2	1
IVTS Small & Gass Stage 3	1
IVTS Small & Gass Stage 4	3
IVTS Medium & Gass Stage 1	0
IVTS Medium & Gass Stage 2	2
IVTS Medium & Gass Stage 3	1
IVTS Medium & Gass Stage 4	6
IVTS Large & Gass Stage 1	0
IVTS Large & Gass Stage 2	3
IVTS Large & Gass Stage 3	4
IVTS Large & Gass Stage 4	39

**Table 3** Demonstrates the crossed-classification of each FTMH considering the parameters of IVTS and Gass classification. A majority of the FTMH's from the study were found to be large, according to the IVTS, and Stage 4, according to Gass.

**Figure 3** Describes the average BCVA values in logMAR with subsequent Snellen conversions. Using the logMAR values and Gass stage, the Spearman's rho correlation coefficient (CC) was calculated to be 0.21, with a p-value equal to 0.15. With a similar calculation of logMAR values and IVTS macular hole sizes, the Spearman's rho CC was found to be 0.497 with a p-value equal to 0.01.

## DISCUSSION

The Spearman's rho correlation coefficient calculations for full-thickness macular holes categorized by Gass were not significant, whereas the correlation coefficient value of macular holes categorized by IVTS were considered to be statistically significant.

IVTS demonstrates a monotonic relationship with the logMAR Snellen visual acuities for patients with the macular holes: as the size of the macular hole increases, the logMAR increases as well.

## CONCLUSION

Unlike Gass stages, the IVTS stages are significantly correlated with visual function, which potentially aids in the diagnosis and pre-surgical management of FTMH.

There are several advantages of classifying FTMH on the basis of OCT-defined minimum linear distance, including its high correlation with successful surgery and the accuracy of the visual function correlation with macular hole size.

With a larger patient base, further studies can be conducted to determine the prognosis of full-thickness macular hole visual function while comparing Gass and IVTS methods of classification.

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# Loss of s-Cone and Scotopic Visual Function in Individuals with Retinitis Pigmentosa

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## Introduction

- Some retinitis pigmentosa (RP) patients present early in life with normal rod and cone function, while others have loss of both, and some have only cone function at a young age. The progression rate varies across patients<sup>1</sup>.
- Over 20 years ago, it was reported that some RP patients initially lose short wavelength cones<sup>2,3</sup> (i.e., s-cones for blue-violet colors).
  - At that time, it was hypothesized that a mechanism might affect a shared biochemical pathway vulnerability found in both rods and s-cones selectively, but not other cones.<sup>2</sup>
  - However, no further research on the relationship between loss of s-cone function and rod-mediated scotopic night vision has been published.
- Greater loss of s-cone function, compared to l- (long wavelength) and m- (medium wavelength) cones, has also been previously documented in patients with other eye diseases; e.g. diabetic retinopathy and glaucoma<sup>4</sup>.
- In this current study, we explored whether history of vision loss or current visual function status may predict which RP patients are susceptible to s-cone loss.

## Methods

- Participants**
- A total of 22 participants with a confirmed diagnosis of RP and VA better than 20/400.
  - Ages ranging from 25-70 years; Mean of 46 yrs.
  - 11 Caucasians, 6 Hispanic/Latinos, 3 African Americans, and 2 Asians
  - 12 males and 10 females
- Vision Tests**
- Cone Contrast Test (CCT) by Innova
    - Staircase rapid test
    - Evaluates red, green & blue cones
    - 5-16 letters per cone type
    - ~7-12 minutes total for OD & OS
  - Visual Acuity (VA; ETDRS Chart):
    - measured at 1 or 3 meters (depending on level of vision) with best correction
  - Goldmann Visual Field (VF): Manual Kinetic Perimetry; calculated log retinal area seen by the patient for V4e and III4e target sizes
  - Quick Contrast Sensitivity Function (qCSF) by Adaptive Sensory Technology:
    - computerized test using a Bayesian adaptive algorithm to determine contrast sensitivity at various spatial frequencies (letter sizes) and calculates the area under the curve (AUC)
  - AdaptDx by Maculogix:
    - Dark Adaptation Testing (i.e., rate and sensitivity; also determines if scotopic sensitivity is mediated by cones only, or both cones and rods) at 5 degrees from fixation
- Test Frequency**
- Two visits within 4 weeks apart, each lasting about 5-6 hours.
  - At each visit, the CCT was administered twice and all other vision tests were completed once.
  - All tests were administered to each eye individually and both eyes were tested.
- Self-reported history of vision loss**
- Age of onset for night vision loss and peripheral vision loss
  - Ability to see a sky full of stars at night in a rural area as a child
- Data Analyses**
- For analysis of CCT results, we used the best score from either visit (of 4 tests) for each eye.
  - According to the test manufacturer, reductions in CCT scores are significant when there is a difference of  $\geq 15$  points. We used this criteria to determine if s-cone (blue) sensitivity loss was significantly greater than the other cone types.
  - For the analysis of the other vision test data, we used the better of the scores between the two visits, for each eye.
  - We used multilevel model linear or logistic regressions to account for the correlation between subjects' eyes (STATA 13).



## Hypotheses: Why are cones lost in RP, which is due to a rod-specific mutation?

A long standing question in the field of RP research is why cones depend on rods for survival.

Listed below are a few of the hypotheses that may explain secondary cone loss in a disease that primarily results in widespread rod death:

- Toxic agents released with rod death: either directly from rods or from activated retinal microglia that migrate to the photoreceptor layer<sup>4,5</sup>.
- Rods produce a neurotrophic factor<sup>4,5</sup> that is required by cones for survival.
- Oxidative damage from rod loss may cause death of cones<sup>6,5</sup>.
- Reduced activity in the mTOR pathway (affects nutrition, results in starvation of cones)<sup>4</sup>.
- Loss of structural support (due to rod loss, causes reduced contact between the cones and retinal pigment epithelium (RPE)) may result in a decrease in glucose supply and the starvation of cones<sup>4</sup>.

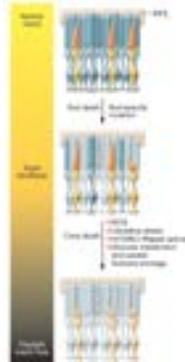


Figure 3: Rod death (represented by blue photoreceptors) is caused by a rod-specific mutation, however, subsequent cone death (represented by orange photoreceptors) can be caused by multiple factors, including increases in reactive oxygen species (ROS), increased oxidative stress, a decrease in the activity of the mTORC1 pathway, and a decrease in the supply of glucose metabolism and uptake, resulting in nutrient shortages<sup>4</sup>.

## Hypotheses: Why is there a greater loss of s-cones versus l- and m-cones in RP?

- S-cones may be more susceptible to changes occurring at the level of the RPE (retinal pigment epithelium) due to morphologic differences when compared to l- and m-cones.
  - S-cones (blue) have a longer / larger inner segment and deeper innervation projecting further into the sub-retinal space towards the RPE (i.e., closer to the RPE).
- S-cones are the least numerous, representing only represent 10% of all cones, thus a loss of a few cells could have a greater impact on visual function.
- S-cones are located parafoveally, outside of central vision, which is where RP patients tend to develop scotomatous and constricted visual field loss.

Figure 4

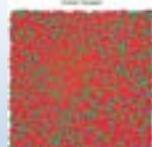


Figure 5

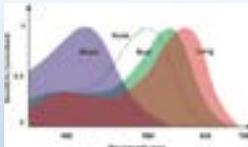
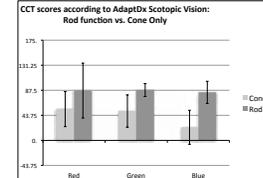
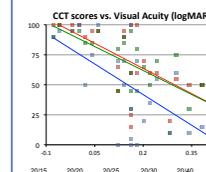


Figure 4: The image displays the location of the cone photoreceptors in the macula. S-cones are represented by the blue dots, m-cones are green dots, and the l-cones are red. Figure 5: Displays sensitivity in terms of wavelength for each cone type, as well as rods.

## Results

- Out of 22 RP patients tested, only 15 (68%) patients had measurable cone function with the CCT.
- How much VA loss was noted in patients who had no measurable CCT results? Worse than 20/50
- How much VA loss was noted in patients with measurable loss of CCT sensitivity?
- Since VA is mediated by cones, its not surprising that with early VA loss (20/25-20/40) there were measured reductions in CCT scores (<75).

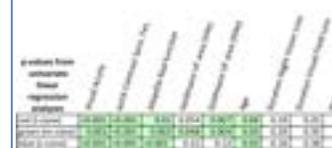


- For the 15 presumed RP patients with measurable CCT results, all of those who did not have any measurable rod function (i.e., reduced night vision mediated by cones only; n=10), had the greatest loss for s-cone sensitivity (blue) when comparing the 3 cone types, while those with rod function (n=5) had either normal s-cone function or relatively equal reductions for all 3 cones [see above figure to the right].

This case represents more advanced visual function loss in one eye due to the progression of RP.

- In a subgroup analysis of the 10 patients without rod function, 2 patients (4 eyes) who had normal red and green CCT scores but reduced blue CCT results were compared to 8 patients (13 eyes) who had decreased red, green and blue CCT scores. The 2 patients who maintained normal red and green cone function had statistically significantly larger Goldmann VF log retinal area with the V4e (larger) test target (p=0.01) and significantly shorter duration of VF loss (i.e., mean 12.5 vs. 23.4 years; p=0.003).

•How well does current visual function loss and/or history of vision loss predict reduced CCT results?



In multivariate regression analyses, statistically significant predictors of reduced CCT sensitivity of s-cones (blue) were: reduced VA (p=0.01) and absence of current rod function (p=0.001). While reduced l-cone (red) sensitivity was significantly predicted by reduced qCSF and decreased Goldmann VF log retinal area with the III4e test target (both p<0.01).

## Discussion / Conclusions

- Greater reduction in s-cone function than l- and m-cone function was found only in RP patients without current measurable rod function; some of these patients never had rod function (i.e., never seen stars), while others lost rod-mediated vision with disease progression.
- These various patterns of rod and cone function loss in RP patients represent different stages of disease progression and/or different subtypes of RP<sup>11</sup> which is genetically and phenotypically heterogeneous.
- The greater loss of sensitivity for s-cones than l- and m-cones in the absence of functional rods may be likely explained by a combination of multiple mechanisms studied in animal models of RP (e.g., structural, toxic, nutritional, trophic, and/or oxidative factors). While it is not currently possible to measure these factors in the retina of human subjects, we hope that future clinical trials will explore potential therapeutic approaches related to these mechanisms and measure the responsiveness of specific cone types, which may reveal valuable insights about s-cone loss, thus furthering our knowledge of RP pathophysiology.

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## Acknowledgments

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# Neovascular Glaucoma with Prominent Iris Bombe Status–Post Funnel Retinal Detachment

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## ABSTRACT

This case represents chronic iris neovascularization status-post funnel retinal detachment, resulting in angle closure secondary to posterior synechiae and prominent iris bombe. Evisceration recommended preventing future phthisis bulbi.

## PERTINENT FINDINGS

22-year-old African American Male presents with a red, painful right eye with photophobia and intermittent headaches for 1-2 months.

**Ocular History:** Constant Right Esotropia (congenital), Strabismus surgery (3 years old), (+) degenerative myopia OD, (+) retinal detachment longstanding, (-) trauma, (+) amblyopia OD

**Medical History:** (-) Diabetes, (+) Hypertension, (-) autoimmune disorders

**Medications:** None

**VA (sc):** OD: Light Perception, NIPH OS: 20/40 PH: 20/25

**1 day follow-up:** OD: Light Perception, NIPH

**2 week follow-up:** OD: NLP

### Slit Lamp Findings:

- 1+ injection OD
- 2+ corneal edema with superficial punctate keratitis OD
- Angle: Grade 0, closed 360 degrees
- AC: 1+ cell, possible flare
- Iris: NVI, posterior synechiae 360 degrees, iris bombe
- Lens: opacified lens, no red reflex present

**IOP:** OD: 26 OS: 18

**1 day follow-up:** OD: 17 OS: 18

**2 week follow-up:** OD: 34 OS: 17

**DFE:** OD: not visible OS: within normal limits, (-) vitritis, (-) neovascularization

**B-scan analysis:** funnel retinal detachment with possible vitreous hemorrhage OD

**Diagnosis:** Neovascular Glaucoma  
Phthisis Bulbi  
Funnel Retinal Detachment

**Differential Diagnosis:** Chronic Angle Closure Glaucoma, Acute Angle Closure Glaucoma, Inflammatory Glaucoma, Uveitis

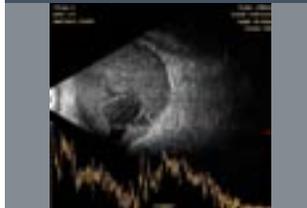
## DISCUSSION

- Most common causes: diabetic retinopathy, central retinal vein occlusion, and branch retinal vein occlusion
- Other possible causes: ocular ischemic syndrome, tumors, chronic inflammation, chronic retinal detachment, and radiation therapy
- Treatment:
  - Initial: Combigan BID OD, referral to retinal specialist
  - 1 day follow-up: Continue Combigan BID OD until next appointment
  - Retinal Specialist appointment (2 weeks)
    - Continue Combigan BID OD, start Cyclopentolate 1% BID OD, and start Pred Forte BID OD to help improve discomfort
    - Recommend evisceration OD due to suspected phthisis bulbi
    - Monitor OS annually

**FIGURE 1:** Anterior Segment Ocular Coherence Tomography OD demonstrating angle closure with prominent iris bombe and posterior synechiae



**FIGURE 2:** B scan demonstrating funnel-retinal detachment with possible vitreous hemorrhage OD



**FIGURE 3:** Chronic neovascularization and lens yellowing OD



**FIGURE 4:** Chronic iris neovascularization and iris bombe OD



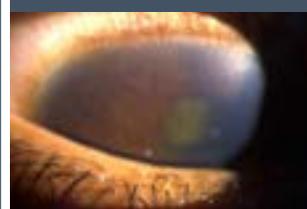
**FIGURE 5:** Chronic iris neovascularization, iris bombe, and lens yellowing OD



**FIGURE 6:** Chronic iris neovascularization and iris bombe OD



**FIGURE 7:** Iris neovascularization extending to the angle



## PATHOGENESIS

- Retinal ischemia causes the release of vascular endothelial growth factor (VEGF), causing the growth of new, leaky blood vessels. These leaky blood vessels travel from the retina forward through the pupil, onto the iris and into the angle. The blood vessels as well as the fibroblastic membranes form and block the trabecular meshwork.

## CONCLUSION

- Although IOP lowering is important for the comfort of the patient, even with IOP control, 3-48% will still lose light perception, so identifying the cause of neovascularization and starting treatment, such as anti-VEGF or PRP, are most pertinent in these cases
- If no light perception or no visual potential of the eye, evisceration/enucleation is recommended to prevent phthisis bulbi and to improve cosmesis
- Patients must understand the importance of treatment and maintenance nature of neovascular glaucoma or vision may be lost

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# Meibomian Gland Dysfunction in Pre- and Post-Menopausal Women

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## INTRODUCTION

Meibomian Gland Dysfunction (MGD) is commonly associated with the signs and symptoms of ocular surface disease in dry eye patients (up to 86%). In 2010, the International Workshop on Meibomian Gland Dysfunction developed a recommended definition for MGD:

*"Meibomian gland dysfunction (MGD) is a chronic, diffuse abnormality of the meibomian glands, commonly characterized by terminal duct obstruction and/or qualitative/quantitative changes in the glandular secretions. This may result in alterations of the tear film, symptoms of eye irritation, clinically apparent inflammation, and ocular surface disease."*

The production of lipids and meibum from the meibomian glands is controlled by neuronal, vascular, and hormonal factors. Common perception among practitioners is that females may be particularly affected in peri- and post-menopausal years secondary to these hormonal regulatory controls. This study looked to compare differences for MGD patients of pre- and post-menopausal ages.

## METHODS

- Subjects over age 18 were enrolled.
- Subjects were administered the Ocular Surface Disease Index (OSDI), Total Ocular Symptom Score (TOSS), and a 9 symptom questionnaire that asked both frequency and intensity of dry eye.
- Subjects were then classified based on Meibomian Gland (MG) expression grade (1.0 or worse).
- The worst eye results were taken.

**Figure 1:** Meibomian Gland Dysfunction Grade 1



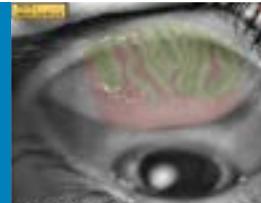
## RESULTS

- 56 females older than 40 years of age were enrolled.
- 42 females younger than 40 years of age were enrolled.
- None of the clinical tests nor symptom surveys showed a statistical significance between the two cohorts for MGD expression grade (p=0.285). Figures 2-4

**Figure 2:** Meibography in a Patient Younger than 40

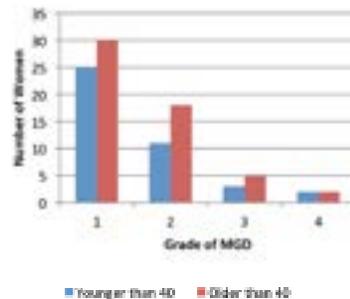


**Figure 3:** Meibography in a Patient Older than 40

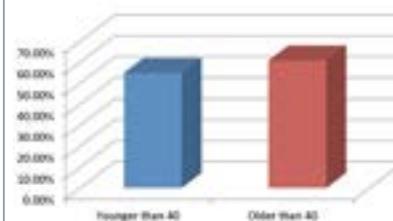


- None of the clinical tests nor symptom surveys showed a statistical significance between the two cohorts for OSDI (p=0.146).
- In the younger than 40 group, 23 out of 42 subjects had MGD (54.8%) and in the older than 40 group, 34 out of the 56 subjects had MGD (60.7%). Figure 5
- The over than 40 females reported more constant frequency of their worst symptom and higher sensitivity to bright light compared to the under 40 females.

**Figure 4:** Grade of MGD in Pre- and Post- Menopausal Women



**Figure 5:** Presence of MGD in Females by Age



## CONCLUSIONS

Past perceptions of dry eye patient demographics may age bias clinicians to under diagnose Meibomian Gland Dysfunction (MGD). No significant differences found between the two cohorts suggest that hormonal controls might play a more minor role in MGD than as previously thought. As MGD is one of the most common causes for the development of ocular surface disease symptoms (specifically those associated with dry eye conditions), younger women with dry eye symptoms need careful ocular surface evaluation, just like their older cohort.

## DISCLOSURES

J Harthan: Bausch +Lomb, Contamac, Metro Optics  
J Kwan: Johnson & Johnson Vision Care, Inc.  
D Opitz: Allergan, Bausch + Lomb, Glaucox, NiCox  
M Hom: Abbott Medical Optics , Alcon, Allergan, Bausch + Lomb, Shire, Valeant

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# Differential Diagnosis of 'Hordeolum' in an HIV+ Patient

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## BACKGROUND

Kaposi Sarcoma (KS) is the second most common malignancy encountered in HIV occurring in 20-30% of HIV+ population. Epidemic KS is an angio-proliferative cancer caused by human herpesvirus-8 typically occurring in an immunosuppressed host.

KS presents as proliferative multi-centric lesions, often with neovascularization resulting in a violaceous appearance. The lesions may initially present as flat 'patch lesions' up to several centimeters in size but progress over time to a more nodular, edematous appearance.

Lesions evolving in the pulmonary system are the most life threatening form of KS and the second most common non-cutaneous site of involvement after the oral cavity and GI tract. Early in the AIDS epidemic, KS was the presenting sign of almost 50% of AIDS patients and KS had a high mortality rate; before highly active antiretroviral therapy (HAART), pulmonary epidemic KS reportedly had 90% 5 year mortality rate. After HAART the incidence and risk of mortality from epidemic KS has decreased significantly with a 5 year survival rate of 72%; however, both remain elevated in areas with poor access to treatment.

Prevalence of ocular lesions in KS is approximately 20% and may affect the eyelid, conjunctiva, plica semilunaris, caruncle, and both the lacrimal sac and gland. Ocular KS may clinically appear similar to chronic subconjunctival hemorrhage, pyogenic granuloma, hordeolum, nodular scleritis, lymphangioma, hemangioma, and malignant melanoma.

## CASE REPORT

A 52 year old presented to the IEL urgent care clinic with concern over a hard, elevated purple red nodule on his right lower lid. A month prior he presented to a local hospital's Emergency Department with the same complaint and was diagnosed with a hordeolum for which he was prescribed gentamicin ointment. He reported using the ointment for the last month without improvement in lid appearance. He reported the lesion progressing in size over a two month period and noticed a smaller hard, elevation on his right nasal upper lid two weeks ago.

Examination of the right eye revealed a multi-nodular purple red subdermal elevation (12 mm length x 10 mm width x 7 mm elevation) on the right lower lid with lesion extension through the septum to an associated red, flat patch lesion on the palpebral conjunctiva. A hard subdermal mildly erythemic elevation was noted on the nasal upper lid without conjunctival involvement. Examination of the left eye uncovered two flat, red patch lesions on the nasal palpebral conjunctiva. The patient was unaware of the left side conjunctival lesions. Neither set of lesions were painful nor any discharge present.

Systemic history included HIV diagnosed eleven years ago with self-reported CD4 count 78 cells/mm<sup>3</sup> and undetectable viral load measured three months before. Current medications included Norvir, Isentress, Efavirenz, and Prezista; all components of HAART. The patient denied dermal lesions elsewhere on the body.

Lesion biopsy confirmed the tentative diagnosis of epidemic KS. An attempt was made to excise the largest lesion on the right lid but was stopped secondary to extensive bleeding of the lesion mid-procedure. The patient was referred to oncology and received 30 Gy over several irradiation sessions.

## DIFFERENTIAL DIAGNOSIS

Chronic sub-conjunctival hemorrhage: acute, often painless, superficial bright-red patch presenting between the bulbar conjunctiva and the sclera resulting from bleeding from conjunctival vessel; associated with increased venous pressure, trauma, and vascular diseases, recurrent hemorrhages may be associated with hematologic disease

Hemangioma: congenital vascular malformation densely composed of blood-filled capillaries, presents at birth as raised painless purple red lobulated lesions; may be found cutaneous, subcutaneous, on the conjunctiva, and on the retina

Pyogenic granuloma: acquired capillary hemangioma, usually conjunctival, result of trauma, friction, or hormonal changes; presents as painless, smooth, raised, red lesion which bleeds easily

Lymphangioma: congenital or acquired lymphatic malformation composed of grape-like clusters of lymph or serosanguinous filled vesicles; presents as pinkish to dark red raised clusters; spectrum of presentation

Hordeolum: acute suppurative inflammation of meibomian gland or glands of Zeiss or Moll; presents as tender erythematous nodule at or near lid margin; benign in nature, may be recurrent

Nodular scleritis: acquired painful, sectoral, red-violet injection of deep scleral vessels with immobile elevated nodule; half of all cases associated with systemic disease, often immune-mediated disease

Malignant melanoma: acquired irregular tan to dark lesion of the skin or conjunctiva, may be elevated; significant risk for metastatic disease and mortality

## CONCLUSION

Ocular KS is important to include as a differential of external ocular lesions in HIV+ patients. Incidence of epidemic KS has decreased with HAART; however, the disease is often multi-centric and may be life threatening if pulmonary, visceral, or extensive lymph node involvement occurs as well as potentially being the first presenting sign of AIDS.

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KS EXTERNAL OD



KS EXTERNAL



KS OS 1



KS OS 2



KS OD 1



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# A COMPARISON OF TWO CLASSIFICATION METHODS FOR FULL THICKNESS MACULAR HOLES: A RETROSPECTIVE ANALYSIS

ICO LOGO HERE

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## BACKGROUND

A full-thickness macular hole (FTMH) is defined as a foveal lesion that interrupts all macular layers from the internal limiting membrane to the retinal pigmented epithelium. In the 1980's, Donald M. Gass began classifying FTMH's based on clinical presentation. In 2013, the International Vitreomacular Traction Study (IVTS) started a new classification scheme, meant to eliminate any contradictions within (and modifications to) the Gass scheme and find a consensus using an Optical Coherence Tomography (OCT)-based system that used size rather than anatomy to classify macular holes.

While approximate estimates of visual function have long been noted with the Gass staging of FTMH's, the IVTS method has not yet correlated staging with visual function on a quantifiable basis, and the two methods have yet to be compared.

It is the purpose of this study to directly compare the IVTS and Gass systems for classifying idiopathic FTMH's with regard to visual function.

## METHODS

The charts of 276 patients at the Illinois Eye Institute from 2010 to 2014 were reviewed for idiopathic FTMH's, 64 of which met our criteria of patients with no concurrent macular disease (mean age=70yr).

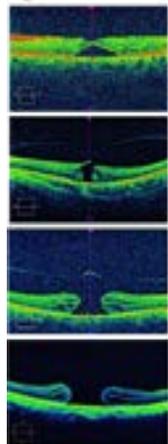
Each FTMH was categorized into Gass stage 1, 2, 3, or 4 based on anatomical presentation and IVTS stage small, medium, or large (see **Tables 1 and 2**) using measurements and anatomical analyses of the minimum hole width from OCT images (see **Figures 1 and 2**). The size of each macular hole was measured at its narrowest opening using the OCT program caliper.

The average best-corrected visual acuity (BCVA) for each stage and method was calculated and then the correlation between the BCVA logMAR and FTMH stage for each method was obtained.

Stage	# of macular holes
Stage 1	4
Stage 2	6
Stage 3	6
Stage 4	48

Stage	# of macular holes
Small	9
Medium	9
Large	46

Figure 1



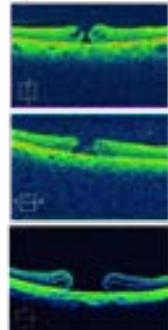
**Gass Stage 1 FTMH**  
(No separation between the vitreous and the fovea)

**Gass Stage 2 FTMH**  
(Foveal dehiscence and formation of pseudo-operculum)

**Gass Stage 3 FTMH**  
(Continued vitreo-macular traction; operculum lies anterior to foveal plane)

**Gass Stage 4 FTMH**  
(Smoothing of retinal tissue edges; complete separation of vitreous from fovea)

Figure 2



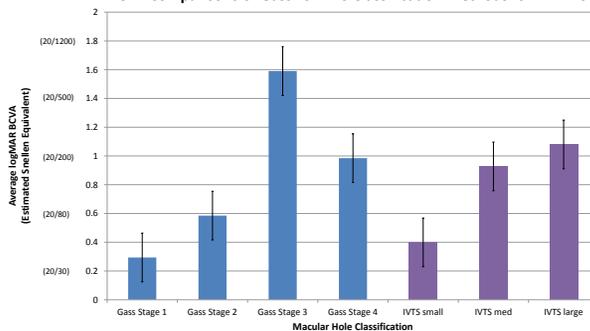
**IVTS Stage Small FTMH**  
(≤ 250µm)

**IVTS Stage Medium FTMH**  
(> 250µm but ≤ 400µm)

**IVTS Stage Large FTMH**  
(> 400µm)

## RESULTS

BCVA Comparisons of Gass vs. IVTS Classification Methods for FTMH's



Stage	# of macular holes
IVTS Small & Gass Stage 1	4
IVTS Small & Gass Stage 2	1
IVTS Small & Gass Stage 3	1
IVTS Small & Gass Stage 4	3
IVTS Medium & Gass Stage 1	0
IVTS Medium & Gass Stage 2	2
IVTS Medium & Gass Stage 3	1
IVTS Medium & Gass Stage 4	6
IVTS Large & Gass Stage 1	0
IVTS Large & Gass Stage 2	3
IVTS Large & Gass Stage 3	4
IVTS Large & Gass Stage 4	39

**Table 3** Demonstrates the crossed-classification of each FTMH considering the parameters of IVTS and Gass classification. A majority of the FTMH's from the study were found to be large, according to the IVTS, and Stage 4, according to Gass.

**Figure 3** Describes the average BCVA values in logMAR with subsequent Snellen conversions. Using the logMAR values and Gass stage, the Spearman's rho correlation coefficient (CC) was calculated to be 0.21, with a p-value equal to 0.15. With a similar calculation of logMAR values and IVTS macular hole sizes, the Spearman's rho CC was found to be 0.497 with a p-value equal to 0.01.

## DISCUSSION

The Spearman's rho correlation coefficient calculations for full-thickness macular holes categorized by Gass were not significant, whereas the correlation coefficient value of macular holes categorized by IVTS were considered to be statistically significant.

IVTS demonstrates a monotonic relationship with the logMAR Snellen visual acuities for patients with the macular holes: as the size of the macular hole increases, the logMAR increases as well.

## CONCLUSION

Unlike Gass stages, the IVTS stages are significantly correlated with visual function, which potentially aids in the diagnosis and pre-surgical management of FTMH.

There are several advantages of classifying FTMH on the basis of OCT-defined minimum linear distance, including its high correlation with successful surgery and the accuracy of the visual function correlation with macular hole size.

With a larger patient base, further studies can be conducted to determine the prognosis of full-thickness macular hole visual function while comparing Gass and IVTS methods of classification.

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# Effect of Occlusion Therapy in Eyes with Unilateral Optic Nerve Hypoplasia – A Case Series

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## BACKGROUND

- Optic nerve hypoplasia (ONH) is a congenital condition that results in a small optic disc and/or diminished number of nerve fibers. Visual acuity (VA) can vary widely in patients with ONH, ranging from 20/20 to NLP.
- Amblyopia may develop in patients with ONH. Vision loss from amblyopia is in addition to that from ONH. Occlusion therapy is often performed as part of the treatment of amblyopia.

## PURPOSE

- To investigate visual acuity outcome following a trial of occlusion therapy for amblyopia in children with unilateral optic nerve hypoplasia (ONH)

## METHODS

- Three patients with unilateral ONH were prescribed a 12-week trial of patching treatment (of the dominant eye). Two hours per day of patching were prescribed for patients with VA from 20/40 to 20/80 and six hours per day for patients with VA of 20/100 or worse.
- VA was measured with single letters or symbols with crowding bars.

## CASE REPORTS

### CASE 1

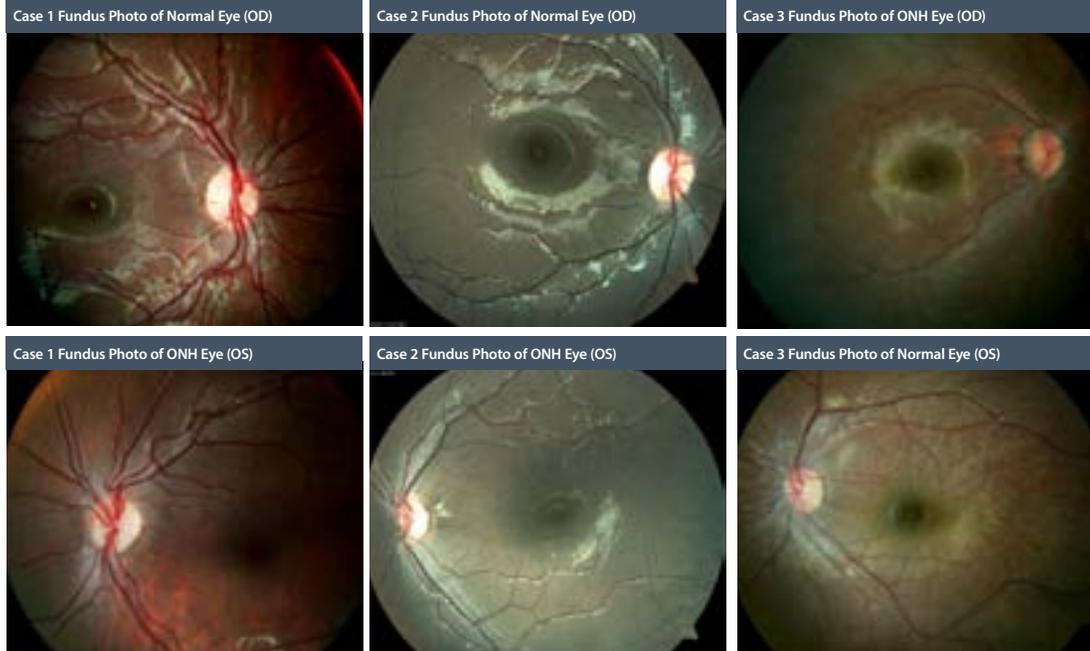
A 5-year-old Hispanic girl with ONH OS started with VA of 20/25 OD and 20/100 OS, through cycloplegic refraction of +0.50-1.00 x 180 and -2.50-0.50 x 180 respectively. There was no strabismus present. Pupillary function was normal. After patching was completed with a reported compliance of >80%, the patient's VA was 20/25 OD and 20/40 OS. Amblyopia was attributed to the anisometropia.

### CASE 2

A 4-year-old African American girl with ONH OS had VA of 20/25 OD and 20/800 OS, through cycloplegic refraction of +2.25 sph. and +2.50-0.50 x 180 respectively. Ocular alignment testing revealed a 45Δ constant left esotropia at distance and near. Visuoscopy showed unsteady eccentric fixation OS. There was a left relative afferent pupillary defect (RAPD). After patching was completed with a reported compliance of >80%, the patient's VA was 20/20 OD and 20/160 OS. Amblyopia was attributed to the unilateral strabismus.

### CASE 3

A 13-year-old African American girl with ONH OD had VA of 20/160 OD and 20/40 OS, through cycloplegic refraction of -8.50-2.75 x 180 and -3.75-3.25 x 180 respectively. A right RAPD was present. No strabismus was observed. Following patching with poor reported compliance of <15%, the patient's VA was not improved OD. Amblyopia was attributed to the anisometropia.



### Summary of Clinical Findings of Unilateral ONH patients

Case #	Age	ONH Eye	Strabismus	APD	Cycloplegic Refraction	Visual Acuity of ONH Eye		Compliance with Patching
						Pre-Tx	Post-Tx	
1	5	OS	No	No	OD: +0.50-1.00 x 180 OS: -2.50-0.50 x 180	20/100 vs. 20/40	>80%	
2	4	OS	45Δ CLET	Yes	OD: +2.25 sph OS: +2.50-0.50 x 180	20/800 vs. 20/160	>80%	
3	13	OD	No	Yes	OD: -8.50-2.75 x 180 OS: -3.75-3.25 x 180	20/160 vs. 20/160	<15%	

## CONCLUSIONS

- Patching treatment improved VA in the two younger patients with unilateral ONH.
- Compliance and age appeared to be more important in contrast to presence of strabismus, refractive error, or pupillary abnormality.
- A trial period of patching may be advisable for patients with unilateral ONH.

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# Third Nerve Palsy Caused by Giant Cell Arteritis in an Elderly Hispanic Female

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## BACKGROUND

Giant cell arteritis (GCA) is a systemic immune-mediated vasculitis that most commonly affects individuals greater than 50 years of age and affects women 2-6 times more often than men. The most common associated symptoms include headache, scalp tenderness and jaw claudication. Although GCA is a rare etiology of a third nerve palsy, it is crucial to be able to identify, understand and manage these patients promptly to decrease the risk of vision loss. This case represents GCA with third nerve palsy in an elderly Hispanic female.

## PERTINENT FINDINGS

- 70 y/o Hispanic female presents with total ptosis and periorbital pain of the left eye for three days. Her symptoms have been getting progressively worse. She rates the pain as 9/10 and described it as a dull ache that extends to the forehead and temporal region
- She denies double vision or a decrease in vision
- She reports falling two days prior to onset of symptoms, however denies hitting her head

**Medical History:** Diabetes mellitus Type 2, hypertension, hypercholesterolemia

**Ocular History:** Cataract extraction OD, OS, dry eye syndrome OD, OS

**Medications:** Terbinafine, Aspirin, Metformin, Lisinopril, Klor-Con 10, Gabapentin, Furosemide, Famotidine, Clopidogrel, Carvedilol, Atorvastatin

**Allergies:** Penicillin

**Exam Findings:**

**Initial exam (Day 1)**

• **Distance unaided Visual Acuity:**

OD: 20/25 PH: NI  
OS: 20/40+2 PH: NI (with lid holding)

• **Pupils: PERRL (-) APD**

• **EOMs:** FROM OD

Moderate exotropia present, moderate adduction, elevation and depression deficits OS (see figures 1, 2, 3, 4)

• **CVF:** FTFC OD, OS with lid holding

• **Slit lamp findings:**

**Anterior:** OD-centered PCIOI  
OS-total ptosis with lower lid lag (see figure 5), PCIOI with 1+ PCO

**Posterior:** OD-pseudo pallor of optic nerve, small heme at inferior/nasal part of disc, AV nicking, scattered microaneurysms. (-) edema of optic nerve  
OS-pseudo pallor of optic nerve, (-) edema of optic nerve (photo OU)

- **Intraocular pressures:** 17 mmHg OD, OS
- **Blood pressure:** 143/69 mmHg
- **Blood sugar:** 269 mg/dL as of that morning, HbA1C: 8.0 % as of 1 month prior
- Patient was referred to ER for GCA work-up

**Pertinent findings from ER visit (Day 1)**

- ESR: 52mm/hr (elevated)
- Blood Glucose: 318 mg/dL (elevated)
- CRP: unknown
- Platelet count 219 K/UL (within normal limits)
- CT head scan without contrast: No acute intracranial abnormality

**Follow-up exam (Day 4)**

• **Distance unaided Visual Acuity**

OD: 20/25 PH: NI  
OS: 20/40+2 PH: NI (with lid holding)

• **Pupils: PERRL (-) APD**

• **EOMs:** FROM OD

Moderate exotropia present, moderate adduction, elevation and depression deficits OS

• **CVF:** FTFC OD, OS with lid holding

• **Slit lamp findings:**

**Anterior:** OD-centered PCIOI  
OS-total ptosis with lower lid lag, PCIOI with 1+ PCO, white mucous strands in tear film

**Posterior:** OD-pseudo pallor of optic nerve, small heme at inferior/nasal part of disc, AV nicking, scattered microaneurysms, (-) edema of optic nerve  
OS-pseudo pallor of optic nerve, (-) edema of optic nerve

• **Intraocular pressures:** 17 mmHg OD, 18 mmHg OS

• **Blood pressure:** 170/90 mmHg

**Primary Diagnosis:** Incomplete third nerve palsy without pupil involvement secondary to GCA

**Differential Diagnoses:** Complete pupil-sparing third nerve palsy, complete pupil-involving third nerve palsy, orbital malignancy, syphilis

**Potential Etiologies of Third Nerve Palsy:** Aneurysm, trauma, neoplasm, multiple sclerosis, microvascular ischemia secondary to diabetes, hypertension or giant cell arteritis

**Secondary Diagnoses:** Mild non-proliferative diabetic retinopathy OD, dry eye syndrome OS>OD, posterior capsular opacification OS

## DISCUSSION

- Incomplete third nerve palsy due to presence of limited motility superiorly, nasally and inferiorly with total ptosis
- Microvascular ischemia secondary to poorly controlled blood sugar and blood pressure as the etiology was considered, however need to rule out GCA based on patient demographics and new onset of periorbital pain
- Referred patient to ER for GCA work-up including Westergren SED rate (ESR), C-reactive protein (CRP), platelet testing and temporal artery biopsy
- ER doctor did not order temporal artery biopsy for this patient
- SED rate was elevated. Patient was diagnosed with giant cell arteritis by ER doctor and referred to PCP for management
- Patient needs to be monitored closely to rule out pupillary involvement, which can be delayed by seven days after initial onset of symptoms
- CT scan was performed on this patient, however MRI and MRA of the brain and circle of Willis can be performed to rule out mass or aneurysm
- It would be prudent to run an MRI and MRA on this patient because of her recent fall just prior to onset of symptoms



## TREATMENT

- Giant cell arteritis is treated with corticosteroids
- Typically a high dose of intravenous methylprednisolone, followed by 80-100 mg/d po of prednisone is recommended
- Immediate treatment is important to decrease risk of vision loss associated with arteritic anterior ischemic optic neuropathy
- Patient was treated initially with 20 mg/d po of prednisone and referred to her PCP for management
- The ER physician may have prescribed a lower dose of prednisone to reduce the potential risk of diabetic complications
- Recovery after onset of treatment can be as rapid as several days

## CONCLUSION

- Monitor for pupillary involvement for up to seven days
- Laboratory testing to determine ESR and CRP levels should be performed on all patients greater than 50 years of age who present with a third nerve palsy to rule out GCA
- Not all patients with GCA will have periorbital pain, scalp tenderness or jaw claudication at initial exam
- Prompt diagnosis and treatment is vital in decreasing risk of vision loss

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# Visual Structure Biomarkers of Traumatic Brain Injury (TBI) in Contact Sport Athletes using Optical Coherence Tomography (OCT)

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## INTRODUCTION

It is estimated sports-related concussion afflicts 3.8 million American athletes each year. Recent research suggests that repeated head trauma may be associated with the development of neurodegenerative changes in the brain. These changes in the brain can begin months, years, or even decades after the last brain trauma and are not visible on standard brain scans. As the eye is an extension of the brain, this study investigates the retinal structure in active and retired professional contact sport athletes with a high risk of concussion (football, boxing, hockey, soccer and rugby) compared to age-matched controls to determine if visual structure measures may be an in vivo clinical biomarker to detect nerve damage related to repeated head trauma.

## METHODS

Patients with history of elite-level, high TBI risk contact sport exposure (Boxing, n=13), moderate TBI risk contact sport exposure (Football, n=20) and elite-level, non-contact sport athletes or non-athlete controls (n=25) underwent Spectral-Domain OCT optic nerve and macula scans.

## RESULTS

Boxing athletes demonstrated significant thinning in average Retinal Nerve Fiber Layer (RNFL) compared to Controls (Boxers:  $84.31 \pm 9.81 \mu\text{m}$  vs. Controls:  $94.72 \pm 11.10$ ,  $p = 0.01$ , Figure 1) and Football athletes (Boxers:  $84.31 \pm 9.81 \mu\text{m}$  vs. Football:  $93.70 \pm 9.03$ ,  $p = 0.04$ ). Average Ganglion Cell Complex (GCC) thickness was thinner in Boxing athletes compared to Football athletes (Boxers:  $76.85 \pm 7.85 \mu\text{m}$  vs. Football:  $80.10 \pm 8.16 \mu\text{m}$ ), and compared to Controls (Boxers:  $76.85 \pm 7.85 \mu\text{m}$  vs. Controls:  $82.16 \pm 5.86 \mu\text{m}$ ) however these differences were not statistically significant. No significant differences in average macular thickness or volume were observed between groups.

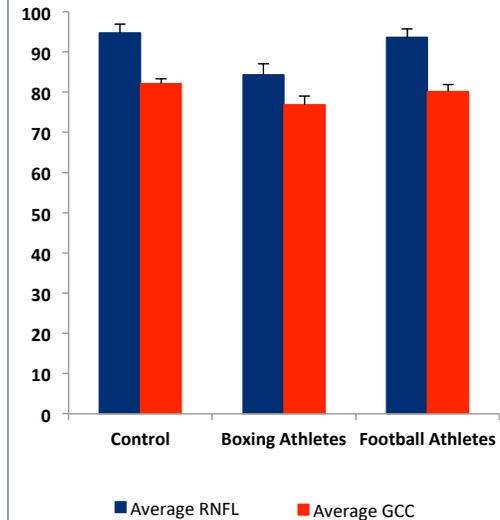
## CONCLUSIONS

Retinal structure thinning quantified using non-invasive OCT imaging is evident in high TBI risk contact sport athletes and may serve as an important biomarker of sport-related TBI exposure. Continued investigations will examine correlations with visual function and determine changes over time.

## ACKNOWLEDGEMENTS

Study supported by the Illinois Society for the Prevention of Blindness Research Grant.

Figure 1. Average Retinal Nerve Fiber Layer and Ganglion Cell Complex Layer Thickness ( $\mu\text{m}$ ) by Group



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# King-Devick Test-Retest Reliability in Normal Adults and Adolescents

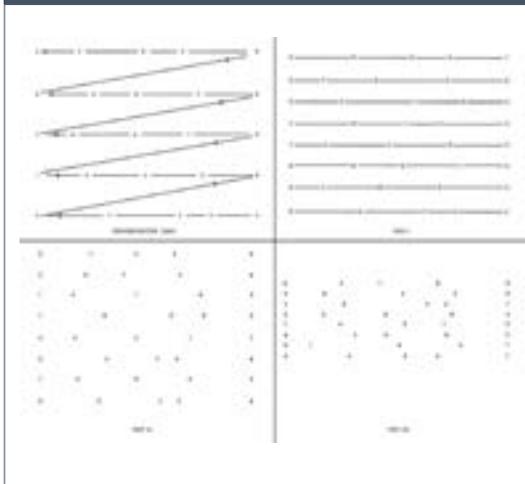
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## INTRODUCTION

The King-Devick(K-D) test (Figure 1), a 2-minute timed assessment of rapid number naming, has been studied as a rapid, quantitative screening tool for neurological dysfunction associated with concussion, hypoxia, Parkinson’s disease, multiple sclerosis and extreme sleep deprivation. The purpose of this study was to report the test-retest variability of the K-D test performance in normal adults and adolescents.

**FIGURE 1**  
King-Devick (K-D) Test Cards

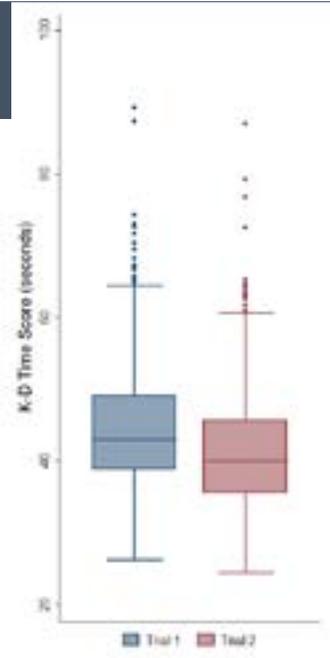


## METHODS

In this cross-sectional, multi-center study, subjects ≥15yrs old with near vision better than or equal to 20/30 completed two trials of the K-D test protocol. Exclusion criteria included concussion within 3-months, post-concussion syndrome, dyslexia or neuro-degenerative disorders. History of concussion, amblyopia, strabismus as well as demographic variables of education, race/ethnicity, gender and age were assessed by subject interview. Intraclass correlation coefficients (ICC) and descriptive statistics were calculated.

**FIGURE 2**  
King-Devick Median Scores for Trial 1 and 2 (n= 691).

Box plots show the median K-D scores for trial 1 and trial 2 for all study participants demonstrating improvement of scores (p<0.001, student’s t-test) likely consistent with learning effects. The lines in the box represent the medians, and boxes delineate the interquartile range (25th to 75th percentiles). Whiskers represent the range of observations minus outliers; the circles represent outliers.



## RESULTS

Subjects (n=691, mean age 39.8±17.7 years) were enrolled across 5 sites. High levels of test-retest reliability were observed between the two K-D trials (ICC=0.93 [95% confidence interval: 0.62, 0.97]). There was an average improvement from trial 1 to 2 of 3.53 seconds (±2.83).

## CONCLUSION

The K-D test shows excellent test-retest reliability in adults in the absence of neurological disease and/or neurological events such as concussion. The K-D test requires visual processing, saccades, language, attention, and has been proposed as a marker of integrated neurological function. Knowledge of test-retest reliability will have application to future studies of K-D test performance in neurologically diseased populations and potential future application to clinical settings.

## ACKNOWLEDGEMENT

We would like to thank the American Academy of Optometry Fellows Doing Research SIG for their guidance and support to our study team.

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# Shaken Baby Syndrome Sequelae in the Adult

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## BACKGROUND

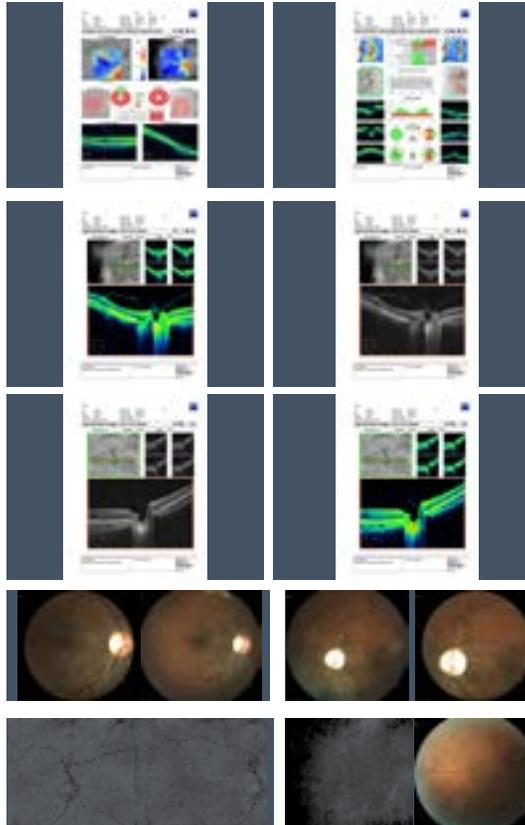
Although the outcome sequelae of Shaken Baby Syndrome (SBS) in young children have been extensively documented, not a single reference was found in the literature of the long term outcomes of this traumatic experience in the adult. Children who experience SBS demonstrate characteristic anatomical, neurological and ocular anomalies. These anomalies include but are not limited to subdural, subarachnoid and retinal hemorrhages.

## Clinical Findings



### Shaken Baby Syndrome in the Adult

Optic atrophy OS >> OD; OD:Mild NFL/severe GCC thinning.  
OS: Severe NFL/GCC thinning.



## CASE REPORT

SB is a 28 year old female with a history of SBS. According to her adoptive mother SB experienced retinal detachments at 2 months of age due to the SBS. She is able to see with her right eye but cannot see out of her left eye because of a retinal hole. SB was recently diagnosed with diabetes (DM). She has worn glasses since 18 months of age and has not noted any changes in her vision. SB takes medication for her DM, high blood pressure and cholesterol. Her last eye examination was several years ago. She has no allergies and denies smoking and drinking. Her aided distance visual acuities were 20/30 OD and 3.5/400 OS and 20/25 (Snellen) at near. Anomalous findings included confrontation visual field restrictions, constant left XT, poor oculomotor skills, ptosis and moderate to high myopia and astigmatism. Biomicroscopy was unremarkable. The dilated fundus noted numerous retinal changes and optic atrophy OS. Fundus photographs, OCT and a follow up by an IEL retinal specialist were completed.

## CONCLUSION

Infants and young children with SBS are characterized by subdural hemorrhages, retinal hemorrhages, damage to the spinal cord and neck, and fractures of the ribs and other bones. Other signs of SBS include lethargy, decreased muscle tone, irritability, and seizures. Poor oculomotor ability and other visual anomalies may be present as well. Since no papers have been published on the sequelae of SBS in the adult, we do not know if these signs carry over into adulthood. Our diagnosis included myopia, optic atrophy OS >> OD. Suspected TON because of the resultant neurological trauma. There was also evidence of left upper lid ptosis (long standing by history). Many of the expected findings associated with Shaken Baby Syndrome were present in this adult patient. Since she has also been diagnosed with diabetes, we will be following her routinely and monitoring and changes. This poster may be the first description of the long term outcomes for adults with SBS. It appears that those ocular anomalies frequently noted in the child with SBS are also present in the adult.

## ACKNOWLEDGEMENT

My thanks to Dr. Leonard Messner for his consultation on this case.

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# Student Performance examining patients with visual impairments for the first time

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## INTRODUCTION

The Illinois College of Optometry educates students for Low Vision Rehabilitation through twelve 2 hour lectures, eight 2 two hour laboratories and five 3.5 hour clinic observation sessions during their third professional year. They have one laboratory practical, 2 tests and one final examination to determine their grade. A second practical exam was added to the course this past year. The practical was designed to have students work one on one with a patient with visual impairment. The results of the student's performance on the practical demonstrate the value of working directly with patients to verify competency in preparing students for this specialty care.

## METHODS

Seven long time experienced patients of the Low Vision Rehabilitation clinic were selected to sit as patients for the practical. Their diagnoses included: aphakia, myopic degeneration, Stargardt (SMD), retinitis pigmentosa (RP), aniridia and albinism. The practical occurred at the end of the quarter, after all the lectures and laboratories had taken place. The students had 45 minutes for the practical. Students were given 10 minutes before and 10 minutes after working with the patient and 25 minutes with the patient. In the 10 minutes before working with the patient, the students were given a history on the patient including age, vocation, diagnosis and which eye to test. During the 25 minutes with the patient, the student was asked to: obtain 3 visual goals, perform retinoscopy and trial frame refraction on one eye, document best corrected VA at distance and near on that eye, predict/calculate magnification for distance and equivalent power for near and select appropriate device to demonstrate/train patient with the device.

## RESULTS

150 students completed the practical. Students did very well in a number of areas as seen in Figure 1. Documentation of low vision acuities at distance and near was done well. Accuracy of the distance acuities was less successful; this was dependent on student's ability to refract the patients. Performance on refraction was evaluated by their accuracy. There were 4 grading categories for each of the 3 refraction components, within 9% of actual refraction, 10-19%, 20-29% and greater than 30% off. Figure 3 shows the accuracy of the student refractions. The spherical component was the most accurate, followed by the axis then cylinder. This data also shows that some conditions were more challenging to work with. Students had the most difficulty refracting patients with retinitis pigmentosa and the least difficulty with myopic degeneration and aphakia. For these 2 conditions, the accuracy was the worst. When calculating magnification, choosing and training with appropriate devices, the students did very well (see Figure 1). In selecting near devices, the majority of student selected hand held magnifiers, followed by microscopes then stand magnifiers. The lower percentage of prescribing stand magnifiers may be due to less comfort with the slightly more complicated equivalent power calculation. Some vision impairments challenged students more. Students had the most difficulty with RP to prescribe a near device; only 38% did so correctly. The most difficulty for telescopes was with SMD, 74% prescribed correctly.

FIGURE 1: Practical Results-% Correct/Accurate

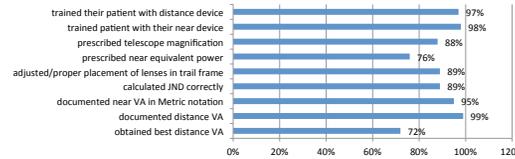


FIGURE 2: What near devices were prescribed

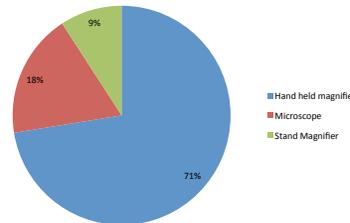
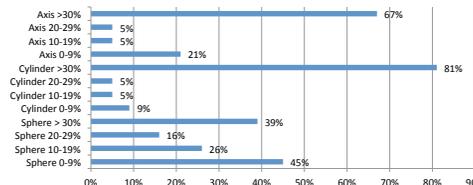


FIGURE 3: Trial frame refraction results % accurate



## DISCUSSION

Students did well on tasks more related to calculations while not as well on tasks which involved applying low vision knowledge to clinical situations. This project supports continued use of actual patients to assess student's low vision skills. During the laboratory series, students have labs dedicated to all the skills tested, including retinoscopy and trial frame refraction on each other while one student is wearing contact lenses with higher prescription and simulated/frosted visual impairments. Even with this preparation, trial frame refraction of a patient was the most challenging for students and could benefit from additional preparation. Ideally this preparation would include actual patients with visual impairment. Also, assessment would include students encountering patients with a variety of conditions. Due to the nature of this practical each student had the opportunity to examine only one patient and therefore only work with one type of vision loss. This would be even more important if students do not have the opportunity to rotate through a Low Vision Rehabilitation clinic during their fourth year of optometry school. Students do experience stress in testing situations, but overall, student comments were positive on the experience of working with a patient and helping them to improve their vision.

## CONCLUSION

Students did well most tasks on this practical. This project supports the use of patients with visual impairment in didactic programs to aid in assessing the topics that may need to be reinforced during Low Vision clinical rotation.

## ACKNOWLEDGMENT

This project was made possible by the 2014 Fredric Rosemore Low Vision Education Grant. We would like to thank the Fredric and Marion Rosemore Family Foundation for their generous support allowing Low Vision Educators to increase student's abilities and awareness in the field of Low Vision Rehabilitation.

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# The Case of the Missing iStent

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## BACKGROUND

Minimally invasive glaucoma surgeries (MIGS) have been gaining popularity due to a better safety profile compared with traditional glaucoma surgery. Although these surgical approaches have fewer complications, they do not achieve the dramatic intraocular pressure (IOP) reduction seen with the gold standard trabeculectomy. MIGS procedures are best for patients that require moderate IOP reduction.

The iStent is a trabecular micro-bypass stent which lowers IOP by direct cannulation of Schlemm's canal to enhance aqueous outflow. It is the smallest medical device to be implanted into the body. The iStent is made of surgical-grade nonferromagnetic titanium. The most common postoperative complications reported are early stent occlusion and malposition. The rate of stent malposition ranges from 2.6% to 18%.

## CASE REPORT

A 79 year old African American male presented for a one day post op visit after cataract extraction and iStent placement OD

### Ocular History:

Primary Open Angle Glaucoma OU  
Cataracts OU

### Ocular Medications:

Combigan BID OU  
Travatan Z QHS OU

### Surgical Notes:

The iStent was unsuccessfully inserted in the trabecular meshwork and lost in the uveal-ciliary cleft upon sudden eye movement upward.

### Exam Findings:

BCVA: 20/150 OD, 20/50 OS  
Biomicroscopy: corneal edema, a well-placed posterior chamber intraocular lens (PCIOL), and inflammatory cells, blood (no hyphema), and pigment in the anterior chamber OS  
IOP: 12mmHg  
Gonioscopy: bleeding in the angle inferiorly

### Follow Up Visits:

BCVA improved to 20/100 OD  
IOP: 16mmHg  
Biomicroscopy: inflammatory cells in the anterior chamber  
Gonioscopy: previous blood had resolved, iStent was not observed

### MRI and iStent

Glaukos, the company that produces the iStent, states the iStent is MR-Conditional. This means it poses no known hazard in a specific MRI environment with specified conditions of use. The patient can be scanned if: static magnetic field of 3-Tesla or less and maximum spatial magnetic field gradient of 720-Gauss/cm or less. Due to the unknown location of the iStent, and because it may be free floating and not implanted, this poses a possible risk to the patient.

## CONCLUSIONS

There are no reports in the literature on the loss of an iStent during surgical placement. Due to unknown location of the iStent the patient has been educated that a MRI may be contraindicated because of the potential ocular complications. Fortunately for this patient no other ocular complications have been observed.

IMAGE 1: Implanted iStent



IMAGE 2: Ultrasound Biomicroscopy no iStent seen



IMAGE 3: B scan no iStent seen

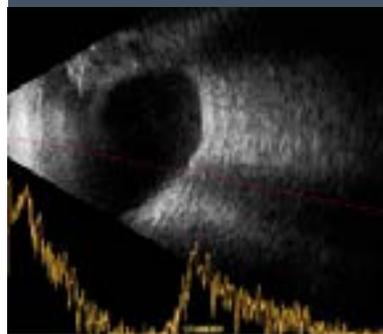


IMAGE 4: Anterior Segment OCT no iStent seen



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# Considerations in the Management of a 15 year old Anterior Chamber Foreign Body

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## ABSTRACT

Penetrating intraocular foreign bodies can pose a major threat to vision and require prompt evaluation and management. High speed metallic foreign bodies, after penetrating through the cornea, may potentially lodge in any ocular structure and only about 15% are retained in the anterior chamber. These objects must nearly always be surgically removed in order to prevent infection, inflammation, or siderosis.

After performing gonioscopy on a 58 year old male with narrow angles, a lesion was noted within the nasal portion of the angle and anterior to the iris. He reported trauma more than 15 years prior when a metal shaving hit his eye. The lesion was presumed to be the encapsulated shaving. CT confirmed likelihood of metallic substance, while ERG found no evidence of siderosis.

Peripheral iridotomies were performed on both eyes, but at this time the foreign body has not been removed. It is felt that surgery would leave, at the least, a wide cyclodialysis cleft, which would increase his risk for future complications. Given the longstanding presence of this foreign body and absence of sequelae in this case, treatment, unlike the majority, is not warranted.

While the majority of acute intraocular foreign bodies necessitate removal, special consideration should be given to longstanding foreign bodies. When inert, or otherwise tolerated, surgical correction may pose a greater risk and potential complications need to be carefully weighed against any perceived benefit.

## KEYWORDS

intraocular foreign body, penetrating trauma, metallic foreign body

## INTRODUCTION

The majority of patients with confirmed intraocular foreign bodies require surgical removal of the foreign material. Complications secondary to retained material are frequently reported with serious and often irreversible consequences including cataracts, endophthalmitis, retinal detachment, and siderosis.<sup>1</sup> Intraocular foreign bodies may be organic or inorganic, although the majority are metallic and result from striking metal on metal. Working-aged males account for up to 94% of patients presenting with intraocular foreign bodies.<sup>2</sup>

## CASE REPORT

Initial visit: A 58 year old male presented for diabetic eye exam and complaining of distance blur OU. He also reported a history of metallic foreign body OS 15 years prior, when a piece of lug nut broke off and hit his eye. He denied ever receiving treatment. Best corrected vision with a hyperopic refractive error was 20/20 OD, OS. Entrance tests were normal. Slit lamp was remarkable for narrow angles open to anterior TM OD and TM OS with a superior area of synechiae. No cell or flare was present. Intraocular pressure measured 20mmHg OU. Dilation was performed, noting mild diabetic retinopathy OU. Post-dilated IOP elevated to 31mmHg OD and 28mmHg OS. The patient was referred for an LPI consult.

Narrow angle consult: Gonioscopy revealed anterior TM OU, and the previously noted angle synechiae was believed to

be an encapsulated cyst (Figures 1-6). It was theorized that the metallic foreign body from the reported trauma 15 years prior had lodged in the angle and remained inert since that time. The patient was scheduled for laser peripheral iridotomy OU. Ultrasound biomicroscopy and anterior segment OCT were performed, offering further evidence the lesion was a retained metallic foreign body. An orbital CT was ordered and later confirmed that the foreign body was indeed metallic. To rule out retinal siderosis, an ERG was ordered, which showed no evidence of retinal toxicity (Figure 7). LPIs were performed and remain patent OU and IOP is stable at 15mmHg OU. After careful consideration of the risks and benefits of surgical removal, the decision was made to monitor the angle foreign body for now, as it is not causing any secondary complications. The patient will be monitored at three month intervals for now, and knows to return if complications arise.

FIGURE 1 & 2  
Slit lamp photographs showing lesion extending into anterior chamber.



FIGURE 3&4  
Gonioscopic views of encapsulation within anterior chamber angle.

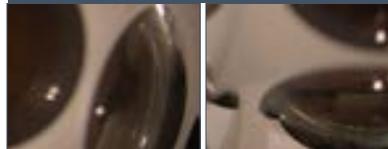


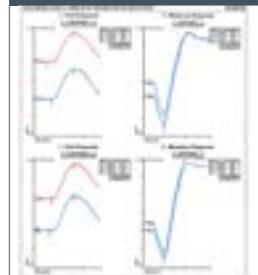
FIGURE 5  
Ultrasound biomicroscopy detailing lesion in anterior chamber angle. High reflectance supports theory of metallic foreign body.



FIGURE 6  
Visante OCT detailing lesion in anterior chamber angle. Fluid-filled cyst or shadowing from foreign body.



FIGURE 7  
ERG responses. No evidence of siderosis OS due to longstanding ocular metallic foreign body.



## DISCUSSION

Most intraocular foreign bodies are detected at initial presentation when an open globe injury is suspected, either via patient history or clinical findings. Retained foreign bodies pose a risk of infection, inflammation, mechanical complications and, if metallic such as iron or copper, retinal degradation.<sup>3</sup> Surgical removal is preferred. Reports do exist, however, detailing cases where longstanding foreign bodies in the anterior segment,<sup>4,5,6</sup> lens,<sup>7</sup> and retina<sup>8</sup> were monitored, as the surgical extraction was deemed to pose a greater risk of complications.

Visualization of an intraocular foreign body and suggestions as to the composition may be enhanced through x-ray, ultrasound, OCT, CT, and MRI. Utilization of each is dependent upon location and material.<sup>9</sup>

Siderosis is an additional ocular risk if the foreign body is an iron-containing metal. Iron ions dissolve into the ocular tissues and can extinguish rod and cone responses. Visual dysfunction may be irreversible, but has been shown to improve somewhat after foreign body extraction.<sup>10</sup>

## CONCLUSION

The presence of a longstanding metallic foreign body as an incidental finding during an eye exam has rarely been reported in the literature. When making the decision to monitor the patient rather than surgically remove the material, consideration must be given to the current ocular health state as well as the risk and benefit of such surgery. In this case, encapsulation by surrounding ocular tissue is presumed to have rendered the metal fragment inert, and the patient has remained asymptomatic for 15 years. Future testing will include periodic gonioscopy, with ERG repeated as necessary. Clinicians should be aware of such a scenario given a patient history of trauma or open globe.

The authors would like to thank Lili Farrok-Siar MD, Mary Flynn Roberts OD, and Rebecca Tudor CRA for their involvement in this case.

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# Ocular Dysmetria in a Patient with Charcot-Marie-Tooth Disease

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## BACKGROUND

A 74 year-old Caucasian male presents with ocular dysmetria and a known medical history of Charcot-Marie-Tooth disease (CMT), an inherited neuropathy. CMT is caused by a gene mutation that results in impaired nerve conduction, either due to abnormal myelin or axons. There are a limited numbers of case reports that highlight the ocular manifestations of CMT and few, if any, report abnormalities of ocular motility. The absence of other systemic causes, as well as a normal MRI in this patient, suggests that ocular dysmetria is a possible manifestation of CMT. Table 1 summarizes the known systemic and ocular manifestations of CMT

SYSTEMIC MANIFESTATIONS	OCULAR MANIFESTATIONS
<ul style="list-style-type: none"> <li>Weak handgrip and difficulty with fine motor skills</li> <li>Foot deformities like foot drop</li> <li>Abnormal gait</li> <li>Difficulty with balance and sensory ataxia from proprioceptive sensory loss and vestibular impairment</li> <li>Diagnostic testing               <ul style="list-style-type: none"> <li>History and physical examination</li> <li>Electromyogram</li> <li>Nerve conduction studies</li> <li>Genetic testing</li> </ul> </li> </ul>	<ul style="list-style-type: none"> <li>Pupil abnormalities have been reported in 17% of patients with CMT, especially CMT2               <ul style="list-style-type: none"> <li>Tonic pupils</li> <li>Argyll-Robertson pupil</li> <li>Horner's syndrome</li> <li>Anisocoria</li> </ul> </li> <li>Optic atrophy</li> <li>Macular degeneration</li> <li>Glaucoma</li> <li>Cataracts</li> <li>Case report of bilateral vitritis</li> <li>Subclinical optic neuropathy based on reduced contrast sensitivity and visual evoked potential</li> </ul>

## CASE DETAILS

OCULAR HISTORY	MEDICAL HISTORY	FAMILY HISTORY
<ul style="list-style-type: none"> <li>Mild cataracts OU</li> <li>Dry eye syndrome OU</li> <li>Refractive error OU</li> </ul>	<ul style="list-style-type: none"> <li>Charcot-Marie-Tooth disease</li> <li>Asthma</li> <li>Hypercholesterolemia</li> <li>Herpes zoster</li> <li>Chronic lower back pain</li> <li>Dermatitis</li> <li>Obstructive sleep apnea</li> </ul>	<ul style="list-style-type: none"> <li>(+) CMT: Father and grandfather</li> </ul>

EXAM FINDINGS		
EXAM	OD	OS
VA's	20/20 <sup>2</sup>	20/20 <sup>1</sup>
Pupils	NL, (-)JNLD	NL, (-)JNLD
OM's	Full OD	Restricted lateral and inferior-temporal gaze (Fig 1 A)
	Ptosis OD (Fig 1B)	
Lids/lashes	MRD 1: 3mm	MRD 1: 5mm
	MRD 2: 8mm	MRD 2: 4mm
Cornea	NL	NL
IOP	13	13
Lens	2+ NS, trace ACS	2+ NS, trace ACS
Optic Nerve	0.25/0.25, (-) pallor	0.25/0.25, (-) pallor
Macula	NL	NL
Periphery	NL	NL



Figure 1



Figure 2

### OTHER FINDINGS

- Mixed hypometric and hypermetric saccades with intermittent disconjugate movement
- Borderline reduced contrast sensitivity OD, mildly reduced contrast sensitivity OS (Figure 2)
- Abnormal gait
- EMG (01/2006): mild distal, primarily axonal sensorimotor polyneuropathy, consistent with positive family history of CMT
- MRI (04/2013): no intracranial mass or acute infarcts seen, no evidence of cerebellar abnormality

## DISCUSSION

### ABOUT CMT

- Inherited genetic neuropathy that affects peripheral motor and/or sensory nerves
  - Prevalence is estimated to be 1 in 2500
  - Symptoms present in the first two decades of life
- Cause: gene mutations that affect myelin sheath or axons leading to reduced nerve conduction.
  - The most common mutation is a duplication in peripheral myelin protein 22 (PMP22) on chromosome 17 - expressed mainly in Schwann cells and is involved in myelin production
  - Mutation can in the gene MFN2 causes an inability for mitochondria to travel along axons, resulting in clusters of mitochondria that block synaptic function

CMT 1	Demyelinating condition	Conduction velocity < 38 m/s Autosomal dominant
CMT 2	Axonal condition	Conduction velocity > 38 m/s Autosomal dominant
CMT 3	Early onset	
CMT 4	Usually demyelinating	Autosomal recessive
CMT X	Usually demyelinating	X-linked

### DIFFERENTIAL DIAGNOSIS

- Ocular dysmetria is typically associated with cerebellar dysfunction, but the absence of other systemic causes, as well as normal MRI in this patient, suggests a possible manifestation of CMT.
- Other differentials include:
  - Traumatic brain injury
  - Multiple sclerosis
  - Amyotrophic lateral sclerosis
  - Intracranial mass

## TREATMENT & MANAGEMENT

- Ocular findings can generally be observed and other etiologies ruled out.
  - For this patient, a neurology follow-up with gait evaluation was recommended by the primary care provider
- One study has suggested that "rehearsal by eye movement" can improve visuomotor function and reduce saccadic dysmetria in patients with cerebellar conditions.
  - Rehearsals involved making saccadic eye movements to a series of foot targets before walking. Given the absence of cerebellar lesions in this patient, the same rehearsals may not be beneficial.
- Multi-disciplinary management of patients with CMT should involve:
  - Neurology
  - Orthopedic surgeons
  - Occupation or physical therapy
  - Consideration for psychiatry to address a patient's quality of life
- Studies have shown that diabetes mellitus can exacerbate peripheral neuropathy and CMT patients should undergo routine diabetes screening
- Genetic counseling may be considered to evaluate the risk for future generations

## CONCLUSION

- Current literature about the ocular manifestations of CMT is limited to a small sample of case reports, none of which have identified ocular dysmetria as a likely association. This report highlights an additional finding in which a demyelinating condition may manifest itself ocularly.
- Patients identified as having CMT should be managed by a multi-disciplinary approach and appropriate referrals should be made, including routine screenings for diabetes.
- Should a patient present with CMT, optometrists ought to be aware of possible ocular manifestations and perform appropriate testing, especially pupil abnormalities and subclinical neuropathy based on VEP and contrast sensitivity.

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### ACKNOWLEDGEMENTS

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# Pigmentary Retinopathy in an Adult Hispanic Female with Bardet–Biedl Syndrome

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## BACKGROUND

Bardet-Biedl Syndrome is an autosomal recessive disorder with a diverse clinical presentation. The primary features of the syndrome include pigmentary retinopathy, truncal obesity, polydactyly, intellectual impairment, hypogonadism, and renal abnormalities. This case represents an illustration of pigmentary retinopathy in an adult Hispanic female with Bardet-Biedl Syndrome.

## PERTINENT FINDINGS

23-year-old Hispanic female presents with gradual worsening of vision, first noted at age nine, and longstanding nyctalopia and photophobia.

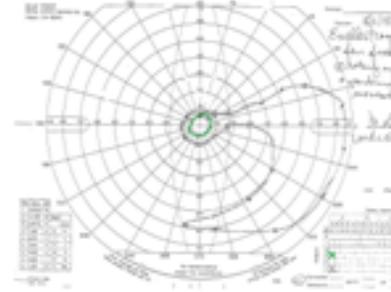
- **Ocular history:**
  - Diagnosed with pigmentary retinopathy at age 12
- **Medical history:**
  - Genetic testing revealed homozygous variation of IVS2-IG>A in BBS5; diagnosis of Bardet-Biedl Syndrome
  - Secondary obesity, polydactyly; history of urinary problems and kidney infections
  - (-)Diabetes mellitus, reproductive system abnormalities, cognitive disability
  - No reported consanguinity
- **Clinical findings:**
  - BCVA:
    - OD: 4/250, Near: 20/800 at 10 cm
    - OS: 4/300, Near: 20/400 at 5 cm
  - Gradual worsening of visual fields from ring scotoma at initial visit to central and temporal islands on most recent visual field done 8/2008
  - (+)Horizontal and pendular nystagmus OU
  - Fundus examination:
    - Lens: 1-2+ PSC OU
    - Macula: mild atrophy with relative foveal sparing OU
    - Optic nerve: mild, diffuse waxy pallor OU
    - Vessels: mild attenuation OU
    - Periphery: modest granular and bone spicule pigmentation and diffuse RPE and choroidal atrophy with hypopigmentation in midperiphery OU

## DISCUSSION

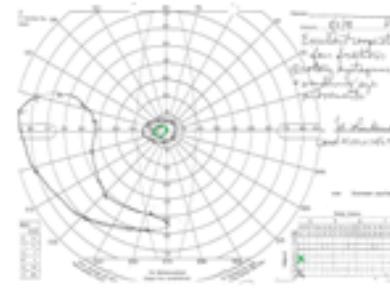
- Differential diagnosis: Alström syndrome, Lawrence-Moon syndrome, retinitis pigmentosa
- Diagnosis based on presence of four of the five primary features or three primary plus two secondary features
- Pigmentary retinopathy leads to progressive decrease in visual acuity in first decade of life; nearly 50% are legally blind by age 20
- Renal failure is most life-threatening complication of BBS
- Treatment and management:
  - Yearly ocular examination with refraction, fundus examination, and kinetic visual field

- Low vision evaluation:
  - Implement orientation and mobility training
  - Electronic magnification necessary at an early age
  - Patient currently using ZoomText, CCTV, Braille, Talking Book program, smartphone with accessibility features, white cane
  - Consider statement of legal blindness, communication with educational programs, and referrals to psychologist, vocational services, and occupational therapist, as needed
- Routine general health examination:
  - Manage the systemic effects of the condition, including diabetes, hypertension, and renal complications

**FIGURE 1:**  
Kinetic visual field of right eye done 8/2008



**FIGURE 2:**  
Kinetic visual field of left eye done 8/2008



**FIGURE 3:**  
Fundus photo of right eye depicting diffuse hypopigmentation and vessel attenuation; date unknown



**FIGURE 4:**  
Fundus photo of left eye depicting diffuse hypopigmentation and vessel attenuation; date unknown



## PATHOPHYSIOLOGY

- Mutation of one of twelve specific genes; p.M390R is the most common mutation in BBS1 that is found in 18-32% of individuals with BBS
- All of the known BBS proteins are parts of the centrosome and/or basal body that play a role in ciliary transport
- Mutation leads to degenerative loss of retinal photoreceptors and later, the inner retinal layers and RPE

## CONCLUSION

- Bardet-Biedl is an autosomal recessive disorder that has a significant effect on many of the body's systems
- Pigmentary retinopathy is the most commonly seen manifestation of the condition; this has a marked effect on both peripheral and central vision.
- Intervention should begin at birth
- Co-management is important; these patients should have routine care by ocular health and primary care physicians. Additionally, these patients should be referred to low vision providers, occupational therapists, psychologists, and other specialists to help them adapt to vision loss and other physical disabilities.

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# Performance and Reliability of the Sanet Vision Integrator (SVI) in Athletes versus Non-Athletes

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## INTRODUCTION

There is currently great interest in finding sensitive, reliable tests to detect and monitor concussive injuries. Evidence strongly suggests a direct correlation between concussive injuries and eye-hand coordination impairment. The SVI offers five programs to evaluate the visual guidance of motor performance, one being "eye-hand." The "eye-hand" program offers a series of subtests designed to assess the speed and accuracy of motor responses to visual stimuli. If any of these tests are reliable and repeatable then the SVI has the potential to assess both the presence of concussion and the time to recovery. As these variables have not been previously investigated, the first goal of this study was to measure the performance and reliability of the SVI. Since eye-hand coordination may differ between athletes and non-athletes we also measured the reliability of SVI subtests between Division I athletes and non-athletes (controls). Our second goal was to determine if the performance on any of the SVI subtests differed between the athletes and the non-athletes.

## METHODS

Twenty-two visually normal subjects most of whom were students at the Illinois College of Optometry (ICO) and nineteen members of the DePaul women's softball team were tested on three subtests of the SVI eye-hand program; Proactive, Reactive and Hand Speed. The subjects were instructed to quickly and accurately hit the stimulus once it was presented using one finger of their dominant hand. Subjects' speed and accuracy were recorded for each of the three subtests. Pilot data indicated the number of trials needed for each test for the learning curve to plateau; thus four trials were run for the Proactive program, one trial was run for the Reactive program, and three trials were run for the Hand Speed program. Only the data from the last trial was used. Performance was measured on two visits at least two weeks apart.

## RESULTS

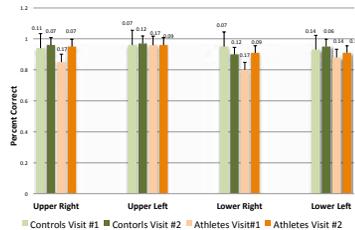
The reliability of each subtest was analyzed using the intraclass correlation coefficient (ICC). Speed and accuracy measurements were compared between groups and visits using a mixed design repeated-measures Analyses of Variance (ANOVA; IBM SPSS, V 21). The ICC values for various measures of each subtest are reported in Table 1. ICC values are greater for the non-athletes than the athletes for all subtests. The analyses reported below are only for those subtest variables with an ICC score greater than 0.5.

TABLE 1	ICC	Controls	Athletes
PROACTIVE:	Upper Right Visit 1 vs. Visit 2	0.42 (p=0.03)	0.35 (p=0.027)
	Upper Left Visit 1 vs. Visit 2	0.67 (p<0.000)	0.12 (p=0.26)
	Lower Right Visit 1 vs. Visit 2	0.24 (p=0.14)	0.179 (p=0.208)
	Lower Left Visit 1 vs. Visit 2	0.29 (p=0.10)	0.52 (p=0.007)
REACTIVE:	Reaction Time	0.8 (p=0.000)	0.41 (p=0.149)
	Percent Correct	0.57 (p=0.001)	0.57 (p=0.006)
Hand Speed: Overall Data Only	Reaction Time Visit 1 vs. Visit 2	0.64 (p=0.001)	0.28 (p=0.06)
	Hand Speed Visit 1 vs. Visit 2	0.83 (p<0.000)	0.4 (p=0.02)
	Percent Correct Visit 1 vs. Visit 2	0.24 (p=0.173)	0.34 (p=0.062)

## PROACTIVE TEST

Both athletes and non-athletes were significantly more accurate when tested in the upper left quadrant (F=6.09, p=0.001; see Figure 1). No practice effect was observed over the two visits and the mean results were the same for the two groups (F=1.63, p=0.21).

FIGURE 1: Percent Correct by Quadrant and Visit



## REACTIVE TEST

Figure 2 plots the mean reactive time for the two groups and the two visits. There is no significant difference between group means (F=1.01, p=0.3) nor was there a practice effect (F=0.39, p=0.54).

Figure 3 plots the percent correct for the two groups over the two visits. The non-athletes are significantly more accurate (F=4.61, p=0.04). Both groups show a significant practice effect (F=4.95, p=0.03).

FIGURE 2: Reaction Latency

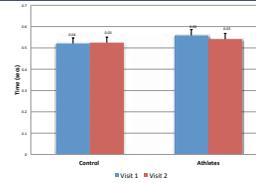
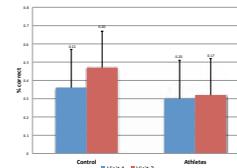


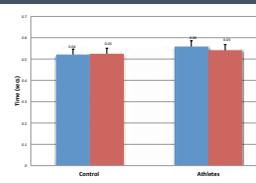
FIGURE 3: Percent Correct



## HAND SPEED TEST

Figure 4 plots the mean reaction latency for the two groups and the two visits. The reaction time latency was significantly shorter for the non-athletes (F=5.9, p=0.04) while only the athletes showed a significant decrease in reactive latency on Visit 2. The two groups did not differ in hand-speed (F=1.42, p=0.24), although the athletes showed a significant decrease on Visit 2.

FIGURE 4: Reaction Latency



## CONCLUSIONS

- The reliability of all SVI subtests was greater for the non-athletes than for the athletes. Possibly the non-athletes may have focused more carefully on the tasks given they were aware of the importance of research in general. Another possible explanation is that the athletes were tested in a common space where distractions sometimes occurred. The results obtained from ICO subjects (non-athletes) should be compared with those obtained from a general, non-athletic group to determine if they are representative of non-athletes.
- The athletes were significantly slower or significantly less accurate than the control group on each subtest (Proactive F=7.1 p=0.01; Reactive F=4.6 p=0.04; Hand Speed F=5.9 p=0.02).
- Of the three subtests that showed good or better reliability, only one was both reliable and able to differentiate between athletes and non-athletes. That was the Reaction latency on the Hand Speed test.

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# Broadened Foveal Base Detected by Spectral Domain Optical Coherence Tomography – A Case Series in Patients on Hydroxychloroquine Therapy

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## BACKGROUND

Hydroxychloroquine (HCQ) is an antimalarial drug that is often used to successfully treat a wide variety of autoimmune disorders including systemic lupus erythematosus, various connective tissue disorders and other rheumatological diseases.<sup>1</sup> Though HCQ is generally well-tolerated, irreversible retinal toxicity is a serious potential side effect.<sup>1</sup> The “Revised Recommendations on Screening for Chloroquine and Hydroxychloroquine Retinopathy” published in 2011 promote earlier recognition of retinal toxicity in order to limit the level of vision loss.<sup>2</sup>

## CASE REPORTS

This case series presents three female patients, ranging in age from 24 to 53 years old, who currently receive HCQ therapy. Two patients have systemic lupus erythematosus and the third has mixed connective tissue disease. All three patients were on a maintenance dose of 200 mg HCQ daily with medication duration ranging from 1.5 years to over 20 years. Baseline examination included a comprehensive dilated fundus exam, 10-2 Humphrey visual field and Spectral Domain Optical Coherence Tomography (SD-OCT). Best corrected visual acuities ranged from 20/20 to 20/25 OD, OS. Dilated fundus examination revealed the absence of bull’s eye maculopathy OD, OS and visual field testing was negative for any evidence of paracentral scotoma OD, OS in all patients. The SD-OCTs for all three patients demonstrate alteration of the normal foveal contour OD, OS. A broad and flat foveal base is visualized with an intact photoreceptor integrity line and no evidence of perifoveal thinning of the outer retinal layers. (See Figures 1 thru 3.)

## DISCUSSION

HCQ retinopathy is characterized by an insidious onset that makes early detection and diagnosis very difficult.<sup>1</sup> For that reason, there has been an effort to determine if the earliest signs of HCQ toxicity could be detected with SD-OCT prior to symptomatic vision loss.<sup>3</sup> Inner retinal thinning observed by SD-OCT has been reported in the absence of clinically detectable HCQ retinopathy.<sup>4</sup> A study by Pasadhika and Fishman determined that once clinically visible RPE changes or the classic bull’s eye retinopathy is observed, thinning of the inner retina, including ganglion cells and nerve fiber layer, is already present.<sup>2</sup> Cessation of HCQ does not generally result in improvement of vision and progression of vision loss has been shown after the discontinuation of

Figure 1a) Patient 1 OD

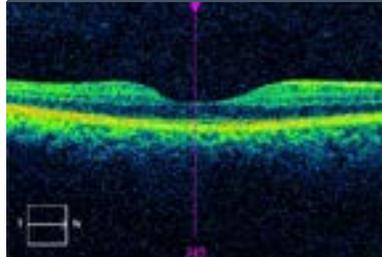


Figure 1b) Patient 1 OS

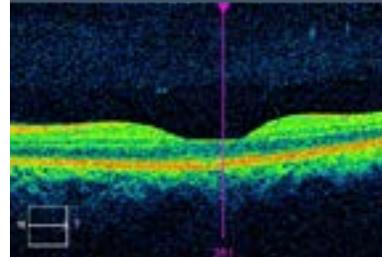


Figure 2a) Patient 2 OD

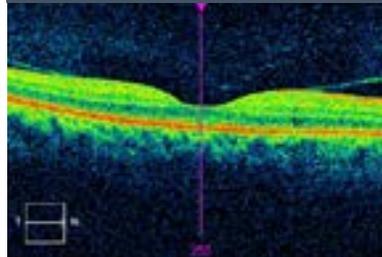


Figure 2b) Patient 2 OS

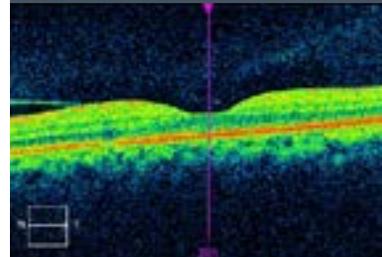


Figure 3a) Patient 3 OD

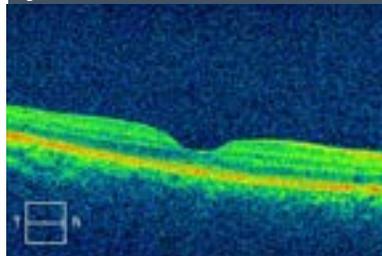
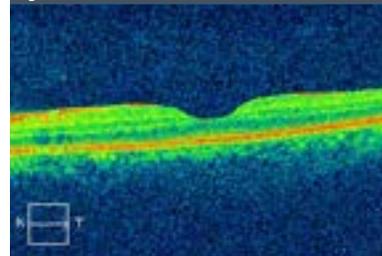


Figure 3b) Patient 3 OS



HCQ.<sup>5</sup> Though the exact mechanism of HCQ retinopathy is unknown, detection of retinal toxicity at the earliest stage is important to minimize any functional vision loss.<sup>5</sup> A recent study demonstrated flattening of the foveal contour with loss of normal central foveal depression on SD-OCT for a group of patients on chloroquine or hydroxychloroquine therapy.<sup>7</sup> The authors of this same study propose this perifoveal retinal thinning may be an early predictor of antimalarial toxicity.<sup>7</sup>

## CONCLUSIONS

Thinning of the inner retinal layer on SD-OCT in the absence of fundus changes has been documented for patients on HCQ therapy. SD-OCT has been proposed as a viable quantitative measurement for early diagnosis of retinal changes. Appearance of a broadened foveal base may represent a variation of perifoveal inner retinal thinning that could indicate early hydroxychloroquine retinopathy.

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# Choroidal Melanoma in a Patient with Ocular Melanocytosis

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## BACKGROUND

Ocular melanocytosis is characterized by unilateral hyperpigmentation of the uveal tract and sclera, presenting as heterochromia, melanosis oculi, and/or darker fundus pigmentation of the involved eye. Patients with these findings are at a greater risk for uveal melanoma, including choroidal melanoma. This case describes the diagnosis and treatment of choroidal melanoma in an adult male with ocular melanocytosis.

## PERTINENT FINDINGS

54 y/o Caucasian male presents for comprehensive eye exam with no visual complaints.

**OCULAR HISTORY (+)** longstanding heterochromia

**MEDICAL HISTORY (+)** hypertension, controlled

**VA (cd)** 20/15 OD, OS

**LEE** 2 years prior

### SLIT LAMP

- Heterochromia – OD blue, OS green/brown
- Diffuse 1-2+ melanosis oculi OS only

### POSTERIOR POLE FINDINGS

- New, elevated amelanotic lesion along inferior arcades (see Figures 1a & 1b)
- Humphrey 30-2 SITA Standard (see Figure 2)
  - o Good reliability OU
  - o OD: full field
  - o OS: superior defect
- Retinal OCT OS (see Figure 3)
  - o Elevated lesion with separation between lesion and overlying retina
- B scan & A scan OS
  - o Focal lesion inferior to disc
  - o 11.05 x 8.88 mm in size
  - o 2.95 mm deep
  - o Low internal reflectivity

**DIAGNOSIS** Choroidal melanoma with congenital ocular melanocytosis OS

**DIFFERENTIAL DIAGNOSES** Choroidal nevus, choroidal osteoma, metastatic carcinoma

## DISCUSSION

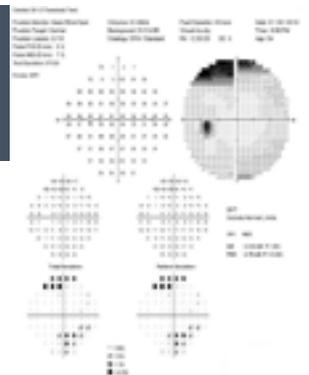
### OCULAR MELANOCYTOSIS (OM)

- Unilateral hyperpigmentation of sclera and uveal tract
  - o Heterochromia, melanosis oculi, darker fundus pigmentation
- 0.04% of Caucasians
- 1 in 400 Caucasian patients with OM develop uveal melanoma in their lifetimes
  - o Increased risk vs. patients without melanocytosis
  - o 3% of all uveal melanoma patients display ocular or oculodermal melanocytosis (similar to OM, but with hyperpigmentation of periocular skin)

FIGURE 1A: Fundus image OS from 2013



FIGURE 2: 30-2 SITA Standard visual field OS



### CHOROIDAL MELANOMA

- 5 to 6 cases per million people in the US each year
- Most common differential is choroidal nevus
  - o Suspect melanoma if lesion is thick (>2 mm), patient is symptomatic, and lesion includes subretinal fluid, orange pigment, or is within 3 mm of disc
- Most common sites of metastasis are liver (90%), lung (24%), and bone (16%)

### PROGNOSIS

- Risk for metastasis 1.6 times greater in patients with OM
- Melanoma-related death and risk of recurrence are unrelated to presence of OM
  - o Most strongly correlated with tumor size

FIGURE 1B: Fundus image OS from 2015

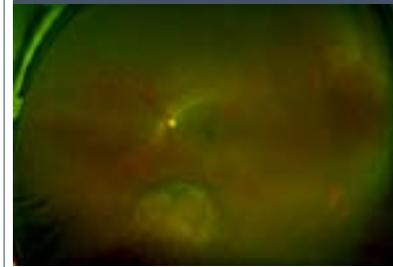
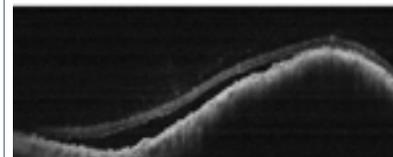


FIGURE 3: OCT raster scan through tumor



## TREATMENT

### TREATMENT OPTIONS

- Enucleation
  - o Rarely performed
  - o Only used on blind/painful eyes, or if tumor is too big or too near the disc for other options
- Plaque brachytherapy
  - o Most common
  - o Radioactive plaque sutured to external sclera in region of tumor
  - o Removed after 3-7 days
- Charged particle / proton beam therapy
  - o Good alternative to plaque brachytherapy
  - o Charged particle beams directed at lesion over 7-9 days

### MANAGEMENT

- Proton beam therapy
  - o Plaque brachytherapy not performed because of proximity of tumor to inferior oblique muscle and concerns about post-treatment diplopia
  - o 50 Cobalt Gray Equivalents (CGE's) in fractions of 10 over a period of 8 days

### OUTCOME

- Chest CT and liver function tests normal, indicating no metastasis
- At 3 month follow-up, patient showed no evidence of disease and no visual changes

## CONCLUSION

- Ocular and oculodermal melanocytosis are relatively rare findings that can significantly increase a patient's risk of developing uveal melanoma and the likelihood of metastasis
- Patients with ocular melanocytosis should have thorough examinations of the posterior and anterior segments (with imaging) on a twice-yearly basis

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# Herpes Zoster Ophthalmicus in African American Octogenarians: A Comparative Study

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## INTRODUCTION

We compare two patients with Herpes Zoster Ophthalmicus. Both patients present with a diagnosis and prescription for oral antivirals by outside physicians. One heals quickly while the other experiences complications prior to resolution.

## CASE 1

An 84 yo African American female presented with shingles on the right side of face x6 days, 3/10 pain across eye and down face, blurry vision OD. Her PCP had evaluated her one day prior and diagnosed HZV. The patient was prescribed acyclovir 400mg QID PO and gabapentin 100mg 2 capsules TID PO.

POHx: Cataracts OU.

PMHx: Hypertension, hypercholesterolemia.

Systemic Medications: Amlodipine, aspirin, pravastatin, acyclovir, gabapentin.

## PERTINENT FINDINGS

Initial Presentation	OD	OS
VA (ac)	20/50-1 PH:20/30-2	20/50-2 PH:20/30-2
Adnexa	Vesicles along face, lid edema (Figure 1)	WNL
Cornea	Clear	Clear
Anterior Chamber	Deep and quiet	Deep and quiet
Plan	Acyclovir dose increased to 800 mg 5x/day for 7 days	

Follow Up	Visit	IOP (mmHg)	Adnexa	Treatment
Day 1	13/13		Mild lid edema	Increase oral antiviral
Day 2	9/7		Significant lid edema; unable to open eyelid	Continue oral antiviral. Begin cool compresses
Day 5	13/9		Improvement in swelling	Monitor
Day 14	13/10		Improvement in comfort; crusting of vesicles	Monitor

FIGURE 1



## CASE 2

An 89 yo African American female presented with shingles on the right side of face x2 days. She denied eye pain, but reported tingling and pain on top of head. She also reported a fever, a sore throat, and a headache beginning earlier that day. The patient was evaluated by PCP earlier the same day, diagnosed with HZV, and prescribed Valacyclovir 1000mg BID x 7 days.

POHx: POAG, retinal hemorrhage, lattice degeneration, s/p cataract extraction, all OU.

Ocular Medications: Combigan, Lumigan.

PMHx: DMII, gout, hypertension, s/p hysterectomy.

Systemic Medications: Allopurinol, amlodipine, Coumadin, furosemide, valacyclovir, Vesicare.

## PERTINENT FINDINGS

Initial Presentation	OD	OS
Visual Acuity (ac)	20/50-1 PH: 20/40-1	20/40-2 PH: NI
Adnexa	Vesicles along forehead, eyelids, cheek, nose, and upper lip (Figure 2)	WNL
Cornea	Trace diffuse guttatae	2+ diffuse guttatae
Anterior Chamber	Deep and quiet	Deep and quiet
Plan	Valacyclovir dose increased to 1000 mg TID for 7 days	

Follow up Week 1	OD	OS
Visual Acuity (ac)	20/50-1 PH: 20/40-1	20/40-2 PH: NI
Adnexa	Vesicles scabbing along right forehead, eyelids, cheek, nose, and upper lip	WNL
Cornea	Trace diffuse guttatae	2+ diffuse guttatae
Anterior Chamber	1+ cells, trace flare	Deep and quiet

Follow up	Visit	IOP (mmHg)	Anterior Chamber	Treatment
Day 1	20/22		(-)cells, (-)flare	Increase oral antiviral
Day 9	14/13		1+ cell, trace flare	Prednisolone acetate QID OD
Day 16	15/17		1+ cell, trace flare	Prednisolone acetate 6x/day OD
Day 21	14/14		0.5+ cell, trace flare	Prednisolone acetate 6x/day OD Taper to QID OD after 1 week
Day 38	18/14		1+ cell, trace flare	Prednisolone acetate 6x/day OD
Day 45	17/10		0.5+ 1+ cell, (-)flare	Prednisolone acetate 6x/day OD Discussed re-initiating oral antiviral

## DISEASE COURSE

Herpes Zoster Ophthalmicus (HZO) is the result of reactivation of the varicella zoster virus along the ophthalmic division of the trigeminal nerve (CN V.). Following resolution of infection with varicella (chicken pox), the virus lies dormant in a ganglion-- in the case of HZO the trigeminal ganglion-- until reactivated by an insult, including illness, stress, and immunosuppression. Ten to 20 percent of people will develop zoster over their lifetime, with HZO accounting for one in four cases.

Typical extraocular presentation consists of a rash involving vesicles spreading along the top of the head, forehead, and eyelids. Hutchinson's sign, involvement of the tip of the nose, indicates a higher likelihood of ocular involvement. Prodromal phase includes influenza-like symptoms, including low grade fever preceding the lesions.

Patients may also present with blepharoconjunctivitis, corneal disease (including punctate epithelial keratitis, stromal disease, and neurotrophic disease), uveitis, and retinal necrosis. Uveitis is a complication which can present in HZO. Typical presentation occurs approximately one week after rash presentation.

Elderly patients are more likely to have zoster, as well as develop post-herpetic neuralgia. Treatment of HZO consists of a course of antivirals with supportive topical therapy. In some cases topical antivirals may also be prescribed. The best results are achieved when treatment is started within 72 hours of onset of symptoms.

FIGURE 2



## DISCUSSION

Recommended treatment for HZO is a regimen of oral antivirals: acyclovir 800mg 5x/day x 7-14 days, valacyclovir 1000mg TID x 7-14 days, or famciclovir 500mg TID x 7 days. Starting antiviral therapy within 72 hours has been shown to decrease length of active zoster as well as decrease likelihood of PHN. PHN can also be treated with narcotics, such as oxycodone, or gabapentin, which should also be started within the first 72 hours of infection.

## CONCLUSION

HZO manifests with a distinctive vesicular rash that extends from the forehead to the lower eyelid and respecting the midline. Additionally, the condition may lead to a number of ocular complications, more frequently seen in elderly patients. Patient 1 exhibited a classic pattern of vesicles which followed a normal course of resolution, with the exception of severe lid edema. Patient 2 exhibited an atypical vesicular pattern involving V<sub>1</sub> and V<sub>2</sub> dermatomes. She also developed an anterior uveitis.

Both patients showed signs of one of the most debilitating symptoms associated with this condition: pain. This may last for months following resolution of acute symptoms. Immediate treatment with full-dose antiviral medication has been shown to decrease the likelihood and duration of PHN. Both our patients were initially prescribed antivirals less than the recommended dose at external clinics, which may have contributed to their PHN. Additionally, the literature suggests that elderly patients may develop residual complications even months after the acute episode and are more likely to be physically debilitated by their symptoms, warranting close monitoring of these individuals.

## REFERENCES

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# CLASSIFICATION OF LONG ANTERIOR LENS ZONULE CLINICAL PATTERNS

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## INTRODUCTION

Long anterior lens zonules (LAZ) are characterized by zonules that extend more central than usual on the anterior lens capsule (Fig. 1).<sup>1</sup> One LAZ type occurs with gene mutation and late-onset retinal degeneration.<sup>2,3</sup> Another variety, which may have prevalence near 2%, has unknown etiology and is associated with age >50 years, female sex, hyperopia, shorter axial length, and iris pupil strands.<sup>4,7</sup> Due to rubbing against the posterior iris, LAZ may be pigmented and associated with pigment dispersion signs, including Krakenberg's spindles and angle pigment.<sup>1,5,8</sup>

LAZ have also received some attention because they might have association with open- and narrow-angle glaucoma,<sup>6,9</sup> and because they cause smaller zonule-free zones (ZFZs), which has relevance to cataract surgery.<sup>10-12</sup> Recently, we observed that LAZ have distinct forms, one in which zonules can be visually traced from an anterior tip all the way to their obscuration at the pupil border and another in which fibers end abruptly, without visible extension all the way to the pupil border (Fig. 1). To gain more understanding, we more systematically studied these different clinical presentations.

## METHODS

Subjects for this analysis have been studied previously and were part of an existing dataset accumulated within an urban, academic eye care facility in Chicago, Illinois, USA.<sup>6</sup> The criterion for LAZ was zonule fibers present >10 mm central to the normal zonule insertion zone. Only eyes with >5 LAZ fibers were included to insure definitive cases. Only African-Americans were included due to facility demographics.

To document LAZ, retro-illumination photos of the anterior capsule were taken (Haag-Streit BX900<sup>®</sup>, Haag-Streit AG, Koeniz, Switzerland) with a light beam just inside the nasal and temporal pupil borders (Fig. 2, top). To merge unobstructed image halves and enhance them for LAZ tracing, we developed a custom program using MATLAB<sup>®</sup> 6.1 (The MathWorks, Inc., Natick, MA, USA). After LAZ tracing by a masked observer (CM) (Fig. 2, middle), three observers (DR, TN, CM) categorized eyes via consensus into one of three overall LAZ patterns based on the predominant type of LAZ fibers present (Fig. 2, bottom). "Non-segmental" LAZ fibers were considered present if the most peripheral portion of a zonule could be traced to the border of the dilated pupil, where it becomes obscured by the iris. If 75% or more of the visible LAZ fibers were non-segmental, then the eye was classified as having the "non-segmental LAZ pattern." "Segmental" LAZ fibers were considered those in which the most peripheral portion of a zonule appeared to end abruptly, without extension to the border of the dilated pupil. If 75% or more of the visible LAZ fibers were segmental, then the eye was classified as having the

"segmental LAZ pattern." If neither non-segmental nor segmental LAZ fibers comprised 75% or more of the visible LAZ fibers, then a "mixed LAZ pattern" was considered present.

We explored relationships between LAZ patterns and other variables, including ZFZ size. Our calculation of ZFZ size has been reported elsewhere,<sup>13</sup> which was estimated only for subjects with available lens optic section photos that also yielded view of the overall iris (Fig. 2). A masked observer

FIGURE 1

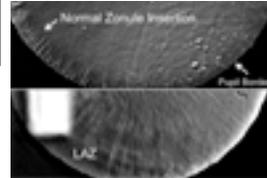
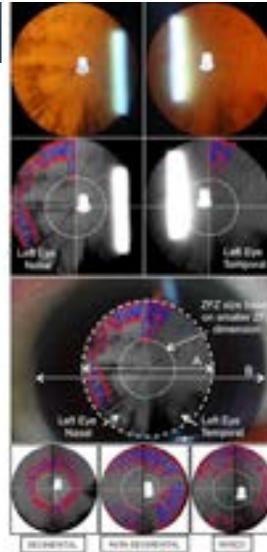


FIGURE 2



(SF) used EyeCap™ SL V3 software (Haag-Streit AG, Koeniz, Switzerland) to calculate a "pupil-to-iris diameter ratio." This enabled calculation of ZFZ size, which we based on three iris diameter assumptions (11.5, 12.0, 12.5 mm).<sup>14</sup> ZFZ outlines were drawn from each eye's most anteriorly-extended LAZ fiber. Analyses were conducted using the SAS<sup>®</sup> System, 9.3 for Microsoft Windows<sup>®</sup> (SAS Institute, Inc., Cary, NC, USA). P-values <0.001 were considered significant. Institutional Review Board approval was obtained for the investigation.

TABLE 1 SUBJECT CHARACTERISTICS AND LAZ PATTERNS

Total (N=59 (54 female, 5 male) Median Age = 70 (53-91) years	
<b>Right Eyes</b> N=51 (47 female, 4 male) Median Age = 70 (53-91) years *Median SR = +1.89 D (-1.88 to +5.38 D)	<b>Left Eyes</b> N=51 (48 female, 3 male) Median Age = 69 (53-91) years *Median SR = +1.56 D (-1.38 to +7.38 D)
<b>LAZ Patterns</b> Non-segmental: 47.1% (N=24) Segmental: 35.3% (N=18) Mixed: 17.6% (N=9)	<b>LAZ Patterns</b> Non-Segmental: 51.0% (N=26) Segmental: 29.4% (N=15) Mixed: 19.6% (N=10)

\*Abbreviations: D, diopters; LAZ, long anterior zonules; mm, millimeters; N, number; SR, subjective refractive error; †Spherical equivalent refractive error

TABLE 2 DISTRIBUTION OF LAZ<sup>†</sup> PATTERN TYPES RELATIVE TO OTHER VARIABLES<sup>‡</sup>

	NON-SEGMENTAL (N=24/26, RE/LE)	SEGMENTAL (N=18/15, RE/LE)	MIXED (N=9/10, RE/LE)
<b>‡ZFZ, mm</b>			
**RE	4.4 (2.6-5.8)	2.6 (1.9-3.8)	2.9 (2.6-3.8)
**LE	4.2 (1.7-6.9)	2.4 (1.6-3.3)	3.1 (2.1-5.2)
<b>RE</b>	65 (5-207)	39.5 (5-149)	35 (7-142)
<b>LE</b>	76.5 (10-267)	29 (9-157)	58 (8-142)
<b>Age (years)</b>			
RE	71 (56-88)	66.5 (53-80)	73 (59-91)
LE	68.5 (56-88)	68 (55-91)	74.5 (53-86)
<b>‡Refractive Error (D)</b>			
RE	+1.94 (-1.88 to +8.38)	+1.44 (-0.50 to +7.25)	+2.50 (1.13 to +7.75)
*LE	+1.88 (-1.88 to +5.13)	+0.50 (-3.50 to +7.38)	+1.63 (-1.03 to +4.63)

\*Abbreviations: D, diopters; LAZ, long anterior zonules; mm, millimeters; N, number; RE/LE, right/left eye; †ZFZ, zonule-free zone; ‡ZFZ data available for N=42 right eyes and N=41 left eyes.  
\*Median values (range)  
†Spherical equivalent refractive error  
‡Significant differences between segmental and non-segmental groups (P<0.01)  
§Significant differences between segmental and non-segmental groups (P<0.001)  
¶Significant differences between segmental and non-segmental groups (P<0.0001)

## RESULTS

Fifty-nine African-Americans (54 females) with median age=70 years (53-91 years) were included. Distribution of the 3 LAZ pattern types relative to the right and left eyes and their characteristics are shown in Tables 1 and 2. With the univariate analysis, when we excluded the mixed LAZ pattern type and compared eyes with the non-segmental LAZ pattern against eyes with the segmental LAZ pattern, strong relationship was found between the segmental LAZ eyes and smaller ZFZs (P<0.0001/P<0.001, right/left eyes). The segmental LAZ pattern eyes tended to have fewer LAZ per eye than the non-segmental LAZ pattern eyes but differences did not reach statistical significance (P<0.07/P<0.10, right/left eyes). Age and refractive error did not show consistent relationship with LAZ pattern type.

## CONCLUSIONS

We found that LAZ commonly exhibit different patterns, one type predominantly having LAZ fibers that can be visually traced from their anterior tip all the way to their obscuration at the pupil border (non-segmental LAZ), and another type mainly having fibers that end abruptly, without extension all the way to the pupil border (segmental LAZ). A relatively equal mixture of both LAZ types also occurs. Eyes with the segmental LAZ pattern tended to have smaller ZFZs than eyes with the non-segmental LAZ pattern. Future study should consider these pattern differences to help further understand relationships they may have.

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# Use of Diagnostic Drops in Pregnant and Nursing Women

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## INTRODUCTION

Little has been published on the use of topical anesthetic and dilating drops in pregnant or nursing women. During pregnancy, any drug that is absorbed systemically can affect both the pregnant woman and the fetus. Therefore, the clinician must consider the adverse effects of the drug itself as well as its potential for teratogenicity. Potential toxicity of drugs that are excreted in breast milk may also be difficult to detect. Virtually all investigations of milk secretion have been carried out in animals. There are considerable differences in the composition of milk in different species. Human milk is a suspension of fat and protein in a carbohydrate-mineral solution. Drug excreted into milk binds to proteins or onto the surface of the milk fat globules. Drugs that are present in the maternal circulation are therefore transferred into milk. [1]

Proparacaine, tropicamide, & phenylephrine are categorized as Pregnancy Category C. [2] The package insert for these drops states that they should be used in pregnancy "only if clearly indicated." No controlled studies have been performed for these drops. Animal reproduction studies have not been performed with these drops either. When used systemically, an association has been made between the first-trimester use of phenylephrine and minor fetal malformations which were not life-threatening. [3] Systemic phenylephrine may also cause fetal hypoxia and bradycardia. [4] It is unknown whether mydriatics are excreted in human breast milk. However, since it is known that low-weight infants (less than 1600 grams) are particularly susceptible to systemic hypertension when 10% or 2.5% phenylephrine eye drops are used, it is safest to avoid these drugs in a nursing mother. [5] Pseudoephedrine, a pharmacologically similar vasoconstrictor, decreases milk production in nursing mothers after oral use. [6]

The aim of this study was to take a poll of optometrists at Illinois Eye Institute who are involved in patient care to assess their use of diagnostic drops in pregnant and nursing women.

## METHODS

A survey was completed by 55 optometric faculty members at the Illinois College of Optometry. The survey was anonymous and inquired about years of practice. The subjects were asked questions about their practice habits with the use of proparacaine, tropicamide, and phenylephrine in pregnant and nursing women.

Figure 1: Drops used on a pregnant woman during a ROUTINE exam

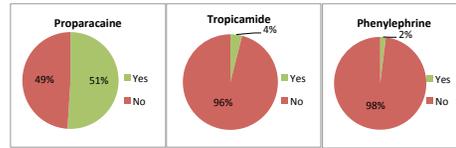


Figure 2: Drops used on a pregnant woman during a PROBLEM-BASED exam

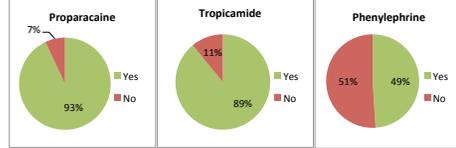


Figure 3: Drops used on a nursing woman during a ROUTINE exam

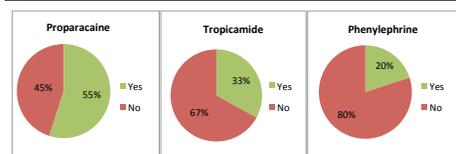


Figure 4: Drops used on a pregnant woman during a PROBLEM-BASED exam

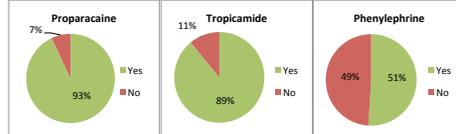


Figure 5: Concern that drops may cause a problem with the fetus

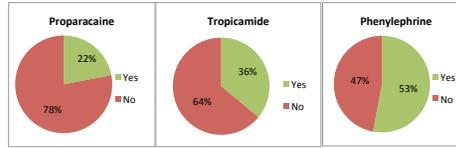


Figure 6: Concern that drops may cause a problem with a nursing infant through breast milk

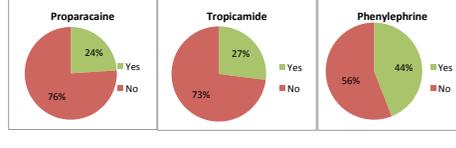
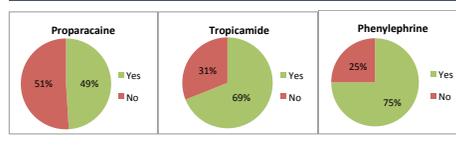


Figure 7: Medical-legal concern with using the drops



## RESULTS

Less than 4% of optometrists would use tropicamide or phenylephrine on a pregnant woman during a routine exam [Figure 1]. Over 85% of optometrists would use proparacaine or tropicamide on a pregnant patient during a problem-based exam where dilation or tonometry is indicated, yet only 49% would use phenylephrine [Figure 2]. Less than 35% of optometrists would use tropicamide or phenylephrine on a woman who is nursing during a routine exam [Figure 3]. Over 85% would use tropicamide or proparacaine on a woman who is nursing during an problem-based exam where dilation or tonometry is indicated [Figure 4].

Junior faculty (those in practice 5 years or less) were more aggressive in using tropicamide and proparacaine on pregnant or nursing women than senior faculty. There were no other significant trends based on years of practice.

Over 60% of optometrists surveyed had no worry about proparacaine or tropicamide causing a problem with the fetus or the nursing infant [Figure 5, Figure 6]. More than 65% of optometrists were concerned for medical-legal reasons about using phenylephrine or tropicamide in pregnant and nursing women [Figure 7].

## CONCLUSIONS

Overall the optometrists surveyed were conservative with using diagnostic drops on pregnant and nursing women on routine exams. There was the greatest concern with using phenylephrine even in urgent situations. While the majority of optometrists were not worried about problems with the fetus or nursing infant, most were still hesitant to use drops based on medical-legal concerns.

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# Therapeutic Use Of Scleral Lenses In A Patient With Superior Limbic Keratoconjunctivitis

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## BACKGROUND

Superior limbic keratoconjunctivitis (SLK) can present with mild clinical findings that pose a disproportional amount of patient discomfort. The etiology of the condition is not clearly understood and various causes such as eye rubbing, conjunctivochalasis, viral infection, localized tear deficiency, blink-related microtrauma and autoimmune disease have all been proposed. Hyperthyroidism is a systemic disease which has also been implicated in SLK<sup>4</sup>.

Patients typically show signs of sodium fluorescein and rose bengal staining of the superior cornea and limbus and can also have upper corneal filament formation. Classic treatment is with silver nitrate. More commonly used topical treatments include autologous serum and cyclosporine A. Vitamin A drops, ketotifen fumarate, and topical tacrolimus ointment have been suggested as adjunctive topical treatment options<sup>5-8</sup>. Procedures such as punctal occlusion and/or bandage contact lens application can also provide relief<sup>9,10</sup>. More invasive treatments include thermal cauterization, conjunctival resection, liquid nitrogen cryotherapy, conjunctival fixation sutures, supratarsal triamcinolone injections, and botulinum toxin injections<sup>9,14</sup>.

### Clinical Findings Associated with SLK

Blepharospastic eyelid  
Bulbar conjunctival inflammation  
Conjunctivochalasis  
Conjunctival keratinization  
Corneal filaments (especially in the upper cornea)  
Punctate staining adjacent to the superior limbus

## CASE

A 67-year-old Chinese female with no history of thyroid disease or contact lens wear presented with bilateral eye pain. She recalled a history of lid surgery (presumably blepharoplasty) approximately 10 years prior. She has a systemic history positive for hypertension which is diet/exercise controlled and also noted dry mouth symptoms. Entering distance VA cc was OD 20/70 and OS 20/60. Phenol red thread testing showed reduction at OD 10mm and OS 7mm. She was initially diagnosed with filamentary keratitis, keratoconjunctivitis sicca, blepharospasm and significant bilateral pterygium (see Figures 1 and 2).

Figure 1. Diffuse filamentary keratitis noted at an early visit.



Figure 2. Prominent pterygium which complicated bandage soft contact lens therapy.



She had limited relief of symptoms and had multiple recurrences of superior corneal filaments with the following treatment options:

- non-preserved artificial tears every hour (persistent symptoms)
- topical steroid and steroid/antibiotic combination drops (persistent symptoms)
- repeated filament debridement (recurrence)
- topical cyclosporine A emulsion 0.05% (persistent symptoms)
- soft bandage contact lens (ptyegium-related failure)
- cryopreserved sutureless amniotic membrane (recurrence)

Figure 3. SLK diagnosis: superior conjunctival staining with lissamine green dye.



After consult with ophthalmology for possibly pterygiectomy, the patient was formally diagnosed with superior limbic keratoconjunctivitis (see Figure 3) and referred back for scleral contact lens fitting. She was successfully fit into Jupiter 18.2mm scleral lenses OU (Visionary Optics, Front Royal, VA) with toric peripheral curves to vault the significant pterygia. With this liquid bandage scleral lens, vision improved to 20/25 OD and OS with improved ocular surface condition and reduction in severity of blepharospasm. She continued use of topical cyclosporine emulsion twice daily as well as preservative-free artificial tears hourly when not wearing the lenses.

The patient notes the condition has improved with continued scleral lens wear (see Figure 4). She no longer uses artificial tears every hour and enjoys comfortable wear of the lenses for 6-8 hours per day.

Figure 4. With scleral lens in place, improvement in the ocular surface can be noted.



## DISCUSSION

While this patient does not have any noted thyroid dysfunction, she still exhibits many of the classic signs of SLK including the characteristic superior arcuate conjunctival staining. Bloodwork was requested through her primary care physician for both Sjogren's markers (due to the dry mouth complaint) and thyroid function (due to its associated with SLK). All bloodwork came back within normal ranges except a mildly elevated serum IgG level and mildly reduced hemoglobin and hematocrit levels.

## CONCLUSION

Traditional SLK interventions can still provide ocular relief for some patients. Mini-scleral and scleral lens designs have been important in the therapy of ocular surface disease through their use as a tear reservoir and can represent alternate therapy for SLK before more invasive management techniques are attempted. In the presence of significant pterygia, toric peripheral systems can improve scleral lens fitting success and patient comfort.

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# Bacterial (gonococcal) Conjunctivitis in an Adult Caucasian Male

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## INTRODUCTION

Bacterial conjunctivitis is rare in adults, if bacterial is suspected the pathogen is typically staphylococcal species, *Streptococcus pneumoniae* or *Haemophilus influenzae*. In rare cases, the conjunctivitis can be due to a sexually transmitted disease. Gonococcal conjunctivitis presents with a mucopurulent discharge, conjunctival chemosis, eyelid swelling and may involve the cornea. This case represents bacterial (gonococcal) conjunctivitis in an adult Caucasian male.

## PERTINENT FINDINGS

29 y/o Caucasian male presents with red, swollen shut right eye with discharge for 3 days (see figure 1)

**OCULAR HISTORY:** LEE 5 y/o, no previous occurrences, no contact lens wear

**MEDICAL HISTORY:** unremarkable, denies any known sexually transmitted diseases

**MEDICATIONS:** Tobradex 6x/day OD x 3 days (prescribed QID by outside O.D.)

### VA (cd)

- OD 20/200, PH 20/150
- OS 20/25+2

### SLIT LAMP OD

- Lids/lashes: 2+ upper lid edema, mild tender to touch, 2+ MGD, thick mucus and dried on lashes (see figure 2)
- Conjunctiva: 4+ bulbar injection and 3+ edema, mucopurulent discharge, palpebral petechial hemes
- Cornea: 1+ corneal folds and edema
- A/C: D & Q
- Fundus: unremarkable

Figure 1: Self-reported onset of symptoms, early discharge and lid swelling



Figure 2: Presentation to clinic, 3 days after onset (post saline rinse), eye swollen shut



**Other:** (+) preauricular lymphadenopathy on affected side, denies pain on urination or genital sores

### LABORATORY TESTING

- STD testing performed 11 days after initial presentation
- Tested positive for gonorrhea (throat), negative for chlamydia, syphilis and HIV

**DIAGNOSIS** Bacterial (gonococcal) conjunctivitis OD

**DIFFERENTIAL DIAGNOSES** Other bacterial conjunctivitis other than gonococcal: *Staphylococcus aureus*, *Staphylococcus epidermidis*, *Streptococcus pneumoniae*, and *Haemophilus influenzae*

## DISCUSSION

Common ocular signs/symptoms

- Profuse, hyperacute purulent discharge
- Severe conjunctival chemosis, conjunctival vessel dilation
- Eyelid swelling, eye pain on palpation
- Epithelial or stromal keratitis (variable), decreased vision

Complications of gonococcal conjunctivitis: corneal perforation within 24 hours without treatment

Patient follow up schedule after initial presentation: 3, 7, 20 and 26 days

### TREATMENT

- Initial: Azithromycin 500mg PO x 3 days, Besivance TID OD, saline wash, PFATs, d/c Tobradex
- Day 11: 250mg IM Rocephin/Ceftriaxon and 1000mg PO Azithromycin (walk-in clinic)
- Day 20: Lotemax QID OD, PFATs, d/c Besivance
- Day 26/maintenance: PFATs OU

### RESPONSE

- Day 3: improvement of vision to 20/50+
- Day 7: 20/20 vision, no corneal involvement
- Day 8: resolution of discharge
- Day 20: resolution of bulbar conjunctival edema
- Day 26: resolution of conjunctival injection
- Steady improvement of injection/chemosis throughout follow ups (see figures 3-5)

Figure 3: Day 2 follow-up: 2+ bulbar conjunctival injection and chemosis, improvement of lid swelling



Figure 4: Day 7 follow-up: improvement of injection, chemosis and lid swelling



Figure 5: Day 20 follow-up: trace conjunctival injection remains



## DISEASE COURSE

- Incidence of gonorrhea in adults (2013 CDC): 0.26% of men ages 25-29 in the US test positive for gonorrhea. Highest prevalence age group (0.54%) is men 20-24.
- Etiology
  - *Neisseria gonorrhoeae* is a gram-negative diplococci
  - Incubation period of gonococcal ocular infection generally ranges 3-19 days
  - Contaminated fingers with urine or genital secretions, oculo-genital spread, and contaminated fomites are common routes of transmission
  - Urethral symptoms typically precede ocular symptoms from one to several weeks – not in this case
- CDC recommended treatment for gonorrhea:
  - For uncomplicated gonococcal infections of the: Ceftriaxone 250mg IM in a single dose plus Azithromycin 1g orally in a single dose
  - For gonococcal conjunctivitis: Ceftriaxone 1 g IM in a single dose plus Azithromycin 1 g orally in a single dose, and consider one-time lavage of the infected eye with saline solution

## CONCLUSION

- Although rare, bacterial conjunctivitis can occur in the adult population
- Do not rule out STD infections if patient denies them
- Systemic symptoms do not need to precede ocular symptoms
- Refer when necessary to co-manage with primary care physician or infectious disease specialist for systemic treatment prevent permanent vision loss

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# Endogenous Endophthalmitis comparison DDX in Known vs. Unknown immunocompromised patients

Endophthalmitis has potential grave consequences whether it is exogenous or endogenous<sup>1</sup>. The latter requires an extensive systemic workup often looking for novel etiologies<sup>2</sup> compared to the former<sup>3</sup>. Analysis of vitreous fluid that includes staining, PCR and both bacteria and fungal culture is necessary.

Thomas R Stelmack, O.D. FAAO Jesse Brown VAMC Illinois College of Optometry

## Case #1 Known Immunocompromised Pt

48 year old, WHITE, MALE presents in triage clinic states that 4 days ago he started going completely blind in OS (can only see light out of the eye) and it is spreading to OD. Pt denies getting anything in his eye or any trauma to the eye prior to this starting.

Personal and family ocular are essentially negative. LEE unknown.

### PAST MEDICAL HISTORY:

Has been seeing PCP outside VA.  
Patient denies history of HTN, DM, MI, CVA, COPD.  
HIV dx 1995 sexual risk; viral load: 343 (02/15); CD4 230 (05/15) wide variation;  
off HAART x 3 yrs genotype no resistance (darunavir/ritonavir/truvada)  
2. Recent tinea infection; PCP pneumonia and oral thrush  
3. Multiple STDs (rei-nfections as well)  
NKMA

### SLIT LAMP EXAMINATION:

LIDS/LASHES:  
OD: NL  
OS: NL

CONJUNCTIVA:  
OD: NL  
OS: NL

CORNEA:  
OD: NL  
OS: scattered KPs on endothelium

ANTERIOR CHAMBER:  
OD: D/Q  
OS: Deep, 3+ cell

IRIS:  
OD: NL  
OS: NL

INTRAOCULAR PRESSURES:  
OD: 15  
OS: 12 @ 3:00  
By: GAT  
1% Mydracil and 2.5% Phenylephrine @ 3:01 pm

LENS:  
OD: tr NS  
OS: tr NS

### VISUAL ACUITIES: sc

OD: 20/20-1  
OS: LP c projection PH: NI

**PUPILS:** + Direct and consensual without APD  
**EXTRAOCULAR MUSCLES:** Full Range of Motion, both eyes  
**CONFRONTATION FIELDS:** Full to Finger Counting, both eyes

**DDX: VZV vs. CMV vs. opportunistic bacteria / fungal**

### DILATED FUNDUS EXAMINATION:

CUP/DISC RATIO: OD: 0.3/0.3 OS: no view

OPTIC NERVE HEAD:  
OD: NL

MACULA:  
OD: NL

VITREOUS:  
OD: NL  
OS: 4+ vitreous cell

VESSELS:  
OD: NL

PERIPHERY:  
OD: very small choriorretinal scar at 6 o'clock - likely old

OS: difficult view but superior retina intact with no evidence of ARN or CMV retinitis; hazy view inferior-temporal; cannot rule out peripheral patchy areas of necrosis; posterior pole with no gross view

B-scan: no evidence of RD in any quadrant; has thick/dense vitritis condensing primarily along hyaloid face in all quadrants.



## Case #2 Unknown Immunocompromised Pt

50 year old, WHITE, MALE presents for DFE and f/u for likely ischemic partial CN III palsy. MRI / MRA normal. Pt reports double vision resolved about a week ago. He was a no show for carotid duplex. Carotid testing scheduled for 05/26/15 He now complains of left red eye/ blurry vision that started a week ago with 3/10 pain (like someone pushing on eye), denies irritation, denies trauma, denies allergies, (+) valsalva 2 weeks ago lifting a futon mattress and (+) chest cold, cough one month prior.

Family Ocular hx neg.

### PAST MEDICAL HISTORY:

IDDM poorly controlled A1c: 10.6 BG 250+ dx 2.5 yrs ago  
Htn borderline control  
Recent UUTI from urethral stricture tx IV Ab  
Followed by bacteremia and fungemia (Candida)  
NKMA

Ocular Hx: prior PPDR + CWS

### SLIT LAMP EXAMINATION:

LIDS/LASHES:  
OD: NL (-) ptosis  
OS: NL

CONJUNCTIVA:  
OD: NL  
OS: deep injection diffuse, subconj heme nasal

CORNEA:  
OD: NL  
OS: endothelial haze (+) tearing

ANTERIOR CHAMBER:  
OD: D/Q  
OS: Deep 4+ wbc, (+)fibrin (+) light sensitivity

IRIS:  
OD: (-) NVI OU  
OS: NL

INTRAOCULAR PRESSURES:  
OD: 18  
OS: 14 @ 3:31 pm  
By: GAT  
1% Mydracil and 2.5% Phenylephrine @ 3:32 pm

LENS:  
OD: 1+ NS  
OS: 1+ NS, post synechiae 360, (+) fibrin on ant lens capsule

### VISUAL ACUITIES: cc, habitual

OD: 20/20  
OS: 20/400 PH: NI

**PUPILS:** + Direct and consensual without APD  
**EXTRAOCULAR MUSCLES:** FROM OD, OS (-) diplopia ;CT at distance (sc):  
CLXT  
**CONFRONTATION FIELDS:** Full to Finger Counting, both eyes

**DDX: Candida vs. Gm - bacillus**

### DILATED FUNDUS EXAMINATION:

\*\*\*poor view OS 2/2 to poor dilation and vitritis  
CUP/DISC RATIO:  
OD: 0.25V/H  
OS: 0.40 c dense vitritis overlying

OPTIC NERVE HEAD: (-) NVD OU  
OD: NL  
OS: (-) hyperemia

MACULA: (-) CSME OU  
OD: NL  
OS: NL

VITREOUS:  
OD: NL  
OS: (+) dense vitritis

VESSELS: (-) hemes, (-) infiltrates OU  
OD: NL  
OS: NL to view

PERIPHERY: (-) NVE OU  
OD: NL  
OS: dense snow balls inferior temporal

B-scan:  
OS: (+) vitritis (-) RD, (-) mass

### Initial Tx:

Procedure: OS PPV/Cx/Intravitreal Injection of Antibiotics (Ceftazidime 2.25 mg/0.1cc, Vancomycin 1 mg/0.1 ml, Voriconazole 100 mCg/0.1cc)

Specimens: TWO, One Vitreous Sample, Second Vitreous

### ENDOPHTHALMITIS CASE #1 LAB RESULTS:

Known HIV: CD4 & PCR quant  
Vitreous: Neg for bacteria /fungal Including AFBs  
Creatinine: mildly elevated

**RPR: Reactive 256 dils! (10 day course IV PCN resolved Casette in Blood Culture**

### WORK UP FOR ENDOGENOUS ENDOPTHALMITIS

#### Bacteria / parasite: (serum)

1. Quantiferon Gold
2. Toxoplasmosis: IgG/ IgM
3. RPR / CIA

#### Viral: (serum)

1. HSV: IgG/IgM
2. VZV: IgG / IgM
3. CMV (IgG/IgM)

#### Urine analysis / cultures (prior UTI case #2)

1. Ph, Protein, glucose, ketones, bilirubin, nitrite, urobilinogen, color, appearance

2. Leukocyte esterase, RBC, bacteria, WBC/HPF, Epithelial cells

**Chem (serum):** eGFR, BUN, Na, K, Cl, CO<sub>2</sub>, glucose, creatinine, anion gap, Ca

**CBC (blood):** Platelets, PMNs, IG%, WBC, RBC, Hgb, Hct, MCV, MCH, MCHA, RDW, MPV  
%: LYPHS, MONOCYTES, GRANULOCYTES.  
ABS: LYPHS, MONOCYTES, GRANULOCYTES

**AC / Vitreous tap:** PCR (HSV, VZV, CMV), Culture / stain for Bacterial / fungal

### Initial Tx:

Declined Tap / PPVx / Infusion

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### ENDOPHTHALMITIS CASE #2 LAB RESULTS:

Urine:  
Leukocyte esterase, ++  
Occ Bacteria  
Trace protein  
Blood moderate  
WBCs > 50+++  
Epithelial cells: few  
Albumin / protein very high; however Serum BUN / Creatinine nl

Serum:  
Glucose 311!  
Creatinine moderately high  
HSV2 IgG 75.00 high! HSV 1 neg  
CMV IgG 3.26 H++

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# An Emotional Thygeson's SPK

Andrew Ta, OD

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## INTRODUCTION

In 1950, Thygeson first described a condition consisting of transient, bilateral presence of multiple elevated, white-grey, granular, intraepithelial corneal lesions without associated stromal involvement, corneal edema, or conjunctival inflammation. Thygeson's SPK is a chronic condition characterized by exacerbations and remissions with symptoms including photophobia, blurred vision, foreign body sensation, tearing, and irritation.

A 54 year old male presents with alternating unilateral and bilateral flare-ups of large Thygeson's sub-epithelial infiltrates exacerbated by emotional stressors for the past 17 years.

## CASE HISTORY

A 54 year old African American male reports a flare-up of symptoms OS starting yesterday, which includes 5/10 pain and constant blurry vision. He believed it began after a reading of elevated blood pressure.

Ocular History	Medical History
<ul style="list-style-type: none"> <li>Thygeson's SPK ~ 1998</li> <li>Normotensive glaucoma OU x 2006 - +Hb: mother</li> <li>10/0 steroid response</li> <li>HSV epithelial keratitis OD (01/12, 07/13)</li> <li>Recurrent episcleritis OS (12/10, 01/11, 02/11)</li> <li>Trauma OD (flat) without subconjunctive (07/13)</li> <li>DES without keratopathy OU</li> <li>Corneal abrasion OS (01/15)</li> </ul>	<ul style="list-style-type: none"> <li>CAD +/- PR cardiac stress 2004</li> <li>Orthostatic hypotension</li> <li>Neurotic depression</li> <li>Myocardial infarction (12/03)</li> <li>Hyperlipidemia</li> <li>Lower back pain</li> <li>NIDDM (05/10)</li> <li>Peripheral artery disease</li> <li>Nummular dermatitis</li> <li>HTN</li> </ul>

### Current ocular medications:

- 2gtts Restasis BID OD, OS
- 1gtt Vexol qid OD, OS
- 1gtt preservative-free artificial tears q2h OD, OS
- 1gtt latanoprost qPM OD, OS
- 1gtt Celluvisc qid OD, OS

## FINDINGS

	OD	OS
BCVA	20/20	20/20-2
Pupils	PERIL, (-) RAPD	PERIL, (-) RAPD
EDMs	FROM	FROM
CVF	FTFC	FTFC

Anterior Segment	OD	OS
LL	NL	NL
Conjunctiva	NL, (-) injection	NL, (-) injection
Cornea	Clear	~15 focal areas of epithelial edema central and superior, (+) negative staining, some in a vague linear pattern
Anterior Chamber	D/Q, (-) cells/flare	D/Q, (-) cells/flare
Iris	Diffuse atrophy, (-) NVI	Diffuse atrophy, (-) NVI
HOP (mmHg) by GAT	11	11

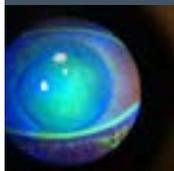
Posterior Segment	OD	OS
Leas	Tr NS	Tr NS
C/D	0.85V/0.90H, deep cupping, thin temp rim	0.80V/0.85H, deep cupping, thin temp rim
Macula	perifoveal soft druse, no CSME	NL, no CSME
Vitreous	NL	NL
Vessels	NL	NL
Periphery	NL	NL

Auxiliary Testing	OD	OS
OCT RNFL	SSR, Red (S,T,I), Yellow (N)	SSR, Red (S,T), Yellow (I), Green (N)
HVF	Isolated non-specific defects	Isolated non-specific defects

Figure 1



Figure 2



## DIFFERENTIAL DIAGNOSIS

- Thygeson's SPK
- HSV keratitis
- Dry eye syndrome with keratopathy
- Marginal infiltrative keratitis
- EKC
- Map-Dot-Fingerprint Dystrophy
- Exposure keratopathy
- Recurrent corneal erosion
- Bacterial keratitis
- Nummular keratitis

## DIAGNOSIS & DISCUSSION

On initial presentation, our patient was diagnosed with HSV keratitis and enrolled in the HEDS study. However, Dr. Joel Sugar, a corneal specialist at UIC, had evaluated the patient in 1998 and determined our patient had an atypical form of Thygeson's SPK. Our patient then began treatment with prednisolone acetate 1% susp qid OU, which improved his symptoms. This confirmed that the patient did not likely have HSV keratitis, as symptoms should have worsened with steroid treatment. Recurrent flare-ups of his symptoms were common and were often linked with emotional stressors. For example, self-monitored blood pressure readings that were elevated would cause the patient distress, as well as divorce and child custody, and result in a worsening of his condition. The patient's symptoms did not match with recurrent corneal erosions because our patient's symptoms were constant throughout the day during flare-ups, whereas symptoms of recurrent corneal erosions typically occur at the time of awakening. Also, the superior corneal staining pattern did not correlate well with keratopathy associated with dry eye syndrome or exposure. The lack of conjunctival injection ruled out marginal infiltrative keratitis, bacterial conjunctivitis, nummular keratitis, and epidemic keratoconjunctivitis. Map-dot-fingerprint dystrophy is consistently bilateral, usually asymptomatic, and does not improve with topical corticosteroid use, all of which did not match well with our patient's case. Because of the presentation, recurrences, and improvement with immunosuppressants, we solidified the diagnosis of recurrent Thygeson's SPK.

Thygeson's SPK is a chronic condition characterized by exacerbations and remissions. It consists of transient, bilateral presence of multiple elevated, white-grey, granular, intraepithelial corneal lesions without associated stromal involvement, corneal edema, or conjunctival inflammation. In a retrospective review of 40 patients by Nagra et al. 2004, the age of onset of symptoms or initial diagnosis was 28.7 years with a range of 2 to 60 years. Only 15% had unilateral disease and most patients (78.9%) had visual acuities of 20/30 or better, while the remaining were between 20/40 and 20/50 in their worse eye. The average disease time course was 11.1 years with the longest course being 40 years reported by Tanzer and Smith.

Currently, there is no clear etiology for Thygeson's, but it has been shown to be associated with HLA DR3. Thygeson's is not a bacterial etiology because of absence of bacteria, as well as lack of improvement with antibiotic treatment. There is question of viral etiology due to resemblance of epithelial lesions to those seen in other known viral conditions; however, Connell et al. 2007 was able to show the absence of HSV, VZV, and adenovirus in the epithelium of patients with Thygeson's SPK. After ruling out bacterial and viral etiologies and reports of efficacy of cyclosporine A in achieving long-term resolution, it is suggested that there is an immunological component.

## TREATMENT & MANAGEMENT

Thygeson's SPK has a good visual outcome, but Fintelmann et al. (2012) presented three case reports of scarring causing a reduction in vision. Given the potential of scarring, there is support for the use of topical corticosteroid to treat symptoms and reduce the risk of corneal scarring from a chronic inflammatory disease. Topical corticosteroid is the therapy of choice, but has risks and side effects from chronic use, such as secondary infections, cataract, and glaucoma. Reinhard and Sundmacher (1999) reported that cyclosporine A 2% achieved permanent resolution of lesions in up to 31% of patients with active disease. Tacrolimus, another topical immunomodulatory agent, was shown to be effective for controlling Thygeson SPK for a long period with good tolerance and no noticeable side effects (Marquezan et al. 2015). However, over the course of 6 years, tacrolimus was not curative.

In the past, our patient had been responsive to cyclosporine A 1% when it was formulated in the pharmacy, but after it became commercially available, it was difficult to control the flare-ups with Restasis (cyclosporine 0.05%) alone. Flare-ups were better managed when Vexol was added, but emotional triggers continued to spark flare-ups. Restasis was also increased to 2 drops twice a day to attempt to increase the concentration of cyclosporine. This combination has been effective in management of our patient, though flare-ups persist, as he remains sensitive to emotional stressors.

## CONCLUSION

Thygeson SPK is a chronic condition that requires constant management as recurrences and flare-ups may occur frequently. Patients should be monitored closely when being treated with topical corticosteroids, especially in those who are steroid responders or diagnosed with glaucoma. More studies into the pathophysiology of Thygeson's will help guide treatment, but it will be difficult given the rarity.

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## ACKNOWLEDGEMENTS

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# A Comparison of Four Different Measures of Accommodation

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## INTRODUCTION

Accommodation, convergence and miosis play an important role in the ability to focus and perform near work<sup>1</sup>. As a result, near point stress and visual complaints at near can be related to accommodation, specifically insufficient accommodation, excess accommodation, or inflexible accommodation<sup>2</sup>. Various methods of measuring accommodation are used clinically. Four methods routinely used include three subjective measures: push up, pull away, minus lens amplitude, and one objective measure, using an open field auto-refractor (Grand Seiko WAM-5500 Optometer). We evaluated these four methods to determine if they provide comparable results.

## METHODS

Thirty-seven Illinois College of Optometry students aged 22-32, with best corrected vision of 20/20 were tested. Each participant's accommodative amplitude was measured with the Grand Seiko WAM-5500 Optometer (WAM), push up, pull away, and minus lens methods in randomized orders. Tests were performed on the right eye, by convention. If the right eye was not corrected to 20/20, tests were performed on the left eye. See Figures 1 through 3 for more detail. The analysis of variance (ANOVA) was used to compare the four different methods of accommodation.

**TABLE 1**  
Mean Accommodative Amplitude of the Four Methods

Measures of Accommodation	Average Accommodative Amplitude (Diopters)
WAM	5.962 ± 0.252D
Minus Lens	8.478 ± 0.310D
Pull Away	8.910 ± 0.350D
Push Up	10.178 ± 0.571D

**FIGURE 1A &1B:**  
Grand Seiko WAM-5500 Optometer.  
Measuring accommodation with a 20/40 line target at 40 cm, in full illumination with minus lenses.



**FIGURE 2:**  
Minus Lens Method.  
Measuring accommodation using a 20/40 line at 40 cm until first sustained blur.



**FIGURE 3:**  
Push Up/Pull Away Method.  
Measuring accommodation using a box of 20/20 letters at near at a rate of 1-2cm/s.



**FIGURE 4:**  
Mean Accommodative Amplitude of the Four Methods



## RESULTS

Mean accommodative amplitudes ranged from lowest to highest as follows: WAM, minus lens, pull away, and push up (Fig. 4 and Table 1). ANOVA showed a statistically significant lower amplitude of accommodation with the WAM than the three subjective methods and a statistically significant higher amplitude of accommodation with the push up than the three other methods.

## CONCLUSION

Previous research by Taub and Shallo-Hoffmann compared the three subjective methods and found the push up and pull away methods correlated with Hofstetter's norms, while minus lens amplitudes underestimated accommodative amplitude<sup>3</sup>. In our study, the push up method showed the largest accommodative amplitudes. This may suggest that the push up method overestimates accommodative amplitudes. Our study also indicates lower accommodative amplitudes measured objectively than subjectively, which mirrors a previous study<sup>4</sup>. As a result of these findings, further research needs to be completed to develop objective normative values of accommodative amplitudes.

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# Co-managing Optic Neuritis, in the Absence of a Systemic Diagnosis, in a Travelling Patient

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## BACKGROUND

Optic neuritis is an acute inflammatory disorder of the optic nerve, which typically presents in young adults, more commonly in women. Optic neuritis manifests with sudden monocular visual loss and eye pain. It is a common initial manifestation of multiple sclerosis (MS). Therefore, a high incidence of optic neuritis is correlated with MS diagnosis. This case represents co-management of optic neuritis, in the absence of a systemic diagnosis, in a travelling adult Asian female.

## PERTINENT FINDINGS

31 y/o Asian female presents with blurry vision, eye pain, and visual field loss for 7 days in the left eye

**OCULAR HISTORY** (+) Myopia. No previous episodes of current symptoms

**MEDICAL HISTORY** (+) Diabetes mellitus type 2, onset 2013, controlled with medication  
(-) Prior diagnosis of Multiple Sclerosis  
• (-) Numbness  
• (-) Tingling  
• (-) Weakness or fatigue  
• (-) Pain

**MEDICATIONS** Tobradex 4x/day OS (prescribed by an OD at an unaffiliated practice), Metformin

**VA** (cc) OD 20/20  
OS 20/100, PH 20/60

**PUPIL TESTING** ERRL OU, (+) APD 2+ OS

**EOMs** (+) Pain on right and left gaze OS; FROM OD, OS

**RED CAP DESATURATION** Desaturation by 70% OS, (+) Reduced perception of light intensity

**BP** 118/74 mmHg, RAS

**SLIT LAMP** Unremarkable OD, OS

**DFE** Unremarkable OD  
(+) Disc edema, indistinct margins OS; (-) Disc hemorrhage

**VISUAL FIELD TESTING** Unremarkable OD, Generalized visual field defect OS

**CIRRUS OCT** Unremarkable OD  
(+) Disc edema OS, (+) Retinal axonal loss OS

**DIAGNOSIS** Optic Neuritis OS without formal diagnosis of Multiple Sclerosis

**DIFFERENTIAL DIAGNOSES** Ischemic optic neuropathy, Acute papilledema, Severe systemic hypertension, Orbital tumor compressing the optic nerve, Intracranial mass compressing the afferent visual pathway

## DISEASE COURSE

Optic neuritis is an inflammatory demyelinating disease of the optic nerve. The inflammatory process involves perivascular cuffing, edema in the myelinated nerve sheaths, and myelin breakdown. Myelin loss typically exceeds axonal loss.

Optic neuritis typically affects Caucasian females between ages 18-45. In the US, the annual incidence is 6.4 per 100,000. There is a high correlation between optic neuritis and MS, a demyelinating condition characterized by white matter lesions noted on MRI scans. In fact, the risk of developing MS after one episode of optic neuritis is 50%. Other less common etiologies include childhood infections or vaccinations such as measles, mumps, chickenpox, viral infections, granulomatous infections, and idiopathic etiologies. Clinically, in unilateral or asymmetric presentations, a relative afferent pupillary defect is present. Decreased color vision and pain on eye movement are also commonly noted. Additionally, only one-third of patients exhibit swollen optic nerves; the other two-thirds manifest with retrolubar optic neuritis. Treatment involves IV corticosteroids, which speed visual recovery.

## DISCUSSION

Our patient was advised to discontinue Tobradex. As this condition is not treated with topical medications, it was essential to re-establish patient expectations. The patient was also immediately referred to a local ER. Despite initially requesting to delay treatment, she agreed to pursue the immediate ER visit after a detailed discussion regarding the findings, urgency, impact of delay on ocular health, and the need to address a potential systemic etiology. A phone consult with the ER physician helped relay our findings and recommendations, as well as transition the patient's care. Though a neurologic consult was scheduled for the same day, the ER was unable to coordinate both the MRI and the administration of IV steroids. Thus, the patient returned to her home state and was guided to coordinate all necessary care without delay. Pertinent exam findings, including imaging results, were forwarded to her ophthalmologist. A diagnosis of MS or other underlying etiology has not yet been confirmed.

## CONCLUSION

It is imperative to refer a patient with optic neuritis for an immediate ER visit and treatment with IV steroids. Corticosteroids speed up visual recovery without affecting the final visual outcome. Given the correlation between optic neuritis and MS, MRI testing is beneficial in ruling out other differentials, confirming MS, or documenting baseline if white matter lesions are not noted. Once an MS diagnosis is confirmed, optometrists may significantly aid neurology in monitoring progression through OCT imaging, specifically optic nerve cubes and ganglion cell analyses. Throughout all phases of management, an open line of communication between all providers is necessary to ensure both comprehensive care for the patient and ease the transition of care. As seen in our case, this is even more imperative when the co-management extends beyond state borders.

## REFERENCES

Available upon request

FIGURE 1A. Visual Field OD

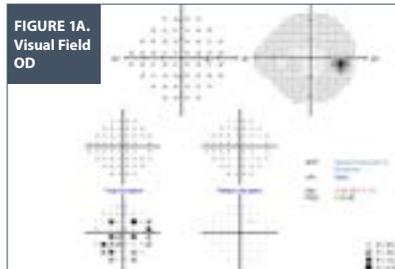


FIGURE 1B. Generalized visual field loss OS

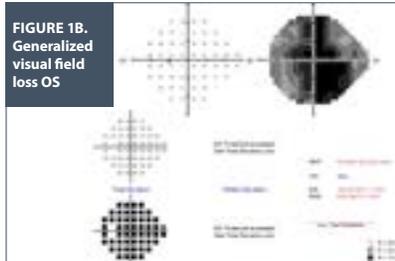


FIGURE 2A. Normal optic disc appearance OD



FIGURE 2B. Optic disc edema with indistinct margins OS

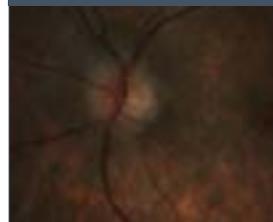
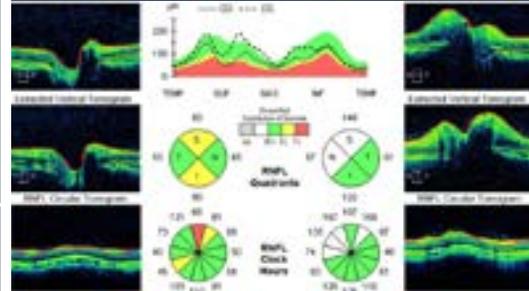


FIGURE 3. Optic disc edema OS



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# Painful Isolated CN III Palsy with Muscle Contracture Headache Comorbidity

Shannon Ver Woert, OD

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## BACKGROUND

A 59 YO AAM was sent to the eye clinic from the emergency department with an isolated, pupil-sparing cranial nerve palsy. The patient had a history of eye pain and photophobia, followed two days later by sudden onset diplopia, and four days later by stabbing 10/10 pain radiating from his right eyebrow over the top of his head to his neck. The pain was so severe that the patient had difficulty sleeping even with maximum dose Motrin (600 MG TID PO). Initial neurology evaluation documented a contralateral MR palsy, which was not noted by optometry or neurology on follow-up examination. Neurology initiated Gabapentin for ischemic pain.



Ocular and Medical History	
<b>Ocular History</b>	<b>Medical History</b>
<ul style="list-style-type: none"> <li>Moderate NPDR OD, OS                             <ul style="list-style-type: none"> <li>sip focal laser tx by outside MD for DM retinopathy OS</li> </ul> </li> <li>sip focal laser at Jesse Brown VAMC for CSME OD</li> <li>Early NS OD, OS</li> <li>Refractive error with presbyopia OD, OS</li> <li>Blepharitis OD, OS</li> <li>Mild DES 2/2 MGD OD, OS</li> </ul>	<ul style="list-style-type: none"> <li>IDDM dx and treated since 1997                             <ul style="list-style-type: none"> <li>LBG 204 (07/15)</li> <li>LAIC 8.2% (06/15)</li> </ul> </li> <li>HTN dx 1976 and treated since 1997                             <ul style="list-style-type: none"> <li>last BP 170/83 (06/15)</li> </ul> </li> <li>Hyperlipidemia</li> <li>Neuropathy</li> <li>Obesity</li> <li>Vitamin D deficiency</li> <li>Lower back pain</li> <li>Onychomycosis</li> <li>Right elbow pain</li> </ul>

Pertinent Clinical Ocular Findings		
	OD	OS
<b>Pupils</b>	+ Direct and consensual without APD	+ Direct and consensual without APD
<b>Pupil Size</b>	Bright: 1.5 Dim: 2.5	Bright: 1.5 Dim: 2.5
<b>EOMs</b> <i>*see figures 2-10</i>	Restricted nasal, superior and inferior movement, intorsion intact	FROM
<b>MRD 1/2</b> <i>*see figure 1</i>	-5mm/0mm	2mm/5.5mm
<b>Iris</b>	Flat, (-) NVI	Flat, (-) NVI
<b>Lens</b>	1-2+ NS	1-2+ NS

Fundus Evaluation:		
	OD	OS
<b>C/D Ratio</b>	0.60 V/H	0.65 V/H
<b>Optic Nerve</b>	Pink with distinct margins, (-) NVD	Pink with distinct margins, (-) NVD
<b>Macula</b>	large laser scar temp to macula, perifoveal D/B hemes and CWS, (-) CSME	focal laser scars, scattered MAS, perifoveal D/B hemes and CWS, (-) CSME
<b>Vessels</b>	scattered MAs and D/B hemes in arcades	scattered MAs in arcades
<b>Periphery</b>	Flat, intact X 360, (-) NVE	Flat, intact X 360, (-) NVE

Other Significant Information:	
<b>COVER TEST: CC at distance in primary gaze</b>	Horizontal: 18 PD BI Vertical: 8 PD BU OS
<b>Cranial nerve testing</b>	I, II, IV, V, VI-XII intact bilaterally
<b>Contraction furrows on right forehead in frontal region even with eye patch in place</b>	



Diagnostic workup:			
<b>Ordered by PCP</b>	<b>Ordered by ER on initial presentation</b>	<b>Ordered by Optometry</b>	<b>Ordered by Neurology: (all WNL)</b>
<ul style="list-style-type: none"> <li>HbA1C: 8.2%</li> </ul>	<ul style="list-style-type: none"> <li>MRI/MRA: no ICA, vascular malformations, or masses</li> </ul>	<ul style="list-style-type: none"> <li>ESR (WNL)</li> <li>CRP (WNL)</li> </ul>	<ul style="list-style-type: none"> <li>Lumbar puncture</li> <li>T4</li> <li>TSH</li> <li>ANA</li> <li>ANCA</li> <li>RPR</li> <li>Anti-TPO</li> <li>Anti-microsomal ab CXR</li> <li>CT head and neck</li> </ul>

Diagnoses:	
<b>DDx by Neurology</b>	<b>Diagnosis by Optometry</b>
<ol style="list-style-type: none"> <li>Pupil-sparing CN III palsy – ischemic</li> </ol>	<ol style="list-style-type: none"> <li>Painful pupil-sparing isolated right CN3 palsy, likely ischemic</li> </ol>
<ol style="list-style-type: none"> <li>Inflammatory process such as sarcoidosis</li> </ol>	<ol style="list-style-type: none"> <li>Right HA/periorcular ischemic pain with comorbid component of muscle contraction/tension HA from patient squeezing right eye shut while not wearing patch</li> </ol>
<ol style="list-style-type: none"> <li>Connective tissue diseases such as inflammatory ophthalmopathy</li> </ol>	<ol style="list-style-type: none"> <li>Moderate NPDR s CSME OU</li> </ol>
<ol style="list-style-type: none"> <li>Thyroid ophthalmopathy</li> </ol>	<ol style="list-style-type: none"> <li>Early Cataracts OU</li> </ol>

## TREATMENT & MANAGEMENT

- Patient was presented to neuro-ophthalmology conference, and the doctor concurred with the optometry diagnosis.
- Recommended full time patching and Motrin 600 mg TID PO for inflammation by optometry without relief, and then gabapentin by neurology.
- Educated patient as to muscle contracture headache comorbidity.
- Pain gradually improved over the next few weeks with patch use.
- Patient was educated on the cause of the condition and importance of strict BG control and follow-up with PCP.

## DISCUSSION

- CN III palsy is the ocular motor nerve palsy most commonly associated with pain.
- Ischemia is the most common etiology for a painful CN III palsy.
- When maximum dose Motrin does not manage pain, Gabapentin is an effective drug to use for pain management.
- Gabapentin is a drug used to control epilepsy that is now a mainstay in pain management.
- The exact MOA of Gabapentin as an analgesic and antiepileptic medication is unknown.
- Onset of ischemic pain may precede or be associated with onset of diplopia.
- Binocularity is commonly believed to promote healing of CN III palsies; however in this case, it was an aggravating factor.
- While it is important to rule out an aneurism as a causal factor current literature suggests that in patients with small vessel disease, it is appropriate to wait 4-8 weeks and monitor for resolution before initiating imaging studies.<sup>6,7</sup> However, it is still not widely agreed upon as to which palsies should be imaged and when.<sup>1,4,7</sup>
- Multiple headache etiologies may also be present; such was the case with this patient, who developed a muscle contraction HA after initial onset of eye pain.
- Our patient's persistent severe pain caused by tension headache and muscle contraction complicated his clinical picture.

## ACKNOWLEDGEMENTS

Charles W Kinnaird, OD, FAAO  
Michelle M Marciniak, OD, FAAO  
Thomas R Stelmack, OD, FAAO

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# The Effect of an Ocular Motility Diagnosis and the Subsequent Diplopia on a Patient with Early Onset Parkinson's

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## INTRODUCTION

Parkinson's disease (PD) is a neurodegenerative disorder affecting middle aged and elderly people. The etiology lies in a deficiency of dopamine in areas of the midbrain causing movement problems such as akinesia, rigidity and tremor. PD can also have non motor symptoms that affect a patient's well-being. Ocular motility issues are often seen in PD.

A 51 year old male was referred by his neurologist to treat his diplopia. No past history of diplopia or strabismus was noted. Our patient is a police officer and is unable to participate in work activity due to issues with gait and stability as well as issues with vision and diplopia. He was diagnosed with PD 3 years ago and diplopia onset was 2 years ago. Diplopia presentation is horizontal and vertical and noted most at distance. Frequency of diplopia has worsened since the onset and is now constant. Patient is unable to drive. The main goal of the patient is to be reinstated to work as a police officer in the next 10 months. Our patient is currently being treated with Sinemet.

### Parkinson Disease and the Ocular System

Visual Signs and Symptoms of Parkinson's Disease
Blurred Vision; Difficulty Focusing
EDM Related Diplopia
Difficulty with EOMs and Blinking
Difficulty with Reading or Driving a Vehicle
Difficulty Seeing in Dim Light
Decreased Color Vision and Contrast
Visual Spatial Disorientation
Hallucinations, Illusions, and Visual Misinterpretations
Reduced Convergence
Reduced Magnocellular Function and RNFL Loss
Reduced Accommodative Flexibility
Reduced Range of Clear Vision
Visual Midline Shift Syndrome
Narrowing of Ocular Veins

Ocular Treatments Options for Patients with Parkinson's Disease
Prism to Alleviate Diplopia
Horizontal Yoked Prism for Ocular Midline Shift
Base Down Yoked Prism to Increase Near Range of Clear Vision
Vision Therapy

## CLINICAL FINDINGS/OUTCOME

Our patient's diplopia was eliminated with the prism prescriptions. The IXT was reduced to a phoria at all distances. The single vision Rx allowed him to sustain fusion at the distance he was working at for longer periods of time. Prior to using a SV near Rx our patient was struggling with asthenopia, headaches, loss of place when reading and general fatigue with near work. The bifocal segment worsened the issues by decreasing the frequency of fusion due to the small progressive segment. He was unable to drive or complete work tasks due to diplopia at distance. The combination of the prism Rx and a much larger segment to work through proved successful. Nine months after the presentation of our patient his systemic and visual symptoms of Parkinson's were well controlled. He returned to his work and was able to reach the career milestones he desired in this time frame.

Initial Presentation	
Current Rx/VA	-3.75 -0.75 x 010 3ABU 20/40 -4.75 -1.25 x 135 20/20 +2.00 Add 29/20 VA OU
Distance Cover Test (cc)	20Δ IAXT, 7Δ LHT <i>High frequency XT, RE Fixation preference</i>
Near Cover Test (cc)	30Δ IRXT, 7Δ LHT <i>High frequency XT, RE Fixation preference</i>
Stereopsis	(-) Forms, (-) Fly
Worth 4 Dot	Diplopia response <i>Neutralized with 18Δ at near and 12Δ at distance</i>
Pupils	PERRLA (-) APD
EOMs	FROM OD, OS

Follow up	
Distance Cover Test (cc)	40Δ IAXT, 6Δ LHT <i>Variable deviation</i>
Near Cover Test (cc)	25Δ IRXT, 6Δ LHT <i>Variable deviation</i>
Stereopsis	(-) Forms, (-) Fly
Fresnel prism Rx	15Δ Fresnel application to OD

5 months of after initial presentation	
Case history	<i>Patient esotropia has stabilized, diplopia is intermittent, onset is typically at 2PM daily</i>
Distance Cover Test (cc)	16Δ IRXT, 6Δ LHT <i>Low frequency XT, no fixation preference noted</i>
Near Cover Test (cc)	16Δ IRXT, 16Δ LHT <i>Low frequency XT, no fixation preference noted</i>
Stereopsis	7/9 circles, (+) Forms, (+) Fly
Worth 4 Dot	Fusion at all distances
Final Rx I Single vision distance	-3.75 -0.75 x 075 3ABU, 1.5ABU -5.25 -1.00 x 145 3ABU, 1.5ABD
Final Rx II Single vision near	-2.00 -0.75 x 075 3ABU, 1.5ABU -3.50 -1.00 x 145 3ABU, 1.5ABD
Final results	<i>Patient returned to work 9 months after initial presentation. Our patient felt that his visual symptoms were his primary obstacle. His return to work was precipitated by the collaboration of his primary care physicians in conjunction with optometric treatment</i>

Variability of Horizontal Deviation		
Visit	Magnitude of deviation cc	Optometric Treatment Progress
Visit 1	20Δ IAXT 20Δ IAXT*	Monitor
Visit 2	16Δ IAXT 20Δ IAXT*	Monitor
Visit 3	40Δ IAXT 25Δ IAXT*	Monitor
Visit 4	20Δ IAXT 12Δ IAXT*	Fresnel prism dispensed
Visit 5	20Δ IAXT 16Δ IAXT*	Single vision Rx with ground in prism dispensed
Visit 6	16Δ IAXT 16Δ IAXT*	Decreased Diplopia Frequency; Decreased Symptoms
Visit 7	10Δ XP 10Δ XP*	Patient capable of being reinstated at work

### Intermittent Exotropia Left Eye Fixation



### Intermittent Exotropia Right Eye Fixation



## CONCLUSION

The presentation of a strabismic deviation in conjunction with a systemic diagnosis should be approached in a similar manner as a non-pathologic etiology. Our testing of the ocular motor system involved the use of cover testing, stereopsis assessment, Worth four dot and Maddox rod evaluation and most importantly case history regarding the needs of the patient. Small modifications to the spectacle prescription options aided in increasing the patient's abilities and moving him toward his goals. The optometric practitioner is critical in the treatment of an adult with Parkinson's to monitor and treat their visual symptoms.

Our patient viewed his visual symptoms as the greatest obstacle returning to his career. The control of diplopia with the proper prism Rx along with proper systemic medications and treatment contributed to his success.

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### Fusion with Single Vision Rx



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# Assessment of Smoking Status among Urban Adults Presenting for Eye Examination who were Covered by Medicaid/Medicaid Managed Care Plans

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## INTRODUCTION

Currently, Illinois and 29 other states / the District of Columbia have adopted Medicaid Expansion which has allowed many adults to qualify for Medicaid/ Medicaid Managed Care Plans (MCP) that were not previously eligible for health insurance.

This population may have faced/continue to face barriers to obtaining health care /health related education.

Smoking has been associated as a causative/exacerbating factor in ocular/ systemic diseases.

The 2012 National Health Interview Survey (NHIS) reported that **18.1%** of US adults currently smoke. A higher proportion was identified for males (20.5%), 'below poverty status' (27.9%) and age 25- 44 yrs. (21.6%)/45-64 yrs. (19.5%).

The purpose is to assess prevalence of smoking in this vulnerable population that presented for an eye exam at the Illinois Eye Institute (Chicago, IL).

## METHODS

Medical records were reviewed retrospectively:  
New patients/ comprehensive eye examinations (exam coded as 92004)  
≤18 years of age  
Examinations took place July 1 through December 31, 2014

Medical records were assessed for self-report of smoking status, age, gender, and health insurance. Those with Medicaid/MCP as a primary or secondary insurance were classified as Medicaid/MCP. Those whose smoking status was listed as 'unclassified' or whose insurance status was listed as ICO employee/student/family were excluded from analysis.

## RESULTS

There were 5004 adults included in analysis. 2379 were covered through MCP and 1480 through traditional Medicaid. In total, **3859** (77.1%) were **Medicaid/MCP**.

See Figure 1-4 for analysis of these 3859 patients.

Mean (SD) age was 49.0 years (14.4) for the group (n=3859). Mean (SD) age of those who reported current smoking was 50.4 years (14.4).

No significant difference in proportions between traditional Medicaid and MCP was found. Differences in smoking status and age were statistically significant.

Figure 1: Smoking Status

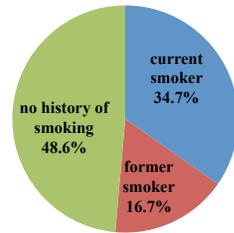


Figure 2: Smoking Subclassification

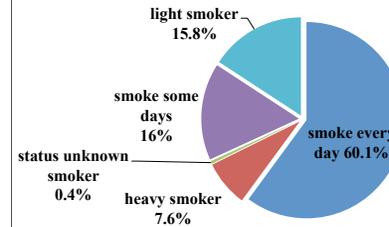


Figure 3: % Report Current Smoking By Age Group

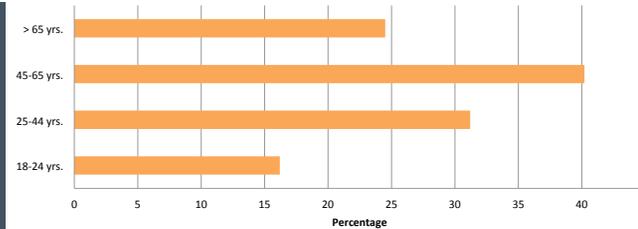
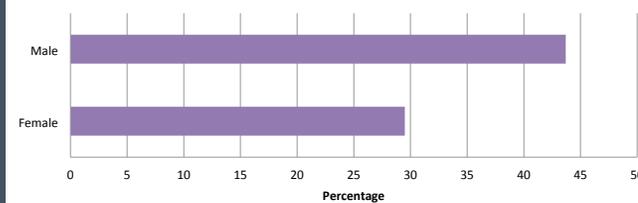


Figure 4: % Report Current Smoking By Gender



## DISCUSSION

More than 1/3 of the adults examined reported current history of smoking. Of those, more than 2/3 characterized their smoking as 'heavy' or 'smoking every day'. Although results cannot directly be compared, these findings are much higher than the national average of 18.1%. Although income was not directly assessed, due to insurance status of these patients it can be assumed that the group in general is lower income. Higher prevalence of smoking has been found among lower income groups.

A higher prevalence was found among males versus females which is consistent with national data. When age is considered, there was a large difference in smoking prevalence between age groups (range = 16.2% - 40.2%). The highest prevalence was found among those 45-65 years. The findings of this study are somewhat inconsistent with national data.

This study assessed new patients only. Although no specific data was available, many of these patients may not have had health insurance previously and so may not have received education from a health care provider on the importance of smoking cessation. This study demonstrates that high prevalence of smoking in this group and the need for eye care providers to promote smoking cessation especially among groups similar to the one studied.

This study is based solely on patient self-report of smoking. It is possible that report of smoking was not accurate. It seems likely if there were inaccuracies that smoking would be under-reported by patients therefore the prevalence of smoking might be higher than assessed.

## CONCLUSIONS

More than one-third of Medicaid/MCO adults reported a current smoking. The prevalence was higher among men and those 45-64 yrs. These findings cannot be directly compared to NHIS data however prevalence seems higher. These finding underscore the importance of healthcare coverage and smoking cessation programs/education for this population.

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# Hyphema and Iridodialysis Secondary to iStent Implantation

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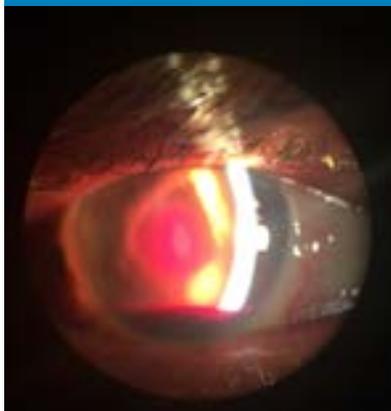
## INTRODUCTION

The treatment and management of glaucoma is evolving. As our ability to diagnose and monitor the disease has improved, the quest has continued for alternative treatment options. Due to the arduous nature of traditional glaucoma surgery, the search has been out for newer procedures that may suffice as a safer alternative to trabeculectomy for patients with mild to moderate glaucoma. Minimally invasive glaucoma surgeries (MIGS) are a subset of newly developed procedures that typically have a faster recovery, lower complication rate, minimal impact on refractive error, and preserve the conjunctival tissue should filtering surgery be warranted in the future. These procedures are optimal for patients with a target Intraocular pressure (IOP) in the mid- high teens to low twenties, with better IOP control seen when the target IOP is at the higher end of that range<sup>1</sup>. They are most commonly combined with cataract surgery, but may also be done in pseudophakic and less commonly phakic patients. Currently, the iStent is one of two FDA approved MIGS devices. The foundation of the iStent is to bypass the trabecular meshwork, which is thought to be the major source of resistance to outflow.

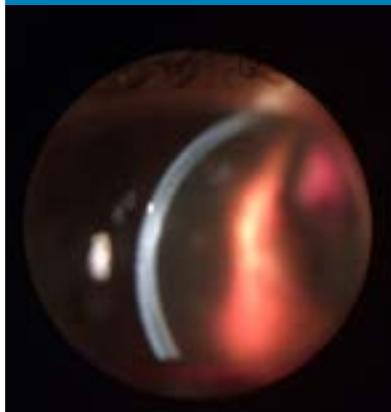
## CASE REPORT

A 76 year old Caucasian male with a history of angle closure glaucoma was referred for evaluation 23 days post phacoemulsification with intraocular lens implantation and attempted iStent insertion in the right eye. During the procedure, the iStent became dislodged from the insertion device and rested upon the iris. An iridodialysis was inadvertently created upon removal of the iStent, resulting in hyphema. The patient presented with light perception vision, iridodialysis from 2-4 o'clock and a non-clearing 1.4mm hyphema with 4+ suspended red blood cells (**photo**) and concomitant vitreous hemorrhage. His IOP was 25mmHg in the right eye while taking prednisolone BID OD, ketorolac BID OD, timolol/brimonidine BID OD, bimatoprost qhs OD and acetazolamide 250mg BID by mouth. It was determined that the patient would need additional surgery to stabilize him and he underwent surgery for a glaucoma drainage device (Ahmed), pars plana vitrectomy, iridodialysis repair and anterior chamber washout.

Hyphema secondary to iStent implantation



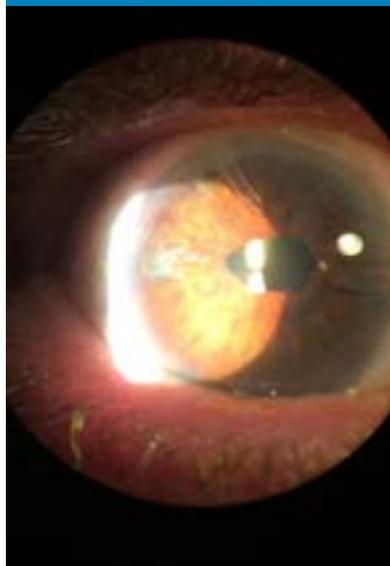
Iridodialysis secondary to iStent implantation



## DISCUSSION

The iStent is a 1mm heparin coated stent that is inserted with an applicator through a clear corneal incision and implanted through the nasal aspect of the trabecular meshwork into Schlemm's canal. The iStent can be difficult for surgeons to place, especially in restless patients or patients with peripheral anterior synechia or corneal edema. The most common complications of the iStent include elevated IOP and stent obstruction or malposition<sup>2</sup> which was most recently observed in 2.6%–18% of study subjects, of which 4.5% of these patients required surgical intervention<sup>3</sup>.

S/P GDD, pars plana vitrectomy, iridodialysis repair and anterior chamber washout.



## CONCLUSIONS

The ultimate goal of treatment strategies should be to achieve target IOP with the least amount of risk and impact to the patient. With the arrival of the MIGS, safe and effective surgical alternatives are now available to patients with early to moderate glaucoma. These procedures are potentially opening the door to mitigate earlier glaucoma related vision loss. Although these procedures are gaining acceptance and utilization by comprehensive ophthalmologists, the potential complications could arguably indicate the need for subspecialty in patients with coexisting cataracts and glaucoma, with patient care being the ultimate priority.

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# Relationship between Visual Acuity and Retinal Structures Using Spectral Domain Optical Coherence Tomography in Patients with Optic Nerve Hypoplasia

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## PURPOSE

Optic nerve hypoplasia (ONH) is a developmental disorder characterized by a small optic disc and/or a reduced number of retinal nerve fibers. We have reported that VA in ONH eyes has a moderate correlation with DM: DD ratio; worse VA in patients with higher DM:DD ratio.<sup>1</sup> Cirrus spectral domain optical coherence tomography (Cirrus SD-OCT) has been used successfully to measure parameters of both optic disc and retina. The purpose of this study was to evaluate the relationship between visual acuity and retinal structures measured by Cirrus SD-OCT in patients with ONH.

## METHODS

Twenty-two ONH subjects were recruited with a mean age of 28.60 (±18.23) years. All subjects had full eye examinations and were diagnosed with either unilateral or bilateral ONH, resulting in 30 ONH eyes. Cirrus SD-OCT obtained retinal nerve fiber layer (RNFL) thickness and macular ganglion cell analysis (GCA), which included GCA sector map, average, and minimum thickness of macular ganglion cell layer-inner plexiform layer (GCL-IPL). The correlations between best-corrected visual acuity (BCVA) and OCT parameters were evaluated using Pearson's correlation test and regression analysis.

## RESULTS

**Table 1** lists the demographic characteristics of ONH subjects. **Table 2** shows the OCT parameters of the ONH subjects.

- BCVA was statistically significantly correlated with RNFL thickness (**Figure 1**).
- BCVA was statistically significantly correlated with GCA parameters (average (**Figure 2**), minimum, superior, nasal, inferior, and temporal GCL-IPL thickness).
- Average GCL-IPL thickness showed a slightly higher correlation with BCVA ( $r = -0.718$ ,  $P < 0.001$ ) than did average RNFL thickness ( $r = -0.702$ ,  $P < 0.001$ ).
- BCVA was not statistically correlated with disc area ( $r = 0.282$ ,  $P = 0.146$ ) (**Figure 3**).

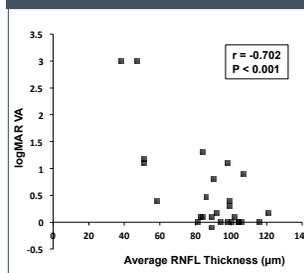
**TABLE 1**  
Demographic Characteristics of ONH Subjects (n = 22)

	Number of Subjects (%)
<b>Gender</b>	
Female	17 (77.3)
Male	5 (22.7)
<b>Race</b>	
Black	11 (50.0)
Hispanic or Latino	5 (22.7)
White	2 (9.1)
Asian	4 (18.2)
<b>Age (years)</b>	
Range	7.5 – 66.4
Mean (SD)	28.6 (18.2)

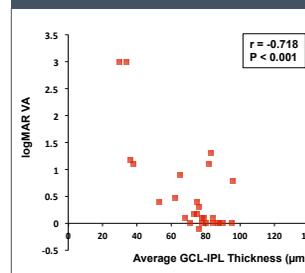
**TABLE 2**  
Correlation between logMAR VA and Optical Coherence Tomography Parameters in ONH Subjects

	Mean (SD)	Correlation with logMAR VA	
		r value	P Value
<b>Optic Disc Parameter</b>			
Disc area (mm <sup>2</sup> )	1.5 (0.4)	.282	0.146
Rim area (mm <sup>2</sup> )	1.3 (0.4)	.005	0.980
Average C/D ratio	0.3 (0.2)	.228	0.244
Average RNFL thickness (µm)	87.6 (21)	-.702	<0.001
<b>GCL-IPL Thickness (µm)</b>			
Average	71.9 (18.1)	-.718	<0.001
Minimum	61.4 (22.2)	-.599	0.001
<b>GCA Sector Map (µm)</b>			
Superior	74.2 (21.4)	-.639	<0.001
Superior nasal	72.7 (22.6)	-.723	<0.001
Inferior nasal	69.1 (25)	-.493	0.008
Inferior	70.9 (20.5)	-.573	0.001
Inferior temporal	72.6 (15.4)	-.732	<0.001
Superior temporal	71.6 (17.5)	-.718	<0.001

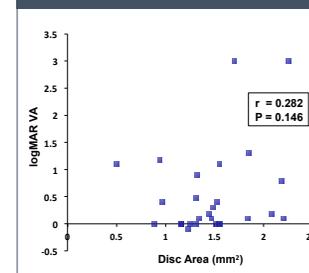
**FIGURE 1**  
Correlation between logMAR VA and Average RNFL Thickness in ONH Subjects



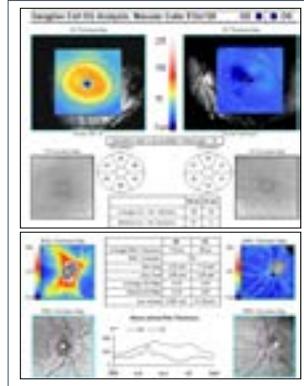
**FIGURE 2**  
Correlation between logMAR VA and Average GCL-IPL Thickness in ONH Subjects



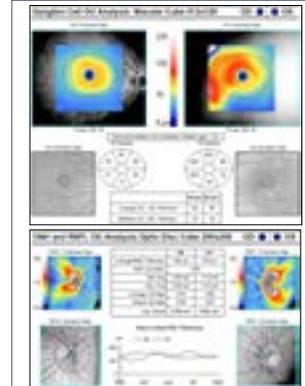
**FIGURE 3**  
Correlation between logMAR VA and Disc Area in ONH Subjects



**FIGURE 4**  
Reduced RNFL and GCL-IPL Thickness in an ONH Subject with NLP Vision (OS) (11 year old African American girl, BCVA OD: 20/20, OS: NLP, Refraction OD: +1.25-0.50 x 180, OS: +3.25-0.50 x 180. Normal disc area OD and ONH OS)



**FIGURE 5**  
Normal RNFL and GCL-IPL Thickness in a bilateral ONH Subject with 20/20 Vision (11 year old Hispanic girl, BCVA OD: 20/20, OS: 20/20, Refraction OD: -3.50-0.50 x 005, OS: -1.75-0.25 x 175. Small disc area OU)



## CONCLUSION

- BCVA was highly correlated with average RNFL thickness and GCL-IPL thickness, which can help to account for reduced BCVA in ONH patients.
- Although we found that BCVA was moderately correlated with the DM:DD ratio in a previous study,<sup>1</sup> BCVA was not correlated with disc area measured with Cirrus SD-OCT.

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## CONTACT INFORMATION

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# Non-arteritic Anterior Ischemic Optic Neuropathy in a Young African American Male

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## BACKGROUND

Non-arteritic anterior ischemic optic neuropathy (NAION) represents the most common acute onset optic neuropathy in patients over the age of 50, though no age is immune. Symptoms of NAION include sudden, painless vision loss, most frequently upon awakening. Visual acuity may be reduced and an arcuate or altitudinal visual field defect, particularly inferior, is common. In the acute setting, an ischemic event involving the anterior portion of the optic nerve head leads to unilateral optic disc edema. Disc hemorrhaging typically accompanies the edema. On average, the optic disc edema resolves within 4-8 weeks with subsequent retinal nerve fiber layer (RNFL) loss and diffuse or segmental optic disc pallor. There is no well accepted treatment for acute NAION.

## PERTINENT FINDINGS

32 y/o African American male presents with painless, sudden onset blurry peripheral vision and dulling of colors OS for 2 weeks.

**OCULAR HISTORY/MEDICAL HISTORY/MEDIATIONS:** unremarkable

**SOCIAL HISTORY:** former smoker, heavy alcohol use

**VA (sc):** 20/20-1 OD, OS

**CVF:** FTFC OD, OS

**EOMs:** FULL, (+) discomfort in right and left gaze OD, OS

**Pupils:** PE(3)R/L, 1+ APD OS

**HRR screening:** 6/6 OD, OS

**DFE:** Well perfused, flat and pink; not a "disc at risk" OD (Figure 1). Supernasal hyperemia with mild edema without disc hemorrhaging OS (Figure 2).

**HVF:** No field defects OD. Superior altitudinal field defect OS (Figure 3).

**Optic disc cube OCT:** Average NRR and RNFL thickness OD. Elevated RNFL OS (Figure 4).

**DIFFERENTIAL DIAGNOSES:** Non-arteritic anterior ischemic optic neuropathy (NAION), optic neuritis, compressive lesion, toxic optic neuropathy

**MRI:** No longitudinal enhancement of the left optic nerve (Figure 5).

Figure 1. Initial presentation right eye.



Figure 2. Initial presentation left eye.



Figure 3. Initial HVF of the left eye.

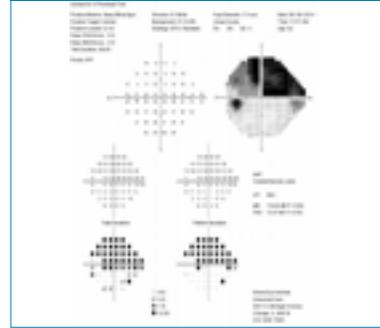


Figure 4. Initial optic disc cube OCT.

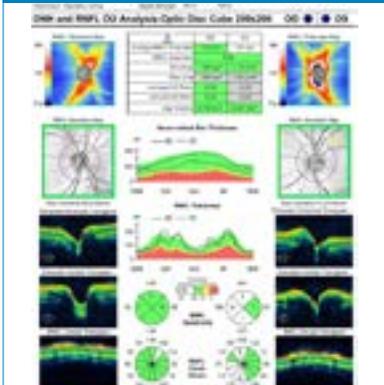
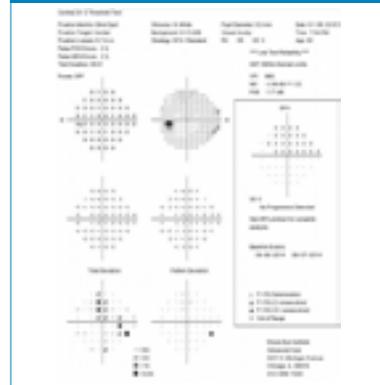


Figure 5. Baseline T2 MRI of the orbits post-gadolinium.



Figure 6. HVF of the left eye four months following initial event.



## DISCUSSION

Laboratory testing for syphilis, diabetes, and hyperlipidemia were unremarkable, and the patient did not have hypertension as assessed by his PCP. While an urgent MRI was recommended to rule out optic neuritis or compressive lesions, insurance complications significantly delayed the testing. MRI testing was unremarkable, demonstrating no enhancement of the left optic nerve post gadolinium, no compressive lesions and no ischemic lesions. For this reason, the diagnosis of atypical NAION was concluded. A coincidental granuloma of the right lung without bilateral hilar lymphadenopathy was identified with chest CT, while serum ACE levels were within normal limits. Using OCT technology, the optic disc edema persisted longer than average for NAION, resolving at the four month follow up. Without treatment, the visual field defect spontaneously improved to near normal levels four months following the initial event (Figure 6).

## CONCLUSION

Though NAION typically presents in patients over the age of 50, younger individuals are not immune to the condition. This case represents atypical NAION in a young adult male, without optic disc hemorrhaging, optic disc edema persisting beyond the typical two months, and significant visual field improvement. In the atypical acute setting of NAION in young patients, other organic causes for unilateral optic disc edema, including optic neuritis and compressive lesions, must be ruled out.

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## BACKGROUND

Idiopathic intracranial hypertension (IIH) is a condition that most commonly presents in obese females. The condition occurs most frequently in the 3rd to 4th decade. IIH often presents with symptoms of headache, transient visual obscurations, tinnitus, and pain. The biggest threat of IIH is vision loss. Additional presenting signs can include: visual field defects, papilledema, elevated intracranial pressure, specific neuroimaging signs, and a cranial nerve six palsy. The following supportive tests and studies should be ordered to support a diagnosis of IIH: MRI/MRV, lumbar puncture, optic nerve OCT, and Humphrey visual field.

## PERTINENT FINDINGS

44 y/o African American female presented with constant, left retrobulbar pain that had been occurring for the past nine months. The patient also reported tinnitus greater on the left side.

**PATIENT OCULAR & MEDICAL HISTORY:** A record review revealed that the patient presented 7 months previously to an outside provider with retrobulbar pain but no observed papilledema. She was diagnosed and treated unsuccessfully for cluster headaches. The patient most recently saw an eye care provider 2 months previously; she was prescribed reading glasses that did not clear the vision in her left eye. Additional medical history includes: hypertension, asthma, and seizure disorder.

**BCVA:** OD: 20/20-; OS: 20/20-

**BLOOD PRESSURE:** 120/75

**BMI:** 41.2

**SLIT LAMP EXAM:** within normal limits OU

### DFE:

- OD: mild optic nerve elevation (-) hemorrhages, (-) vessel obscurations (Figure 1); C/D ratio: 0.2V/0.2H
- OS: mild optic nerve elevation (-) hemorrhages, (-) vessel obscurations (Figure 2); C/D ratio: 0.2V/0.2H

### 24-2 HUMPHREY VISUAL FIELD:

- OD: scattered changes, blind spot enlargement (Figure 3)
- OS: scattered changes (Figure 4)

### SUPPORTIVE STUDIES:

- MRI/MRV: normal
- Lumbar puncture: Opening pressure: 240 mm H<sub>2</sub>O, normal components; Patient reported immediate relief of the pain behind her left eye

### DIFFERENTIAL DIAGNOSIS:

- Optic nerve drusen
- Tilted discs
- Congenitally crowded discs
- Optic discs with anomalous branching and tortuosity

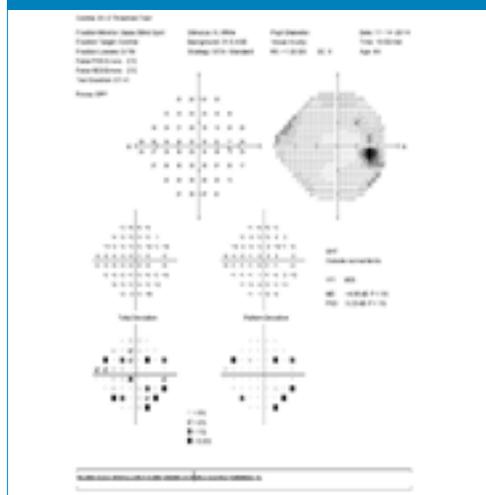
**Figure 1.** Cirrus™ OCT of optic nerve: elevated optic nerve, outward deflection of retinal pigment epithelium/Bruch's membrane layers, OD



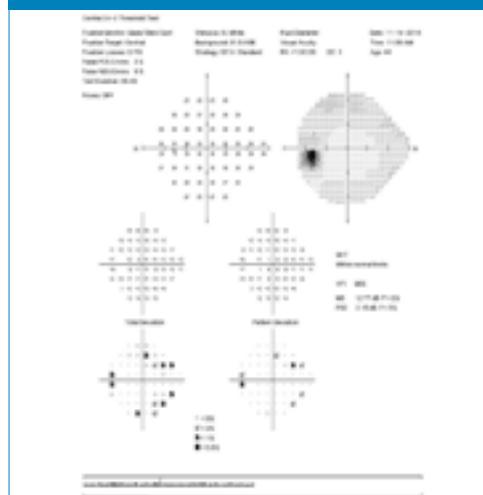
**Figure 2.** Cirrus™ OCT of optic nerve: elevated optic nerve, outward deflection of retinal pigment epithelium/Bruch's membrane layers, OS



**Figure 3.** 24-2 Humphrey visual field: scattered changes, blind spot enlargement, OD



**Figure 4.** 24-2 Humphrey visual field: scattered changes, OS



## DISCUSSION

Following a normal MRI and slightly elevated lumbar puncture opening pressure, the patient was started on 500 mg BID PO of acetazolamide. The patient reported initial relief, but her headaches and eye pain reoccurred over the next week. She also reported worse pain with sitting up. The acetazolamide was subsequently increased to 1000mg BID PO. She reported relief of symptoms with the increased dosage.

## CONCLUSION

This case is an example of idiopathic intracranial hypertension. It is important to control the signs and symptoms of the condition, as permanent vision loss can occur if it is left untreated. A prompt and appropriate work up can rule out other serious differentials and help to maintain good vision. The role of the eye care provider can be key in early identification and proper management.

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# Motion Sickness Symptomology and Occurrence among Visually-Impaired Adults

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## INTRODUCTION

Motion sickness (MS) can be caused by riding/reading on a form of transportation or be visually induced. MS can cause a variety of symptoms and affect health related quality of life. Assessment of MS may be difficult in VI adults due to travel/reading patterns, visual acuity and variable symptomology. VI adults were surveyed to assess MS occurrence/symptomology.

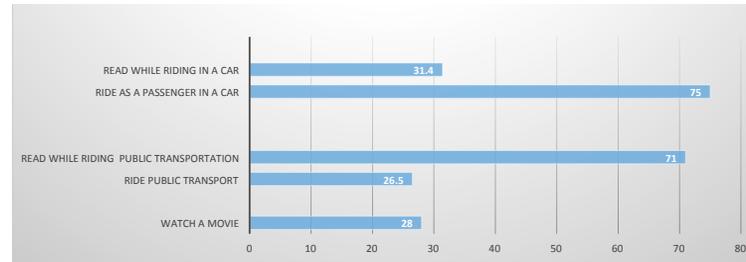
## METHODS

VI patients attending two urban Vision Rehabilitation clinics were surveyed. MS symptomology was assessed using the 'Motion Sickness Assessment Questionnaire'. In addition to occurrence of motion sickness, symptomology was also queried. Responses of gastrointestinal, central, peripheral, and sopite-related questions (See Table 1) were tabulated.

**TABLE 1**  
Subclassification of Motion Sickness Assessment Questionnaire statements

Gastrointestinal	Central	Peripheral	Sopite
I felt sick to my stomach	I felt faint-like	I felt sweaty	I felt annoyed/irritated
I felt queasy	I felt lightheaded	I felt clammy/cold sweat	I felt drowsy
I felt nauseated	I felt disorientated	I felt hot/warm	I felt tired/fatigued
I felt like I may vomit	I felt dizzy		I felt uneasy
	I felt like I was spinning		

**FIGURE 1** Activities Which Caused Motion Sickness



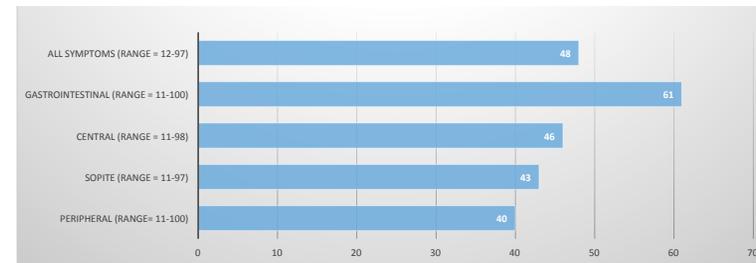
## RESULTS

MS was reported in 24.6% (N=73) of VI patients surveyed. The majority of those who reported MS were female (68.5%) Age categorization for those who reported MS was: 23.3% 18-39 years, 49.3 % 40-59 years and 27.4% 60 yrs and older. See Figure 1 for activities causing MS symptomology. Interestingly the majority of those who reported MS when riding in a car or on public transportation (bus or train) reported that they could not read while riding on the mode of transportation (59% and 56%) The majority reported they did not fly in an airplane or sail on a ship/boat (65% and 52.8%). See Figure 2 MS symptomology rating.

## CONCLUSIONS

There were a large number who reported MS. Many did not take forms of transportation or read on the car/bus/train. Most did not experience MS watching a movie. There is a variety in types and severity of MS symptomology. VI may play a role in these findings. Professionals/public familiarization with these findings as with MS symptoms is important so that MS can be diagnosed/treated and effect on health related quality of life limited.

**FIGURE 2** Mean Rating: Symptomology (Range= 0-100)



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# Comparison of Academic Performance Data in Chicago Public Schools (CPS) Students Gathered Prior to and After A Vision and Eye Health Exam

Sandra S. Block<sup>1</sup>; Melissa Suckow<sup>1</sup>; Adrianna M. Hempelmann<sup>1</sup>; Julia Gwynne<sup>2</sup>; Valerie Michelman<sup>2</sup>; Paul Moore<sup>2</sup>

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## BACKGROUND

The Illinois College of Optometry partnered with the Chicago Public Schools (CPS) to open the Illinois Eye Institute at Princeton Vision Clinic in January 2011. The clinic was opened in response to the lack of follow through for children who failed vision screenings and were referred and who were not seeking appropriate comprehensive vision care. CPS has developed a sophisticated system to ensure vision screenings of children at regular times in their academic lives, more specifically, they are screened at kindergarten, second and eighth grades, as well as, children who are considered at risk for a vision problem. This gap in ensuring that children received needed eye care led to the opening of the IEI at Princeton School-based Vision Clinic.

## PURPOSE

The purpose of this retrospective study was to compare standardized academic test scores, course grades and attendance in school for CPS students who attended the IEI at Princeton Vision Clinic during 2011 through 2013. Academic data from prior to the exam was compared to performance on standardized tests after the student received an eye exam was used. For analysis purposes, subjects were limited to those who were seen while they were in third grade or above since standardized test scores were unavailable for students in earlier grades.

## METHODS

The IEI at Princeton has comprehensive eye exam data on 14,663 CPS students who presented for eye during 2011 – 2013. The vision clinic serves primarily low-income students in schools in the geographic area surrounding the clinic which is located on the south side of Chicago in a low-income area that is predominantly Black and Hispanic. Children are brought by either their schools (bus) or with their parent or guardians (walk-in). The clinic provides comprehensive exams including refraction and cycloplegic/dilated exam.

The data important for this study included the following:  
 Previous History of Spectacle Correction  
 If they had reported wearing glasses before and were not wearing them currently – they reported whether the glasses were they lost, broken, at home or school  
 Entering Monocular Visual Acuity  
 Best Corrected Visual Acuity  
 Presence of Strabismus and type  
 Refractive Error – dry and cycloplegic

The Consortium on Chicago School Research (CCSR) has a Memorandum of Understanding with CPS allowing them access to longitudinal CPS administrative data from 2007 through 2014. For purposes of this study the focus was on reading and math standardized test scores as well as grade point average.

Student academic data was matched to individual vision data. Matching occurred through the subject's name and birthdate in order to link clinic patients to their unique identifier – i.e. student identification number. CCSR did the analysis and preliminary modeling using a student fixed effects model to estimate within-student change in outcome following attending the clinic to see if the vision intervention had any effect on student performance.

Prior to this analysis, a review of the subjects was made in which it was determined that the population of students seen at the IEI at Princeton Vision Clinic do not match the students within the entire district. Students who visited the clinic had test scores approximately 0.31 SD below the district average before visiting the clinic and average GPAs that were 0.24 SD below the district average. Any extrapolation beyond the subject pool cannot be made. The students seen at IEI at Princeton were more likely to be special needs patients and come from neighborhoods with higher concentrations of poverty than both the district averages and averages within their own schools. The lack of a suitable comparison group of non-patient students necessitated the student fixed-effects estimation strategy.

Please note that this stage of the analysis is on the aggregate data and not subgroups that clinically we would expect to benefit from a correction, ie, those with significant improvements in visual acuity or significant uncorrected refractive errors. This will be investigated in the next stage of analysis.

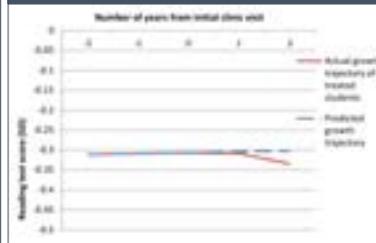
## RESULTS

It is felt that the estimates may be considered as causal if it is assumed that students who visit the clinic are not more likely to improve their performance concurrently with visiting the clinic for unrelated reasons. Preliminary models control for changes that may be associated with outcomes of interest and with attending the clinic (e.g., eligibility special ed. and school mobility). All standard errors are clustered by student. These models reveal that students who attended the clinic made increased relative gains in test scores and course grades in the year after attending the clinic.

Table 1. Summary of Results

	Standardized Reading Test Scores	Standardized Math Test Scores	Standardized Grade-Point Average
Main effect	-0.007 p = 0.334 (0.008)	0.025 p = 0.000 (0.007)	0.027 p = 0.006 (0.01)
Change in growth trend following visit	-0.021 p = 0.000 (0.005)	-0.023 p = 0.000 (0.005)	-0.029 p = 0.001 (0.009)
Students (N)	12,601	12,606	13,144

Figure 1. Average Effect of Clinic Visit on Reading Test Scores



No effect of visiting the clinic was seen on students' reading test scores (see Table 1).

Considering students' math test scores, there was a small and significant positive effect of visiting the clinic (0.025 SD).

There was a small and significant positive effect of visiting the clinic on students' GPAs (0.027 SD).

Figure 2. Average Effects of Clinic Visit on Math Test Scores

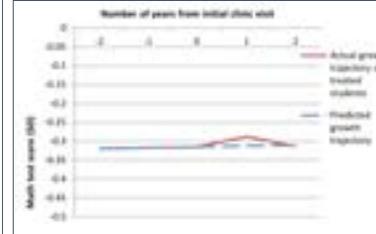


Figure 3. Average Effects of Clinic Visit on GPA



Table 1 represents the point estimate from Table 1 graphically in Figures 1, 2, and 3, 4. The level of the lines at one year before initial clinic visit is set as the average value observed in students the year before they visited the clinic.

The annual growth of students who would go on to visit the clinic may be seen by observing the slope of the line from two years before visiting the clinic to the year of visitation. The main effect of visiting the clinic can be seen by comparing the value of the red line to the blue line at one year after clinic visitation. A change in trend following clinic visitation may be seen by comparing the slope of the red line to the slope of the blue line between one and two years after visiting the clinic.

Analysis on attendance revealed that the rates aren't normally distributed like test scores and GPA, so even after fitting a Poisson distribution the estimates were unstable. Therefore additional analysis was not completed on attendance rates.

Note: The data for attendance could not be analyzed. N

## CONCLUSIONS

The small relative gains seen in math scores are preliminary and it is hoped that as the analysis looks in more depth to specific type and magnitude of refractive error, previous experience history of refractive correction, along with other visual diagnoses the effect will demonstrate more specifically the impact of vision corrections on academic performance in specific vision problems.

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# The Effect of Room Length on Perceived Egocentric Distance in Darkness

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## INTRODUCTION

In the dark the perceived egocentric distance of a self-illuminated target has been reported to be underestimated by a number of laboratories. This underestimation is of about the same magnitude whether perceived distance is measured using a verbal magnitude estimation task or a blind-walking motor task. In addition, self-illuminated targets are usually perceived as significantly elevated even though they are placed on the ground. This perceived elevation of the target increases with target distance.

The foreshortened distance and target elevation have been attributed to the presence of an implicit surface (Ooi, Wu and He, 2006) and to an over-estimation of angle of gaze (AoG) (Li and Durgin, 2012). There is empirical support for each of these proposals. In this study we have investigated the effect of the room dimensions on the perceived egocentric height and distance of a self-illuminated target viewed in darkness and found this variable to be significantly affected by room length.

## METHOD

Two groups of visually normal subjects were tested, all of whom had an eye exam within the past year. One group was tested in a room 3.8m wide and 8m long (n=20). The other group was tested in a room 11m wide and 9m long (n=15). Subjects clearly viewed the testing space prior to the testing session.

After obtaining informed consent, subjects were instructed to monocularly observe the perceived distance and height of a small, self-illuminated, red target placed on the floor of an otherwise dark room. The target was randomly located at 4 distances between 1.5 and 7.5m. When ready, subjects occluded their eye and blind-walked to the remembered target location. The walked distance indicated the perceived distance. The perceived height was indicated by instructing subjects to bend at the knees and gesture with their index finger the perceived height of the target. Subjects made two such estimates for each of the four target distances.

## RESULTS

A two-way analysis of variance (mixed design) was used to compare the perceived distances for the two testing spaces (IBM SPSS, V21.0). There was a significant effect of room size ( $F=13.01$ ,  $p=0.001$ ). See Figure 1. The interaction effect of Distance\*Room was also significant ( $F=7.48$ ,  $p<0.000$ ). Independent t tests with Bonferroni correction indicated that perceived target distances varied significantly between the two testing spaces except at 1.5m. Figure 2 plots the perceived height as a function of perceived distance. It can be seen that there is a significant effect of spatial layout on these two variables.

Figure 1: Perceived Distance vs Target Distance

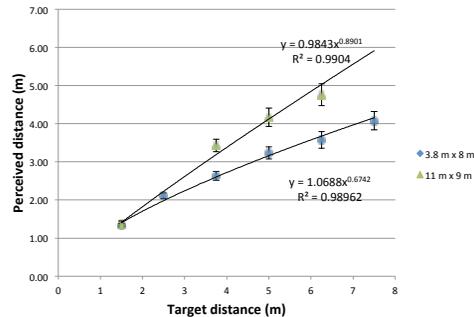
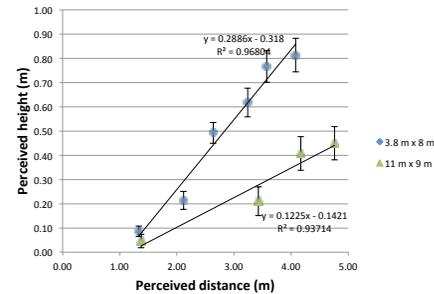


Figure 2: Perceived Height vs Perceived Distance



## CONCLUSIONS

1. The perceived distance is significantly shorter when measured in a shorter room than in a longer testing space.
2. The perceived height of self-illuminated target is significantly higher in a shorter room than in a longer testing space.
3. The exponent for perceived distance is smaller the shorter the testing space. This indicates that perceived distance is more compressed the shorter the room.
4. One possible interpretation of these results is that the slope of the implicit surface varies with the perceived dimensions of the room. In our study the slope was much greater in the shorter room than in the longer room.
5. We have also recently measured the Angle of Gaze to the test targets using a magnitude estimation procedure in each of the testing spaces described above. As predicted by Durgin et al, subjects did indeed overestimate the AoG to self-illuminated targets viewed in darkness, but the overestimations did not differ as a function of room size.
6. Our results support the hypothesis that the underestimation of distance and overestimation of target height may be due to a default implicit surface which can vary in slope depending on the dimensions of the testing space.

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# GPS usage in a Population of Low Vision Drivers

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## PURPOSE

- Low vision practitioners often recommend their patients drive with a passenger to assist with navigation. Often, this is not possible and global positioning systems (GPS) can serve as an additional navigator while driving.
- GPS have markedly improved since their development in 1985.
- Audio navigation instructions and street sign identification have been incorporated into stand-alone navigation units (i.e. Garmin or TomTom), smart phone applications, and built-in car navigation systems for the purpose of improving driver comfort and safety.
- We surveyed bioptic and non-bioptic Illinois low vision drivers to determine their GPS device usage.

## METHODS

- 27 low vision patients completed an IRB-approved phone survey
- Of the 27 low vision patients, 12 were bioptic drivers and 15 were non-bioptic drivers.
- Survey included questions of driving demographics (i.e. frequency of driving, motivation for driving) and GPS usage (i.e. type of device, helpful features, factors for not using)
- Patients were eligible to participate if they were 18 years of age or older, and met one of the following criteria:
  - best corrected visual acuity (BCVA) worse than 20/40
  - central or significant peripheral visual field defects
  - combination of above
- Participants were also required to be active drivers with an Illinois driver's license (Day and Night, Day Only restriction, or Bioptic restriction; as defined by the state of Illinois vision requirements for driving).

Subject #	Age	Diagnosis	BCVA Better Eye	Visual Field	Bioptic
1	91	ARMD	20/50	central scotoma	no
2	66	Myopic degeneration	20/100	full	yes
3	90	POAG, ARMD	20/150	central scotoma	no
4	58	Cerebral vascular accident (stroke)	20/20	hemifield	no
5	82	ARMD	20/100	central scotoma	yes
6	86	POAG, ARMD	20/50	central scotoma	no
7	84	POAG, ARMD	20/40	arcuate	yes
8	57	Ocular Albinism	20/80	full	yes
9	76	ARMD	20/80	central scotoma	yes
10	52	Retinitis Pigmentosa	20/40	arcuate/nasal step	no
11	76	ARMD	20/60	full	yes
12	57	ARMD, POAG, Myopic degeneration	20/30	central scotoma	no
13	88	POAG	20/40	arcuate	no
14	86	POAG, ARMD	20/40	central scotoma	no
15	26	Optic atrophy	20/60	arcuate/nasal step	yes
16	55	POAG, Myopic degen	20/50	central scotoma	no
17	62	AION	20/20	altitudinal defect	no
18	84	ARMD	20/70	full	no
19	74	Hereditary retinal	20/40	full	no
20	47	NC	20/60	gen constrict	no
21	78	POAG, Exudative ARMD	20/80	central scotoma	no
22	43	Ocular Albinism	20/50	gen constrict	yes
23	61	Bilateral AION	20/50	gen constrict	no
24	42	NC	20/60	full	yes
25	86	Central scotoma	20/80	gen constrict	yes
26	71	ARMD	20/80	full	yes
27	84	ARMD	20/70	full	no

POAG: Primary Open Angle Glaucoma • ARMD: Age-Related Macular Dystrophy  
NC: North Carolina Macular Dystrophy • AION: Anterior Ischemic Optic Neuropathy

Figure 1: Screen shots from "Google Maps" indicating appropriate lane choice before upcoming turns. 40% of patients using GPS to drive report smart phone as the modality of GPS used

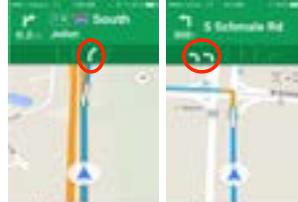


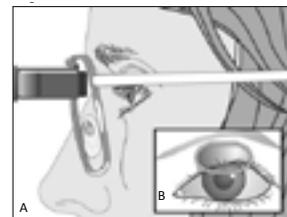
Figure 2: GARMIN GPS naviga7on device. 30% of patients using GPS to drive report a separate device, such as a GARMIN, as the modality of GPS used



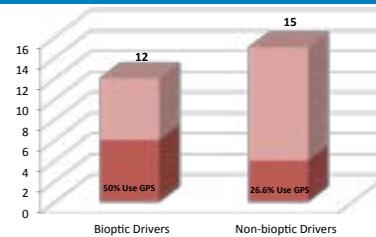
Figure 3: Built-in car GPS naviga7on system. 30% of patients using GPS to drive report this modality as the type of GPS used.



Figure 4: Schematic detailing proper bioptic positioning for viewing through the telescope(4a) and through the carrier lens (4b). Only 5-10% of time should be spent viewing through the telescope for distance spotting.



Graph 1: Number of Drivers Using GPS



Graph 2: Reasons for Not Using GPS to Drive



Graph 3: GPS Usage of Bioptic vs. Non-bioptic Drivers



## RESULTS

- 10 of the 27 patients (37%) report using a GPS while driving (graph 1)
- 6 of 12 (50%) bioptic drivers reported using GPS
- 4 of 15 (27%) non-bioptic drivers reported using GPS
- All 10 subjects who use GPS while driving report both of the following:
  - it improves comfort or safety level while driving
  - GPS is used specifically when driving to unfamiliar destinations rather than all the time or for night driving only.
- There is a positive correlation between those that own a GPS device (i.e. Garmin or TomTom) and those that use it to drive (Spearman rank correlation coefficient = 0.686, p<0.002); there is a less robust positive correlation between those that own a smartphone and those that use GPS to drive (0.394, p=0.04).
- The average age of drivers was 54.3 years for GPS users, and 77.6 years for non-GPS users.
  - Average age of bioptic drivers was 52.5 years for GPS users and 80.6 years for non-GPS users.
  - Average age of non-bioptic drivers was 57 years for GPS users and 75.9 years for non-GPS users.

## CONCLUSIONS

- The results of the study suggest that older low vision patients are less likely to use GPS while driving as compared to younger patients.
- Most useful GPS qualities included which lane to stay in before preparing for a turn and audio stating each passing street name.
- Given that 100% of those who use GPS while driving report increased comfort and safety level, low vision practitioners should educate patients on the use of GPS as an additional navigator while driving.
- Investigations are needed to analyze further enhancement and accessibility features that may benefit a low vision driver using GPS as an aid.

## SPECIAL THANKS



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# Factors Predicting King-Devick Test Performance in Adults and Adolescents

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## INTRODUCTION

The King-Devick(K-D) test (Figure 1), a <2-minute timed assessment of rapid number naming, has been studied as a rapid, quantitative screening tool for neurological dysfunction associated with concussion, hypoxia, Parkinson's disease, multiple sclerosis and extreme sleep deprivation. We have reported the K-D test performance in normal adults and adolescents in our previous study.<sup>1</sup> However, potentially confounding variables have not been studied in a large sample. The purpose of this study was to determine important confounding variables that are associated with K-D test performance.

## METHODS

In this cross-sectional, multi-center study, subjects ≥15yrs old with binocular near visual acuity better than 20/30 completed two trials of the K-D test protocol. Exclusion criteria included concussion within 3-months, post-concussion syndrome, dyslexia or neuro-degenerative disorders. History of concussion, amblyopia, strabismus as well as demographic variables of education, race/ethnicity, gender and age were assessed by subject interview. Multiple linear regression analysis was performed. Independent variables were modeled as categorical (age (< or > 40 years), race/ethnicity, gender, education, concussion, amblyopia, strabismus) and continuous (age in years greater than 40 years) terms. Education levels included high school graduate, some college training, college graduate, and post-college graduate.

Figure 1. King-Devick (K-D) Test Cards

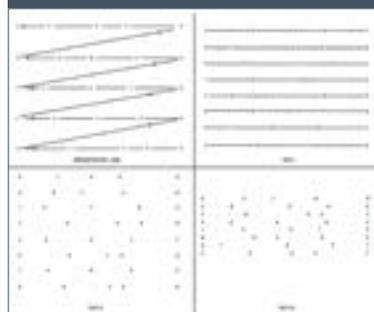


Table 1. Multiple Regression Analysis for Factors Predicting the K-D Test Time

	Standardized Coefficient	T	P-Value
Age	0.45	11.41	<0.001*
Education	-0.32	-7.95	<0.001*
African-American race	-0.12	-3.56	<0.001*
Hispanic race	0.07	2.09	0.04*
White race	0.03	0.28	0.78
Asian race	0.02	0.31	0.76
History of concussion	-0.01	-0.19	0.85
Gender	0.00	-0.01	1.00
Amblyopia	0.04	0.97	0.33
Strabismus	-0.01	-0.19	0.85

Figure 2. K-D Time in Different Age Groups

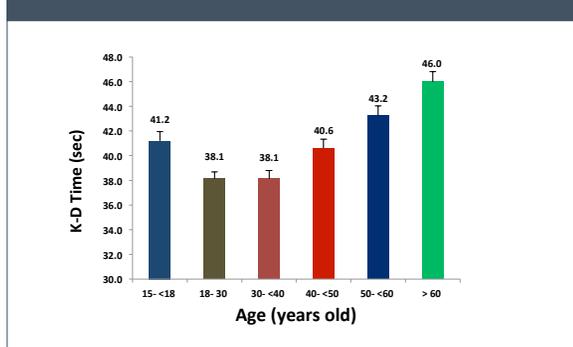


Figure 3. K-D Time in Different Races

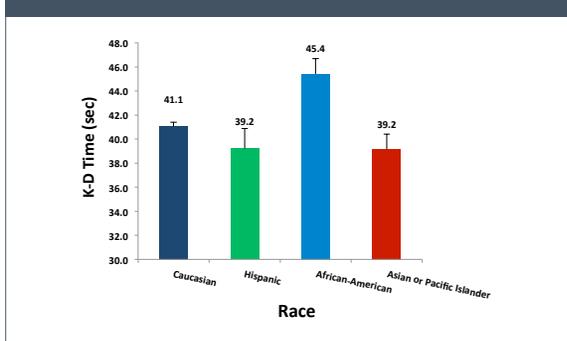
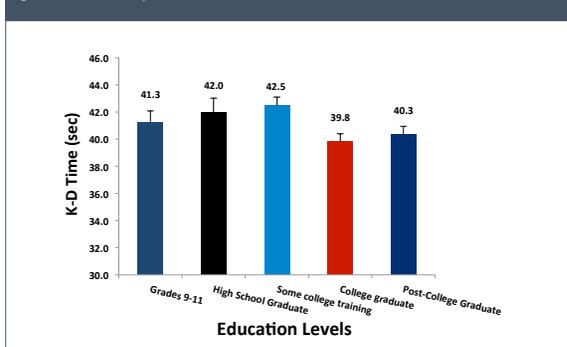


Figure 4. K-D Time in Subjects with Different Education Levels



## RESULTS

Subjects (n=691, age 39.8±17.7 years) were enrolled across 5 sites. The average best K-D time was 41.2±8.2s. Table 1 shows the results of multiple regression analysis. The final multiple regression model included age beyond 40 years, education, African-American race and Hispanic race. With other variables held constant, the K-D test time worsened by 0.27s for each year in age above 40, improved by 1.94s for each category of higher education level achieved, worsened by 3.67s for African-American race and improved by 2.38s for Hispanic ethnicity. Gender, White race, Asian race, amblyopia, strabismus, or history of concussion was not associated with K-D test performance.

## CONCLUSION

- Older than age of 40 years and African-American race were associated with poorer K-D test performance.
- Higher education and Hispanic ethnicity were associated with better K-D test performance.
- Gender, White race, Asian race, amblyopia, strabismus, or history of concussion had no impact on the K-D test performance.
- Knowledge of these confounding variables is important for applications of K-D test in neurologically diseased populations and its development as a clinical measurement tool.

## ACKNOWLEDGEMENT

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# Analysis of Health Associations with Long Anterior Lens Zonules

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## BACKGROUND

Long anterior zonules (LAZ) are characterized by zonular fibers that extend more central than usual on the anterior lens capsule (Fig. 1).<sup>1,8-12</sup> One variety occurs with genetic mutation and late-onset macular degeneration,<sup>8,9,10</sup> and another type with unknown etiology is associated with age >50 years, female gender, hyperopia, shorter axial length, and remnants of the tunica vasculosa lentis.<sup>11,16</sup> LAZ can cause a unique type of pigment dispersion and are being studied as a potential risk factor for open and narrow angle glaucoma.<sup>13,14,15,17</sup> There is also interest in LAZ relative to cataract surgery. The purpose of this study was to assess potential health associations with LAZ.

## METHODS

Patients presenting for existing scheduled appointments at an urban academic eye care facility in Chicago, IL were examined for LAZ if they were dilated, and questionnaires were administered to help collect information about health and lifestyle including education level, hypertension, diabetes, use of cholesterol lowering medications, height/weight, alcohol use, smoking, second hand smoke, and drug use. Regression analyses were conducted to evaluate potential associations with LAZ. Backward, forward, and stepwise elimination methods were used to identify variables for final models. Variables were included in final models if significant levels were <0.10. Only right eyes were included in the analysis, and subjects with <5 LAZ (trace) in both eyes were excluded to ensure definitive cases. LAZ were defined as zonule fibers present approximately >1.0 mm central to the normal zonule insertion zone.

## RESULTS

After exclusions, the total number of subjects in the final analysis was 1,826 (63.4% female, 82.6% African-American) with a mean age=54.2 +/- 16.3 years (18-95 years). Included were 66 LAZ subjects (55 females, 11 males). Categorical and continuous variables that were assessed for a potential relationship with LAZ are summarized in Table 1. Although a number of variables showed relationship with LAZ presence via the bivariate analyses, most of the relationships did not persist with simultaneous control for age, race, and gender. Final models are illustrated in Tables 2 and 3. While controlling for age (in decades) (OR=1.7, P<0.0001) female gender (OR=2.7, P=0.006), African-American vs. other race (OR=3.8, P=0.07), and hypertension (OR=1.8, p=0.09), associations persisted with ocular parameters including hyperopic refractive error (in diopters) (OR=1.2, P=0.005) and presence of a pupillary membrane remnant (OR=3.4, P<0.0001). Associations were not detected with the other non-ocular variables studied except for marginal association with hypertension (OR=1.7, P=0.09).

## DISCUSSION

Similar to previous investigations, this dataset also showed associations between LAZ and age, female gender, refractive error, and persistent pupillary membrane iris strands. Association has not previously been demonstrated between African-American race and LAZ,<sup>13</sup> but this potential relationship should be interpreted with caution because there were so few non-African-American LAZ subjects included. Due to the population from which the subjects were derived, race association has also been minimally studied. An association with hypertension has not previously been detected, although it has been studied.<sup>12,13</sup> Thus, due to this, as well as the marginal p-value, we also interpret this potential relationship with caution. Of interest is the fact that no other relationships were found between LAZ and the variables studied. Although not an exhaustive study of potential health relationships, this information is important toward further understanding of the nature of the LAZ trait.

Table 1: UNIVARIATE ASSOCIATIONS WITH LAZ

Variable	LAZ Subjects N=66	Controls N=1,760	P-value
Gender - Female	83%	63%	**0.001
Race			
African-American	97%	82%	**0.007
Other	3%	18%	
Education - College degree (Yes)	30%	31%	0.90
Alcohol use			
Current	41%	48%	
Past	16%	12%	0.31
Never	43%	40%	
Smoking			
Current	24%	33%	
Past	30%	18%	0.60
Never	46%	49%	
Diabetes (Yes)	35%	21%	**0.009
Hypertension (Yes)	79%	49%	**<0.0001
Cholesterol medication (Yes)	48%	29%	**0.001
Drug use (Yes)	6%	13%	0.12
Asthma (Yes)	6%	9%	0.39
Lung disease - any (Yes)	14%	13%	0.85
Heart disease - any (Yes)	15%	6%	**0.002
Cancer - any (Yes)	5%	4%	0.78
Persistent pupil iris strands (Yes)	30%	15%	**0.001
	Mean +/- SD	Mean +/- SD	
Age, years	64.5 +/- 11.9	51.5 +/- 15.4	**<0.0001
Refractive error (SE), D	+0.70 +/- 2.0	-0.86 +/- 3.1	**<0.0001
BMI	31.3 +/- 8.0	30.4 +/- 7.8	0.34

Abbreviations: D, diopters; SE, spherical equivalent; LAZ, long anterior zonules; SD, standard deviation

Figure 1: Normal zonule insertion (top-left), LAZ in retroillumination (top-middle and bottom), and pigmented LAZ in direct illumination (top-right).

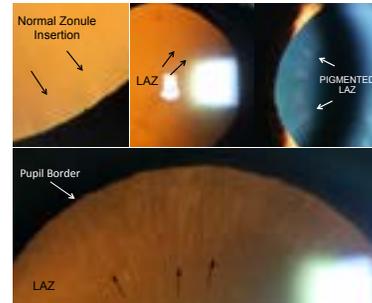


Table 2: MULTIVARIATE RELATIONSHIPS WITH LAZ\*

Variable	Coefficient	Standard Error	P-value	Odds Ratio (95% CI)
Intercept	-9.93	1.14		
Age, in decades	0.52	0.11	<0.0001	1.7 (1.3 to 2.1)
African-American race	1.34	0.74	0.07	3.8 (0.9 to 16.3)
Female gender	0.98	0.36	0.006	2.7 (1.3 to 5.4)
Hypertension	0.58	0.34	0.09	1.8 (0.9 to 3.5)
Refractive error (SE), D	0.19	0.07	0.005	1.2 (1.1 to 1.4)
Pupillary iris strands	1.23	0.31	<0.0001	3.4 (1.9 to 6.2)

\*Abbreviations: CI, confidence interval; D, diopters; SE, spherical equivalent; LAZ, long anterior zonules;

Table 3: MULTIVARIATE RELATIONSHIPS WITH LAZ\* WITHOUT OCULAR VARIABLES

Variable	Coefficient	Standard Error	P value	Odds Ratio (95% CI)
Intercept	-9.43	1.06		
Age, in decades	0.51	0.10	<0.0001	1.7 (1.4 to 2.0)
African-American race	1.33	0.72	0.07	3.8 (0.9 to 15.8)
Female gender	0.93	0.34	0.006	2.5 (1.3 to 4.9)
Hypertension	0.54	0.33	0.10	1.7 (0.9 to 3.7)

\*Abbreviations: CI, confidence interval; D, diopters; SE, spherical equivalent; LAZ, long anterior zonules;

## CONCLUSIONS

Although this analysis finds consistent associations between LAZ and age, female gender, hyperopia, and persistent pupillary membrane iris strands, definitive associations were not detected with general health parameters that were studied.

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# Comparison of Baseline Visual Function for Patients in VA Low Vision Intervention Trials I and II

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Robert W. Massof, PhD, Baltimore, Maryland  
and LOVIT II Study Group

## BACKGROUND

- Two RCTs were conducted to evaluate the outcomes of outpatient low vision service delivery provided by the Department of Veterans Affairs.
- LOVIT I studied severely visually impaired veterans with macular diseases; habitual visual acuity (better seeing eye) <20/100 and >20/500, mean 1.1 log MAR SD (0.2)
- LOVIT II studied veterans with macular disease and less severe visual impairments; best corrected visual acuity <20/50 and >20/100, mean 0.6 log MAR SD (0.2).

## PURPOSE

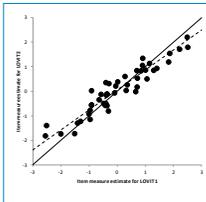
- To determine if the LOVIT II participants have greater visual function at baseline than the LOVIT I participants.

## ANALYSES

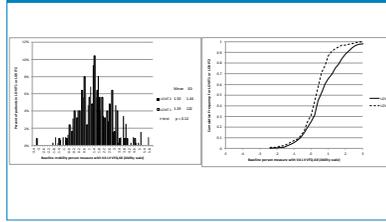
- Performed Rasch analysis with Andrich rating scale model on VA LV VFQ-48 responses at baseline for LOVIT I and LOVIT II participants combined.
- Looked for evidence of study-related DIF (different groups with the same ability have a different probability of giving a certain response on a questionnaire).
- Compared visual ability measures for the different functional domains.

## STUDY-SPECIFIC DIF

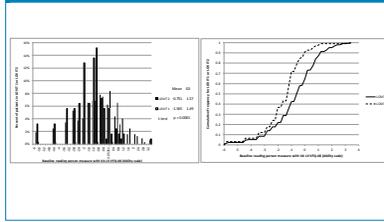
- There is some minor study-specific DIF (departure of points from solid identity line).
- The DIF will become insignificant if a shallower slope line is fit (dashed line). Shallower line would be interpreted as higher intrinsic noise in the LOVIT II participants.
- Conclusion: no meaningful study-specific DIF



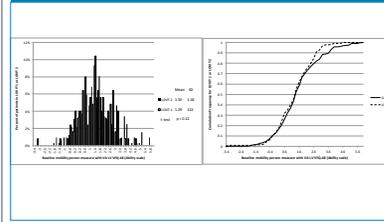
Person measures estimated from all 48 items in the VA LV VFQ-48 at baseline



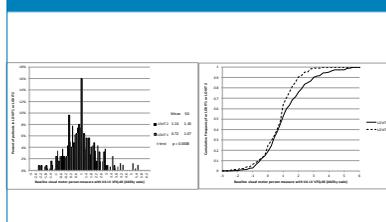
Person measures estimated from VA LV VFQ-48 reading items



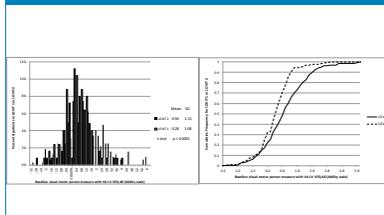
Person measures estimated from VA LV VFQ-48 mobility items



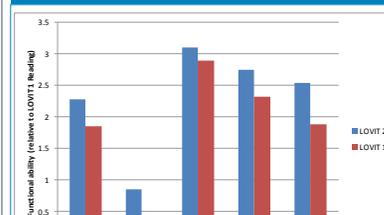
Person measures estimated from VA LV VFQ-48 visual motor items



Person measures estimated from VA LV VFQ-48 visual information items



Summary of comparisons of LOVIT 2 to LOVIT 1 baseline functional ability



## COMPARISON

Comparison of visual ability measures at baseline for the different functional domains

- Overlaid histograms of person measures estimated from VA LV VFQ-48 difficulty ratings at baseline for LOVIT I and LOVIT II patients.
- Inset lists means and standard deviations of the 2 person measure distributions and the p value for the t-test.
- Higher numbers indicate more visual ability.
- Same comparison of LOVIT I and LOVIT II baseline person measure distributions as in the left panel, but illustrated as cumulative distributions.
- Slope of function reflects the standard deviation (small SDs have steeper slopes) and separation of functions corresponds to differences between the means.

## CONCLUSION

- LOVIT II participants have significantly more visual function at baseline (less difficulty performing activities) in all areas except mobility than patients in LOVIT I.

## FINANCIAL SUPPORT

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# Comparison of M&S smart system II single letter contrast test and iPad letter test results with Pelli-Robson in low vision patients

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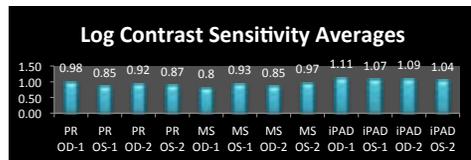
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## PURPOSE

The purpose of our study is to measure the reliability of two new tests that measure the contrast sensitivity (CS); the M&S smart system II single letter contrast test and the iPad letter test. These results were compared with those obtained from the Pelli-Robson (PR) test which is well-established and clinically accepted but no longer in production. In addition we also asked subjects if amber/yellow lenses improve their subjective perception of text.

## METHODS

24 low vision patients diagnosed with either myopic degeneration or advanced glaucoma were selected from the Illinois Eye Institute. Inclusion criteria included 20/70 VA or worse in the better-seeing eye or a visual field of 20 degrees or less in the better-seeing eye. Patients were examined at two separate visits 1 month apart (+/- 1 week). During each visit, patient CS was measured using three different tests: iPad version of PR, M&S contrast sensitivity and PR.



### M&S Smart System II



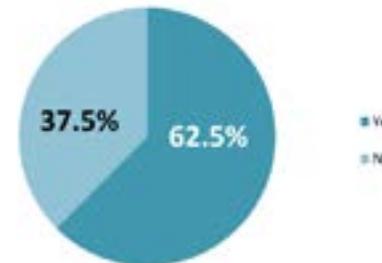
### iPad Letter Test



### Single Letter Contrast Test



### Filter Preferred



## RESULTS

A two-way repeated measures analysis of variance (ANOVA) reveals that the absolute CS values are lowest when obtained from the M&S test but the CS values do not differ significantly between the three tests ( $F=2.96, p=0.06$ ). The same ANOVA analysis indicated that there is no difference in the CS of visit 1 versus visit 2 for any of the three CS tests ( $F= 0.81, p=0.38$ ). Reliability was assessed with the intraclass correlation coefficient (ICC). The ICC values for the 3 tests are: PR=0.82, MS=0.91 and IPAD=0.93 ( $p=0.001$ ). We also found that low vision patients subjectively prefer to have their specs tinted; we did not find a preference for yellow or amber tint but in general any sort of tint was preferred.

## CONCLUSIONS

Two new CS tests; the M&S smart system II single letter contrast test and the iPad letter test, were compared with the well-established PR test in a low vision sample population. Our results indicate that the CS obtained on the first visit was statistically the same as that obtained from the second visit and that the absolute CS values do not differ significantly between the tests. We also found that the reliability of the two new tests was very similar to that of the PR test, which is the gold standard in terms of CS tests. We conclude that these two new tests offer excellent reliability when used to test CS in this low vision sample. We also found that 62.5% of the tested low vision patients subjectively prefer to use yellow (60%) or amber (40%) tinted specs.

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# Establishing a Model to Investigate Anterior Epithelial Cell Division in Whole Pig Lenses

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**Background:** Presbyopia is a progressive loss of accommodation that occurs naturally in people starting around the age of 40. It is one of the most common conditions encountered by optometrists and yet the only treatment is compensatory, rather than directly addressing biological changes in the eye. While the exact etiology is not fully understood, the predominant theory implicates the gradual stiffening of the intraocular lens—rather than ciliary muscle degeneration—as the reason for the decreased ability to stimulate accommodation effectively with age.

Lens stiffness most likely originates from physical changes produced by the germinative zone lens epithelium (GZ LE), which is the only site of active cell division in the mammalian lens. It is here that lens fibers are constantly added to the adult lens, ultimately leading to changes in lens morphology, zonular insertion, and capsule rigidity. Because the germinative zone plays an important role in lens development and presumably presbyopia too, it is an obvious target for studying lens growth and possible manipulation.

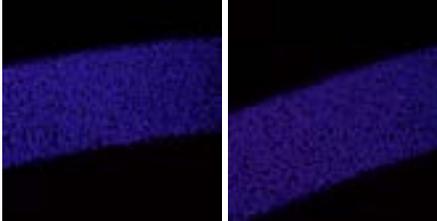
**Purpose:** The goal of this study is to establish a reliable method to determine the specific location and mitotic rate of germinative zone lens epithelium in whole pig lenses. To isolate actively dividing epithelial cells, intact lenses are exposed to 5-ethynyl-2'-deoxyuridine (EdU), which is a DNA precursor analogue that is incorporated into a growing DNA strand during the S phase of the cell cycle. Cells that take up EdU are visualized using click chemistry, a copper-catalyzed chemical reaction that covalently binds an alkyne (EdU) with an azide attached to a fluorescent dye (Alexa 594 azide). With a model to measure GZ LE cell division in healthy tissue, it may then be possible to study physiologic responses to laser ablation as a potential treatment for presbyopia.

**Methods:** Fresh pig eyes are obtained from a local abattoir and dissected to extract whole lenses. Lenses are kept warm and bathed in 3mg/mL chymotrypsin to remove excess ciliary body and then exposed to 0.6mM EdU for 2 hours. Negative controls are concurrently performed without EdU to assess specificity of the technique. All samples are then fixed, washed, and permeabilized, followed by incubation with a fluorescent marker (Alexa 594) to visualize EdU uptake. Lenses are then counterstained with a nonspecific DNA marker (Hoechst 33342) to define cell nuclei as landmarks for co-localization with mitotic cells that have incorporated EdU. Whole lenses are secured to a stage so that the anterior lens surface is flat and pictures are taken in each lens quadrant using a Nikon AR1 multi-photon microscope.

Cell quantification is completed using Fiji imaging software and is based upon careful selection of those cells that demonstrate spatial co-localization of both the red (Alexa 594 representing EdU uptake) and blue (Hoechst DNA marker) emission signals. To set up images for analysis, red and blue signals were merged into one overlay channel and run under the program function "Z projection" to stack cells from many consecutive planes into a single two-dimensional image that represents the three-dimensional dataset of the region. Any cells that demonstrate co-localization by mere observation are manually scanned using a measurement tool to record peak signal intensity. In early trials, cells that emit at least 30% more red light than local red background staining are consistently detected by the naked eye during image analysis. This observed value now serves as the threshold criterion to distinguish those cells that show significant fluorescence—and therefore EdU uptake—above background noise. An objective criterion is necessary to minimize selection bias and variability across different lens samples. Therefore, cells are deemed positive for active mitosis only if they meet this threshold; that is, if their maximum point intensity emits at least 30% more light in the red channel (i.e. Alexa-EdU) than that of an adjacent cell with a maximum intensity representative of the target region (i.e. background staining).

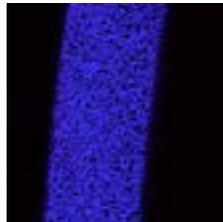
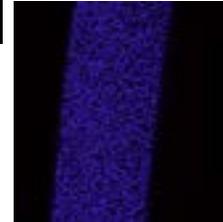
**Results:** The germinative zone of the adult pig lens is approximately 500 microns from the lens equator in a band that is roughly 500 microns wide. All experimental lenses show staining (see table 1). The range in percentage of dividing cells per quadrant is 0 – 1.10%. The average percentage across lenses is 0.33±0.08 (SEM), while for the control lenses the range is 0-0.01%.

**Figure 4 (left and right): Lens 1, Quadrant 4**  
Both show staining, with the total sample yielding 0.85% of cells that pass the threshold criterion and are counted as positively co-localized cells.

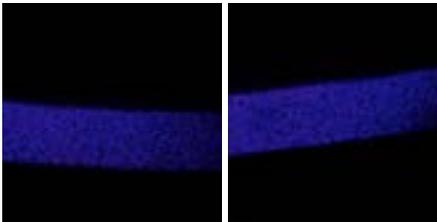


**Figure 1 (top and bottom): Lens 1, Quadrant 1**  
(\*) EdU, (\*) Alexa 594, (\*) Hoechst 33342

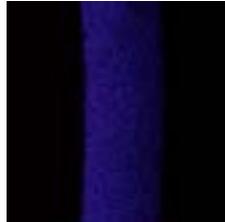
Each image is a three-dimensional composite depicting 31 adjacent slices of cells stacked together for analysis. Positive co-localization appears purple; the summation of red and blue signals over the same focal cell area. Figure 1 (top) does not have any staining that meets the threshold criterion, whereas Figure 1 (bottom) has staining most notably inferior in the image.



**Figure 5: Control Lens 1**  
(\*) EdU, (\*) Alexa 594, (\*) Hoechst 33342 – Control lenses underwent the exact same conditions as experimental lenses, except that EdU is excluded from treatment. Red signal is uniformly dim throughout the region to confirm specificity.



**Figure 2 (left and right): Lens 1, Quadrant 2**  
Despite the appearance of positive co-localization in the upper region of Figure 2 (right), there are no cells in this entire quadrant (left nor right) that are considered positive for staining based on the 30% threshold criterion.



**Figure 3 (top and bottom): Lens 1, Quadrant 3**  
(top) There is no significant staining in this area of the quadrant; however, the adjacent area in (bottom) demonstrates significant staining. Taken together, the entire quadrant shows 1.10% staining. Notice the minor disruption in tissue integrity towards the top of Figure 3 bottom.



**Figure 6A: ICTN plugin – Fiji** includes a program that facilitates cell quantification analysis. It relies on parameters taken from a sample cell to define and record the amount of cells in a given region of interest. In this particular image, it measures 1111 cells in a 124,548.43 square micron area.

Quadrant	# of Dividing Cells (%) by Location				
	Q1	Q2	Q3	Q4	Global
Lens 1	0.46	0	1.10	0.85	0.60
Lens 2	0.96	0.35	0	0	0.13
Lens 3	0.17	0.43	0.48	0.91	0.49
Lens 4	0.03	0	0	0.55	0.13
Lens 5	0.69	0	0.34	0	0.28
Lens 6 (control)	0	0.05	0	0	0.01

Table 1: Cell Quantification – EdU and Alexa Interaction

**Conclusions:** This method provides a way to establish the germinative zone's location and size in a whole pig lens. As can be seen from the data per quadrant, dividing cells are not equally distributed within the germinative zone. The observed staining pattern may support a concept of spiral or bidirectional lens fiber development.

The protocol requires additional adjustments to improve the signal-to-noise ratio and maximize visualization quality and yields of desired staining. It is important to recognize the possibility that active cell division drops to a level so low in vitro and post-mortem that the sensitivity of this model to detect mitosis may have reached a limit.

**Future Directions:** Given that changes to the concentrations of EdU and Alexa no longer increase staining, the next step is to promote greater mitotic rates by enhancing EdU uptake and preserving the natural lens environment. Instead of extracting lenses from the globe prior to treatment, EdU could be injected into the eye with the lenses undisturbed. In addition to preserving the physiological environment, this new approach may reveal more about patterns of growth because it maintains the proper orientation that is otherwise lost once lenses are removed from the eye.

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# Expression of Cx46fs380 Causes Significant Changes in Lens Fiber Membrane Structural Organization

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## Purpose:

Inherited human cataracts have been linked to mutations in connexin46 (Cx46) including Cx46fs380. Mice expressing Cx46fs380 (generated by a knock-in strategy) show nuclear cataracts that are evident at 2 months in homozygotes and at ≥4 months in heterozygotes. These animals have substantially reduced levels of fiber cell connexins before cataracts become detectable. The experiments were designed to characterize the structural changes in the organization of lens fiber cell membranes that occur in these mice with age.

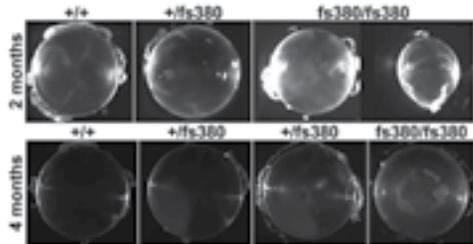
## Methods:

Lenses from the different genotypes at 2 and 4 months of age were photographed using dark-field illumination. Additional lenses from young (≤3 months) and older (>3 months) wild type (+/+), young (n=9, older: n=6) heterozygous (+/fs380), young (n=5, older: n=6) and homozygous (fs380/fs380), young: n=10, older: n=5) Cx46fs380 mice were examined by scanning electron microscopy (Hitachi S-3000N). Both lenses from each animal were fixed and dissected into approximately 250 μm thick peels to allow examination of nuclear, inner, and outer cortical regions. Broad faces of fiber cells were classified by organization of interdigitations, ball and socket joints, and furrows. Data are presented as the percentage of fiber cells showing organization of these features. Statistical significance was determined using nonparametric t-tests using SPSS comparing percentage. A Kruskal-Wallis test was used to compare the three genotypes, while a Mann-Whitney test was used for all post-hoc analyses as well as the age comparisons.

## Results:

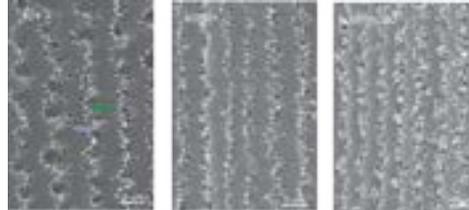
**Expression of Cx46fs380 causes cataracts, evident at 2 months in homozygotes and at ≥4 months in heterozygotes.**

**Figure 1:** Dark-field photomicrographs of lenses from the different genotypes at 2 and 4 months of age

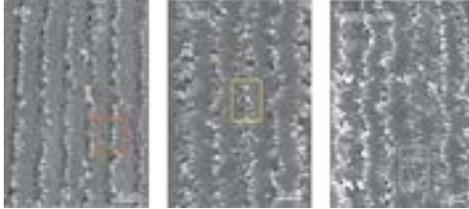


The percentages of fiber cells with organized ball and socket joints were reduced in both young and old heterozygous and homozygous Cx46fs380 mice. Organized ball and socket joints decreased with age in heterozygous Cx46fs380 mice.

**Figure 2:** Fiber cells showing ball and socket joints in young mice. The green arrow points to a ball and the purple arrow point to a socket in a wild type lens



**Figure 3:** Fiber cells showing ball and socket joints in older mice. The red box encloses linearly organized ball and socket joints in a wild type lens. The yellow and blue boxes enclose the ball and socket joints from heterozygotes and homozygotes that do not follow a specific pattern of organization



**Table 1:** Quantification of organization of ball and socket joints in lens fibers from young and old animals of the different genotypes

Age	Ball & Socket Joints (%)	+/+	+/fs380	fs380/fs380	Overall p value
Young	Unorganized	26.9	36.4	87.6 <sup>f</sup>	0.001
	Organized	73.1	63.6	12.7 <sup>f</sup>	
Older	Unorganized	21.7*	66.3*	70.2**	0.001
	Organized	78.3*	33.7**	29.8**	

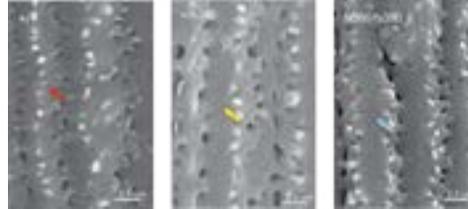
\* = p<0.05 as compared to wild type in the same age group

<sup>f</sup> = p<0.05 as compared to wild type and heterozygotes in the same age group

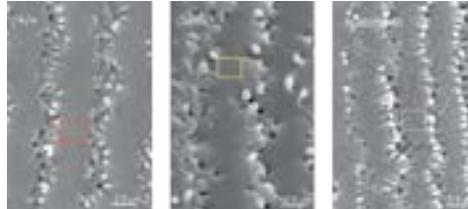
\*\* = p<0.05 as compared to the young group in the same genotype

The percentages of fiber cells showing a consistent pattern of furrows were reduced in both young and old heterozygous and homozygous Cx46fs380 mice.

**Figure 4:** Fiber cells showing furrows in young mice. The red arrow points to furrows forming a uniform linear pattern (linear pairs of membrane folds) in a wild type lens. The yellow and blue arrows point to disordered membrane folds in heterozygous and homozygous Cx46fs380 lenses



**Figure 5:** Fiber cells showing furrows in older mice. The red box encloses furrows in a linear pattern in an older wild type lens. Yellow and blue boxes enclose regions with irregular membrane folds in heterozygous and homozygous Cx46fs380 lenses



**Table 2:** Quantification of organization of furrows in lens fibers from young and old animals of the different genotypes

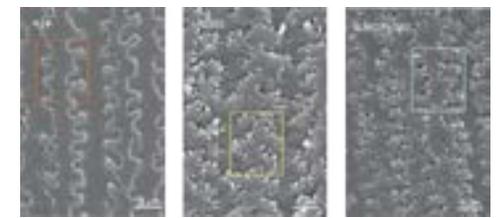
Age	Furrows (%)	+/+	+/fs380	fs380/fs380	Overall p value
Young	No organization	7.7	22.7	23.6*	0.001
	Partially organized	3.8	9.1	0*	
	Random linear arrangement	3.8	9.1	0*	
	Uniform linear arrangement	15.4	0	0*	
	None present	69.2	59.1	76.4*	
Older	No organization	2.9	26.7*	26.3**	0.001
	Partially organized	20.3	19.8*	17.5**	
	Random linear arrangement	37.7	0*	7.0**	
	Uniform linear arrangement	17.4	0*	0**	
	None present	21.7	53.5*	49.1**	

\* = p<0.05 as compared to wild type in the same age group

\*\* = p<0.05 as compared to the young group in the same genotype

The percentages of fiber cells with organized interdigitations were significantly decreased in older heterozygous and homozygous Cx46fs380 mice.

**Figure 6:** Fiber cells showing interdigitations in fiber cells from older mice. The red box shows the smooth regular pattern of interdigitations seen in wild type compared with the disorganization of this pattern in heterozygotes and homozygotes (yellow and blue boxes)



**Table 3:** Quantification of organization of interdigitations in lens fibers from young and old animals of the different genotypes

Age	Interdigitations (%)	+/+	+/fs380	fs380/fs380	Overall p value
Young	Unorganized	0	9.1	5.5	0.111
	Organized	15.4	9.1	1.8	
	None present	84.6	81.8	92.7	
Older	Unorganized	4.3	26.7*	38.6*	0.001
	Organized	30.4	18.6*	19.3*	
	None present	65.2	54.7*	42.1*	

\* = p<0.05 as compared to wild type in the same age group

## Conclusions:

The organization of fiber cell membrane structure in lenses from mice expressing Cx46fs380 show significant changes compared with that of wild type lenses. These changes may be caused by the near absence of Cx46 and reduction of Cx50 in Cx46fs380 lenses.

Because some biochemical and membrane structural alterations precede the appearance of opacities, they likely contribute to cataract formation.

## Future Directions:

We plan to study the relationship between the structural and biochemical changes as well as the impact of the structural changes on the optical quality of the lens.

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ICO

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## 2 ICO PRESENTATIONS

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# Does Pachymetry Correlate with Corneal Steepness, Vision and Sequela in Patients with Keratoconus?

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## PURPOSE

While we know that keratoconus is a multifactorial disease that is characterized by corneal ectasia and steepening, it is unclear as to the correlation between these two. Is a steep cornea always a thin cornea? Conversely, can a flatter and seemingly milder keratoconic cornea be significantly thinned? Therefore, we were interested in determining if steeper keratometric readings correlated with thinner corneas. This was of particular interest to us as our patients of the IEI tend to have steeper corneas than estimated by the Collaborative Longitudinal Evaluation of Keratoconus with less than 12 percent of patients being classified as mild and 39 percent as severe. This data was presented at BCLA 2011.

We hypothesized that our patient population would have significantly thin corneas and that this would indeed correlate with their K readings. We also hypothesized that thinner corneas would have worse acuity with spectacles and contact lenses and greater prevalence of corneal scarring and staining.

## METHODS

A study of the keratoconic population of the Illinois Eye Institute was conducted. We reviewed over 600 charts. Ultimately those patients that had been evaluated with the Orbscan2 were included in this evaluation. This study was also limited to patients wearing corneal GP lenses.

The charts were pulled and data was collected regarding their lens designs, vision, and topographical indices. Some 121 patients were evaluated with the orbscan with a total of 217 eyes. Some patients had failed scans for one eye.

### Data collected included

Age (12-64 years, mean 33 std 12)

Gender (Figure 1)

Race (Figure 2)

Spectacle acuity

Contact lens acuity

Flat k

Steep k

Minimum pachymetry

Presence/absence of corneal staining

Presence/absence of corneal scarring

Statistical evaluation included descriptive statistics as well as t-test and pearson correlation. The non parametric data was evaluated with a spearman's rho.

FIGURE 1: Gender Distribution

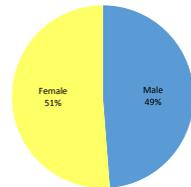
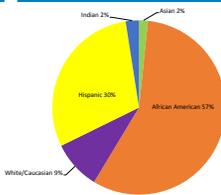


FIGURE 2: Racial Distribution of Keratoconic Population



## RESULTS

Pachymetry readings ranged from 108 to 583microns with an average of 393.4 ±76.6microns. Flat Ks averaged 49.15±6.08D and steep Ks were 54.50±7.63D. Pachymetry was moderately correlated with keratometric readings (p<0.0001, r=-0.55) for flatK and r=-0.46 for steepK. (Figures 3 and 4) For the 175 eyes for which spectacle acuity had been recorded there was mild correlation with pachymetry (r=-0.19 p=0.01). Contact lens acuity and pachymetry were not correlated (r=-0.07, p=0.32). Corneal scarring (scar r=-0.28, p<0.001) and staining (stain r=-0.20, p=.001) occurred more frequently in thinner corneas. Corneal staining occurred in only 29 percent of corneas thicker than 400 microns, but for those thinner than 400 staining occurred at a rate of 43 percent. Similarly staining also occurred more often in thinner corneas). For corneas greater than 400 the rate was only 20 percent, under 400 the rate increased to 39 percent. For the extremely thin cornea (<300microns), the rate increased to 59.5 percent. Correlations are summarized in Figure 5.

FIGURE 3: Correlation between minimum corneal thickness and flat K readings

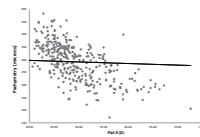


FIGURE 4: Correlation between minimum thickness and steep K readings

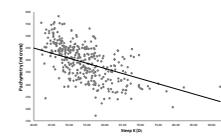


FIGURE 4: Correlation summarization

Pearson correlations					
	FlatK	SteepK	min pachy	spec VA	CLVA
FlatK		0.579	-0.548	0.338	0.162
SteepK			-0.464	0.292	0.181
min pachy				-0.198	-0.067

Spearman correlations		
	corneal stain	scar
min pachy	-0.196	-0.282

correlation is significant at the 0.01 level

## CONCLUSIONS

Pachymetry is moderately negatively correlated with both the flat and steep simulated K, supporting the widely accepted concept that thinner corneas are likely to be steeper. Thinner corneas do tend to have steeper K readings in both the flat and steep meridians. Additionally, they tended to have reduced spectacle acuity. However, having a thin cornea did not correlate with contact lens acuity. This suggests that thinness of the cornea does not impact best corrected contact lens acuity. However, thinner corneas were more likely to have staining and scarring. This may suggest that while patients with very thin can achieve improved vision with lenses they should be monitored more closely for the sequelae of corneal scarring and staining.

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# Rosacea as a differential diagnosis in corneal scarring

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## BACKGROUND

Rosacea is a dermatological condition characterized by changes in the sebaceous glands and the blood vessels of the face as well as the adnexa of the eye. Rosacea is often diagnosed by both dermatologist and eye care providers. Studies have shown that both groups are successful in diagnosis. There are four main subtypes of rosacea: erythematotelangiectatic, papulopustular, phymatous, and ocular.

Recently, it has been noted that patients with rosacea have increased concentration of Demodex. It is estimated that Demodex elevation occurs in 30% to 100% of patients suffering with rosacea. It is believed that this elevation in Demodex leads to an increased innate immunity response with resultant increase in inflammatory mediators. These inflammatory factors include interleukin-1, interleukin-6, interleukin-8, interleukin-17, interleukin-22, interleukin-23, interleukin-25, interleukin-36, interleukin-37, interleukin-38, interleukin-39, interleukin-41, interleukin-42, interleukin-43, interleukin-44, interleukin-45, interleukin-46, interleukin-47, interleukin-48, interleukin-49, interleukin-50, interleukin-51, interleukin-52, interleukin-53, interleukin-54, interleukin-55, interleukin-56, interleukin-57, interleukin-58, interleukin-59, interleukin-60, interleukin-61, interleukin-62, interleukin-63, interleukin-64, interleukin-65, interleukin-66, interleukin-67, interleukin-68, interleukin-69, interleukin-70, interleukin-71, interleukin-72, interleukin-73, interleukin-74, interleukin-75, interleukin-76, interleukin-77, interleukin-78, interleukin-79, interleukin-80, interleukin-81, interleukin-82, interleukin-83, interleukin-84, interleukin-85, interleukin-86, interleukin-87, interleukin-88, interleukin-89, 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These inflammatory mediators appear to be implicated in changes to the ocular surface. Patients with rosacea have exhibited many ocular complications including:

- Scarring
- Bacterial keratitis
- Blepharitis
- Chalazion/hordeolum
- Chronic cicatrizing conjunctivitis
- Corneal thinning and perforation
- Dendritic keratopathy
- Episkeratitis/scleritis
- Madarosis
- MGD
- Phlyctenule formation
- Pseudokeratomoclonus
- Pyogenic granuloma
- Vasculization

The treatment of rosacea frequently includes topical preparations as well as oral tetracyclines. Many studies have demonstrated the efficacy of various treatment modalities. Most common topical preparations include metronidazole and azelaic acid. Oral tetracyclines have also been shown effective and may be used in combination with topical preparations. Doxycycline has become the drug of choice with effective doses ranging from 20mg a day to 50mg twice a day.

Presented here are two cases in which patient had been previously diagnosed by dermatology with rosacea. However neither patient had disclosed this information at prior optometric visits and neither was using previously recommended treatments for their rosacea. Both patients were Caucasian females who exhibited corneal scarring. Patient one had been diagnosed with Terrien's and patient two with Salzmann's. Upon confirming their diagnoses of rosacea and based upon the appearance of their facial skin and ocular adnexa both patients were treated with oral doxycycline and topical metronidazole. Presented below are their initial and subsequent appearances.

## CASE ONE

A 64-year-old white female exhibited scarring of the superior cornea. It had a mildly arcuate appearance in the patient was preliminarily diagnosed with Terrien's marginal degeneration. She was wearing by Biofinity soft torics and complained of ocular discomfort, redness, and instability of vision. Best corrected vision was 20/25 in each eye and binocularly. Areas of redness and 4-5 pustules were noted on her face including a small ulcerated area just inferior to the lid margin of the left eye.

Figure 1 shows the changes to the lower lid of patient one. There is telangiectasia as well as madarosis. On the lower lid you can see the small ulceration.



Figure 2 is a photograph of the upper lids of patient one. On the right eye you can see the telangiectasia as well as collarettes appearance indicative of elevated Demodex. On the image of the left eye you will notice pustule on the side of the nose as well as induration of the lower lid.



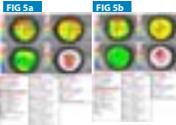
Figure 3 is Meibography of the lower right lid. Due to the swelling of the lid it was unable to get reliable Meibography. You will note that Meibography of the right lid shows a grade three changed meibomian glands with dilation, shortening and loss of the glands.



Figure 4 are corneal pictures. 4A shows the right cornea with its superior corneal scarring that follows the lid margin. Similarly for B shows the left cornea and its corneal scarring along the upper lid.



Figure 5 just patients once corneal topographies which exhibit mild irregular astigmatism with superior steading. For this reason the patient was refit into a Purevision2 for astigmatism. It was hoped that the aspheric optics and the larger overall diameter would improve quality of vision and stability.



At this visit on June 4, the patient was started on doxycycline 20 mg twice daily and metronidazole applied to the erupted plaques on the face as well as a thin layer applied to the upper and lower lids at that time. She was also given Tobradex ointments to apply to the ulcerated lower left lid twice a day. She returned on July 9. At that time she reported marked improvement in vision and comfort with her contact lenses and decreased facial and ocular redness. Vision had improved to 20/20 in the left eye and was stable at 20/25 in the right. Binocularly she had also improved to 20/20. She reported good compliance with her treatment regimen. The ulceration of the lower left lid was resolved.

Figure 6 shows the marked improvement in the appearance of the ocular adnexa her redness in telangiectasia are markedly less noticeable and the pustule on her nose is almost completely resolved. Her conjunctiva is significantly less injected.

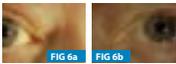


Figure 7 are the Meibography of both lower lids. At this visit their left lower lid was significantly less swollen and a better lower lid eversion and therefore a reliable Meibography was able to be performed. Note the marked improvement in the meibomian glands. She now is only a great to in both eyes.



Figure 8 is a high mag photograph of the left cornea showing the areas scarring however there is less surrounding haze. At this visit, her Tobradex was discontinued and her doxycycline was reduced to once a day. She continued to use the metronidazole daily. She remains stable with this combination treatment and has experienced no adverse events related to either medication.



## CASE TWO

A 59 year old white female was referred by a corneal specialist OMD for a scleral lens fitting having failed with her Air Optix Multifocal contact lenses. She had been diagnosed with Salzmann's nodular degeneration. She presented with concerns of ocular discomfort and a desire to return to contact lens wear

Figure 9 shows the superior lids of the patient which exhibit prominent telangiectasia and mild collarettes formation.



The telangiectasia is consistent with that of ocular rosacea and the collarettes confirm suspicions of Demodex elevation.

Figure 10 shows the inferior lids of patient one. The lower lids have a mildly scalloped lid margin. Some of the meibomian gland orifices appear to be dilated and there is mild telangiectasia.



Figure 11 shows the corneal appearance of the patient one. Figure 11a shows a flat round nasal scar likely a result of a previous staph marginal ulcer or phlyctenule. 11b shows the areas of superior scarring that run parallel to the superior lid of the right eye. Figures 11B and C show the changes to the left cornea with and without fluorescein. Superior nasal on the left cornea is the area that has been considered a Salzmann's nodule that we now believe is likely a corneal phlyctenule.



Figure 12 are the meibographies of the two eyes. The right lower lid is a grade 4 loss. All other lids are a grade 3. The glands

are noticeably truncated and dilated with multiple areas of atrophy.

Figure 13. Topographies showed mild irregularity of the corneal surface with no evidence of corneal thinning. She was fit into a OneFit P+A MP in the right eye and an MSD lens in the left eye.

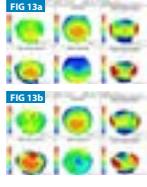


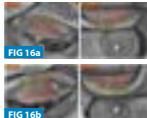
Figure 14. Both eyes exhibited improved redness and telangiectasia.



Figure 15. The lenses were dispensed they cleared well centrally as seen on both optic section and OCT. There was limbal clearance and appropriate landing on the conjunctiva. The left lens allowed for adequate clearance of the phlyctenule/nodule. The lenses were dispensed and the patient noted markedly improved vision and comfort. She was reading 20/20 binocularly at distance and 20/30 paragraph at near. She was continued on her regimen of metronidazole and doxycycline. At subsequent visits, the nodule had begun to decrease in size and the blood vessels surrounding it were beginning to recede.



Figure 16 are the meibographies taken on December 4th showing significant improvement of the meibomian glands.



The patient remains stable on her combination of oral and topical medications. She has achieved 12 hours of successful lens wear. She has exhibited no adverse effects from her treatment regimen.

## CONCLUSION

Both of these patients showed rapid, marked improvement in their ocular surface and symptoms with treatment of their rosacea. Both patients were able to achieve successful contact lens. They will be monitored every 3-6 months for additional sequelae related to their rosacea.

These cases demonstrate the importance of considering rosacea in the differential diagnoses of corneal scarring in close proximity to the lids. Suspicion of rosacea keratoconjunctivitis with subsequent scarring should be heightened in the presence of telangiectasia of the face and/or lids, demodex over growth, and MGD. Traditional rosacea treatments of oral doxycycline and topical metronidazole can provide improvement in ocular signs and symptoms. The patients here were able to safely use metronidazole on the lids with careful attention not to get the medication in the eye or on the lid margin. Additional ocular treatments with warm compresses and lid scrubs to control demodex and improve MGD may also be helpful in providing relief. Both patients expressed appreciation for their improved appearance, vision and comfort.



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## 1 ICO PRESENTATION

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# Overcoming Barriers to Publication – A Case Report of a Chiropractic Researcher

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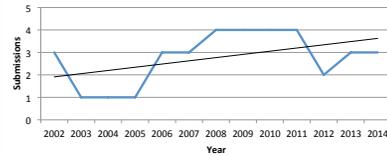
## INTRODUCTION

A chiropractic researcher has a complaint of frequent delays in getting very developed but partially incomplete manuscripts into the publication pipeline. He does not lack productivity and in fact is relatively prolific. However, he has been accumulating developed manuscripts at a rate that exceeds his submission rate. This publication lag has been a source of significant concern and now threatens to diminish productivity, because the cognitive dissonance it creates is affecting him mentally and physically.

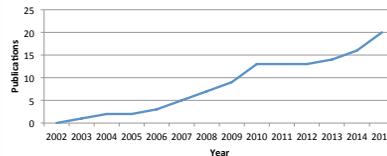
The subject had decided to attend 2013 ACC/RAC meeting for other purposes than addressing his issue. In fact, the issue was rather ill-defined in his own mind. While at the meeting, the researcher decided to attend a platform presentation entitled: Publication Rates of Abstracts Presented at Association of Chiropractic Colleges/Research Agenda Conference Meetings Between 2002 and 2008.<sup>1</sup> At this presentation, he learned that the publication rates of abstracts from ACC/RACs were about 32%, which is on the low end of the range of publication rates from other spine and orthopedic surgery national organizational meetings: 34-59%.<sup>2-12</sup> It was at this point that the subject had an epiphany of sorts and recognized in himself what the issue was and that he was probably not alone in experiencing it.

The subject made contact with the first author to share his personal experiences. In contacting the first author, the subject hoped that generalizing his experience and, more importantly, his plan to take up the first author's suggestions at the conference may be of interest to other researchers who also experience the phenomenon of presenting studies at scientific meetings but failing to follow up and publish the studies in peer reviewed journals.

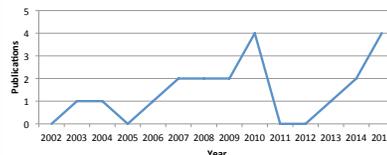
ACC/RAC Submissions



Cumulative Publications



Publications Per Year



## BEHAVIORAL CHANGES

The subject heard the ACC/RAC presentation in Spring of 2013, so it was not until the Fall of 2013 that the subject resolved to make behavioral modifications to improve his publication rate. These included:

- Arranging with family members for lifestyle modifications to free up more time.
- Reducing (he stated "regrettably") his effort to maintain let alone develop his private practice.
- The head research administrator became more supportive of submitting to journals that were the most appropriate given the content of the projects, irrespective of their impact factors.
- The subject (somewhat reluctantly) took over the responsibility of finalizing and submitting manuscripts that were languishing in the hands of colleagues who were supposed to have done so.

The subject resolved to attempt submitting one manuscript per month in 2014, including but not limited to prior ACC/RAC submissions in limbo.

## OUTCOMES

As of August 2014, the subject has submitted 7 manuscripts in 7.5 months. He also hopes to submit 3-4 manuscripts to the 2015 ACC/RAC conference. Of those submitted in 2014, 4 are either published or in press, and 3 are in review as of this writing. As the subject works through the backlog of manuscripts, he is experiencing less mental stress and is feeling better about his situation.

## DISCUSSION

The problem of conducting research, presenting the information at a scientific meeting but failing to complete the process by submitting it for journal publication, is a phenomenon that can be recognized in many researchers – somewhere between 1/3 to 2/3 of chiropractic and spine and orthopedic surgery researchers. It is possible that the cognitive dissonance created by this behavioral pattern can actually cause mental/physical problems, as seen in this case, but the frequency that this happens is unknown.

After the initial presentation on ACC/RAC publication rates in 2013,<sup>1</sup> those same authors presented a survey at the 2014 ACC/RAC on the self-reported barriers to publication experienced by chiropractic researchers. It showed that lack of time and low priority to publish were most frequently cited as reasons for not publishing in this population.<sup>13</sup> These seemed fairly similar to self-reported barriers to publication for a cohort of orthopedic surgeons.<sup>14</sup> The presenters concluded that these barriers were largely institutional in nature. In this case, it seemed to the subject that, although some of the barriers may be institutional (e.g., the expectation to publish in journals with a certain minimum impact factor and lack of support staff), many of the barriers were basically time management issues. Once this researcher recognized the manifestation of the phenomenon in himself, how to manage the problem became apparent. He decided he needed better time management with a rearrangement of priorities to making finishing manuscripts more important.

## LIMITATIONS OF THE STUDY

The experience of this researcher in this case report does not necessarily reflect the experience of other individuals whose personal circumstances may differ in important ways. It is presented for the purpose of illustrating how awareness of the research bottle-necks may under certain circumstances provide an impetus for behavioral modification leading to greater publishing success. The greater emphasis on publishing came at the expense of developing his private practice, a choice which may not be considered appropriate for publishing researchers in general.

## CONCLUSION

This case report illustrates how one researcher, having become more fully aware of why and how he had failed to fully realize his potential and having better understood the barriers to publication that were most amenable to amelioration, was able to translate research on barriers to publication into actually breaking through such barriers.

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ICO

# COVD

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# When An Eye Turn Is More Than Strabismus: A Case Report

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## INTRODUCTION

A six-year-old African American male presented for his first eye exam with parental concern regarding a longstanding intermittent left eye turn. The eye turn was described as stable, and had been occurring since birth. Birth and developmental history were reported as unremarkable. Patient was in 1st grade with no complaints regarding his vision.

## RESULTS

The patient's best-corrected distance visual acuity was 20/20 OD and 20/200 OS. The best corrected near visual acuity was 20/20 OD and 20/30 OS. Pertinent and concerning exam findings prior to dilation included leukocoria OS (**figure 1**), a grade 1+ afferent pupillary defect and unequal Bruckner reflex as shown in **table 1**. An intermittent 25 prism diopter left exotropia at near was observed (**table 2**) with an abnormally slow refixation of the OS on alternating cover test. These findings were accompanied by an abnormal head turn to allow for eccentric viewing of the OS when the OD was occluded, along with poor fixation of the OS. Retinoscopy was +0.25-1.00x180 OD and +3.50-4.00x180 OS with a poor quality retinoscopy reflex in the OS. Anterior segment was normal with equal corneal diameters OU. Non-contact tonometry was 13mmHG OD and 14mmHG OS. The dilated fundus evaluation OS revealed diffuse and coalesced intra-retinal and sub-retinal exudates adjacent to tortuous and telangiectatic vasculature with a fibrotic nodule over the macula (**figure 2a**). The OD was unremarkable (**figure 2b**). The ophthalmic B-scan ultrasonography of the OS revealed an elevation over the macula without any apparent calcifications or retinal detachment (**figure 3**).

Figure 1: Leukocoria in the OS secondary to Coats disease.



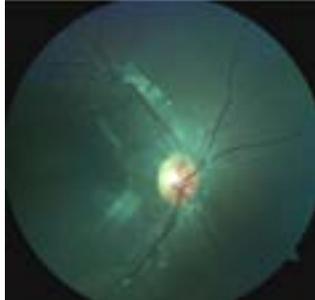
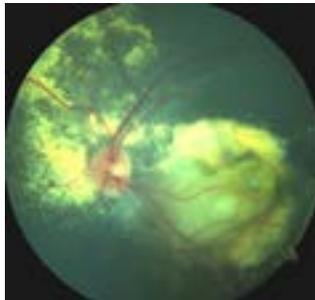
Table 1: Entrance testing at patient's first eye exam.

Test	OD	OS
VA: D	20/25+2	20/200, PHNI
N	20/20	20/30-
Pupils	PERRL (-)APD	PERRL 1+APD
ECOMs	FULL	FULL
CVF	FTFC	FTFC
Bruckner	Red	Unequal
HIX	Centered	Nasal 1mm

Table 2: Binocular vision testing at patient's first eye exam.

Test	Results
Cover Test	20 PD Intermittent Left Exotropia 25 PD Intermittent Left Exotropia
Stereo	Suppression OD, I / Fusion/FLY
CVME	89 OD, 49 OS
Retinoscopy	OD +0.25-1.00x180 OS +3.50-4.00x180 (poor quality)

Figure 2a: Fundus photo of OS showing diffuse and coalesced exudates throughout posterior pole, retinal telangiectasia, and fibrotic nodule over the macula. 2b: Unremarkable fundus photo of the OD.



## DISCUSSION

Differential diagnosis for sensory strabismus and leukocoria include retinal detachment, retinoblastoma, Coats disease, toxocariasis, persistent hyperplastic primary vitreous, retinopathy of prematurity, retinal capillary hemangioma and retinal astrocytoma.<sup>1,2</sup> A referral to a pediatric ophthalmologist confirmed the diagnosis of Coats disease.

Figure 3: B-scan ultrasonography of the OS showing elevation over the macula. No apparent calcifications or retinal detachment.

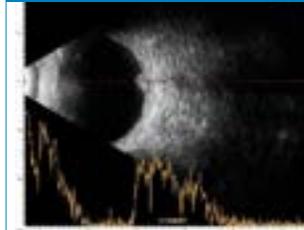
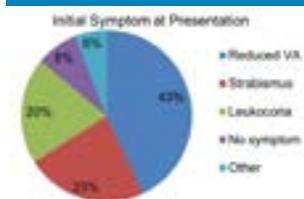


Figure 4: Presenting symptoms for patients with Coats disease from the largest Coats disease retrospective review available. N=150, from the oncology service at Wills Eye Hospital between 1976-2000. Other symptoms include pain, heterochromia and nystagmus.<sup>2</sup>

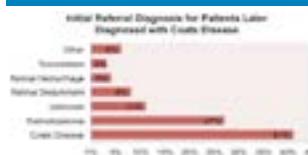


Coats disease is an exudative retinopathy that has no racial predilection.<sup>3,4</sup> It is found in males 75% more often than females.<sup>2,4</sup> There are no known systemic associations and presentation is usually during the first decade of life.<sup>3,4</sup> The typical presentation is unilateral, however in rarer cases, it can be bilateral.<sup>2,4</sup>

The initial symptoms at presentation of Coats disease is displayed in **figure 4**. Strabismus is the initial symptom in 23% of cases according to a retrospective review from the Wills Eye Hospital.<sup>2</sup> Characteristic retinal presentations include retinal telangiectasia or light-bulb aneurysms, exudates that characteristically accumulate in the macula, and exudative retinal detachments in the posterior pole.<sup>2</sup> More advanced stages of Coats disease can lead to anterior segment findings like cataract, iris neovascularization, glaucoma and orbital cellulitis.<sup>4</sup>

The most common differential diagnosis for Coats disease is retinoblastoma (**figure 5**).<sup>2</sup> Similar to Coats, the most prevalent initial symptom for retinoblastoma are leukocoria (60% of cases) and strabismus (20% of cases).<sup>1</sup> Unlike in Coats disease, retinoblastoma clinical findings include uniformly dilated blood vessels that may taper near the tumor, as well as white fluid exudate.<sup>5</sup> Accepted practice in either condition is to enucleate when the etiology of the mass cannot be determined.<sup>6</sup> Coats is the 2nd most common cause of pseudoretinoblastoma that leads to enucleation.<sup>6,7</sup>

Figure 5: Initial diagnosis at time of referral for patients later diagnosed with Coats disease. N=150, from the oncology service at Wills Eye Hospital.<sup>2</sup>



## CONCLUSION

The patient was referred to a pediatric retinal specialist for later treatment to eradicate the telangiectasia and allow the exudate to resolve. The patient was also given full time wear glasses with polycarbonate for protection. The parent was educated about the eye turn and future treatments. Coats has been deemed a lifetime disease and requires frequent monitoring, as recurrence after treatment is common into adulthood.<sup>8</sup> This case is an example of an intermittent eye turn in the presence of a thorough and signifies the importance of a thorough health examination with an atypical strabismus.

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# Wolf-Hirschhorn Syndrome: A Case Report

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## ABSTRACT

**BACKGROUND:** Wolf Hirschhorn Syndrome (WHS) describes a series of malformations caused by a deletion of one copy of the distal short arm of chromosome 4p (called 4p-). WHS is characterized by anomalies that occur in the first trimester that result in intellectual disability, delayed development, seizures, and a characteristic face appearance referred to as the "Greek Warrior Helmet Face." The extent of the 4p deletion determines the occurrence of microcephaly, midline defects, cardiac and renal anomalies. To provide appropriate ophthalmic and medical care to children with Wolf Hirschhorn syndrome, it is important to understand this rare genetic abnormality. A case study of a nine year and seven month old Caucasian male with WHS is presented.

**CASE REPORT:** A 9 year 7 month old Caucasian male child with Wolf-Hirschhorn syndrome presented with a large angle exotropia, possible glaucoma and high astigmatism. He also has hypertelorism, developmental delays and a history of occupational and physical therapy.

**CONCLUSION:** Wolf-Hirschhorn syndrome diagnosis is determined by phenotypical expressions, such as classic facial abnormalities and confirmed by detection of a deletion of the Wolf Hirschhorn critical region (WHCR) (chromosome 4P16.3). The developmental problems vary based on the size of the deletion while the pathogenesis is determined by chromosome abnormalities, extent of seizures, and systemic deviations. WHS patients can benefit from genetic counseling, systemic treatment, ocular, and management of psychosocial issues. This will help the patient and families to deal with common problems like limited speech and comprehension. And all oculo-visual problems should be monitored, diagnosed and treated as is appropriate.

**KEYWORDS:** Partial deletion 4p-, microcephaly, Greek warrior helmet face, hypertelorism, wolf-hirschhorn syndrome, seizures

## INTRODUCTION

The etiology of Wolf Hirschhorn syndrome (WHS) is partial deletion of the short arm of chromosome 4p (4p16.3). The WHS critical region is the smallest region of overlap with WHS patients and is limited to 165kb. A 25 exon 90kb gene called WHSC1 has been mapped to the WHS critical region. It is expressed in early development and encodes a 136kDa protein that has four domains that are found in other development proteins. WHSC1 is expressed with a preference for fast growing embryonic tissues in a method that corresponds to affected organs in patients with WHS.<sup>1</sup> There is an estimated 30% percent of WHS patients with anomalous ophthalmic manifestations.<sup>2</sup> The estimated prevalence of the disease is 1:50,000 births with a 2:1 female:male ratio.<sup>3</sup> WHS appearance in patients has been described as the 'Greek Warrior Helmet Face' because of the broad nasal bridge accompanying ocular hypertelorism.<sup>2</sup> Also, this Greek warrior face describes an enlarged glabella, high arched eyebrow, wide nasal bridge, proptosis with a downturned mouth, epicanthal folds and bilateral cleft lip or palate (especially in infancy).<sup>2</sup> Microcephaly and micrognathia (smaller lower jaw that often interferes with normal tooth development) are frequently noted as well.<sup>4,5,6</sup> Other anomalies include hypotonia, intra-uterine and postnatal growth retardation, intellectual disability and cardiac problems.<sup>5,7</sup>

## CASE REPORT

The 9 year 7 month old child with WHS has a noteworthy medical history. He was born full-term after 38 weeks of gestation, birth weight of 907 grams and reported oxygen therapy for 2-3 months in NICU after birth. Despite being at risk for retinopathy of prematurity based on the screening criteria reported by the American Academy of Ophthalmologists (birth weight of less than or equal to 1500 grams, gestational age of 30 weeks or less or birth weight between 1500 and 2000grams or gestational age of more than 32 weeks with an unstable clinical course), no retinopathy of prematurity was noted in this patient.<sup>8</sup> His family history was unremarkable with no alcohol, tobacco or illicit drug use.

Table 1: Case Summary

Chief complaints	Wolf-Hirschhorn Syndrome, School for special needs, occupation and physical therapy
History of Present Illness	Referred for strabismus and refractive care, no squinting or other anomalies noted
Ocular History	Glaucoma Suspect Low kidney function Seizure disorder
Systemic History	Enalapril Maleate and Phenytoin Latanoprost gts OU
Podiatric History	Gestational age: 38 weeks, premature delivery. Birth weight: 2 lbs. No ROP Oxygen required: 2-3 months duration in NICU
Entrance Test Findings	Abnormal most, Oriented x3 Pupils: pupils, dark = light OD, OS CVF: unable to test, poor fixation Mobility: poor fixation, torsional nystagmus
Anterior Segment	Unremarkable Peri-iridial shadow test: Angles open
Posterior Segment	Vitreous Clear Macula Clear OD: Large CID, cupping OS: Large CID, cupping, PPA 0.7 AVN ratio
Periphery	Limited periphery secondary to poor fixation
Additional Testing	Hirschberg-60 prism diopters Alternating Exotropia (XT) OD fixation preference Kappa Central, steady OD, OS Krimsky: -50 prism diopters MEM: Attempted, poor fixation

The patient was referred from another clinic for a refractive assessment and a possible strabismus of the right eye. The last eye examination was 2 months prior as described in the table below. Visual acuity was not recorded at that visit, perhaps due to limited cooperation, receptive/express speech and/or comprehension. The mother of the patient had not noticed the eye-turn prior to the referral but had noticed frequent voluntary bulging of eyes. His mother also denied seeing any squinting or other visually/ocular induced anomalous behaviors. The patient participated in occupational and physical therapy at the time of this visit.

Table 2: Characteristic Anomalies Associated with Wolf Hirschhorn Syndrome and those characteristics noted in our patient

Anomalies associated with WHS	Anomalies present in our patient
Exodeviation	Large angle Exotropia
Nasolacrimal obstruction	Torsional Nystagmus
Shallow Orbits	Seizure Disorder
Epicanthal folds	Intellectual delay/Developmental Delay
Foveal hypoplasia	
Upper lid coloboma	
Optic Disc Anomalies	
Down-slanting Palpebral fissures	
Hypertelorism	
Nystagmus	
Congenital and juvenile glaucoma	Glaucoma suspect-low risk
Facial Abnormalities:	Hypertelorism
Epicanthal folds	
Hypertelorism	
Proptosis	
Microcephaly	
Wide nasal bridge	
Micrognathia	
Enlarged glabella	
Ocular Abnormalities:	Large CID optic nerves
Shallow orbits	
Anterior segment anomalies,	
Optic disc abnormalities	
Nasolacrimal obstruction	
Chorioretinal colobomas	
Foveal hypoplasia	
Down slanting palpebral fissures	
Microcosmes	

Table 3: Ophthalmic Manifestations noted in literature for Wolf Hirschhorn Syndrome<sup>13</sup>

Ophthalmic Presentations with WHS	Patient description
Out of 10 patients aged 4 months to 11 years with ophthalmic presentations studied <sup>14</sup>	8 patients with 4p- showed interruptions that ranged from band 4p14 to 4p16.3
Exodeviation	9 patients
Nasolacrimal obstruction	6 patients
Shallow Orbits	3 patients
Epicanthal folds	3 patients
Foveal hypoplasia	3 patients
Upper lid coloboma	2 patients
Optic Disc Anomalies	2 patients
Down-slanting Palpebral fissures	2 patients
Hypertelorism	1 patient
Nystagmus	1 patients

## DISCUSSION/ CONCLUSION

Wolf et al. and Hirschhorn et al. individually described Wolf-Hirschhorn syndrome as occurring because of a contiguous gene syndrome with differing phenotypes.<sup>2,3</sup> The most common ophthalmic finding is exotropia, with bilateral nasolacrimal obstruction being the second most frequently encountered anomaly in patients with WHS.<sup>3</sup> There are several other frequently encountered features many of which were not noted in our patient.<sup>2,11</sup> This particular patient had a limited ability to communicate and cooperate during the assessment.<sup>5</sup> However, there are instances where speech and language skills have developed.<sup>6</sup>

The more common ophthalmic presentation with Wolf Hirschhorn syndrome are strabismus (exodeviations), nasolacrimal obstruction, shallow orbits, epicanthal folds, foveal hypoplasia and nystagmus. We provided an appropriate refractive correction and am monitoring the other areas of concern (glaucoma suspect). The challenges of patients with WHS will require the optometrist to work within a multi-disciplinary environment so that management of these patients can best increase their quality of life to

the fullest extent possible by providing single, clear, comfortable, binocular and pathology free vision. The plan for this patient included a prescription for full time wear of polycarbonate spectacles for astigmatism and monitoring alternating exotropia. The patient was to be monitored at a home clinic and scheduled to return to us in three months. Unfortunately, the patient did not return for that follow up appointment.

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ICO

# EUNOS

(EUROPEAN NEURO-OPHTHALMOLOGICAL SOCIETY)

1 ICO PRESENTATION

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# GPS usage in Bioptic and Non-Bioptic Neuro-Ophthalmic Low Vision Drivers

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## AIM

- Low vision practitioners often recommend their patients drive with a passenger to assist with navigation. Often, this is not possible and global positioning systems (GPS) can serve as an additional navigator while driving.
- GPS have markedly improved since their development in 1988.
- We surveyed bioptic and non-bioptic neuro-ophthalmic low vision drivers to determine their GPS (global positioning system) device usage

## METHODS

- After IRB approval, 27 low vision patients were recruited for a phone survey.
- Patients were eligible to participate if they were 18 years of age or older, and met one of the following criteria:
  - best corrected visual acuity (BCVA) worse than 20/40
  - central or significant peripheral visual field defects
  - combination of above
- Participants were also required to be active drivers with a valid Illinois driver's license (Unrestricted, Daylight Only restriction, or Bioptic restriction; as defined by the state of Illinois vision requirements for driving).
- Survey included questions of driving demographics (i.e. frequency of driving, motivation for driving) and GPS usage (i.e. type of device, helpful features, factors for not using)
- The results of the entire study were presented at the 2015 ARVO meeting. The present poster focuses solely on four patients with neuro-ophthalmic conditions.

## RESULTS

- The patient ages were 26, 58, 61, and 62.
- Two had AION, one optic atrophy, and one homonymous hemianopia.
- One was a bioptic driver currently, and another was in the process of training with a bioptic.
- All four patients used GPS and reported improved comfort and/or safety level.
- All four also reported using GPS only when driving to unfamiliar locations

## CONCLUSIONS

- Given that 100% of those who use GPS while driving report increased comfort and safety level, neuro-ophthalmologists and low vision specialist should educate patients on the use of GPS as an additional navigator while driving.
- Investigations are needed to analyze further enhancement and accessibility features that may benefit a low vision driver using GPS as an aid.
- GPS provides an additional navigational tool for both bioptic and non-bioptic neuro-ophthalmic low vision drivers.

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FIGURE 1: Screen shots from "Google Maps" indicating appropriate lane choice before upcoming turns.

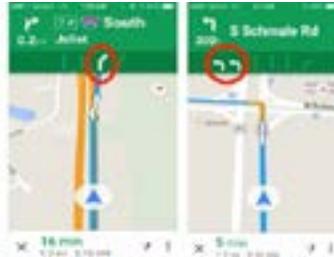


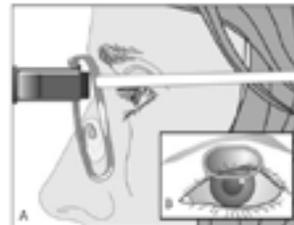
FIGURE 2: GARMIN GPS naviga7on device.



FIGURE 3: Built-in car GPS navigation system



FIGURE 4: Schematic detailing proper bioptic positioning for viewing through the telescope(4a) and through the carrier lens (4b). Only 5-10% of time should be spent viewing through the telescope for distance spotting.





ICO

# GSLs

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# ICO

# Localized Corneal Graft Rejection from Scleral Contact Lens Wear with Excessive Limbal Clearance

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## BACKGROUND

Contact lens wear is known to decrease oxygen transmission to the cornea. Many studies have been performed to determine what the minimum criteria transmission is to prevent corneal hypoxia. The Holden and Mertz criteria recommend 23 for open eye and 89 for closed. While Harvitt and Bonano revised this recommendation to 35 and 125. For irregular corneas which have undergone incisional surgeries oxygen needs may be greater to prevent ocular sequelae. These highly irregular and challenged corneas are being fit more frequently in advanced designs especially scleral lenses. Recent studies suggest that similar concerns exist for oxygen transmission under these lenses. Michaud et al. hypothesized that Fatt's formula of "resistance in series" which has been previously applied to piggyback lenses and other designs can also be applied to sclerals where the second lens is the tear lens. This formula is

$$Dk = \frac{1}{(t1/Dk1) + (t2/Dk2)}$$

In addition to overall oxygen concerns in scleral lens fitting, there is concern for mechanical irritation to the delicate stem cells at the limbus. Therefore, limbal clearance is recommended. The lens should not impinge on the conjunctiva but rather allow some flow of tears to minimize stagnation. We present here a complicated case where central clearance was achieved but the limbus was severely vaulted with conjunctival compression. As a result this patient developed a focal rejection in his graft in the area of greatest clearance.

## CASE

A 54 year old patient with keratoconus and a history of penetrating keratoplasty OS presented for a contact lens refitting. He had previously been seen in urgent care with a red, painful left eye for about a week. It worsened with lens wear. He was wearing a Jupiter reverse geometry lens in 6.49 base curve, 16.0 diameter, with a -15.75 sphere. The lens was 18 months old and he had a similar episode 6 months prior but did not return for refit. He had a central corneal abrasion and was treated accordingly. When he presented for the lens fitting, he had been out of lens wear for two weeks and the eye was white and quiet.

Evaluation of his old lens revealed significant central touch of greater than 2mm in diameter. It cleared the limbus but then impinged on the conjunctiva. It was determined that this poor

fitting lens was the likely etiology of his red eye and a new lens fitting was initiated. However vision was quite good at 20/30. His keratoconic eye was seeing 20/50 through a similar scleral lens. His corneal transplant was clear with only one small area of vascularization along the 2 o'clock incision where he had a previous localized rejection when a suture had ruptured.

The corneal transplant was extremely proud and initial efforts were made to fit the eye with a piggyback as the patient had brought in an old corneal lens and was anxious to return to lens wear due to the poor vision (20/300). Seven different soft lenses were tried however all fluted excessively and dislodged with just a few blinks. Therefore, a new scleral lens design was chosen with adjustable limbal vault in hopes of clearing the donor host junction.

The first lens chosen was a 540 micron sag with a standard limbal profile and standard edge, 15.8 diameter MSD lens in -8.00. This lens continued to touch severely but provided 20/30 vision with a -8.75 over-refraction. Mild conjunctival compression was noted. The next lens in the set was a 560S. It still touched the cornea and impinged the conjunctiva. The lens was imaged and the data was sent to the lab to custom design a significantly deeper lens.

The lab designed a 600II (600 micron sag with a double increased limbal profile) with a double flat edge. The power was -15.75. Remarkably, the patient saw 20/40 with the lens and there was only a -0.25 over-refraction. It cleared the apex and the limbus. It was not imaged but was dispensed. The patient did not show for his one week follow-up but rather returned urgently at five

weeks complaining of pain and redness. The redness had started a few days after receiving the lens but would go away overnight for the first few weeks. However in the last week the eye had been becoming increasingly red and tender. He was wearing the lens all waking hours as he loved the vision (20/40+)

Evaluation of the contact lens revealed excessive clearance over the limbus. It was more than double the thickness of the cornea. Central clearance appeared to be near 500 microns. OCT analysis confirmed excessive clearance. In addition there was compression about 0.75mm outside the limbus and inside the scleral lens.

The cornea showed generalized haze and prominent injection with vascularization along the 7 to 8 o'clock hour donor/

host junction. The lens had also left a prominent conjunctival impression. The patient was diagnosed with rejection and started on topical loteprednol gel four times a day with copious tears. He was instructed to discontinue lens wear.

He was seen twice over the next week and resolved quickly. A new lens was ordered with reduced limbal clearance. The new lens had parameters of 600S (standard limbal profile) with a double flat edge. The lens provided improved comfort and excellent vision. Limbal clearance was markedly reduced. The patient did not return for follow up but was contacted and reports no redness with the new lens.

## DISCUSSION

While scleral lenses provide excellent vision for patients with highly irregular corneas they are not without risk. In this case the high limbal clearance and heavy conjunctival compression resulted in significant decrease in oxygen to the cornea. The calculated Dk/t for 1.4mm of limbal clearance is only 4.1 which falls well short of both the Holden-Mertz and Harvitt-Bonano criteria. In addition, the conjunctiva was severely compressed where the very steep limbal curve met the very flat peripheral curve choking off the conjunctival vessels. This likely exacerbated the hypoxic condition of the cornea. Removal of the lens and aggressive anti-inflammatory treatment resulted in rapid resolution.

The new lens was designed to reduce vault across the cornea and minimize the junction between the limbal and peripheral curves allowing a more gentle landing on the cornea. The patient was educated to keep his lens wear to 10-12 hours a day. A lengthy discussion regarding rejection and the risk of continuing to wear the lens when there are abnormal signs like redness could result in loss of the graft. Unfortunately patient continues to no show. We have been able to email and speak to him so we are hopeful that he remains compliant and reports no adverse effects from the new lens.

FIGURE 1  
OCT of corneal transplant

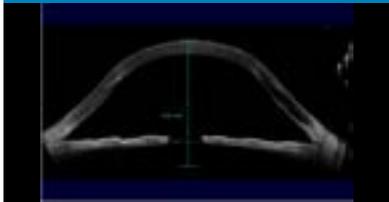


FIGURE 4A AND 4B  
OCT Images of the lens' central clearance (a) and nasal limbal clearance (b)

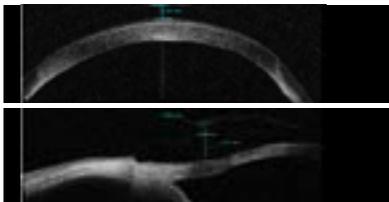


FIGURE 2A AND 2B  
Topographies of right (a) and left (b) eye

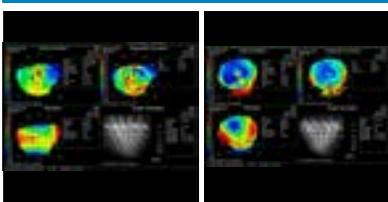


FIGURE 5A AND 5B  
Low (a) and high (b) mag corneal with visible haze and nasal injection located at point where clearance exceeded 1mm.



FIGURE 3A AND 3B  
Optic section views of the deep lens centrally (a) and nasal limbus (b)

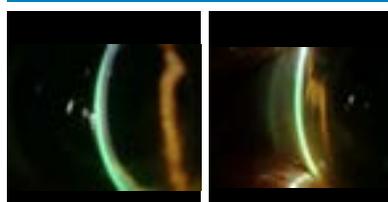


FIGURE 6  
Resolved corneal appearance



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# Differential Diagnosis of Terrien's Marginal Degeneration

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## BACKGROUND

Terrien's marginal degeneration was identified in the early 1900s as a rare peripheral corneal degeneration. The condition is found most often in younger men (3:1). It begins with peripheral corneal haze and leads to peripheral corneal thinning, vascularization and irregular astigmatism. The affected areas show a central channel of thinning with normal cornea on either side. Within this thinned zone, stromal cavity formation develops which is believed to be the reason for the thinning and risk of perforation. Over time, lipid will begin to deposit along the edges of the lesions.

Optical coherence tomography is helping better identify the corneal thinning and the changes within these lesions. Technology further aids in categorizing the condition into one of its five stages:

- Stage 1 - haze and thinning
- Stage 2 - lipid deposition
- Stage 3 - irregular astigmatism
- Stage 4 - pseudokeratoconus
- Stage 5 - perforation

Rosacea is a relatively common inflammatory disease of the skin. It is found in as high as 22 percent of patients of Northern European descent. It occurs in approximately three percent of the general US population and comes in four serotypes:

- Subtype 1 - erythematotelangiectatic (ERT)
- Subtype 2 - papulopustular (PPR)
- Subtype 3 - phymatous
- Subtype 4 - ocular

TRADITIONAL OCULAR SIGNS OF ROSACEA
Demodicosis
Meibomian gland dysfunction
Blepharitis
Corneal staining
Generalized thinning of cornea
Peripheral corneal infiltrates (usually inferior)
Lid telangiectasia

ADVANCED OCULAR MANIFESTATIONS OF ROSACEA DESCRIBED IN THE LITERATURE
Chronic cicatrizing conjunctivitis
Dendritic keratopathy
Pseudokeratoconus
Pyogenic granulomas
Bacterial keratitis
Phlyctenular keratoconjunctivitis
Corneal vascularization
Corneal scarring

We describe below a case that had previously been diagnosed as Terrien's marginal degeneration. However, upon closer slit lamp evaluation many of the signs of rosacea were identified. A more in depth history led to a new diagnosis and a new treatment plan.

## CASE

### VISIT 1 (05.04.2014)

A 64 year old Caucasian female presented noting hazy vision around lights and discomfort with her current PureVision Toric (8.7/14.0) lenses. She has type II diabetes, hypertension, and hyperlipidemia for which she is well-controlled with oral medication. Her habitual CLRx was OD -3.75 -1.75 x 070, OS -5.50 -0.75 x 100. Her new spectacle refraction was OD -4.50 -1.75 x 069 and OS -5.75 -1.25 x 105 with an add of +2.25. Slit lamp examination revealed lid margin telangiectasia OU, mild papillary response OU, meibomian gland dysfunction OU, pannus 360° OU, and superior peripheral corneal scarring. Topography was taken showing superior steepening consistent with Terrien's marginal degeneration.

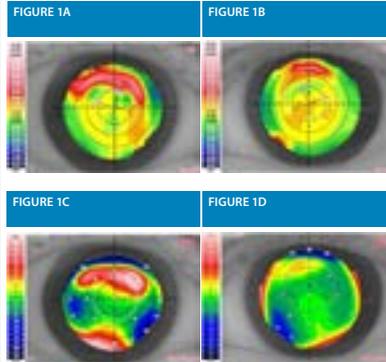


Figure 1. ReSeeVit topography maps (tangential, top and elevation, bottom) showing superior steepening and scarring in each eye.

She was re-fit into PureVision2 for Astigmatism 8.9/14.5 OD -4.00 -1.25 x 080 and OS -5.00 -1.25 x 100 to try and reduce ocular discomfort. The patient was to return in 2 weeks.

### VISIT 2 (05.22.2014)

At follow-up the patient reports a flare up of skin irritation she describes as "acne" which was previously diagnosed by dermatology. She reports she was prescribed sulphur gel (Proscace) by a dermatologist but has not been taking it recently.

Slit lamp examination reveals lid margin telangiectasia OU, inferior madarosis OS>OD, injection and papillary response OU, meibomian gland dysfunction OU, tear film debris OU, follicles OS with discharge and crusting OS, pannus 360° OU, peripheral thinning and superior scarring. The patient was dilated at this visit with findings of peripheral retinal atrophy in a single quadrant of each eye.

The patient was diagnosed with a bilateral flare up of rosacea conjunctivitis and concomitant blepharitis OD>OD. She was started on tobramycin/dexamethasone ointment QID OU. The contact lenses were finalized. She was to return in 2 weeks.

### VISIT 3 (06.04.2014)

The patient reports discharge is persistent OS and that she feels the condition has gotten worse. She has not seen dermatology and is still not using the sulphur gel. She feels the outbreak is worst from the left nasal canthal region down to the nasolabial fold. SLE confirms minimal improvement from last exam with new hordeolum developing on the left lower lid centrally.

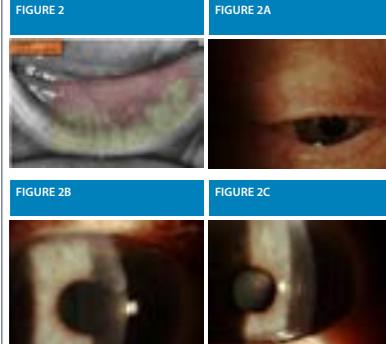


Figure 2. Baseline anterior segment photographs of the right eyelid and both the right and left corneas (see Figure 4 for baseline left eye lid).

At this time, the diagnosis was determined to be rosacea, likely mimicking Terrien's marginal degeneration. The patient was started on topical metronidazole to the skin twice daily. She was also started on oral doxycycline 20mg twice daily. She was to continue tobramycin/dexamethasone ointment QID OU.

### VISIT 4 (07.09.2014)

The patient denies any further redness, discharge or eye pain and notes the symptoms have improved by taking the oral doxycycline, topical metronidazole BID and tobramycin/dexamethasone ointment BID.

SLE reveals trace lid margin telangiectasia OU, meibomian gland dysfunction OU, pannus 360° OU, peripheral thinning and superior scarring (improved).

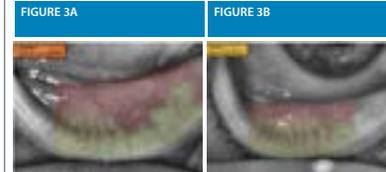


Figure 3. Meibography images of the right lower lid from before treatment (left) and after (right).



Figure 4. Anterior segment photographs of the left eye showing marked improvement from before treatment (left) and after (right).

The patient was instructed to decrease the tobramycin/dexamethasone to once daily until gone and continue oral doxycycline and metronidazole for maintenance.

## DISCUSSION

Our patient did not fit the profile for Terrien's, being female and over 40. She did not have any signs of lipid deposition along her corneal lesions nor were there any cavities visible in the stroma. Close slit lamp evaluation revealed thickening of the superior cornea along the scars representing hypertrophy rather than thinning. A closer examination of her adnexa revealed significant erythema and eruptions on the lids with telangiectasia leading to a suspicion of rosacea. When she was specifically questioned about rosacea, she confirmed that she had previously been treated. Standard treatment of PPR subtype rosacea includes concurrent oral and topical therapies. Recommended oral treatment is a subantimicrobial dose of doxycycline. Recent studies show equal effectiveness of 40mg and 100mg daily. An emerging alternative to doxycycline is azithromycin 500mg taken three times a week. Studies have not been done with azithromycin to determine if a lower dosing provides comparable response. Both treatment regimens show improvement at 6 weeks with protocols extending from 4-10 months. Topical treatment is considered additive and is usually comprised of metronidazole or azelaic acid. For dermatology treatment, maintenance is often topical. Standard treatment of ocular subtype rosacea also includes oral therapy. The standard is also oral doxycycline in the same dosing as for PPR. Topical cyclosporine has been shown to be additive in these patients.

As our patient exhibited the PRT subtype and significant ocular signs, we chose to treat with the concurrent oral and dermatological approach. The patient exhibited significant and rapid improvement of her rosacea signs. Her ocular discomfort resolved and contact lens awareness resolved. To our knowledge, this is the first case reported where dermatological metronidazole was applied around the eye.

As our patient exhibited the PRT subtype and significant ocular signs, we chose to treat with the concurrent oral and dermatological approach. The patient exhibited significant and rapid improvement of her rosacea signs. Her ocular discomfort resolved and contact lens awareness resolved. To our knowledge, this is the first case reported where dermatological metronidazole was applied around the eye.

## CONCLUSION

Rosacea can mimic many different ocular disease entities. The scarring and resultant irregular astigmatism may resemble such conditions as keratoconus and Terrien's marginal degeneration. This irregular astigmatism may result in their presentation to a contact lens clinic for improved vision. However, close evaluation will likely reveal erythematous and telangiectatic changes to the face and lids. In cases of PPR subtype, pustules may also be visualized on the face and/or lids. Traditional rosacea interventions can provide both dermatological and ocular relief. It is our hope that by managing this patient's rosacea, we will stabilize her cornea and vision.

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# NANOS

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# Normative Database for the King-Devick Test in Adults and Adolescents

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## INTRODUCTION

The King-Devick(K-D) test (Figure 1), a <2-minute timed assessment of rapid number naming, has been studied as a rapid, quantitative screening tool for neurological dysfunction associated with concussion, hypoxia, Parkinson's disease, multiple sclerosis and extreme sleep deprivation. All studies to date have either compared within subjects against individual baselines or between affected subjects and age-matched neurologically-normal control subjects. A large normative database has not been previously established. The purpose of this study was to determine the distribution of K-D test performance in normal adults and adolescents and evaluate the effect of potential confounding variables.

## METHODS

In this cross-sectional, multi-center study, subjects ≥15yrs old with binocular best-corrected near visual acuity better than 20/30 completed two trials of the K-D test protocol. Exclusion criteria included concussion within 3-months, post-concussion syndrome, dyslexia or neuro-degenerative disorders. History of concussion, amblyopia, strabismus as well as demographic variables of education, race/ethnicity, gender and age were assessed by subject interview. Analysis of Covariance was used to determine if age, education, or race affected performance while controlling other confounders.

FIGURE 1. KING-DEVICK (K-D) TEST CARDS

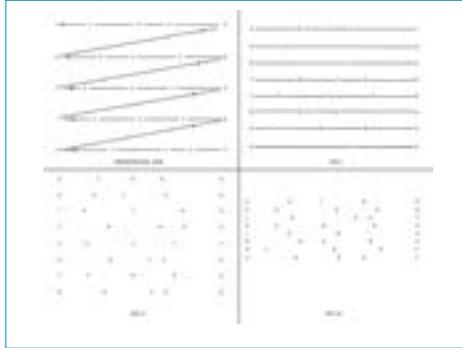


TABLE 2. K-D TIME IN DIFFERENT RACES

Race Groups	n	Mean (seconds)
Caucasian	542	41.06 ± 8.00 <sup>†</sup>
Hispanic	14	39.21 ± 6.26 <sup>§</sup>
African American	57	45.42 ± 9.70 <sup>§</sup>
Asian or Pacific	42	39.15 ± 8.11
Other	18	39.88 ± 5.68

<sup>†</sup>Comparison of Caucasian vs. African American: p ≤0.001

<sup>§</sup> Comparison of Hispanic vs. African American: p ≤0.001

TABLE 1. K-D TIME IN DIFFERENT AGE GROUPS

Age Groups	n	Mean (seconds)
15- <18 Years Old	95	41.18 ± 7.57 <sup>§</sup>
18- 30 Years Old	153	38.13 ± 6.66 <sup>§</sup>
30- <40 Years Old	97	38.13 ± 6.73
40- <50 Years Old	108	40.61 ± 7.65
50- <60 Years Old	123	43.23 ± 8.87
> 60 Years Old	115	45.98 ± 8.81 <sup>†</sup>

<sup>†</sup>p≤0.001

<sup>§</sup>p≤0.05

TABLE 3. K-D TIME IN SUBJECTS WITH DIFFERENT EDUCATION LEVELS

Education Groups	n	Mean (seconds)
Grades 9-11	115	41.27 ± 8.52
High School Graduate	73	41.97 ± 8.86
Some college or technical	182	42.50 ± 8.17
College graduate	186	39.81 ± 7.80
Post-College Training	131	40.33 ± 7.07

## RESULTS

Subjects (n=691, age: 39.8±17.7yrs, 58%female) were enrolled in 5 sites. Best K-D times were 41.2±8.2s. Difference between K-D trials was 3.1±3.3s. Performance did not vary by history of concussion, gender, amblyopia, or strabismus. Younger age, higher education and Caucasian or Hispanic race/ethnicity had better K-D times. K-D times were stable in the subjects aged ≤39yrs and demonstrated worsening ≥40yrs. African American race demonstrated significantly worse K-D times on average (45.42s ± 9.70) compared to Caucasian (41.06s±8.00, p ≤0.001) and Hispanic (39.21s ± 6.26, p≤0.001).

## CONCLUSIONS

The K-D test requires visual processing, saccades, language, attention, and has been proposed as a marker of integrated neurological function. We report normative data in a large adolescent/adult cohort and report associations with age, education and race. Knowledge of these confounding variables is important for design and interpretation of future K-D studies. Our database will have application to future studies of K-D test performance in neurologically diseased populations and potential future application to clinical settings.

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# NEUROSCIENCE

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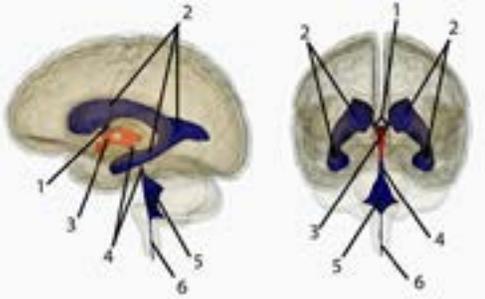
# The Interventricular Foramen – An Example of a Scientific Argument Carried Out in Public

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Illinois College of Optometry - Chicago, Illinois 60616

21.06 SA

## INTRODUCTION

Political correctness is a concept that is practiced by many in the early part of the 21<sup>st</sup> Century. This seems especially true for scientific writing, which is currently characterized by rare use of the first person, customary employment of the passive voice, and often, usage of qualifying phrases to soften statements. This was not always the case. One example where a scientific argument was personal and carried out in the open using publications concerned the interventricular foramen of Monro, which is a passage through which the lateral and third ventricles of the brain communicate.<sup>1</sup>



- 1. Interventricular Foramen
- 2. Lateral Ventricle
- 3. Third Ventricle
- 4. Cerebral Aqueduct
- 5. Fourth Ventricle
- 6. Central Canal

## ALEXANDER MONRO SECUNDUS (1733–1817)

Three generations of Monro's continuously held the prestigious Chair of Anatomy at the University of Edinburgh from 1720 to 1846: Alexander *primus*, Alexander *secundus*, and Alexander *tertius*.<sup>2,4</sup> Alexander *secundus* was perhaps the greatest of the three.<sup>5</sup> He first described the interventricular foramen in 1764.<sup>6</sup> In his most famous book: *Observations of the Structure and Functions of the Nervous System*, published in 1783, Monro gave a fuller account of this structure.<sup>7</sup> Here he was honest in pointing out that the presence of a communication between the ventricles was well known by others preceding him (even Galen). Monro then went on to claim that he described it in more detail than anyone before him and that prior descriptions had no value.<sup>6,8</sup> This appears to have given rise to challenges from his contemporaries, especially those in London. In response, Monro had eminent physicians write letters that would leave no doubt that he well deserved the acknowledgment,<sup>9</sup> and he wrote a second article in 1797 that substantiated and defended his anatomical descriptions of these communications.<sup>10</sup>



Fig. 1. The Interventricular Foramen. See the text.

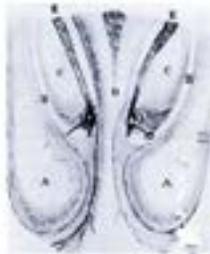


Fig. 2. The Interventricular Foramen. See the text.

## CHARLES BELL (1774–1842)

Charles Bell was another Scottish anatomist that came from a family of physicians.<sup>11</sup> He, along with his brother, John, was forced to leave Edinburgh for London because of political reasons. In 1802 he published a book, *The Anatomy of the Brain*, that has some of the most beautiful neuroanatomical renderings of the time.<sup>12</sup> In a famous drawing he showed the interventricular foramen but gave no credit to Monro. In fact in an appendix of this text, Bell specifically called Monro to task. This was the first published critique of Monro and was more of a personal attack on Monro for presuming to describe something that was already well known than an attempt to show that he was mistaken, although he actually was.

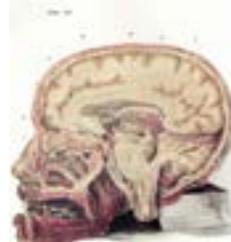


Fig. 1. The Interventricular Foramen. See the text.

## CONCLUSION

Monro described a direct communication between the lateral ventricles and a separate foramen opening into the third ventricle.<sup>7</sup> Subsequently it has been established that each lateral ventricle communicates separately with the third ventricle, so each brain has a pair of interventricular foramina, and there is no direct communication between the two lateral ventricles.<sup>13</sup> Essentially Monro was mistaken. Even though most of his vitriolic criticism of Monro was on a personal level, Bell was correct that Monro probably does not deserve the eponymous term since not only did he not discover the foramen, he incorrectly described it.



- 1. Interventricular Foramen
- 2. Lateral Ventricle
- 3. Third Ventricle
- 4. Cerebral Aqueduct
- 5. Fourth Ventricle

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# SPECIAL OLYMPICS

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# The Importance of Considering Vision and Eye Health in Children with Neurodevelopmental Anomalies (ID/DD)

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## SO HEALTHY ATHLETES

Launched in 1997, Healthy Athletes works to identify problems that may need additional follow-up.

- Despite a mistaken belief that people with intellectual disabilities (ID) receive the same or better health care than others, they typically receive sub-standard care, or virtually no health care at all.
- More than 136,000 health care professionals have been trained to provide improved care to millions with ID.



Special Olympics - Lions Clubs International Opening Eyes® (Opening Eyes)

The mission of Opening Eyes is to optimizing their vision, eye health and visual skills.

## OBJECTIVES

Provide screenings to Special Olympics athletes; educating athletes, parents and coaches about the importance of eye care; educating eye care professionals about the vision care needs of persons with ID; and increasing surveillance of visual and eye health of persons with ID.

Opening Eyes screening includes distance and near visual acuity, cover test (strabismus), color vision, autorefraction (refractive error testing), eye health (internal, external, and intraocular pressure).

## DATA

Since 2007 Opening Eyes has provided more than 134,514 assessments:

Age Group	N	%
8-17 years	53,439	39.7%
18-39 years	65,477	48.7%
40 and older	15,598	11.6%

What has the results shown about the vision of individual with Intellectual Disability?

- 10.1% - Strabismus (esotropia, exotropia, and hypertropia)
- 28.0% - Visual acuity poorer than 6/12
- 56.7% - Needed for lens correction (significant hyperopia ( $\geq 2.00D$ ) myopia ( $\leq 1.00D$ ) and astigmatism ( $\geq 1.00D$ ))
- Only 14, 561 reported having glasses ever

## CONCLUSION

Vision is important to evaluate since children may have vision problems.

Recommendations from National Center for Children's Vision & Eye Health.

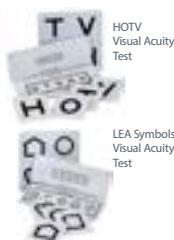
Children at high risk for vision disorders should be referred directly. This includes children with the following:

- Readily observable ocular abnormalities
- Neurodevelopmental disorders, preterm birth (<32 weeks)
- Systemic conditions that have associated ocular abnormalities
- First-degree relatives with strabismus or amblyopia
- Parents who believe their child has a vision problem

Children who do not have a neurodevelopmental problem should have vision screened as follows:

Best Practice PDR Visual Acuity Testing
Single crowded (logMAR) HOTV letters or Lea symbols surrounded by crowding bars at a 5-foot (1.5 meter) distance. A
Passing score - is 6/15 for 36 - 47 months & 6/12 - 48 to <72 months
Acceptable Practice
HOTV or LEA symbols at 20-foot (3 meter) test distance or for older children use this distance.
Best Practice for Instrument Based Screening
- Retinomax (Right MG, Co Ltd - Tokyo, Japan)
Acceptable Practice
- Plusoptix Photoscreener (Plusoptix - Nuremberg, Germany) or Welch Allyn Spot VS 100 (Welch-Allyn Inc., Skaneateles Falls, NY)

- Instrument-based screening refers to using automated technology (autorefraction or photoscreening) – it is quick and requires minimal cooperation and identifies the presence and magnitude of refractive error rather than providing a measurement of visual acuity.
- Abnormal refractive error is a significant risk factor for amblyopia or strabismus.
- Screening for refractive error alone is often successful in identifying children with constant strabismus and moderate to severe levels of amblyopia.



Plusoptix Photoscreener (Plusoptix - Nuremberg, Germany)



Welch Allyn Spot VS 100 (Welch-Allyn Inc., Skaneateles Falls, NY)