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# Pituitary Macroadenoma in a Pre-fixed Optic Chiasm with Incongruous Left Hemianopsia

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## BACKGROUND

- The anatomical variations of the optic chiasm and the size and shape of pituitary adenomas can elicit visual field defects that deviate from the classical bitemporal hemianopsia.
- The majority of the normal population have optic chiasms that lie directly above the diaphragma sellae, but a minority have post-fixed optic chiasms in which the chiasms are displaced posterior and lie over the dorsum sellae. A pre-fixed optic chiasm is an anterior displacement of the chiasm overlying the tuberculum sellae and is present in 10-15% of the normal population.
- A patient presents with an incongruous left hemianopsia secondary to a pituitary macroadenoma in a likely pre-fixed optic chiasm.

## CASE HISTORY

- A 79-year-old white male presented with complaints of reduced vision and loss of peripheral visual field.
- The veteran was diagnosed with a pituitary adenoma after work-up was performed following visual field defects detected by an outside eye doctor.
- The patient's medical history includes pituitary adenoma, schizophrenia, auditory loss, Barrett's esophagus, sinusitis, benign prostate hyperplasia, and osteoarthritis.
- MRI revealed an intrasellar, parasellar, and suprasellar 4.5 x 3.0 x 4.0 cm lobulated pituitary macroadenoma with compression of the optic nerve more on the right side (Figures 3-6). There were mass effects on the hypothalamus, corpus callosum, right carotid artery, right lateral ventricle, and third ventricle.
- Prolactin levels were slightly elevated at one point in time then returned to normal levels without treatment. TSH, GH, ACTH, cortisol, and testosterone levels were normal.

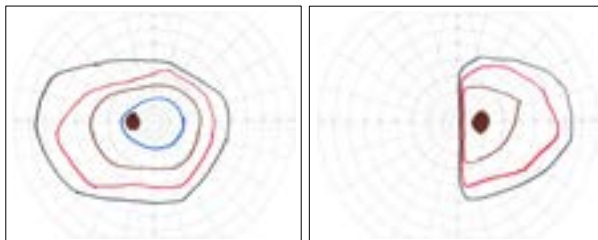


Figure 1. Goldmann visual field OS. Isopter colors: Black: V4e, Red: III4e, Brown: I4e, Blue: I3e.

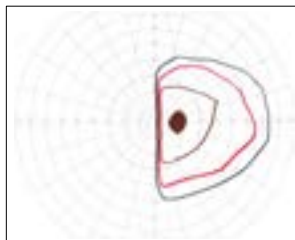


Figure 2. Goldmann visual field OD. Isopter colors: Black: V4e, Red: III4e, Brown: I4e.

## FINDINGS

- The BCVA OD and OS were 20/400 and 20/25, respectively. There was a 1+ RAPD OD. Ishihara color vision was reduced with 0/14 plates OD and 5/14 plates OS.
- The right optic nerve had 1+ diffuse and 3+ temporal pallor; the left optic nerve head was pink without pallor.
- Goldmann visual fields, which were reliable with good fixation, showed complete loss of nasal field OD and mild temporal visual field depression OS (Figures 1 and 2)

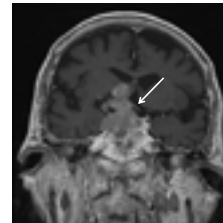


Figure 3. Pituitary macroadenoma in coronal MRI scan. White arrow: left optic tract.

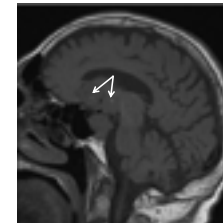


Figure 4. Pituitary macroadenoma in sagittal MRI scan. White arrows: micro infarcts within macroadenoma

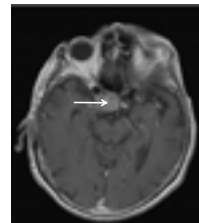


Figure 5. Pituitary macroadenoma in axial MRI scan.



Figure 6. Pituitary macroadenoma in axial MRI scan. Note the midline shift.

## TREATMENT

- Asymptomatic non-secreting pituitary adenomas may not require treatment and can be closely monitored. Secreting adenomas can alter hormone levels and may shrink with medical therapy such as dopamine agonists. Symptomatic adenomas and those that fail to respond to medical intervention or radiotherapy can be removed surgically, often through a transphenoidal route.
- This patient's non-secreting macroadenoma did not warrant medical therapy. However, the patient has repeatedly refused surgical intervention against medical advice. Follow-up visits showed stable decreased vision. Goldmann visual fields revealed slow, progressive overall constriction of right and left visual fields. Unfortunately, the patient's mental status limits his ability to perform visual field testing.

## CONCLUSIONS

- Pituitary adenomas are found to be present in approximately 17% of the population and are the most common intrasellar tumor in adults.
- Visual field defects and optic atrophy are ocular findings that may be present. Invasion of the cavernous sinus can cause cranial nerve palsies.
- Pituitary apoplexy is hemorrhaging or infarct within a pituitary adenoma. It is a rare occurrence but can be life-threatening if untreated.
- Post- or pre-fixed chiasms are present in 30% of the population so one must keep in mind that visual field defects can be highly variable in patients with pituitary adenomas and should be correlated with imaging.
- Pituitary adenomas may grow to a large size with varying morphologies that can invade different structures causing systemic and ocular problems that should be correlated with imaging.

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There was no conflict of interest.

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# Corneal epithelium defect due to rheumatoid arthritis and staphylococcal marginal keratitis

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## BACKGROUND

Rheumatoid arthritis (RA) is a common inflammatory condition that often affects small joints throughout the body especially the hands and wrists. Common ocular manifestations of RA are keratoconjunctivitis sicca, episcleritis, scleritis, and peripheral ulcerative keratitis. The diagnosis of RA includes four of the following criteria: morning stiffness lasting greater than 1 hour, swelling of the soft tissue of three or more joints, swelling of the soft tissue of three or more hand joints, symmetrical soft tissue swelling and subcutaneous nodules.

Staphylococcal marginal keratitis begins with an infiltrate that has an intact epithelium but may ulcerate with prolonged inflammation. Staphylococcal marginal keratitis is a hypersensitivity reaction to staphylococcal antigens. It is mostly occurs bilateral and occurs adjacent to the limbus with a clear zone of cornea between the lesion and the limbus. The presence of chronic staphylococcal bacteria on the lid margins is thought to trigger an immune response in a sensitized cornea. The immune response is most likely a type III hypersensitivity reaction in complex deposition in the peripheral cornea.

Keratoconjunctivitis sicca also known as dry eye syndrome is divided into two categories based on etiology, which are aqueous deficient and evaporative dry eye. There are many causes of evaporative dry eye including but not limited to meibomian gland dysfunction/posterior blepharitis, contact lens wear, environmental factors, usage of certain medications, aging, or lagophthalmos. K. sicca has long been recognized to have a largely inflammatory component, with inflammation occurring in the meibomian glands which leads to inflammation on the ocular surface.

Dry eye disease is very common in the US, affecting approximately 25%-30% of the population.

## CASE DETAILS

A 61 year old Hispanic female presented to the Urgent care clinic with complaints of severe burning, pain, redness, and irritation in both eyes, with the right eye more affected than the left eye for the past three days. The patient's medical history is positive for hypertension, diabetes, and rheumatoid arthritis.

## EXAMINATION

Figure 1: Initial Visit- Slit lamp examination

Meibomian Gland Dysfunction 2+, Lid notching	Lids/Lashes	Meibomian Gland Dysfunction Lid notching
1+ injection	Conjunctiva	1+ injection
2+ injection	Sclera	2+ injection
1.5 sup/nasal epithelium defect 0.5 mm infiltrate sup/nasal Micropannus and neovascularization with scarring 360 Large diffuse confluent SPIK	Cornea	Micropannus and neovascularization 360 Mild peripheral corneal scarring Large confluent SPIK concentrated inferiorly

PICTURE 1:

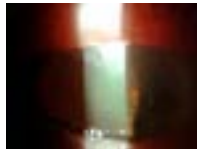


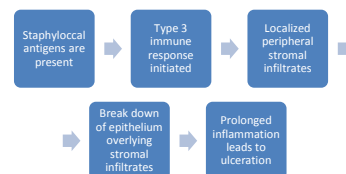
Figure 1 shows the findings of the slit lamp examination.

Picture 1 shows superior nasal aspect of the patient's right eye.

PICTURE 2:



Figure 2: Process leading to corneal thinning/ulceration from staphylococcal hypersensitivity



## DIAGNOSIS

Due to right eye's superior corneal epithelium defect, the patient was diagnosed as having Sclerosing keratitis with a concomitant keratoconjunctivitis sicca and staphylococcal marginal keratitis which was secondary to her rheumatoid arthritis. Once the corneal epithelium defect healed with the mentioned management plan, the patient has been followed for several weeks in order to successfully treat the concomitant keratoconjunctivitis sicca and staphylococcal marginal keratitis. The pathogenesis of how chronic staphylococcal hypersensitivity can lead to ulceration is shown in Figure 2. As is the case of this patient there are several concomitant diagnoses, as Figure 3 shows there is a long list of differential diagnoses of a patient that presents with peripheral corneal thinning. However, all the patient's ocular diagnoses are inflammatory and immune mediated, so the root of the cause must be controlled in order to adequately provide relief for the patient.

Figure 3: Differential Diagnoses of Peripheral Corneal Thinning/Ulceration

Keratoconjunctivitis Sicca
Staphylococcal Marginal Keratitis
Connective tissue disease, ie Rheumatoid Arthritis
Mooren ulcer
Keratoconus/Pellucid Marginal Degeneration
Terrien Marginal Degeneration
Ocular Rosacea
Peripheral Ulcerative Keratitis

Figure 4: Management

Week 1	Doxycycline 50 mg BID Lotemax BID Erythromycin ointment NP Artificial Tears q1h
Week 2-3	Doxycycline 50 mg BID Lotemax BID Refresh pm ung
Week 3-4	Doxycycline 50 mg BID Restasis BID Refresh pm ung NP Artificial Tears
Week 5-6	Continue prescribed regimen Scleral lens fitting

## RESULTS

The management plan is shown in Figure 4. Initially the patient was put on Doxycycline in order to help control the lid inflammation, the non-preserved artificial tears and erythromycin ointment to help lubricate the eyes, and the Lotemax was to help the infiltrate dissipate and epithelial defect heal. After the epithelial defect healed, the patient was switched to Restasis to help further control long term inflammation. At this time, the patient is continuing the prescribed ocular medications and will ultimately be fit in a scleral contact lens. A scleral contact lens will serve to lubricate the eyes by vaulting the cornea which creates a tear filled vault and further preserves corneal integrity. The patient is currently receiving weekly Enbrel injections to treat her severe rheumatoid arthritis, due to the prolonged inflammation; the patient has been informed that further treatment by her primary care practitioner may need to be done to control her RA.

## CONCLUSION

Patient's with staphylococcal marginal keratitis often present with redness, pain, foreign body sensation, and photophobia. It is important to rule out potentially devastating vision causing disorders such as peripheral ulcerative keratitis and mooren's ulcer. Copious patient education is important because ocular manifestations will wax and wane based on the systemic condition. A scleral lens is a long term treatment plan for this patient in order to insure comfort. Overall, the goal is to increase patient comfort throughout the day. It is imperative to thoroughly educate each patient about their ongoing condition, while tailoring treatment options based on signs observed.

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Special thank you to Dr. Jennifer Harthan

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# Effect of Room Illumination on Manifest Refraction and Patient Preference

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## PURPOSE

During observation of student exams at the Illinois College of Optometry (ICO), various levels of room lighting were being utilized during the manifest refraction, ranging from full brightness to total darkness. The purpose of our study was to determine if illumination significantly affected manifest refraction. Subjective preference on the illumination condition was also investigated.

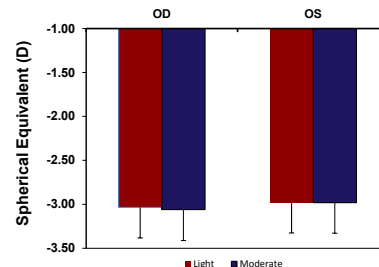
Figure 1: Post Refractions Questionnaire

- Under which lighting condition were you most comfortable:
  - Dim
  - Bright
  - No difference
- If there was a difference in comfort between the lighting conditions, please select all reasons that apply:
  - Greater amount of light
  - Less amount of light
  - Greater amount of glare
  - Less amount of glare
  - Greater contrast
  - Less contrast
- Under which condition did you feel your vision was the most clear:
  - Dim
  - Bright
  - No difference

## METHODS

Seventy-one subjects free of any significant ocular disease were recruited from the staff, student and faculty of ICO and refracted by two doctors in adjoining exam rooms. One exam room was brightly illuminated (overhead room lights on) while the other room was moderately lit (overhead room lights off, dimmable pocket lights on behind the exam chair). Illuminance was measured prior to each examination with a Sekonic L-758Cine DigitalMaster Light Meter. The brightly lit room was set to 320 lux, and the dim room to 3.5 lux. Subjective manifest refractions were performed in both rooms with an M & S Smart System 2020 computerized visual acuity chart. The identical refractive procedure was performed by each examiner. Nidek autorefractometer measurements were used as the starting point for both refractions. Pupil sizes were measured prior to each refraction with an infrared Colvard pupillometer. All subjects were asked to complete a 3-question survey post-examination to evaluate subjective preference. Paired T-test was performed to compare the spherical equivalent (SE) refraction and pupil sizes of right and left eyes in the bright illumination to the SE refraction and pupil sizes in the moderate illumination.

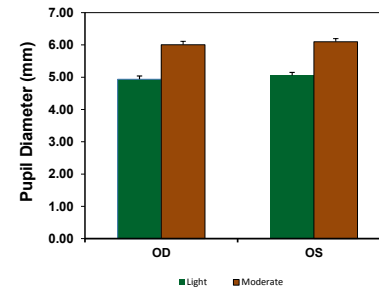
Figure 2: Spherical Equivalent Refractions in Bright and Moderate Illumination



## RESULTS

No significant difference was found in the SE refraction between the two illuminations for either eye (OD:  $p=0.40$  and OS:  $p=0.92$ ). Average SE was  $-3.04$  in bright illumination and  $-3.06$  in moderate illumination, OD, and  $-2.99$  in bright illumination and  $-2.98$  in moderate illumination, OS. As expected, pupil size in the two illuminations was significantly different ( $p<0.001$ ). There was no patient preference for light level when evaluated for comfort (29% preferred bright, 36% preferred moderate, 36% had no preference), or clarity of vision (29% preferred bright, 39% preferred moderate, 33% had no preference).

Figure 3: Pupil Sizes in Bright and Moderate Illumination



## CONCLUSION

The results indicate that subjective manifest refractions do not significantly differ whether they are obtained in bright or moderate room illumination. In addition, patients have no preference for one lighting condition over the other. These findings may impact how this procedure is taught in an optometric curriculum or performed in daily clinical practice.

Figure 4: Patient Preference for Comfort in Bright and Moderate Illumination

### Subjective Response: Comfort

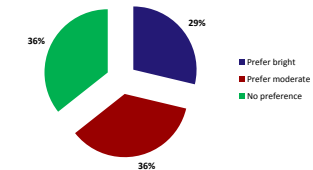
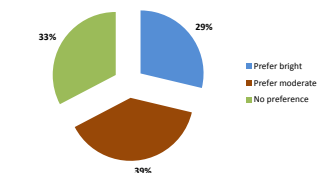


Figure 5: Patient Preference for Vision in Bright and Moderate Illumination

### Subjective Response: Clarity of Vision



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# Progressive Optic Neuropathy in Systemic Lupus Erythematosus



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## INTRODUCTION

Approximately one third of patients with systemic lupus erythematosus (SLE) have ocular manifestations ranging from dry eyes to optic neuropathies. Of these, optic neuropathy is a rare finding and includes optic neuritis and ischemic optic neuropathy. A case of progressive ischemic optic neuropathy presents in a patient with work-up and symptoms indicating an active autoimmune disease process.

## CASE HISTORY

A 65-year-old African American male presented with complaints of recurrent painless, gradual vision loss OS. His ocular history consisted of optic neuropathy OS with previously decreased vision of 20/70, reduced red-green color vision, and trace RAPD. A work-up was performed at the initial presentation in 2010 with the results summarized in Table 1. The positive autoimmune lab results led to a rheumatology referral where the patient was deemed likely to have undiagnosed and subclinical SLE. He had no other clinical symptoms of SLE. He was treated with Cellcept, an immunosuppressant and his vision and ocular presentation remained stable.

His medical history consists of HTN, SLE, hyperlipidemia, anemia, multiple aortic dissection surgeries with complications resulting in a St. Jude aortic valve replacement, COPD, BPH, gout, and treatment for venereal disease in 1967.

**Table 1. Original Lab and Imaging Work-Up**

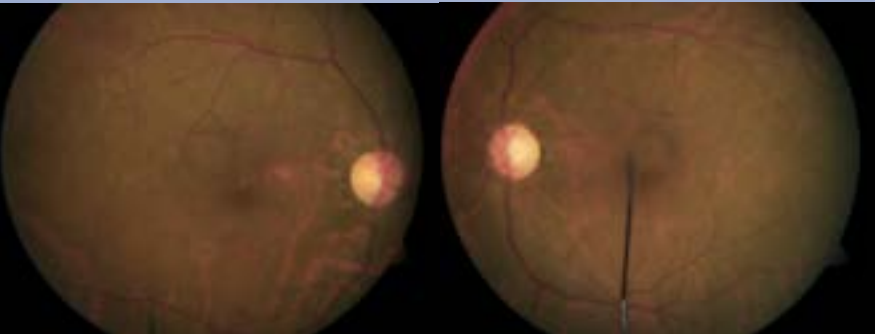
Test	Results
Blood work-up	<b>Elevated/Positive:</b> ANA, anti-SSA, anti-SSB, anti-Sm, anti-RNP, CRP, MHA-TP, polyclonal hypergammaglobulinemia, hyperalbuminemia, and hyperproteinemia <b>Negative:</b> anti-cardiolipin antibodies, anti-dsDNA, ESR, ACE, RPR, lysozyme, Rf
CT head/orbits	No mass effect or evidence of MS *MRI contraindicated due to St. Jude valve
Carotid duplex	No evidence of hemodynamic carotid or vertebral disease

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Thank you to everyone who helped review and edit the contents of this poster. There were no conflicts of interest.

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**Figure 1.** Posterior pole fundus photos. Note optic nerve head pallor OS.

## FINDINGS

TEST	OD	OS
BCVA	20/20	20/200
Pupils	+ Direct & consensual	+ Direct & consensual, 1+ RAPD
EOMS	Full, unrestricted	Full, unrestricted
Ishihara color vision	14/14	5/14 9/14 (2010)
Anterior Seg IOP	WNL	WNL
Posterior Segment	10-13 mmHg	10-12 mmHg
HVF 24-2 (Figure 2)	Question of trace diffuse ONH pallor, (-) ONH edema	1+ diffuse ONH pallor, greater temporally, (-) ONH edema
Updated lab results	Full	Inferior arcuate defect; superior edge defects
Normal levels of anti-SSA/SSB, anti-dsDNA, anti-RNP, and CRP. However, anti-Sm antibodies level was markedly elevated to 32.08 EU (reference range 0-19)		
The patient also began developing skin rashes despite the use of Cellcept. Rheumatology switched to hydroxychloroquine, which resolved his skin rashes and mildly improved his vision to 20/100.		

**Figure 2.** HVF 24-2 inferior arcuate OS (left), ess full OD (right)



## DISCUSSION

Optic neuropathy as the first presenting sign of SLE is a rare finding. It is estimated that 1-2% of SLE patients will have optic neuropathy. Its presentation can vary greatly and in the case of ischemic optic neuropathy, can present with painless vision loss, optic disc edema, arcuate visual field defects, and have unilateral or bilateral presentations. Visual acuities can also range with studies indicating 37-61% of patients having VAs of 20/200 or worse.

SLE ischemic optic neuropathy can occur when immune complexes deposit in the small vessels supplying the optic nerve, which leads to focal thrombotic changes and optic nerve head ischemia. This ischemic event is believed to cause subsequent demyelination and axonal necrosis.

A unique feature of this case is the association between the elevated levels of anti-Sm antibodies and optic neuropathy progression. Anti-Sm antibody levels along with a positive ANA plays an important role in diagnosing SLE and indicating disease activity. Anti-Sm antibodies are seen in 5-30% of SLE patients and have high specificity for SLE, making it one of the diagnostic criteria for the disease. Our patient had elevated levels of anti-Sm antibodies and new skin rashes at the time of his optic neuropathy progression, likely correlating with an active systemic and ocular disease process.

Typical management of ischemic optic neuropathy involves monitoring and reducing risk factors that can cause recurrences. In our case, this required co-management with rheumatology in treating the patient. During an acute phase, systemic corticosteroids, and even IV steroids in severe cases, may be used in addition to immunosuppressant therapy.

## CONCLUSION

- Optic neuropathy in SLE is a rare finding and can occur in 1-2% of patients with SLE.
- The visual prognosis can vary with up to 61% of patients ending with 20/200 or worse.
- Positive ANA and Anti-Sm antibody levels have a large role in diagnosing SLE and may be contributing to active ocular disease in our patient.
- Co-management with different medical specialties in maintaining a patient's overall systemic health is crucial in preventing vision loss.

## DIFFERENTIAL DIAGNOSIS

Diagnosis	Supporting	Not supporting
SLE associated ischemic optic neuropathy	Positive lab work-up. SLE criteria met: urine abnormality, anemia, skin rashes, positive ANA, positive anti-Sm antibodies. Responded and improved with SLE treatment	Negative labs: anti-cardiolipin antibodies, anti-dsDNA, ESR. Asymmetric presentation versus unilateral presentation.
Post-operative posterior ischemic optic neuropathy	History of complicated aortic dissection surgery prior to first noted vision decrease	Recurrence 2 years post surgical intervention
Optic neuritis	Unilateral presentation and positive autoimmune disease work-up	No evidence of MS on imaging.
Luetic Optic Neuropathy	Positive MHA-TP	Time frame inconsistent & negative RPR

## DIAGNOSIS & MANAGEMENT

Given the pain-free course, positive autoimmune disease work-up, and improvement in symptoms with SLE treatment, our patient was diagnosed with progressive ischemic optic neuropathy most likely secondary to an autoimmune disease process likely SLE. The patient's vision remained relatively stable on Plaquenil. We are continuing to monitor the patient closely and will continue co-management with rheumatology.

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SPECTRIOS INSTITUTE  
for Low Vision

# A Ciliopathic Syndrome initially identified as Retinitis Pigmentosa alone

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## BACKGROUND

Bardet-Biedl syndrome is a rare, genetically heterogeneous condition occurring in between 1:100K and 1:160K people. The autosomal recessive ciliopathy causes retinitis pigmentosa, obesity, post-axial polydactyly, renal and cardiac abnormalities, learning disabilities, and hypogonadism, among other systemic manifestations.

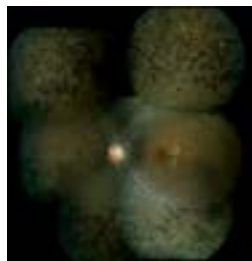
## CASE SUMMARY

An 11 year-old African-American male who received a diagnosis of retinitis pigmentosa two years prior by another ophthalmologist presented for a comprehensive eye examination. He had subjective complaints of worsening nyctalopia and peripheral field constriction but otherwise reported having normal central and color vision. His medical history included his 5 week prematurity with no other renal or cardiac history.

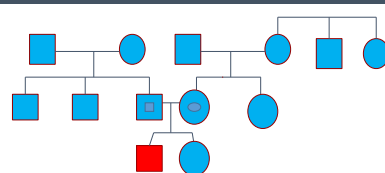
Upon gross observation, the patient was overweight and presented with scars on the ulnar side of each hand and also post-axially on his left foot. The patient revealed he had a history of polydactyly with subsequent removal of the additional digits. His ocular history was significant for bilateral isometric amblyopia. His best corrected visual acuity was 20/60 in each eye. No other individuals in his family

pedigree were affected with retinal disease, and any history of consanguinity was denied. Upon dilated fundus examination, the patient showed typical signs of retinitis pigmentosa including 360 degrees of bone spicule hyperpigmentation and clumps of hypopigmentation, waxy optic disc pallor, and mild vessel attenuation in each eye. There was relative foveal sparing without any sign of a large atrophic macular lesion in either eye. Electrophysiologic testing revealed a non-detectable rod and cone response on the full-field electroretinogram of each eye.

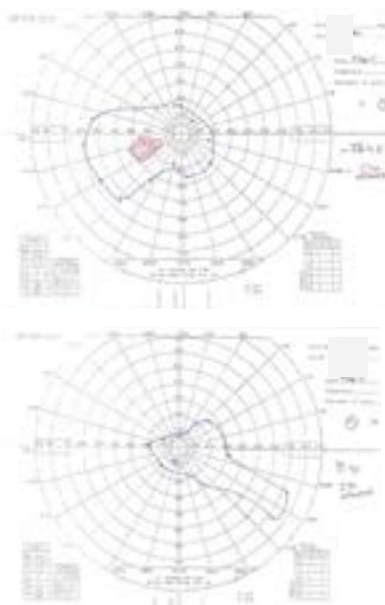
After being referred back to his internist to rule out any other systemic abnormalities, the patient was found to show signs of hypogonadism. No other significant findings were confirmed after being referred for a cardiology and nephrology consult. Based on the



### Pedigree Analysis



### Goldmann Visual Field



patient's retinal findings, polydactyly, obesity, and hypogonadism, a diagnosis of Bardet-Biedl syndrome was made. The patient was educated on the diagnosis, prognosis, and option for genetic testing for BBS mutations. He was sent for a low vision rehabilitation consultation.

## CONCLUSION

This case represents the importance of completing a thorough review of systems and medical history with pediatric populations presenting with inherited retinal disease, as to not overlook syndromes like Bardet-Biedl in obese pediatric patients. Signs of obesity and post-axial polydactyly presenting with retinitis pigmentosa are key signs for making the diagnosis.

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# Diagnosis and Management of Ring Scotoma Associated With Macular Degeneration

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## BACKGROUND

Ring scotomas often result in well preserved objective visual acuity with disproportionate subjective reports of significant difficulty with daily living tasks. They are reportedly found in 13% to 17.4% of patients with AMD.<sup>1-3</sup> Traditional low vision (LV) rehabilitation strategies for macular degeneration (AMD) often rely on high powered magnification; however these strategies often fail to improve functionality in patients with ring scotoma. Failing to diagnose a ring scotoma can result in frustrated patients and clinicians. Several tests are available to map central and paracentral scotomas and/or evaluate fixation preferences. It is important to accurately assess scotomas in order to prescribe appropriate rehabilitation.

## CASE DISCUSSION

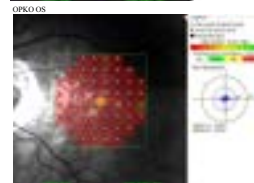
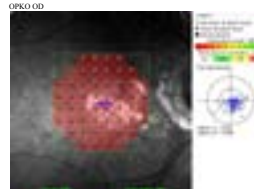
- LV follow-up exam of 81 year old Caucasian female
- CC: Continued severe difficulty reading despite previous LV interventions. Good distance acuity and continued driving without difficulty.
- POHx: AMD OU Exudative s/p Avastin x 7OD, Atrophic OS with ERM OS
- Previous Devices: +2.75 Add for reading, CCTV "not helpful", task light "most helpful"
- DBVA 10/100 OD, 10/25 OS, 10/20 OU NBVA 0.3M/0.8M OU slow
- Functional DVA Slow10/80, Fast 10/40, Slow 10/20, Near 0.1M/2M fast
- MARS Contrast sensitivity 0.36 log Mar, Profound deficit
- Saccadic dysfunction, 2+ undershoots and overshoots
- Pursuits and EOM's wnl.
- SLX and DFE: Consistent with AMD diagnosis with optical changes OD>OS, ERM OS and Pseudophakia OU
- Assessment: Probable Ring Scotoma OS, Central Scotoma OD 2o AMD
- Management: Order OPKO OCT-SLO Microperimetry and California Central Visual Field Test (CCVFT) then evaluate LV devices for individual rehabilitation plan

**FIGURE 1:** Visual acuity was evaluated at distance and near using suprathreshold and subthreshold measures. Testing in this manner can reveal a slow acuity on supra and sub threshold measures indicating a pattern typical of ring scotoma.



**FIGURE 2:** Microperimetry/ Fundus Monitored Perimetry allows a real time retinal image or OCT to be viewed during visual field testing, to allow correlation between the retina and visual function. The OPKO OCT-SLO is such a device. Other devices include the Nidek MP-1 and the MAIA.<sup>4</sup>

OPKO OCT-SLO OD confirms complete central scotoma and poor fixation without a preferred retinal locus (PRL). OPKO OS shows a response to a 2dB stimulus with paracentral fixation and surrounding absolute scotoma.



**FIGURE 3:** The CCVFT is an inexpensive, quick to administer, objective (fixation tracking) and subjective clinical test that may be used to detect and map central scotomas. This test is ideal for patient education as they can visualize the scotoma once mapped. As the test is done at a near working distance, you may overlay this on printed material to demonstrate to patients and family effects of the scotoma on reading.<sup>5</sup>



**FIGURE 4:** CCVFT OS shows good correlation with automated OPKO OS confirming ring scotoma.



## LOW VISION DEVICE & MANAGEMENT PLAN

Figure 5a: Education and scotoma awareness – We demonstrated the scotoma and reviewed risk of visual deterioration. Ring scotomas are reported to progressively deteriorate in up to 50% of patients.<sup>6</sup>

Figure 5b: Lighting - A clip on spectacle light for portable increased luminance was prescribed in addition to continued use of stand lamp. A yellow typoscope/bar magnifier was also used for scotoma management and functional contrast enhancement.

Figure 5c: Mild Magnification – +5.00D prismatic spectacles achieved .4M and a 1.5x bar magnifier were preferred and prescribed. Occupational therapy training with scrolling material using the patients CCTV X-Y table on lowest magnification setting to reduce saccadic eye movements was ordered.

At follow-up the patient reported a moderate decrease in frustration and much greater acceptance of her vision loss.

**FIGURE 5a, 5b, 5c:**



## CONCLUSIONS

- Given its prevalence, when subjective complaints of ADL difficulty are disproportionate to measured acuity ring scotoma should be considered as a differential
- Appropriate initial diagnosis of ring scotoma is imperative to rehabilitation success.
- Microperimetry is useful in correlating retinal structure and function when mapping scotomas
- The CCVFT is clinically useful and helpful in practices where access to microperimetric studies is limited
- Measurements of acuity, contrast sensitivity and saccades are important in the assessment and management of ring scotoma.
- Rehabilitation strategies for a ring scotoma should include education, scotoma awareness training, increased lighting and low magnification.<sup>7,8</sup>

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Financial Disclosures: None

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# Stickler Syndrome: Overview and Fundamentals of Patient Care

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## INTRODUCTION

Stickler Syndrome (hereditary arthro-ophthalmopathy) is an autosomal dominant genetic disorder that affects connective tissue, specifically collagen tissue. There are three main classifications to the syndrome. Types 1 and 2 present with the classic ocular and systemic features. Type 1 is the most common. Type 3 demonstrates only systemic findings and no ocular manifestations. Systemically this disease causes facial, oral and skeletal anomalies as well as deafness. Ocular findings include high myopia (non-progressive), cataracts (pre-senile), glaucoma and retinal detachment.

## CASE REPORT

A 9-year-old Hispanic female presents with complaints of blurry vision in her left eye at distance and near since she experienced a non-traumatic retinal detachment one year ago. Her medical and ocular history is significant for Stickler Syndrome and a retinal detachment in her left eye. Her spectacle prescription is -9.75-0.75x175 and -9.75-1.25x177. Entering visual acuities are 20/30-1 in the right eye and counting fingers improving to 20/800 with pinhole in the left eye. Pupil is slow to react in the left eye and there is a superior defect on confrontation fields. Retinoscopy and auto-refraction with trial frame refraction was performed. Her best corrected visual acuities are 20/25-2 in the right eye with -9.50-1.25x180 and 20/150 in the left eye with +8.25-0.75x180. Dilated fundus exam in the right eye shows peripheral laser scarring. In the left eye she is aphakic and has dense laser scarring and fibrosis in the periphery.

## RESULTS

The amount of anisometropia present is too large for spectacle correction, thus contact lenses are the first line of treatment. At the contact lens fitting, extended wear silicone hydrogel lenses were chosen. With a -8.00D lens, visual acuity is 20/30- in the right eye. With a +4.50D lens and a +3.50D over-refraction, visual acuity is 20/125 in the

left eye. The final contact lens prescription is -7.50D for the right eye and +6.00D for the left eye with Air Optix Night and Day lenses. The remaining refractive error, -2.25-1.25x180 and +2.25-0.75x180 is placed in polycarbonate spectacles. This will encourage her to wear the glasses for protection. Occlusion therapy of

the right eye is a future option to promote improved vision in her left eye. The patient will return for a three month follow-up, then yearly dilated fundus evaluations. She was educated on the signs and symptoms of retinal detachments.

## CONCLUSION

Stickler Syndrome is the most common inherited disorder to cause rhegmatogenous retinal detachments in children, most occurring within the first decade of life. The detachments result from giant retinal tears presenting as a circumferential break at the pars plana from the splitting of the posterior hyaloid membrane. Over 50% of patients with this syndrome will suffer from a retinal detachment and almost 70% of those will be affected bilaterally. It is imperative for children of family members with this syndrome to have a comprehensive eye exam at an early age due to the gene's strong penetrance. Affected parents have a 50% chance of passing it to their offspring. Patients with this disease should have annual dilated eye examinations. Early and even prophylactic treatment along with education is the key for managing this syndrome.

Table 1: Classifications of Stickler Syndrome

Type	Gene	Clinical Findings
Type 1	COL2A1	Vitreoretinal degeneration (optically empty vitreous), retinal detachment, cataracts, myopia, mid-facial hypoplasia, cleft palate, deafness, osteoarthritis (early onset), hypermobility of joints and mitral valve prolapse
Type 2	COL11A1	Vitreoretinal degeneration (fibrillary and beaded appearance), retinal detachment, cataracts, myopia, cranial abnormalities, cleft palate, deafness, osteoarthritis (early onset) and hypermobility of joints
Type 3	COL11A2	Mid-facial hypoplasia, cleft palate, deafness, osteoarthritis (early onset) and short stature

Table 2: Clinical Findings of Stickler Syndrome

Ocular Findings	Systemic Findings
High myopia -non-progressive Vitreoretinal degeneration Presenile cataracts -wedge or fleck opacities, non-progressive Retinal detachment -50% within first decade Glaucoma -angle anomaly, 5-10% of cases Ectopia lentis -uncommon	Facial anomalies - mid-facial hypoplasia - depressed nose bridge - short nose Oral anomalies -cleft and high-arched palates - bifid uvula Skeletal anomalies - spondyloepiphyseal dysplasia - joint hypermobility - osteoarthritis Deafness

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# A Comparison of the Binocular Vision Assessment Program to Outcomes from a Visual Efficiency Examination

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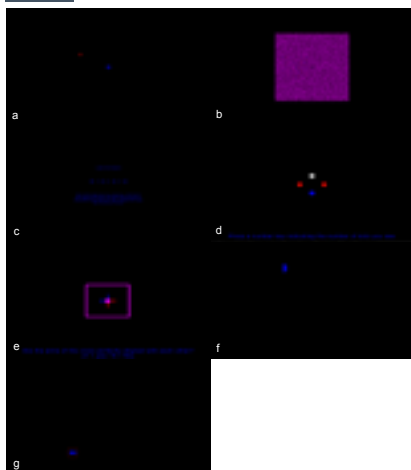
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## BACKGROUND

Binocular vision, accommodative and oculomotor dysfunctions are frequently encountered in optometric practice, second only to that of refractive error.<sup>1-4</sup> These disorders have been associated with a decreased quality of life and poor academic performance. Symptoms noted by those with the disorders range from asthenopia and headaches to diplopia and decreased reading comprehension.<sup>5,6</sup> Given their prevalence and the impact they may have on daily living and learning, it is important to accurately and efficiently assess these functional disorders in optometric practice and vision screenings so patients can seek appropriate care. *Binocular Vision Assessment* software (BVA) by Home Therapy Solutions is used by optometrists in-office to screen for and identify binocular vision, accommodative and oculomotility disorders. BVA claims it is an excellent pass/fail assessment to use in-office or on location at schools, and learning centers or health fairs.<sup>7</sup> The purpose of this study is to compare the *Binocular Vision Assessment* as a screening tool to a standard visual efficiency exam (VEE).

Figure 1 BVA test screens: a) Lateral and Vertical Phorias; b) Positive and Negative Fusional Vergences; c) Monocular Accommodative Facility; d) Worth 4 Dot; e) Fixation Disparity f) Pursuits; g) Saccades

Fig 1a-g



## METHODS

Binocular, oculomotor and accommodative functions were measured in 39 optometry students between the ages of 18 and 30 using both the BVA and a standard VEE. Exclusion criteria included any history of strabismus, amblyopia or extra ocular muscle palsy. The BVA program was configured using a 13-inch MacBook Pro monitor with brightness set to full and subjects positioned 40 cm from screen for consistency. The BVA measured heterophorias, vergences, monocular accommodative facility, Worth 4 dot, fixation disparity, saccades, and pursuits (Figure 1a-g). The VEE also included tests for these seven functions. Data was assessed using Pearson correlations and Bland Altman analysis for heterophorias, vergences and monocular accommodative facility. The data for saccades, pursuits, Worth 4 Dot and fixation disparity was evaluated using the Fisher's exact test.

Figure 2a: Regression analysis indicated fair correlation between BVA and each of the three standard phoria measurements. b: Bland-Altman Plots for horizontal phorias show fair agreement between BVA and both Von Graefe and Cover Test measurements, but poor agreement between BVA and Modified Thorington.

Figure 3a: Regression analysis for NFV break indicates fair correlation between BVA vergences and both Prism Bar and Risley Prism vergences. b: Bland-Altman Plots for NFV break shows poor agreement between Prism Bar and BVA, but fair agreement between Risley Prism and BVA.

Figure 4a: Regression analysis for NFV recovery indicates fair correlation between BVA vergences and both Prism Bar and Risley Prism vergences. b: Bland-Altman Plots for NFV recovery shows poor agreement between Prism Bar and BVA, but fair agreement between Risley Prism and BVA.

Figure 5a: Regression analysis for PFV break shows poor correlation between Prism Bar vergences and BVA, but fair correlation between Risley Prism vergences and BVA. b: Bland-Altman Plots for PFV break indicate poor agreement between both Prism Bar vergences and Risley Prism vergences as compared to BVA.

Figure 6a: Regression analysis for PFV recovery shows poor correlation between Prism Bar vergences and BVA, but fair correlation between Risley Prism vergences and BVA. b: Bland-Altman Plots for PFV recovery indicate poor agreement between both Prism Bar vergences and Risley Prism vergences as compared to BVA.

Figure 7a: Regression analysis comparing Monocular Accommodative Facility testing to the BVA indicates fair correlation between the two methods. b: Bland-Altman Plot for Monocular Accommodative Facility shows fair agreement between the BVA and standard method.

Fig 2a

Fig 2b



Fig 3a

Fig 3b



## RESULTS

■ The BVA showed fair correlation for assessing horizontal heterophorias as compared to objective and subjective phoria measurements (Figure 2a), but only poor to fair agreement.

■ Bland-Altman plots reveal that the BVA tends to underestimate the magnitude of exophorias and esophorias (Figure 2b).

■ There is fair correlation between BVA and prism bar/Risley Prism methods for measuring negative fusional vergence (NFV) and positive fusional vergence (PFV) break/recovery (Figures 3a-6a) with the exception of positive fusional prism bar vergences showing poor correlation.

■ Overall, the high mean difference and broad limit of agreement on Bland-Altman plots indicates poor agreement between the BVA and VEE on NFV and PFV break/recovery (Figures 3b-6b).

■ Monocular accommodative facility measured by the BVA and VEE has fair correlation and agreement (Figures 7a-b).

■ The Fisher's Exact Test indicated no significant difference between the two methods in assessing fixation disparity, Worth 4 dot, saccades and pursuits.

Fig 4a

Fig 4b



Fig 5a

Fig 5b



Fig 6a

Fig 6b



Fig 7a

Fig 7b



## CONCLUSION

This study demonstrated that non-eye care professionals might be able to use the *Binocular Vision Assessment* as a screening tool to determine the presence of binocular, oculomotor and accommodative disorders in a young-adult population. In general, if they failed the BVA program, a functional vision problem is present and should be referred for further diagnosis and treatment.

However, caution should be used when assessing numerical values for heterophorias, vergences and monocular accommodative facility collected from the BVA. These values are not directly comparable with those found during a standard clinical assessment. The BVA tends to underestimate the magnitude of horizontal heterophorias (both exophorias and esophorias) and overestimate negative fusional vergence recoveries. The BVA may also serve great purpose in the hands of optometric clinicians and paraoptometric professionals. BVA is simple to administer and patient instructions are clear, making it an easy and accurate screening test. The only drawback is that it can take up to 10 minutes to complete, which may pose a problem in some office and/or screening settings. Further research is needed to assess the repeatability and reliability of the BVA on children, special populations, and those with known binocular vision disorders.

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# Avellino Dystrophy and Recurrent Corneal Erosions – A Scleral Solution

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## CASE HISTORY/ INTRODUCTION

A 34 year old African American female presents to the urgent care clinic complaining of pain and decreased vision in the right eye more than the left. Ocular history is significant for a previous diagnosis of Avellino dystrophy in both eyes. Family history indicates that three of the patient's children also have Avellino dystrophy.

Figure 1: Corneal amyloid and hyaline deposits, in parallelepiped (right eye)

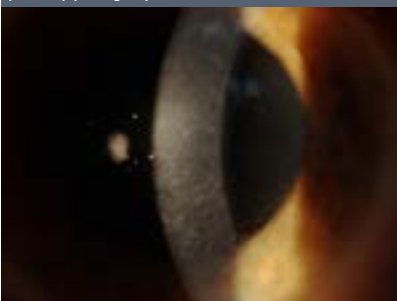


Figure 2: Corneal amyloid and hyaline deposits (left eye)



## OCULAR EXAMINATION

On clinical examination, the patient had decreased acuity in both eyes, right greater than left (20/100 OD, 20/60 OS). Slit lamp examination revealed corneal amyloid and hyaline deposits OU [Figures 1 and 2]. Sodium fluorescein staining of the cornea showed central epithelial erosions OU with adjacent areas of negative staining [Figures 3 and 4]. The epithelium was noted as loose and irregular with underlying stromal haze. Also noted was 1+ conjunctival injection. The posterior segment was not assessed.

Figure 3: Sodium fluorescein staining of the right eye at initial presentation

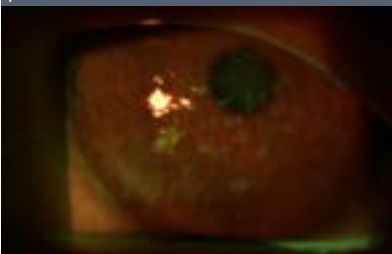
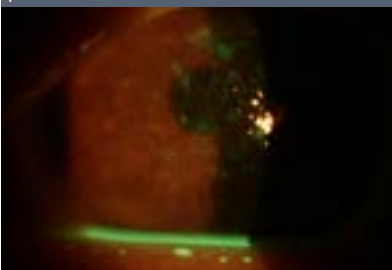


Figure 4: Sodium fluorescein staining of the left eye at initial presentation



## RESULTS

The previous diagnosis along with the clinical examination led to a presumed diagnosis of recurrent corneal erosion secondary to a congenital corneal dystrophy. The acute corneal condition was treated with antibiotic drops QID OU, artificial tears OU, and bandage soft contact lenses OU. At follow-up, the patient had developed severe corneal edema and a mild anterior chamber reaction secondary to bandage soft lens-induced hypoxia. The lenses were removed, and a cycloplegic, steroid (Q1h) and oral doxycycline were added to the treatment regimen. With improvement the following day, the patient began a steroid taper (5 days), continued the topical antibiotic for 5 days then stopped, and oral doxycycline was continued QD indefinitely. With complete resolution of the acute corneal condition (1 month), the patient was fit with scleral lenses OU [Figures 5 and 6] to improve vision (20/25 OD, OS) and decrease likelihood of future recurrent erosions.

Figure 5: The patient's right cornea and scleral lens as viewed with anterior-segment OCT

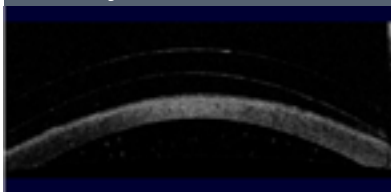
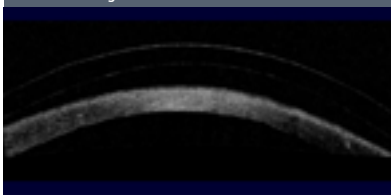


Figure 6: The patient's left cornea and scleral lens as viewed with anterior-segment OCT



## CONCLUSION

Avellino corneal dystrophy displays features of both granular and lattice dystrophy. Anterior stromal discrete gray-white opacities and lattice lesions in mid- and posterior stroma are pathognomonic for the condition. Anterior stromal haze is also often noted in these patients. The condition has an autosomal dominant inheritance pattern with very high penetrance, and has been linked to a mutation in the Big-h3 gene. Current treatments include PTK and PKP, with frequent recurrence after these interventions. Soft bandage lenses are often useful in preventing episodes of erosion in these patients, though erosions are seen more rarely than in either of the contributory corneal degenerations in isolation. Scleral lenses can also be useful both in preventing recurrences of erosions as well as improving vision. While the patient failed in a soft bandage lens, the liquid bandage and high Dk afforded by a rigid bandage lens has improved the patient's vision and comfort and delayed the need for surgical intervention.

Special thanks to Elyse L. Chaglasian, OD, FAAO

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# An Observational Study to Determine the Prevalence of Binocular Distance Visual Acuity among Lifeguards

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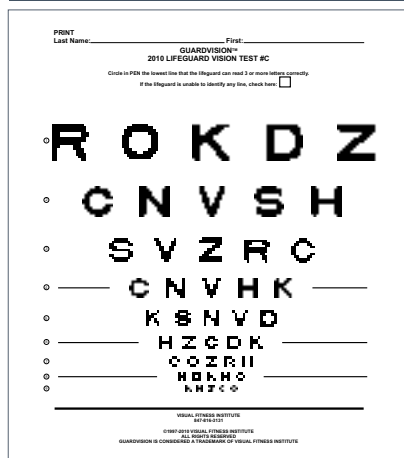
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## BACKGROUND

In most communities, lifeguards do not have a required minimal visual acuity (VA) in order to be certified. Various agencies have attempted to define the visual acuity requirements needed for effective lifeguarding; some have advocated a 20/30 threshold. Others have advocated a more rigorous requirement of 20/20 visual acuity. There are no data available as to what the average visual acuity is in a cohort of lifeguards. Without this data, it is difficult to predict the number of lifeguards that would be excluded from duty if various visual acuity requirements were initiated. The purpose of this study was to determine the prevalence of usual-corrected distance binocular visual acuity among a group of lifeguards.

FIGURE 1: The GuardVision™ visual acuity chart is a LogMAR-based chart modeled after the EDRS chart.



## METHODS

The GuardVision™ Self Testing Vision Screening Program provides distance VA screening materials that aquatic facilities can administer to their lifeguards. The program is the singular commercial product available for this purpose.

The program uses a LogMAR-based distance VA chart to assess the lifeguard's usual-corrected binocular VA at ten feet. In addition to identifying the smallest chart line read, the screening program also involves participants completing a self-report survey to identify age, gender, race, and usual-corrected refractive correction.

Analysis of variants (ANOVA) was used to identify any VA differences among race. Pearson correlation was used to determine any correlations between age and VA or between gender and VA. Statistical Package for Social Sciences (SPSS) was used for statistical analysis.

TABLE 1: Race frequencies of lifeguards studied..

Race	Frequency
Asian	6.8%
Black or African American (Non-Hispanic Origin)	4.8%
Hispanic or Latino	3.4%
White (Non-Hispanic Origin)	79.5%
Other	2.7%
Unknown	2.8%

## RESULTS

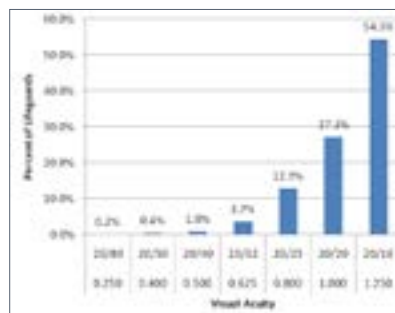
From 2,002 lifeguards, the mean age was  $20.0 \pm 4.73$  years (median of 19.0 years) with a range from 15.0 to 69.1. Lifeguard gender was 51.1% female, 48.4% male, and 0.5% unknown. The usual vision correction worn by the lifeguards was 28.6% contact lenses, 8.8% spectacles, and 62.6% not wearing any refractive correction.

The mean decimal VA for all lifeguards was  $1.09 \pm 0.20$  (median of 1.25) with a range from 0.25 to 1.25 corresponding to a mean Snellen equivalent of 20/18.

ANOVA showed a statistically significant difference in VA versus race ( $F=2.675$ ,  $p=0.02$ ). However, this was determined not to be clinically significant, with the mean acuity for each race group ranging only from 20/19 to 20/18. There is no correlation between age and VA (Pearson correlation= $-0.031$ ) or between gender and VA.

Snellen Equivalent passing criteria were evaluated at 20/20, 20/25, and 20/30 cut points.

FIGURE 2: Usual-corrected binocular distance visual acuity measures of lifeguards..



## CONCLUSION

This study indicates that a usual-corrected binocular distance acuity passing threshold of 20/20 would restrict a larger number of lifeguards from duty than a visual acuity passing threshold of 20/25 or 20/30. Although the current study makes no attempt to qualify the impact of visual acuity on lifeguarding, it should be recognized that nearly one-fifth of the lifeguards in this sample would be unable to be employed if a 20/20 visual acuity passing threshold were used. It should also be recognized that other visual skills such as contrast sensitivity, visual field, and stereopsis may also play a role in lifeguarding besides just the visual acuity studied here.

FIGURE 3: Distance visual acuity screening pass and fail rates using Snellen Equivalent pass rates of A) 20/20, B) 20/25, and C) 20/30.



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# TearLab Evaluation of Dry Eye Disease Related to Tear Quantity in a Normal Population

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## INTRODUCTION

A good case history is critical in the doctor's approach to treatment of dry eye. Factors contributing to dry eye include: age, sex, medications, diet, disease, surgery, contact lens wear and environmental factors. A patient may have evaporative dry eye, aqueous deficient dry eye or both conditions. Dry eye disease can be classified as mild, moderate or severe. TearLab Corp has designed an instrument to measure tear osmolarity and classify the severity of the disease. The purpose of this pilot study was to measure tear osmolarity in a young population (<40 years). The quantity of tear production was then measured to evaluate if aqueous deficiency was related to elevated tear osmolarity in this population of subjects.

## METHODS

134 second year optometry students were tested using a calibrated TearLab instrument. Subjects were instructed not to wear contact lenses or put any drops in their eyes 2 hrs prior to being tested. Both the eyes of each subject were tested and recorded. Values > 309 (MOSMS/L) were considered the lower limit for dry eye disease with the instrument for this study. Following the testing tear quantity was assessed by performing a Zone Quick test on both eyes of each subject. Results were analyzed using a 2 way repeated Analysis of Variance. Tear break up time (TBT) was performed on subjects with an elevated tear osmolarity level.

## RESULTS

1. Only a small percentage of subjects in this study had elevated tear osmolarity as classified by TearLab. (94.8% normal 5.2% elevated)
2. Both groups of subjects showed normal tear quantity production using Zone Quick testing.  
Normal 22.61 mm OD, 23.27 mm OS  
Elevated 24.16 mm OD 24.50 mm OS  
( $F=0.57$ ,  $p=0.45$ )
3. Subjects with elevated osmolarity exhibited a reduced TBT.  
5.8 mm OD 5.4 mm OS
4. No subjects in the study were classified as severe dry eye (>335)

### TearLab



## DISCUSSION

Appropriate therapy for DES depends on appropriate diagnosis of the cause(s) of the disease. Clinical examination of the patient to determine the cause(s) of the problem is imperative in prescribing and managing the appropriate therapy for the patient. Tear hyperosmolarity can be caused by a number of factors and has been linked to dry eye disease. Simply obtaining an elevated tear osmolarity does not address the cause of the problem. TearLab analysis uses osmolarity values of <308 as normal, 309-320 as mild, 321-335 as moderate and >335 severe when classifying dry eye. 5.2 % of the normal young subjects in this study had elevated tear osmolarity. There was no difference in tear quantity measurements between the normal and elevated tear osmolarity groups. All of the patients with elevated osmolarity levels showed a decreased TBT. Blepharitis and meibomian gland dysfunction were not evaluated in the study.

## CONCLUSIONS

1. Supplemental drops containing mucimimetics might be a good approach for younger patients with mild to moderate dry eye as classified by TearLab
2. Future studies should be proposed using mucimimetics to monitor tear osmolarity levels.
3. Research should develop new methods to increase mucin production.
4. Tear production is not reduced in all patients with elevated tear osmolarity.
5. A patient with elevated osmolarity levels without an evaporative component (blepharitis) and normal tear production might be classified as mucin deficient

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# Atypical Presentation of Endophthalmitis After Cataract Extraction

George Hanna, O.D.

Illinois College of Optometry, Chicago, IL

## BACKGROUND

Endophthalmitis is a rare but serious complication following cataract surgery. Without prompt treatment, irreversible vision loss can occur. This is a presentation of an atypical case of staphylococcus epidermidis positive endophthalmitis presenting without pain or redness one week after cataract extraction.

## PERTINENT FINDINGS

70 y/o African American female presented for her scheduled one week follow-up for an uncomplicated cataract extraction complaining of blurry vision and cob webs for two days in the operated eye. She denied any pain, discomfort or redness and was using her post-operative drops as directed (pred acetate QID OD, ofloxacin QID OD, ketorolac QID OD).

**Medical History:** HTN, DM, CAD

**Medications:** aspirin, lisinopril, warfarin, chlorthalidone, Lopressor, simvastatin

**BCVA:** OD: HM

**Slit Lamp Exam:** OD: 3+ cells, 2+ flare, 1mm hypopyon, no conjunctival injection.

**DFE:** OD: 4+ vitritis

**Diagnosis:** OD: endophthalmitis positive for staphylococcus epidermidis

**Differential Diagnosis:** Toxic Anterior Segment Syndrome (TASS)

**Treatment:** Pars plana vitrectomy and intravitreal vancomycin OD

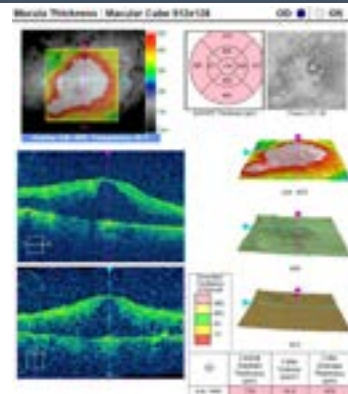
## PATHOPHYSIOLOGY

Endophthalmitis is an inflammatory condition involving the aqueous and vitreous. Destruction of the intraocular tissue occurs by direct invasion of the organism and by the inflammatory mediators of the immune response. Ultimately, destruction of the neurosensory retina and RPE occurs.

**Table 1:**  
Expected findings in endophthalmitis from the EVS study

Sign/Symptom	Incidence (EVS Study)
Blurred vision	94%
Red eye	80%
Progressive vitritis	80%
Hypopyon	75%
Pain	73%
Lid swelling	34%

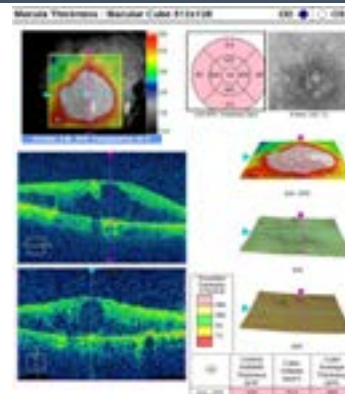
**FIGURE 1:** Macular edema one month after PPV and intravitreal vancomycin. CF VA.



## DISCUSSION

Inflammation is an expected outcome after any surgical procedure due to the breakdown of the blood aqueous barrier. One study shows that the expected amount of anterior chamber inflammation after uncomplicated anterior segment surgery is 2+ on day 1 and 1+ on day 14. Inflammation out of proportion to this should heighten the suspicion of endophthalmitis. Endophthalmitis has an incidence ranging from 0.08%-0.7% following cataract extraction. Approximately 20-30% of patients will present without pain or redness (see **table 1**). Prompt treatment is essential in attempting to preserve vision. Treatment typically includes intravitreal antibiotics including vancomycin and ceftazidime. According to the EVS study, patients presenting with light perception vision or worse also benefit from an immediate vitrectomy. Approximately 1 month after treatment, the patient returned to clinic with reduced vision and macular edema (see **figure 1**). Bromday once daily was initiated and the macular edema is slowly diminishing (see **figure 2**).

**FIGURE 2:** Macular edema decreasing 2 weeks after initiation of Bromday once daily in the operated eye. 20/600 VA.



## CONCLUSION

Patients presenting with decreased vision, hypopyon and vitritis after cataract surgery, with or without the presence of redness or pain, need immediate evaluation for endophthalmitis. It is also important not to overlook other potential causes of decreased vision such as macular edema.

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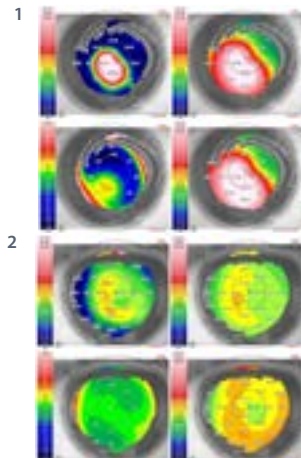
# The Atlantis Scleral Lens Design: A Case Series on Twin Sisters

Jennifer S Harthan OD, FAAO  
Chicago, IL

## BACKGROUND

Patients with asymmetric irregular corneas can be very challenging to fit with contact lenses secondary to the displacement of the corneal apex or due to the high levels of astigmatism, which may cause the lenses to de-center, ultimately impacting vision and comfort. Scleral lenses have long been an important option in the management of the irregular cornea and in the rehabilitation of ocular surface disease. Today, there are many scleral lens designs available that offer patients excellent vision and comfort. We present here the cases of forty-eight year old identical twin sisters that were diagnosed with corneal ectasia 6 months apart from each other. Each was successfully fit with the Atlantis™ Scleral Lens Design after failing with other lens modalities.

Figures 1 and 2: Topographies of the right and left eyes showing ectasia OD>OS for Patient 1



## CASES

**Patient 1** presented with complaints of blurred vision, distorted vision, and discomfort with her current hybrid lenses. She had experienced discomfort with all other lens modalities. She was diagnosed with keratoconus ten years prior OD>OS. Her refraction was -6.00-0.50x075, 20/200 OD and +1.25-0.75x110, 20/25 OS. Topography showed K readings of: 57.82/66.89 @128 OD and 43.65/44.61 @142 OS (Figures 1 and 2). The diagnosis of keratoconus OD>OS was confirmed and she was fit with the Atlantis™ lens based on the fitting guide (Figure 3).

Her vision improved to 20/20 OD, OS and she immediately noticed improved quality of vision. The patient has been able to wear her lenses comfortably for 14 hours per day.

**Patient 2** was diagnosed with keratoconus 6 months after her sister and reported that her left eye always had poorer vision than her right eye. She reported slight discomfort with her current contact lenses, and wanted to try the same design that her sister spoke so highly of. Her refraction was -2.50-3.50x042, 20/30 OD and -2.75-2.25x160, 20/100 OS.

Figure 3: Xcel's Atlantis™ Fitting Zones



K readings were: 45.49/49.36 @125 OD and 46.22/50.52 @043 OS (Figures 6 and 7). The patient's topographic analysis was consistent with pellucid marginal degeneration OS>OD. Patient 2 was also fit with the Atlantis™ lens and initial lenses provided excessive central clearance. After flattening the base curves, the fit was improved and the patient achieved comfortable 20/20 vision during all waking hours.

## DISCUSSION

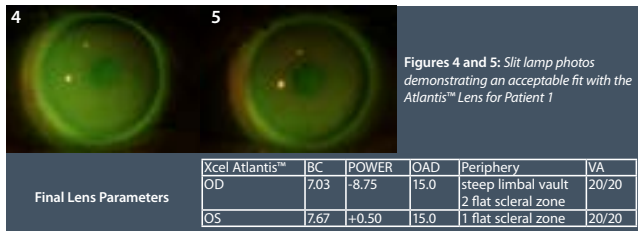
Both patients presented with keratoconus and desired improved comfort with their contact lenses. Clear and comfortable vision may be achieved using mini-scleral or scleral gas permeable lenses. These lenses provide refractive correction for the irregular and regular astigmatism. The larger lens designs

and a high Dk gas permeable material provide much success in the treatment of those with severe corneal disease and ectasias. The size of these lenses also improves comfort as the lid margin interacts with the surface of the lens rather than the edge. The Atlantis™ lens design has three fitting zones: the Base Curve or Central Zone, the Limbal Vault Zone, and the Scleral Zone. When fitting patients with this lens design, each zone can be manipulated to customize the lens fit for the patient. As with any lens fit, potential complications that may arise include corneal hypoxia, conjunctival and corneal staining, neovascularization, corneal infiltrates and microbial keratitis. These complications can be avoided with proper patient selection, fitting techniques, patient education, and with close observation. When prescribing these lenses, careful consideration is needed to weigh the potential benefits and risks.

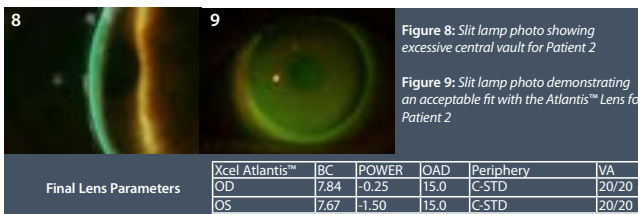
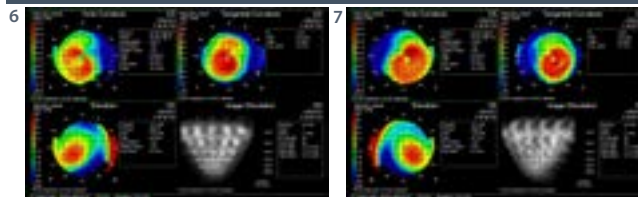
## CONCLUSION

Both patients were diagnosed with corneal ectasia, with each having asymmetry in disease severity between their eyes. In such patients who have failed with other lens modalities, scleral lenses may be considered as an appropriate management option. In both cases, the patients reported good vision and markedly improved comfort with the large diameter lenses. To date, both are wearing their Atlantis™ lenses successfully all day long.

References: Available upon request.



Figures 6 and 7: Topographies of the right and left eyes showing ectasia OS>OD for Patient 2



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# A Quarter Century of Residency Programs

Janice M. Jurkus, OD, MBA; Rebecca K. Zoltoski, PhD  
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## INTRODUCTION

In the quarter century of its existence, the residency program at the Illinois College of Optometry (ICO) has worked with 240 people. Knowing the former resident's thoughts about residency programs, as well as how it influenced their career will help to understand and share the value of completing a residency. This study investigates what people thought of the residency, as well as how their careers were influenced by residency programs from a private Optometry school.

## METHODS

A link to an online survey (SurveyMonkey) was sent via email to 225 of the past residents. The survey had nineteen multiple choice and open ended questions. Information on demographics, schooling, and residency program completed was collected. The reason for selection of a specific program, advantages and disadvantages, satisfaction and comments on the best and worst thing about the residency were requested. Lastly, post residency career and academic contributions were investigated.

## RESULTS

Ninety-three people (40%) responded to the survey. The majority of those that responded were women (68.8%) with an average age during the residency of 26.6 (24-36). The programs that were represented in the responses were:

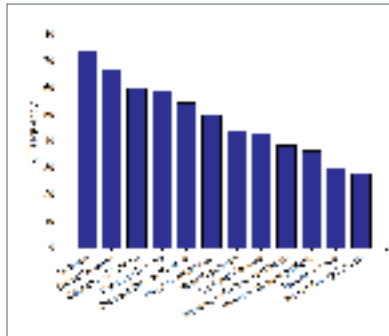
At the Illinois College of Optometry: Primary care/ Ocular Disease (52.1%), Pediatrics/Binocular Vision (16.0%), Cornea/ Contact Lenses (7.4%) and Low Vision Rehabilitation/Ocular Disease (7.4%).

Affiliate programs were VA Ocular Disease/Low Vision (7.4%), Primary Care (1.1%) and Refractive Surgery Co-Management/Anterior Segment Disease (8.5%).

The top five reasons for selecting a specific residency were:

1. Location (72%)
2. Clinical diversity (65.6%)
3. Educational options in the program (59.1%)
4. Variety of doctors to work with (58.1%)
5. Being able to work in different clinical settings (53.8%)

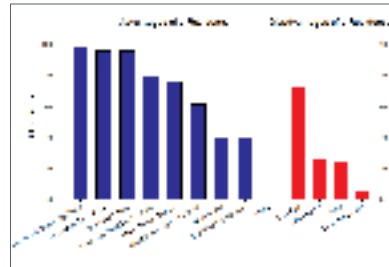
FIGURE 1: Reasons for Selecting a Specific Residency



When asked what the main advantages of completing a residency, over 95% responded the increased knowledge base, clinical skill and confidence. The ability to treat and diagnose eye disease (79.8%), having more career options (75.5%), interdisciplinary interactions (61.7%), networking (40.4%) and certification credit (39.4%) were also indicated. 1.1% of the respondents indicated there was no advantage to doing a residency. Common themes as to what the best thing about the resident included increased skill and confidence due to patient diversity and working with mentors as well as the friendships and interaction with faculty, staff and students.

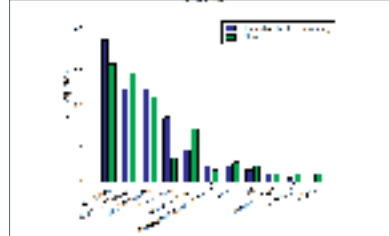
The greatest disadvantage was the low pay (73.6%) while over a quarter of the respondents thought there were no disadvantages.

FIGURE 2: Advantages/Disadvantages



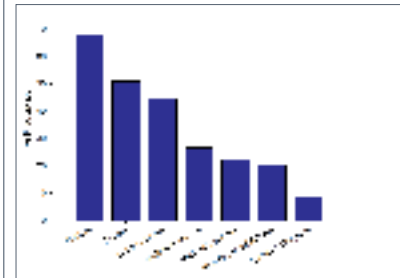
Residents practiced in a variety of settings upon completion of the residency. They also tended to change positions. The average resident reported practicing in 3 different sites, with the most reporting 10 different places. Initially, the largest number (38.2%) worked in education. This number decreased to 33% when asked where they are currently practicing. A move away from OD/MD practice (24.7% to 22.7%) and commercial optometry (16.9% to 6.8%) was also noted. More residents went to group practice (25.8% to 31.8%), solo practice (6.7% to 13.6%), VA optometry (3.4% to 4.5%) or research (1.1% to 2.3%).

FIGURE 3: Practice Setting



Graduates of a residency program continue to contribute to the profession. 79.7% have presented posters while 59.5% presented papers. 62.1% have published articles or text book chapters and 77.2% have lectured at national or local meetings.

FIGURE 4: Academic Contributions



When asked if they would encourage an optometry graduate to do a residency, one (1) person was undecided while the overwhelming majority (98.9%) answered yes.

## CONCLUSIONS

The field of optometry has seen some dramatic changes since the residency program at ICO was established in 1987. Residency programs increase confidence in providing patient care. Despite the low pay, majority find the advantages to far outweigh the benefits. Most of those who completed residencies programs become educators or private practitioners. They provide academic contributions to the profession.

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# A Hereditary Retinal Disease Masquerading as Anisometropic Amblyopia in a Pre-school Patient

Valerie M. Kattouf O.D., Leonard Messner O.D., Mary Flynn-Roberts O.D., Jacqueline Williams, Emily Lemburg, Sasha Murphy

## INTRODUCTION

A 2 year old male is diagnosed with anisometropic amblyopia (refractive error of OD +2.00 sph, OS +4.50 sph). A spectacle Rx was prescribed. Visual acuity (VA) after one month of Rx wear was OD 20/60, OS 20/125 with poor cooperation secondary to age and ability. Dilated fundus exam was unremarkable. Occlusion therapy and then Atropine therapy were prescribed; poor compliance was noted with each. Little progress was made in regard to VA improvement therefore a diagnosis of pathology was more ardently pursued.

FIGURE 1: 5 year old patient



## METHODS

Repeat cycloplegic refraction one year after initial diagnosis was OD +2.00, OS +3.50. At this point the diagnosis of anisometropic amblyopia was questioned. A repeat dilated fundus examination showed no significant findings. VA was stable. At 4 years old a small anterior cataract was noted on retinoscopy. Visual acuity was OD 20/125, OS 20/125. This was the first decrease noted in visual acuity OD. A repeat cycloplegic retinoscopy revealed OD +3.00 sph, OS +2.25 sph. An OCT and retinal photography were performed. The dilated fundus exam noted cystic/petaloid appearance to the macula OD> OS. Changes to the retinal periphery were also noted. Fundus photos and OCT OD revealed a spoke wheel pattern of the macula, retinal splitting and cystic changes of the macula. Each of these findings were present but less apparent OS. A diagnosis of X-linked retinoschisis was suspected.

FIGURE 2: Posterior Pole Images OD, OS

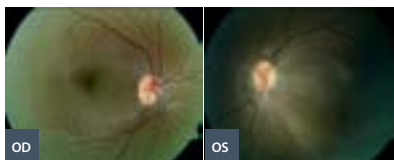
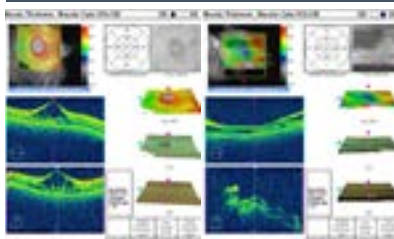
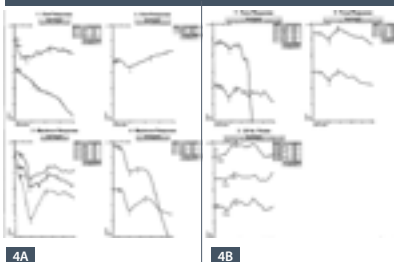


FIGURE 3: OCT Images OD, OS



An ERG was performed to confirm the diagnosis. The ERG revealed a selective loss of the b-wave in combined response and confirmed the diagnosis of X-linked retinoschisis OU. An unspecified hereditary retinal dystrophy was also detected.

FIGURE 4A: Scotopic ERG, 4b: Photopic ERG



Patient History

Age	Visual Acuity	Comments
2.5	none	Cyclo Rx OD +2.00 D OS +4.50 D Rx given OD +1.00, OS +3.50
2.9*	OD 20/60 OS 20/200	No stereo fly, no RDS Stereo smile occlusion started 2-4 hrs/day OD
3.0*	OD 20/60 OS 20/125	occlusion done 1-2 hrs/day OD Resume occlusion 2-4 hrs daily
3.1	OD 20/40 OS poor responses	occlusion 2-4 hrs/day OD Poor compliance, was using pirate patch, stressed importance of adhesive occlusion
3.5	OD 20/60 OS 20/125	occlusion 2-4 hrs/day OD Poor compliance
3.9	OD 20/60 OS 20/125	Dry Ret OD +0.50 D, OS +2.50 Occlusion d/c Atropine started (1 gtt OD Saturday and Sunday )
3.11	OD 20/70 OS 20/80	Atropine Installation wrong eye
4.1	OD 20/80 OS 20/125	Atropine Installation now appropriate
4.5	OD 20/60 OS 20/80	VA not making sense, repeat DFE at FU, r/o pathology, repeat cyclo
4.8	OD 20/125 OS 20/125	Cyclo Ret OD +1.50 D, OS +2.75 Cataract noticed on retinoscopy Poor dilation with spray, no detectable macula findings
4.9	OD 20/100 OS 20/125	Cyclo Ret OD +3.25, OS +4.00 Schedule posterior pole photos and OCT, suspect retinal disease

FIGURE 5A – 5C, Low Vision Devices



## RESULTS/TREATMENT

Treatment with topical dorzolamide was initiated with a goal of decreasing the macular edema and improving the visual acuity. Our patient underwent a Low Vision Evaluation. His grandmother was educated on the dispensing and use of magnification devices at distance and near, reading stand use, safety precautions in gym, Vision Itinerant Teacher, front row seating, testing accommodations and adaptive technology. He continues to wear his hyperopic spectacle correction and his refractive error is monitored regularly.

## CONCLUSION

X Linked Juvenile Retinoschisis is an X linked recessive disorder affecting mostly males. It typically displays symmetric bilateral macular involvement with onset in 1st decade of life. The prevalence of X-linked juvenile retinoschisis ranges from 1 case per 5,000 population to 1 case per 25,000 population. Visual acuity typically deteriorates during 1st-2nd decade of life and remains stable until 5th-6th decade of life. Patients have been diagnosed as early as age 3 months most patients are seen at 5 years or older.

"X-linked juvenile retinoschisis often presents in a young boy with slightly decreased vision that cannot be corrected fully by refraction. Diagnosis is easily missed during early onset".

The initial diagnosis of anisometropic amblyopia in this 2 year old pediatric patient was correct and warranted. The inability to get accurate/repeatable visual acuity on a 2-3 old and the poor compliance with occlusion and atropine treatment influenced the diagnosis. Once a consistent lack of visual acuity improvement and a change in refractive error were noted the X-linked retinoschisis became more evident. Posterior pole photos and OCT technology were able to reveal what was not appreciable with a standard assessment of the poster pole with a binocular indirect ophthalmoscope. An ERG confirmed the diagnosis with specificity. Referral within the optometric community provided the most up to date and complete care for this patient. As the diagnosis of our patient was being finalized it was revealed/discovered that he has two maternal cousins with a diagnosis of X-linked retinoschisis.

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# Late Stage Coat's Disease in a 19-month-old male with a presumed diagnosis of Retinoblastoma

Valerie M. Kattouf O.D., Kelsey Beck, Katie Davis

## CASE HISTORY / INTRODUCTION

A 19-month-old African American male presents for his first eye exam. His mother notes an intermittent eye turn inward for several months. History notes a full term birth and no developmental delays. The patient has previously been diagnosed with Fabry disease (an X linked lysosomal storage disease), which is being managed by his primary care physician.

FIGURE 1: Leukocoria OS



## OCULAR EXAMINATION

The patient was able to fix and follow OD but unable to fixate and follow OS. Kappa-Hirschberg findings revealed no strabismus PRE dilation. Evaluation of alignment status with a trans illuminator also revealed leukocoria in the left eye (Figure 1: Leukocoria OS). Retinoscopy findings were +1.00 sphere in the right eye, no retinoscopy reflex could be obtained in the left eye. The anterior segment examination was unremarkable, no corneal clouding coincident with Fabry's disease was apparent. A 25 prism diopter intermittent left esotropia was apparent POST dilation (Figure 2: Esotropia OS). Fundus examination of the right eye was unremarkable. Fundus examination of the left eye revealed a large mass obscuring the majority of the posterior pole (including the macula and optic nerve). The mass was vascularized and projecting significantly anterior in the globe (Figure 3: Ocular Mass).

FIGURE 2: Esotropia OS



FIGURE 3: Ocular Mass OS



FIGURE 4: Examples of a CT scan in a Coat's Disease patient



## RESULTS

The size and appearance of the mass led to a presumed diagnosis of Retinoblastoma. Additional differentials for leukocoria were considered. Congenital cataract, ROP and PHPV were ruled out. The vascular appearance of the mass and the apparent retinal detachment led to the consideration of Coat's Disease as a differential. Coat's disease, a rare congenital, nonhereditary eye disorder, results in the exudative retinitis and retinal telangiectasis that were apparent in our patient.

Table 1: Characteristics of Coat's Disease

Exudative retinitis, retinal telangiectasis
Inheritance pattern unknown
Very rare; young males (M:F, 3:1)
80% unilateral
Characterized by abnormal vessel development
Poor prognosis in advanced stages
Retinal detachment in advanced stages

Table 2: Stages of Coat's Disease

I	Abnormal dilation of retinal blood vessels
II	Telangiectasia and exudation
III	Exudative retinal detachment
IV	Total retinal detachment
V	Characterized by irreversible blindness

A consult with our pediatric ophthalmologist and a CT were scheduled to determine the etiology of the retinal findings. The CT scan confirmed a diagnosis of Stage IV Coat's disease, a very severe presentation in a 19-month-old patient (Figure 4: Example of a CT scan in a Coat's Disease). Referral was made to a pediatric retina specialist who treated our patient with injections and laser therapy. The treatment spared the eye and enucleation was not necessary.

## CONCLUSION

Leukocoria presents with a varied list of differentials. The determination of the proper etiology is crucial as some differentials are sight and/or life threatening. Imaging work ups are often necessary to determine the most precise diagnosis. Presentation of Coat's disease at 19 months old is not expected (the peak age of onset for Coat's Disease is 6-8 years of age). This case is a reminder that not all patients fall into classic age guidelines. The inheritance pattern of Coat's Disease is unknown and affects males in a 3:1 ratio compared to females. 80% of cases are unilateral and characterized by abnormal blood vessel development as seen in our patient. Early diagnosis is important to prevent a poor visual outcome and possible retinal detachment.

Table 3: Clinical Comparison of Retinoblastoma vs. Coat's Disease

	Retinoblastoma	Coat's Disease
Leukocoria	Grey/white pupillary reflex	Yellow pupillary reflex
Strabismus	Possible presentation	Possible presentation
Retinal Blood Vessels	Uniform dilation Disappear into adjacent neoplasm	Irregular, (+) aneurysms Remain visible from posterior pole to peripheral fundus
Anterior Chamber	Typically clear	Typically clear
Laterality	70% unilateral	80% unilateral

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# Optic Disc Edema Associated with Sudden Onset Panuveitis

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## BACKGROUND

Optic disc edema in association with sudden onset panuveitis is a rare clinical entity. This entity was first reported by Monheit and Read in 2005 as optic disc edema associated with anterior uveitis and no posterior uveitis. To our knowledge, the clinical features and course of optic disc edema associated with sudden onset panuveitis have not been reported.

Figure 1:  
Nodular Episcleritis OD

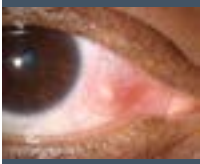


Figure 2:  
Nodular Episcleritis OS



Figure 3:  
Optic Disc Edema OD



Figure 4:  
Optic Disc Edema OS



Figure 5:  
increased RNFL thickness OD and OS

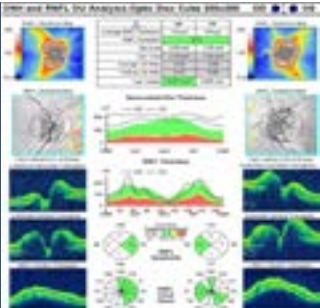


Figure 6 & 7: positive deflection of RPE-Bruch's complex suggestive of papilledema OD and OS.

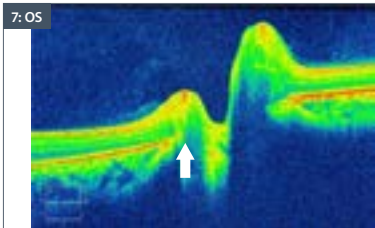
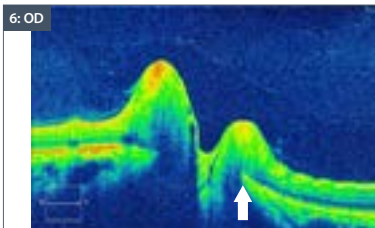
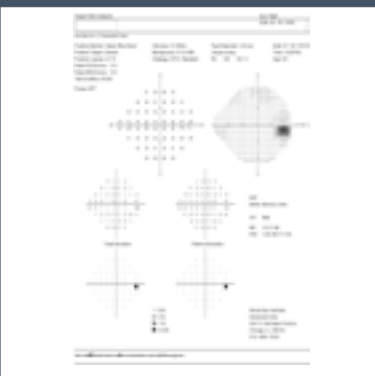


Figure 8: enlarged blind spot OD



## PERTINENT FINDINGS

35 y/o African American female presented with bilateral red eyes for three days.

**MEDICAL HISTORY:** fibromyalgia, asthma and anemia

**MEDICATIONS:** Proventil inhaler

**BCVA:** 20/30 OD, OS

**PUPILS/SLIT LAMP EXAM:** PERRL (-) APD/bilateral nodular episcleritis (see Figures 1 & 2), AC deep & quiet OU

**DFE:** bilateral optic disc edema (see Figures 3 & 4), vitreous clear with no cells present

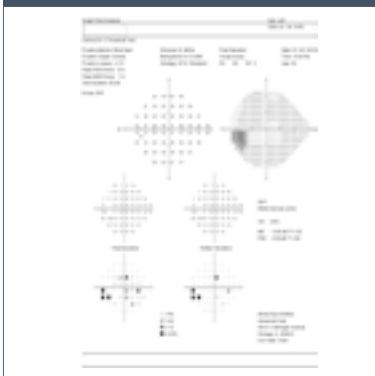
**B-SCAN ULTRASONOGRAPHY:** negative for posterior scleritis

**CIRRUS™ SPECTRAL DOMAIN OCT:** increased RNFL thickness (see Figure 5) and positive angle of deflection of the RPE-Bruch's complex suggestive of papilledema (see Figures 6 & 7)

**HUMPHREY VISUAL FIELD:** enlarged blind spots in both eyes (see Figures 8 & 9)

**MRI/MRV:** brain and orbits with and without contrast unremarkable

Figure 9: enlarged blind spot OS



Five days later, the patient presented with panuveitis in the left eye.

**SEROLOGY:** negative for toxoplasmosis, sarcoidosis, syphilis, lupus and rheumatoid arthritis

Seventeen days after the initial presentation, visual acuities had declined to PH: 20/60

**DIAGNOSIS:** optic disc edema associated with sudden onset panuveitis

**DIFFERENTIAL DIAGNOSIS:**

- Ocular toxoplasmosis
- Ocular sarcoidosis
- Ocular syphilis
- Systemic lupus erythematosus
- Rheumatoid arthritis

## DISCUSSION

After ruling out infectious etiology, eighty milligrams oral steroids were initiated and tapered over three weeks. Resolution of the panuveitis occurred within three weeks and the vision returned to 20/20 in each eye, while optic disc edema lagged by one month. The positive deflection of the RPE-Bruch's complex ultimately returned to a neutral position, suggesting normalization of intracranial pressure.

## CONCLUSION

To our knowledge, this is the first report of the clinical features and course of optic disc edema associated with sudden onset panuveitis. Resolution of the disc edema lagged that of the uveitis. In the presence of normal pupil function absence of visual field loss, return of normal vision, and absence of infectious etiology, this case suggests that treatment may be directed towards the uveitis rather than prolonging treatment until optic disc edema resolves.

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# Choroidal Ischemia with Serous Retinal Detachment in Undiagnosed Preeclampsia

Kaitlyn Keller, O.D., Stephanie Klemencic, O.D., F.A.A.O., Leonard Messner, O.D.

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## BACKGROUND

Preeclampsia is a potentially fatal condition of elevated blood pressure, proteinuria and edema after 20 weeks of gestation. The only known cure is delivery of the placenta. The acute elevation in blood pressure with preeclampsia can also result in severe vision loss. We present a case of presumed choroidal ischemia and serous retinal detachment in a pregnant woman with severe retinal detachment.

## PERTINENT FINDINGS

26 y/o African American female presented with sudden blurry vision in her infero-nasal field of the right eye of one day duration. She also reported recent onset right upper eyelid swelling and bilateral lower leg edema

**MEDICAL HISTORY:** 34 weeks pregnant without known complications

**MEDICATIONS:** Prenatal vitamins

**BCVA:** 20/40 OD, 20/20 OS

**BLOOD PRESSURE:** Elevated at 188/125 mmHg RAS

**PUPILS/SLIT LAMP EXAM:** PERRL (-)APD/unremarkable

**DFE:** Right eye revealed serous retinal detachment and widespread presumed choroidal infarctions (see Figure 1). Left eye was unremarkable (see Figure 2). The optic nerves were healthy without edema.

**CIRRUS™ SPECTRAL DOMAIN OCT:** Macular involved serous retinal detachment in the right eye (see Figure 3). Left eye was unremarkable (see Figure 4).

**DIAGNOSIS:** Serous retinal detachment with presumed choroidal infarctions in the presence of elevated blood pressure in a pregnant woman

**DIFFERENTIAL DIAGNOSIS:**  
Central serous chorioidopathy

## DISCUSSION

The patient's obstetrician could not be reached, and she was immediately referred to the emergency room. She was diagnosed with preeclampsia and underwent an emergency cesarean section that night. The patient remained in the hospital for 5 days and was prescribed oral hypertensive medication. One week later, vision improved to 20/25+ OD and the right upper eyelid edema resolved. There was flattening of the serous retinal detachment (see Figure 5) and improvement of the presumed choroidal infarctions with remaining mild disseminated RPE changes temporal to the macula (see Figure 6).

Figure 1: Serous retinal detachment and widespread presumed choroidal infarctions OD



Figure 2: Normal fundus OS

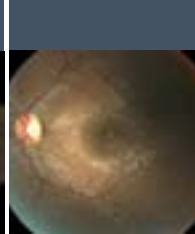


Figure 3: Serous retinal detachment OD

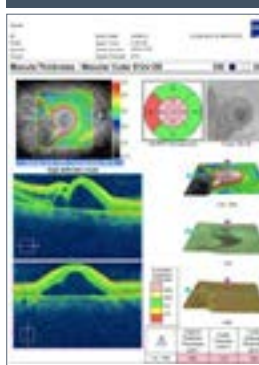
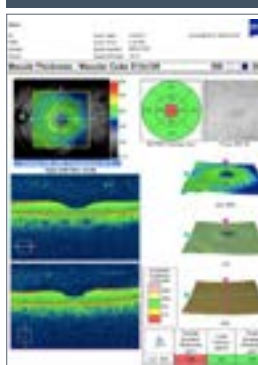


Figure 4: Normal macular OCT OS



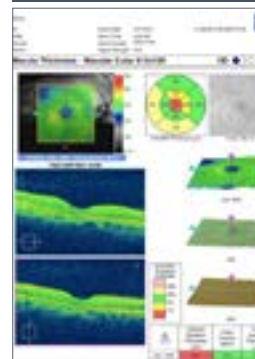
## PATHOPHYSIOLOGY

- Choroidal infarctions due to an acute rise in BP
  - Constriction of the choroid and choriocapillaris leads to choroidal ischemia
  - Damage to the endothelium of the choriocapillaris causes increased permeability and disruption of the blood-retinal barrier allowing macromolecules to pass through and deposit within the retinal layers.
- Serous retinal detachment occurs secondary to choroidal ischemia
  - Disruption of the vascular supply in the choroid caused by intense arteriolar vasospasm.
  - Lack of choroidal perfusion decreases function of RPE and allows for accumulation of subretinal fluid

Figure 6: Improvement of presumed choroidal infarctions with remaining mild disseminated RPE changes temporal to macula OD



Figure 5: Flattening of serous retinal detachment OD



## CONCLUSION

This case represents a rare case of unilateral presumed choroidal infarctions and serous retinal detachment in a pregnant patient due to undiagnosed preeclampsia. Patients with preeclampsia may present with the isolated symptom of blurred vision. Checking blood pressure is key for diagnosis. Elevated blood pressure in pregnant patients should be communicated to the obstetrician and the presence of associated retinopathy requires immediate referral to the emergency room. Recognizing the signs and symptoms of this condition can save the lives of the mother and her baby.

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# Assessment of Motion Sickness Among Patients with Visual Impairment

Tracy L Matchinski, OD, FAAO and Janis E Winters, OD, FAAO

## BACKGROUND

Motion sickness (MS) typically occurs when there is a neural mismatch between the sensory system (vision) and the vestibular system. A survey was developed to determine incidence of motion sickness in patients with no vision or patients losing their vision and what these patients did to alleviate their symptoms. Another objective of the survey was to determine if patients with vision impairments experience MS when receiving new glasses or using low vision devices.

## METHODS

All patients with visual impairment entering 2 different Low Vision Rehabilitation clinics were asked to participate in a 31 item Motion Sickness Questionnaire. The questionnaire sought to characterize vision loss, MS and changes in MS over time.

## RESULTS

Of the 263 VI patients surveyed, 25.5% (67) reported a history of motion sickness (MS). See **Figure 1** and **2**. Of those with MS,

- **Demographics: 23.9% Age 18-39 yrs, 47.7% 40-59 yrs and 28.3% 60 yrs and older; 67% female**
- **35% reported no MS prior to vision loss however 37% no memory /congenital VI.**
  - ➔ **The majority felt MS has remained stable (54%) and their balance has worsened (53%) as VI had progressed.**
- **59% reported noticing MS when riding in a car. The most common method of relief was to 'think of something besides MS' followed by 'do nothing'. Unable to assess other modes of transportation, when reading on a mode of transportation or watching a movie due to limited number of patients performing those activities.**
  - ➔ **The majority did not notice of MS with glasses or low vision devices (66% and 77% respectively).**
  - ➔ **If they did experience MS with devices, the most common categories patients reported experiencing MS with included electronic magnifiers and hand held magnifiers.**

## DISCUSSION

We are unable to assess if this study shows that people with vision impairment or blindness have increased incidence of MS or demographic trends follow the general population since data from the general population is unknown. However since 1:4 VI patients did report MS; surely MS is not rare among those who are VI.

Patients with a range of visual impairments and ocular conditions causing visual impairments reported a history of motion sickness. A large number reported that they developed MS after becoming VI.

Although the majority did not report motion sickness with new glasses or low vision devices, motion sickness still should be a consideration when caring for visually impaired patients. More rehabilitation training may be needed especially with hand held magnifiers or electronic magnifiers. Patients may discontinue use of devices to the discomfort.

## CONCLUSIONS

Clinicians should be aware that the potential for motion sickness in VI patients. Since the majority did not physically do anything to relieve MS, clinicians should be aware of common remedies and provide appropriate patient education about common treatment modalities of alleviating motion sickness. In addition, this survey showed that 53% reported that their balance had worsened with their vision loss and fall prevention education should be done.

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Figure 1: Classification of VI among those with Motion Sickness

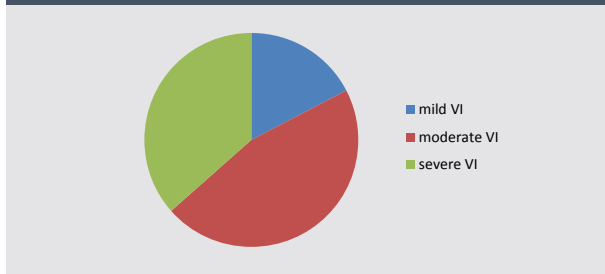


Figure 2: Cause of VI in those with Motion Sickness

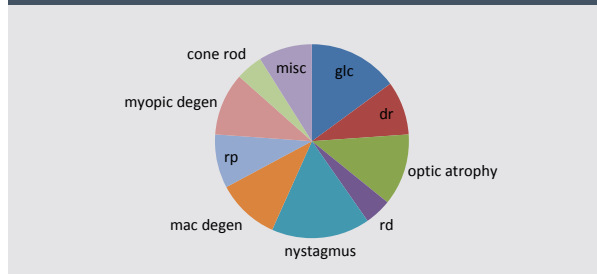


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# Atypical symptoms of profound dyschromatopsia and visual disturbance, without pain on eye movement in a case of retrobulbar optic neuritis

Christina E. Morettin, O.D. and Leonard V. Messner, O.D. F.A.A.O. • Illinois College of Optometry, Chicago, Ill

## BACKGROUND

Optic neuritis is an inflammatory, demyelinating lesion of the optic nerve. The exact cause is unknown. It is the most common neuropathy in persons under 50 y/o and is the first presenting symptom of multiple sclerosis (MS) in 15-20% of cases. Patients in whom optic neuritis is the first manifest of MS have shown to have a low rate of disability, compared to other MS patients. The presence of brain MRI abnormalities at the time of optic neuritis is a strong predictor for the 15 year progression to MS. Overall, 38% of patients will develop MS in 10 years.

## PERTINENT FINDINGS

42 y/o Caucasian woman presented with sudden onset blurred vision w/ profound dyschromatopsia OS for 2 weeks (reported as 20/60), w/ resolved eye strain OS for two days the week prior. She reported a history of migraine with aura since 19; however, the new symptoms were not consistent with her previous aura. No other neurological symptoms.

**Medical history:** (+) migraines with aura since 19 y/o

**Medications:** (+) Tretinex prn

**BCVA:** 20/20 OD, OS

**Pupils:** (+) RAPD OS

**Color Vision:** R/G defect OS

**Red cap desaturation:** 90% desaturation OS

**Clinical Exam/DFE:** unremarkable; optic nerves pink and well-perfused, margins in tact OU (Figure 1, 2)

**Cirrus™ Spectral Domain OCT:** Unremarkable and symmetric (Figure 3, 4)

**24-2 Sita-Standard Visual Field:** Full OD (Figure 5), Centrocecal defect OS (Figure 6)

**MRI:** T1 through orbits with fat suppression show enhancing lesion of the optic nerve distal to the optic chiasm, consistent with optic neuritis (Figure 7)

Corresponding coronal T1 image with enhancing lesion of the optic nerve (Figure 8)

Two hyperintense non-enhancing isolated periventricular white matter lesions on FLAIR, most likely ischemic events from migraine (Figure 9)

The 15 year risk of developing MS with retrobulbar optic neuritis is 25% when there are no active demyelinating lesions on MRI, which is exemplified. With one or more enhancing demyelinating lesions on MRI, the risk increases to 72%.

**Diagnosis:** Retrobulbar optic neuritis OS

**Differential diagnosis:** Posterior ischemic optic neuropathy OS, migraine related

FIGURE 1: Posterior pole unremarkable OD

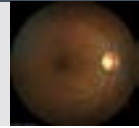


FIGURE 2: Posterior pole unremarkable OS

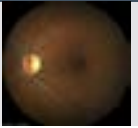


FIGURE 3: GCC OCT analysis unremarkable and symmetric OU at consult

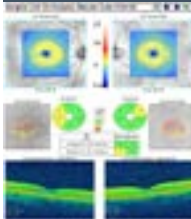


FIGURE 4: RNFL OCT analysis unremarkable and symmetric OU at consult

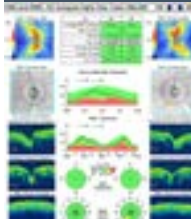


FIGURE 5: Unremarkable visual field OD



FIGURE 6: Centrocecal defect OS

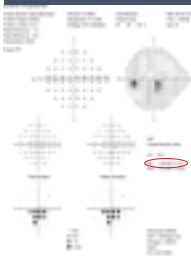


FIGURE 7: Transverse T1 image through orbits with fat suppression show enhancing lesion of the optic nerve distal to the optic chiasm



FIGURE 8: Corresponding coronal T1 image with enhancing lesion of the optic nerve



## TREATMENT

Steroid treatment was not initiated. Self-limiting recovery, with regular follow up.

Patient was referred for neurological consult. Neurologist agreed that two non-enhancing periventricular white matter lesions are likely ischemic and non-demyelinating. Interferon therapy not initiated.

## FOLLOW-UP

**2 Week:** Symptoms stable. GCC superior and temporal thinning OS noted (Figure 11).

**2 Month:** Symptoms resolving. Diffuse GCC thinning OS (Figure 11). Repeat visual field shows improving Mean Deviation, with central field depression OS (Figure 10).

FIGURE 9: FLAIR, isolated periventricular white matter lesion right side



FIGURE 10: Two month visual field shows improvement of Mean Deviation, with central depression OS

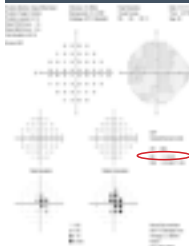
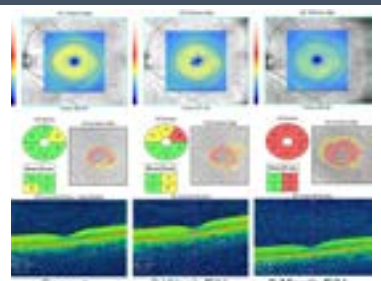


FIGURE 11: Thinning of GCC over time, as visual acuity paradoxically improves in optic neuritis



## DISCUSSION

This case exemplifies atypical symptoms in a case of retrobulbar optic neuritis. She presented without classic pain on eye movement; described as eye strain for two days the previous week. The patient presented with profound dyschromatopsia and desaturation. Given her past history of migraines with aura, posterior ischemic optic neuropathy was ruled out with MRI.

A paradoxical relationship exists between visual acuity and GCC in resolving optic neuritis. This relationship is seen in this case; as visual acuity improves, GCC thickness decreases.

The MRI of the brain showed an enhancing lesion of the left optic nerve distal to the optic chiasm, which was consistent with optic neuritis. Her two non-enhancing periventricular white matter lesions were not characteristic of demyelinating lesions, but of previous ischemic events from migraine. Her risk of developing MS in 15 years without accompanying active demyelinating lesions at the time of the optic neuritis was 25%. It was important to assess her risk of developing MS, as it is important not to incorrectly give a patient the diagnosis of MS and commit the patient to lifelong disease modifying treatment.

## ACKNOWLEDGEMENTS

Dr. L.V. Messner

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# Physical, Systemic and Ocular-Visual Findings Associated with Angelman Syndrome

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## BACKGROUND

Angelman syndrome (AS) is a developmental disorder characterized by ataxia with hypotonia, severe mental retardation, epilepsy, paroxysmal laughter and severe speech impairment. An obsolete term "happy puppet syndrome" was once coined for this syndrome, as the children appear as if they are walking like puppets on a string due to the ataxic gait. Prevalence is estimated between 1/10,000 to 1/20,000. Genetic studies most frequently reveal a deletion of the long arm (q) on chromosome 15. Ocular findings consist of hypopigmentation of the fundus and iris, nystagmus, strabismus and refractive error.

## CASE STUDY

A 5-year-old Hispanic female was referred to the developmental disabilities clinic at the Illinois Eye Institute from an outside optometrist for a comprehensive eye examination. A review of medical history indicated full term birth with seizures noted before 24 months. At 26 months, a diagnosis was made for AS by seizures, developmental delay, ataxia, tongue protrusion and hypotonia. Genetic studies revealed a deletion of chromosome 15q. A course of therapy with occupational, developmental, physical and speech therapists was initiated.



### Visit #1:

At the initial visit, the patient was currently on Keppra for seizures. She was unable to walk independently secondary to hypotonia and ataxic gait. The patient exhibited severe mental retardation, language delay, macrostomia, tongue protrusion and paroxysmal laughter. Visual acuity measurements were attempted but unobtainable using Teller visual acuity cards. Hirschberg was central steady OU. Bruckner displayed equal reflexes OU. Pupils were equal, round and reactive to light. No strabismus was noted upon cover test. The extraocular muscles revealed full range of motion. The cycloplegic refraction revealed +0.50 -3.75 X 165 OD and +2.00 -3.00 X 015 OS. External health was unremarkable. Dilated fundus exam revealed a hypopigmented fundus. The patient was diagnosed with ametropia, astigmatism and ocular hypopigmentation. Glasses were prescribed for full-time wear and parents were educated about sunwear.

### Visit #2:

Follow-up examination occurred 1.5 months after glasses were prescribed. The patient had been wearing the glasses for most of the course of a day. Mother noted that she seemed more aware of her visual surroundings with glasses on. At this visit, Teller acuity was found to be 20/400 with no aversion to covering of either eye. Over-retinoscopy with the glasses on revealed low plus OD and OS. No strabismus was noted upon cover test. The patient was scheduled for follow up in 4 months.

## DISCUSSION

AS is often difficult to diagnose with a developmental delay noted first at about 6 months but other characteristics of AS not becoming apparent until over the age of 1. Laboratory testing is used to confirm the clinical diagnosis. As the patient enters adulthood, the clinical features of AS change: Seizures typically remain but with decreasing frequency and hyperactivity and sleep issues improve. Patients with AS have a near normal life expectancy.

Table 1: Clinical Features

<b>Systemic Features</b>	<ul style="list-style-type: none"><li>-seizures</li><li>-severe mental retardation</li><li>-microcephaly</li><li>-abnormal EEG</li></ul>
<b>Ocular-Visual</b>	<ul style="list-style-type: none"><li>-hypopigmentation of fundus and iris</li><li>-strabismus</li><li>-nystagmus</li><li>-amblyopia</li><li>-astigmatism</li><li>-keratoconus</li><li>-optic nerve atrophy</li><li>-optic disc pallor</li><li>-atrophy ptosis</li></ul>
<b>Physical Characteristics</b>	<ul style="list-style-type: none"><li>-ataxia</li><li>-hyperactivity/attention deficit</li><li>-frequent bouts of laughter</li><li>-severe speech impairment</li><li>-tongue protrusion</li><li>-excessive chewing</li><li>-macrostomia</li><li>-sleep disturbances</li><li>-hypopigmentation of skin</li><li>-sensitivity to heat</li></ul>

## CONCLUSION

The physical, systemic and ocular-visual findings are reviewed in a five year old with AS. The ocular findings in addition to the physical and systemic findings can help aid in the early diagnosis of AS. Eye care professionals should work in conjunction with medical professionals and special educators in managing the care for this patient population.

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# Sudden Onset Reduced Vision in Acute Viral Meningoencephalitis

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Jesse Brown Veteran's Administration Medical Center, Chicago IL



## INTRODUCTION

A case of sudden onset vision reduction presented with systemic malaise and headache warranting a work-up that indicated meningoencephalitis. Symptoms and further testing suggested a viral etiology.

## CASE HISTORY

A 68 y/o AA male presented to the triage clinic with complaints of a frontal headache and fever worsening over the previous 17 days with associated reduced vision OS only. The patient had a past ocular history significant for OAG OU treated with latanoprost, which he self-discontinued when his visual symptoms began. The patient had his yearly DFE 8 weeks prior to onset at which he was correctable to 20/20 OD, OS and was recommended to RTC in 4 months for a HVF.

## PERTINENT FINDINGS

During the clinical exam, the patient had an altered mental status and suffered from malaise. He also admitted to an unintentional weight loss of 10 lbs over the past few weeks and night sweats. There was no temporal artery tenderness or jaw claudication.

A CT, MRI and Carotid Duplex were performed and came back within normal limits.

On the day of presentation, the patient had an ESR of 113 and a CRP of 4.60 (NL: 0.0-1.0mg/dl).

VDRL, RPR, HCV, HIV, RF, ANCA, ANA, anti-DNA, CCP, anti-SSA/SSB, SPEP, Quantiferon, West Nile CSF, VZV CSF, Enterovirus CSF, CMV CSF, EBV CSF, HSV CSF were all negative.

## DIFFERENTIAL DIAGNOSIS

- Acute Meningoencephalitis: Viral vs Bacterial
- Giant Cell Arteritis

	OD	OS
BCVA	OD: 20/30-2	OS: CF @ 2ft
Pupils	+ Direct & Consensual	+APD OS
EOMs	FROM	FROM
CVF	FTFC	Constricted to fixation 360°
IOP	19 mmHg	17 mmHg
ONH	Pink, distinct	1-2+ temporal pallor
FANG	NL	Delayed Filling



Figure 1. Fundus photo right eye



Figure 2. Fundus photo left eye with temporal pallor

## DIAGNOSIS & MANAGEMENT

The patient was referred immediately to the ER and later admitted to be co-managed with rheumatology and neurology. Diagnosis was originally based on medical history and examination. Initial treatment included IV: ampicillin, vancomycin, ceftriaxone, acyclovir and methylprednisolone. Neuroimaging was performed followed by an LP. CSF analysis found elevated WBCs and steroids were discontinued. Protein, glucose levels, cellular analysis, and serology were all WNL. Negative bacterial cultures lead to the antibiotics being stopped after 3 days. Acyclovir was continued for 7 days followed by a 7 day course of Valtrex due to presumed HSV.

Three days after initial presentation, BCVA improved to 20/20-3 OD, OS and the patient's CVF were FTFC OD, OS. A temporal artery biopsy was initially considered but was deemed unnecessary due to rapid recovery with antiviral therapy.

## DISCUSSION

Varied symptoms, course and outcome are possible with meningoencephalitis depending on the causative agent. Most commonly, headache, fever and malaise are present on initial exam which are also consistent with GCA. This patient, however, did not have jaw claudication, scalp tenderness, upper body stiffness, or a temporally located headache. Asymmetric reduced VA and optic neuropathy is rare and has not been correlated to severity of course in meningoencephalitis. Early detection and treatment is vital to reduce sequelae and mortality.

Given negative bacterial cultures, rapid recovery, focal neurological changes and altered mental status the patient was presumed to have HSV meningoencephalitis. If left untreated mortality rates are as high as 70% in this condition, making it the most fatal CNS condition of viral cause. HSV is the most common cause in the Western hemisphere, followed by VZV and EBV. Treatment should not be based on viral cultures, but rather initiated quickly based on clinical presentation.

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## ACKNOWLEDGEMENTS

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# Structure-Function Relationship of an Arachnoid Cyst as seen with MRI, Macular Ganglion Cell-Inner Plexiform Layer and Visual Field

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## BACKGROUND

Early detection of structural and functional changes in patients with glaucoma is necessary to prevent or delay disease progression which could lead to blindness. Historically, critical evaluation of the optic nerve and the retinal nerve fiber layer (RNFL) has been the primary methods for structural assessment in glaucoma. Optical coherence tomography (OCT) has greatly enhanced our ability to detect early structural changes to the RNFL often before functional loss of vision is detected with visual field testing. Assessment of the macular ganglion cell-inner plexiform layer (GCIPL) is possible with spectral domain optical coherence tomography (SD-OCT)<sup>1</sup> and studies have demonstrated the diagnostic accuracy of the GCIPL thickness and glaucoma.<sup>2,3</sup> Additionally, one study suggests that the GCIPL thickness is more valuable than the peripapillary RNFL thickness for detecting glaucomatous eyes with parafoveal VF defects.<sup>4</sup> GCIPL thinning has also been reported in patients with multiple sclerosis and may offer insight for monitoring for disease progression.<sup>5,6</sup> Presently, there is little evidence demonstrating structure-function correlation with other non-ophthalmic brain disorders. We present a case of a glaucoma patient with visual field loss resulting from a prominent arachnoid cyst.

Figure 1 and 2: Optic nerve photo of the right eye and left eye, respectively.



Figure 3 and 4: Humphrey visual field 24-2 showing a bilateral right hemianopia.



Figure 5: Cirrus SD-OCT nerve fiber layer analysis with diffuse thinning of the RNFL OD and temporal thinning OS.

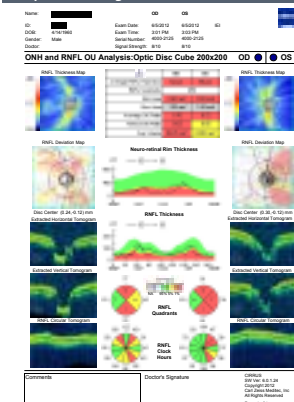


Figure 6: Cirrus SD-OCT Ganglion Cell Analysis shows nasal thinning OD correlating with the temporal hemianopia VF defect OD. The left eye GCA shows temporal thinning correlating with the nasal hemianopia VF defect OS.

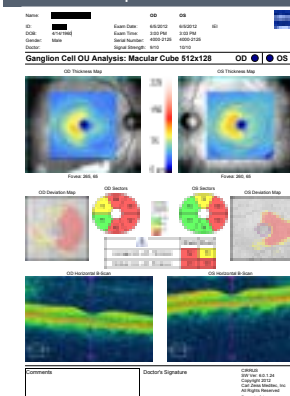


Figure 7: Axial view of a T<sub>1</sub> MRI image showing a fluid filled cyst contiguous with the posterior horn of the lateral ventricle.

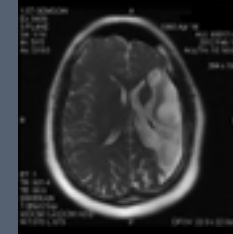
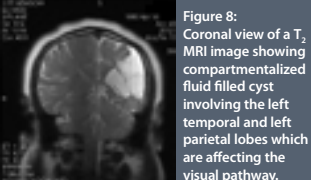


Figure 8: Coronal view of a T<sub>1</sub> MRI image showing compartmentalized fluid filled cyst involving the left temporal and left parietal lobes which are affecting the visual pathway.



## CASE REPORT

A 53 year-old African American male presented to the Illinois Eye Institute for a glaucoma evaluation. Medical history is significant for cerebral palsy, seizure disorder, hypertension and hypercholesterolemia. The patient reported taking the following medications: hydrochlorothiazide, Flomax, carbamazepine, and Zocor. The patient denied previous eye injury or surgery. Upon evaluation, best corrected visual acuity was 20/20 OD, OS at both distance and near. Pupils and EOMs were normal OU, but a bilateral right hemianopia visual field defect was elicited during confrontation fields. Slit lamp examination was normal. IOP was 38mmHg OD, 39 mmHg OS. Gonioscopy was open to the ciliary body in each eye. Optic nerve photos are shown in figures 1 and 2. Threshold perimetry results are shown in figures 3 and 4. Results of the retinal nerve fiber layer analysis and the ganglion cell analysis with the Cirrus SD-OCT are shown in figure 5 and figure 6, respectively. The patient was diagnosed with open angle glaucoma. Travoprost 0.004% was prescribed every evening in both eyes. Brimonidine

0.1% was added bid OU on a subsequent visit to reach a target IOP of the mid-teens. MRI of the brain with and without contrast was obtained. Figures 7-10 show a large left middle cranial fossa arachnoid cyst affecting the visual pathway. When compared to a previous MRI, no progression or changes were noted. The patient's IOP has been 12-14mmHg OU with his current treatment.

## CONCLUSION

Visual field loss can occur with arachnoid cysts. Monitoring for progression of VF loss can be challenging especially when significant VF loss has already occurred as with our patient. Our case demonstrates the structure-function relationship of the arachnoid cyst as seen by MRI, Cirrus SD-OCT ganglion cell analysis (GCA), and automated perimetry. The GCIPL thickness as measured by the GCA may offer an alternative method to monitor for structural changes to monitor for progression of functional loss of vision that result from non-ophthalmic disorders of the brain especially those that affect the visual pathway.

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# Value of Heidelberg Retina Tomograph in Diagnosing Optic Nerve Hypoplasia

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## PURPOSE

A common test for confirming optic nerve hypoplasia (ONH) is to evaluate the disc-macula distance to disc diameter ratio (DM:DD). We have published a case report showing Heidelberg Retina Tomograph (HRT) to be useful for assisting in diagnosis of ONH. The purpose of this study was to determine if HRT accurately diagnoses ONH, and the proper cutoff value for disc area to diagnose ONH. Furthermore, use of HRT was compared to DM:DD in diagnosis of ONH.

## METHODS

Thirty subjects were recruited. All subjects had comprehensive eye examinations and diagnosis of either unilateral or bilateral ONH, resulting in 42 eyes with ONH. Fundus photography and HRT were performed by one technician. DM:DD ratios were measured by one of the authors who was masked to subjects' other clinical data. A clinical cutoff of  $>3$  for DM:DD was used to assist in diagnosis of ONH. Three cutoffs for HRT disc area were chosen:  $1.60 \text{ mm}^2$  (= mean disc area-SD, used by HRT to indicate a small optic disc),  $1.32 \text{ mm}^2$  (=mean-1.50 SD), and  $1.07 \text{ mm}^2$  (=mean-1.96 SD). Pearson Chi-square test was used to determine the accuracy of HRT and DM:DD in diagnosing ONH.

FIGURE 1: Subject #27: Asian male, 12 years old, bilateral ONH, BCVA OD: 20/20 OS: 20/20

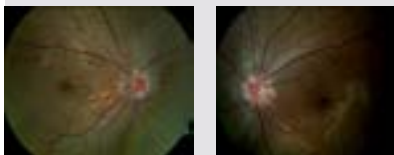


FIGURE 2: Subject #21: AA female, 8 years old, unilateral ONH (OS), BCVA OD: 20/20 OS: NLP

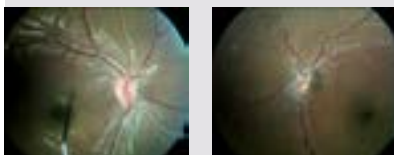


FIGURE 3: ROC Curve for HRT and DM:DD Ratio

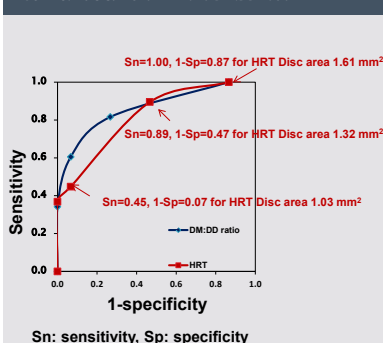


Table 1: Demographic Characteristics of ONH Subjects (n = 30)

	n (%)
<b>GENDER</b>	
Female	21 (70)
Male	9 (30)
<b>RACE</b>	
African American	19 (63.3)*
Hispanic or Latino	5 (16.7)
Caucasian	4 (13.3)
Asian	2 (6.7)
<b>AGE (YEARS)</b>	
Range	4.2 - 60.5
Mean (SD)	22.3

\*80.6% of the clinic population from which ONH subjects were enrolled was African American

Table 2: Classification of ONH (n=30)

	n (%)
<b>UNILATERAL ONH</b>	18 (60)
Right eye	2 (11)
Left eye	16 (89)
<b>BILATERAL ONH</b>	12 (40)

Table 3: General Ocular Characteristics of ONH Subjects (subjects = 30, ONH eyes= 42)

	n (%)
<b>MEAN VA</b>	20/80 (ranged from 20/20 to NLP)
<b>STRABISMUS</b>	8 (26.7)†
Exotropia	2 (25.0)
Esotropia	5 (62.5)
Hypertropia	1 (12.5)
<b>NYSTAGMUS</b>	3 (10.0)
Stereoacuity positive	14 (46.7) Stereo Fly
	10 (33.3) Preschool Random Dot (ranged from 20" to 400")
<b>APD (+)</b>	5 (11.9)
<b>(42 ONH EYES)</b>	
Double Ring Sign	26 (61.9)
Full	7 (26.9)
Partial	19 (73.1)
Vessel Tortuosity (42 ONH eyes)	10 (23.8)

† Cover test could not be performed in 3 subjects because of poor VA (no light perception).

## RESULTS

The demographic characteristics of our subjects are listed in Table 1. Table 2 shows the classification of ONH subjects. The general ocular characteristics of ONH subjects are described (Table 3). Figures 1 and 2 show fundus photos of two of our subjects. The mean DM:DD for ONH eyes was  $4.00 \pm 1.22$ . The mean HRT disc area was  $0.86 \pm 0.36 \text{ mm}^2$ . The cutoff of 3 for DM:DD was significant to diagnose ONH ( $\chi^2=19.55$ ,  $P<0.001$ ). Disc area of  $1.60 \text{ mm}^2$  was not significant to diagnose ONH ( $\chi^2=5.27$ ,  $P=0.08$ ); however, disc area of  $1.32$  was significant ( $\chi^2=11.25$ ,  $P<0.001$ ) as well as disc area of  $1.07 \text{ mm}^2$  ( $\chi^2=7.63$ ,  $P=0.01$ ). The accuracy of a HRT cutoff of  $1.32$  in diagnosing ONH was 89% vs. 68% for a HRT cutoff of  $1.07$ . In addition, there was a significant association between a HRT cutoff of  $1.32$  and a DM:DD ratio of 3 in diagnosing ONH ( $\chi^2=5.83$ ,  $P=0.03$ ). Figure 3 shows the Receiver Operating Characteristic (ROC) curves for DM:DD ratio and HRT. Area under the ROC curve was  $0.80$  ( $P=0.001$ , 95% CI:  $0.66-0.94$ ) for HRT and  $0.88$  ( $P<0.001$ , 95% CI:  $0.80-0.97$ ) for DM:DD, which indicates that both HRT and DM:DD ratio are valuable tests for diagnosing ONH.

## CONCLUSION

- Both HRT and DM:DD ratio are effective for detecting HRT.
- HRT can be used to assist in ONH diagnosis and the cutoff should be  $1.32 \text{ mm}^2$ , not  $1.60 \text{ mm}^2$ .
- The DM:DD ratio remains a useful clinical tool for diagnosis of ONH.

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# Bulbar Conjunctival Chemosis and Choroidal Folds in Posterior Scleritis Patient

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## BACKGROUND

Posterior scleritis is a rare, severe inflammatory disorder. Patients often present with severe periorcular pain, loss of vision, and redness. The fundus may demonstrate optic nerve swelling, retinal detachments, macular edema and/or choroidal folds. While 40-50% of scleritis patients have an associated autoimmune systemic disorder, less than 10% of scleritis cases have an infectious cause. Treatment for scleritis can include oral non-steroidal anti-inflammatory drugs (NSAID), oral steroids and/or immunosuppressive agents. Early diagnosis is essential to these cases in order to prevent permanent vision loss.

## PERTINENT FINDINGS

A 63 y/o African American female presented with severe pain and bulbar conjunctival chemosis in the right eye.

- Medical History: Hypertension, Diabetes Mellitus, Thyroid disorder, Osteoarthritis
- Medications: synthroid, ranitidine, metformin, lovastatin, lisinopril
- Allergies: Iodine
- Clinical
  - BCVA: 20/20 OD, OS
  - PERRL (-) APD
  - EOM FROM OU with pain on right gaze
  - SLE: 2+ bulbar injection with 4+ bulbar conjunctival chemosis OD (**Figure 1**), 1+ bulbar conjunctival chemosis OS (**Figure 2**)
  - DFE: Right eye reveals radiating alternating light (peaks) and dark (troughs) bands characteristic of choroidal folds in posterior pole. (**Figure 3**) Left eye shows drusen within the posterior pole. (**Figure 4**)
  - Hertel Exophthalmometry: 18 OD, 18 OS (base 10S)
  - SD-OCT: Right eye highlights choroidal folds. (**Figure 5**) Left eye is unremarkable. (**Figure 6**)
  - B scan: Right eye shows a positive T sign highlighting a thickened sclera. (**Figure 7**) Left eye is unremarkable. (**Figure 8**)
- Laboratory studies ordered:
  - FTA Abs, RPR, ANA, RF, ACE, Lysozyme, Quantiferon Gold
- Diagnosis: Posterior Scleritis
- Differential Diagnosis
  - Orbital Tumor
  - Retrobulbar Mass
  - Thyroid Eye Disease
  - Hypotony

FIGURE 1: 4+ bulbar conjunctival chemosis



FIGURE 2: 1+ bulbar conjunctival chemosis



FIGURE 3: radiating choroidal folds OD

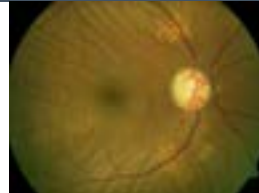


FIGURE 4: drusen in the posterior pole OS



FIGURE 5: choroidal folds OD

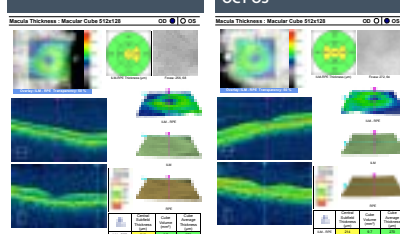
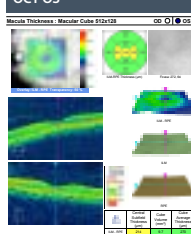


FIGURE 6: normal macular OCT OS



## DIAGNOSIS/TREATMENT

The patient was referred to ophthalmology for systemic blood work up to rule out autoimmune and/or infectious cause of posterior scleritis. Ophthalmology confirmed the diagnosis of posterior scleritis and began 800mg of ibuprofen three times a day. The patient's blood workup tested negative for all laboratory tests ordered with the exception of quantiferon gold. The patient was then referred to infectious disease for further evaluation. A chest x-ray was ordered and was normal. As a result, isoniazid was initiated for latent tuberculosis. One week follow up showed resolving choroidal folds and bulbar conjunctival chemosis with subjective improvement of periorcular pain.

## DISCUSSION

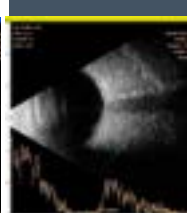
The presence of choroidal folds is an important sign of posterior scleritis. Choroidal folds are most often idiopathic; however, non-idiopathic causes must be ruled out in acquired choroidal folds, especially if presented unilaterally. Furthermore, ancillary testing, specifically a B scan ultrasound, is integral in the diagnosis of posterior scleritis. B scans of posterior scleritis demonstrate a characteristic T-sign, highlighting the thickened, inflamed sclera and choroid. In addition to early diagnosis, further autoimmune and systemic work up is pertinent due to the strong association to systemic disorders.

Conventional therapy includes oral NSAIDs, oral steroids and/or immunosuppressive agents. According to a clinical survey by Calthorpe et al, all patients are usually given an NSAID for treatment of posterior scleritis. If patients do not respond to the NSAID, an oral steroid, such as prednisolone 80-120mg is administered. Patients are started at this dose and tapered as the condition resolves. In their clinical

FIGURE 7: positive T-sign highlighting thickened sclera



FIGURE 8: normal B scan OS



survey, 14.9% of patients resolved with NSAID use alone. Of these patients, none had an associated systemic disease. 62.7% of patients were administered a systemic steroid, in which 36% had an associated systemic disease. Lastly, 23.4% were administered an immunosuppressive agent, in which 54% were associated with a systemic disease. To target treatment appropriately, infectious etiologies need to be ruled out before initiating therapy. Our patient responded well to NSAID use alone and did not require oral steroids. However, secondary to the diagnosis of latent tuberculosis, isoniazid treatment was initiated.

## CONCLUSION

The early diagnosis and treatment of posterior scleritis is essential in order to prevent permanent vision loss. B scan ultrasonography depicting an inflamed sclera and choroid is a key part in the diagnosis. Posterior scleritis has a strong association with systemic disease; therefore, non-infectious and infectious etiologies must be ruled out prior to initiating therapy. Conventional treatments include NSAIDs, steroids and/or immunosuppressive agents. Most patients with infectious etiology do not resolve with NSAID use alone and need an oral steroid or an immunosuppressive agent. It is important to address and treat the underlying systemic etiology of scleritis. Posterior scleritis can recur or become a chronic condition; therefore, it is necessary to monitor patients regularly.

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# TWO CASES OF LEVATOR DEHISCENCE CONFOUNDED BY CONTACT LENSES

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## BACKGROUND

Dehiscence of the levator aponeurosis (LDH) may occur normally in association with aging. In a study of anatomical variation in cadavers by Lim, 13 of 20 eyes had some level of LDH, most often nasal. LDH can be further complicated by orbital fat atrophy also seen in aging, prostaglandin use and HIV. In the presence of these conditions, the fornix enlarges creating a trap for debris and trap for bandage contact lenses(BCLs) as presented here.

In severe cases giant fornix syndrome (GFS) may develop. GFS was first identified by Dr. Rose in 2004. More recent case series offer CT scans to aid in diagnosis. In addition new surgical techniques have been developed to normalize the fornix and alleviate the large pocket. GFS usually occurs in the elderly. The condition is characterized by recurrent or recalcitrant infections. Cultures are most often positive for Staphylococcus species with only one case in the literature attributed to Pseudomonas.

## CASES

### PATIENT 1

On September 15, 2010 patient one presented complaining of having to pry her lids open in the morning. She was an 86-year-old African-American female who had lost vision in her left eye due to chronic angle closure glaucoma in 2003. In 2004 she developed a sclerosing keratitis or PUK that was treated with a bandage contact lens. Extensive bloodwork was also ordered and the diagnosis of Paget's disease was made. Ultimately the patient underwent an amniotic membrane transplant in late 2004. She was lost to follow-up and there is no note as to whether the BCL was ever removed from the eye. She had also been treated for filamentary keratitis, keratoconjunctivitis sicca and blepharitis. Her dry eye regimen included loteprednol, cyclosporine, doxycycline, moisture chamber goggles, bacitracin ointment, and punctal occlusion.

Upon examination, severe mucopurulent discharge was noted. She was started on ofloxacin every two hours. When there was no improvement in one week, she was cultured. Her cultures revealed many Pseudomonas aeruginosa with multiple resistance. **Figure 1.** She was changed to ciprofloxacin drops with tobramycin ointment at bedtime as suggested by the culture sensitivities. On October 13, she had significantly less discharge however her lower lid was red and severely swollen. Differentials were dacryocystitis versus preseptal cellulitis. Her primary care physician was consulted and systemic treatment was started with Cefdinir, a third generation cephalosporin. On October 16, she had only mildly improved so the culture was repeated and revealed less pseudomonas. She was followed closely and on November 10, 2010 her condition finally resolved.

The patient did well for several months but returned on April 7, 2011 with mucopurulent discharge. She was again positive for Pseudomonas and was given ciprofloxacin drops. In addition, the corneal ophthalmologist swabbed the patient's fornix with povidone iodine. On 4/21, she appeared to be about 60% better and the medicine was continued. However on 4/27 the dacryocystitis returned and she was started on oral ciprofloxacin. She was referred to oculoplastics where no nasolacrimal reservoir was found. Unfortunately her problems continued necessitating multiple fornix sweeps and oral antibiotics to minimize the discharge. On her third fornix sweep on July 7, 2011 what appeared to be a very old soft contact lens was retrieved from the superior fornix. **Figure 2** The swab went approximately 3 inches back, essentially behind the eye. Several additional fornix sweeps were performed and after two months all discharge resolved. She remains quiet.

Figure 1: Culture and sensitivity results for Patient 1



Figure 2: Contact lens recovered on fornix sweep of Patient 1



### PATIENT 2

An 82-year-old African-American male had been previously diagnosed with orbital fat atrophy likely secondary to prostaglandin use. **Figure 3.** His left eye was blind from glaucoma. In addition, he suffered from recurrent erosions(RCE) and filamentary keratitis for which BCLs had been used in the past. When he presented to clinic on March 6 complaining of grittiness, he had a small RCE and several filaments. **Figure 4.** The filaments were removed with a golf club spud and a BCL was applied. He was already using ofloxacin, so it was continued. Over the next two months, he presented with multiple RCEs for which BCLs were applied. However, at each follow-up visit no lens was noted. The caregivers reported never finding a lens. On April 24, there was significant mucous and filaments but again no BCL. Thus a fornix sweep was recommended to prevent further infection and determine if any lenses had been "lost" in the fornix. A povidone iodine swab was inserted into the upper fornix nasally and swept temporally, three times. It went in approximately 2.5 inches and swabbing recovered five BCLs which were still in good condition. Subsequently, patient has been taping his lids shut and punctal plugs have been inserted. His staining and filaments have improved. Referral for acetylcysteine therapy may be necessary if filaments return as bandage lenses are now contraindicated.

Figure 3: Profile image of patient 2. Deep set eye with orbital fat atrophy



## CONCLUSIONS

It is frequently stated that "a contact lens cannot go behind the eye". However in certain situations it can become captured in the enlarged fornix of patients with LDH and orbital fat atrophy. In these cases, a contact lens can become trapped resulting in GFS. This severe form of the condition is demonstrated in patient one. Performing fornix sweeps and removing five BCLs from the fornix of patient 2, has likely prevented GFS in this patient.

When managing the elderly who have need of bandage lenses suspicion should be raised when multiple lenses are lost and not recovered. Likewise elderly patients with recalcitrant infections should be evaluated for GFS. Diagnosis can be aided by CT scans looking for a pocket of air in the fornix. Fornix sweeps with povidone iodine can recover lost lenses, minimize the risk of and treat infection associated with GFS. In patients where infection is not controlled with fornix sweeps referral for surgical intervention to reconstruct the fornix may provide significant relief.

Figure 4: A large RCE on patient 2's left eye



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## CONTACT INFORMATION

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# Longitudinal Analysis of Iris Transillumination Defects in Pigment Dispersion Syndrome Using Infrared Iris Imaging

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## INTRODUCTION

Although mid-peripheral iris transillumination defects (ITDs) are a hallmark of "classic" type pigment dispersion syndrome (PDS), little is known about the natural history and stability of this clinical sign, mostly likely because sensitive methods to detect and record ITDs have not been readily available. We therefore analyzed serial iris photographs of PDS subjects belonging to an iris pathology database that had been collected during investigations of near infrared (NIR) iris transillumination imaging.

## METHODS

An NIR iris transillumination database was accumulated at a single urban eye care facility in Chicago, IL, U.S.A., during the development and study of digital imaging systems to examine iris disorders. We searched this database for any "classic" type PDS subjects who had archived images collected on more than one occasion, with at least 4 years between the earliest and latest imaging date. Analysis included side by side comparison of early/late images (Figure 1) using a three person consensus panel, as well as ITD mapping and quantification using custom designed MATLAB computer algorithms (Figure 2). This process involved alignment and adjustment of initial and late images so that direct comparisons could be made, selection of ITD areas, and then calculation of ITD area as a percentage of sector and total iris area.

## RESULTS

Analysis included 7 PDS subjects (2 females, 5 males), ranging from 28.4 to 67.6 years (median=56.1 years) at initial imaging (Tables 1 & 2). Length of time between initial and last imaging ranged from 4.4 to 7.0 years (median=5.4 years). At initial imaging, the median ITD area as a percentage of total iris area was 7.4% (1.1 to 16.1%) for right eyes and 3.2% (0 to 12.6%) for left eyes. Difference in ITD total area between the early/late imaging was median=-0.4% (-3.0 to 6.6%) for right eyes and +0.3% (-2.0 to 4.1%) for left eyes. Using gross visual inspection, there was agreement among the consensus panel that overt differences in ITD appearances could not be detected between the early/late images for each subject.

## DISCUSSION

To our knowledge, no other work has attempted to evaluate the stability of ITDs associated with PDS. Thus, although this dataset is small, it helps gain some initial insight into the natural history of ITDs, as well as methods that can be used for further study. Questions currently exist regarding the variability of ITD patterns, their age of onset, change with time, and whether they have any predictive value toward level of disease activity and/or the development and progression of associated glaucoma. Since laser iridotomy has been advocated to eliminate reverse pupillary block in PDS, evaluation of ITDs using the methods we describe may have value in the study of such procedures that may help lessen posterior iris bowing and subsequent iris pigment debris.

## CONCLUSIONS

In this initial attempt to longitudinally study PDS-related ITDs using NIR iris imaging, we observed relative stability of ITDs over a several year period. These initial observations, along with methods used, may be helpful to future studies of PDS.

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FIGURE 1: Initial and late ITD images belonging to subjects used in the analysis.

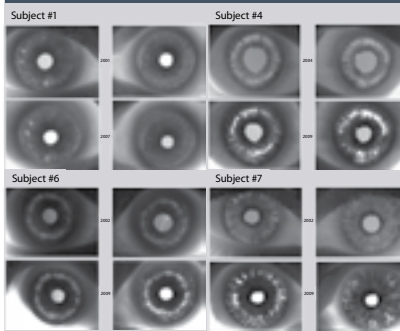


TABLE 1: ITD Percent AREA DIFFERENCES BY SUBJECT (Area at Age 1 minus Area at Age 2)

Subject	Gender	Race	Age1	Age2	Years Elapsed	ITD Area Difference Right Eye	ITD Area Difference Left Eye
1	Male	AA	67.6	73.0	5.4	-0.4	0.0
2	Male	W	51.6	56.0	4.4	-1.3	+
3	Female	W	28.4	32.8	4.4	-0.5	0.0
4	Male	W	56.1	60.8	4.7	1.5	3.7
5	Male	H	57.7	63.1	5.4	-3.0	-2.0
6	Female	AA	62.0	69.0	7.0	1.8	4.1
7	Male	W	49.1	56.1	6.9	6.6	0.6

\*Abbreviations: AA, H, W, African-American, H, Hispanic, W, White; ITD, iris transillumination defect; <sup>+</sup>Excluded Subject 2 left eye comparison due to poor Age 2 image.

FIGURE 2: Method of image/ITD analysis.

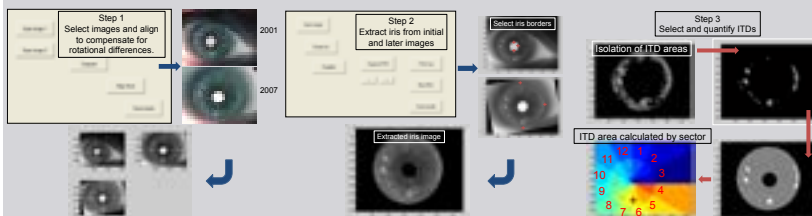


TABLE 2: SUMMARY OF TOTAL ITD AREAS AND DIFFERENCES (Total ITD Area / Total Iris Area) x 100

Eye	Number of Eyes	Image Collection	Mean ITD Area	Median ITD Area	Std Dev	Min	Max
Right	7	Age 1	7.6	7.4	5.4	1.1	16.1
Right	7	Age 2	8.2	9.1	5.9	0.6	1.8
Right Difference	7	-	0.7	-0.4	3.1	-3.0	6.6
Left	6	Age 1	4.6	3.2	5.3	0.0	12.6
Left	6	Age 2	5.6	2.6	7.0	0.0	16.3
Left Difference	6	-	1.1	0.3	2.4	-2.0	4.1

\*Abbreviations: ITD, iris transillumination defect; Min, Max, minimum, maximum; Std Dev, standard deviation; <sup>+</sup>Excluded Subject 2 left eye comparison due to poor Age 2 image.

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Support: NEI EY015604  
Conflict of Interest: Employee, consultant, patent

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# Putting the Pieces Together: A Case of Inherited Retinal Disease

Josh Robinson, O.D.; Robert Chun, O.D.

Illinois College of Optometry

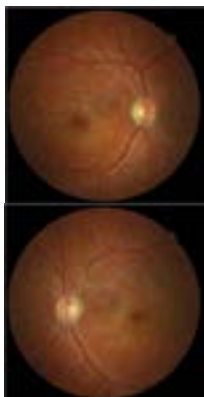
## BACKGROUND

In 1909, Karl Stargardt described the clinical appearance of yellow, pisciform-shaped lesions or flecks in the macula of seven patients from two families. The macular dystrophy that he described generally presents between the ages of 8 and 16 years and became known as Stargardt disease. It is the most common form of juvenile macular dystrophy and results in a bilateral reduction in visual acuity, central scotomata, photostereopsis, dyschromatopsia, and dark adaptation difficulty. The autosomal recessive form of Stargardt disease is associated with a mutation in *ABCA4*, while the much less common dominant form involves *ELOVL4*.

## PATIENT PRESENTATION

A 34 y/o Caucasian female was referred by her retinal specialist for "salt and pepper retinopathy." She reported a progressive decline in central vision, depth perception,

FIGURE 1: Fundus photos exhibiting bull's eye macular appearance with partially resorbed lipofuscin flecks OD, OS



and color vision. She denied any symptoms of nyctalopia or peripheral field defects. There was no history of ocular trauma or surgery. Her medical history was unremarkable. She denied taking any medications. There was no family history of retinal disease.

## CLINICAL TESTING

**BCVA:** 20/120 OD, OS

**ISHIHARA:** Sees only test plate OD, OS

**SLIT LAMP EXAM:** Unremarkable

**DFE:** Bull's eye macular appearance with diffuse partially

FIGURE 2: OCT results showing macular thinning and disrupted inner segment ellipsoid band with peripapillary sparing OD, OS

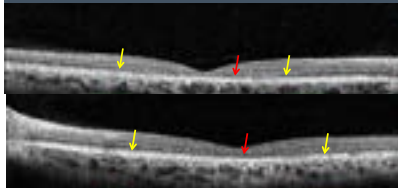
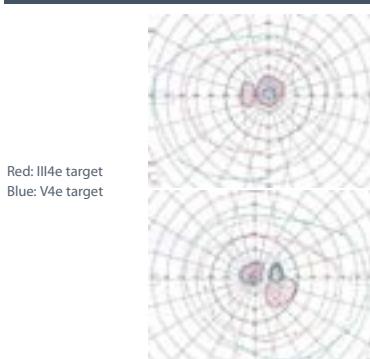


FIGURE 3: Central scotoma on Goldmann Visual Field testing OD, OS



resorbed fundus flecks extending anterior to the vascular arcades OD, OS (see Figure 1)

**SPECTRAL DOMAIN OCT:** Marked macular thinning with disrupted photoreceptor inner segment ellipsoid band in the fovea and peripapillary sparing OD, OS (see Figure 2)

**GOLDMANN VISUAL FIELDS:** Central scotoma with mild constriction to the size III4e target OD, OS (see Figure 3)

**ERG:** Reduced rod and cone function OD (see Figure 4)

**GENOTYPING:** Compound heterozygous *ABCA4* (L541P exon 12; A1038V exon 21; D576H exon 12)

**DIAGNOSIS:** Stargardt disease – stage 2/3

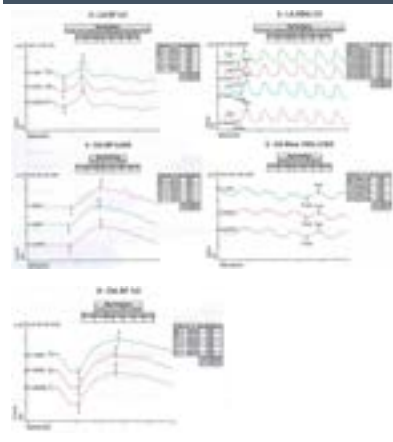
## DISCUSSION

Patients with Stargardt disease may be classified according to four distinct stages outlined by Fishman in 1976:

Stage 1 - flecks limited to macula with or without an atrophic macular lesion

Stage 2 - diffuse flecks anterior to vascular arcades, some partially or totally resorbed

FIGURE 4: ERG results indicating reduced rod and cone function OD, OS



Stage 3 - diffuse, totally resorbed flecks often with choriocapillaris atrophy in the macula

Stage 4 - diffusely resorbed flecks with extensive choriocapillaris and RPE atrophy

Based on the clinical presentation, findings, OCT imaging, visual field results, and genotyping, a diagnosis of Stargardt disease was made. Specifically, this patient was categorized as having late stage two or early stage three Stargardt disease.

## PATHOPHYSIOLOGY

A genetic mutation in the *ABCA4* gene causes a rim protein dysfunction that results in an accumulation of toxic bisretinoid compounds in the outer segments of photoreceptors. As retinal pigment epithelial (RPE) cells phagocytose the outer segments of photoreceptors, the compound causes subsequent cellular damage to the RPE. Histological studies show that the pisciform lesions or flecks represent aggregates of swollen RPE cells inflated with lipofuscin. Intravenous fluorescein angiography (IVFA) often reveals a "silent choroid" as lipofuscin in the RPE cells blocks underlying choroidal fluorescence.

## CONCLUSION

Goldmann visual field testing, electroretinography, SD-OCT imaging, and genetic analysis are valuable tools to aid in making a diagnosis of Stargardt disease. Proper treatment and management includes patient education regarding prognosis, low vision rehabilitation options, genetics, and emerging clinical trials.

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# Diagnosing Goldenhar Syndrome by the Presence of Dermoid Cysts

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## BACKGROUND

Epibulbar dermoid cysts are congenital choristomas that are fairly rare. The cyst wall can contain dermal appendages such as hair shafts, sebaceous glands and sweat glands. They may present unilaterally or bilaterally and are most frequently located on the inferior temporal limbus. While dermoid cysts are benign in nature, a correct diagnosis is important because they can be associated with Goldenhar syndrome which has the classic triad of signs of epibulbar dermoids, preauricular appendages and pretragal fistulas. Goldenhar syndrome also has multiple associated systemic findings such as vertebral, cardiac, renal and pulmonary defects.

## CASE REPORT

A 12-year-old Hispanic male presented for a comprehensive eye exam to evaluate the "bumps" on both eyes that had been present since birth but had recently started becoming itchy and red. He reported a history of esotropia with surgical correction, surgery on his mouth as a newborn because it wasn't fully formed, and heart problems noted shortly after birth. There was no family history of facial or other congenital anomalies. All entrance testing was normal. His refractive error was myopic with no astigmatism in either eye. The anterior segment exam showed orbital neoplasms on the temporal bulbar conjunctiva OU with multiple hairs in the center OD (Figure 1) and cysts on the surface OS (Figure 2). The eyes were otherwise healthy. On cutaneous examination, a surgical scar for a probable facial cleft was noted on the right side of his mouth to his right ear and he had two auricular appendages on the right ear (Figure 3). His facial features were typical of right hemifacial microsomia (Figure 4). The patient was diagnosed with conjunctival dermoid cysts. We started him on olopatadine hydrochloride 0.2% ophthalmic solution and referred him to an oculoplastics doctor at his request for possible cosmetic removal of the dermoid cysts. We told the patient he likely had Goldenhar syndrome and it was important that he see his physician to do further radiologic scans to rule out other associated systemic abnormalities. His father later confirmed that his medical doctor agreed with the diagnosis. Besides the congenital heart defect, no other systemic abnormalities were found.

## DISCUSSION

Epibulbar dermoids can be unilateral or bilateral and are most commonly found at the inferior temporal limbus. They are marginally vascularized, smooth, white or yellow lesions that grow very slowly if at all. They are thought to arise from an early embryological anomaly (occurring at 5-10 weeks gestation) resulting in metaplastic transformation of the mesoblast between the rim of the optic nerve and surface ectoderm. Histologically, they contain choristomatous tissue such as hair follicles, fat, muscle, cartilage, brain, teeth and bone. The most common ocular complications of epibulbar dermoids are amblyopia, cosmetic concerns, exposure keratopathy, ocular irritation and dellens. The major indication for treatment is cosmesis. If the lesion is amblyogenic, either by occlusion of the visual axis or by induction of astigmatism, surgical excision should be strongly considered. They are frequently an isolated anomaly but in about one-third of cases they are seen in association with congenital abnormalities such as Goldenhar syndrome.

Figure 1



Figure 2



Goldenhar syndrome was identified by Maurice Goldenhar, a Swiss ophthalmologist, in 1952 and included preauricular appendages, pretragal fistulas (malformed ears) and epibulbar dermoids. Gorlin and associates included vertebral anomalies as signs of the syndrome and suggested the name oculoauriculovertebral (OAV) dysplasia for the condition in 1963. It has been established to be sporadic and non-inherited; occasional evidence of familial transmission and genetic or chromosomal abnormalities has been described. The incidence of the syndrome has been reported to be between 1:3500 and 1:5600 with a male to female ratio of 3:2. It is a congenital anomaly involving the first and second branchial arches. The exact etiology is unknown, but it is believed to be due to abnormal embryonic vascular supply to the first arch and abnormality of mesoblastic development affecting the formation of the branchial and vertebral system. It is a disorder where the patient's facial features are incompletely developed on one side, resulting in eye, ear and jaw abnormalities as well as multiple systemic anomalies such as cardiac, pulmonary, central nervous system, renal and vertebral abnormalities (Figure 5).

Figure 3



Figure 4



Figure 5: Classic Features of Goldenhar Syndrome

**Ocular anomalies** (~ 60% of cases)  
epibulbar dermoid or lipodermoid, coloboma, microphthalmia, strabismus  
**Ear anomalies** (~ 40% of cases)  
hearing loss, preauricular skin tags or blind fistulas, microtia  
**Facial anomalies**  
hemifacial microsomia, cleft lip & palate, prominent forehead  
**Vertebral anomalies** (~ 40% of cases)  
scoliosis, hemivertebrae, hypoplastic or fused ribs  
**Systemic abnormalities** (~ 50% of cases)  
**cardiac** - ventricular septal defect, atrial septal defect, Fallot's tetralogy  
**pulmonary** - tracheoesophageal fistula  
**central nervous system** - occipital encephalocele, microcephaly  
**renal** - ectopic kidneys, ureteropelvic junction obstruction

## CONCLUSION

Clinicians must be able to diagnose conjunctival dermoid cysts and know that they can be associated with Goldenhar syndrome in about one-third of cases. Goldenhar syndrome has many systemic malformations such as congenital cardiac anomalies that are potentially lethal and others including deafness, scoliosis, and cleft lip or palate which may hinder the normal developmental progress of these children unless detected and treated through a multidisciplinary approach at an early age.

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# Oculomotor Nerve Palsy Due to Metastatic Space Occupying Lesion and Concomittant Vasogenic Edema of the Pons and Midbrain

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## BACKGROUND

Oculomotor nerve (CN III) palsy resulting secondarily from diabetes mellitus, aneurysmal compression, or cavernous sinus lesion is well documented. Our patient presented with CN III palsy secondary to a metastatic lesion involving the lateral aspect of midbrain and pons.

## CASE REPORT

A case of a 49-year-old female with history of stage 4 breast cancer 10 years prior is being reported who presented with a complaint of intermittent recent onset diplopia. Pupil and extraocular motility testing revealed a partial right CN III palsy and relative pupil sparing. Figures 3a – 3i show the partial CN III palsy in the 9 positions of gaze.

**Figure 1: CNIII Pathway:** The CNIII nucleus (red arrow) lies within the dorsal midbrain. CNIII (green arrow) exits the ventral midbrain and enters the Cavernous Sinus (CS) passing between the superior cerebellar artery and posterior cerebral artery and lies most superiorly within the CS. In the orbit, the preganglionic parasympathetic fibers to the ciliary ganglion travel with the inferior division of CNIII. The trigeminal ganglion (blue arrow) is located in the Meckel's cave inferior to CNIII.  
Reference: AMIRSYS STATdx, www.statdx.com



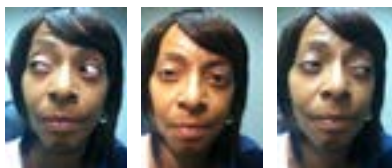
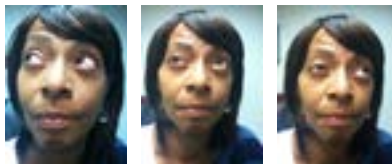
**Figure 2: CNIII Nucleus:** The CNIII nucleus (red arrow) lies in the dorsal midbrain at the level of the superior colliculus (green arrow) and is located anterior to the cerebral aqueduct (blue arrow). CNIII (yellow arrow) exits the ventral midbrain and enters the interpeduncular fossa.  
Reference: AMIRSYS STATdx, www.statdx.com



Trigeminal nerve (CN V) involvement was also suspected based on patient's complaint of sensory loss along her oral cavity and weakness of muscles of mastication on her right side. The atrophied muscles of mastication can be appreciated in all the facial photos (Figures 3a – 3i).

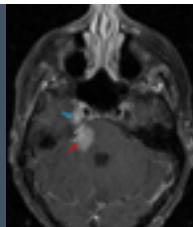
Patient was referred to her primary care physician and oncologist to rule out an aneurysm or brain or orbital metastasis. Imaging studies revealed a well-circumscribed, homogeneously enhancing solid mass, consistent with neoplasm, centered within the right lateral aspect of the midbrain and pons (See Figures 4, 5a and 6a). Tumor extension through the right Meckel's cave along CN V3 was also noted (see Figures 4, 5b, 5c, 6b and 6c).

Figures 3a – 3i: Nine positions of gaze: show an incomplete CN III Palsy.

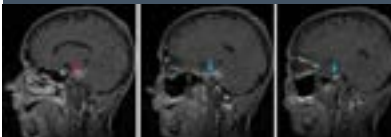


Local mass effect resulting from concomitant vasogenic edema of the entire right half of the midbrain (See Figure 7a) and pons (See Figures 7b and 7c) was also noted. Severe atrophy of the muscles of mastication on the right side including the masseter, temporalis, medial pterygoid and lateral pterygoid, with fatty infiltration as a result of chronic neuropathy of the mandibular division of trigeminal nerve secondary to tumor invasion, was also observed (See Figure 8). The orbits including the globes, extraocular muscles and optic nerves appeared normal.

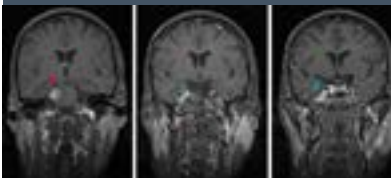
**Figure 4: MRI T1 post-contrast axial image:** shows an enhancing mass centered in the right midbrain / pons (Red arrow) with extension along the right Meckel's cave (Blue arrow).



**Figures 5a, b, c: MRI T1 post-contrast sagittal images:** show an enhancing mass centered in the right midbrain / pons (red arrow) with extension through the right Meckel's cave along V3 (blue arrow).



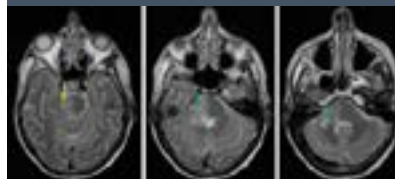
**Figures 6a, b, c: MRI T1 post-contrast coronal images:** show an enhancing mass centered in the right midbrain / pons (red arrow) with extension through the right Meckel's cave along V3 (Blue arrow).



## CONCLUSION

Midbrain space-occupying lesion at the level of oculomotor nerve nucleus or fascicle should be considered as a differential diagnosis for patients presenting with CN III palsy, especially with a history of carcinoma and/or signs of involvement of multiple cranial nerves.

Figures 7a, b, c: MRI T2 axial images: show hyper-intensity in the right midbrain, pons and brachium pontis consistent with vasogenic edema in the right midbrain (orange arrow) and the right pons and brachium pontis (green arrow) due to the mass.



**Figure 8: CT scan with contrast - Axial Image:** CT post-contrast image shows severe atrophy of the muscles of mastication including the right lateral pterygoid (Red arrow), the right medial pterygoid (Blue arrow) and the masseter muscle (Green arrow) due to chronic V3 denervation / neuropathy. Compare to the normal left side.



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# Therapeutic Scleral Lenses: Treatment of Ocular Graft-Versus-Host-Disease

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## BACKGROUND

In patients with Graft-versus-host-disease (GVHD) secondary to stem cell or bone marrow transplant, host antigens activate donor immuno-competent cells, instigating inflammatory processes. Nearly 60-90% of these patients develop debilitating ocular complications, which may include severe chronic keratoconjunctivitis sicca, cicatricial lagophthalmos, persistent corneal epithelial defects, corneal ulceration, and keratinization of the conjunctiva and cornea. Further complications may include perforation, scarring, and permanent vision loss. Current treatment for ocular GVHD focuses on supportive therapy with topical lubricants, steroids, and immune-modulators in addition to punctal occlusion and lid hygiene. This case summarizes a patient with ocular GVHD who was fit with scleral lenses to improve vision and rehabilitate her ocular surface.

Fig. 1: Topography OD, minimal astigmatism

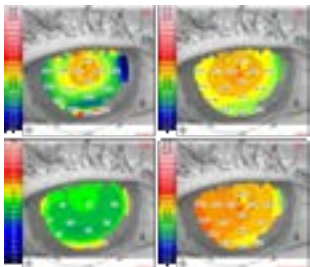
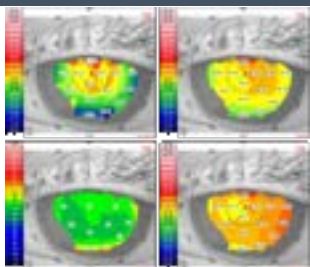


Fig. 2: Topography OS, two diopters of with-the-rule astigmatism



## CASE SUMMARY

A 51 year old Caucasian female was referred for a therapeutic contact lens fit secondary to ocular GVHD. She had a bone marrow transplant to treat leukemia five years prior. She had complaints of foreign body sensation, pain, mucus discharge, and extreme photophobia in both eyes, which was persistent for the last six months. She reported no relief with topical lubricants and punctal plugs. Ocular history was remarkable for corneal abrasions in both eyes three weeks prior. Corrected visual acuities with the patient's habitual glasses were 20/25 OD and 20/40 OS, no improvement with pinhole. Manifest refraction was stable to the patient's glasses: OD -2.00 -0.50 x 125 with a VA of 20/25 and OS -2.00 -0.50 x 090 with a VA of 20/40. Topographies showed a relatively spherical cornea OD and two diopters of with the rule astigmatism OS (Figures 1 and 2). Corrected visual acuities throughout the follow up period fluctuated consistently between 20/80 and 20/40 for both eyes. Biomicroscopy findings in both eyes revealed meibomian gland dysfunction, conjunctival chemosis and

injection, microcystic corneal edema, and diffuse punctate epithelial erosions with sodium fluorescein and lissamine green staining (Figures 3 and 4). Posterior segment findings were within normal limits.

Assessments of keratoconjunctivitis sicca and graft versus host disease were made. The patient was instructed to continue supportive therapy in both eyes – including Restasis twice per day, preservative free tears every hour, and lubricating ointment at bedtime. She was also fit with 18.2-millimeter diameter Jupiter lenses. The peripheral zones were flattened to achieve optimal conjunctival landing and 200-250 microns of vault were maintained to ensure an optimal tear reservoir (Figures 5-7). Within months of scleral lens wear, the patient's VA stabilized to a consistent 20/40 OD, 20/30+ OS, anterior segment inflammation calmed (Figure 8), and the patient's symptoms improved drastically. Therapeutic treatment is ongoing and the patient is still being monitored regularly.

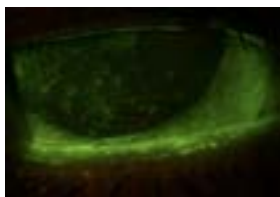


Figure 3: Diffuse corneal punctate epithelial erosions staining with sodium fluorescein



Figure 4: Conjunctival chemosis and injection



Figure 7: Adequate lens edge



Figures 5 and 6: Scleral lens demonstrating 200 to 250 microns of apical clearance

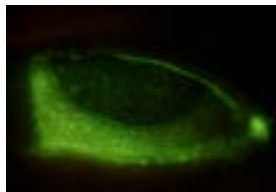


Figure 8: Post-lens staining

## DISCUSSION

Clear and comfortable vision may be achieved using mini-scleral or scleral gas permeable lenses. These lenses can contribute to the management of ocular surface disease by allowing more tear exchange from the vaulting of the lens and providing increased comfort due to the larger size. These lenses also provide refractive correction for the irregular and regular astigmatism. The tear film reservoir accumulated under the lens provides optical assistance for clearer vision. Irregular corneal changes from ocular surface disruption can often times make it more challenging to fit with regular contact lenses. Larger diameter gas permeable contact lenses provide protection to the eye, acting as a barrier for insubstantial corneas against inflamed tissue of the lids. The size of these lenses also improves comfort as the lid margin interacts with the surface of the lens rather than the edge. The use of mini-scleral and scleral contact lenses has become a major indication for individuals who not only need a refractive correction but also suffer from irregular corneas and severe ocular surface disease.

## CONCLUSION

Our findings suggest that scleral contact lenses are a promising management options for patients with ocular GVHD. Due to the severe complications associated with this condition, patients with GVHD require a comprehensive management approach. In addition to supportive therapy, scleral lenses can aid in rehabilitating the ocular surface, minimizing patient symptoms, and improving the quality of vision; thus helping sustain optimum vision potential.

References: Available upon request.

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3241 South Michigan Avenue, Chicago, Illinois 60616

# Comparison of Visual Field and Contrast Sensitivity in TBI and non-TBI Subjects

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## INTRODUCTION

Injury to the visual system is common in traumatic brain injury (TBI) and can occur by several mechanisms. Deficits in contrast sensitivity and visual field, although not apparent to the patient, may occur following a TBI. If left undiagnosed, these can have significant consequences on rehabilitation and quality of life. The purpose of this study is to determine if there is a difference in contrast sensitivity (CS) and visual field (VF) between patients that have sustained a mild or moderate TBI and those that have not.

Table 1				
	All Subjects	Non-TBI	TBI	p-value
n=60	n=30	n=30		
Age, years, median (IQR)	31 (25-37)	31(27-36)	29 (24-39)	0.554
Gender (male), n (%)	53 (88%)	23 (77%)	29 (97%)	0.052
Months from TBI to Testing, median (IQR)	N/A	N/A	4.7 (3.3-19.5)	
Visually Symptomatic, n (%)	24/54 (44%)	5/25 (20%)	19/29 (66%)	0.001
PTSD, n (%)	10 (17%)	1 (3%)	9 (30%)	0.012

\*Visually Symptomatic: double vision/blurred vision/photophobia/decreased peripheral vision/asthenopia/eye movement difficulty.  
\*Post Traumatic Stress Disorder (PTSD) diagnosed at the time of testing.

Table 2	
CAUSE OF TBI	
Blast	20
Fall	3
Motor Vehicle Accident	4
Parachute/Jump Training Related	2
Explosively Formed Projectile	1
PTSD, n (%)	10 (17%)

Table 3			
	Non-TBI n=30	TBI n=30	p-value
Contrast Sensitivity			0.619
OD	1.72 (1.68 - 1.76)	1.72 (1.68 - 1.84)	0.724
OS	1.76 (1.68 - 1.76)	1.76 (1.64 - 1.84)	0.757
OU	1.82 (1.76 - 1.88)	1.76 (1.72 - 1.88)	0.196
Mean Deviation			0.001
OD	0.23 (-0.39 to 1.94)	-2.47 (3.69 to -0.63)	0.001
OS	0.23 (-2.16 to 1.62)	-3.05 (-6.10 to -0.02)	0.003
Pattern Standard Deviation (PSD)			0.009
OD	2.71 (2.35 - 3.13)	3.42 (2.91 - 3.74)	0.004
OS	2.83 (2.59 - 3.10)	3.62 (3.05 - 4.78)	0.021
Total PSD			0.056
OD	1 (0 - 2)	2 (0 - 4)	0.080
OS	1 (0 - 2)	3 (0 - 9)	0.035

Data presented as median with IQR (25th percentile - 75th percentile) or count with (%).

\* p value for GEE includes both OD and OS outcomes in a multivariate model

## METHODS

A cross-sectional study design was used to compare differences in contrast sensitivity and visual field between the mild and moderate TBI subject population and the non-TBI population. Subjects from the active duty and retired military population were recruited in each group, 34 in the TBI group and 32 in the non-TBI group. CS, Mean Deviation (MD), Pattern Standard Deviation (PSD) and location of defects were compared using the Wilcoxon rank sum test, Fisher's exact test (univariate analysis per eye) and generalized estimating equations (GEE) for both eyes. Confounding effects of visual symptoms, PTSD, and injury by blast were explored using GEE.

**Table 4: Examined the 30 subjects with TBI, those with PTSD demonstrated significantly higher PSD**

	TBI No PTSD n=21	TBI With PTSD n=9	p-value
Contrast Sensitivity			0.922
OD	1.72 (1.68 - 1.76)	1.84 (1.72 - 1.88)	0.234
OS	1.76 (1.64 - 1.76)	1.88 (1.48 - 1.88)	0.385
OU	1.76 (1.72 - 1.84)	1.88 (1.76 - 1.88)	0.244
Mean Deviation			0.392
OD	-2.30 (-3.55 to 1.14)	-3.32 (-7.26 to -1.89)	0.216
OS	-3.66 (-4.10 to -1.06)	-2.99 (-5.82 to -2.88)	0.759
Pattern Standard Deviation			0.023
OD	3.21 (2.60 - 3.57)	3.71 (3.52 - 5.94)	0.012
OS	3.28 (2.45 - 4.52)	4.06 (3.72 - 4.99)	0.062
Total PSD			0.043
OD	1 (0 - 3)	2 (2 - 18)	0.091
OS	2 (0 - 8)	7 (8 - 15)	0.068

Data presented as median with IQR (25th percentile - 75th percentile)

\* p value for GEE includes both OD and OS outcomes in a multivariate model

**Table 5: TBI subjects injured by a blast related incident had similar CS as those that had been injured by other mechanisms, such as a fall or motor vehicle accident. However, the MD trended lower in blast related TBI patients with a significantly higher PSD (P=0.024).**

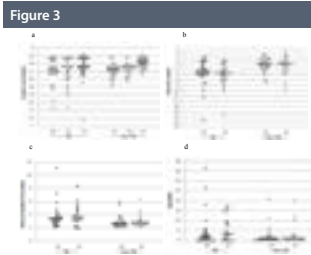
	TBI No Blast n=10	TBI Blast n=20	p-value
Contrast Sensitivity			0.876
OD	1.74 (1.68 - 1.76)	1.72 (1.68 - 1.84)	0.768
OS	1.76 (1.64 - 1.76)	1.76 (1.62 - 1.86)	0.800
OU	1.76 (1.72 - 1.88)	1.76 (1.72 - 1.88)	0.974
Mean Deviation			0.088
OD	-2.29 (-3.31 to 1.34)	2.89 (-5.54 to -1.39)	0.274
OS	-1.61 (-4.22 to 1.30)	3.96 (-6.10 to -2.88)	0.041
Pattern Standard Deviation			0.065
OD	3.12 (2.91 - 3.74)	3.45 (2.96 - 3.63)	0.601
OS	3.05 (2.39 - 3.37)	3.85 (3.42 - 4.99)	0.046
Total PSD			0.024
OD	1 (0 - 3)	2 (1 - 5)	0.485
OS	1 (0 - 2)	5 (3 - 15)	0.041

Data presented as median with IQR (25th percentile - 75th percentile)

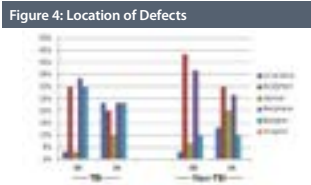
\* p value for GEE includes both OD and OS outcomes in a multivariate model

## RESULTS

Of the 66 subjects enrolled, there were 30 subjects in both the TBI group and non-TBI group between the ages of 21 and 57. Of the TBI group, all were classified as mild TBI (mTBI) except for 2 that were moderate. The median interval from the time of TBI to study testing was 4.7 months (3.3-19.5). The median age in the TBI group was 29 years (range: 21-57) and 31 years (range: 21-54) for the non-TBI group. There was no statistically significant difference in contrast sensitivity between the groups (median CS in OD eye for both groups=1.72, p=0.72). Analysis of visual fields showed a significant difference in MD (TBI median in OD eye=-2.47 vs. non-TBI=-0.23, p=0.001; TBI median OS=-3.05 vs. non-TBI=-0.23, p=0.003) and PSD (TBI median in OD eye=3.42 vs non-TBI=2.71, p=0.004). There was no significant difference in location or categorical defects (quadrant, hemianopia, etc) between groups.



**Table 3 and Figure 3** summarize the results of the visual field and contrast sensitivity (CS) testing between the two groups. Median contrast sensitivity was similar between groups, but there was a significant difference in the variability of CS values between TBI and non-TBI subjects for both the OD (P=0.038) and OS (P=0.001) eyes, Figure 3a. With regard to visual field, the TBI group had a significantly depressed Mean Deviation (MD) and elevated Pattern Standard Deviation (PSD).



**Figure 4: Location of Defects**

## DISCUSSION

Visual field defects have long been associated with TBI but are likely to occur in the more severely injured and those sustaining a cerebral vascular accident.[1, 2-6] This study did find a significant difference between Mean Deviation (MD) and Pattern Standard Deviation (PSD) but no significant difference in location of the defects and no categorical (ie, quadrantanopia) defects in either group. A reduction in sensitivity can accompany many ocular and neurologic pathologies and optic nerve damage has been noted to occur in TBI.[7] However, our study participants had ocular pathology ruled out on enrollment.

The two most likely causes are an overall neurological disruption from the TBI slowing or altering the visual pathways or a lack of focus during testing by the patient reducing the reliability of testing.[8-10] The results of this study were affected by the presence of Post Traumatic Stress Disorder (PTSD) in the TBI group. PTSD is an anxiety disorder characterized by re-experiencing, avoidance, and hyperarousal symptoms following exposure to a traumatic event. [11] PTSD has been linked to TBI and Post-Concussion Disorder (PCD). [13] Exploratory analysis in this study showed patients with both TBI and PTSD had significantly worse visual field results than those with TBI alone.

**Table 6: Those subjects with visual symptoms documented in their records had significantly lower CS and depressed MD, and tended to have elevated PSD and total PSD when compared to those without visual symptoms. However, there were no classical defects such as quadrantanopia or hemianopia.**

	TBI No Visual Symptoms n=10	Non-TBI Visual Symptoms n=19	p-value
Contrast Sensitivity			0.031
OD	1.74 (1.69 - 1.84)	1.72 (1.52 - 1.79)	0.288
OS	1.76 (1.72 - 1.84)	1.72 (1.52 - 1.84)	0.256
OU	1.76 (1.76 - 1.88)	1.76 (1.68 - 1.88)	0.234
Mean Deviation			0.022
OD	-1.76 (-2.47 to 0.94)	-3.44 (-6.82 to -1.89)	0.031
OS	-1.08 (-4.25 to -1.06)	-3.66 (-6.28 to -2.22)	0.283
Pattern Standard Deviation			0.194
OD	2.80 (2.42 - 3.68)	3.49 (3.30 - 3.95)	0.072
OS	2.39 (2.01 - 3.26)	3.97 (3.28 - 4.99)	0.015
Total PSD			0.160
OD	1 (0 - 2)	3 (1 - 5)	0.076
OS	0 (0 - 3)	4 (1 - 10)	0.085

Data presented as median with IQR (25th percentile - 75th percentile)  
\* p value for GEE includes both OD and OS outcomes in a multivariate model  
A central defect was defined as having 75% of the overall visual field squares (c) located within the central 16 squares. Peripheral defects were defined as 75% of squares (c) lying outside the central 16 squares. A random defect was any defect not meeting peripheral or central criteria. There was no significant difference found between the two groups in location of defects although there was a tendency for more random defects for the TBI group.

## CONCLUSION

TBI subjects in this study exhibited no statistically significant difference in contrast sensitivity compared to their non-TBI counterparts. However, visual field outcomes did show a significant reduction in MD and increase in PSD in the TBI population. Although there were no categorical defects, visual field testing should be taken into consideration as an important addition to the examination protocol for the mild or moderate TBI patient.

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# Treating High Myopia in a Child

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## INTRODUCTION

A 2 year 10 month old African American female presented with a chief complaint of a close working distance and possible eye turn. The patient was a twin, 37 weeks gestation, 5 lbs 8 oz, no oxygen required and all developmental milestones were reached. Health history was positive for eczema.

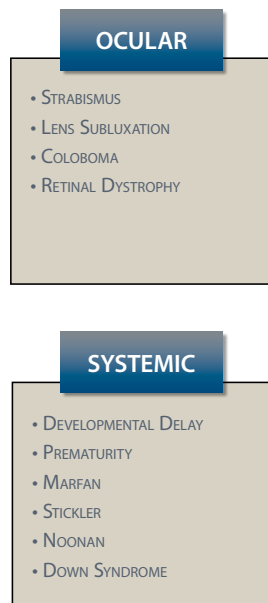
Fig. 1: Visual Acuity/Comments

Date	Visual Acuity	Comments
Visit One	OD: F&F OS: F&F	OD: -15.00 DS OS: -7.50DS
2 mo f/u	OD: 20/60 OS: 20/30 Lea Matching	Poor compliance with SpecRx Suppression OD in W4D Continue FTW of Rx
4 mo f/u	cc OD: 20/100 cc OS: 20/60 w/ TF SRx	Lost glasses 2 weeks prior; very poor compliance; average wear 10 min at a time. Discussed option of SCL fit.
5 mo f/u	CL fit OD: 20/125 OS: 20/100 *Poor Cooperation	CLs Biofinity OD: -12.00 OS: -6.00 Extended wear x 6 days, remove Sat., reinsert Sun.
1 wk CL f/u	Stable VA	CLs fell out without knowledge, were reinserted at this visit. *Mom feels that the patient's visual behavior improved at home with CLs
2 wk CL f/u	With CLs Dva Nva OD: 20/60; 20/40" OS: 20/40+; 20/25	Biofinity CL fit was too loose Switched to Acuvue Oasys extended wear CL trials were under-corrected Stereo: +Lang 3/3, +Fly
1 mo CL f/u	Stable VA	Polycarbonate over CL SRx given OD: -0.75 -0.75 x 180 OS: -0.50 -0.75 x180 Next follow-up expect further improvement in VA, will determine need for occlusion treatment

## METHODS

A cycloplegic retinoscopy revealed high anisometropic myopia. Amblyopia was present OD and suppression is evident with spectacle Rx. No strabismus was noted at the time of examination and follow-up. Slit lamp and fundus examination was unremarkable. Furthermore, no known systemic associations were found. The diagnosis of anisometropic amblyopia was made.

Fig. 2: Ocular &amp; Systemic Diagnosis Associated with High Myopia



## RESULTS/TREATMENT

At the initial exam the patient was prescribed SRx for full time wear. However, patient compliance with the SRx was poor secondary to age and likely visual discomfort due to anisometropia. The patient was then fit with extended wear soft contact lenses (Acuvue Oasys) to promote compliance and decrease anisocoria. Her mother was educated/trained on contact lens insertion and removal. At multiple follow-ups it was noted that patient rubs out her contact lenses likely secondary to the history of eczema and dry eye. Artificial tears were prescribed in order to promote comfort of the lenses. In addition, polycarbonate glasses were prescribed to wear over the contact lenses for protection and avoidance of eye rubbing.

Fig. 3: Why Contact Lenses

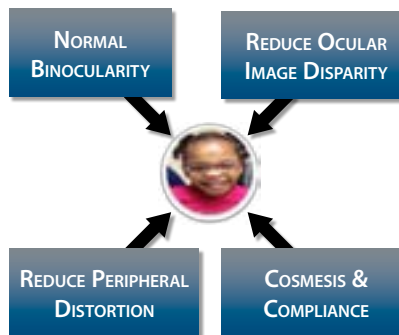


Fig. 4: Tips To Improve Pediatric Contact Lens Fitting

1. Talk directly to children	6. Give children some control
2. Encourage lens handling	7. Rinse lenses with PF saline
3. Exude confidence	8. Have children practice touching their eyes
4. Have children wash their hands with you	9. Reward success
5. Teach breathing exercises	10. Teach lens removal first

## CONCLUSION

This is a unique case of high myopia with an anisometropic amblyopia component in a young child. In such cases it is important to rule out pathologic etiology and to make an appropriate referral. A study done in London, UK found that only 8% of 112 children with high myopia had simple myopia with no associated ocular or systemic conditions. The remaining children in the study either had systemic (54%) or ocular problems along with high myopia (38%). At this time, the patient did not present with any ocular or systemic findings related to high myopia. However, it is crucial to follow her with annual dilated exams

It was essential that the patient was compliant with SRx wear due to the anisometropic amblyopia OD> OS. Although it is a challenge to fit a pediatric patient with contact lenses, there is substantial benefit in a patient with high refractive error or anisometropia; including compliance, cosmesis, normal binocularity, reduction in ocular image disparity, peripheral distortion and amblyopia. The patient's compliance improved significantly with contact lenses versus spectacles. Visual acuity will be monitored and if no improvement is seen in 2-3 months, occlusion treatment of the left eye will be initiated.

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# Successive Anterior Ischemic Optic Neuropathy in a Healthy, Young Male

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## BACKGROUND

Non-arteritic anterior ischemic optic neuropathy (NAION) is an ischemic vascular disorder of the optic nerve. It is the most common cause of acute optic neuropathy in patients over 50 years of age, and affects approximately 2-10 individuals per 100,000 in the United States annually. Long-term follow-up studies have shown an incidence of new NAION in the fellow eye of 14.7% over a median follow-up period of 5.1 years. Like the arteritic form, NAION was previously thought to be a condition exclusive to middle-aged and elderly patients, but recent research shows that as many as 15% of cases occur in patients under the age of 45. The underlying mechanism is thought to result from ischemic infarction of the posterior ciliary arteries supplying the optic nerve. Treatment trials have been aimed at reducing axonal edema to prevent progressive cell loss.

## CASE REPORT

A 38 year old Caucasian male (MF) presented to the Illinois Eye Institute (IEI) complaining of sudden onset vision loss in the left eye (OS) beginning one week earlier and worsening upon awaking that morning. The patient denied pain, headache, scalp tenderness, and jaw claudication. He reported a similar occurrence in the right eye (OD) one-month prior and sought care with a neuro-ophthalmologist at that time. Two days before presenting to the IEI, MF was started on a 16-day course of oral prednisone by a different neuro-ophthalmologist. MF reported that the following tests were all performed twice and read as normal: complete blood count with differential, carotid duplex, erythrocyte sedimentation rate, C-reactive protein, echocardiogram and magnetic resonance imaging of the brain and orbits. He was seeking a second opinion.

Medical history was positive for depression and hypothyroidism, which were being managed by his primary care practitioner with bupropion HCl and levothyroxine, respectively. The patient denied any history of smoking or use of recreational drugs including phosphodiesterase type-5 inhibitors.

Initial and follow-up exam data with serial photographs, Humphrey perimetry (HVF), and Cirrus optical coherence tomography (OCT) are depicted to the right.

Our findings confirmed the diagnosis of sequential NAION OD, followed by OS. MF was advised to complete the oral prednisone course and follow-up for observation in 3 weeks, then monthly for 2 months. We recommended a sleep-study to investigate for nocturnal blood pressure fluctuation.

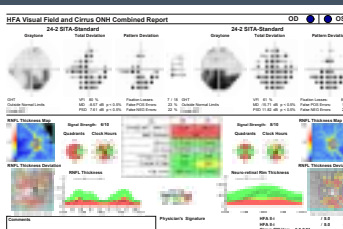
**FIGURE 1:** Humphrey visual field (HVF) and Cirrus OCT combined report at presentation. OD: Inferior altitudinal depression with scattered depressions superior nasally corresponds to retinal nerve fiber layer (RNFL) thinning present on OCT and diffuse disk pallor. OS: Inferior altitudinal depression with scattered depression superior temporal on HVF. OCT shows RNFL elevation consistent with disk edema.



**FIGURE 2:** Combined report at 3 week follow up. OD: Relatively stable HVF and OCT. OS: General depression of visual field, more dense below the horizontal. OCT shows the RNFL returning to normal thickness with thinning superior-temporally and only mild thickening inferiorly.



**FIGURE 3:** Combined report at 3 month follow up. OD: Relatively stable HVF and OCT. OS: Inferior altitudinal depression with continued RNFL thinning on OCT.

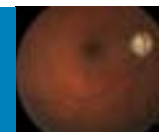


## DISCUSSION

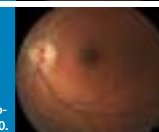
The pathophysiology of NAION is complex and highly controversial. The underlying mechanism is thought to be ischemic infarction of the optic nerve head, likely owing to hypoperfusion of the blood supply from the posterior ciliary arteries (PCAs). The hypoperfusion in these cases is believed to be transient in nature, due to delayed filling of the PCAs seen on fluorescein angiography without evidence of permanent occlusion. Ganglion axonal edema occurs in response to the ischemia, causing a compartment syndrome leading to cell death by apoptosis, especially in a structurally crowded optic disc.

There has been much research exploring potential systemic cardiovascular risk factors for NAION, including but not limited to: diabetes mellitus, hypertension, hyperlipidemia, chronic obstructive pulmonary disease, sleep apnea syndrome and smoking. Another possible associated factor in development of NAION is the use of phosphodiesterase type 5 (PDE5) inhibitor drugs such as Viagra and Cialis. These medications potentiate nitric oxide and are thought to create a hypotensive state at the site of the optic nerve head in associated NAION.

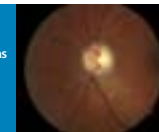
**IMAGE 1:** OD at presentation. Vision measured 20/20, the optic nerve head was flat with diffuse disk pallor and, while difficult to judge in this photo, the cup-to-disk ratio was estimated to be 0.20/0.20.



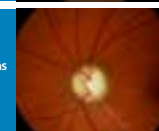
**IMAGE 2:** OS at presentation. Vision measured 20/30 and a 2+ relative afferent pupillary defect was present. The optic nerve head was edematous with indistinct margins and splinter hemorrhages at 4 and 6 o'clock. Cup-to-disk ratio was estimated to be 0.20/0.20.



**IMAGE 3:** OS at 3 week follow up. Vision measured 20/40. Optic nerve was flattening with mild edema remaining inferiorly. Splinter hemorrhages had resolved.



**IMAGE 4:** OS at 3 month follow up. Vision measured 20/40. Optic nerve was flat with distinct margins and diffuse pallor.



Treatment of NAION in the acute phase is aimed at reducing axonal edema to prevent progressive cell loss. The Ischemic Optic Neuropathy Decompression Trial Research Group failed to show a beneficial effect with optic nerve decompression surgery. Neuroprotection with brimonidine tartrate 0.2% has not been shown to have any beneficial effects in visual acuity or visual field outcome in NAION. The role of systemic corticosteroid therapy has been extensively debated. A prospective study by Hayreh and Zimmerman compared patients who voluntarily opted for oral prednisone therapy to those who opted for no treatment. The results demonstrated a benefit in visual acuity outcome in eyes with initial visual acuity of 20/70 or worse, as well as visual field outcome in eyes with an initial visual field defect described as moderate to severe with Goldmann perimetry. These results have been contradicted by other authors.

## CONCLUSION

NAION is a disorder of complex pathogenesis with continued debate over contributory variables and treatment recommendations. Once believed to be a disease of older adults, this case highlights its presence in a younger population with fellow eye involvement over a short period of time. Patient education on the natural history of the disease, including clinical course and risk of progression and fellow-eye involvement are critical to manage patient expectations.

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The authors have no disclosures.

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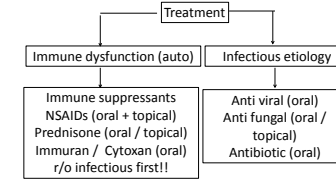
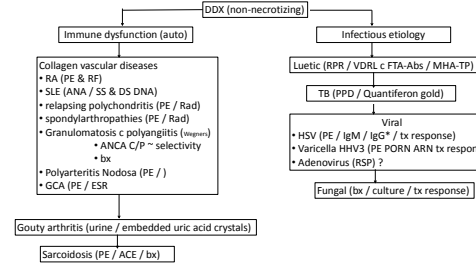
3241 South Michigan Avenue, Chicago, Illinois 60616

# Scleral Uveitis: Differential Diagnosis and Approach to Treatment Options

Thomas R Stelmack, O.D., FAAO

## ABSTRACT

The differential diagnosis of Scleral Uveitis can be difficult but very important to guide treatment options. It is very important to delineate infectious from autoimmune etiology. A series of cases will demonstrate the clinical approach to make this differentiation.



## CASE #1

**50 y/o** CAM c/o OS painful photophobia & globe sensitive to touch

20/20 OD OS with small pupils + LRAPD & light – near dissociation; Motility / VF full

Sclera showed diffuse injection

OS AC rxn 2+ cell c 2+ flare

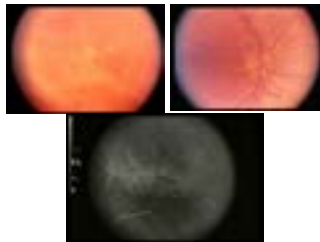
+ serum VDRL & FTA – Abs

Fundus findings see below:

Hx intercourse c prostitute x 1 mo

IV Pen x 10 days 1x106 units

Case #1



## CASE #2

**60 y/o** NAM c/o OS painful photophobia & globe sensitive to touch; recurrent following successful tx with oral NSAID (Indomethacin 50 mg x3 x 1 mo)

20/30 OD 20/20 OS

Motility / VF full / pupils normal s APD

OD Sclera diffuse injection c AC 2+ cell / flare

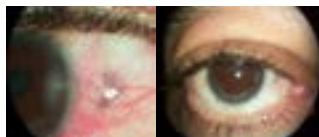
OD inferior infiltrate dx staph marginal keratitis

Poor response to topical pred / e-mycin ung / moxi gtt

Progressed to Wesley immune ring (see below)

Dx changed to HSV; responded well to acyclovir 800x5x4 mos then maintenance dose.

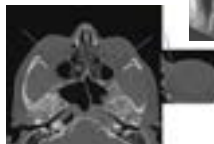
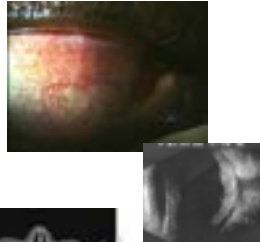
Case #2



Case #3



Case #4



## CASE #4

**41 y/o** AAM c/o bilateral NON – painful muco-purulent red eye x 1 wk. Prior hx glaucoma suspect (CD / CCT).

20/20 OD OS initially treated for allergic then bacterial blepharoconjunctivitis s resolution.

Motility / VF full / pupils normal s APD

Scleral injection noted but w/o pain. Started on 25 mg x3 indomethacin & Zaditor. Labs ordered: CBC c diff, RF, ANA, ESR, ACE, Lysozyme, HLA, ANCA, Quantiferon Gold

Seen in ER noted lid edema & started Augmentin following CT (? Peri-orbital cellulitis).

After 1 month no resolution and one occasion pain 4/10.

NoeOD Scleral diffuse segmental injection

AC: OU 2 flare c oc cell.

Labs negative but on MRI bilateral scleral thickening; T-sign on b-scan. MRI noted fat stranding. On just indomethacin / topical pred acetate. Chest CT negative for hilar adenopathy.

Presented to uvea rounds asking for opinion starting oral pred and diagnosis. Dx sarcoid from fat stranding. Votes on oral steroids both for and against. Decision was made not to start oral steroids as patient was not symptomatic

## CASE #5

**65 y/o** AAM c/o OD painful photophobia & globe sensitive to touch; h/o ulnar drift and morning joint pain.

20/20 OD OS with motility / VF full / pupils wnl – RAPD

Scleral diffuse injection OD 2-3+ cell mild flare. Labs ordered: CBC c diff, RF, ANA, ESR, ACE, Lysozyme, HLA, ANCA, & claimed – PPD in recent past. RF = 500. Dx RA already being seen by Rheumatology. They weaned of oral Pred (steroid responder) and started Imuran because of scleral uveitis.

## BECAME CASE #3

Case #5 responded poorly to immunosuppressant tx (Imuran max daily dose). Fundus nodule noted below and Quantiferon Gold ordered and was positive.

ID consulted but since chest CT negative and patient denied night sweats, weight loss and lethargy, was started only on INH 300 mg daily. Simultaneously was started on anti-tnf Humira sub c q 2 wks. Sclerouveitis resolved.

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# ICO

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# The Benefits of Expanding Optometric Services Offered at a School Based Vision Clinic.

Melissa A. Suckow, O.D., FAAO; Sandra S. Block O.D., M.Ed, FAAO, FCVOD  
Illinois College of Optometry, Chicago, Illinois

## INTRODUCTION

The Illinois Eye Institute at Princeton School (IEI at CPS) opened in 2011 as a collaboration between the Illinois Eye Institute (IEI) and the Chicago Public Schools. The goal of this program was to decrease the number of children in Chicago that were failing to receive vision services. The clinic operates year round, and has served more than 15,000 Chicago children as of October 1, 2013. During its first year, we found that we were still referring a large number of students to the Illinois Eye Institute, main campus for follow up care. Because compliance rates with these follow up appointments were very low, we looked for ways to incorporate more of the testing into our clinic.



Vision therapy



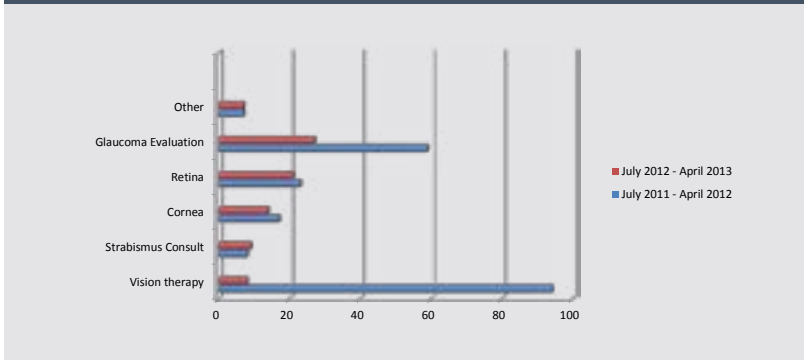
Fundus

## RESULTS

Between July 1, 2011 and April 30, 2012, IEI at CPS referred 208 of 4571 patients for follow up care. Reasons included vision therapy (94), strabismus consult (8), cornea (17), retina (23), glaucoma evaluation (59), and other (7). Because of the large unmet need, we felt it would be beneficial to extend our hours and add specific times for vision therapy. In July 2012, we expanded our clinic hours to include afternoon vision therapy appointments. We added a visigraph and new vision therapy equipment to help with training. In February 2013, we were able to obtain a retinal camera and an OCT.

Between July 1, 2012 and April 30, 2013, IEI at CPS referred only 86 of our 5016 patients. Reasons for referral included vision therapy (8), strabismus surgery consult (9), cornea (14), retina (21), glaucoma evaluation (27), and other (7). Other referrals included 2 for diagnostic testing and 5 for headache evaluations. All but two of the glaucoma evaluations occurred before we received our camera and OCT. Vision therapy referrals were for patients who preferred weekend or evening appointments.

Figure 1: Referrals to Illinois Eye Institute Main Campus



OCT



Visagraph

## CONCLUSION

Expanding the services available at IEI at CPS has allowed us to greatly reduce our outside referral rate. Increasing our afternoon appointment slots and including contact lens care may be ways to further reduce our number of referrals. However, many patients that we ask to return to IEI at CPS for strabismus and amblyopia follow ups are still not receiving the care they need. During the next year, we plan to focus our efforts in this area.

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# Exposure Keratopathy Secondary to Prostaglandin Associated Periorbitopathy

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## INTRODUCTION

Prostaglandin analogues (PGAs) are used in the treatment and management of glaucoma by increasing aqueous outflow via the uveoscleral pathway. Common known side effects of these drugs include increased pigmentation of irides and lashes, hypertrichiasis, blurred vision and conjunctival hyperemia. Periorbitopathy is a more recently documented side effect that has been noted with an absence of dermatochalasis, levator dehiscence, ptosis of upper lid, decreased prominence of orbital fat pads and relative enophthalmos. This case report presents three cases of prostaglandin associated periorbitopathy (PAP) that resulted in exposure keratopathy.

## CASE REPORTS

**PATIENT 1:** 90 yo CM with POAG treated with bimatoprost 0.03% bilaterally at bedtime for the past eight years. Bilateral PAP observed with more significant changes to the left ocular adnexa (Photo 1). Slit lamp examination revealed a 6mm x 2mm sterile corneal ulcer in the 5-7'clock positions in the left eye (Photo 2).

**PATIENT 2:** 90 yo CM with POAG was initially treated bilaterally at bedtime with bimatoprost for three years and was switched to travoprost for the past 7 years due to insurance coverage. Bilateral PAP was observed (Photo 3) by external evaluation. Slit lamp exam revealed neovascularization of the cornea and an overlying 5mm x 2mm sterile corneal ulcer in the 4-6 o'clock position in the left eye (Photo 4).

**PATIENT 3:** 80yo CM with primary open angle glaucoma (POAG) taking bimatoprost .01% bilaterally at bedtime for the past 3 years. Upon external examination, bilateral PAP was noted (Photo 5). Slit lamp examination revealed pannus and punctate surface irregularities in the 4-6 o'clock position in the right eye (Photo 6).

All three patients were treated with bacitracin ointment at bedtime to the affected eye and were instructed to use copious amounts of artificial tears. The PGAs were discontinued. Improvement of the ocular surface was noted within 2-3 weeks in each of the cases and alternative

therapy was initiated to manage the intraocular pressure. After changing topical glaucoma therapy, all three patients demonstrated resolution of PAP within the year and no recurrence of exposure keratopathy was noted.

Photo 1



Photo 2



Photo 3



Photo 4



Photo 5



Photo 6



## DISCUSSION

PAP was initially reported in 2004 with bimatoprost and subsequent cases were then documented with travoprost, latanoprost, tafluprost, and unoprostone. It is thought to occur most frequently with bimatoprost. The exact mechanisms of these side effects have not yet been established. These effects have been documented to appear after a period of several weeks and have been reported to be reversible after discontinuing use of PGAs.

## CONCLUSIONS

It is important to recognize PAP as a side effect of prostaglandin medications in the treatment and management of glaucoma. In addition, this side effect should be considered with unilateral administration of these medications. Patients with these side effects may note increased ocular surface irritation and have an increased risk for exposure keratopathy. As this case demonstrated, it has been reported that PAP resolves after discontinuation of the offending medication. Consideration should be given for topical medication adjustments should this side effect become problematic.

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# Atypically Located Unilateral Optic Disc Pit with Serous Detachment and Evidence for Cerebrospinal Fluid Source Theory

Eric A. Woo, OD<sup>1</sup>, Stephanie Klemencic, OD, FAO<sup>2</sup>

1. MCPHS University School of Optometry, 2. Illinois College of Optometry

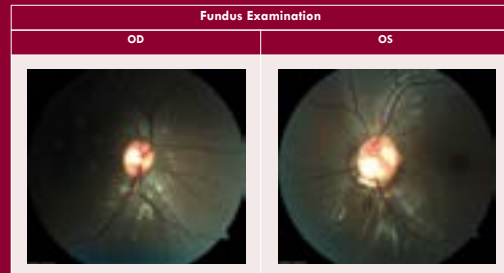
## BACKGROUND

Optic disc pits are congenital anomalies predominantly affecting the temporal optic disc. The pathophysiology behind optic disc pits is controversial and has been a topic of debate. There is also controversy in treatment due to debate over the source of the fluid under the retina. This case presents a child with a rare inferonasal optic disc pit and adjacent serous detachment. Spectral domain OCT images may give insight to the source of the fluid.

## CASE REPORT

- 12-year-old African American male
- Routine eye examination without complaints. First eye exam ever.
- No systemic conditions and was taking no medications. NKDA/NKMA.
- No prior ocular history. Social history unremarkable.
- Best corrected vision: 20/20 OD, OS.
- EOMs: FROM OU, CVF: FTFC OU, Stereo & Color: unremarkable. PERLL(-)JAPD.

Slit-Lamp Examination		
	OD	OS
Adnexa/Lids:	Normal	Normal
Conjunctiva/Sclera:	W&Q	W&Q
Cornea:	Normal endothelium, epithelium, stroma and tear film	Normal endothelium, epithelium, stroma and tear film
Angles:	4+T/4+N	4+N/4+T
Anterior Chamber:	D&Q	D&Q
Iris:	Normal, round pupil	Normal, round pupil
Lens:	Clear lens capsule, cortex and nucleus	Clear lens capsule, cortex & nucleus
IOP (Goldmann):	15mmHg	16mmHg



Optic disc photo OD (Left): Unremarkable. Optic disc photo OS (Right): Large optic nerve pit inferonasal without margin of disc involvement with oval shaped serous detachment inferonasal to optic nerve. Small scattered retinal pigment epithelium (RPE) changes within area of detachment. Non-macula involved. No apparent retinal/choroidal coloboma. Of note, the two cilioretinal arteries superotemporal and temporal.

Three month follow-up examination showed stability in area of detachment. The patient was scheduled to return for DFE and to repeat Humphrey 24-2 SITA Standard visual field OS.

## DISCUSSION

The appearance of optic disc pit excavations can vary wildly, not only in color (30% white/yellow, 60% grey, and 10% black) but in location (predominantly temporal/inferotemporal, 20% central, and 10% elsewhere)<sup>1,2</sup>, and size (can vary from 0.1-0.7DD and 0.3-2.5D in depth with an average of 5D in depth).<sup>1,2</sup> Goktas noted that optic discs with pits present were statistically significantly larger than those without ( $p = 0.038$ ).<sup>3</sup> The greater the disc size, the greater unilateral occurrence of optic disc pits, however, only 10-15% of optic disc pit cases were found to be bilateral with most cases having predominantly a single pit per nerve.<sup>1,4</sup> The presence of cilioretinal arteries have been noted in 64% (16 of 25) of optic disc pit cases studied, of which 86% had two.<sup>5,6,7</sup> (Continued Far right)

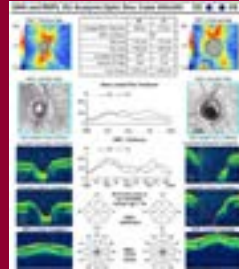


Fig. 1

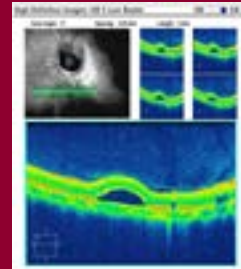


Fig. 2

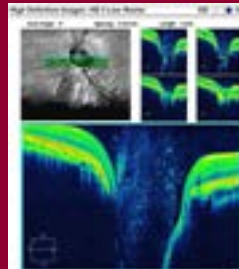


Fig. 3



Fig. 4

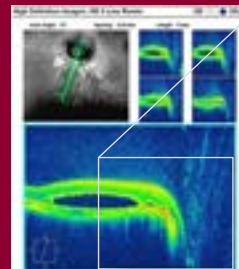


Fig. 5

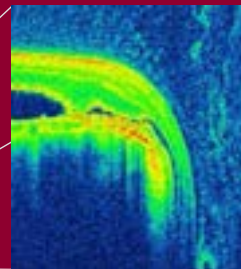


Fig. 6

## DISCLOSURE

No financial disclosures/conflicts of interest.

## DISCUSSION continued

This finding is stated to reinforce the relationship between optic pits and other congenital anomalies including optic disc colobomas of which they note cilioretinal arteries emerge from the inferior margin of the optic disc. This study showed that cilioretinal arteries of their optic pit cases emerged from the bottom or margin of the pit themselves.<sup>7</sup> Condensed vitreal tissues or glial tissue has been noted to be seen in 78-89% of optic disc pits extending from within the excavation.<sup>1,4</sup> The possibility of serous detachments caused by vitreal traction from the condensed vitreous has not been determined. However, the resolution of retinal detachments in cases where the condensed vitreous/glia tissue was removed has occurred and should be of interest for future studies.<sup>1</sup>

Optic disc pit maculopathy is the greatest risk with 25-75% of cases developing serous macular elevations such as serous detachments/schisis, cystic degeneration, and degenerative macular detachments.<sup>1</sup> possibly in a classic teardrop pattern adjacent to the location of the optic disc pit.<sup>8</sup> The treatment options for optic disc pit maculopathy is presently considered necessary but controversial, partly because of the rarity of the condition but also due to the unknown nature of the source of the fluid involved.<sup>1,4,8</sup> Many treatment modalities have been attempted with differing visual outcomes but most with successful anatomical attachment of detached retina. Such techniques are pars plana vitrectomy (PPV) with or without internal limiting membrane peel (ILM) peel and with or without photocoagulation and/or gas tamponade. Laser photocoagulation had been used previously in some cases along the temporal margin of the disc closest to the optic pit to potentially seal off a theoretical connection between the pit and subarachnoid space and minimize fluid movement between the pit and the retina.<sup>4,9</sup> The success of this treatment ranged from completely unsuccessful to five of six cases with reattachment with underlying debate of how long the reabsorption of fluid will truly take if successfully sealing off such a connection.<sup>4</sup> In the study by Gregory-Roberts et al, they note that the removal of hyaloid traction may have the best outcome as found in a study by Hiraoka et al and that multiple other small studies showed better visual outcomes (average of 0.8logMAR)<sup>10</sup> when vitrectomy and/or removal of vitreal/glia tissue in the optic pit were involved.<sup>4</sup>

## FLUID SOURCE THEORIES

- **Leaky blood vessels at base of pit or under subretinal space:**  
Mostly disproven with several fluorescein angiograms negative for leakage in optic disc pit or subretinal space.<sup>4</sup>
- **Vitreous fluid source (physical link between vitreous and fluid cavity hypothesis):**  
No large scale evidence in humans. Recent study utilizing scanning electron microscopy displayed holes in the membrane overlying the edge of the optic pit to create an area of schisis in combination with histological preparations showing a lamellar defect allowing communication into the subretinal space.<sup>11</sup>
- **Cerebrospinal fluid (CSF) from the subarachnoid space:**  
Krivoy et al. found, using the Carl Zeiss Meditec OCT 1, a space of direct communication from schisis/subretinal cavity leading towards the subarachnoid space to which they attributed as consistent with the CSF theory.<sup>12</sup>

## CONCLUSION

This case presents an optic disc pit with serous detachment in an atypical location that one would more often attribute with optic disc coloboma; thus, contributing to the debate between the link between the two conditions. While optic disc pits may be rare, the biggest concern is an adjacent serous detachment of the macula occurring in 25-75% of cases, especially when pits are located temporally. Serous detachments are more likely to occur in the patient's 30s and 40s but can occur at any age. Extensive education on possible retinal detachments is crucial for these patients. Though the best type of treatment is debatable, treatment that involves vitrectomy and/or removal of the vitreal/glia tissue within the optic pit is most favorable.

With the utilization of Cirrus OCT imaging to display a potential connection of subretinal fluid to the subarachnoid space, this case documents evidence that may support the cerebrospinal fluid source theory. Utilization of high definition OCTs in cases of optic disc pits with adjacent serous detachments may be useful for potential treatments in cases where the macula is threatened.

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# DECREASED VISUAL ACUITY AND FOVEAL RETINAL THINNING IN A 66 YEAR OLD MALE VEGETARIAN AND SERIAL BLOOD DONOR

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## Abstract

Unexplained decrease in best corrected visual acuity (BCVA) can be frustrating. Sometimes common etiologies with atypical signs are the culprit, while in other cases it is a rare condition that could be treated or stabilized, if properly identified. This unique case began as a diagnosis of non exudative Age Related Macular Degeneration (AMD), but with second opinions, a retina referral, blood tests, and somewhat novel functional and imaging studies a diagnosis and management plan were reached.

## Details

A 66 year old white male previously diagnosed with AMD presented for a two month follow up appointment. Since his last appointment, he had lost one line of BCVA OD and three lines OS. He had a history of fluctuating blood pressure, quiet macular scars OD>OS presumably from a diving accident 30 years ago, and one incident of transient monocular blindness OD for 45 seconds.

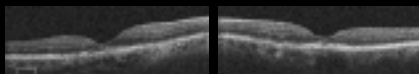
**Medications:** Daily Vitamin B Complex Capsule  
25 mg Hydrochlorothiazide

**Unique Lifestyle History:** Strict vegetarian, regular blood donor (150 lifetime pints)

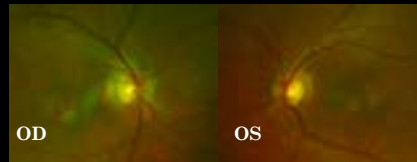
### Clinical findings:

- Quiet macular scarring OD>OS with RPE mottling OU
- Accelerated retinal arteriolar sclerotic changes OU (~1/3 A/V ratio) with wide  $\alpha$  and  $\beta$  zone peripapillary atrophy (PPA) OU, BCVA 20/40 OD, OS (down from 20/30 OD, 20/20 OS)

## Imaging



Zeiss Cirrus SD OCT images taken 08/12/2013 show disruption in the photoreceptor integrity line (PIL) OD>OS. The foveal pits are widened and central thickness is decreased. Central thickness: OD: (210um), OS (200um)



Wide field Optos Imaging OD and OS taken 08/12/2013 show some RPE irregularity,  $\alpha$  and  $\beta$  zone PPA OU, but no drusen characteristic of AMD. The dark circular areas OD>OS are likely a result of his aforementioned diving accident.

QuantifEYES MPS II showed decreased pigment optical density OS>OD, which was confirmed by ARIS 3D profile

Macular Pigment Density OD: 1142 AUC (LOW)

Macular Pigment Density OS: 770 AUC (LOWER)

\*Normal Macular pigment density is ~3000 AUC

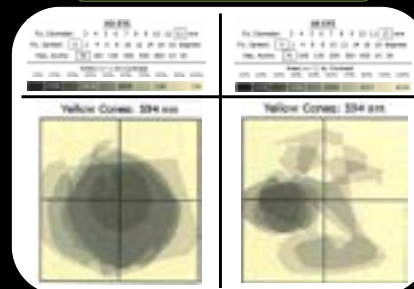
## Lab Testing

TEST	RESULTS	NORMAL RANGE
• FERRITIN	7 (LOW)	26 – 388 ng/ml
• FIBRINOGEN	187.3 (LOW)	200 – 400 mg/dl
• IRON	49 (LOW)	65 – 175 ug/dl
• B12 VITAMINS	286 (LOWER)	211 – 911 pg/mL
• HOMOCYSTEINE	15.2 (HIGH)	3.2 – 10.7 umol/L
• MAGNESIUM	2.1 (NORMAL)	1.8 – 2.4 mg/dL
• FOLATE	13.38 (NORMAL)	Ref.: >5.39 ng/mL
• MMA	154 (NORMAL)	87 – 318 nmol/L
• RDW	13.1 (NORMAL)	11.0 – 14.0 %
• RBC	4.46 (NORMAL)	4.20 – 5.7 M/uL
• Bleeding time	4.5 (NORMAL)	3.0 – 9.0 minutes
• Von Willebrand Factor	143 (NORMAL)	50 – 160
• Riscocetin Cofactor	93 (NORMAL)	46 – 154
• Factor VIII	102 (NORMAL)	61 – 184
• Factor IX	83 (NORMAL)	63 – 146

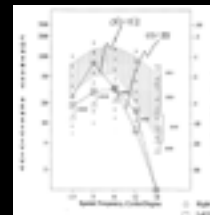
**Explanation of Lab Testing:** Vegetarian diet and frequent blood donation initially indicated some type of anemia. Serum analysis was performed including iron, homocysteine, and B12 levels. With a borderline low B12 level, methylmalonic acid (MMA) level was checked. Elevated MMA with elevated homocysteine indicates B12 deficiency anemia. Normal MMA with Elevated homocysteine can indicate folate deficiency. Both MMA and folate levels were normal. Normal red blood cell distribution width (RDW) and red blood cell (RBC) count further rule out anemia. The patient is iron deficient and will likely benefit from less frequent blood donation.

\*He was also tested for clotting disorders, given a carotid Doppler, and an echocardiogram, but found no explanation for his single episode of transient monocular blindness. MRI revealed mild chronic small vessel ischemic disease.

## Functional Testing

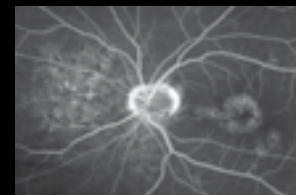


Kinetic Fields Test (above) revealed a central scotoma OD, and a paracentral scotoma OS, which was not seen on the HVF. The Kinetic Fields Test measures a 10 degree field.



Stereo Optical Contrast Sensitivity: OD is poor at all points save 1, (at 3 cycles per degree) (153 AUC). OS is poor at all points (88 AUC). Normative Range is 200-300 AUC.

## Retina Consult



Fluorescein angiography (FA) during a retina consult on 10/10/13 revealed RPE atrophy, and window defects. Slit lamp examination showed a subtle cream colored sheen to the retina at the macula OU.

## Diagnosis

Iron deficiency without anemia and Macular pattern dystrophy resulting in disruption of PIL, decreased BCVA, decreased contrast sensitivity, and central/paracentral scotomas OS and OD.

## Treatment/Management

- Start Martek vegetarian EPA/DHA supplement
- Start two aspirin 81 mg per day
- Recommended reduced frequency of blood donation
- Patient will self monitor Amsler Grid
- Monitor. Follow up in 8 months

## Conclusion/Discussion

Pattern dystrophy encompasses several autosomal dominant retinal dystrophies causing variations in macular RPE and abnormal lipofuscin accumulation. It is often misdiagnosed as AMD. Pattern dystrophy is rare, with mild symptoms presenting later in life. It can present with a variety of patterns. This case had creamy-white pigment disturbances; however black, grey, brown, orange, or yellow patterns are also possible. Patients are often monitored for signs and symptoms of choroidal neovascular membrane, and in rare cases can develop geographic atrophy. The FA performed revealed a stippling/mottled pattern of hyperfluorescence at the macula with window defects characteristic of pattern dystrophy. On careful examination of the fundus, a classic creamy-white sheen was observed. The white/yellow flecks previously thought to be drusen were actually areas of RPE atrophy also characteristic of pattern dystrophy.

## Acknowledgements

This case report poster is based on original clinical work supported by the Optometry/Ophthalmology sections of Captain James Lovell Federal Health Care Facility, DVA-Naval Medical Center, North Chicago, IL, USA. There are no conflicts of interest.

## Sources

Garrison, Cheryl (Editor), *The Iron Disorders Institute Guide to Anemia: Second Edition*. Cumberland House Publishing. 2009

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# Central Retinal Artery Occlusion and Resistant Hypertension

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## Background

- Central retinal artery occlusion (CRAO) is an acute event that results in profound and often permanent vision loss.
- Because these patients are at higher risk for stroke and ischemic heart disease, it is imperative for eye and health care professionals to determine the source of the occlusion.
- Controlling risk factors such as hypertension (HTN) and diabetes mellitus (DM) will improve health outcomes and, in patients with unilateral disease, prevent vision loss in the fellow eye.
- However, when HTN is not sufficiently controlled, a concern for resistant hypertension arises.

## Case

### Case History

A 56-year-old Japanese male veteran presented to the emergency room for evaluation of sudden vision loss in the right eye occurring one day prior. He reported his vision returned for one hour, then was lost again. He was admitted for hypertensive crisis with blood pressure (BP) of 238/126 and was also diagnosed with Type 2 DM. His personal medical history was unknown due to the patient's lack of medical care for the past 35 years. His body mass index was 27.0.

### Exam Findings

OD	Relevant findings	OS
Hand motion	<i>Best corrected visual acuity</i>	20/200
2+ RAPD	<i>Pupils</i>	Normal
2+ PSC	<i>Lens</i>	2+ PSC
Distinct margins, no pallor	<i>Optic nerve</i>	Distinct margins, no pallor
Superficial retinal whitening, cherry red spot, multiple cotton wool spots, dot/blot and flame hemorrhages	<i>Posterior pole</i>	Multiple cotton wool spots, dot/blot and flame hemorrhages
Diffuse macular thickening	<i>Macula</i>	Central macular thickening
Diffuse retinal thickening	<i>Optical coherence tomography (OCT)</i>	Large foveal cysts
Delayed filling and significant late leakage	<i>Fluorescein angiography</i>	Late hyperfluorescence centrally

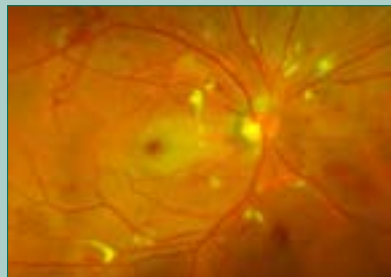


Figure 1.

Optos widefield fundus photography OD and OS pictured in Figures 1 and 2, respectively, were acquired six days after initial presentation. Figure 1 shows a distinct cherry red spot and a surrounding area of whitening, indicating that most of the retina has re-perfused. The quality of Figure 2 is limited by a posterior subcapsular cataract.

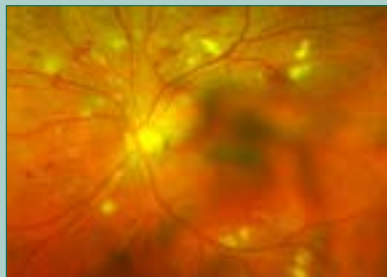


Figure 2.

### Imaging/Labs tests

Carotid duplex study revealed no significant stenosis but did reveal moderate plaques on the right side. Transesophageal echocardiogram (TEE) was unremarkable. Other imaging and lab tests ruled out giant cell arteritis (GCA), hyperviscosity syndrome, pheochromocytoma, renal artery stenosis, and Moyamoya disease.

### Assessment

- Non-arteritic CRAO OD, with the source of the embolus likely originating from the carotid artery
- Grade 3 hypertensive retinopathy OU
- Diabetic macular edema (DME) OS
- Severe non-proliferative diabetic retinopathy OU

All the above conditions are secondary to longstanding undiagnosed essential HTN, DM, and chronic kidney disease (CKD).

### Management/Outcome

Treatment was held in the left eye to allow for spontaneous resolution of macular edema. Visual acuity in both eyes remained stable at all subsequent visits to the clinic. OCT two months later revealed diffuse retinal atrophy in the posterior pole of the right eye and significantly improved macular edema in the left eye.

The medical center's ophthalmology, primary care, cardiology, and nephrology clinics are closely following this patient. Due to the rapid decline in his kidney function in the last two months, his BP control regimen has changed drastically. He is currently taking hydralazine 50 mg po for HTN. Meanwhile, his home monitor BP measurements remain high, measuring as high as 209/109. While the appearance of his retina has improved and a cataract evaluation is pending, aggressive intervention in his BP and blood sugar (BS) control are still needed to give this patient the potential to return to normal vision.

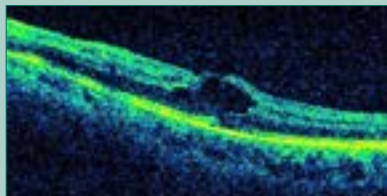


Figure 3.

Spectral-domain Zeiss OCT image of the left eye six days after initial presentation, displaying large cystic changes and a disruption of the IS/OS junction at the fovea.

The absence of an evident embolus in the central retinal artery does not mean the occlusion was not caused by an embolus. It could have migrated and disappeared by the time the eye was examined.

### Resistant HTN

Strict glycemic and blood pressure control remain the most effective ways of reducing DME. However, when HTN is not controlled even with multiple oral anti-hypertensive medications and optimal compliance, a concern for resistant hypertension arises. The exact prevalence is unknown, but data from population studies and clinical trials suggest that it is a relatively common problem. Prevalence is expected to rise given the aging population and trends in obesity and CKD. Diagnosis of this condition is complicated by "pseudo-resistance," i.e. improper BP measurement technique, the white-coat effect, and poor compliance with medications and lifestyle/diet adjustments, such as sodium restriction, weight loss if obese, and reduction of alcohol intake.

Appropriate lab work and imaging should be considered in malignant and resistant HTN to rule out pheochromocytoma, renal artery stenosis, and hyperviscosity syndrome.

Optimal BP control can only be achieved if the treatment regimen is directed toward the cause, i.e. volume overload should be treated with the appropriate dose and type of diuretic.

## Take Home Messages

- In CRAO, determining the etiology is important in improving remaining quality of life and vision.
- The resolution of carotid duplex and TEE may not be sensitive enough to detect small plaques significant enough to cause CRAO.
- Eye care professionals should advocate for strict BP and BS control for patients with macular edema.

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## Acknowledgements

This case report poster is based on original clinical work supported by the Optometry/Ophthalmology sections of Captain James Lovell Federal Health Care Facility, DVA-Naval Medical Center, North Chicago, IL, USA. There are no conflicts of interest.

## Discussion

### CRAO

Fluorescein angiography should be performed in all CRAO patients 50 years and older. It plays a crucial role in the diagnosis of arteritic CRAO and GCA; in almost every patient with GCA, fluorescein angiography discloses occlusion of the posterior ciliary arteries.

Following a diagnosis of CRAO, a carotid duplex and echocardiogram should be ordered to determine the origin of the embolus. Negative results do not mean that these structures were not involved because the resolution of these images may not be sensitive enough to detect very small plaques or lesions that could be significant enough to cause CRAO.

- Carotid duplex:** The internal carotid artery should be evaluated not only for stenosis but also for the presence of plaques. The carotid duplex only evaluates the carotid artery within the neck, possibly missing plaques above or below.
- TEE:** TEE produces superior images to the transthoracic echocardiogram (TTE) in detecting heart abnormalities.



ICO

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## 14 ICO PRESENTATIONS

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3241 South Michigan Avenue, Chicago, Illinois 60616

# Eye-Movement Analysis in Relationship to Birth Order in Children

#182

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## PURPOSE

The purpose of this study was to evaluate the relationship that birth order may have in relationship to the development of eye movement skills. We previously reported on the results of 63 children (Allison C. Schlange D, ARVO 2011). The relationship between IQ and birth order has been looked at in a number of studies in the past, with many finding that first born children scored higher on intelligence tests, as well as in behavioral development. In a study by Clark and Rice, they found that firstborn children were overrepresented in Nobel Prize winners. A recent study also showed that first born children spend 3,000 more quality hours with their parents between the ages of 4 and 13 than the next born child in the family when going through the same age span.

Accurate eye movement skills are an essential part in the process of reading, and therefore learning in a classroom environment. In a recent study by Quid and Simpson comparing 50 Canadian children with reading based Individualized Education Plans (IEP) with 50 children without reading complaints or IEPs, they found the children with reading difficulties showed statistically significantly increased numbers of fixations, excessive eye movements, and decreased reading speed when compared with the control group. An Austrian study by Dusek, Pierschione, and McClelland, also showed a slower reading speed and a larger number of errors in children referred from an educational assessment center with reading difficulties versus those children with no reading difficulties. A study by Tassinari showed that untreated oculomotor dysfunction does not resolve on its own, and a number of studies show that vision therapy can improve oculomotor dysfunction. Oculomotility can be objectively evaluated with the use of computerized tests such as the Visagraph or RealDyler. Our theory is that first children, and children with no siblings, may exhibit better eye movement skills prior to entering Kindergarten due to the types of play activities that they perform, as well as the amount of time spent reading with adults one on one. The types of activities that are performed in oculomotor vision therapy are often similar to the types of activities that children perform during solitary play. Children who perform more activities similar to those that may be prescribed during vision therapy (mazes, puzzles, activity books, computer games) at an early age, may have better oculomotor function upon entering school. Children with multiple siblings may participate in more imaginative play, and spend less time doing certain types of eye movement activities. Thus, birth order and/or number and age of siblings may be a factor in the development of eye movement skills, including accurate saccades and better fixation control.

## METHODS

Ninety-three children were examined the summer prior to entering Kindergarten. The age range of these children was 4 to 6 years old depending on their birthdates, and the date of the exam. The children were given a full comprehensive eye examination including tests of accommodation and vergence, as well as a full ocular health evaluation. See **Table 1** for a list of the tests performed during the examination, and the mean values for those findings. The parents filled out a survey (See **Figure 1**) regarding the number and ages of the siblings, prior school history, and the amount of time the children spent on specific tasks such as reading books with a parent or playing near vision type games. See **Figure 2** for the scale used to interpret the survey results in **Figure 3**.

The subjects also received eye movement recordings using the Visagraph III (Taylor), an infra-red system (60 Hz) with goggles the subject wears while viewing words, numbers and symbol targets. (**Figures 4 & 5**). The system software, combined with manual analysis of the recorded data is used with this younger population to evaluate the results. The following three procedures were completed.

### Fixation Control:

Task I determines how successful our subject is at holding fixation on a target (20/30 equivalent letter or face target, 33 cm viewing dist.) for 15 seconds and inhibiting all eye and head movement. Fixation drifts, attention loss and off-target saccades are recorded and evaluated for frequency and amplitude (**Figure 6**).

### Saccadic Speed:

Task II determines how quickly and accurately the subject can complete horizontal saccades, as fixation is alternated between two 15 deg. separated targets for 20 sec. duration. The number of saccadic excursions is a saccadic speed score. Targets similar to Task 1 are used (**Figure 7**).

### Saccadic Accuracy:

Task III determines saccadic accuracy by recording the number of refixations (corrective saccades) required to regain fixation after completing a 15 degree saccadic excursion (similar to Task II). (**Figures 8 & 9**).

Our sample size included three sub-groups, compared according to Fixation Control, Saccadic Speed and Saccadic Accuracy.

- Only child
- First born with one sibling
- May not be first born, with multiple siblings

Table 1: Comprehensive Exam Tests Performed

TESTS PERFORMED	MEAN VALUES
Case History	Unremarkable findings
Snellen Visual Acuity, distance	OD 20/27, OS 20/21
Snellen Visual Acuity, near	OD 20/21, OS 20/21
Cover Test, distance and near	DCT = 0.00 exp. NCT = 0.00 exp
Refraction	Normal
Near Point of Convergence	Break = To The Nose
Stared	42.5"
Extraocular Muscles	Full
Gross Pursuits with a moving target	3"
Gross Saccades - horizontal	3"
Confrontation Visual Fields	Full
Post-dilated Stereometry - Cyclotest	OD +1.03 D, OS +1.14 D
Cyl	OD -0.64 D, OS -0.30 D
Post-dilated Spherical Equivalent	OD -0.69 D, OS -1.00 D
Accommodation of Accommodation - Pull Away	OD 14.05 OS 14.61
Monocular Estimate Method	OD 0.56 OS 0.51
Near Base Out Vergence (Phoria Bar Method)	Blue = 0, Break = 17.25, Recovery = 11.98
Near Base Out Vergence (Phoria Bar Method)	Blue = 0, Break = 27.75, Recovery = 18.31
Non-contact Tonometry	Unremarkable
Anterior Segment examination with Slit Lamp	Unremarkable
Binocular Indirect (BNI) with TriVision	Unremarkable
Color Vision	One with color deficiency
Visualizing Glasses and/or Vision therapy	2 recommended treatments after exam

Figure 1: Parent Survey

Figure 1: Parent Survey

Child Name: \_\_\_\_\_

Child's Age: \_\_\_\_\_

Parent's Name: \_\_\_\_\_

Parent's Occupation: \_\_\_\_\_

Parent's Education: \_\_\_\_\_

Parent's Marital Status: \_\_\_\_\_

Parent's Religion: \_\_\_\_\_

Parent's Ethnicity: \_\_\_\_\_

Parent's Language: \_\_\_\_\_

Parent's Address: \_\_\_\_\_

Parent's Phone: \_\_\_\_\_

Parent's Email: \_\_\_\_\_

Parent's Occupation: \_\_\_\_\_

Parent's Education: \_\_\_\_\_

Parent's Marital Status: \_\_\_\_\_

Parent's Religion: \_\_\_\_\_

Parent's Ethnicity: \_\_\_\_\_

Parent's Language: \_\_\_\_\_

Parent's Address: \_\_\_\_\_

Parent's Phone: \_\_\_\_\_

Parent's Email: \_\_\_\_\_

Figure 2: Survey Question Scale

Figure 2: Survey Question Scale

Parent Name: \_\_\_\_\_

Child Name: \_\_\_\_\_

Child's Age: \_\_\_\_\_

Parent's Name: \_\_\_\_\_

Parent's Occupation: \_\_\_\_\_

Parent's Education: \_\_\_\_\_

Parent's Marital Status: \_\_\_\_\_

Parent's Religion: \_\_\_\_\_

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Parent's Marital Status: \_\_\_\_\_

Parent's Religion: \_\_\_\_\_

Parent's Ethnicity: \_\_\_\_\_

Parent's Language: \_\_\_\_\_

Parent's Address: \_\_\_\_\_

Parent's Phone: \_\_\_\_\_

Parent's Email: \_\_\_\_\_

Figure 3: Parent Survey Responses (Mean values)

	All Groups Means	All Groups Std. Deviation	Only Child N = 12	First Born 1 Sibling N = 29	May Not 1st Born ≥ 2 Siblings, N = 20
Prior Exam	1.90	.297	1.88	1.87	1.95
Preschool years	1.99	.810	1.93	2.05	1.98
Daytime reading to	5.86	1.673	5.41	5.97	5.89
Mixday reading to	1.84	.715	1.76	1.82	1.79
Puzzles	1.69	.752	1.65	1.82	1.42
Activity books	2.19	.958	2.06	2.46	1.96
Coloring/drawing	2.80	.964	3.52	2.88	2.51
Computer use	1.92	.947	2.21	1.99	1.76
Handheld games	1.42	.863	1.31	1.48	1.40
TV viewing	3.93	.938	3.42	3.11	2.81
Play outdoors	3.69	.876	4.11	3.88	3.82
Imaginative play	3.51	.895	3.64	3.43	2.99
Reading skills	1.78	.415	2.01	1.82	1.85
Sports	1.27	.446	1.38	1.31	1.29
Ballgames	1.13	.887	0	1	1.69
Birth order	1.23	1.023	1	1	1.85

Figure 4 & 5

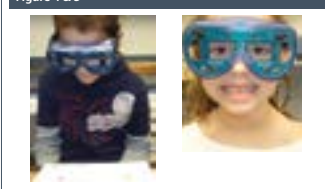


Figure 6: Fixation Control - Inhibit Drift

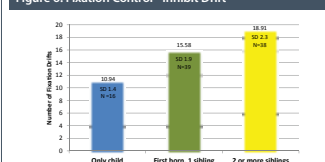


Figure 7: Saccadic Speed

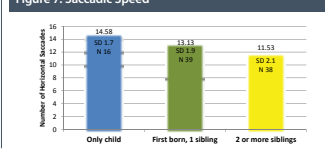


Figure 8: Saccadic Accuracy=Post-Saccade Refixations

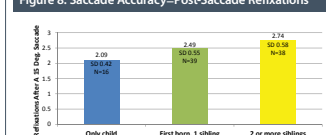


Figure 9: Saccadic Accuracy-Post Saccade Refixations (F 15.07, p=.001)

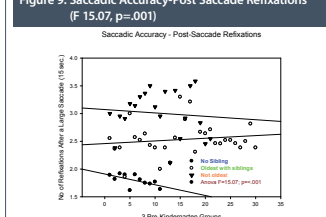
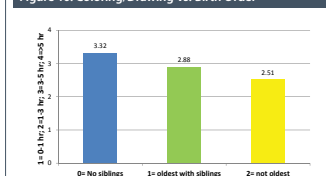


Figure 10: Coloring/Drawing Vs. Birth Order





2341 South Michigan Avenue, Chicago, Illinois 60616

# Retrospective Review of Refractive Error in Two Age Groups of Hispanic and Black Children Seen in a Chicago Vision Clinic

#5685

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## PURPOSE

The study was designed to compare refractive error findings in Hispanic and Black children aged 6-7 years and 11-12 years who were seen in a community based vision clinic serving the Chicago Public School students.

## METHOD

This retrospective cross-sectional study reviewed autorefractometer findings and compared the outcomes from the Hispanic subjects to the Black children seen at the Illinois Eye Institute at Princeton School-based vision clinic from Jan 2011 through Aug 2012. Two states were evaluated: dry and Cycloplegic. Cycloplegic autorefractometer was attained with 1% tropicamide, 2.5% phenylephrine, and 1% cyclopentolate (2 qts). Autorefractometer was conducted at least 30 minutes after the drops were instilled. The study was limited to 6-7 year olds and 11-12 years old children seen for exams during the time period.

### Study Demographics

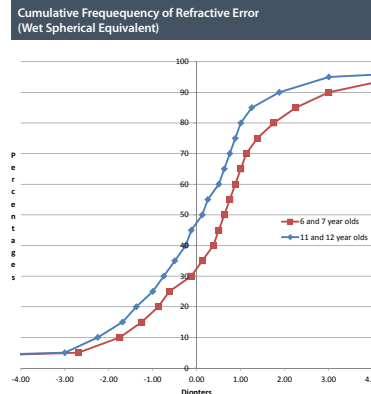
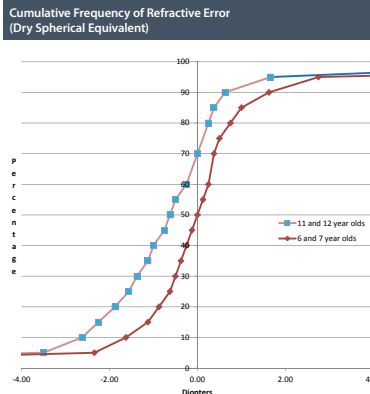
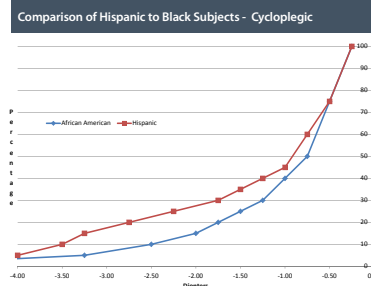
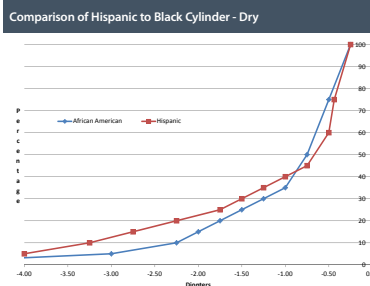
Breakdown of Children by Race and Age Group

Race	Age Group	Frequency	Percentage
Black	6 or 7 years of age	552	19.7
	11 or 12 years of age	1090	39.0
Hispanic	6 or 7 years of age	356	12.7
	11 or 12 years of age	618	22.1
Other	6 or 7 years of age	81	2.9
	11 or 12 years of age	100	3.6

A univariate analysis of right eye only was done for dry and wet cylinder, sphere, and spherical equivalent. In addition, a comparison of each variable with age (younger subjects versus older subjects) was included to observe if shifts in refractive error occurred over time.

Comparison of Autorefractometer Results Between Hispanic Subjects to Black Subjects						
Autorefractometer Type	Race	N	Mean	Std. Deviation	Levene's Test for Equality of Variances	
					F	Sig.
Sphere (Dry)	Black	1610	-0.08	1.95	3.01	.083
	Hispanic	955	-0.15	1.95		
Cylinder (Dry)	Black	1225	-0.95	0.89	117.13	.000
	Hispanic	738	-1.29	1.28		
Sphere (Cycloplegic)	Black	1265	0.68	2.21	2.90	.089
	Hispanic	797	0.55	2.36		
Cylinder (Cycloplegic)	Black	1012	-0.99	0.92	79.88	.000
	Hispanic	623	-1.30	1.28		
Spherical Equivalent (Dry)	Black	1611	-0.44	1.92	0.34	.559
	Hispanic	955	-0.65	1.78		
Spherical Equivalent (Cycloplegic)	Black	1265	0.29	2.14	0.61	.435
	Hispanic	797	0.04	2.18		

Comparison of Autorefractometer Results From Younger Subjects to Older Subjects						
Autorefractometer Type	Age	N	Mean	Std. Deviation	Levene's Test for Equality of Variances	
					F	Sig.
Sphere (Dry)	6 or 7 years of age	964	0.49	1.97	2.37	.124
	11 or 12 years of age	1772	-0.43	1.89		
Cylinder (Dry)	6 or 7 years of age	784	-1.27	1.15	32.69	.000
	11 or 12 years of age	1302	-0.96	1.00		
Sphere (Cycloplegic)	6 or 7 years of age	768	1.09	2.58	11.62	.001
	11 or 12 years of age	1429	0.36	2.05		
Cylinder (Cycloplegic)	6 or 7 years of age	644	-1.25	1.20	26.68	.000
	11 or 12 years of age	1098	-1.02	0.99		
Spherical Equivalent (Dry)	6 or 7 years of age	964	-0.02	1.90	4.52	.034
	11 or 12 years of age	1773	-0.78	1.81		
Spherical Equivalent (Cycloplegic)	6 or 7 years of age	768	0.57	2.47	6.12	.013
	11 or 12 years of age	1429	-0.03	1.95		



## RESULTS

Subjects included 974 Hispanics (n=356, 6-7 year olds, n=618, 11-12 year olds) and 1642 Black (n=552, 6-7 yr olds, n=1090, 11-12 year olds).

Univariate analysis comparing findings between each age group and each race showed a significant difference ( $p < 0.001$ ) in refractive cylinder in both dry and wet states but no difference in sphere or spherical equivalent with the Hispanic subjects showing more cylinder. The data also showed a significant myopic shift from the younger age group to the older group ( $p < 0.05$ ).

## CONCLUSIONS

Hispanic children show significantly more cylinder in both dry and Cycloplegic states than Black children. This finding is in agreement with literature on refractive error in these races reported in other geographic areas in the US.

In addition, a myopic shift is seen in the subjects from the age of 6-7 years to the age of 11-12 years. Limitations include the fact that this is a cross-sectional and not a longitudinal study.

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# A Comparison of the MacuScope and QuantifEye Macular Pigment Densitometers in Two Distinct Population Types

#3777

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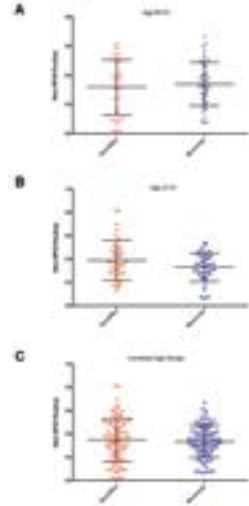
## BACKGROUND

Studies have suggested that reduced levels of macular pigment (MP) may increase risk for developing age-related macular degeneration (AMD). There are two compact commercially available heterochromatic flicker photometry instruments that measure MP in the USA. Previous studies revealed significant variability between instruments. Our aim was to determine if the same variability would be found in a young, healthy, population compared to an older population for which these instruments have more significance.

## METHODS

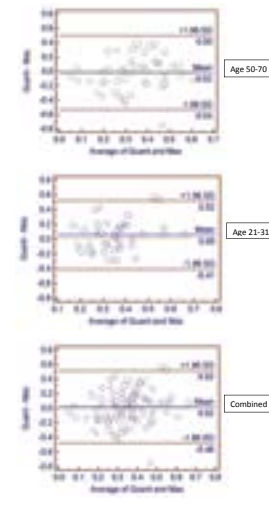
Twenty-one young healthy adults (21-29 years old) and 28 older adults (50-70 years old) with and without early signs of AMD were recruited from the Illinois Eye Institute patient base. Macular pigment optical density (MPOD) was measured using the MacuScope and QuantifEye. Data was collected for each patient in one session. A single, but different operator collected data for each of the patient populations. Whether a subject was first tested on the MacuScope or QuantifEye was randomly determined. Two measurements per eye were taken on each instrument and each eye was used as a separate data point. If the difference was greater than 0.04 absorbance units between two measurements on a single instrument, a third measurement was taken. Invalid readings were excluded. Of the 98 eyes tested, 85 paired eyes provided valid data for comparison of the mean MPODs while 89 (MacuScope) and 88 (QuantifEye) individual eyes provided valid data for comparison of the individual subjects standard deviations. Paired t-tests and ANOVA were done to compare the statistical significance of the results from both instruments and age groups. Bland-Altman plots were done for an added comparison.

Figure 1



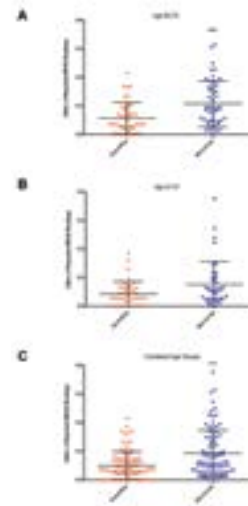
**Figure 1:** Comparison of the mean MPOD level in the eyes of subjects using the QuantifEye™ and MacuScope. **A.)** The mean MPOD reading for adults aged 50-70 (n=46 eyes) was  $0.317 \pm 0.191$  for the QuantifEye™ and  $0.341 \pm 0.149$  for the MacuScope. **B.)** The mean MPOD reading for the adults aged 21-29 (n= 39 eyes) was  $0.3870 \pm 0.171$  for the QuantifEye™ and  $0.331 \pm 0.121$  for the MacuScope. **C.)** The mean MPOD for the combined age groups (n= 85 eyes) was  $0.349 \pm 0.184$  for the QuantifEye™ and  $0.336 \pm 0.136$  for the MacuScope. A paired student's t-test was performed using GraphPad Prism 5.0 software and there was no significant difference found between the two machines in any of the groups.

Figure 2



**Figure 2:** Repeated measures Bland-Altman plot comparison of the QuantifEye™ and the MacuScope. **A.)** 46 eyes from adults aged 50-70 years old with at least 1 and no more than 3 MPOD readings per eye were used in the comparison of the instruments. **B.)** 39 eyes from adults aged 21-29 years old with at least 1 and no more than 4 MPOD readings per eye were used in the comparison of the instruments. **C.)** 85 eyes from the combined age groups with at least 1 and no more than 4 MPOD readings per eye were used in the comparison of the instruments. The differences in the MPOD measurements between the two techniques are plotted against the average MPOD reading of the two techniques. The mean of the differences (bias) was  $-0.0193$  from the 50-70 group,  $0.0555$  from the 21-29 group, and  $0.0175$  from the combined groups and reflects the systematic error. The upper and lower limits of agreement ( $\pm 1.96$  SD) were  $0.498$  and  $-0.536$  for the 50-70 group,  $0.5181$  and  $-0.4071$  for the 21-29 group, and  $0.5162$  and  $-0.4812$  for the combined groups. These represent the precision of the instruments.

Figure 3



**Figure 3:** Comparison of the mean Standard Deviations (SD) of repeated MPOD measurements from individual eyes on the QuantifEye™ and MacuScope. **A.)** Eyes were measured 2-3 times with the QuantifEye™ (n=46) and with the MacuScope (n=50) from adults age 50-70. The mean SD of MPOD measurement from each eye was  $0.0577 \pm 0.0544$  from the QuantifEye™ and  $0.1074 \pm 0.0798$  from the MacuScope. An unpaired student's t-test was performed using GraphPad Prism 5.0 software. The  $p=0.0006$  indicates a significant difference when looking at the individual subject variability between the 2 instruments. **B.)** Eyes were measured 2-3 times with the QuantifEye™ (n=42) and with the MacuScope (n=39) from adults age 21-29. The mean SD of MPOD measurement from each eye was  $0.0425 \pm 0.0425$  from the QuantifEye™ and  $0.0749 \pm 0.0815$  from the MacuScope. The  $p=0.0596$  signifies no significant difference between the instruments. **C.)** Eyes were measured 2-3 times with the QuantifEye™ (n=88) and with the MacuScope (n=89) from the combined groups. The mean SD of MPOD measurement from each eye was  $0.0505 \pm 0.0494$  from the QuantifEye™ and  $0.0932 \pm 0.0817$  from the MacuScope. The  $p=0.0004$  indicates a significant difference when looking at the individual subject variability between the 2 instruments.

## RESULTS AND CONCLUSIONS

- There was no significant difference between the mean MPOD readings of the two instruments for either age group (**Figure 1**).
- A repeated measures Bland-Altman analysis revealed that overall, both machines are sufficient to measure MPOD in both age groups (**Figure 2**) with a relatively even distribution above and below the mean. However, the spread of the paired observations between the limits of agreement demonstrates a lack of precision between the instruments.
- There is a significant difference in intra-instrument measurement variability when considering individual subject variability in the 50-70 age group and the combined age group (**Figure 3**). The individual subject variability for each instrument with the 21-29 age group, while not statistically significant, demonstrated a wide range variability.
- Is this variability acceptable?
- The clinician must take this variability into account if using MPOD as an indicator for AMD risk and/or clinical care.
- If MPOD is being monitored clinically to assess risk of AMD with the possibility of causing altered treatment regimens, the need for reliable data measurements is imperative.

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# Using EdU in Whole Pig Lenses to Establish Anterior Epithelial Cell Division Rates

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## Background:

In 2005, it was estimated that 1.04 billion people were presbyopic globally, and that 517 million are not adequately corrected. This number will only increase with an aging population. Current treatments for presbyopia include spectacle lenses and accommodating IOLs, but no treatments fully return the natural accommodative abilities of the younger lens. Presbyopia creates both a financial and psychological detriment to the patient, and therefore there is a need for improved correction<sup>1</sup>. As the crystalline lens grows throughout life, the germinative zone lens epithelial (GZ LE) cells located close to the equator divide and differentiate into lens fibers. New fibers overlay the existing lens mass, with multiple layers of younger fibers compacting the fibers beneath and increasing the diameter of the lens. It is believed that compaction plays a significant role in decreased flexibility of the lens with age, and therefore presbyopia<sup>2</sup>. It is plausible that decreasing the rate of cellular division of the GZ LE cells may also preserve lens flexibility and subsequent accommodation.

We are currently investigating methods to alter lens growth rates. In this pilot study, we are quantifying the cellular division rate and specific location of the GZ LE cells of whole porcine lenses exposed to the thymidine analog, 5-ethynyl-2'-deoxyuridine (EdU), which labels for active DNA synthesis in proliferating cells. In this manner, cells that were undergoing DNA synthesis will be marked following mitosis<sup>3</sup>.

## Methods:

Fresh porcine eyes were obtained from a local abattoir (Park Packing Company, Chicago IL). Lenses removed within 2 hours of collection. Using modifications of previously published methods<sup>3</sup>, 18 lenses were treated with chymotrypsin (3mg/mL) and washed before incubating in EdU (2.52 µg/µL) for 2 hours at 37°C. The lenses were fixed and permeabilized before tagging with "click" chemistry. The EdU labeled nuclei were conjugated to the Alexa594 fluorophore during a 1 hour incubation at room temperature protected from light. Lenses were washed before being counterstained with SYTO 9 to visualize all background nuclei. Samples were viewed using a Nikon A1R-MP+ multiphoton microscope (Northwestern University Cell Imaging Facility-Nikon Imaging Center). Collected images were viewed and analyzed using Fiji image J (NIH).

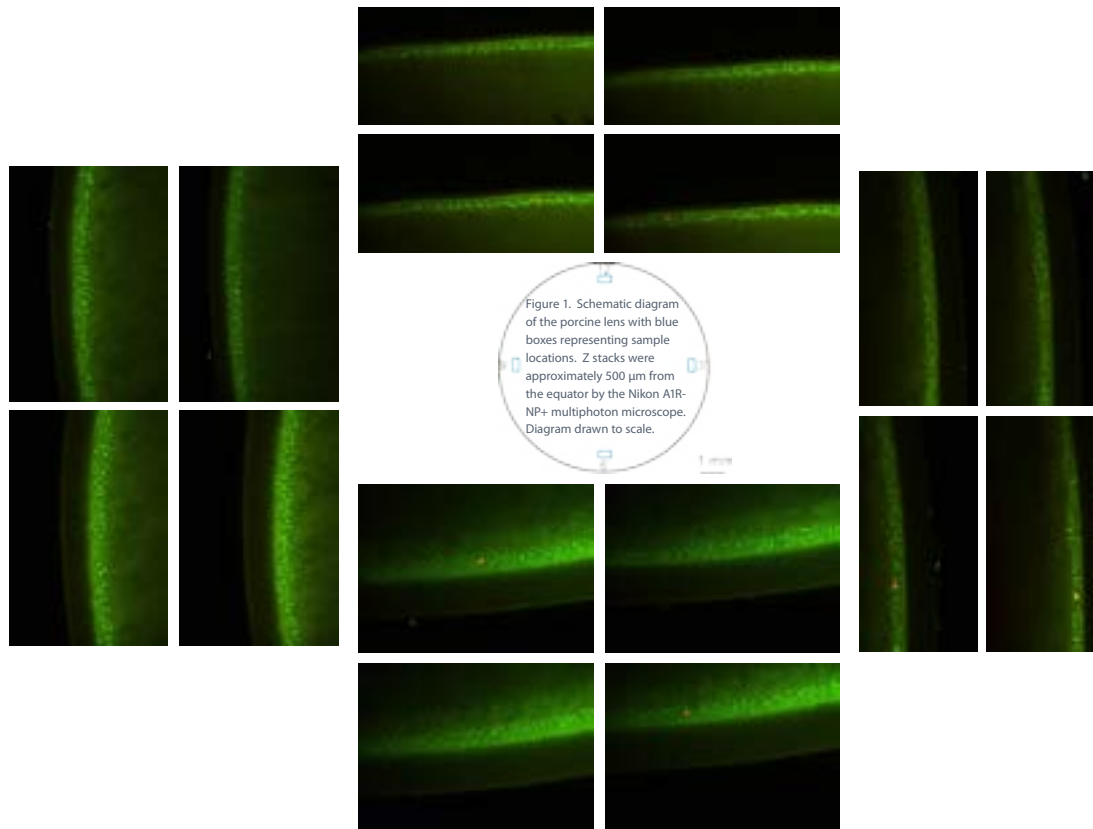
## Results:

In our preliminary assessment of the lenses, approximately 0.31% of the GZLE cells were labeled with EdU. These cells were located approximately 500 µm anterior to the equator of the lens and the width of the area measured was 400 µm.

	Number of EdU Labeled Cells	Total Number of Cells	Percentage of EdU Labeled Cells
3'	16	3352	0.48%
6'	12	3317	0.36%
9'	7	4510	0.16%
12'	6	2273	0.23%
TOTAL	41	13452	

**Table 1.** Cell counts per Z stack of 400 um2 and percentage of EdU labeled cells as taken by the Nikon A1R-MP+ multiphoton microscope.

All results reported are preliminary and discoveries in the future may alter these results.



## Discussion:

- We were successfully able to identify dividing cells in the GZLE cells using EdU as a marker. The 0.31% of mitotic cells located 500 µm from the equator that we report is comparable with other literature<sup>4,5</sup>.
  - Although the geometric patterns are slightly different, porcine lenses are a viable model to their human counterparts due to similar protein composition to human lenses<sup>6,7</sup> and easy availability from nearby slaughterhouse.
  - We will be utilizing this procedure to establish baseline values for comparisons to treated lenses.
- Future Directions:
- Create a more accurate and repeatable method for quantifying EdU labeled cells, representative of the entire lens epithelium.
  - Consider counterstaining background nuclei with Hoechst 33342 to reduce interference between the red and green fluorescent channels.
  - Compare our reported percentage of mitotic GZLE cells to laser treated lenses.

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## Funded by:

ICO RRC  
Lenticular Research Group

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# Is Peripheral Retinal Ischemia Associated with Systemic Atherosclerosis?

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#194

## INTRODUCTION

Diabetes and carotid intima media thickness (IMT) are both well-known risk factors for cardiovascular disease and atherosclerosis. The Framingham Study showed a nearly 15x increase risk in cardiovascular events in middle-age patients with diabetic retinopathy (DR). Technological advances in recent years have produced smaller, portable ultrasound devices making screening of IMT in the carotid arteries easier and more accessible. Fluorescein angiography technology has evolved, providing up to 200 degree fundoscopic views in a single, high-resolution image. These images have allowed improved visualization of the extent of peripheral retinal nonperfusion. Previous studies have found that diabetic patients presenting with retinopathy have a higher IMT than diabetic patients without retinopathy and patients with severe DR are more likely to have subclinical vascular disease.

The purpose of this study was to investigate whether there was a correlation between peripheral retinal nonperfusion and IMT thickness of the common carotid artery in patients with DR. A positive correlation might suggest a significant relationship between microangiopathy in the retina and systemic macroangiopathy. This association might allow for more diverse and non-invasive means to screen for retinal nonperfusion. Conversely, identification of peripheral retinal nonperfusion may warrant further cardiovascular evaluation in diabetic patients.

## METHODS

In this cross-sectional pilot study, 15 subjects with diabetes and advanced DR were enrolled. No subject had PRP treatment. Ultrasound images of both common carotid arteries were taken using the HeartSmart IMTplus<sup>®</sup> portable ultrasound device (Figure 1). The scans were electronically transferred and read by an off-site reading facility. A grade of IMT was assigned by the reading facility based on comparison to a database of over 40,000 patients mapped and categorized based on age, gender and race. A grade of normal represents the mean IMT for age and gender, and mild, moderate and severe categories represent 2, 4, and 8 standard deviations above the mean, respectively. Ultra-

widefield fluorescein angiography (UWFFA) images on both eyes were obtained using Optos C200 scanning laser ophthalmoscope (Figure 2). The same certified retinal photographer obtained all images. Using area measurement function in the V2 Vantage Review Software (Optos, PLC), one of the coauthors determined both the total area of gradable fundus and area of nonperfusion seen in the arteriovenous phase of the UWFFA. Ischemic index (ISI) was calculated by dividing the nonperfused retinal area with the total retinal area. An ISI of 0 indicates a fully perfused retina while an ISI of 1.0 indicates complete absence of retinal capillary perfusion. One-way ANOVA was performed to test if there was a difference in ISI among normal, mild, moderate, and severe IMT groups both in the right and left side.

Figure 1: Intima-Media Thickness Report

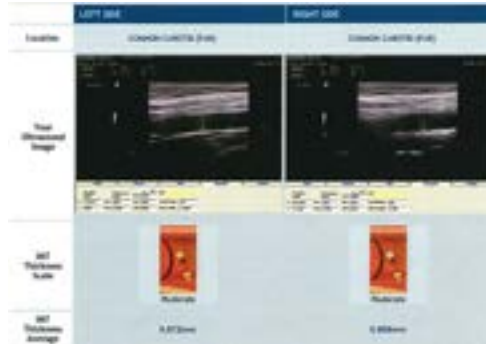


Figure 2: Ultra-Widefield Fluorescein Angiogram. The white border outlines the extent of gradable fundus and the blue border delineates the extent of peripheral nonperfusion. The ISI is 0.21.

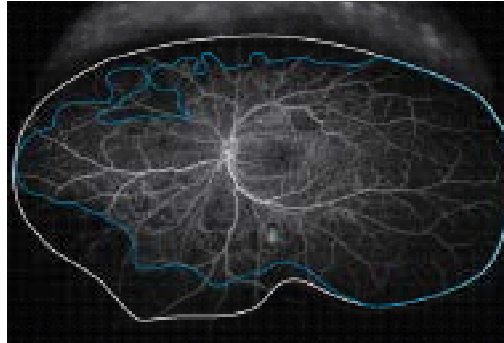


Table 1: Demographics

Age (yrs)	Race	Sex	Length of diagnosis (yrs)	LBG (mg/dl)	Smoking history	HTN
56	AA	M	5	200	Never	Yes
50	AA	M	12	107	Current	No
58	AA	F	16	160	Former	Yes
50	W	M	1	160	Former	Yes
44	H	M	unknown	160	Never	Yes
41	H	F	unknown	185	Never	No
66	AA	F	11	170	Never	No
46	H	F	14	120	Never	No
26	H	M	12	120	Never	No
71	C	M	22	89	Former	Yes
63	AA	M	4	144	Former	Yes
78	AA	F	21	118	Never	Yes
49	C	M	14	160	Never	Yes
77	AA	F	unknown	127	Current	Yes
58	AA	M	unknown	72	Never	Yes

## RESULTS

Compiled demographic data from the 15 subjects is presented in Table 1. Average ISI in the right eye was 0.145 (± 0.134) ranged from 0.01 to 0.40, and 0.144 (± 0.144) in the left eye ranged from 0.01 to 0.47. ISI values classified by IMT group are presented in Table 2. No subjects were classified as severe. No statistically significant difference was identified in right eye ISI among normal, mild, and moderate IMT groups in the right carotid ( $F_{2,12} = 1.31, P = 0.31$ ). However, there was significant difference in left eye ISI among all IMT groups in the left carotid ( $F_{2,12} = 7.63, P = 0.007$ ). Post hoc analysis reveals the difference in left eye ISI was between the normal and mild IMT groups ( $P = 0.006$ ).

Table 2: ISI values for IMT grade

	Normal	Mild	Moderate
OD	0.0267	0.2291	0.0512
	0.2276	0.2708	0.0948
	0.0059	0.0253	0.0184
	0.0099	0.3636	0.2352
		0.1595	0.3985
			0.0620
n	4	5	6
Mean	0.0675	0.2097	0.1433
OS	0.0386	0.4664	0.1063
	0.1428	0.2979	0.0925
	0.0131	0.0126	0.1431
	0.0326	0.2829	0.1134
	0.0164	0.3692	
	0.0298		
n	6	5	4
Mean	0.0456	0.2858	0.1139

## CONCLUSION

- In this pilot study we looked for a correlation between peripheral retinal nonperfusion and IMT grade of the common carotid artery in patients with DR.
- We found no statistical significant difference in ISI among IMT groups on the right side.
- We found significant difference in ISI among normal and mild IMT on the left side.
- Our study is limited by a small sample size and a narrow range of ISI (0.01-0.47) and IMT grade (normal – moderate). Previous studies investigating ISI have found values from 0-1.0.
- Our study supports a common pathogenic mechanism for both macro and microangiopathy but the data set may be too narrow to identify significance between groups.
- Further investigation with a larger cohort may provide more accurate results.

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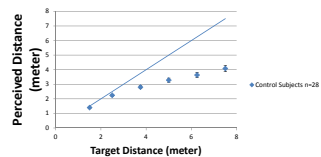
## INTRODUCTION

A number of authors have reported that human observers can accurately judge the physical distance of visual targets if they are located on a flat ground surface. Distance judgments measured with the blind-walking technique correlate highly with a target's physical distance in lighted conditions. However distance judgments of self-illuminated targets in darkness are very inaccurate.

Ooi and colleagues have proposed that in both darkness and light, observers judge an object's distance as the intersection between its angle of declination and the perceived slant of the ground surface. They report that in darkness the human visual system relies on a default ground surface called the implicit surface that tilts upwards towards the horizon with a slant of around 12 deg.

Our experiments have revealed another aspect of this implicit surface; it is triggered by the absence of light in the visual field and suppressed by the presence of light in the visual field.

Figure 1: Perceived Distance Measured by Blind-Walking in Darkness

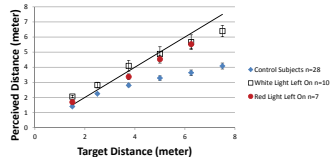


## METHODS

### Measuring Perceived Distance

- Subjects were instructed to look at a small, illuminated target placed on the floor of a large dark space.
- Subjects viewed the target monocularly for as long as needed to identify and remember the target location.
- They were then instructed to occlude both eyes and walk to the remembered target location.
- This technique is referred to as **BLIND-WALKING**.
- They were asked to place their toes at the point where they felt the small target had been.
- Once they stopped at the remembered location the experimenters measured the perceived distance of the target.
- A few practice trials were administered to each subject so they were clear on the instructions.

Figure 2: Perceived Distance vs Physical Distance as Measured by Blind-Walking Technique



## SUBJECTS

- Three groups of different subjects participated in the study.
- All subjects in each of the three groups were in their twenties and students at the Illinois College of Optometry (ICO). All subjects were naive with respect to the purpose of the study.

### Control Group: Blind-Walking in Darkness

- The first group was comprised of 28 subjects. They viewed the small illuminated target in darkness then blind-walked to the remembered location in darkness.
- When they indicated the remembered location the lights were turned on and perceived distance was measured by the experimenters.

### Blind-Walking in White Light

- The second group was comprised of ten ICO students.
- These subjects were also instructed to view the small target in the dark, under monocular conditions.
- After they viewed the target they occluded both eyes and blind-walked to the remembered location.
- However, when they performed the blind-walking task the room was illuminated with white light.
- The luminance reflected off the floor directly in front of the target was approximately 30 cd/m<sup>2</sup>.

### Blind-Walking in Red Light

- The second group was comprised of seven ICO students.
- These subjects were also instructed to view the small target in the dark, under monocular conditions.
- After they viewed the target they occluded both eyes and blind-walked to the remembered location.
- However, when they performed the blind-walking task the room was illuminated with 3 goose-neck lamps each fitted with a red 40 watt compact fluorescent bulb.
- This provided a low photopic luminance.

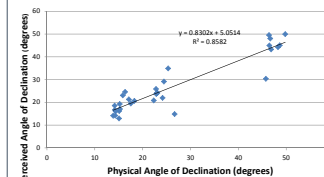
## RESULTS

- When subjects are asked to blind-walk to the remembered location in complete darkness they significantly underestimate target distance as shown in **Figure 1**.
- Note that when the target is actually 7.5m from the subject the subjects tend to perceive it as only about 4m away.
- Our data confirm the target underestimation previously reported by Ooi et al. when subjects were asked to blind-walk to the remembered target location in darkness.
- However, when subjects viewed the target in darkness and then blind-walked to the remembered location with the room illuminated with white light the results were much more accurate as shown in **Figure 2**.
- When subjects were asked to blind-walk to the remembered target location in the presence of red light, the results did not differ from those obtained in the white-light condition.
- A two-way analysis of variance (ANOVA; distance x lighting) was used to compare the mean distance estimates obtained under different conditions of illumination while blind-walking (SPSS version 17.0, Chicago, IL).
- The overall F value = 18.85, p<0.001.
- Pairwise comparisons indicated that there was no difference between the distance estimates obtained in either lighted condition (p=0.21) but both these distance estimates differed significantly from the dark condition (p<0.01).

## CONCLUSIONS

- The distance perception of targets viewed in darkness is dramatically affected by the presence or absence of room lighting; when subjects blind-walk to the remembered target location in darkness we assumed they relied on the implicit surface which according to the theory proposed by Ooi et al (2006) would result in significant **underestimates** of target distance which should get worse the farther away the target. As can be seen in Figure 1 this assumption proved true.
- We assumed that if subjects **blind-walked** to the remembered target location with room illumination present (but they were still blindfolded) then there would again be a default to the implicit surface and distance perceptions would be underestimated.
- However, as Figure 2 clearly indicates the expected underestimation does not occur. Despite the fact that the subjects are blindfolded and received no visual cues other than that light was left on the room, they blind-walked to the correct location.
- One conclusion that may be drawn from this result is that the visual system fails to default to the implicit surface unless there is darkness.

Figure 3: Perceived Angle of Declination vs Physical Angle of Declination



- Li and Durgin (2012) offered a different explanation for the foreshortened distance perceptions subjects exhibit during blind-walking in a dark environment; they suggest that subjects overestimate the angle of gaze declination (while looking at the self-illuminated target in the dark) and thus when asked to blind-walk to the remembered location they exhibit a foreshortening. They further suggest that the angle of gaze declination **appears** to be accurate (Ooi et al, 2006) because subjects underestimate their perceived self-motion and thus underestimate their walked distance. Thus because their action (walking) is calibrated to their underestimation of optic flow they will act as if their gaze declination is accurate.

- Although the hypothesis of Li and Durgan is interesting and has evidence to support it, it doesn't explain why blind-walking is accurate when subjects have knowledge of light in the room. Our results also do not support their general claim that gaze declination is exaggerated as we found the gaze declination to be accurate when blind-walking occurred in the presence of a light as shown in **Figure 3**.

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# Effect of Amblyopia Treatment on Macular Thickness in Eyes with Myopic Anisometropic Amblyopia

#3991

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## INTRODUCTION

In a previous study, we reported that amblyopic eyes with unilateral high myopia had significantly thicker minimum and average foveas but thinner inner and outer maculae compared to sound eyes.<sup>1</sup> In addition, we found that both refractive correction and patching significantly improved visual acuity (VA) of the amblyopic eye associated with myopic anisometropia.<sup>2</sup> The purpose of this study was to investigate whether amblyopia treatment (both refractive correction and patching) affected macular thickness in the amblyopic eyes with unilateral high myopia. Furthermore, the association between VA improvement and the change of macular thickness in the amblyopic eyes was determined.

Table 1: Eligibility and Exclusion Criteria

### ELIGIBILITY CRITERIA

- Age: 4 to < 14 years
- Best-corrected VA in the amblyopic eye 20/40 to 20/400 inclusive
- Best-corrected VA in the sound eye 20/40 or better
- Myopic anisometropia more than 3.00 D
- Inter-eye acuity difference >3 logMAR lines
- Amblyopia associated with myopic anisometropia
- No amblyopia treatment (other than spectacles) in the past month and no more than one month of amblyopia treatment in the past 6 months

### EXCLUSION CRITERIA

- Presence of ocular pathology causing reduced visual acuity
- Prior ocular surgery
- Current vision therapy
- Known skin reaction to patch or bandage adhesive

## METHODS

A total of 20 children diagnosed with myopic anisometropic amblyopia were recruited at the Illinois Eye Institute, an urban eye clinic, and 17 children (range of VA in the amblyopic eye: 20/80 to 20/400) completed the study with the other 3 subjects lost to follow-up. All subjects underwent a comprehensive eye examination including VA at far and near, cover test at far and near, stereoacuity with both the Randot<sup>®</sup> Preschool Stereoacuity Test and Stereo Fly, manifest refraction, dilated fundus examination with indirect ophthalmoscopy, cycloplegic refraction, monocular fixation using visuscopy, and A-scan ultrasound biometry. The eligibility and exclusion criteria are listed in **Table 1**.

Direct patching, 2 hours per day for moderate amblyopia and 6 hours per day for severe amblyopia, was initiated 16 wks following the initiation of refractive correction. The patching time was consistent throughout the study. Optical coherence tomography (Stratus OCT3; Carl Zeiss Meditec, Dublin, CA) was performed on all subjects at the enrollment visit and at the end of 16 weeks of patching. All scans had signal strength of at least 6. The following ten parameters were measured for each subject: foveal minimum thickness, average foveal thickness, inner nasal, superior, temporal, and inferior macular thickness, outer nasal, superior, temporal and inferior macular thickness. **Figure 1** demonstrates the locations of fovea, inner macula, and outer macula. The foveal area is defined as the central circle of 1 mm diameter. Inner macula refers to the ring around the foveal area of 3 mm diameter and outer macula refers to the ring around the inner macula of 6 mm diameter.

Paired t-test was performed to compare macular thickness of the amblyopic eye before and after amblyopia treatment. Spearman correlation was used to test the relationships between macular thickness change and VA improvement in amblyopic eyes. For multiple comparisons and correlations among the 10 OCT parameters, Bonferroni's correction was applied with a resultant significance level of  $P < 0.005$ .

## RESULTS

Mean ( $\pm$ SD) baseline VA in the amblyopic eyes was  $0.96 \pm 0.27$  logMAR and improved to  $0.71 \pm 0.30$  after amblyopia treatment, an average improvement of 2.5 logMAR lines ( $P < 0.0001$ ). **Table 2** shows the macular thickness measurements before and after amblyopia treatment. No statistically significant difference was identified in macular thickness before and after treatment, although there was a trend toward decreasing minimum and average foveal thickness after treatment. No correlation was found between macular thickness change and VA improvement. A sample OCT image from one of the subjects is shown in **Figure 2** (before amblyopia treatment) and **Figure 3** (after amblyopia treatment).

Figure 1: Schematic diagram to demonstrate the locations of fovea, inner macula, and outer macula

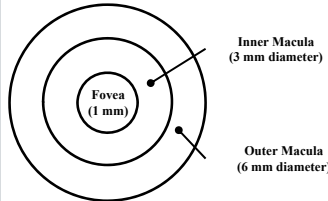
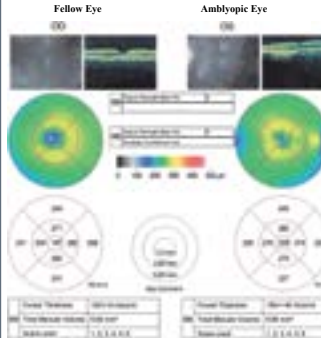


Table 2: Macular Thickness ( $\mu$ m) of the Amblyopic Eyes before and after Amblyopia Treatment (n = 17)

Location	Before Amblyopia Treatment (Mean $\pm$ SD)	After Amblyopia Treatment (Mean $\pm$ SD)	P Value
Foveal minimum	171.25 $\pm$ 23.97	157.81 $\pm$ 24.81	0.12
Average foveal (1 mm)	198.19 $\pm$ 16.66	189.06 $\pm$ 16.79	0.05
Inner macula (3 mm)			
Nasal	259.69 $\pm$ 16.52	255.81 $\pm$ 11.36	0.34
Superior	258.25 $\pm$ 17.86	261.25 $\pm$ 14.42	0.14
Temporal	247.25 $\pm$ 17.40	248.19 $\pm$ 16.41	0.52
Inferior	254.13 $\pm$ 17.66	257.63 $\pm$ 11.30	0.14
Outer macula (6 mm)			
Nasal	245.81 $\pm$ 15.19	243.00 $\pm$ 14.85	0.85
Superior	232.69 $\pm$ 13.92	225.63 $\pm$ 18.28	0.87
Temporal	207.06 $\pm$ 12.89	207.88 $\pm$ 20.89	0.41
Inferior	213.50 $\pm$ 11.39	213.63 $\pm$ 13.88	0.96

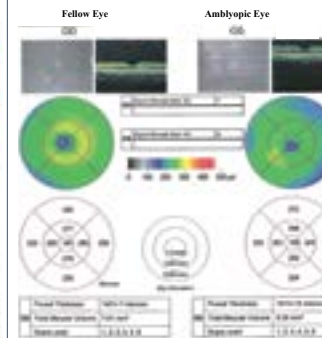
SD= standard deviation. Bonferroni's correction for multiple correlations, significant at  $P < 0.005$ .

Figure 2: OCT Image of One of the Subjects before Amblyopia Treatment.



Subject information: 7 years old, female, AA. VA: 20/25 OD, 20/250 OS, 10° CLXT, 6° XP°. Cyclo-refraction: OD: PL OS: -13.00sph. After amblyopia treatment: VA: 20/20 OD, 20/160 OS

Figure 3: OCT Image of One of the Subjects after Amblyopia Treatment.



## CONCLUSION

- Amblyopia treatment improved VA in myopic amblyopic eyes, but anomalies in macular thickness of these eyes remained.
- These findings suggest that VA improvement in myopic amblyopic eyes is not due to macular thickness changes.

## COMMENTS

This study was supported by the Illinois College of Optometry Research Fund, Illinois Society for the Prevention of Blindness, and CIBA Vision. The authors thank Rebecca Tudor for her assistance in performing OCT on all subjects.

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# The Effect Of Age On The Lens Ultrastructure During Accommodation As Measured Using Slit Lamp Photos And Wave Front Analysis

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## Purpose:

During dynamic focusing, the shape, as well as the ultra-structure of the lens is changed. Our lab is investigating these changes, specifically at the sutures of the lens during accommodation. We have hypothesized that unique structural features and organization of fiber cells enables them to interface at the sutures resulting in a change in surface curvature of the lens, as well as an increase in thickness, allowing near focus to occur. We are reporting data on lens slit lamp photos, OCT of lens thickness changes, and sequential ray tracing analysis of the patterns associated with the lens sutures resulting in a change in surface curvature of the lens, as well as an increase in thickness, allowing near focus to occur. We are reporting data on lens slit lamp photos, OCT of lens thickness changes, and sequential ray tracing analysis of the patterns associated with the lens sutures resulting in a change in surface curvature of the lens, as well as an increase in thickness, allowing near focus to occur.

## Method:

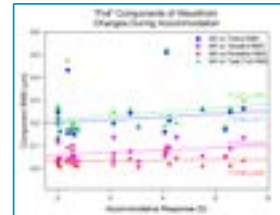
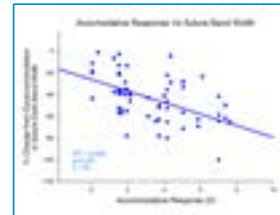
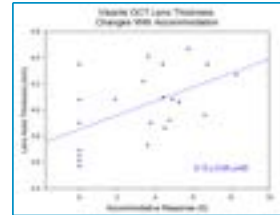
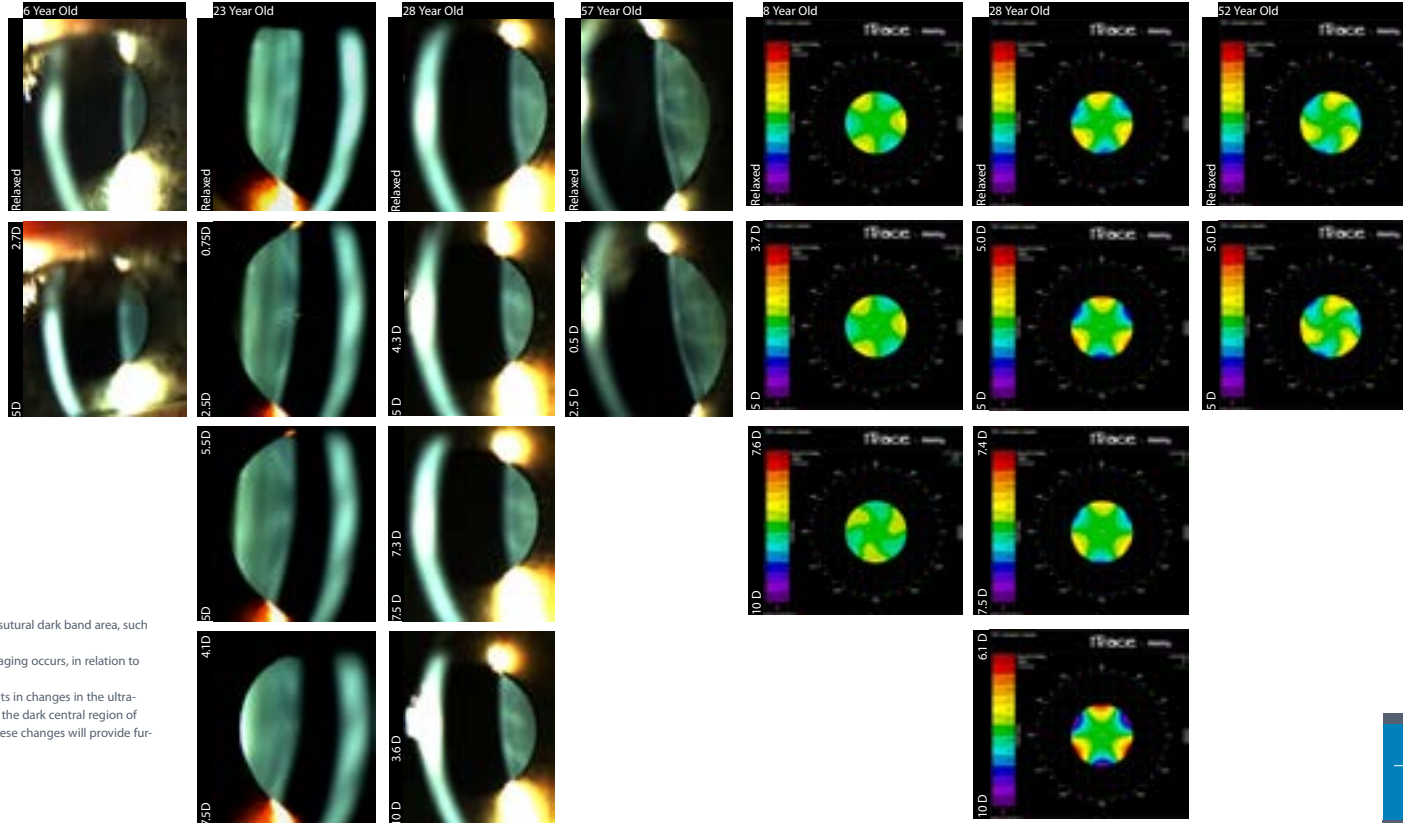
OCT (VisanteTM), wavefront analysis (iTraceTM), and slit lamp photos (Haag Streit, 16X magnification, dilated eye) were collected on normal subjects, between the ages of 7 - 63 (n=30). The data was collected on the right eye. Accommodation was stimulated using minus lenses in front of the viewing eye in 2.5 D increments until the subject could no longer clearly view the target. For the photos a prism system was used to keep the eye appropriately oriented. The objective accommodative response was calculated as the change from a distance measurement using the iTrace refractive measurements corrected for stimulation lens power. ImageJ (NIH) was used to analyze the area of the sutural dark central region. Data were analyzed using Systat v11 to correlate accommodative response with total HOA, SA and the foil patterns, as well as changes in slit lamp suture areas. Spearman Rank Correlation coefficients and p values are presented.

## Conclusions:

- In a young lens, accommodation leads to a decrease in the sutural dark band area, such that it is difficult to see
- This decrease in the area of the dark band is maintained as aging occurs, in relation to the objective accommodative response.

Regardless of age, increase in accommodative response results in changes in the ultra-structure of the lens, as evidenced by the decrease in area of the dark central region of the suture and in the total foil patterns. Further analysis of these changes will provide further insight into the anatomical basis of accommodation.

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# Anterior Chamber Depth, Lens Thickness, and Related Measures in African-American Females with Long Anterior Zonules

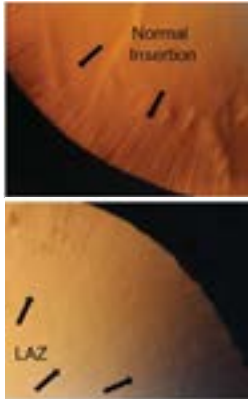
#3504

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## BACKGROUND

Long anterior zonules (LAZ) are characterized by the presence of crystalline lens zonules central to the normal insertion zone on the anterior capsule (Fig. 1).<sup>1,2</sup> They frequently become pigmented due to contact with the posterior iris, and may be detected as radially-oriented, fine brown lines after pupillary dilation. Other signs of pigment dispersion commonly occur. In addition to this unique type of pigment dispersion, at least one subtype of LAZ is associated with female gender, older age, hyperopia, and shorter axial length.<sup>1,2,4,5,14</sup> Observations so far have suggested that this LAZ phenotype may be a risk factor for angle-closure glaucoma, possibly in association with plateau iris configuration.<sup>1,9</sup> The ocular dimensions of LAZ eyes are incompletely understood, and the purpose of this investigation was to further assess certain dimensions in LAZ eyes, with simultaneous control for refractive error.

Figure 1: Normal zonule insertion on anterior capsule (arrows, top) vs. long anterior zonules (arrows, bottom).



## METHODS

LAZ subjects were recruited via mailed invitation from a database developed over several years during the provision of primary eye care within an urban, teaching facility, located in Chicago, Illinois, USA. The criterion for LAZ was the presence of zonular fibers  $\geq 1.0$  mm central to the normal zonule termination zone on the anterior capsule (Fig. 1), with exclusion of subjects with  $< 5$  LAZ to ensure definitive cases. Only African-American females were included due to low numbers of males and people from other racial groups at the study site. Controls were recruited when they presented for routine care within the same setting, and invitation was based on one-to-one matching using age ( $\pm 5$  years), race, gender, and refractive error ( $\pm 1.00$  D spherical equivalent).

All participants had ocular/medical history, extraocular muscle testing, color vision testing, confrontation fields, pupil testing, subjective refraction, Goldmann applanation tonometry, four-mirror gonioscopy, and dilated fundus exam. A-scan ultrasonography (CompuScan<sup>®</sup> LT Biometric Ultrasound System; Storz Instrument Co., St. Louis, MO) was performed on each subject by one of three practitioners who were masked to subject status. Five scans were obtained for each eye and averaged for the final measurements, which included anterior chamber depth (ACD), lens thickness (LT), vitreous body length (VBL), and axial length (AL). The eye tested first was randomized, and the right eye measures were analyzed unless inclusion criteria were not met.

Data analysis was carried out using the SAS<sup>®</sup> System, Release 9.2 for Microsoft Windows<sup>®</sup> (SAS Institute, Inc., Cary, NC, USA). Conditional multiple logistic regression was used to further ensure simultaneous control of age and refractive error. Institutional Review Board approval was obtained for this investigation, and subjects provided informed consent to participate.

## RESULTS

Each of the variables, i.e., age, refractive error, ACD, LT, VBL, and AL, resembled Gaussian distributions with all subjects grouped together and when split into case and control groups. The mean age  $\pm$  SD, range, of the LAZ cases (67.1  $\pm$  7.6 years, 52-85 years) and controls (66.6  $\pm$  8.5 years, 48-85 years) was similar ( $P > 0.1$ ), as was mean refractive error of the cases ( $+1.85 \pm 1.41$ D,  $-1.75$  to  $+4.75$ D) and controls ( $+1.85 \pm 1.31$ D,  $-0.75$  to  $+4.75$ D). The mean values for ACD, LT, VBL, and AL were very similar between the cases and controls, suggesting that any differences were exceptionally small (Table 1). Results did not vary with or without additional control for age and refractive error using regression analysis.

To explore the data further, we evaluated the relationship between ACD and the other variables and found that ACD had the strongest correlations with LT and AL for both groups. In this regard, ACD had a negative correlation with LT (LAZ:  $N=50$ ;  $r = -0.46$ ,  $P=0.009$ ; Controls:  $N=50$ ;  $r = -0.52$ ,  $P<0.0001$ ) and a positive correlation with AL (LAZ:  $N=50$ ;  $r = 0.31$ ,  $P=0.02$ ; Controls:  $N=50$ ;  $r = 0.37$ ,  $P=0.008$ ) (Fig. 2). Multiple linear regression showed similar models containing LT, VBL, and AL as significant explanatory variables for the ACD dimension (Table 2).

## DISCUSSION

Since LAZ are associated with hyperopia in our subject population, and since there is question whether the trait is a risk factor for angle-closure, it is logical to ask whether people with LAZ are likely to have differences in their ocular dimensions when compared to similar people without LAZ who have similar refractive error. In this analysis of African-American females, while controlling for refractive error, we did not find significant differences between the groups relative to the variables of interest.

Despite the fact that we did not observe differences in the dimensions studied, this does not imply absence of any elevated risk of angle-closure among LAZ eyes because it has still been observed from prior studies that LAZ eyes are more likely to have higher grades of hyperopia along with shorter axial lengths. Perhaps the risk of angle-closure is elevated among people with LAZ but no more than similar people with similar refractive errors. On the other hand, questions remain whether LAZ eyes are also more likely to exhibit plateau iris and/or other iridocorneal angle differences that may increase the risk of angle-closure. Therefore, it remains unknown whether there are other characteristics of LAZ eyes that could also contribute to angle-closure risk, and it will be important to continue investigation of these questions going forward.

Figure 2: Scatterplots showing the relationship of central ACD relative to refractive error, age, LT, and AL.

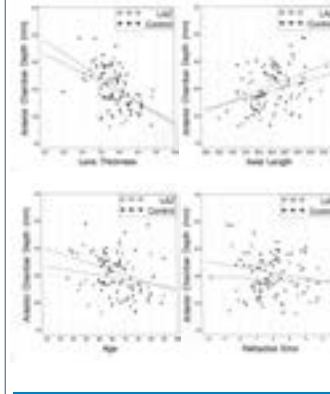


Table 2

MULTIPLE LINEAR REGRESSION MODELS RESPONSE VARIABLE = <sup>1</sup> ACD				
LAZ SUBJECTS (N=50)				
Variable	Coefficient	Standard Error	P Value	
Intercept	0.130	0.170		
X <sub>0</sub> , LT	-0.954	0.019	<0.001	
X <sub>0</sub> , VBL	-0.952	0.022	<0.001	
X <sub>0</sub> , AL	0.952	0.020	<0.001	
CONTROL SUBJECTS (N=50)				
Variable	Coefficient	Standard Error	P Value	
Intercept	-0.174	0.060		
X <sub>0</sub> , LT	-1.000	0.005	<0.001	
X <sub>0</sub> , VBL	-1.002	0.006	<0.001	
X <sub>0</sub> , AL	1.001	0.006	<0.001	

<sup>1</sup>Abbreviations: ACD, anterior chamber depth; AL, axial length; LT, lens thickness; VBL, vitreous body length.  
<sup>2</sup>The explanatory variables age and refractive error were not significant and were removed from the models. Parameter estimates remained stable with and without Age and Refractive Error in the models.

## CONCLUSIONS

This group of African-American females with LAZ did not exhibit clinically significant differences in ACD, LT, VBL, and AL compared to age, race, gender and refractive error matched controls.

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# Contrast Sensitivity (CS) test-retest reliability and consistency with Vector Vision CSV-1000LV and CSV-1000HGT in patients with Albinism

#2760

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## PURPOSE

The ability to accurately monitor the disease status or improve the visual function of low vision patients requires reliable and repeatable vision tests. It is generally accepted that measures of visual function in the low vision patient population are more variable than normally sighted individuals. This may decrease the reliability of standard visual tests when applied to a population such as albinism. The purpose of our study is to determine whether one such test of visual function, contrast sensitivity, could reliably be measured in this population both in the presence of glare and without.

## METHODS

CS tested with Vector Vision CSV-1000 LV which is a low vision letter chart and the CSV-1000HGT (halogen glare test) which tests CS in the presence of glare.

Thirteen subjects with albinism with best corrected logMAR visual acuity (VA) ranging from 0.4 to 0.9, participated in this study. LogMAR VA was tested using the EDTRS chart. Dependent variables including CS under normal test condition and CS with glare, were tested at two different visits, 6 months apart. The CSV-1000LV chart was used to measure CS while the CSV-1000HGT was utilized to measure the effect of glare on CS. Test-retest reliability was assessed with both the correlation of repeatability (COR) and the intraclass correlation coefficient (ICC).

## PROCEDURES

### 1. Measuring best corrected LogMAR visual acuity:

After obtaining informed consent, thirteen subjects with albinism with best corrected logMAR visual acuity (VA) ranging from 0.4 to 0.9 were recruited for this pilot study. LogMAR VA was tested using the EDTRS chart.

### 2. Measuring Contrast Sensitivity

CSV-1000LV-V and CSV-1000 LV-C contrast sensitivity charts were used to provide an appropriate target size for our patient population. Contrast thresholds were obtained at a distance of 4 feet.

### 3. Measuring the effect of glare (Halogen Glare Test)

CSV-1000HGT was used with the CSV-1000 LV test face to evaluate the effect of glare on contrast sensitivity. In medium setting, the unit is precisely calibrated to simulate two on-coming halogen car headlights as seen at night from 150 feet.

## RESULTS

1. **Figure 1a** plots the average LogMAR VA results for the thirteen subjects in our sample for Visit 1 versus the values obtained for the follow-up visit. The average LogMAR acuity was 0.58 for the first visit and 0.63 at the follow-up visit. The slightly higher mean observed at the follow-up visit is not statistically significant (paired t value=-1.72, p=0.06).

2. **Figure 1b** plots the best corrected LogMAR VA for the thirteen subjects for Visit 1 versus the values obtained for the follow-up visit. The solid line has a slope of 1.0. Although the correlation coefficient is fairly high (0.76) correlation coefficients cannot be used to assess test-retest reliability because the absolute value of the correlation varies with the range of the data points. The test-retest reliability was calculated with the ICC and is described below.

3. **Figure 2a** presents the average CS test results (n = 13) under normal test conditions with no glare for test and follow-up visits. Again there is a slight increase in the follow-up mean log CS which in this case is significantly different from the log CS obtained at the initial visit (t= 1.78, p=0.01).

4. **Figure 2b** plots CS test results (n = 13) with Vector Vision CSV 1000LV, under normal test conditions (i.e., dim room illumination and no glare) for visit 1 versus the values from the follow-up visit. Inspection of this scatterplot indicates that retest values fall above or on the line with a slope equal to 1.0 which explains the small but significant learning effect illustrated in Figure 2a.

5. **Figure 3a** presents the Average CS test results (n = 13) with glare. The two means are not significantly different (t=1.16, p=0.13). **Figure 3b** presents CS test results (n = 13) with Vector Vision CSV-1000HGT (with glare) for visit 1 versus the values from the follow-up visit. The solid line has a slope of 1.0 and it can be seen that retest values lie above, on and below this line.

6. **Figure 4** plots log CS (without glare) as a function of logMAR acuity. This graph illustrates that although the logMAR acuity extends over a range of values, the log CS reveals no systematic change. If the patient's entire log CS function was shifted down then we might expect log CS to decrease as logMAR acuity decreases. However as this trend was not observed we can conclude that a drop in high spatial frequency sensitivity is not correlated with a drop in sensitivity to lower spatial frequencies.

Figure 1a: Average results for LogMAR VA

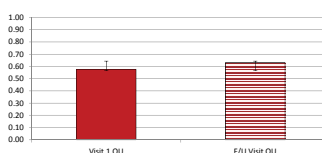


Figure 1b: LogMAR Visual Acuity OU

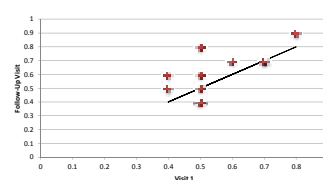


Figure 2a: Average values for CS test results with Vector Vision CSV-1000 LV

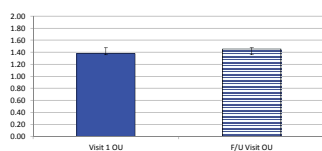


Figure 2b: CS test results with Vector Vision CSV-1000LV (no glare)

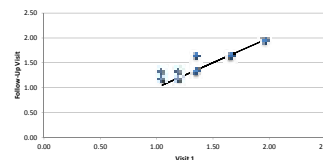


Figure 3a: Average values for CS test results with Vector Vision CSV- HGT

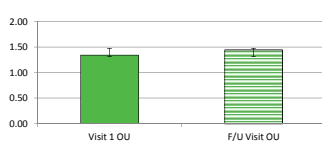


Figure 3b: CS test results with Vector Vision CSV-1000HGT(with Glare)

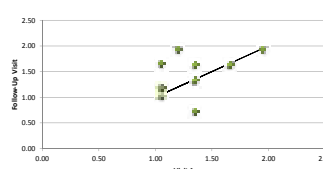
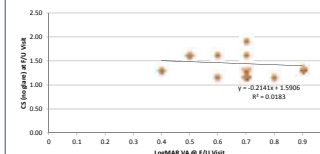


Figure 4: Vectore Vision CS test results with CSV-1000LV versus LogMAR VA



### VECTOR VISION CSV-1000LV

- Designed for CS testing in patients who have visual acuity worse than 20/70.



### VECTOR VISION CSV-1000HGT

- Evaluation of CS in the presence of glare (calibrated to simulate two on-coming halogen car headlights as seen from 150 feet).



7. ICC values were computed to examine test-retest reliability. The ICC value calculated for CS measured without glare was significantly different from zero, indicating very good to excellent reliability (ICC = 0.864, p<0.001).

8. The ICC value calculated for CS measured with glare was also significantly different from zero, although its absolute value was not as high as that observed without glare (ICC = 0.455, p = .048).

## CONCLUSIONS

- CS in patients with albinism, as tested with Vector Vision CSV-1000LV shows a slight but significant practice effect even with the use of two charts and a 6 month test-retest interval.

- The significant ICC value of 0.87 for the CS test without glare indicates that the test-retest reliability is very high and thus acceptable for clinical and research use.

- It is possible that the practice effect could be entirely eliminated if subjects were given a "practice" test first with the second administration's results recorded as the Visit 1 result.

- CS obtained with the CSV-1000HGT does not show a practice effect. Test-retest reliability is good (ICC coefficient = 0.5) but not as high as that observed for the without glare condition.

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# Sustained benefits of Therapeutic Tinted Contact Lenses (CL) in patients with Albinism

#2778

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Illinois College of Optometry, Chicago, IL

## PURPOSE

To determine whether the improvement in visual function and nystagmus eye movements observed in patients with Albinism using tinted CL correction is sustained after 6 months following the initial dispense.

Results on 20 subjects after the initial fit and dispense of the tinted CL, were presented last year (Saeed & Schlange, 2011; 2012). Six month F/U results on 13 subjects that returned to clinic are now being reported. 1 subject discontinued CL wear due to an allergic reaction to the CL material. 2 subjects discontinued CL wear after losing a CL. An additional 2 subjects discontinued CL wear due to a torn lens. The remaining 2 patients were lost to F/U (change of phone number or address).

Figure 1: Albinism patient with tinted CL OS

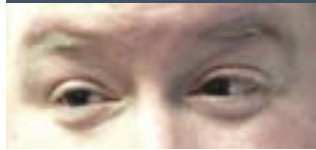


Figure 2: Albinism patient with un-tinted CL



Figure 3: Albinism patient with Tinted CL



## METHODS

20 subjects were initially fitted with soft toric CLs that were then custom tinted to create an artificial iris. In 13 subjects returning for a 6 month FU visit, dependent variables including visual acuity (VA), contrast sensitivity (CS) with and without glare, and nystagmus eye movement (NEM) recordings, were tested while wearing tinted CLs. The EDTRS chart was used to measure visual acuity. The CSV-1000HGT (halogen glare test) and 1000E (Contrast Sensitivity chart) were used to measure the contrast sensitivity function and effect of glare. Nystagmus eye movement characteristics of intensity and foveation were recorded and analyzed with an ISCAN system (RK 826PCI) that uses a video based dark pupil-to-cornea reflection method.

## PROCEDURES

- Prescribing therapeutic tinted Contact Lenses:** After obtained informed consent, toric or spherical, annual replacement soft lenses were fit and custom tinted with an opaque tint and clear pupil to fully mask the iris transillumination defects (ITD's). The pupil diameter for the CL was defined approximately 0.5mm larger than the patient's pupil size measured in dim illumination, to avoid tunnel vision.
- Measuring Contrast Sensitivity Function** CSV-1000LV-V and CSV-1000 LV-C contrast sensitivity charts were used to provide an appropriate target size for our patient population.
- Measuring the effect of glare (Halogen Glare Test)** CSV-1000HGT was used with the CSV-1000 LV test face for the evaluation of glare disability. In medium setting, the unit is precisely calibrated to simulate two on-oncoming halogen car headlights as seen at night from 150 feet.
- Recording Eye Movements** Eye movements were recorded with ISCAN (RK 826PCI), a high speed video based dark pupil-to-cornea reflection method using a remote (ETL-300) camera recording horizontal and vertical eye movements simultaneously at 120 or 240 Hz. The patient viewed at 3 meters, 5 wall mounted targets separated by 15 degrees and at appropriate size for patient's acuity. Three ISCAN runs (5 sec each) in each of the 5 positions (primary, up, right, down, left) were recorded when viewing targets with glasses, un-tinted and tinted contact. Nystagmus waveform amplitude and frequency were recorded for each run and with each optical device. Results from the dominant eye (primary position) used in this presentation were analyzed in MATLAB using OMLAB software (www.omlab.org).

## RESULTS

- The improved logMar VA observed with tinted CLs was still present at the 6 month FU visit, as the difference obtained at the two visits did not differ significantly ( $t = 1.72, p = 0.11$ ).
- Table 1a** presents LogMAR visual acuity for individual subjects with tinted CL at the initial dispense and at the 6 month FU visit. In 5 of 13 subjects, visual acuity was maintained with tinted CL wear at F/U visit. In 2 subjects, visual acuity was further improved with tinted CL at FU visit. In 6 subjects, visual acuity was slightly decreased at the FU visit, but was no worse than previously measured VA with un-tinted CL.
- ISCAN recordings with 3 runs in each of 5 gaze positions showed that the reduced intensity of nystagmus was also maintained with tinted CL wear after 6 months ( $t = 0.61, p = 0.557$ ).
- Table 2a** presents mean intensity of Nystagmus waveforms for each subject's dominant eye in primary gaze. In 6 of 13 subjects, nystagmus intensity further reduced at the FU visit with tinted CL wear. In 7 of 13 subjects, nystagmus intensity increased at the FU visit. Five of these 7 subjects (JC, PM, RG, MM and LM) presented with faded tinting of CL due to improper storage in no-rub CL solution. The average results for the group ( $N = 13$ ) are presented in Table 2b.
- Mean CS under normal testing conditions was further improved at the 6 month follow up visit ( $t = 2.49, p = 0.028$ ).
- Table 3a** presents Contrast sensitivity measurements, under normal testing conditions. Contrast sensitivity was maintained in 8 subjects and slightly improved in 5 subjects. Table 3b shows the average results for the group.
- Mean CS with glare did not differ between the two visits ( $t = 1.17, p = 0.266$ ).
- Table 4a** presents Contrast sensitivity measurements with glare. Contrast sensitivity function with tinted CL was maintained in 7 of 13 subjects, showed further improvement in 5 of 13 subjects, and deteriorated in 1 subject. Table 4b presents the average values for the group. Table 4b shows the average results for the group.
- The subjective evaluation of Sensitivity to light, glare reduction, clarity of vision and comfort of vision are shown in **Tables 5, 6, 7 and 8** respectively. The subjective improvement in visual function previously reported by patients with tinted CL wear continued to be reported at the 6 months FU visit.

Table 1 - 4



Table 5, 6, 7 and 8



## CONCLUSION

We previously reported that VA, Nystagmus intensity and CS with and without glare were significantly improved with tinted CL wear in patients with albinism. The current study indicates that these significant improvements are maintained or even increased for at least six months following initial dispensing of tinted CLs.

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- Saeed F, Schlange D. Effectiveness of Therapeutic Tinted Contact Lenses (CL) in Patients with Albinism. Poster session presented at the annual conference of the Association of Research in Vision and Ophthalmology (ARVO); 2012 May; Fort Lauderdale, FL.
- Saeed F, Schlange D. Factors Influencing Improvement in LogMAR Visual Acuity in Patients with Albinism. Poster session presented at the annual conference of the American Academy of Optometry (AAO); 2012 October; Phoenix, AZ.

## ACKNOWLEDGEMENT

We would like to thank Dr. Susan Kelly for her help with statistical analysis of this data.

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# Is Peripheral Retinal Ischemia Associated with Systemic Atherosclerosis?

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#194

## INTRODUCTION

Diabetes and carotid intima media thickness (IMT) are both well-known risk factors for cardiovascular disease and atherosclerosis. The Framingham Study showed a nearly 15x increase risk in cardiovascular events in middle-age patients with diabetic retinopathy (DR). Technological advances in recent years have produced smaller, portable ultrasound devices making screening of IMT in the carotid arteries easier and more accessible. Fluorescein angiography technology has evolved, providing up to 200 degree fundoscopic views in a single, high-resolution image. These images have allowed improved visualization of the extent of peripheral retinal nonperfusion. Previous studies have found that diabetic patients presenting with retinopathy have a higher IMT than diabetic patients without retinopathy and patients with severe DR are more likely to have subclinical vascular disease.

The purpose of this study was to investigate whether there was a correlation between peripheral retinal nonperfusion and IMT thickness of the common carotid artery in patients with DR. A positive correlation might suggest a significant relationship between microangiopathy in the retina and systemic macroangiopathy. This association might allow for more diverse and non-invasive means to screen for retinal nonperfusion. Conversely, identification of peripheral retinal nonperfusion may warrant further cardiovascular evaluation in diabetic patients.

## METHODS

In this cross-sectional pilot study, 15 subjects with diabetes and advanced DR were enrolled. No subject had PRP treatment. Ultrasound images of both common carotid arteries were taken using the HeartSmart IMTplus<sup>®</sup> portable ultrasound device (Figure 1). The scans were electronically transferred and read by an off-site reading facility. A grade of IMT was assigned by the reading facility based on comparison to a database of over 40,000 patients mapped and categorized based on age, gender and race. A grade of normal represents the mean IMT for age and gender, and mild, moderate and severe categories represent 2, 4, and 8 standard deviations above the mean, respectively. Ultra-

widefield fluorescein angiography (UWFFA) images on both eyes were obtained using Optos C200 scanning laser ophthalmoscope (Figure 2). The same certified retinal photographer obtained all images. Using area measurement function in the V2 Vantage Review Software (Optos, PLC), one of the coauthors determined both the total area of gradable fundus and area of nonperfusion seen in the arteriovenous phase of the UWFFA. Ischemic index (ISI) was calculated by dividing the nonperfused retinal area with the total retinal area. An ISI of 0 indicates a fully perfused retina while an ISI of 1.0 indicates complete absence of retinal capillary perfusion. One-way ANOVA was performed to test if there was a difference in ISI among normal, mild, moderate, and severe IMT groups both in the right and left side.

Figure 1: Intima-Media Thickness Report

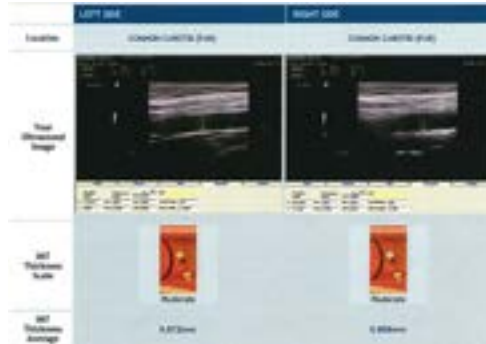


Figure 2: Ultra-Widefield Fluorescein Angiogram. The white border outlines the extent of gradable fundus and the blue border delineates the extent of peripheral nonperfusion. The ISI is 0.21.

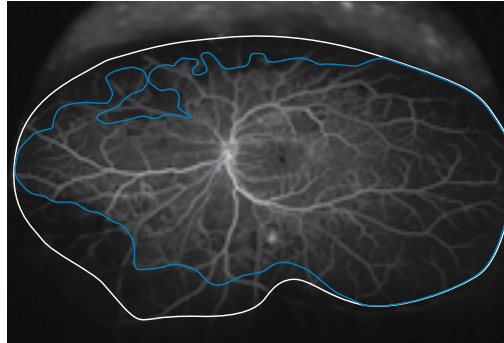


Table 1: Demographics

Age (yrs)	Race	Sex	Length of diagnosis (yrs)	LBG (mg/dl)	Smoking history	HTN
56	AA	M	5	200	Never	Yes
50	AA	M	12	107	Current	No
58	AA	F	16	160	Former	Yes
50	W	M	1	160	Former	Yes
44	H	M	unknown	160	Never	Yes
41	H	F	unknown	185	Never	No
66	AA	F	11	170	Never	No
46	H	F	14	120	Never	No
26	H	M	12	120	Never	No
71	C	M	22	89	Former	Yes
63	AA	M	4	144	Former	Yes
78	AA	F	21	118	Never	Yes
49	C	M	14	160	Never	Yes
77	AA	F	unknown	127	Current	Yes
58	AA	M	unknown	72	Never	Yes

## RESULTS

Compiled demographic data from the 15 subjects is presented in Table 1. Average ISI in the right eye was 0.145 (± 0.134) ranged from 0.01 to 0.40, and 0.144 (± 0.144) in the left eye ranged from 0.01 to 0.47. ISI values classified by IMT group are presented in Table 2. No subjects were classified as severe. No statistically significant difference was identified in right eye ISI among normal, mild, and moderate IMT groups in the right carotid ( $F_{2,12} = 1.31, P = 0.31$ ). However, there was significant difference in left eye ISI among all IMT groups in the left carotid ( $F_{2,12} = 7.63, P = 0.007$ ). Post hoc analysis reveals the difference in left eye ISI was between the normal and mild IMT groups ( $P = 0.006$ ).

Table 2: ISI values for IMT grade

	Normal	Mild	Moderate
OD	0.0267	0.2291	0.0512
	0.2276	0.2708	0.0948
	0.0059	0.0253	0.0184
	0.0099	0.3636	0.2352
		0.1595	0.3985
			0.0620
n	4	5	6
Mean	0.0675	0.2097	0.1433
OS	0.0386	0.4664	0.1063
	0.1428	0.2979	0.0925
	0.0131	0.0126	0.1431
	0.0326	0.2829	0.1134
	0.0164	0.3692	
	0.0298		
n	6	5	4
Mean	0.0456	0.2858	0.1139

## CONCLUSION

- In this pilot study we looked for a correlation between peripheral retinal nonperfusion and IMT grade of the common carotid artery in patients with DR.
- We found no statistical significant difference in ISI among IMT groups on the right side.
- We found significant difference in ISI among normal and mild IMT on the left side.
- Our study is limited by a small sample size and a narrow range of ISI (0.01-0.47) and IMT grade (normal – moderate). Previous studies investigating ISI have found values from 0-1.0.
- Our study supports a common pathogenic mechanism for both macro and microangiopathy but the data set may be too narrow to identify significance between groups.
- Further investigation with a larger cohort may provide more accurate results.

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# Is Vernier Acuity More Sensitive Than Grating Acuity to Visual Impairment in Adolescents?

#2770

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## INTRODUCTION

Teller Acuity Cards grating acuity is one of the most widely used tests of visual acuity in children who cannot perform recognition acuity tests. However, grating acuities may miss amblyopia.

Vernier acuity, often referred to as a “hyperacuity”, may be more sensitive to amblyopia than grating acuity (Drover et al 2010)

Vernier acuity has a longer maturation course, and on several grounds is considered to be cortically mediated (Skoczenski & Good 2004)

## PURPOSE

To compare vernier acuity (VeA) and grating acuity (GA) tests to a recognition acuity (RA) test in adolescents with visual impairment, hearing loss, and cognitive disabilities.

## PARTICIPANTS

### STUDENTS FROM DEAFBLIND PROGRAM

-Perkins School

- N=16 eyes (10 Adolescents)
- Ages 14 - 21 (Median = 19)
- 16 of 20 eyes qualified

#### - Testing Criteria:

- 1) RA must be measureable
- 2) Visual acuity better than 20/600

Primary Ocular Diagnosis	Number
Chorioretinal Coloboma	6
Optic Nerve Abnormality	3
Cerebral Visual Impairment	2
No Known Visual dysfunction	1

Medical Diagnosis	Number
CHARGE association	8
Congenital CMV Infection	1
Hearing Loss (mild-severe)	10
Unknown	1

### NORMAL ADULTS

- students from New England College of Optometry

- N=24 Adults
- Ages 24 - 31 (Median = 25)
- 48 of 48 eyes qualified\*

#### - Testing Criteria:

- 1) BCVA 20/20 in each eye.
- 2) No known ocular pathology

## METHODS

### • 2 separate sessions

(adults retested in 1-2 wks & adolescents by 1 wk -2 mo)

• **RE and LE** were measured for all 3 tests in each session with counterbalanced order

• **Recognition acuity (RA):** Single letter or symbol with crowded bars with 50% separation. Displayed at 12 ft or 6 ft if VA reduced.

### • Grating acuity (GA)

- Teller II Acuity Cards (Setero Optical Co.)

- Tested at 55cm

- 15 cards with gratings

- 2.0–0.10 logMAR range (Step size: 0.15 logMAR)

### • Vernier acuity (VeA) (Drover et al 2010)

- Tested at 55 cm

- 12 Cards: 6 pointed star or flower

- Misalignment ranges: 1.50 – 0.35 logMAR (Step size: 0.16 logMAR)



## RESULTS

### • Mean Acuities and SD (in LogMAR)

#### 1) Adults

Tests	N	Mean	SD
RA	24	-0.11	0.05
GA	24	-0.09	0.05
VeA	24	-0.24	0.15

( RA: 20/15; GA: 20/16; VeA:20/12)

#### 2) Adolescents

Tests	N	Mean	SD
RA	16	0.72	0.33
GA	16	0.70	0.30
VeA	16	0.65	0.41

( RA: 20/105; GA: 20/100; VeA:20/90)

### • Test-Retest Differences

- Using mean difference (S1-S2), SD, and Paired-t test

#### 1) Adults (N=24) RA\*

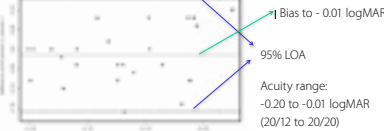
- both sessions RE

Mean Diff	SD	p value
-0.02	0.07	0.29

\* Most adults detected the highest spatial frequencies in both VeA and GA tests. Only RA is analyzed.

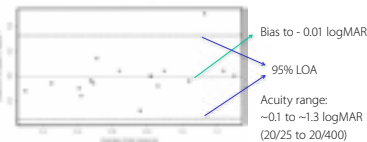
### • Test Retest Reliability for Adults –RE only (Bland-Altman Method)

- RA (jittered): Total 95 % LOA ≈ 0.25 log MAR (2.5 lines)

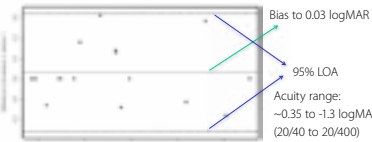


### • Test Retest Reliability for Adolescents (Bland-Altman Method)

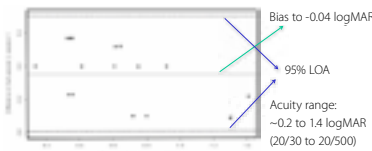
- RA: Total 95 % LOA ≈ 0.66 log MAR (~7 lines)



GA (jittered): Total 95% LOA ≈ 0.56 logMAR (~6 lines)



- VeA (jittered): Total 95% LOA ≈ 0.69 logMAR (~7 lines)



### • Comparison of VeA, RA, and GA normalized value (ΔN) in adolescents\*\*

N=16 eyes

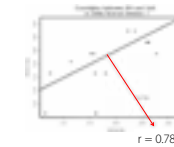
Session 1

Tests	N	ΔN Mean	SD
RA	16	-0.79	0.34
GA	16	-0.76	0.31
VeA	16	-0.89	0.44

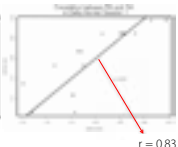
\*\* Adolescents' raw data were normalized taking the difference from the normal value for age (ΔN):  
 - RA: -0.073 logMAR (Ah-Kine et al AAO poster)  
 - GA: -0.06 logMAR (Drover et al 2009)  
 - VeA: -0.24 logMAR (Drover et al 2010)

### • Correlation between VeA and GA to RA using ΔN values in Adolescents

- RA and VeA



- RA and GA



• Detection of shapes was poorer than detection of vernier misalignment: For the 6 adolescents able to identify shapes, median difference was 2 cards (steps). For adults, median difference was 1 card.

## CONCLUSIONS

Test retest reliabilities are similar for RA, VeA, and GA tests in a small sample of adolescents with visual impairment (VI) and other disabilities.

Normalized VeA and GA acuities have a similar relationship to normalized RA and are not significantly different on average.

Therefore, VeA may not be a better predictor of RA than GA in adolescents with VI due to ocular and neurological causes.

VeA may be more sensitive to other visual deficits, specifically amblyopia, than GA (Drover et al 2010).

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ICO

# BCLA

## 3 ICO PRESENTATIONS

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# Disease severity in a keratoconic population within the Illinois Eye Institute

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## PURPOSE

The purpose of his study was to evaluate disease severity in a population of keratoconic patients utilizing corneal gas permeable lenses within the Illinois Eye Institute.

## METHOD

With the approval of the internal review board, a query of our contact lens ordering system was performed searching for all patient for whom any keratoconic lens design had been ordered within the last ten years. All large diameter, scleral, hybrid, and piggyback wearers were excluded. Patients were further excluded if they suffered from other significant ocular disease including glaucoma, corneal transplant, amblyopia, and degenerative myopia. Finally, the charts of 331 patients were reviewed for data related to their keratoconus.

## THE DATA COLLECTED INCLUDED

Topographical data: apical, flat, steep and average K readings, kornea area  
Pachymetry and Aberrations (when available)  
Visual acuity: spectacle and CL  
Presence of scarring  
Age, race, gender  
Lens design and curvature

## RESULTS

Patients were predominantly female, 52.4 percent. The average age was 31.25 ± 11.6 years. Multiple races were represented.

**Figure 1.** Shows the racial distribution within our keratoconus population.

As seen in **Figure 2**, this differs from that of CLEK which has a seemingly opposite racial distribution. However, no significant difference was noted between races for any of the keratometric readings,  $P > 0.05$ .

The average of the mean K readings was 51.84 ± 6.60. The flat K readings ranged from 33.50D to 85.75D with an average of 49.19 ± 6.19. In our population the average steep K was 54.49 ± 7.64 and ranged from 40.38 to 93.14D.

**Figure 3.** Represents a graphical view of the keratometric readings of our patient population.

Utilizing CLEK criteria of 45D for mild keratoconus, only 11.8% of our patients fell into the mild category. Additionally, 39.0% of our

patients were categorized as severe with average K readings greater than 52D. 11.5% of our keratoconus patients exceeded an average K of reading 60D.

**Figure 4** compares our korne population to that of the CLEK study. It demonstrates a mild shift towards more severe keratoconus.

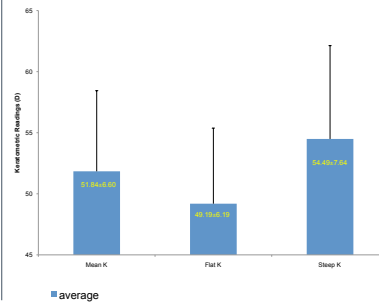
Our patients had average spectacle acuity of 20/70. However 67 percent were able to achieve 20/40 or better vision. Our mean contact lens acuity was 20/30 with patients most often achieving two lines of visual improvement. Ninety-one percent achieved 20/40 or better acuity.

Corneal scarring was only noted in 17 percent of patients.

Figure 2: Racial Percentages

Race	CLEK	IEI
Caucasian	68	9
African-American/Black	21	56
Hispanic	7	32
Asian	3	1
Indian	Not noted	2

Figure 3: IEI Average K Readings



## DISCUSSION AND CONCLUSIONS

Unlike CLEK this is a retrospective study to which enrollment bias does not apply. This may account for some of the differences. This study is limited to traditional, corneal gps and excludes more advanced designs. It does not include seemingly even more advanced cases necessitating transplantation, piggyback, sclerals or hybrids. This might suggest our population may be even more severe than indicated here.

Despite the seemingly steeper K readings within our population only 17% were noted to have scarring. This differs greatly with CLEK which reports 73% had scarring. This calls into question whether this observation was properly documented in our charts or do our patients actually exhibit less corneal signs but steeper corneal curvatures.

Our racial distribution is also different than that of CLEK. A review of our overall patient demographics may be indicated to determine if this finding is purely representative of our total patient base at the eye institute or whether selection bias in CLEK could have occurred.

With its over 1500 patients and greater than 30 publications the Collaborative Evaluation of Keratoconus (CLEK) has become the standard by which we evaluate keratoconus. The utilization of CLEK criteria seems to indicate that our patients frequently experience severe disease, 39% of the time. For our patient population we would suggest shifting these categories each up by 2D. Suggesting less than 47 as mild and greater than 54 as severe. This would result in 24% being categorized as mild compared to only 11.8 percent with the 45D criteria. Similarly using 54D would reduce the number of patients categorized as severe to 29.7 percent.

## PRIMARY REFERENCE

Zadnik et al for CLEK. Biomicroscopic signs and disease severity in keratoconus. Cornea 1996; 15(2): 139-146.

## DISCLOSURES

This study was funded by an unrestricted grant from Metro Optics.

## ACKNOWLEDGEMENTS

Ms. Lauren Hunt for her contribution to data collection and analysis.

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# Corneal Hydrops: Prevalence, Presentation, and Recurrence

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## PURPOSE

Keratoconus may be complicated by corneal hydrops. The purpose of this study was to determine the presentation, prevalence, and recurrence of corneal hydrops in the keratoconus population of the Illinois Eye Institute from 2007 to 2011.

## BACKGROUND

Advanced keratoconus patients may develop acute hydrops. Acute hydrops are breaks in Descemet's membrane that result in corneal edema.

**Figure 1.** Marked corneal edema and bullae as viewed with slit lamp biomicroscopy in a patient with hydrops

**Figure 2.** Visante evaluation showing a large break in Descemet's with edema and bullae formation

Following the break, aqueous humor leaks into the corneal stroma to cause edema and loss of corneal transparency. Clinical findings vary and include circumscribed, microcystic, or bullous edema and anterior chamber reaction.

**Figures 3-5.** Demonstrate the marked change in appearance in the cornea during corneal hydrops.

Patients with corneal hydrops may present with a variety of symptoms such as photophobia, pain, blur, foreign-body sensation, tearing, and redness.

## METHOD

This study was approved by the internal review board. A query of our patient management system was performed, looking for diagnosis codes for keratoconus, irregular astigmatism and corneal ectasia. The charts were reviewed for a definitive diagnosis of keratoconus. The final list contained 446 patients. From these charts, data collected included: race, gender, presence of allergies, and the number of hydrops episodes. If hydrops was present, detailed data regarding date of presentation, presenting symptoms, time to resolution, break location, and outcome were collected. Descriptive statistics and t-tests were performed to identify significant results.

Figure 1: Hydrops



Figure 2: Hydrops



Figure 3-5: Hydrops

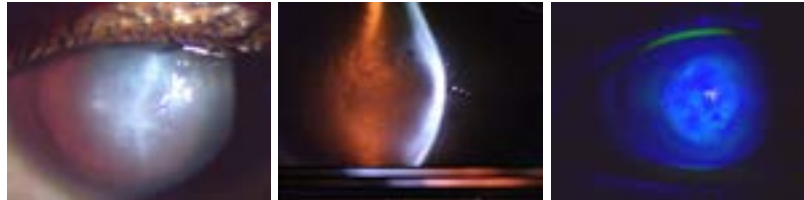


Figure 6: Racial Distribution of Hydrops Events

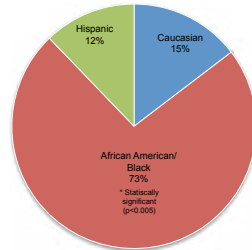
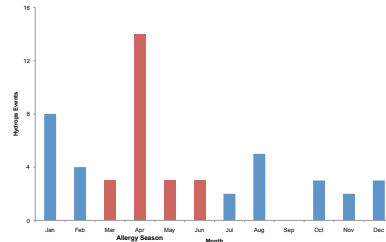


Figure 7: Hydrops Events per Month



## RESULTS

Corneal hydrops occurred in 41 of 446 patients for a prevalence of 9.2 percent. Within the hydrops population, 23 patients were male and 18 patients were female with an average age of 35.53 years. There was no statistical difference between the overall keratoconic population and the hydrops population with regard to gender.

**Figure 6.** Racial distribution of patients with corneal hydrops.

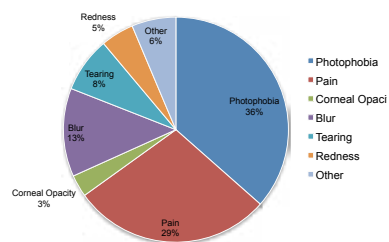
Of the 41 patients, 30 were African American, 6 were Caucasian, and 5 were Hispanic. This distribution was compared to that of our overall keratoconic population and was statistically significant for African-American race,  $p < 0.005$ .

22 percent of the hydrops population experienced a second event. 7 patients suffered an event in the contralateral eye and 2 events were recurrences in the same eye.

Patients with hydrops were noted to have a significantly higher prevalence of atopy ( $p < 0.005$ ). Almost half of the hydrops events (23/50) occurred during Chicago's peak pollen season from March to June. Also, hydrops events were increased in times of stress, with 11 of 50 events occurring during holiday season in December and January

**Figure 7.** Represents this bimodal distribution with a trend in distribution toward pollen season,  $p = 0.07$ .

Figure 8: Presenting Symptoms



Visual acuity at presentation was highly variable ranging from 20/25 to light perception.

Photophobia was the most common presenting symptom, followed by pain and blurred vision.

**Figure 8.** a graphical representation of patients' chief complaints at the onset of hydrops

Contact lens re-fit to steeper a base curve was the most common outcome following hydrops. Despite corneal scarring these patients had an average visual acuity of 20/60. Penetrating keratoplasty was the second most common outcome following hydrops with 10 of 41 patients requiring the procedure. The remainder of patients were refit into a new lens modality which also provided significantly improved vision.

## CONCLUSION

This study confirms a hydrops prevalence of less than ten percent. Patients undergoing hydrops statistically report greater atopy than those who do not experience hydrops. This correlation is further supported by the increased presentation during pollen season.

According to our results, the clinical picture of a keratoconic patient experiencing acute hydrops is a 35 year old, African American of either gender, with a history of atopy.

Recurrences are not uncommon, occurring at a rate of 22 percent of hydrops patients. Therefore patients should be educated on the risk of recurrence and reassured that majority of patients are able to successfully return to contact lens wear with less than one fourth requiring corneal transplantation.

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# Utilizing the RoseK2IC lens as a miniscleral in patients with small corneas

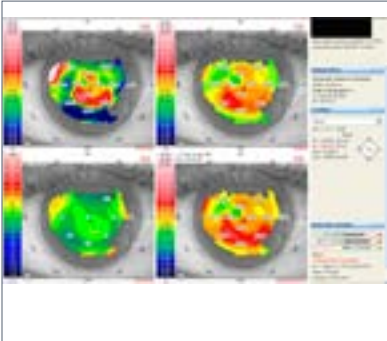
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## BACKGROUND

Patients with microcornea and microphthalmia often have significant refractive error and other ocular co-morbidities that lend themselves to correction with contact lenses. However, lens options have often been limited. Small diameter soft lenses can be utilized but in the US many of these designs are now custom and handling can be difficult for some patients. Corneal GPs are another option but lenses are quite small and some patients with high refractive errors may struggle with lens loss and handling. Recently the trend toward more oxygen permeable GP materials with larger buttons has allowed the resurgence of scleral lens fitting. However many of the traditional scleral lens designs begin at 14mm which can be quite large for these small eyed patients to successfully apply. Employed here is a standard intralimbal design for which the fitting set is available in 11.2mm. Three patients are presented here with aphakia and corneal diameters of less than 10.25mm who were successfully fitting using the intralimbal design, RoseK2IC, as a miniscleral.

Figure 1: Topography of L eye



## METHODS

**Patient 1**, a thirty-one year old African American male with a blind phthisical right eye and an aphakic left eye with a shunt was referred by the department of rehabilitative services for contact lenses. His topographies of the left eye showed over 4D irregular astigmatism. His corneal diameter was only 9.35mm.

We did not have a fitting set with corneal lenses small enough to get within the HVID and the patient had limited dexterity which was concerning for a traditional scleral lens. Adding 2-3 mm to the corneal diameter suggested the need for a diameter of 11.5-12.5 mm to provide appropriate limbal clearance and landing on the conjunctiva. A diagnostic lens of 11.2 with a base curve of 7.34 was applied but did not land far enough out on the conjunctiva. So, a 12.0mm RoseK 2IC with a slightly steeper base curve was ordered for use as a semi-scleral. The lens provided full corneal coverage, central clearance that was approximately 1/3 the thickness of the cornea suggesting approximately 200 microns of clearance, the limbus was cleared and the conjunctiva was unaltered by the lens. The patient was able to successfully apply and remove the lens using a large DMV suction up. His vision improved 3 lines on Feinbloom.

Figure 2: Overall view of scleral lens on L eye



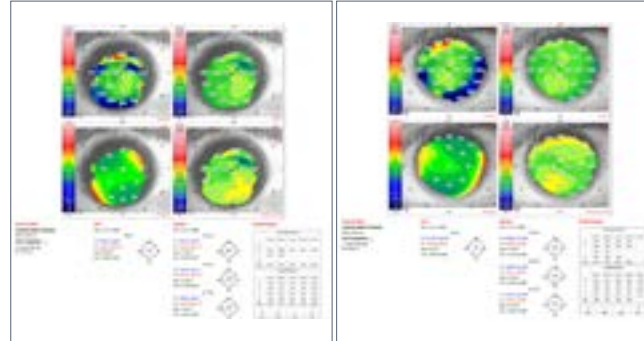
Figure 3: Optic Section of lens on L eye



The **second patient** is a 22 year old Hispanic female who suffered from microphthalmos, nystagmus, and bilateral aphakia secondary to congenital cataract. She had been wearing back toric lenses for several years and noticed that she was having more trouble with comfort and stability especially now that she was taking classes in a standard college lecture hall. The patient was originally fit with an 8.5mm diameter bitoric in both eyes. The right eye was fit with a base curve of 8.30/7.80 and a power of +20.75DS and the left eye was fit with a base curve of 8.50/7.95 and a power of +21.00DS. Her sim Ks were 38.32/42.34 in the right eye with an HVID of 9.83 and the left eye was 41.20/43.19 with HVID 9.67.

Refraction of the right eye was +14.75 -1.00 x 005 providing 20/60 VA and refraction of left eye was +15.75 DS providing VA of 20/400. The right eye was fit with an 11.5mm diameter RoseK2IC lens with a base curve of 7.94 with steep peripheral curves and a power of +21.12DS. The left eye was fit with an 11.5mm diameter RoseK2IC lens with a base curve of 8.04 with steep peripheral curves and a power of +22.87 DS. The lab adjusted the base curve and power to account for changes in the periphery accordingly. Visual acuity improved to 20/50+ and 20/300. Patient reported she had no difficulty with the lens moving around or coming off of the eye.

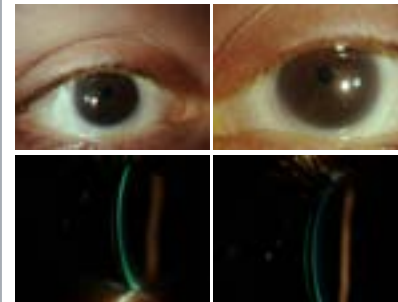
Figure 4 - 5: Topographies of the R and L eye



The **third patient** was a ten year old unilateral pediatric aphake who had failed with multiple lens designs. He was emmetropic in his other eye. He was originally fit with a Silsoft lens in the right eye at the age of 2 but the parents struggled with application. He was then tried in a corneal GP which they also could not get in and he rubbed his eye more with it. The parents eventually gave up and he returned at age 9. They continued to struggle with putting on the small diameter custom siHy lens and the child would not try. So when he returned at age 10, the child decided he wanted to try to do it himself. Unfortunately, he could not hold his eye open wide enough to apply it. When a corneal GP was attempted he screamed and kicked. Therefore, a decision was made to attempt a larger GP that would act like a scleral.

The patient was ordered an empirical 12.0mm RoseK2IC lens as a miniscleral with a 7.4mm base curve and a power of +21.00DS. Evaluation of the fit revealed minimal clearance. Visual acuity was stable but patient reported things were clearer. Over-retinoscopy was +0.50. So a lens with a steeper base was ordered and the patient was trained to to apply and remove the lens himself. Photo attempts were unsuccessful as patient was tired and did not want to sit still. Finally he was referred to our binocular vision service for amblyopia therapy. He now wears his lens every day.

Figure 6 - 9: Views of the contact lens with overall diffuse light demonstrate full coverage of the cornea and landing on the sclera with no impingement. The optic section shows even central clearance.



## CONCLUSIONS

This series of cases demonstrates that patients with small corneas can be provided with improved, vision, comfort and function by utilizing miniscleral fitting techniques and commercially available intralimbal GP lenses. Lenses can be ordered empirically or utilizing a fitting set with good success. Additionally, the customizable peripheral options allow adjustments in the scleral portion as well.

**Disclosures:** Dr. Reeder has been a paid speaker for Blanchard Labs.

## CONTACT INFORMATION

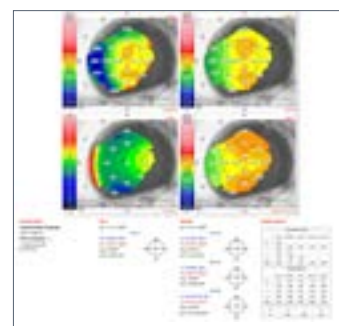
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Figure 10: Topography of the R eye. Showing the HVID was 10mm with simK readings of 43.98/46.15.





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## 2 ICO PRESENTATIONS

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# ICO

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# Sveinsson's Chorioretinal Atrophy

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## BACKGROUND

In 1939, an Icelandic ophthalmologist, Kristjan Sveinsson, was the first to describe Sveinsson's Chorioretinal Atrophy (SCRA), a very rare, bilateral, inherited disorder originally termed chorioiditis areata and later referred to as helicoid peripapillary choroidal atrophy. It has been described as helicoid chorioretinal atrophy, but this is a general term referring to a broad range of retinal disorders, including degenerative and genetic disorders. In order to avoid confusion with similar looking clinical conditions, the term Sveinsson's chorioretinal atrophy is now the standard terminology. SCRA is often misdiagnosed due to its rare nature and similar fundusoscopic appearance to a number of other common disorders. SCRA is slowly progressive, eventually affecting visual acuity, visual field and often electrodiagnostic testing.

## GENOMICS

SCRA is an autosomal dominant condition with no gender predilection in the pedigrees studied and apparently complete penetrance. Exhaustive genetic linkage analysis eventually led genealogists to markers on chromosome 11p15. Fine mapping and screening of the candidate genes revealed a TEAD 1 protein mutation to be the causative allele in SCRA, with a T-to-C transition for base pair 94 of that gene. This Y421H mutation results in the substitution of histidine for tyrosine. While this is not an active phosphorylation site, the C terminus is the connecting position to the protein's cofactor YAP65. The two interconnected proteins are active within cellular nuclei of retinal or choroidal cells, activating the RNA polymerase complex, and hence causing modifications of important structural genes. The co-presence of these two protein genotypes has been verified via the cDNA libraries of public databases within the pedigrees of known affected families by RT polymerase chain reaction testing.

## HISTORY, PATIENT 1

Our first patient is a 41 year-old white male. He initially presented with visual complaints of a blur, near greater than distance in both eyes for six months, and a floater which he could not localize. He had sinus/allergies and depression. He took oral Celebrex. He had an unremarkable family history.

## HISTORY, PATIENT 2

Our second patient is a 70 year-old female who is 50% African American and 50% Native American. She initially presented with visual complaints of a scotoma, and a floater. She could not localize the complaints to a specific eye. She had osteoporosis and a sinus/allergy, neither being treated. She had an unremarkable family history.

Exam Findings	Patient 1	Patient 2
Best Corrected Visual Acuity at Distance	20/20 OD, OS	20/30 OD, OS
Best Corrected Visual Acuity at Near	20/20 OD, OS	20/30 OD, OS
Pupils	Unremarkable	Unremarkable
CVF	Unremarkable	Unremarkable
EOM	Unremarkable	Unremarkable
Amsler Grid	Not Done	Unremarkable
Biomicroscopy	Posterior Blepharitis OU	1+ Nuclear Sclerosis OU
Goldmann tonometry	14 mm Hg maximum OU	14 mm Hg maximum OU
Posterior Pole	Peripapillary atrophy with RPE dystrophy OU (figure1a,b)	peripapillary atrophy OU (figure2a,b)
Humphrey Visual Field	Enlarged blind spot OU (figure3a,b)	Enlarged blind spot OU (figure4a,b)

## FUNDUS EXAM PATIENT 1

The OD dilated fundus exam (figure1a) revealed peripapillary atrophy with pseudopod-like extensions nasal and temporal. Retinal pigment epithelial dystrophy was evident temporal OD more so that nasal. In the OS dilated fundus exam eye there was peripapillary atrophy mostly superior with retinal pigment epithelial dystrophy mostly temporal. (Figure 1b). There was no evidence of subretinal neovascularization or chorio-retinal inflammation during the 2 year follow up of this patient. The areas of peripapillary atrophy remained stable during follow up.

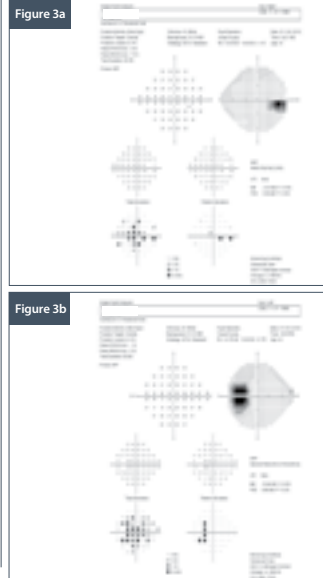
## FUNDUS EXAM PATIENT 2

The OD dilated fundus exam revealed peripapillary atrophy with pseudopod-like extensions outward form the optic nerve in both eyes. In OD there was a temporal extension toward the macula (figure 2a). In the OS there was an extension in the nasal direction (figure 2b). There was no evidence of retinal pigment epithelial dystrophy subretinal neovascularization or chorio-retinal inflammation or retinal pigment epithelial dystrophy during the 7 year follow up of this patient. The areas of peripapillary atrophy remained stable during follow up.



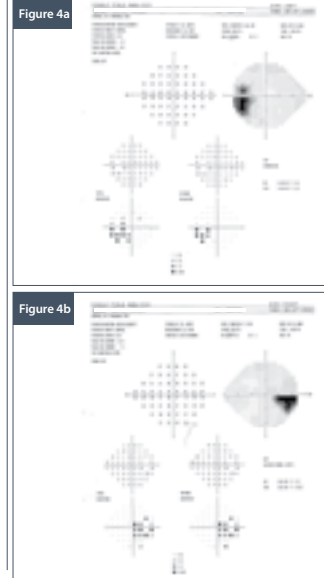
## HUMPHREY VISUAL FIELD PATIENT 1

Humphrey automated visual field examination was performed using a SITA 24-2 testing algorithm. This showed significantly enlarged blindspots OU with a greater extension of the blind spot toward fixation OD. This right eye defect resembled a centrocecal one in nature (figure 3a) and correlated with the temporal retinal extension of the peripapillary lesion in the right eye. The left eye showed a smaller enlargement of the blind spot with little extension toward the macular owing to the nasal extension of the peripapillary lesion in this eye (figure 3b).



## HUMPHREY VISUAL FIELD PATIENT 2

Humphrey SITA 24-2 Standard testing algorithm showed significantly enlarged blindspots OU with a greater extension of the blind spot toward fixation OD. This right eye defect resembled a centrocecal one in nature (figure 4a) and correlated with the temporal retinal extension of the peripapillary lesion in the right eye. The left eye showed a smaller enlargement of the blind spot with little extension toward the macular owing to the nasal extension of the peripapillary lesion in this eye (figure 4b).



## CONCLUSIONS

Recent understanding of the electrophysiological and genetic characteristics of SCRA allow more accurate diagnosis to be made. Inherited in an autosomal dominant pattern, the responsible gene has been mapped to chromosome 11p15. SCRA presents a distinct clinical picture of structure and function. Onset of the fundus lesions is as early as birth and can be slowly progressive throughout life moving from the peripapillary area to the macula. Electrodiagnostic testing supports the clinical observation that there is no edema or inflammation and suggests that the atrophy begins deep at the level of the retinal pigment epithelium only later involving more superficial retina. Visual function can be seen to follows structural change in that visual field loss is consistent with the retina involved and visual acuity and color vision are typically not affected until much later in the course when the fundusoscopic lesions involves the macula. A better understanding of this disorder can aid in making accurate diagnosis.

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# Recurrent Blebitis in Monocular Patient

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## BACKGROUND

Complications of glaucoma filtering surgery may develop months to years post-operatively. With the introduction of antimetabolite usage in filtering surgery, risk of developing a late onset blebitis and bleb-associated endophthalmitis has increased from 0.2% pre-antimetabolite risk to 2.6% risk of blebitis and ~1.7% bleb-associated endophthalmitis<sup>1,2</sup>. Antimetabolites such as 5-fluorouracil and mitomycin C are used in glaucoma filtering surgery to reduce possibility of patient developing subconjunctival scarring which could cause bleb failure, but usage of antimetabolites can cause development of a thin, avascular bleb wall increasing the risk of postsurgical complications<sup>3,4</sup>.

## CASE

A 60-year-old African American male presented to Urgent Care Service with a chief complaint of redness and cloudy vision OS of four days duration. He noted his left upper eyelid had become swollen and reported ocular discomfort of 8/10 on the left side. The patient's ocular history included enucleation OD secondary to end-stage glaucoma complications and severe-stage glaucoma status post glaucoma filtering surgery OS with a previous occurrence the year before of blebitis with positive Seidel sign.

Entering visual acuity was 20/80 OS (PH 20/60) and entering tests were within normal limits excluding 360° constriction on confrontation visual field OS. Slit lamp examination revealed upper lid edema, mild ptosis, diffuse severe conjunctival injection, and a thin elevated, avascular, cystic bleb located superiorly. The peri-bleb area was devoid of injection. Both the cystic bleb and anterior chamber were free of inflammatory cell and protein flare and there was no presence of hypopyon within the anterior chamber. Intraocular pressure was measured at 8 mm Hg with Tono-Pen

Avia™ and application of sodium fluorescein to the bleb site revealed a positive Seidel sign.

A presumptive diagnosis of recurrent late-onset blebitis status post glaucoma filtering surgery was made. A loading dose of gatifloxacin 0.5% (Zymarid®) (1gtt every 5 minutes for 15 minute duration) was given in office and prescribed tobramycin 0.03% and gatifloxacin 0.05% alternating every thirty minutes including overnight. In addition to topical agents, the patient was prescribed 400 mg oral moxifloxacin (Avelox®) qd for seven days.

The patient was followed daily until the bleb leak resolved (day 5). At that time, the topical antibiotics were tapered to QID and continued for nine days. Upon resolution of the blebitis, visual acuity returned to pre-inflammatory level of 20/40 and intraocular pressure had risen to 16 mm Hg.

The patient refused appointment for bleb revision surgery or treatment of bleb site with tissue adhesive, preferring to maintain maximum topical medication therapy and monitor for recurrent bleb leak. He has not experienced another bleb leak or blebitis episode for the last six months.

Figure 1



## DISCUSSION

Glaucoma filtration surgery is not commonly a first line treatment for reducing IOP levels. Surgical intervention is initiated once a patient's IOP levels are no longer properly controlled with available topical therapeutics and/or laser procedures, if a patient has demonstrated chronic compliance issues, if visual field defects or optic nerve damage progresses despite maximum medical therapy, or if hypersensitivity to medication side effects is present. The goal of glaucoma filtration surgery is to reduce IOP by creating an irrigation route from the anterior chamber to a subconjunctival reservoir.

Before the introduction of anti-metabolites during surgery, surgically created irrigation routes or "blebs" could scar shut and cause bleb failure. The use of 5-Fu postoperatively and MMC during filtration surgery has reduced the incidence of fibrotic scarring and bleb failure by reducing fibroblast activity during postsurgical healing<sup>5</sup>. MMC, in particular, disrupts the creation of fibrotic scarring by creating a zone of cell toxicity in which DNA, RNA, and protein synthesis is inhibited secondary to bifunctional alkylation<sup>6</sup>. With

use of anti-metabolites during glaucoma filtration surgery becoming standard, incidence of thin, avascular blebs has increased.

Affectivity of surgical blebs has increased in addition to the risk of developing a leaking bleb with antimetabolite use<sup>7,8</sup>. Leaking blebs put patients at significant risk for developing blebitis and/or endophthalmitis<sup>9,10,11</sup>. Blebitis can progress to potentially blinding endophthalmitis therefore it is important to treat aggressively with topical antibiotics. Blebitis is more likely to progress if the bleb is leaking, if the posterior lens capsule is open, or if infected with an antibiotic resistant strain of bacteria<sup>3</sup>. Culturing a bleb does not always yield accurate results and can take excessive time. Although there is not a preferred antibacterial agent for empirical treatment, third and fourth generation fluoroquinolones are recommended secondary to the broad antimicrobial spectrum and potential concentration within ocular tissues versus non-fluoroquinolones<sup>12</sup>.

Oral moxifloxacin can be of aid for prophylaxis of blebitis progression to endophthalmitis. It is an oral fourth generation fluoroquinolone and studies have

shown that oral moxifloxacin reaches higher levels of vitreal penetration versus oral gatifloxacin<sup>13</sup>. In addition, oral moxifloxacin has been found to exceed minimum inhibitory concentration of 90% of the most common causative bacteria in endophthalmitis. If the patient is at risk of progressing into endophthalmitis, oral moxifloxacin dosed at 400 mg qd x 7 days may be prophylactic. If endophthalmitis does develop, patients should be immediately referred to a retinal specialist for vitreal tap and intravitreal antibiotics.

## SUMMARY

Antimetabolite use in glaucoma filtering surgery has decreased the rate at which surgical blebs fail. Antimetabolites prevent formation of subconjunctival fibrosis by inhibiting fibroblast production. An unfortunate side effect of antimetabolite use is the development of a thin walled, avascular bleb. The thinness of the bleb wall increases the risk of leakage and can lead to transmigration of bacteria through the conjunctival wall, developing into blebitis and/or endophthalmitis.

If treated aggressively with broad-spectrum topical antibiotics, blebitis has a high likelihood of resolving to pre-inflammation visual acuity. Blebitis, if left untreated, may develop into a visually debilitating endophthalmitis, requiring intravitreal antibiotic treatment. Currently there is no agreed upon fluoroquinolone of choice for treatment and cultures are often unreliable leading to empirical treatment.

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# COVD

## 2 ICO PRESENTATIONS

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# Expansion of Vision Services Within the Illinois Eye Institute at Princeton Vision Clinic Beyond Primary Eyecare

Sandra S. Block, OD, M.Ed, FAAO, FCOVD, Medical Director School-Based Vision Clinics

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## BACKGROUND

The IEI at Princeton Clinic is a large school-based vision clinic servicing children from the Chicago Public Schools. The clinic opened in January 2011 and has provided primary eye care for more than 14,000 children in the first 30 months. The clinic is open year round. Third and fourth year students rotate through the clinic for one quarter providing comprehensive primary eye care.

During this period, many patients have required secondary services including referrals for eye health, contact lenses, vision therapy, strabismus, and visual information processing testing.

## METHODS

In the first 18 months, all patients who needed VT, fundus photography, visual information processing and oculomotor testing were referred over to the Illinois Eye Institute main campus. The follow through was minimal. In response to the needs of our patients, we attempted to identify solutions that would increase compliance of the recommended follow up.

### THE APPROACH HAD THREE PRONGS:

1. **Introduction of a staff optometrist to provide vision therapy, visual information processing and oculomotor testing;**

From July 2012 through June 2013, Dr. Kathleen O'Leary provided direct care Tuesday through Thursday afternoon. This included comprehensive eye care as well as vision therapy and visual processing assessment.

From July 2013, Dr. Adrianna Hempelmann has expanded service to Monday through Thursday afternoons.



Ms. Abbey Baus

2. **A ChicagoHealth Corp member (AmeriCorp) was added to our staff to improve the communication between the clinic and families. The term of the member is 9 months and they works closely with us, as well as the vision team within the Chicago Public Schools. The ChicagoHealth Corp members is expected to:**

Work along side the Illinois College of Optometry students, faculty and staff providing entrance testing and monitoring flow each morning.

Identify those patients requiring follow up care and contacting the families to work with them to schedule the needed care. If the care can be provided within the IEI at Princeton clinic, she works to get the appointment scheduled. If the referral is for care on the main campus, the member conferences the staff on campus with the parents to schedule the appointment. She then monitor whether the appointment is kept and does appropriate follow-up.



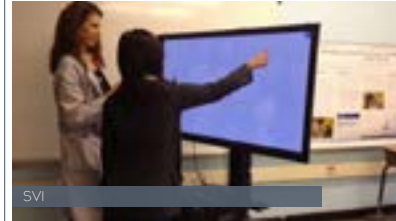
VTS-4

3. **Addition of a current vision therapy technology, a visagraph, an OCT, and a fundus camera.**

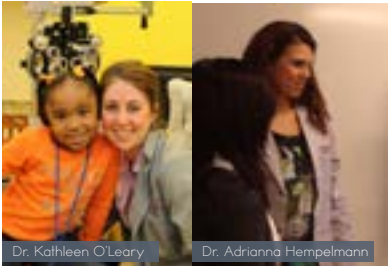
Electronic health records was implemented which allows us to take OCTs and photos on site and share them with staff at the IEI for assistance in diagnosis.

Through the generosity of a funder we were able to purchase a several video based programs that have been instrumental in engaging our patients during vision therapy.

The funder also made it possible to obtain a Visagraph for testing at our clinic.



SVI



Dr. Kathleen O'Leary

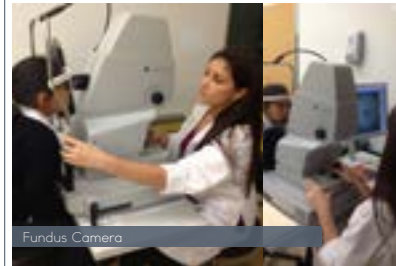
Dr. Adrianna Hempelmann



OCT



Visagraph



Fundus Camera

## RESULTS

Vision therapy was conducted three afternoons a week with 110 visits recorded in the 2012-2013 fiscal year. We have expanded this to four afternoons due to the demand and wait list. Visual information processing assessments have been completed on 25 children.

The OCT and fundus camera had been available since February and we had reduced our referrals for eye health from 106 to 67 during the 2012-2013 fiscal year.

## CONCLUSION

School-based clinics serve children who often do not access appropriate vision care services outside of the school setting. IEI at Princeton began with primary care services but quickly realized the need to expand services to include vision therapy, oculomotor assessments, visual information processing testing, and baseline fundus photos. We will continue to expand services to address the needs of our patients.

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# The Natural History of the Oculo-Visual Anomalies Associated with Traumatic Brain Injury (TBI): A Case Report

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## INTRODUCTION

The Center for Disease Control and Prevention reports that traumatic brain injury (TBI) occurs in 1.7 million individuals in the United States each year. The vision problems routinely reported for those with TBI include binocular vision, accommodative and oculomotor dysfunctions; reduced visual acuity, visual field loss and vision information processing anomalies, as well as oculo-vestibular, midline perceptual shift and attentional issues<sup>1,2,3,4,5</sup>. We know little, however, about the long-term natural history or natural course of the oculo-visual-neuro- anomalies associated with TBI. After an extensive PubMed/Google Scholar search, this case report appears to be the very first of its kind to appear in the literature.

## CASE REPORT CASE HISTORY

TB is now a 31 y/o W/F who has a history of TBI after falling out of a window at age 2. She has been evaluated 26 times over the past 29 years primarily by the first author. At 4 years of age, TB underwent a bilateral lateral muscle resection for an exotropia (07-1986). After surgery it was noted that she was an intermittent esotropie at distance and had a "flick" exotropia at near. Between 1988 and 1993 she also participated in 19 vision therapy sessions. TB's compliance was variable but did exhibit 2<sup>nd</sup> degree fusion and 3<sup>rd</sup> degree fusion towards the end of the therapy program. Visual acuities varied but stabilized after a while at approximately 20/40 OD/OS until the last visit where visual acuities appeared to decrease in the right eye. The case history indicates that she had a tendency to voice fewer complaints as she became an adult. This trend towards adults with intellectual disability and a psychiatric illness voicing fewer complaints has been documented by recently published research<sup>6,7</sup>. She is currently living somewhat independently with her mother and receives services during the day.

## THE EXAMINATION

The examination sequence varied and different tools were used over the years as advances were made in developing examination instrumentation and techniques. Visual acuities were taken using Teller Cards, HOVT, Lea symbols, Snellen and VEP<sup>8</sup>. Objective examination procedures were frequently utilized because of behaviors that would interfere with the standard subjective assessment techniques.

## VISION THERAPY

Post-surgical strabismus vision therapy for remaining binocular vision dysfunctions, oculomotor anomalies, amblyopia and vision information processing (VIP) anomalies was instituted. VIP problems diagnosed at 10 years 11 months of age included: visual discrimination, memory, spatial relations, form constancy, sequential memory, figure ground and closure. Vision therapy can be an effective treatment for those with TBI<sup>9,10,11,12</sup>. During therapy, TB did achieve 2<sup>nd</sup> and 3<sup>rd</sup> degree fusion. Lenses were prescribed to provide best VA and binocular vision.

## DISCUSSION

Research has noted that those adults with intellectual disability and a psychiatric illness tend to offer few complaints when taking a case history even though they are often on numerous medications and exhibit frequent visual and systemic anomalies. This acceptance of visual disabilities was the case for this patient as well. The longitudinal findings seen here suggest that those with TBI may demonstrate variable findings over their lifetimes and require close monitoring of these changes so that appropriate and timely intervention can be provided. It also appears that surgery for strabismus may be of limited intermediate and long term value<sup>2</sup>. Optometric vision therapy appears to have moderate success at least initially. Vision therapy may need to be re-instituted with this now adult patient to help her regain the 2<sup>nd</sup> degree fusion and stereopsis that TB demonstrated after the initial therapy program some years earlier<sup>4</sup>.

The numbers of medications increased significantly as additional diagnoses were added for various systemic and psychiatric disorders. Visual acuities fluctuated significantly, but usually were around 20/40 – 20/50. As a child, her refractive error showed a small amount of hyperopia and astigmatism which later developed into myopia. She also had a tendency to exhibit accommodative excess which could have played a role in the myopia seen as she became older. The oculomotor assessment post-surgical intervention for exotropia also varied from orthophoria to a constant esotropia with a mild upward gaze restriction and nystagmus. Stereopsis and 2<sup>nd</sup> degree fusion was seen after an active course of optometric vision therapy, but then was lost over the years. No major eye health problems were noted. The longitudinal study or those with TBI and the frequently encountered oculo-visual anomalies should be studied to ascertain the natural history and long term effects of various interventions over time. This case study is the first to report such findings. A key element of successful long-term care of patients with TBI must include the comprehensive diagnostic and therapeutic care provided by the behavioral/developmental/functional optometrist.

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Case History*							
Date	01-10/13	09-29-11	05-27-10	05-07-09	12-20-07	11-30-06	11-17-05
Chief Complaint	None	None	Concern for vision	Ocular health assessment	Hx of amblyopia	Needs Glasses	None
Date	09-23-04	08-21-03	10-23-93	11-18-92	10-20-92*	09-10-92	09-05-91
Chief Complaint	None	Itchy eyes	FU OM function Problems reading small print	Strabismus evaluation	Nystagmus	None	Post strabismus surgery exam
Date	08-05-91#	09-15-88	03-10-88	01-07-88	08-16-87*	07-31-86	
Chief Complaint	Afraid of the dark	VEP/Laser interferometry	Rx Follow up	Eye turns in less with Rx	Exotropia	BLRR surgery	

\*Information from vision therapy sessions not noted.  
# Exam at a children's hospital. This is a representative sample of, but not necessarily data from every visit.

Diagnoses*							
Date	2013-2010	2009-2007	2006-2003	2011-1992	1991		
Systemic	Allergies HAs	"	"	"	"		
Neurological	TBI, seizures Mild MR, Emotional/Behavioral Disorder, Speech/Lang Delays	"	"	"	"		
Oculo-visual	CRXT, Amblyopia, CMA, Intermittent Nystagmus	IXT, Amblyopia, CRXT	"	IXT, Esophoria CMA	IXT/IXT CHA		
Date	1991	1988					
Systemic	Allergies HAs	"	1986				
Neurological	TBI, seizures Mild MR, Emotional/Behavioral Disorder, Speech/Lang Delays CAET/IXT	"	TBI, seizures				
Oculo-visual	CAET/IXT		Ortho, Esophoria, OMD	Exotropia (bilateral lateral rectus resection)			
	CMA		CHA				

\*This is a representative sample of, but not necessarily complete data from every visit.

Medications *					
Date	01-2013	09-2011	09-2009	05-2009	06-2007
Medications	Ibuprofen, Potassium, Fenofibrate, Singular, Pepcid, Sertraline, Gabapentine, DetrolLa, Loratadine, Gabapentine, Carbamazepine ER, Prilosec, Tilia FE	Potassium, Singular, Pepcid, Sertraline, Gabapentine, Carbamazepine, Prilosec, Tilia FE, Prochlorperazine, Trilipix, Vesicare, Pseudoephedrine, Naproxen, Niacin, Vitamins	Potassium, Singular, Prilosec, Vesicare,	Lipitor, Zolof, Loratadine, Tilia FE	Potassium, Singular, Gabapentine, Tilia FE, Vesicare, Pseudoephedrine, Pepcid, Lipitor, Ibuprofen, Zolof, Estrostep, Flonase, DetrolLa
Date	11-2006	9-2004	8-2003	02-1987	07-1986
Medications	Singular, Gabapentine, Tilia FE, Vesicare, Pseudoephedrine, Pepcid, Lipitor, Ibuprofen, Zolof, Flonase, DetrolLa	Risperdal, Zolof, Gabapentine, Zolof, Claritin, Flonase, DetrolLa	Risperdal, Zolof, Gabapentine, Zolof, Claritin, Flonase, DetrolLa	Dilantin	Dilantin

\*This is a representative sample of, but not necessarily complete data from every visit.

Refractive Error *		
Date	OD	OS
01-10-2013	-1.75-50X070	-1.25-1.00X180
09-29-2011	-1.75	-2.50 (Stable for 4 years)
11-30-2006	-1.50	-1.75
11-17-2006	-1.25	-1.25
09-23-2004	-1.25	-1.25-50X180
07-03-1993	+2.25-25X180	+1.50-75X180
# 10-20-1992	+1.25-1.00X020	+1.75-1.25X140
09-05-1991	+1.00-1.00X180	+1.75-1.75X180
03-10-1988	+2.25-1.25X175	+2.00-1.25X178

\*This is a representative sample of, but not necessarily complete data from every visit  
# Exam at a children's hospital.

## CONTACT INFORMATION

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# Matching the Visual Needs of an Urban School District and Needs of an Optometric Educational Program

Sandra S. Block, OD, M Ed, FAAO, FCOVD • Valarie L. Conrad, OD, MPH, ARM • Melissa A. Suckow, OD, FAAO • Kathleen P. O'Leary, OD



## PURPOSE

The Illinois College of Optometry (ICO) in partnership with Chicago Public Schools (CPS) opened a school-based vision program to address the unmet need of vision care for their children serving as the Illinois Eye Institute at Princeton School.

The CPS system is one of the largest urban school systems in the United States serving 403,000 children housed in 681 schools.

CPS conducts vision screenings routinely on pre-kindergartners, 2nd grade students and 8th grade students.

Each year, over 100,000 children in the Chicago Public Schools (CPS) fail vision screenings, have broken/lost glasses, or fail to complete a required exam for entry to school.

Lack of follow up and limited access to providers accepting state vision insurance coverage contribute to poor access to eye care. In addition, the Illinois College of Optometry was interested in optometric clinicians working in community based settings.

## METHOD

The Illinois Eye Institute at Princeton School opened in Jan 2011 as a year round school-based vision clinic staffed by faculty, students, staff and opticians from Illinois College of Optometry.

The building is a decommissioned elementary school which is wheelchair accessible located within a 10 minute drive from ICO.

Building a new school-based vision clinic required financial planning. Most of the financial support comes from state insurance. It was also necessary to reach out for significant grant support.

CPS provides a liaison whose responsibility is to schedule schools to attend the clinic. Each day CPS is in session, one school is scheduled to bring up to 45 children to the clinic. In addition, the clinic is open to walk-ins. On days where school is not in session, the clinic serves only walk-ins. The school liaison and ICO have worked hard to market the program to the parents within CPS that have not followed through with recommendation for comprehensive eye care.

Our experience thus far is that the majority of the patients are brought to the clinic with their schools. The schools that are targeted are those that have identified their students as 90% or higher falling below the federal poverty level.

Every 3rd year optometry student rotates through the clinic for 1 session per week for 11 weeks and a number of 4th year students return for a second session.

## RESULTS

A review of the data shows that 74% of children seen need new eyeglasses. 80% of the children fall under the state health insurance. Glasses under this insurance take between 6-12 weeks for delivery.

Lions Clubs International Foundation (LCIF) provided 10,000 polycarbonate lenses to us which will allow the children to receive their glasses in a more timely manner. This pilot program from LCIF is one of four in the United States designed to address uncorrected refractive error.

The clinic has also diagnosed amblyopia, convergence insufficiency and strabismus in numbers that exceed that expected for the general population.

The recommendations for follow up of these problems show less than 20% of the children access appropriate services.

Many of the children provided care in the first two years has included recommendations for vision therapy. Vision therapy for patients with Medicaid is difficult to access. In the summer of 2012, IEI at Princeton added appointments for vision therapy in the afternoons, with a limited numbers of patients accessing this service.

In our first year of service, many children were referred for additional care but did not access the care. A Chicago Health Corp member (AmeriCorp) was appointed to address the need for follow up services. During the year, the member has provided personal contact with families and schools and has slight improvement in follow ups.

The optometric curriculum demands have created challenges on days when students have exams or classes. The clinic is open for walk-ins so it is challenging to know exactly how many patients will be seen.

In addition, the busses from the schools do not always arrive in a timely manner and completing the eye exams in time for the optometric clinicians to arrive on time for their afternoon assignments has provided some challenges.

In an effort to resolve these issues, preceptors may begin and finish exams and fourth year students are assigned to stay 30 minutes later on some days.

## CONCLUSIONS

**During our second year of service to the Chicago Public School students, we have expanded services in the following ways:**

- Offered afternoon appointments for parents to bring children in for eyecare
- Offered vision therapy in our clinic to those with state insurance or no insurance
- Staggered student assignments to address the need of optometry students to leave for other academic or clinical assignments
- Engaged a Chicago Health Corp member (AmeriCorp) to address lack of follow-up recommended
- Received a grant from LCIF to address long delays in receiving prescription eye wear.
- Added OCT, fundus camera and Visagraph so that referrals to the main campus of ICO is unnecessary for these services.
- Continued to market the clinic throughout the Chicago Public School system.

### Future goals will include:

- Expansion of hours and services by becoming an externship site for fourth year optometric students at other institutions.
- Consider adding contact lenses to our services.

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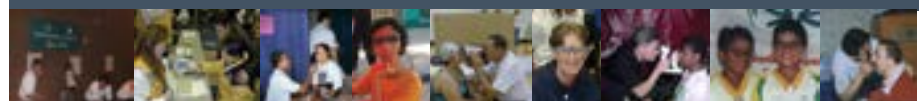
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# Comparison of Visual Findings of Athletes Participating in the Special Olympics Lions Clubs International Opening Eyes by Regions in 2010

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## PURPOSE

This study is a review of the vision screenings of athletes participating in the Opening Eyes by the six regions: Africa, Asia Pacific, East Asia, Europe/Eurasia, Latin America, and North America. Special Olympics is a year round program for individuals who have been diagnosed with intellectual or developmental disabilities. Please note data for the Middle East was unavailable.

## METHOD

A standardized comprehensive vision program is offered to Special Olympics athletes at no cost all around the world. A trained clinical director coordinates the screening, volunteers, and a after the completion of the screening. The Opening Eyes Vision Screening evaluates visual acuity, cover test, color vision, stereopsis, autorefracton, eye health and IOP. Data from the vision program is then entered into a central on-line data system. Data was then extracted for 2010 and analyzed with SPSS 17.0 comparing the visual findings of the athletes by region seen during 2010.

## RESULTS

Data from 21,326 athletes was reviewed. Most of the athletes seen were male (64.1%) with no gender differences found among the regions. Athletes from Africa, Asia Pacific, East Asia, and Latin America were younger than the other regions (Figure 1).

Athletes from Africa, Asia Pacific and Latin America were most likely to report that they never have an eye exam.

A comparison of timing of the last eye exam based on athlete report as compared to the financial status based on World Bank criteria showed the athletes from the regions considered high income appear to access eye care within the

recommendation of every 3 years more often (Figure 2).

The criteria for classification are as follows:

The economies below are divided according to 2011 GNI per capita, which is calculated using the World Bank Atlas Method. Each year on July 1, the World Bank revises the classification of world economies. As of 1 July 2012, the World Bank income classifications by GNI per capita are as follows:

- low-income, \$1,025 - or less;
- lower-middle-income, \$1,026 – \$4,035;
- upper-middle-income, \$4,036 – \$12,475;
- high-income, \$12,476 or more



No regional difference in color vision and strabismus (distance and near) were found. The prevalence of strabismus ranged from 13.2%-14.1% at far and 11.9%-13.1% at near.

Refractive error did reflect regional differences with Asia Pacific showing highest mean spherical equivalent (-1.150 D +/-3.58) while East Asia had the lowest (-0.64 D +/-3.58). Post hoc tests revealed significant differences only between these regions (Figure 3).

## CONCLUSIONS

More significant differences in refractive error were expected to be found between the regions based on diversity of refractive error reported in the literature then we actually found in our study.

It is our interpretation that athletes participating in Special Olympics reflect more similarities in vision findings to each other than to the region that they represent. This is supported by the lack of the similarity in refractive error, strabismus and color vision between them and their respective regions and more similarity to each other.

One difference to note is that athletes residing in North America and Europe/Eurasia were more likely to have had an exam than the other four regions.

Limitations of this study include:

- the history is provided by the athletes,
- the athletes represent only those who were interested in participating first in Special Olympics and also interested in participating in the screening
- our data may exclude those athletes receiving care locally
- data is entered into the data base at multiple locations around the world and we have limited ability to ensure its accuracy in data entry

Further study should include expanding the program to individuals with disabilities that do not participate in Special Olympics to be able to generalize the findings.

This research is made possible by the financial and in kind support of the following:

- Essilor
- Essilor Vision Foundations
- Safilo
- Lions Clubs International Foundation

Figure 1

AGE (YEARS)							
REGION	N	Valid	Missing	Mean (std dev)	Median	Mode	Range
Africa (AF)	955	848	107	16.8 (7.8)	15.2	15.2	3.0-66.5
Asia Pacific (AP)	2933	2822	111	18.7 (8.1)	17	10.6	2.6-85.3
East Asia (EA)	2786	2782	4	19.0 (6.2)	17.6	12.9	4.0-62.0
Europe / Eurasia and Israel (EE)	3856	3781	75	26.3 (11.2)	23.2	17	3.2-94.5
Latin America and the Caribbean (LA)	3053	3020	33	19.2 (9.2)	17.5	7.7*	2.7-98.4
North America (NA)	7777	7597	180	26.1 (12.8)	22.7	17.4*	4.4-76.5

\* Multiple modes exist. The smallest value is shown

Figure 2

REGION	Reported Last Eye Exam	WORLD BANK LEVEL			
		<3 years	>=3 years or never	<3 years	>=3 years or never
AF	<3 years	95	34	24	
AP	>=3 years or never	167	276	41	89
EA	<3 years	510	160		
EA	>=3 years or never	457	111		
EA	<3 years	218	796	54	
EA	>=3 years or never	160	911	84	
EE	<3 years	13	264	1666	
EE	>=3 years or never	3	25	235	1185
LA	<3 years	95	710	13	
LA	>=3 years or never	387	1172		
NA	<3 years	1	5038		
NA	>=3 years or never		4	2371	
Total	<3 years	316 (48.9%)	276 (14.1%)	1975 (41%)	6860 (64.5%)
Total	>=3 years or never	339 (51.1%)	1690 (75.9%)	2840 (59%)	3774 (35.5%)

Figure 3

REGION	Column 1	Column 2	Column 3	Column 4	Column 5
AF	Mean	-0.19	-1.36	-0.82	
AF	Std. Dev.	3.30	1.41	3.42	
AP	Mean	-0.43	-1.47	-1.15	
AP	Std. Dev.	3.46	1.67	3.58	
EA	Mean	-0.01	1.29	-0.05	
EA	Std. Dev.	3.58	1.37	3.58	
EE	Mean	-0.26	-1.36	-0.92	
EE	Std. Dev.	3.59	1.70	3.58	
LA	Mean	-0.32	-1.45	-0.97	
LA	Std. Dev.	3.22	1.53	3.35	
NA	Mean	-0.29	-1.41	-0.99	
NA	Std. Dev.	3.30	1.63	3.45	





ICO

# GLOBAL SPECIALTY LENS SYMPOSIUM

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# ICO

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# Therapeutic Use of Mini-Scleral and Scleral Lenses in Patients with Graves' Ophthalmopathy

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## BACKGROUND

Patients with Graves' ophthalmopathy can be very challenging to manage secondary to the complex nature of their disease presentation. Patients may present with a variety of ocular findings including: lid retraction, periorbital and lid swelling, chemosis, conjunctival hyperemia, proptosis, optic neuropathy, restrictive myopathy, exposure keratopathy and/or keratoconjunctivitis sicca. Patients with these problems who also require refractive correction may experience difficulties in achieving acceptable quality of vision with traditional contact lens designs. Even with maximum topical and systemic therapy, these patients' ocular conditions may be difficult to manage. With the option of mini-scleral and scleral lens designs, patients now have an option to protect the cornea and offer better vision. Scleral contact lenses allow for successful and comfortable fitting and vision in patients with complex corneas. The large gas permeable lenses serve as a pre-corneal fluid reservoir which provides optical correction while rehabilitating the ocular surface.

We present here two patients who had been diagnosed with Graves' ophthalmopathy over ten years prior. With significant ocular surface staining and significant spectacle correction, both patients had never been able to wear contact lenses comfortably. The patients were fit with mini-scleral and scleral contact lenses not only to assist in the rehabilitation of their ocular surface, but also to improve the quality of their vision.

Figures 1 and 2: Topographies showing irregular astigmatism, 10.44 diopters OD and 3.57 diopters OS.



## CASES

### PATIENT 1

A 48 year old Caucasian male was diagnosed with Graves' thirteen years prior and was status-post a complete thyroidectomy. He had previously worn soft, gas permeable, and hybrid contact lenses. He complained of blur at distance and near with his current hybrid lenses, double vision at the end of the day, and significant redness and irritation that did not improve with traditional supportive dry eye therapy. Best corrected vision through the hybrid lenses was 20/30 OD and 20/50 OS. His refraction was -5.50/-4.50x082, 20/80 OD and -11.00/-3.00x115, 20/200 OS. Topographies showed irregular astigmatism that was greater in the right eye than the left, 10.44 diopters OD and 3.57 diopters OS (Figures 1 and 2).

The corneas showed 2+ diffuse sodium fluorescein staining and the conjunctiva showed 3+ lissamine green staining. Schirmer scores without anesthetic were 13mm OD and 8mm OS. The patient was started on Restasis® (cyclosporine ophthalmic emulsion) 0.05% BID, Lotemax® (loteprednol etabonate ophthalmic suspension) 0.5% BID, preservative free artificial tears QID, and preservative free ointment QHS OU to improve his ocular surface and increase tear production (Figures 3-4).

Mini-Scleral Design™ Lenses were ordered which provided acceptable vision and improved the ocular surface.

#### Initial MSD Parameters:

	SAG	Power	OAD	VA	OR
OD	4505	plano	15.8	20/25-	-3.75
OS	4505	plano	15.8	20/30	-4.75

With this first pair of lenses, central touch was noted OD>OS, excessive limbal clearance in the mid-peripheral zone, and slight scleral impingement after about 20 minutes of settling. The left lens demonstrated more central vault with peripheral impingement. Although the first set of diagnostic lenses were not an ideal fit, the patient immediately noted improved ocular comfort, and a significant improvement in the quality of his vision. With the first

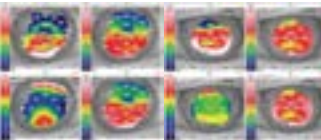
pair of diagnostic lenses, the patient's vision improved to 20/25- OD and 20/30 OS.

The final lenses were ordered with an increased central vault. The periphery was also adjusted to decrease conjunctival impingement OS. The overall pattern improved, the patient's comfort level increased, and the cornea had significantly less staining. The patient has been successfully wearing the lenses for 10 months noting an increase in the clarity of his vision, quality of his vision, and an improvement in ocular comfort. (Figures 5-8).

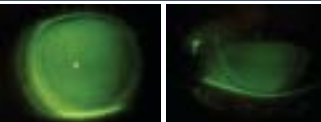
#### Final Lenses Ordered and Dispensed:

	SAG	Power	OAD	VA	Periphery
OD	4805	-2.00	15.8	20/25+	standard
OS	4705	-3.75	15.8	20/25	1 step flat

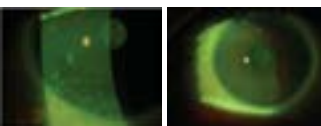
Figures 3 and 4: Lissamine green staining of the cornea and conjunctiva.



Figures 5 and 6: Final MSD lenses OD and OS



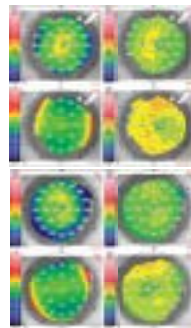
Figures 7 and 8: Sodium Fluorescein staining pre- and post-MSD lens fit.



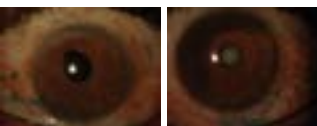
### PATIENT 2

A 46 year old African-American male was diagnosed with Graves' over ten years prior and was status-post orbital decompression and EOM surgery (2011). He had never successfully been able to wear any form of contact lens secondary to significant contact lens discomfort. He presented with complaints of blur, double vision, and ocular discomfort. An abduction deficit was noticed upon EOMs OU. The patient was prototypic and had diffuse conjunctival and corneal staining OU (thyrotoxic exophthalmos OU). The patient was also aphakic OS. Refraction was -32.00DS, 20/400 OD and -2.00/-1.00x180, 20/100 OS. Topographies showed central islands secondary to significant corneal staining from exposure keratitis. (Figures 9-12).

Figures 9 and 10: Topographies OD and OS



Figures 11 and 12: Lissamine green staining of the cornea and conjunctiva.



The patient was started on Restasis® (cyclosporine ophthalmic emulsion) 0.05% BID, preservative free artificial tears QID, preservative free ointment QHS O, and was instructed to wear moisture goggles to improve his ocular surface.

The patient was fit with scleral lenses by another practitioner to decrease image distortion and improved corneal staining.

#### Initial Scleral Parameters:

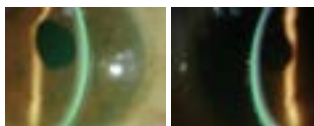
	BC	Power	OAD	VA	Periphery
OD	8.04	-20.50	20.2	20/70	.2mm flat
OS	7.50	-1.25	18.8	20/100	standard

With this first pair of lenses, central touch was noted OD>OS and the periphery had significant edge lift on the right eye and conjunctival impingement of the left eye. The high edge lift OD caused the lens to pop off during evaluation. Secondary to the lenses not providing an adequate fit, the patient was re-fit with the Jupiter reverse geometry scleral lenses by Essilor. After several adjustments, the final lenses ordered and dispensed provided an improved overall pattern, increased ocular comfort, and decreased corneal staining. The patient has been successfully wearing the lenses for 3 months for up to 6-8 hours per day. He has noticed an increase in the clarity of his vision, quality of his vision, and an improvement in ocular comfort. (Figures 13-14).

#### Final Jupiter Parameters:

	BC	Power	OAD	VA	Periphery
OD	8.23	-27.00	18.8	20/40	8.50, 9.35, 16.70
OS	8.23	+0.25	22.0	20/100	9.75, 10.20, 17.00

Figures 13 and 14: Final Jupiter lenses OD and OS.



## CONCLUSIONS

Patients with Graves' ophthalmopathy can be successfully fit with large diameter gas permeable contact lenses that provide adequate oxygen to the cornea, optimal ocular health, maximal comfort, and clear vision. Mini-scleral and scleral lenses may be an option for patients with ocular surface damage and high amounts of astigmatism to optimize comfort and vision quickly. In these patients who have been diagnosed with severe ocular surface disease, scleral lenses may be considered as a therapeutic device to assist in rehabilitating the ocular surface and to determine their optimum vision potential. In these two cases, mini-scleral and scleral lens designs provided improvement in the patients' ocular surface, comfort and vision. However, further investigation is needed regarding the role of scleral lens designs in patients with Graves' Ophthalmopathy.

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# ICO

3241 South Michigan Avenue, Chicago, Illinois 60616

# Therapeutic Scleral Contact Lens: Treatment for Limbal Stem Cell Deficiency

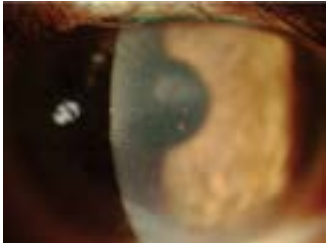
Stephanie Parker, OD, Kaitlyn Keller, OD, Jennifer Harthan, OD, FAAO

## BACKGROUND/PURPOSE

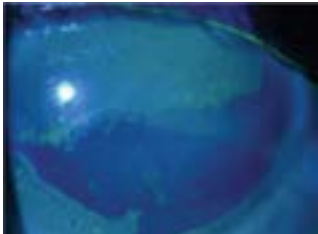
In limbal stem cell deficiency (LSCD), the depletion of limbal stem cells leads to conjunctivalization of the cornea. In sectoral LSCD only part of the cornea is involved, in contrast to diffuse which affects the entire cornea. Secondary stem cell deficiency is caused by external factors, such as ocular surface trauma, where as primary stem cell deficiency is often due to a disease process. Current treatments for secondary partial limbal stem cell deficiency include monitoring and mechanical debridement with or without amniotic membrane transplantation.

## CASE SUMMARY

A 52 y/o CM presented with a complaint of progressive blurred and double vision in the left eye only for the past four years. It began several months after blunt trauma to the left eye. Ocular history was also remarkable for LASIK OU in 1991. Uncorrected

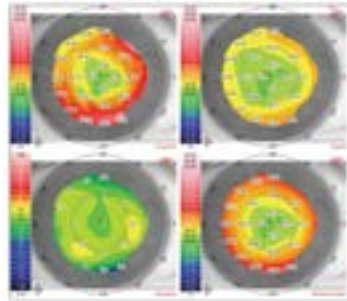


LSCD SLE

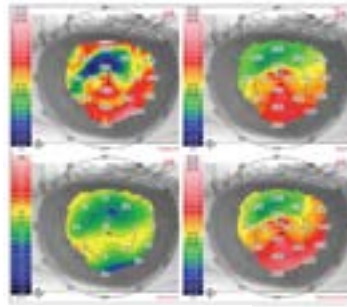


LSCD NaFl Stain

visual acuities were 20/20 OD and 20/250 with monocular diplopia OS. Manifest refraction in the right eye showed -0.25 -0.75 x 140 with a VA of 20/20, and in the left eye +2.25 -1.50 x 105 with a VA of 20/150. There was no clear endpoint on refraction and the horizontal and vertical diplopia remained. Biomicroscopy findings in the right eye revealed trace diffuse punctate epithelial erosion, and a clear LASIK flap. Notable findings in the left eye included mild ptosis, superior and inferior-nasal pannus with diffuse neovascularization, central stromal haze, and traumatic corneal endothelial and anterior capsular pigment. Sodium fluorescein staining revealed a clear demarcation line between the conjunctiva and corneal epithelial cells. Intraocular pressures were 15mmHg OD, OS. Gonioscopy revealed angle recession from one to seven o'clock in the left eye. Slight asymmetry of the cup to disc ratio was noted with 0.55 OD and 0.60 OS.



LASIK Topo



LSCD Topo

Topography findings in the right eye showed a good LASIK outcome, with a clear central ablation zone. In regards to the left eye, the tangential map showed a clear demarcation line, with K values on the superior half of the cornea of 35 and the inferior half of 56. The elevation map also showed a relative shallowness of the superior half of the cornea compared to the inferior half of the cornea. And on the refractive map within the visual axis, a high amount of irregular astigmatism was present with dioptric powers ranging from 44.37 to 52.89.

Assessments of limbal stem cell deficiency secondary to trauma OS and glaucoma suspect OS were made. After discussing treatment options with the patient, a therapeutic scleral contact lens was chosen as a non-invasive option to provide immediate visual improvement. Taking into account the patient's small fissure, he was fit with a -4.00, 47.01, 15.6 Jupiter lens in Boston XO material with standard peripheral curves. With this lens, approximately 200-250 microns of vault was achieved with excellent conjunctival landing. The patient's visual acuity with the lens was 20/20 with no monocular diplopia. The patient was also scheduled to return for a glaucoma work-up including an OCT and HVF.



Conjunctival Landing



Central Vault

## CONCLUSION

Therapeutic scleral lenses are a new promising treatment for LSCD. They offer a non-invasive option for patients and provide the best immediate visual improvement. The tear reservoir is highly oxygenated and helps to stimulate and maintain epithelial cell restoration.

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ICO

# SOCIETY FOR NEUROSCIENCE

1 ICO PRESENTATION

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## Abstract

Lipid rafts are specialized membrane domains rich in cholesterol and intimately associated with cytoskeletal components. G protein signaling is influenced by these domains, but, depending upon the receptor, G protein, and effector enzyme, they either facilitate or attenuate signaling (Allen et al., 2007, 2009). We have demonstrated that, for Gs and Gs-coupled receptors (β-adrenergic, VPAC and 5HT-4, -6, -7), lipid rafts attenuate signaling by separating Gs from adenylyl cyclase. Several lines of investigations from different laboratories suggest a post-synaptic effect of chronic antidepressants and a possible postsynaptic target for these drugs. Data from rats, cultured neural and glial cells, all suggest that the localization of the G protein, Gsα, in lipid rafts is modified by chronic treatment with a number of antidepressant compounds (SSRI, MAOI and tricyclic) (Donati et al., 2005; Toki et al., 1999). Antidepressants facilitate translocation of Gsα from lipid rafts while post mortem studies show increased Gsα in raft fractions from several brain regions of depressed suicide cases relative to controls. In this study, we sought to determine whether raft fractions prepared from platelets of depressed subjects showed enrichment of Gsα in lipid raft fractions. Blood was collected in Ancona Italy, separated into components, coded and shipped to Chicago for assay. Platelet Gsα was extracted, subsequently with Triton X-100 (non-raft fraction) and Triton X 114 (raft fraction). Gsα in the raft fraction was significantly ( $p < .001$ ) greater in platelets prepared from depressed subjects. This suggests the possible development of a simple blood test to indicate the presence of depression. As chronic antidepressants have been shown to translocate Gsα from lipid rafts, it will be interesting to follow Gsα sequestration in depressed patients as they receive and respond to treatment.

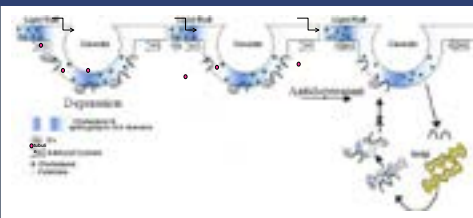
## Introduction

Despite decades of research, no common mechanism has emerged to link the activities of the diverse compounds used in therapy for depression. While primary targets of these agents differ, a shared effect includes increased cAMP production and a cascade of events resulting from sustained increase in cAMP or the activation of Gsα (Malberg and Blendy, 2005).

Antidepressant treatment causes a shift in the localization of the heterotrimeric G protein, Gsα, from a Triton-X 100 insoluble lipid rich domain to a Triton-X 100 soluble non-raft domain, leaving Gsα more available to activate adenylyl cyclase. Both diminished Gsα-adenylyl cyclase coupling and an increase in the proportion of Gsα in lipid rafts are seen in depression, and antidepressants concentrate in lipid rafts (Eisensamer et al., 2005). Thus, it is hypothesized that a) Gsα association with lipid raft will be high during depression and this will lead to decreased cAMP production; b) chronic treatment with therapeutically effective antidepressants will mobilize Gsα to non-raft domains of the plasma membrane and up-regulate the cAMP pathway; c) these central events will be reflected in a peripheral biomarker for both depression and antidepressant response.

We have verified in post-mortem brain that depression correlates with an increased proportion of Gsα in lipid rafts (Donati et al., 2008); a decrease in Gsα-activated adenylyl cyclase in depression has been observed in platelets (Hines and Tabakoff, 2005). We suggest that the ratio of raft-associated to non-raft Gsα is a biomarker of depression, a biomarker readily measured in a standard clinical laboratory.

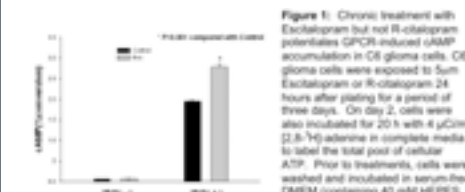
## Chronic antidepressant treatment causes an exodus of Gsα from lipid rafts that has accumulated during periods of depression



**Model.** Gsα localization in depression and after antidepressant treatment. In normal synaptic membranes about 75% of Gsα is localized in Triton-X 100 insoluble lipid rafts where it is also complexed with tubulin. Chronic treatment with a number of antidepressants shifts Gsα out of lipid rafts into membrane domains where it is more associated with adenylyl cyclase and less associated with cytoskeletal components including tubulin. Studies with post-mortem samples from depressed humans (Donati et al., 2007) show that a greater proportion of the Gsα is sequestered in lipid rafts than seen in controls. Note that while this model suggests that chronic treatment with antidepressants shifts Gsα into non-raft membrane domains, the molecular target of the drugs, or the molecular alterations in depression that shift Gsα into rafts, have not been established. We do not suggest that antidepressant drugs bind directly to either Gsα or tubulin.

## Chronic Antidepressant treatment increases coupling between Gsα and adenylyl cyclase and translocates Gsα from lipid rafts.

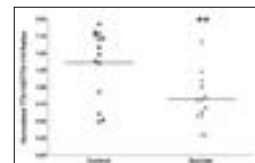
### Chronic antidepressant treatment of C6 cells results in a prolonged elevation of cAMP



**Figure 1:** Chronic treatment with Escitalopram but not R-citalopram potentiates GPCR-induced cAMP accumulation in C6 glioma cells. C6 glioma cells were exposed to 500 nM Escitalopram or R-citalopram 24 hours after plating for a period of three days. On day 3, cells were incubated for 20 h with 4 μM (2,8-14)adenine in complete media to label the total pool of cellular ATP. Prior to treatments, cells were washed and incubated in serum-free DMEM (containing 40 mM HEPES, 40 mM Tris and 1 mM EGTA at pH 7.40), and pre-treated with 500 μM butyl methylxanthine (IBMX), a broad spectrum phosphodiesterase inhibitor, for 5 min. To prevent cAMP-degradation, cells were subsequently exposed to forskolin or isoproterenol at 37°C for 30 min. Reactions were stopped and cells lysed by addition of 5% trichloroacetic acid. Cell lysates were incubated on ice for 1 h and cell debris removed by centrifugation. 1000 cpm each of [<sup>32</sup>P]-o-ATP and [<sup>32</sup>P]-cAMP were added as internal standards to each sample. (2,8-14)-ATP and (2,8-14)-cAMP were separated by sequential Dowex-Alumina ion exchange column chromatography and eluates collected into vials followed by scintillation counting for <sup>3</sup>H, <sup>14</sup>C and <sup>32</sup>P. Normalizing to (2,8-14)-ATP served to control for variations in cell number and metabolism unrelated to adenylyl cyclase activity. Variations in column recovery were corrected for using the [<sup>32</sup>P]-cAMP counts (Solomon 1976). Data are expressed as cAMP formed / (ATP formed + cAMP formed) × 100%.

From Zhang and Rasenick, 2010 see also Ozawa and Rasenick, 1989; Malberg and Blendy, 2005

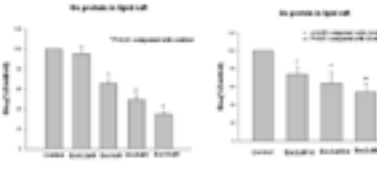
## Post-mortem analysis reveals that Gsα is enriched in lipid rafts of membranes derived from brain of depressed patients



**Figure 3.** Comparison of Gsα localization in cerebellum of control vs. depressed suicide subjects. Human tissue was obtained from Dr. Ghanshyam Pandey (UIC) through the Brain Collection Program of the Maryland Psychiatric Research Center. All suicides had a history of depression; age and post-mortem interval matched controls had no reported psychiatric history. Human cerebellar membrane tissue was sequentially extracted with TTX-100 (soluble Gsα) followed by TTX-114 (insoluble, "lipid raft" Gsα) and the amount of Gsα was compared in the extracts. A TTX-100 to TTX-114 ratio is shown to demonstrate that control subjects (n=14) have a significantly higher proportion of their Gsα localized to detergent soluble membrane domains than do depressed suicide subjects (n=13). [Unpaired t-test  $p < 0.02$ , control mean ratio =  $1.364 \pm 0.5108$  & suicide mean ratio =  $0.8225 \pm 0.3772$ ] (Donati et al., 2008)

Similar results were obtained with membranes obtained from frontal cortex.

### Chronic antidepressant treatment of C6 cells translocates Gsα to a non-raft membrane fraction

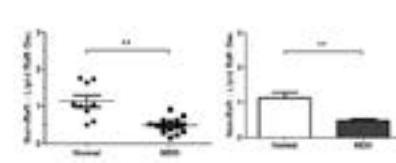


**Figure 2** C6 glioma cells were treated with the indicated dose of escitalopram for the indicated time. After treatment, cells were harvested, membranes prepared and lipid rafts isolated by sucrose gradient sedimentation. Results of densitometry from Western Blots is shown. No changes in overall Gsα were seen. R-citalopram did not translocate Gsα into lipid rafts.

Other agents that have been observed to translocate Gsα after chronic treatment are tricyclic antidepressants (imipramine, desipramine and amitriptyline) SSRIs and SNRIs (fluoxetine, sertraline, venlafaxine) the MAOI, phenelzine. Acute treatment of all compounds was ineffective and other compounds without effect were amphetamine and chlorpromazine. Total Gsα did not vary with any of the treatments).

Data from Zhang & Rasenick, 2010; See also Toki et al., 1999; Donati et al., 2005

## Platelets from depressed human subjects show increased raft association, similar to that seen in postmortem human brain



**Figure 4.** Mean (±SEM) ratios of Gsα in non-raft vs. raft membrane fractions as measured by sequential extraction with the detergents TX-100 and TX-114 for Normal and Major Depressive Disorder (MDD) subjects. Results from a Unpaired t-test show a significant difference between Normal ( $1.141 \pm 0.16$  N=9) and MDD ( $0.4873 \pm 0.051$  N=15) groups (\*\* $p < 0.0001$ ).

## Summary and conclusions

These data suggest the possible development of a simple blood test as a biomarker for depression that may be useful in confirming a clinical diagnosis and in aiding the recruitment of patients for Phase II/III efficacy trials. Furthermore, as chronic (3-5 day) antidepressant treatment translocates Gsα from lipid rafts in cultured cells, it will be interesting to follow Gsα sequestration in depressed subjects as they receive and respond (or fail to respond) to treatment. It is hypothesized that Gsα will translocate from lipid rafts within one week in showing a positive response to antidepressant treatment at 8 weeks.

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ICO

# NAP (NATIONAL ACADEMY OF PRACTICE)

1 ICO PRESENTATION

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# National Center for Children's Vision & Eye Health: An Interdisciplinary Approach to Children's Vision and Eye Health

Sandra S. Block, OD, M Ed, FAAO, FCOVD  
Professor, Illinois College of Optometry

## INTRODUCTION

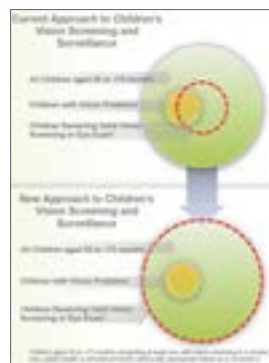
Prevent Blindness America, with support from the Maternal and Child Health Bureau, established the National Center for Children's Vision and Eye Health (Center) to address children's vision screening. The National Center, established in 2009 is looking to create public health infrastructure, training and education addressing the vision and eye health needs of young children. The Center has initially targeted children aged 36 to <72 months.

This center is one of 18 centers supported by MCHB – Division of Services for Children with Special Health Care Needs.

The challenge of the Center was to design a universal vision screening program that is flexible and effective on local, state, and national levels. Challenges include mobilizing stakeholders, building capacity and creating the expertise for success.

An original expert panel met over a 3 year period to determine what form the recommendations for children's eye health and vision should take. The panel had representatives from the following areas:

- Ophthalmology
- Pediatrics/Family Physicians
- Nurse/School Nurse
- Public Health/Title V Program
- Family Advocate
- Head Start/Childcare
- Early Education
- Vision Research



**Figure 1.** The top of the figure depicts the current state of children's vision in which only a portion of the population receives screening and/or eye care.

The bottom portion depicts a vision system in which all members of the target population receive a vision screen or full eye examination.

## PROCESS

The Center was focused on two primary initiatives: First establishing pilots programs in five states, and two, the National Expert Panel work to plan a comprehensive national approach to children's vision screening:

The National Expert Panel formed recommendations around 3 key areas:

1. Performance measures track both provision & receipt of vision screening in children 3-5 years of age.
2. Uniform management data collected during vision screening: demographics, results screening, and capturing follow up to eye exams and treatment outcome.
3. Best practice protocol supported by research evidence.

Recommendations were developed incorporating review of the literature; consultation with states developing their vision screening infrastructure; and consultation with experts in the national and state agencies that are actively involved with performance measure development. The full recommendations are in process to be published in early 2013.

Pilot programs were established in five states (Ohio, Massachusetts, Illinois, Georgia, and North Carolina) to seek out best practices that align with the panel recommendations and to study possible strategies for vision screening, accessing comprehensive eye care and surveillance. Each state developed a program to improve or enhance their children's vision program.

The National Expert Panel recently transitioned to an advisory body under the direction of Dr. M. Kathleen Murphy with three primary objectives:

1. Serve as technical resource for children's vision programs based on scientific evidence, build partnerships with key stakeholders which include physicians, community programs, educators, parents and state government.
2. Assist in creating an improved method for surveillance of children's vision screening, outcomes, follow up and disparities.
3. Develop and disseminate tools and information to promote a comprehensive approach to children's vision and eye health in an effort to improve surveillance.

The Advisory Committee of the Center under the direction of Dr. Kathleen Murphy has three legs: education, technical guidance and policy. These subgroups will work to advocate for implementation of the National Expert Panel recommendations.

## RESULTS

### Vision performance measures:

Well-crafted valid and reliable measures performance measures can help to drive the development of appropriate data systems.

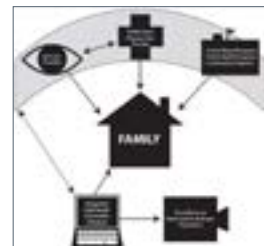
The panel determined all care received by the child should be included.

A child-based measure is preferred, which includes all sources of vision care and removes duplicate counts for children receiving care from more than one provider

### Vision data collection:

Vision programming surveillance needs to incorporate systematic data collection, including child-specific identifiers to ensure that the data are accurately linked to the child without duplications.

Data entry should be simple for community-based as well as health care provider office-based screenings, incorporate



**Figure 2.** Flow of vision screening outcomes in an integrated system.

### Vision programs implementation:

Vision screening of children aged 36 through less than 72 months can be performed using scientifically recommended methods.

Regardless of the method(s) selected, the method is only one part of a comprehensive children's vision program (Image 1). The screening system is only successful when the result of the screening is used in a meaningful way.

**Image 1.** Key features of a comprehensive screening program.



## CONCLUSIONS

Children's eye and vision health has been perpetually challenged by a lack of national standardization, infrastructure, and surveillance.

Vision screening lies at the intersection of multiple health care providers including pediatricians, optometrists, and ophthalmologists as well as many public institutions (Departments of Education, Departments of Public Health, etc).

Each entity has a role in the in addressing children's vision, but often each role is uniquely defined varying by geography and profession.

The Expert Panel to the National Center for Children's Vision and Eye Health has suggested a comprehensive approach including implementation and surveillance with the goal of reducing the number of children suffering from needless vision loss. The Advisory Group will be working diligently to engage the appropriate partners and provide needed support to transition the recommendations into action.

Uniformity in implementation of screening and follow up to eye care, improved data sharing and provider communication, and establishment of state and national level performance measures for children's vision screening and care represent the recommended pathway to healthier vision for children

## ACKNOWLEDGMENTS

We thank members of the National Expert Panel to the National Center for Children's Vision and Eye Health for their countless hours of work to improve children's vision in the U.S. The work represented here was supported by grants (H7MMC15141 and H7MMC24738) from the Maternal and Child Health Bureau of the Health Resources and Services Administration, U.S. Department of Health and Human Services.

## FOR FURTHER INFORMATION

Please contact [kbaldonado@preventblindness.org](mailto:kbaldonado@preventblindness.org). More information on this and related projects can be obtained at <http://nationalcenter.preventblindness.org>.

H7MMC24738

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ICO

# EUNOS

(EUROPEAN NEURO-OPHTHALMOLOGICAL SOCIETY)

1 ICO PRESENTATION

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# The Value of iPads for Low Vision Rehabilitation in Patients with Optic Atrophy

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<sup>3</sup>The Chicago Lighthouse for People Who Are Blind or Visually Impaired, Chicago, Illinois, USA;

<sup>4</sup>Illinois College of Optometry, Chicago, Illinois, USA

## INTRODUCTION

Accessibility features of tablet computers such as the Apple iPad have revolutionized reading rehabilitation for low vision patients. These features include system wide zoom and high reversible contrast. We compared subjective preference as well as reading rates on the Apple iPad and a closed circuit television (CCTV).

## METHODS

After IRB approval, 14 low vision patients, aged 18 years and older, were recruited with best corrected visual acuity (BCVA) of the better eye ranging from 20/50 to 20/200 and minimal prior experience with an iPad or a CCTV. The results of the entire study will be presented as a poster at the 2013 ARVO meeting. The present abstract focuses solely on two patients with optic atrophy, a neuro-ophthalmologic diagnosis. Data collection involved measuring reading rates from a newspaper article and a book individually. Patients read each media for two minutes on each device at their preferred zoom magnification, and a third time on the CCTV with the zoom magnification matched to the iPad's angle of resolution. Physical copies were used on the 24-inch Optelec Clearview CCTV with print size of 1.0 M and electronic copies were acquired on a third generation iPad. Upon conclusion of the reading assignment, patients were surveyed with a questionnaire concerning subjective comfort, performance, and preference.\*

## CONCLUSION

The iPad is a valuable new tool in assisting low vision patients, but may not completely replace the use of CCTV. Patients' primary reasons for preference of the iPad were portability, ease of navigation, and added versatility. Considering these reasons in addition to lower cost and improved social acceptance, the iPad should be considered in the reading rehabilitation of visually impaired patients with optic atrophy.

## SUPPORT

2012 Illinois Society for the Prevention of Blindness Grant



Illinois Society for the Prevention of Blindness  
Empowering and Inspiring People with Vision Loss

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Results							
Patient	Age	BCVA Better Eye	BOOK		NEWSPAPER		Overall Preference
			Faster Reading Rate	Preference	Faster Reading Rate	Preference	
1	63	20/60	iPad	iPad	CCTV	CCTV	iPad
2	76	20/120	CCTV	CCTV	CCVT	CCTV	CCTV



Apple iPad



ClosedCircuit Television (CCTV)

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ICO

# CMSC

(CONSORTIUM OF MULTIPLE SCLEROSIS CENTERS)

## 1 ICO PRESENTATION

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# SD-OCT Imaging of the Retina in Primary Progressive Multiple Sclerosis

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## INTRODUCTION

Multiple Sclerosis (MS) is an autoimmune disease that affects the central nervous system and is characterized by altered permeability of the blood-brain barrier resulting in lesions and neurodegeneration. Primary Progressive Multiple Sclerosis (PPMS) is a type of MS that is characterized by steady worsening of neurologic function without any distinct relapses or periods of remission. Optical coherence tomography (OCT) can be used to assess retinal degeneration in MS. It can provide values that represent the retinal nerve fiber layer (RNFL) and the macular volume. Some of the literature suggests that PPMS may actually have a distinct pathophysiology different than the typical Relapsing Remitting Multiple Sclerosis (RRMS) and may represent a primary neurodegenerative disorder. OCT can be used to study the layers of the retina and may give clues as to the mechanism of this disease.

## PURPOSE

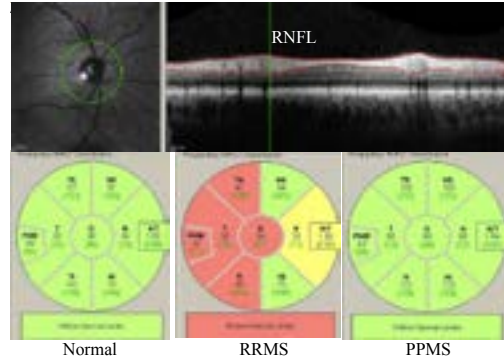
To evaluate the feasibility of using spectral domain OCT (SD-OCT) to evaluate the different retinal layer thicknesses and macular volume measurements in patients with PPMS compared to those with RRMS and normal controls.

## METHODS

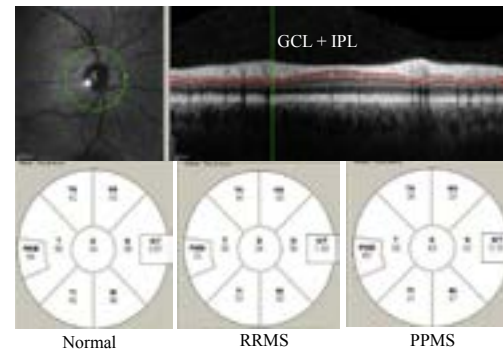
**Subjects:** A group of ten patients diagnosed with Primary Progressive Multiple Sclerosis were scanned and compared to age matched patients with Relapsing Remitting Multiple Sclerosis and normal healthy controls.

**OCT imaging:** RNFL and total macular volume scans were obtained using a spectral-domain OCT (Heidelberg Spectralis SD-OCT, Heidelberg Engineering, Germany) for each eye of the patients. All scans were acquired by experienced operators and were reviewed for sufficient signal strength, correct centering and segmentation. Together the Ganglion Cell and Inner Plexiform layers, and the Inner Nuclear Layer were manually segmented on the RNFL scans for all of the patients.

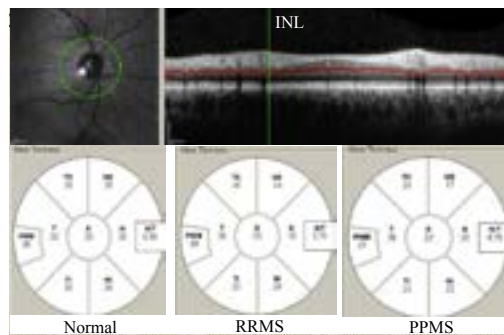
**Figure A:**  
OCT analysis of the RNFL thickness circumferentially in a normal, RRMS, and PPMS patient.



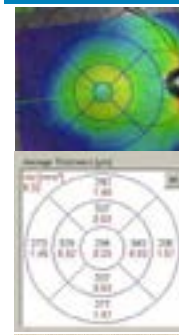
**Figure B:**  
OCT analysis of the segmented GCL + IPL thickness circumferentially in a normal, RRMS, and PPMS patient



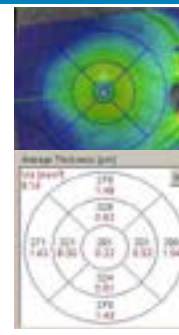
**Figure C:**  
OCT analysis of the segmented INL thickness circumferentially in a normal, RRMS, and PPMS patient.



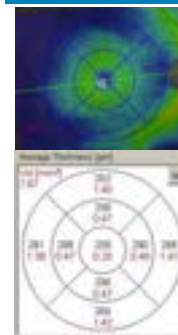
**Figure D:**  
Macular thickness and volume of a normal control patient.



**Figure E:**  
Macular thickness and volume of a RRMS patient.



**Figure F:**  
Macular thickness and volume of a PPMS patient.



## RESULTS

Segmentation of the Ganglion Cell Layer (GCL) and Inner Plexiform Layer (IPL) in patients with PPMS showed that the Papillomacular bundle (PMB) thickness was consistently higher than patients with RRMS. Similar findings were also noted in the temporal segment of PPMS patients. However, these changes were not seen in other segments of PPMS patients (temporal superior, temporal inferior, nasal, nasal superior, nasal inferior) compared to RRMS patients and controls. Segmentation of the Inner Nuclear Layer (INL) in PPMS patients in our study did not confirm the findings stated previously using similar technology (Heidelberg Spectralis SD-OCT), which found a reduction in the INL thickness in PPMS patients compared to RRMS patients and controls.<sup>1</sup> It was also noted that the macular volume and thickness was decreased in patients with PPMS compared to those with RRMS and controls.

## CONCLUSIONS

It is possible to obtain high quality manually segmented SD-OCT images of the GCL, IPL, and INL in patients with PPMS. Further studies are needed using SD-OCT in a larger group of patients with PPMS to determine the mechanism as to why the thickness of the PMB and temporal segments of the GCL and IPL are preserved compared to patients with RRMS.

## REFERENCES

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ICO

# THE TEACHING PROFESSOR CONFERENCE

## 1 ICO PRESENTATION

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# ICO

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# A Novel Approach to Bridge the Gap between Didactic and Clinical Education

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## INTRODUCTION

Traditional problem-based learning (PBL) is a learner-centered educational pedagogy in which students learn through the experience of problem solving in small collaborative groups. Primary Care Conference (PCC) is a hybrid project-based approach to learning that utilizes clinical cases to help bridge the gap between what happens in the classroom and the expectations of clinical performance. The goals of PBL in optometry school are to develop knowledge that is adaptable to a variety of clinical presentations, effective problem solving skills, self-directed learning and intrinsic motivation.<sup>1,2</sup> Unfortunately, traditional problem-based learning is faculty intensive with one faculty member instructing a small group of students. This can act as a financial burden on the institution and prohibits its wide spread adoption.<sup>3,4</sup> The Illinois College of Optometry (ICO) has developed PCC, a didactic course to supplement third year clinical patient care, which takes a novel approach by allowing a single instructor to act as a facilitator for a class size exceeding 150 students. The purpose of developing PCC is to provide our students with the benefits of PBL in a large class setting.

Figure 1: The case information that is posted to the Learning Management System and printed for the In-Class version of the case.



## METHODS

Timed and image-rich clinical cases are presented to students via a learning management system and then later in class with discussion. These cases and questions are deliberately designed to require different levels of analysis. The few straightforward questions have complex partnered questions which involve an enhanced level of clinical decision-making. Ultimately, the multiple choice questions (MCQ) are divided into two groups based on level of analysis required to answer the question. The case and the lower level MCQ are posted on-line for independent open-book completion. Later, the same case and the higher level MCQ are presented in class in an open-book examination format which is immediately followed by a case discussion using an audience response system.

**Figure 2:** The questions that are posted with the case information on the LMS. The questions include commentary to supplement the article text. All multiple response questions are scored as "all or none"-no partial credit is given.

**Figure 2**  
Learning Management System Questions  
SELECT ALL THAT APPLY

1. Which of the following best describes the optic nerve? (SELECT ALL THAT APPLY)

A. 0.25 to 0.30 mm OD, 0.25 to 0.30 mm OS  
B. 0.25 to 0.30 mm OD, 0.25 to 0.30 mm OS  
C. 0.25 to 0.30 mm OD, 0.25 to 0.30 mm OS  
D. 0.25 to 0.30 mm OD, 0.25 to 0.30 mm OS  
E. 0.25 to 0.30 mm OD, 0.25 to 0.30 mm OS  
F. 0.25 to 0.30 mm OD, 0.25 to 0.30 mm OS  
G. 0.25 to 0.30 mm OD, 0.25 to 0.30 mm OS  
H. 0.25 to 0.30 mm OD, 0.25 to 0.30 mm OS  
I. 0.25 to 0.30 mm OD, 0.25 to 0.30 mm OS  
J. 0.25 to 0.30 mm OD, 0.25 to 0.30 mm OS

This question requires interpretation of a visual, which will assist in recognizing the "abnormalities" seen in the patient's optic nerve.

2. Considering the information presented, which of the following is the most likely diagnosis?

A. Pigment dispersion glaucoma  
B. Angle closure glaucoma  
C. Primary open-angle glaucoma  
D. Acute angle-closure glaucoma

This question requires clinical thinking. Putting the case data and image interpretation together to arrive at a diagnosis. This question is PARSED with In-Class question #1.

3. When selecting a glaucoma medication for this patient, which of the following mechanisms of action would be most appropriate?

A. Decreased aqueous humor production  
B. Increased outflow of aqueous humor  
C. Increased outflow of aqueous humor  
D. Increased outflow of aqueous humor  
E. Increased outflow of aqueous humor  
F. Increased outflow of aqueous humor  
G. Increased outflow of aqueous humor  
H. Increased outflow of aqueous humor  
I. Increased outflow of aqueous humor  
J. Increased outflow of aqueous humor

This question builds on LMS question #2 by placing emphasis on the fact that the etiology of the condition must be considered prior to selecting a treatment. It also underscores the importance of pathophysiology and format to introduction to In-Class questions. This question is PARSED with In-Class question #2.

4. Which of the following fields on the visual field print best confirms that this patient has glaucoma?

A. Raw Data  
B. Total Deviation  
C. Pattern Deviation  
D. Residual Deviation Probability Plot  
E. Pattern Deviation Probability Plot

This question serves to remind students of content taught in the didactic curriculum. No analysis of data is required to answer this question. This question is PARSED with In-Class question #3.

5. Assuming the phenomenon is not atypical, your working distance is 60cm, your axial reflex is 100%, and your accommodative response is 100%, what is the most likely cause of the patient's blurred vision?

A. With movement  
B. Against movement  
C. Neutral

This question is PARSED with LMS question #5.

6. Which of the following items should be performed prior to prescribing the subjective refraction and adding the patient will not experience diplopia? (SELECT ALL THAT APPLY)

A. Refraction  
B. Refraction  
C. Refraction  
D. Refraction  
E. Refraction  
F. Refraction  
G. Refraction  
H. Refraction  
I. Refraction  
J. Refraction

This question serves to remind students that patients in clinic often present with multiple problems that involve both knowledge of binocular vision and vision.

7. Which of the following changes would be the best noticeable difference (dB) for the patient's vision?

A. 0.25 dB  
B. 0.25 dB  
C. 0.25 dB  
D. 0.25 dB  
E. 0.25 dB  
F. 0.25 dB  
G. 0.25 dB  
H. 0.25 dB  
I. 0.25 dB  
J. 0.25 dB

This question functions to review when to deviate from one "standard answer". A very small amount of data analysis is required to answer this question. This question is PARSED with In-Class question #6.

## DISCUSSION

Primary Care Conference provides benefits to both students and faculty members. This learner centered module boasts objectives that extend far beyond simple content mastery. The overarching student objectives to these cases include, but are not limited to: lifelong learning, knowledge base enhancement, clinical thinking refinement (data analysis and interpretation of visuals), understanding treatment/management hierarchy, problem solving efficiency and preparation for the board examination. In addition to student objectives, these cases provide faculty and administration with opportunities to develop junior faculty, contribute to curriculum vitae building, and reinforce basic health science integration into clinical practice.

**Figure 3:** The questions that are printed with the case information for the In-Class case. The questions include commentary to supplement the article text. All multiple response questions are scored as "all or none"-no partial credit is given.

**Figure 3**  
In-Class Questions  
SELECT ALL THAT APPLY

1. A geneticopic view of the patient's anterior chamber angle most likely would reveal a:

A. separation of the iris from the lens  
B. contact of the iris with the trabecular meshwork  
C. contact of the iris with the trabecular meshwork  
D. contact of the iris with the trabecular meshwork  
E. contact of the iris with the trabecular meshwork  
F. contact of the iris with the trabecular meshwork  
G. contact of the iris with the trabecular meshwork  
H. contact of the iris with the trabecular meshwork  
I. contact of the iris with the trabecular meshwork  
J. contact of the iris with the trabecular meshwork

This question is a basic science correlate to LMS question #2. Reviewing the case on the LMS would assist the student in answering this question.

2. Which of the following medications would be the most appropriate initial treatment?

A. Timolol  
B. Travatan Z  
C. Flurbiprofen  
D. Gange  
E. Timoptic

This question is a basic science correlate to LMS question #2. Reviewing the case on the LMS would assist the student in answering this question.

3. Which of the following statements best describes this patient's left visual field? (SELECT ALL THAT APPLY)

A. Superior nasal hemianopia  
B. Superior nasal hemianopia  
C. Superior nasal hemianopia  
D. Superior nasal hemianopia  
E. Superior nasal hemianopia  
F. Superior nasal hemianopia  
G. Superior nasal hemianopia  
H. Superior nasal hemianopia  
I. Superior nasal hemianopia  
J. Superior nasal hemianopia

This question is PARSED with LMS question #3. It requires application of the content tested in LMS question #3.

4. Assuming the phenomenon is not atypical, your working distance is 60cm, your axial reflex is 100%, and your accommodative response is 100%, what is the most likely cause of the patient's blurred vision?

A. With movement  
B. Against movement  
C. Neutral

This question is PARSED with LMS question #5.

5. Considering the refractive results, which of the following IBA/PEA results are most likely?

A. 0.75/0.75  
B. 0.75/0.75  
C. 0.75/0.75  
D. 0.75/0.75  
E. 0.75/0.75  
F. 0.75/0.75  
G. 0.75/0.75  
H. 0.75/0.75  
I. 0.75/0.75  
J. 0.75/0.75

Just as we saw with LMS question 5, this question serves to remind students that patients in clinic often present with multiple problems that involve both knowledge of binocular vision and vision.

6. Which of the following is the most appropriate approach to balance accommodation?

A. 300/300  
B. 300/300  
C. 300/300  
D. 300/300  
E. 300/300  
F. 300/300  
G. 300/300  
H. 300/300  
I. 300/300  
J. 300/300

This question is PARSED with LMS question #6. This question also demonstrates that questions can be PARSED to different degrees.

## CONCLUSIONS

Primary Care Conference provides the tools and experiences necessary to improve students' ability to think clinically and apply didactic knowledge to clinical patient care through hybrid project-based learning. The wide variety of cases aid students in identifying areas of weakness in their current knowledge base while the open book nature of PCC allows students to become more familiar with quality reference material and how to access information effectively, which is a critical skill for life-long learning.

Primary Care Conference also provides many benefits to the faculty members involved. It aids in the development of junior faculty, increases their exposure to students, expands clinical knowledge, reinforces importance of basic science, and aids in curriculum vitae building.

Despite the lack of true outcome measures of a change in student clinical thinking aptitude, ICO has adopted this approach based on feedback and general impressions. Problem-based learning has been well studied and has a strong history of positively impacting students,<sup>4,5,6</sup> thus we are pleased that we have discovered a novel format that will allow us to provide our students with the benefits of problem-based learning but in a large class setting.

## KEY WORDS

learner-centered, problem-based learning, critical thinking

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