THE RETINA TIMES

EDUCATION. EXPERIENCE. COMPASSION.

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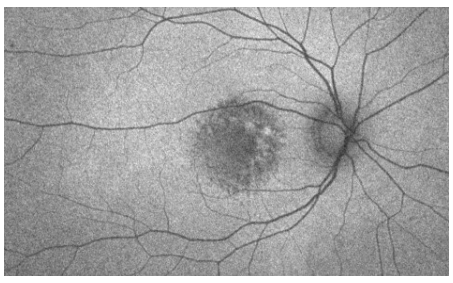
CASE STUDY

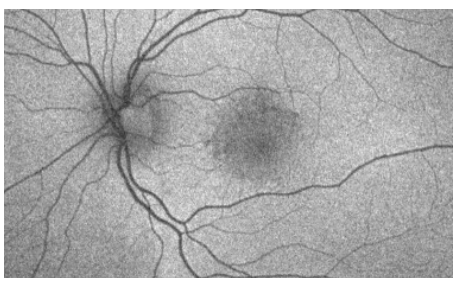
A 34-year-old Caucasian female presented to Retina Associates of Kentucky for blurry vision in both eyes, right eye greater than the left, for the past year, worse at night, causing her to stop driving at night. She also had some floaters in the right eye for the past 2 weeks.

She has a past surgical history of gallbladder removal and a medical history of cystitis. She is a former smoker. Systemic medications include Adderall, Estarylla, Iron, and probiotics.

Her visual acuity is 20/20 in both eyes, with intraocular pressure of 16 in each eye. The slit lamp exam was normal. Her dilated fundus exam showed significant macular changes, including drusen, retinal pigment epithelium (RPE) mottling and hyperpigmentation, and retinal atrophy centered on the fovea in both eyes symmetrically.

Fundus autofluorescence showed areas of predominantly hypoautofluorescence in the macula with small areas of hyperautofluorescence surrounding the fovea in a circular fashion corresponding to the area of atrophy on exam.





OCT showed changes in the RPE resembling drusen OU with focal disruption of the ellipsoidal zone.

She had genetic testing performed which showed non-specific autosomal recessive carrier mutations not related to her retinal pathology.

On a follow up visit, the patient was asked if she had a history of cystitis, or had ever been prescribed Elmiron (pentosan polysulfate or PPS). She recalled a remote history of using this, and after checking her records, found that she was on this medication for 4 years, but had discontinued its use over 5 years ago. Given that she had no apparent genetic basis for her macular disease, and she had a consistent history of Elmiron use, we diagnosed her with Elmiron toxicity.

What is Elmiron?

Interstitial cystitis and bladder pain syndrome are chronic urological pain syndromes that are very challenging to treat with a small number of remedies. The only FDA-approved oral medication for treatment is Elmiron, or pentosan polysulfate sodium (PPS). Elmiron is a synthetic polysaccharide that reduces irritation by adhering to the epithelium of the bladder, producing a protective layer against components of urine.

What is Elmiron Retinal Toxicity?

The ocular toxicity of Elmiron was first described by Pearce et al in 2018. It is also known by the generic name of the medication, Pentosan Toxicity. They described a distinctive pigmentary maculopathy affecting patients who were found to have abnormalities of the retinal pigment epithelium. The patients' symptoms included issues with adjustment to darkness and difficulty reading. All patients had a diagnosis of interstitial cystitis and were being treated with oral Elmiron.

It is unclear how the length of time or the dose of exposure correlates with the degree of maculopathy. Any patient with a current or history of Elmiron use with visual complaints should be screened for maculopathy and should be followed regularly. Like our patient, many patients in the literature with Elmiron maculopathy had stopped the medication in the past so even past exposure is significant.

What are the findings on imaging with Elmiron toxicity?

Fundus autofluorescence (FAF) shows speckled autofluorescence in the posterior pole with peripapillary extension. Optical coherence tomography (OCT) exhibits focal RPE thickening and atrophy in the macula and the periphery.

What are some of the other medications that may be toxic to the retina that we should always consider?

Pigmentary Retinopathy:

- Quinolines: Chloroquine and hydroxychloroquine (Plaquenil)- used to treat rheumatologic diseases and malaria.
- Deferoxamine (Desferal)- an iron-chelating agent used to treat conditions with excessive serum iron levels
- Thioridazine (Mellaril)- antipsychotic medication

Macular edema:

- Latanoprost
- Niacin, or vitamin B3
- Epinephrine or Dipivefrin

For our patient, as she was being followed in the clinic, it appears that her disease was worsening. Although her visual acuity remained stable, her maculopathy worsened on exam and on imaging. The fundus photos showed increased hyper pigmentation of the macular lesion. Fundus autofluorescence showed increased areas of hyper autofluorescence. OCT showed increased areas of atrophy and RPE thickening.

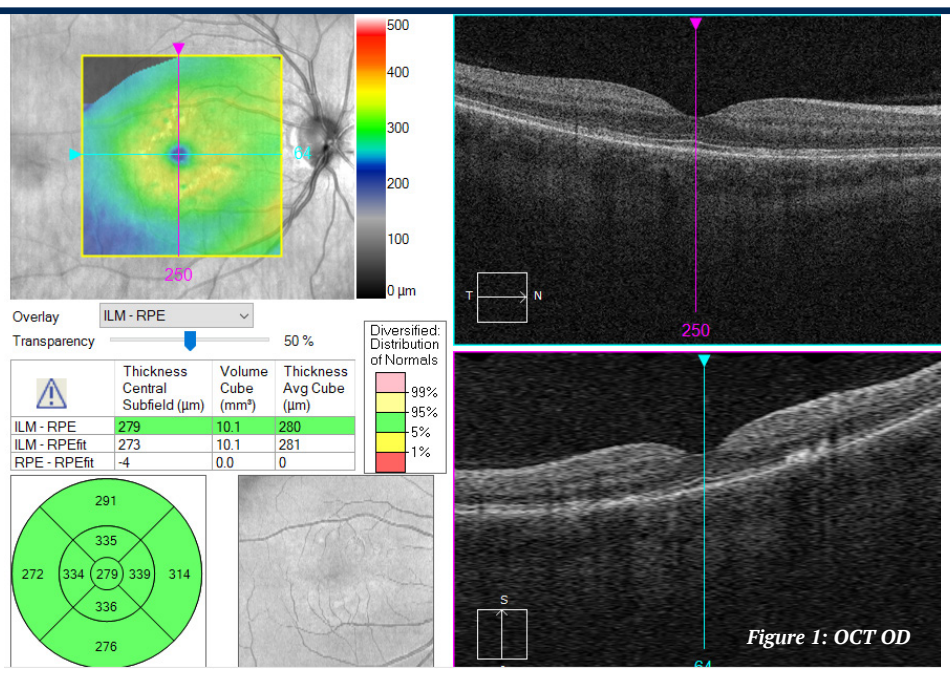


Figure 1: OCT OD

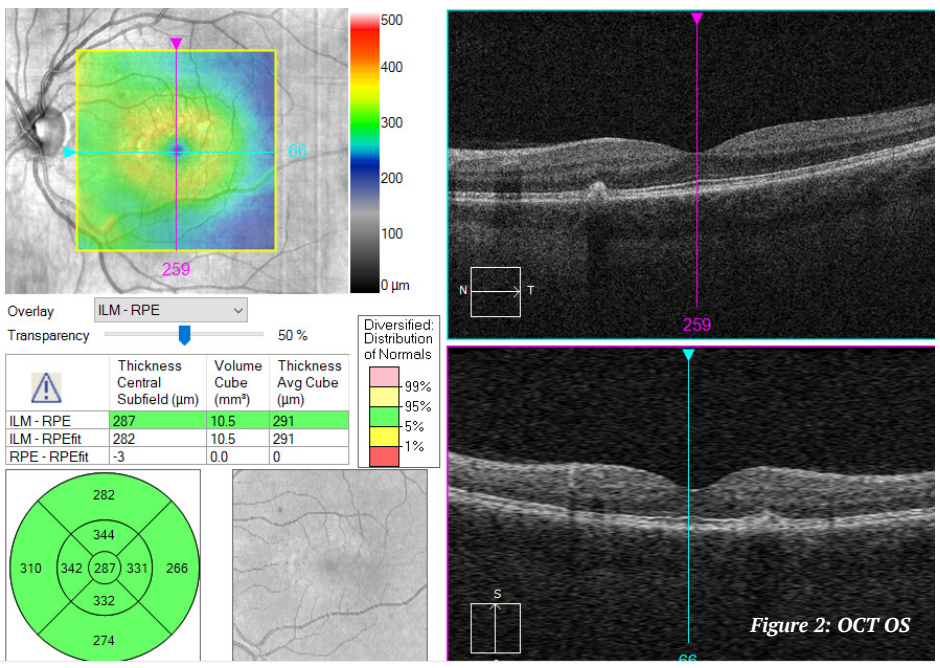


Figure 2: OCT OS

How should I treat and follow patients on Elmiron?

As the specific cause of the toxicity is unknown, there is no currently available treatment for these patients beyond stopping the medication. While research is ongoing, the long term clinical course of patients with this condition is unknown, even if they stop taking Elmiron.

As eye doctors, it is important to recognize this toxicity clinically, to minimize the loss of visual function in those who are affected. At this point, patients who take Elmiron who have no visual complaints and no retinal findings do not need to stop taking the medication, but need to be screened. Exact screening guidelines are being developed, but evaluation twice a year is a typical retinal screening protocol that would be appropriate for patients on this medication until further guidance is determined.

Figure 3: Fundus photo on last follow up right eye

Figure 4: Fundus photo on last follow up left eye

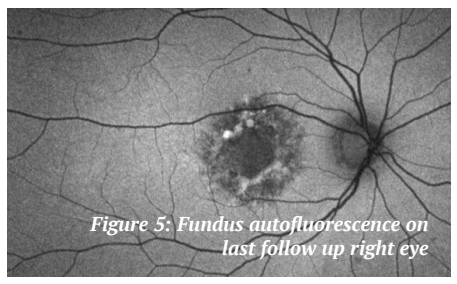


Figure 5: Fundus autofluorescence on last follow up right eye

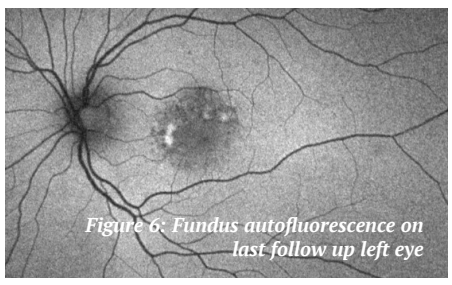


Figure 6: Fundus autofluorescence on last follow up left eye

Submitted by

Thomas W. Stone, MD and Osama Sabbagh, MD

References:

Doiron RC, Bona M, Nickel JC. Possible drug-induced, vision-threatening maculopathy secondary to chronic pentosan polysulfate sodium (Elmiron®) exposure. Can Urol Assoc J. 2020;14(2):10-11. doi:10.5489/cuaj.6401; Pearce WA, Chen R, Jain N. Pigmentary maculopathy associated with chronic exposure to pentosan polysulfate sodium. Ophthalmology. 2018;125:1793–1802. doi: 10.1016/j.ophtha.2018.04.026.; Omar Abbasi, M., & Asheesh Tewari, M. (2009, June 11). Common Medications That May Be Toxic to the Retina. Retrieved August 24, 2020, from https://www.reviewofophthalmology.com/article/common-medications-that-may-be-toxic-to-the-retina

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A NOTE FROM DR. STONE

Dear Colleague,

I know many of you have found your new rhythm caring for patients during the COVID pandemic. We have several updates to share with you as to how we're managing.

Starting with a provider update, our Founder William J. Wood, MD has retired and his patients are being seen by the other doctors in our practice. In addition, Rick D. Isernhagen, MD has become a part time physician, seeing patients one or two days a week. While some of his existing patients will still see him, because he is here less often, we may ask his patients to see other doctors. With these departures, our other physicians have to care for more patients. while we maintain social distancing. As a result, we've made some necessary changes to our Lexington and Louisville offices to streamline patient care.

In Lexington, we have closed our Low Vision office and will no longer see Low Vision patients. We will share a list of statewide resources with our LV patients and will continue selling devices in a limited manner at check-out stations. If your practice offers Low Vision services, please communicate with Kristin Willard (kwillard@retinaky.com) so that we can add your practice as a resource on this statewide listing.

With some of our non-patient care staff working remotely, we have converted our administrative office space at N. Eagle Creek on the 4th floor to a 1-doctor clinic to follow social distancing guidelines. We have our COVID screening team located in the lobby of our N. Eagle Creek offices and they'll guide patients to the applicable office for their appointment.

In the Louisville area, we are excited to expand our reach to Southern Indiana, opening a new satellite in Jeffersonville. Dr. Todd Purkiss and I will be staffing this new office, with our clinical team from Louisville. I am also performing surgery in New Albany Indiana to help keep the Southern Indiana patients close to home for their retinal health needs.

To our Shelbyville colleagues, we are closing our Shelbyville satellite but are happy to serve these patients nearby in our Frankfort office or Louisville branch office. Our frequency is greater in Frankfort, and full-time in Louisville so hopefully this will offer your patients more flexibility with scheduling.

We ask for your support and patience with us as we navigate through these changes. We've always tried to accommodate specific requests when scheduling patient appointments, but during this time as we there may be times where we cannot meet your specific request. We feel fortunate to have a culture here where we physicians share similar views in practicing so that you and your patients will feel the same excellent care and compassion regardless of who the patient sees or the location in which the patient is seen. We're following the COVID precautions in all our offices to minimize the spread. Thank you for trusting us with your patients retinal health.

Thomas Stone, MD
President

UPCOMING CE WEBINAR

Retina Associates partners with

University of Pikeville UPIKE Kentucky College of Optometry

to co-host CE via Zoom Webinar
(3) Credit Hours

11.9.20 • 6-9pm

CORONAVIRUS (COVID-19) Precautions for Safeguarding Our Patients

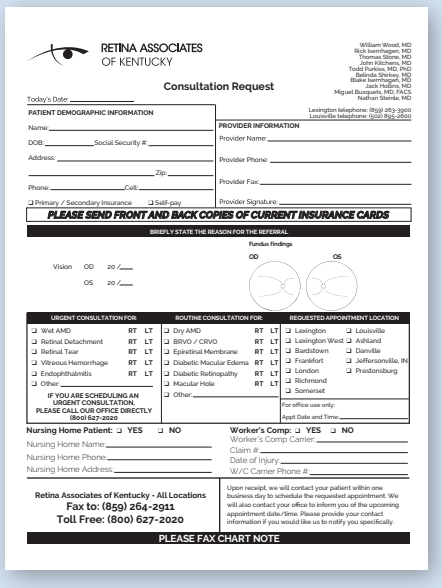
Retina Associates of Kentucky has been open, originally seeing urgent and emergent conditions and now resuming care for our established patients. We are following state and CDC Guidelines to create a maximally safe experience for our patients, staff and physicians.

We have implemented additional precautions in an effort to minimize the spread of this virus.

- We are taking patients' temperatures at point of entry, and physicians & staff at the start of their shift.
- We ask that patients wear a protective mask (surgical or cloth) to their appointment and likewise all RAK physicians and staff will have masks on and other protective gear, as necessary. Protective masks should be worn properly covering nose and mouth to help minimize exposure.
- We ask patients to arrive 15 minutes before their appointment time to help us follow social distancing guidelines. If patients arrive earlier than 15 minutes, they will be asked to wait in their car until closer to the appointment.
- A Retina Associates representative will greet patients at point of entry with a risk survey regarding COVID-19 symptoms, recent travel and will be checking patients' temperature.
- We may ask that patients to wait in their car until their appointment time to follow social distancing guidelines.
- During this time, we have restricted entry to patients only. If patients require assistance, they should call our office (800) 627-2020 prior to their appointment to discuss their needs.
- As we resume care for our established patients, there will be increased demand for appointments. As we follow the social distancing guidelines we have fewer patient appointment openings. **During this time, we are unable to guarantee patient appointments with a specific doctor in our office.** We may change the time, date, location or doctor to accommodate appointments during this time.
- If patients are on a treatment plan, we ask that they keep their appointment, as many of our patients need this treatment to preserve vision. We have and will continue following state and CDC Guidelines for their safety.
- Patients will receive an email or text message with a link to pre-register in the comfort of their own home, using their own device through our self check-in process (Phreesia).
- A Retina Associates representative will call all patients the day before their appointment with a risk survey regarding COVID-19 symptoms and recent travel.
- Patients will be asked to reschedule an appointment if they or a household member have been diagnosed with COVID-19, have a fever, have traveled internationally or on a cruise, have traveled domestically to a hot spot or have had any other COVID-19 symptoms.

We will continue our frequent communication on our Facebook page and Website as new information becomes available. We are always here if you need us, (800) 627-2020. Thank you for trusting us with your retina patients. We look forward to warmer brighter days with you and hope you and your families stay well!

REFERRAL COMMUNICATION WITH OUR OFFICE



RETINA ASSOCIATES OF KENTUCKY

Consultation Request

Today's Date: _____

PATIENT DEMOGRAPHIC INFORMATION
Name: _____
DOB: _____ Social Security # _____
Address: _____
Zip: _____
Phone: _____ Cell: _____
❑ Primary / Secondary Insurance ❑ Self-pay

PROVIDER INFORMATION
Provider Name: _____
Provider Phone: _____
Provider Fax: _____
Provider Signature: _____

PLEASE SEND FRONT AND BACK COPIES OF CURRENT INSURANCE CARDS

BRIEFLY STATE THE REASON FOR THE REFERRAL

Fundus findings
Vision OD 20 /___ OS 20 /___
OD OS

URGENT CONSULTATION FOR			ROUTINE CONSULTATION FOR:			REQUESTED APPOINTMENT LOCATION	
❑ Wet AMD	RT	LT	❑ Dry AMD	RT	LT	❑ Lexington	❑ Louisville
❑ Retinal Detachment	RT	LT	❑ BRVO / CRVO	RT	LT	❑ Lexington West	❑ Ashland
❑ Retinal Tear	RT	LT	❑ Epiretinal Membrane	RT	LT	❑ Bardstown	❑ Danville
❑ Vitreous Hemorrhage	RT	LT	❑ Diabetic Macular Edema	RT	LT	❑ Frankfort	❑ Jeffersonville, IN
❑ Endophthalmitis	RT	LT	❑ Diabetic Retinopathy	RT	LT	❑ London	❑ Prestonsburg
❑ Other:	RT	LT	❑ Macular Hole	RT	LT	❑ Richmond	
			❑ Other:	RT	LT	❑ Somerset	

IF YOU ARE SCHEDULING AN URGENT CONSULTATION, PLEASE CALL OUR OFFICE DIRECTLY (800) 627-2020

For office use only:
Appt Date and Time: _____

Nursing Home Patient: ❑ YES ❑ NO
Nursing Home Name: _____
Nursing Home Phone: _____
Nursing Home Address: _____

Worker's Comp: ❑ YES ❑ NO
Worker's Comp Carrier: _____
Claim #: _____
Date of Injury: _____
W/C Carrier Phone #: _____

Retina Associates of Kentucky - All Locations
Fax to: (859) 264-2911
Toll Free: (800) 627-2020

Upon receipt, we will contact your patient within one business day to schedule the requested appointment. We will also contact your office to inform you of the upcoming appointment date/time. Please provide your contact information if you would like us to notify you specifically.

PLEASE FAX CHART NOTE

As many of you are adapting to a new normal, we would like to take this opportunity to review the Referral Process with our office. We encourage you and your staff to use our Consultation Request form or the online feature for communicating details of the patient when you refer them. Our physicians feel that this form, along with your chart note, will optimize the communication between our offices and ultimately create a better experience for our mutual patients.

Check out the Consultation Request Online feature:

With regard to scheduling, we are always happy to work-in true emergencies same day/next day. However, during this time, any non-emergent referral will be scheduled in relation to their diagnosis.

Our primary goal is to take excellent care of our mutual patients, and we are hoping our renewed emphasis on the Referral Process will help us do that while streamlining the communication for you and your team.

If you have any questions regarding communication with our office, you may call Kristin Willard (502) 649-3681. We'd be happy to give a tutorial of the Online Consultation feature or review the process in its entirety with your team, we can arrange to do so virtually or in-person with PPE!

We are excited about reconnecting with you and streamlining the referral process with our office!

RESEARCH

If you are interested in information regarding past clinical trials or participation criteria in our current clinical trials, please contact our research department:
Diana Holcomb - Clinical Research Manager
PH (859) 264-2905 | dholcomb@retinaky.com

MAIN OFFICES

Lexington
120 N. Eagle Creek Drive, Suite 500
Lexington, KY 40509

Louisville
6420 Dutchmans Parkway, Suite 70
Louisville, KY 40205

Ashland
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Ashland, KY 41101

OUR OTHER LOCATIONS

Kentucky
Bardstown
Danville
Frankfort
Lexington - West
London
Prestonsburg
Richmond
Somerset

Indiana
Jeffersonville

OUR PHYSICIANS

William J. Wood, MD, Founder (Retired)
Rick D. Isernhagen, MD
Thomas W. Stone, MD
John W. Kitchens, MD
Todd J. Purkiss, MD, PhD
Belinda L. Shirkey, MD
Blake A. Isernhagen, MD
Jack L. Hollins, MD
Miguel A. Busquets, MD, FACS