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A Nagging Question...

Paul B. Freeman, O.D.

Since I began practicing low vision rehabilitation in the middle 1970s, there has been an ever-increasing realization that there is a need for these services, and that the need is expected to continue to grow, primarily due to the ocular disease processes associated with the aging population. That said, since I began practicing low vision rehabilitation, and to this very day, there is one nagging question that seems to always escape a rational answer: Where are all the visually impaired people? Where are those people who we know would like the opportunity to function better visually, and why are they not being given the opportunity to do so? Unfortunately, I am not alone in the quest for an answer. Recently, and serendipitously as I was putting the final touches on this editorial, there was an email post to a low vision interest listserve, in which a physician who does low vision rehabilitation commented, “If you asked me what presents the most difficulty in treating a low vision patient, I would have to answer that it is the fact that they don’t get referred!” As I travel around the world lecturing on low vision rehabilitation, and in talking to my colleagues, I find, and they do too, that this is the most difficult part of providing low vision rehabilitation: the patients are not referred for the help we can provide.

Years ago, according to the National Eye Institute, one of the reasons for lack of low vision rehabilitative care was the lack of knowledge on the part of professionals and lay people about the services available. The reality today is that optometry and ophthalmology both have written guidelines that are not considered out of the mainstream of eye care that admonish eye care providers to seek vision rehabilitative services for their patients when conventional optics or medical intervention can no longer satisfy the functional needs of the patient. The lay public is also well aware of pathological eye conditions that can cause a visual impairment (most notably macular degeneration). Such conditions regularly appear in the news media. In fact, recently it was reported that the actress Dame Judith Dench has age-related macular degeneration. Refreshingly, it was also reported, she said that she is not going “blind” and has lenses and glasses and “good old bright light to help her see,” although she has difficulty seeing faces and has people read her scripts to her. The curiosity here is what type of low vision devices she has, and why, based on the fact that both she and her mother have experienced this condition, would she not be asked to become a spokesperson for vision rehabilitative services.

I am not suggesting all eye care practitioners do not refer visually impaired patients, but I am curious about how the selection process works. So, what presumably might be some reasons for the lack of referrals of visually impaired individuals by eye care professionals?

1. **Many visually impaired patients seem not to have pressing visual needs.** This erroneous thinking is based on how the questions are asked of the visually impaired patient: “How are you doing? Are you having any problems?” The answer to those “superficial” questions is typically… “no problem” (implied: “for my age”). Unfortunately, the next level of questioning is not typically asked, undoubtedly for many reasons, i.e., the doctor is too busy to deviate from the task at hand (the medical reason for the visit), or the patient was not forthcoming with a complaint, so “everything must be all right.” Often I ask the same initial question to a visually impaired patient who is referred to me, “How are you doing?” And the response is typically “OK.” However the next question I ask is “How is your reading?” This evokes a completely different response: “I want to be able to see better to read, to do my own checks, etc.” or the equivalent of the conversational flood gate that most other practitioners are not ready to engage in, and to have that chat tie up a “busy day.”

2. **Professionals are too busy, and often, after they have completed their (medical or surgical) service, do not “remember” to recommend further care, or do not want to get into a lengthy conversation about visual rehabilitative alternatives.** Although there is no magic bullet for a resolution to this concern, i.e., “tying up” a busy practitioner with a seemingly endless list of questions, a place to start might be using handout materials (prepared by a low vision service provider or the AOA’s pamphlet written by the Low Vision Rehabilitation Section) explaining low vision rehabilitation services, or using ancillary staff who are given specific guidelines for suggesting additional care. This will, at least, give the patient or family member information to ponder. Identifying those practitioners who provide low vision care, and explaining that the patient can get most of the answers to their questions from them, and then indicating how to reach them, might be all that is necessary to offer the patient a valuable service that might otherwise have gone unrecognized.
Professionals feel if they can’t do everything to help their patients, they might be perceived as failing in their healthcare responsibilities. A number of people have presented this as a subconscious rationale for not referring patients (which I find hard to believe). It seems to me this is somewhat analogous to deciding that when a patient has to be referred for cataract surgery, it suggests that the referring practitioner has failed in being able to help that person see clearly with conventional lenses. This is an obviously preposterous statement!!

Patients will not wear or use the devices recommended as they are not conventional in appearance or in their use, and most are paid for with out-of-pocket dollars, making patients potentially resistant to pursue this type of care. In discussing this with professionals or lay people I use 2 very simple rules. First, do no harm. Second, once the patient has been educated, let the patient decide! In fact, not all patients who have a visual impairment have a visual disability, or need intervention. In a 1994 article, John Tanton, M.D. vividly explained it this way: “Providing rehabilitation services for their visually impaired patients is the medical and moral…responsibility of all ophthalmologists. It is no more acceptable for an ophthalmologist to abandon a patient once the medical treatment is completed but before needed rehabilitation services have been provided, than it would be for an orthopedic surgeon to abandon an amputee to hopping around on one leg.”

Optometry should heed this reproach as well. Patients will make their own decision, once they are given the options, but without those options, they do not have a full complement of choices. And, until any one of us has had the experience that our patients have, imposing our bias is unfair.

When one looks at the perceived dearth of referrals of visually impaired patients for rehabilitative care, the only apparent option low vision providers have left (other than to continue to educate eye care professionals) is to go to others in the medical community whose health care culture is more in tune with offering “ancillary” or rehabilitative services to their patients, i.e., gerontologists, diabetologists, primary care doctors, physiatrists, etc., or to go directly to the public to educate them in a more aggressive way, trying to increase awareness of rehabilitative services and their obvious benefits. The numbers tell us there is a need; the demographic is aging.

And the sooner, the better, for lots of reasons. In a recent small pilot study it was reported, “early intervention using low-vision aids can greatly reduce the severity of depressive symptoms related to the vision loss experienced by people with age-related macular degeneration.” I suppose this applies to any other pathology that can cause a visual impairment. As my colleague from Michigan said, “Even though it may be true that nothing more can be done for the eye, it is almost never true that nothing more can be done for the patient.”

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Our profession lost a great leader and educator in Irvin M. Borish. O.D., DOS. The following is the eulogy that was presented on March 11, 2012, by Dr. Alden N. Haffner, O.D., Ph.D.

Reverend clergy, members of the Borish family, colleagues and friends who adored and respected Dr. Irvin M. Borish:

All of the obituary notices that have been published, or those that will be published, bear the cold accuracy of facts assembled to describe and chronicle Dr. Borish’s long and enormously productive life. And the facts of his lifetime activities are, without any doubt, impressive and inspiring. But, I will not repeat them at this memorial service this afternoon. In fact, there are, in my view, much larger meanings than the recitation of the facts of his life.

I last spoke to Irv about three weeks before he died, and well after his 99th birthday. He pointedly reminded me that I traveled to Boca Raton for his 95th birthday celebration and that I addressed the happy crowd at that event. He then asked (it sounded more like a directive) me to prepare two presentations -- one to celebrate his 100th birthday and the other his eulogy. I made a commitment to do both. Regrettably, only one will do.

After all of the details of his eventful life are read, and their meaning absorbed, 3 facets stand out. The first, and most significant, is that he gave scientific and professional legitimacy to our profession, optometry. For many decades since its initial publication, CLINICAL REFRACTION by BORISH was the fundamental text that unified optometric practitioners throughout the United States and, moreover, in a host of countries throughout the world. It was used as the standard text, and it brought philosophical-scientific unity to optometry. This was, in my view, a monumental achievement. Its impact on optometry cannot be judged as anything less than fundamental and progressive.

Irv was, throughout his career, deeply concerned about the education of optometrists. Indeed, in his lifetime, he was intimately involved in the affairs of three major institutions: the Illinois College of Optometry, the Indiana University School of Optometry, and the University of Houston College of Optometry. The force of his intellect and the force of his personality, both awesome, influenced the leadership of those splendid houses of learning. He urged, cajoled, argued, persuaded and, ultimately, convinced the respective leadership groups that scientific standards should be under constant review in pedagogy. As a person who spent 35 years in a great university, I can attest that Irv Borish’s institutional contribution was to pull optometric education away from vocational education and forcefully to advocate for professional education as the model for our discipline. This contribution was incalculable in importance; that he did so much of this while still engaged in private practice [Kokomo, Indiana] bespeaks the expenditure of enormous personal energies. Irv’s commitment was total and unflagging.

In the next few months, many colleagues in our profession will come forward with personal stories about their encounters with Irv. They will all be different but, many, remarkably the same. His abiding humanity, his ability always to be the teacher, his profound love for his profession, and his thirst for current knowledge about “what’s going on” were hallmarks of his dynamic personality. But, I have a different, and very personal, story to recount. In the mid-1970s, the American Academy of Optometry was holding its annual meeting at the Waldorf-Astoria Hotel in New York City.

On a Sunday morning, I was delivering a paper on public health. While speaking, Dr. William Baldwin came running down the aisle and interrupted my presentation while shouting at me, “Irv is very ill. Come at once.” After turning my paper over to the moderator to complete its reading, I raced with Bill to Irv’s room. Of course, Bea, his loving, devoted (and very tolerant) wife, and partner for more than 60 years, was in a state of anxiety about Irv’s physical distress. (In fact, he was in the midst of a massive coronary dysfunction.) I immediately telephoned my personal internist/cardiologist, Dr. Jerome Schack. Luck was with us because I reached him at Beth Israel Medical Center where he was making rounds.

He advised that we dress Irv warmly, secure a taxicab, and he would meet us in the hospital. Bea, Irv, and I arrived at the hospital within 20 minutes, and Jerry Schack saw him immediately. I sat with Bea until we were notified that Irv promptly was moved to the Cardiac Intensive Care Unit. The seriousness of his illness, and the complications that ensued, required more than a month’s hospital stay. Bea stayed with me at my home, though there were daily hospital visits. Naturally, Irv was frequently conducting seminars with the hospital staff on all sorts of subjects. When he was finally on the mend and discharged, he joined Bea in my home for about 10 days until he was given permission to fly home to Indiana.

About a month later, Bea and Irv called me (though we spoke frequently after their return home) to tell me that they wanted to get me a gift as a token of their appreciation. I resoundingly refused, which provoked a prolonged argument. A few days later, Irv called to say that he was going to execute a painting (he was a prolific artist), and that he and Bea had fixed on that idea. I (graciously) accepted the offer.

About 6 months passed and, one day, the painting arrived. It was a beautiful watercolor of the skyline of lower Manhattan viewed from the Hudson River. But there was something very curious about the skyline. The World Trade Center twin towers were ”broken” and incomplete at the top. When I asked about the towers, Irv’s response was, “well, I saw it that way.” The painting, mounted and framed, still hangs in my study.

The September 11th tragedy, some 35 years after the painting’s creation, served as an eerie and stunning surprise. There still is some mystery about Irv’s conception. The Essilor Company published a book of Irv Borish’s artistic work. The painting of the Manhattan skyline with the broken towers introduced the publication. While Indiana University and the University of Houston have asked for the donation of the painting, it still hangs in my study with its enduring enigma.

I had enormous affection and admiration for Irv and for Bea. He was an extraordinary human being who, without doubt, achieved extraordinary accomplishments. And, our profession and generations of optometrists are in his debt as well. But the welfare of humanity ultimately benefited. One of my heroes, the great Oliver Wendell Holmes, in 1885, prophetically said, “As life is action and passion, it is required of a man that he share the passion and action of his times, lest he be judged not to have lived.” To the members of the Borish Family, I ask what more can be said of any human being!

Abnormal microcirculation of the optic nerve head is an underlying variable contributing to the pathophysiology of primary open-angle glaucoma (POAG), normal-tension glaucoma (NTG), and the pathogenesis of optic disc hemorrhages. The retina and the nailfold of the digits are areas where microcirculation can be observed without performing an invasive procedure. The authors seek to determine if specific nailfold capillary changes are associated with glaucoma.

This observational trial included 108 Korean patients recently diagnosed with glaucoma (86 with NTG, 22 with POAG) and 38 age-matched controls. Patients with glaucoma had glaucomatous optic nerve changes, visual field loss, and open angles on gonioscopy. Glaucoma classification was defined by an intraocular pressure (IOP) above (POAG) or below (NTG) 21 mmHg on different days, without the influence of topical medications. Patients who were identified to have a disc hemorrhage or localized RNFL defect were photographed in color and through a red-free filter.

All subjects underwent a complete physical examination, blood pressure assessment, and nailfold capillaroscopy of the second, third and fourth digits of both hands. The capillary architecture, morphological characteristics, distribution and number of capillary vessels, and presence of splinter hemorrhages were documented.

Analysis of nail bed capillaroscopy revealed 60 (55.6%) patients with tortuous vessels, 38 (35.2%) patients exhibited avascular areas, and 21 (19.4%) had nail bed hemorrhages. A statistically significant difference existed in the presence of nail bed hemorrhages between the glaucoma and control patients. However, there was no difference in the nail bed characteristics between the NTG and POAG patients.

Within the glaucoma group, a disc hemorrhage was found to correlate significantly with avascular areas and hemorrhages in the nail bed. Within the same cohort of subjects, a RNFL defect was significantly associated with avascular zones in the nail bed. Adjusting for age, gender, and IOP, only nail bed hemorrhages were significantly associated with disc hemorrhages.

Nail bed capillary abnormalities are found to be greater in glaucoma patients, specifically in the NTG subgroup, when compared to control subjects. When analyzing the differences between the NTG and the POAG subgroups, evidence suggests NTG patients had greater abnormalities in microcirculation. The authors conclude patients with glaucoma, and with disc hemorrhages, have significant peripheral vascular insufficiency, and that disc hemorrhages are due to these changes and are independent of IOP.

Mark H. Sawamura, O.D.
Duty to Educate

Samuel D. Pierce, O.D.

In 1979, Michael Aldrich invented online shopping.¹ In 1995, Jeff Bezos launched Amazon.com, and in 2011, U.S. Ecommerce and online retail sales were projected to reach $197 billion, a 12% increase over 2010.² There is no denying the Internet and Ecommerce have become an integral part of our existence, both in our personal and professional lives. We use the Internet in our offices to verify coverage and benefits, to market our practices, to order supplies and inventory, for Electronic Medical Records (EMRs), to blog, to email and to update our Facebook or Twitter status.

Our patients use the Internet to order theater tickets, buy books, music, and clothes. Some of our patients order their contact lenses online, and now it seems more and more are ordering their eyeglasses and sunglasses online. The Vision Council estimates between 1.8 and 1.9 million people ordered prescription eyewear in a 12-month period ending September 2010.³

Other than the possible financial impact, are there any other consequences to consumers ordering spectacles online? At the onset, it sure sounds like it would be easier for patients to browse thousands of frames whenever it’s most convenient for them. Then click, click, click, a new pair of glasses are on their way. But what are the realities?

We educate our patients about all the intricacies of acquiring a pair of glasses, ensuring frame selection to ensure suitability to the prescription, insuring accurate measurements, making proper lens choices, etc. We can explain the difficulties of achieving this when ordering online, but is there more to it than that? Are there any safety concerns when ordering glasses online? What are the chances the prescription will be accurate? Prior to the “Safety and compliance of prescription spectacles ordered by the public via the Internet” study published in the September issue of Optometry,⁴ we had no answers to those questions. Now we do. When a patient asks, “What’s wrong with ordering glasses online?” we can refer to the study and inform them that more than 44% of glasses ordered online could have a problem with either the accuracy of the prescription or not meeting minimum U.S. standards for impact-resistance. But are there other concerns with online opticals?

The mere fact this study could take place should raise a red flag. Think about it----orders from fictitious patients with fictitious prescriptions, and the transactions took place with rare verifications, and no consideration of state laws mandating the dispensing of spectacles by licensed professionals. So what keeps consumers from bypassing scheduled eye examinations and just ordering glasses online, tweaking the Rx as they feel (-2.00 D not so good, maybe -2.50 D or -3.00 D would be better)? Apparently nothing, and this is why it falls to us as health care professionals to continually educate our patients about the importance of eye exams, which we all know are vital for better ocular and systemic health, and not just prescription accuracy.

This is not an indictment against all online opticals. Many optometrists have established an online presence in an attempt to offer a greater variety of options to their patients. In these environments the doctor still has control over the prescription, the quality of the materials used, and the final product.

Unfortunately, there is no way for consumers to differentiate the websites that verify the prescription and ensure prescription accuracy, along with using materials that meet the requirements for impact resistance. Googling “online eyeglasses” gets close to 20 million results, with advertised prices as low as $6.95 for a pair of glasses. No doubt this is enticing to patients/consumers, and so, keeping them educated about the potential problems with accuracy and safety with ordering spectacles from “rogue” websites is paramount.

Is the federal government doing anything? The FDA recently updated Import Alert 86-07 to identify additional companies now subject to “Detention Without Physical Examination” (DWPE) relating to imports of impact-resistant lenses either as prescription lenses or as found in sunglasses, reading glasses and monocles.⁵ The companies listed are known to have shipped lenses into the U.S. that failed the impact-resistance test, or where the foreign manufacturer could not validate its impact-resistance test. Also, these shipments were accompanied by impact-resistance certificates stating the lenses passed the drop ball test, and therefore the FDA considers them misbranded and adulterated merchandise. Is this enough? Probably not! The FDA and DEA have been trying to safeguard the public from illegal online pharmacies for more than a decade, and, from the emails that still show up in my spam file, with little or no impact. For example, according to the DEA’s website, just one illegal online pharmacy caught in a drug bust was allegedly

¹ In 1995, Jeff Bezos launched Amazon.com.
² In 2011, U.S. Ecommerce and online retail sales were projected to reach $197 billion.
³ The Vision Council estimates between 1.8 and 1.9 million people ordered prescription eyewear in a 12-month period ending September 2010.
⁴ “Safety and compliance of prescription spectacles ordered by the public via the Internet” study published in the September issue of Optometry.
⁵ The companies listed are known to have shipped lenses into the U.S. that failed the impact-resistance test, or where the foreign manufacturer could not validate its impact-resistance test. Also, these shipments were accompanied by impact-resistance certificates stating the lenses passed the drop ball test, and therefore the FDA considers them misbranded and adulterated merchandise.
distributing approximately 2.5 million dosage units of Schedule II-V pharmaceutical controlled substances, anabolic steroids, and amphetamines per month until shut down by the DEA. In fact, cyber-security experts at Carnegie Mellon University said infected websites are redirecting online shoppers to dangerous unauthorized pharmacies. Nicolas Christin, associate director of the university's Information Networking Institute, said in a Carnegie Mellon news release, "We have known for some time that unauthorized online pharmacies have been using email spam to tap the wallets of unwary online consumers, but that method did not blanket enough customers so now the online thieves are infecting websites to redirect unwary consumers to hundreds of illegal online pharmacies." 

We cannot solely rely on the government or industry to police the Internet and protect our patients and the general public; the task is very daunting. It is going to take each one of us talking to our patients, building trust and developing strong relationships. Out of the strength of those relationships we can educate and protect our patients by making them aware, and keeping them informed of the risks of unsupervised online shopping. It's our job...that's what we do. We are optometrists, doctors on the frontline of eye and vision care.

References

A comparison of short and long reading passages in symptomatic vs. asymptomatic subjects


KEYWORDS
The College of Optometrists in Vision Development Quality of Life questionnaire; Fatigue; Ocular motor dysfunction; ReadAlyzer

ABSTRACT
Background: The visual system is necessary for reading. Understanding the mechanics of eye movements during reading can give insight into the reading process. The ReadAlyzer is an electronic recording system that measures eye movements while reading. Long passages on the ReadAlyzer of 800 words have recently been introduced as a tool to assess the efficiency of reading eye movements. Previously, short passages of 100 words have been used exclusively. This project was designed to determine if passage length influences the quality of reading eye movements.

Methods: Optometry students (N = 40) at Southern College of Optometry were separated into 2 equal groups (symptomatic vs. asymptomatic) based on the College of Optometrists in Vision Development Quality of Life questionnaire score. Each subject then performed two reading passage recordings with the ReadAlyzer: one short, one long. The order of the passages was alternated to reduce fatigue effects. Data were collected based on the ratio of fixations-to-regressions.

Results: A multivariate analysis of variance indicated that the difference between short and long paragraphs for the symptomatic and asymptomatic groups combined was significant (P = 0.001) but was not significant for the symptomatic group vs. the asymptomatic group (P = 0.651). Post-hoc comparisons using estimated marginal means indicated for asymptomatic, short vs. long, P = .036 and for symptomatic, short vs. long, P = 0.008. Additionally, for short length, symptomatic vs. asymptomatic, P = 0.242 and for long length, symptomatic vs. asymptomatic, P = 0.176.

Conclusion: This information indicates that both symptomatic and asymptomatic patients will have more difficulty on longer reading passages. This finding calls into question the use of shorter length reading tests to determine a diagnosis of ocular motor dysfunction and other visual efficiency problems.

Both children and adults are subject to reading difficulties. These reading difficulties are influenced by a myriad of disruptions in the visual system, leading to poor reading eye movements. Research has found 70% of the 2 million school-age children with reading difficulty have some form of visual dysfunction, such as oculomotor, binocular, or perceptual. Similarly, next to refractive conditions, the most prevalent conditions in the pediatric population are binocular and accommodative disorders. Oculomotor dysfunction (OMD) is a general term used to describe a visual system that has abnormal fixation, pursuit, or saccade actions. A list of symptoms can be found in Table I. The prevalence of oculomotor dysfunction has been found to be from 22.6% to 96% in a population of children with reading and learning difficulties. OMD is found in patients with traumatic brain injury, brain lesions, accommodation inefficiencies, and vergence inefficiencies, among other conditions. It is important the specific etiology is diagnosed and treated appropriately. When no organic cause is found, OMD can be considered developmental and functional in etiology. A sequential management approach includes optical correction for ametropia, added lens power for reading, and vision therapy.

Many tests and devices have been developed to assess the oculomotor system, including the Northeastern State University College of Optometry Oculomotor Test of Pursuits and Saccades, Pierce Saccades, Percon Saccades, King-Devick Eye Movement test (K-D), Developmental Eye Movement (DEM) test, Groffman Visual Tracing Test, and

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<th>Table 1</th>
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<td>Frequent loss of place</td>
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<td>Omission of words/skipping lines</td>
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<td>Poor reading comprehension</td>
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<td>Excessive head movement</td>
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<td>Poor sports performance</td>
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<td>Poor coordination</td>
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<td>Poor attention</td>
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<td>Vertigo/dizziness</td>
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<td>Difficulty with column-based math problems</td>
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<td>Trouble copying from the board</td>
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ReadAlyzer Visagraph. The Northeastern State University Oklahoma College of Optometry Oculomotor Test of Pursuits and Saccades is an oculomotor test that assesses pursuits and saccades based on the ability to attend to the task and the accuracy of ocular movements, as well as the amount of head and body movement. The scoring is based on a scale of 1 to 5. K-D replicates reading eye movements with randomly spaced numbers the subject reads as quickly and accurately as possible. Grading for the K-D is based on the number of errors and time taken to read 40 numbers aloud. The DEM is a visual-verbal test that identifies deficiencies in automaticity versus OMD. Its grading is based on number of errors, types of errors, and the time to complete each section (vertical and horizontal). A ratio of the vertical and horizontal times allows for the differential diagnosis. The ReadAlyzer and Visagraph are both instruments that use infrared

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light at the limbus to monitor eye movements while reading. This technology enables the equipment to objectively measure fixations, regressions, and reading rate.\(^{13}\,^{16}\,^{17}\)

Of the tests mentioned, only the ReadAlyzer or Visagraph is objective. The manufacturer of the ReadAlyzer provides passages of about 100 words (short passage) and 800 words (long passage) to be used during testing (see Figures 1 and 2). Each is available in increasing difficulty from level 1 to 10. From these measurements, computer analysis provides different eye movement parameters. Some of the measured parameters include fixations per 100 words, regressions per 100 words, and reading rate. The system calculates several values derived from the recorded measurements, including an equivalent grade level, span of recognition, and average duration of each fixation.\(^{13}\) The normative values based on age and grade level were completed by Taylor et al.\(^{20}\)

Measuring temperature is a way of determining whether one thing is warmer or colder than something else. When two objects are in contact, heat from the warmer one will flow into the cooler one until both are the same temperature. Temperature is the result of the activity of the atoms in an object or within a space. The greater the activity of the atoms, the higher the temperature. While there is no known maximum temperature, there is a minimum. If there is no movement to the atoms, the object has reached a temperature of absolute zero. Nothing can get colder than this. Scientists have tried to reach absolute zero but without success, getting only to within one-thousandth of a degree of absolute zero. One problem with such low temperatures is how to measure them, especially since everything freezes before reaching absolute zero. Magnets currently provide the most accurate way of measuring super-cold temperatures.

### Figure 1
An example of the short passage used during testing.

Colby et al.\(^{9}\) evaluated the repeatability of the Visagraph II in a group of 50 first-year optometry students. Each read 5 standard level-10 passages during each of 2 sessions. Between session correlations ranged from 0.15 to 0.90 with most falling within the 0.50 to 0.80 range. This indicated moderate to strong reliability.\(^{13}\)

Ciuffreda et al.\(^{16}\) evaluated 30 visually normal adults ranging in age from 20 to 59 years with a mean age of 28 years using the Visagraph II. All had a binocular visual acuity of 20/20 or better at distance and near, and none had self-reported reading disabilities or neurological/ocular/binocular problems that could interfere with reading performance. Each subject read 5 passages appropriate for the high school/adult level. Based on their results, it was suggested that at least 3 practice sessions be administered to ensure reliable and stable baseline performance. Borsting et al.\(^{17}\) investigated the repeatability of the Visagraph II in children in grades 3 to 8 ranging in age from 8 to 15 years. Subjects were correctable to 20/20 visual acuity at 40 cm, did not have a diagnosis of reading disability, read English fluently, and had no reported neurologic or ocular condition that would interfere with the recording.

The grade-level passage chosen was determined for each subject by using the San Diego Quick Assessment. Four trials were performed at each of 2 sessions, and no passage was used more than once. The first trial at each session was used as practice and not included in data analysis.

The intraclass correlation coefficient (ICC), used to evaluate within-session repeatability, ranged from 0.48 to 0.86 in session 1 and 0.15 to 0.90 in session 2. The lowest value for each range corresponded to comprehension questions. The ICC values for between-session repeatability for session 1 and 2 were as follows: number of fixations (0.71, 0.83), number of regressions (0.71, 0.82), span of recognition or words per fixation (0.77, 0.89), duration of fixation in seconds (0.75, 82), reading rate in words per minute (0.86, 0.90), and percentage correct on comprehension questions (0.48, 0.15). All values were considered to be strong, with the exception of the comprehension questions, which are considered moderate for session 1 and weak for session 2.

In looking at the between-session repeatability, no significant difference was found for any of the measures (P < 0.086 for all measures). The ICC was moderate to strong for all measures, ranging from 0.56 to 0.92. Reading comprehension was once again the lowest ICC. The 95% limits of agreement showed large intra-subject variability. The authors put forth that the primary limiting factor for repeatability appeared to be the relatively small separation between normative values for different grade levels used in the study. The use of the grade equivalent value when assessing repeatability was cautioned. It was proposed that looking at absolute values would be of greater worth.

Marrs and Patrick,\(^{21}\) in a review of the Reading Plus program (which utilizes the Visagraph to objectively measure eye movements), found reading efficiency was correlated significantly with the students’ starting level of reading skill on the Iowa Test of Basic Skills. Reading Plus is a series of computer-administered programs and exercises designed to develop silent reading fluency. Some believe that even though there is a correlation between functional eye movements and reading skills, OMD is not the cause of good or poor reading.\(^{22}\) Support against this conclusion is put forth by Solan et al.\(^{32}\) who found Visagraph eye movement results identified poor or good readers in middle-school students.

In this study, the ReadAlyzer was used as an alternative to the Visagraph II, as this was the available equipment in our clinic. The ReadAlyzer is essentially an identical device to the Visagraph II because it was created by the same programmer, uses the same analysis engine, and compares recordings to the same normative values. The goggles are interchangeable with an adapter. The passages used are dissimilar but follow the same pattern of increasing level of difficulty.

Symptom questionnaires are used to help clinicians determine if a patient is suffering from an underlying visual disorder. Many symptoms can greatly hamper a patient’s academic or social development. The College of Optometrists in Vision Development Quality of Life (COVID-QOL) questionnaire was developed to assess changes in patient lifestyle.\(^{24}\) It has been shown to have good test reliability\(^{13}\) and has been used to document the benefits of vision therapy.\(^{26-28}\) This short (originally 30 questions but reduced to 19 questions) questionnaire assesses 4 areas pertaining to quality of life: physical/occupational, psychological, social interaction, and somatic sensation.\(^{28}\) Scoring 20 or higher on either version
indicates a symptomatic patient in need of further testing. 29

The Readalyzer goggles (see Figure 3) were placed on the subject, and the near pupillarity distance was set using the vertical markings found on the goggles. Participants were then shown reading passages created for use with the Readalyzer. Participants were next shown both a long and a short demonstration of the level-10 passage and were allowed to scan it to become familiar with the size and word type. Recordings were not obtained from familiarization trials. 16, 17 Patients were given the following instructions:

1. Hold the reading material at a comfortable working distance.
2. You will be reading a passage that consists of either approximately 100 or 800 words.
3. Look at the dot on the top of the page and begin reading when instructed.
4. Read silently and, when done, close your eyes to indicate you are finished reading.
5. Be prepared to answer either 10 or 20 “Yes/No” questions after completing the passage.

Figure 2 An example of the long passage used during testing.

Our investigation was undertaken to ascertain if the 19-question COVID-QOL questionnaire could be correlated with the fixation-to-regression ratio or direction of attack measurement taken on the Readalyzer using both short and long passages. The COVID-QOL questionnaire was used to assign asymptomatic and symptomatic groups. The information gained from the Readalyzer was then compared by symptom group to ascertain if there were significant differences between them.

Materials and methods

Professional-level students were recruited from the Southern College of Optometry (SCO). Participants were first requested to fill out a COVID-QOL questionnaire (see Appendix). Scoring was performed as was in previous studies. 23–28 Each question is scored up to 4 points depending on the frequency of the symptoms. If the symptom was checked “always,” it was given a score of 4 points; “a lot,” 3 points; “sometimes,” 2 points; “once in a long while,” 1 point; and “never,” no points. Twenty participants scoring 15 or fewer were assigned randomly to the asymptomatic group and 20 participants scoring 16 or greater were assigned randomly to the symptomatic group. All participants who scored between 15 and 23 were not included in the study.

Each participant signed a consent form in accordance with the standards of the Human Subjects protocol at the college. All participants had received an optometric examination at the college before participating in the study. Exclusion criteria included amblyopia, strabismus, nerve palsy, and nystagmus. Because these were professional-level students, it was assumed that none suffered from a reading/learning disability, including dyslexia.
passage, an alternative passage would have been used, but this did not occur in this testing sequence.

A repeated measures Multivariate Analysis of Variance (MANOVA) was performed to analyze potential interaction effects. The software used was SPSS 16.0 for Windows, and the alpha level was 0.05. Post-hoc comparisons were performed using estimated marginal means to compare the following groups:
1. Short vs. long passages within the symptomatic group
2. Short vs. long passages within the asymptomatic group
3. Symptomatic vs. asymptomatic during the short passage reading
4. Symptomatic vs. asymptomatic during the long passage reading

### Table 3 Study results

<table>
<thead>
<tr>
<th></th>
<th>Mean</th>
<th>SD</th>
<th>N</th>
<th>P value</th>
</tr>
</thead>
<tbody>
<tr>
<td><strong>Asymptomatic</strong></td>
<td></td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>Short</td>
<td>8.75</td>
<td>4.82</td>
<td>20</td>
<td>0.0809</td>
</tr>
<tr>
<td>Long</td>
<td>10.60</td>
<td>6.17</td>
<td>20</td>
<td>0.0019</td>
</tr>
<tr>
<td><strong>Symptomatic</strong></td>
<td></td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>Short</td>
<td>10.90</td>
<td>6.50</td>
<td>20</td>
<td></td>
</tr>
<tr>
<td>Long</td>
<td>13.30</td>
<td>6.22</td>
<td>20</td>
<td></td>
</tr>
</tbody>
</table>

### Results

The demographic data were as follows: symptomatic group age range, 25 to 33 years, mean age, 26.85 years, and male-to-female ratio, 7:13; asymptomatic group age range, 22 to 38 years, mean age, 27.60 years, and male-to-female ratio, 9:11; entire treatment group age range, 24 to 38 years, mean age, 27.23, and male-to-female ratio, 16:24. There were 20 participants included in the asymptomatic group and 20 in the symptomatic group. The mean fixation-to-regression ratio when reading the short passage was 8.75 (standard deviation [SD] = 2.82) and 10.90 (SD = 6.17) for the asymptomatic and symptomatic groups, respectively (Table 2). Likewise, the mean fixation-to-regression ratio for the asymptomatic group on the long passage was 10.90 (SD = 6.50) and 13.30 (SD = 6.62) for the symptomatic group (Table 3). These numbers show the symptomatic group generally has a higher number of regressions per number of fixations, suggesting less-efficient eye movements.

A repeated measures MANOVA indicated the difference between short and long for the symptomatic group was not significantly different than the difference between short and long for the asymptomatic group (P = 0.651). Post-hoc comparisons using estimated marginal means indicated for asymptomatic, short vs. long, P = 0.036; for symptomatic, short vs. long, P = 0.008. For both asymptomatic and symptomatic subjects, the difference in fixation-to-regression ratio between passages exceeded 2. Additionally, for short length, symptomatic vs. symptomatic, P = 0.242; for long length, symptomatic vs. asymptomatic, P = 0.176. Although the observed difference between patient groups also exceeded 2, the increased variability related to between-subject comparisons resulted in nonsignificant findings.

### Discussion

Analysis using estimated marginal means indicated a significant difference between using a short and long passage for both patient groups (symptomatic and asymptomatic). This is corroborated by the repeated measures MANOVA, which indicated that for both groups combined, the ratio scores were significantly different between the long and short paragraph and there was no significant difference in the change in ratio scores between the 2 groups. The use of shortened passages with patients may, in fact, cause one piece of the visual efficiency puzzle to not be considered, assessed, and treated.

Why this difference exists is a complex question. In looking at the writings of Skeffington and Birnbaum, one could hypothesize there is a mismatch in the different visual efficiency systems that becomes more evident as visual stress increases over time, with prolonged reading. The patient who is not symptomatic either has no issue or has made an adaptation to address this problem and does not exhibit symptoms. The patient who has symptoms does not have an adaptive internal mechanism in place to deal with the building visual stress. Thus, the symptomatic patient exhibits visual efficiency difficulty. Because we are not evaluating only 1 aspect of the visual efficiency triad (vergence, accommodation, and ocular motor), we cannot say that this lower performance by the symptomatic group is caused by a particular vision skill in the triad.

Fatigue has long been considered an issue in academic performance, especially with near activities. Symptoms can include blurred or foggy vision far or near, difficulty focusing at near, difficulty tracking lines, diplopia, poor short-term memory, and headaches. Early on, there were many studies directed at the correlation of accommodative dysfunction with asthenopia. It has also been reported that 60% of college students with moderate to severe symptoms feel discomfort in as little as 30 minutes or less and 30% in less than 15 minutes. These symptoms were also shown to have a negative impact on academic performance. Therefore, it has been suggested that objective visual measurements and extended viewing times are critical to assessing visual fatigue.

Interestingly, our study was not able to answer whether the long passage could or could not be used to determine if the patient had a high level of symptoms as measured by the COVID-QOL questionnaire. A simple explanation for this difference might be that the questionnaire is not specific to ocular motor deficiency but assesses an overall function of vision and how it affects a patient.

A correlation between visual symptoms and eye movements has been investigated previously. Cohen et al. investigated the relationship between asthenopic symptoms using a symptom survey and different measurements of convergence, reading comprehension, and saccadic fixation eye movements. The Asthenopic Symptoms Questionnaire, a 9-question survey, was used. The eye movement test used was the DEM. The DEM ratio score was found to correlate with the asthenopic symptoms.
score (r = 20.32; P = 0.01). They hypothesized that asthenopic symptoms may affect saccadic fixation eye movements.

Conclusion

The longer passage for the ReadAlyzer has been shown to be a more sensitive tool in objectively assessing reading eye movements. Because most readers do not only read 100 words at a time, the long passage better corresponds to “real world” reading conditions. These findings call into question the use of the shorter passage as a tool to accurately help make a diagnosis of ocular motor dysfunction. Although the overall procedure time will increase, our results suggest use of longer passages will help in the diagnosis and treatment of ocular motor deficiency.

References


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### 19 Item COVID-QOL Checklist Questionnaire

Check the column which best represents the occurrence of each symptom

<table>
<thead>
<tr>
<th></th>
<th>NEVER</th>
<th>ONCE IN A LONG WHILE</th>
<th>SOMETIMES</th>
<th>A LOT</th>
<th>ALWAYS</th>
</tr>
</thead>
<tbody>
<tr>
<td>1.</td>
<td>Headaches with near work</td>
<td>-</td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>2.</td>
<td>Words run together reading</td>
<td>-</td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>3.</td>
<td>Burn, itch, watery eyes</td>
<td>-</td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>4.</td>
<td>Skips/repeats lines reading</td>
<td>-</td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>5.</td>
<td>Head tilt/close one eye when reading</td>
<td>-</td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>6.</td>
<td>Difficulty copying from chalkboard</td>
<td>-</td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>7.</td>
<td>Avoids near work/reading</td>
<td>-</td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>8.</td>
<td>Omiss small words when reading</td>
<td>-</td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>9.</td>
<td>Wires up/down hill</td>
<td>-</td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>10.</td>
<td>Misalign digits/columns numbers</td>
<td>-</td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>11.</td>
<td>Reading comprehension down</td>
<td>-</td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>12.</td>
<td>Holds reading too close</td>
<td>-</td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>13.</td>
<td>Trouble keeping attention on reading</td>
<td>-</td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>14.</td>
<td>Difficulty completing assignments on time</td>
<td>-</td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>15.</td>
<td>Always says &quot;I can't&quot; before trying</td>
<td>-</td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>16.</td>
<td>Clumsy, knocks things over</td>
<td>-</td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>17.</td>
<td>Does not use his/her time well</td>
<td>-</td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>18.</td>
<td>Loses belongings/things</td>
<td>-</td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>19.</td>
<td>Forgetful/poor memory</td>
<td>-</td>
<td></td>
<td></td>
<td></td>
</tr>
</tbody>
</table>

X0 ________ X1 ________ X2 ________ X3 ________ X4 ________

TOTAL SCORE __________
Lipemia retinalis: A combination of genetics and the American diet and lifestyle

Samantha L.C. Nogales, O.D., Amy L. Grimes, O.D., and Huey-Fen Song, O.D.

**KEYWORDS**
Lipemia retinalis; Hypertriglyceridemia; Pancreatitis

**ABSTRACT**

**Background:** Lipemia retinalis is a well-documented but rare ocular finding directly associated with serum triglyceride levels. The clinical presentation varies with the amount of triglycerides in the blood and completely resolves with lowering triglyceride levels, making this condition transient with no visual symptoms.

**Case Report:** A 46-year-old white man presented with lipemia retinalis and, hours later, to the emergency department with acute pancreatitis secondary to hypertriglyceridemia.

**Conclusion:** Lipemia retinalis warrants an immediate complete blood count and lipid panel to determine triglyceride levels with referral to a primary care provider.

Lipemia retinalis is an ocular condition that was first described by Heyl in 1880 to be associated with hypertriglyceridemia. The retinal vasculature is salmon pink to creamy white in color, and the arteries and veins are distinguishable only by size of the vessels. The overall fundus appears lighter in color because of the effect on the choroidal vasculature. The lightening of vessel content begins in the periphery and extends to the posterior pole with markedly elevated levels of serum triglycerides that exceed 2,500 mg/dL. A grading system used to describe the stages of this disease process was first established by Vinger and Sachs (see Table 1).

This unique fundus appearance is thought to be caused by the visualization of chylomicrons in the circulating retinal blood. Chylomicrons are large lipoproteins that transport triglycerides and the size is 0.25 wave-lengths of visible light (0.1 mm) and thus can be seen in the visible light range. However, other factors, such as hematocrit levels, may contribute to the fundus appearance and change the triglyceride level at which a particular grade of lipemia retinalis might be present; very low-density lipoproteins (VLDL), which are slightly smaller in size than chylomicrons, are also involved.

Lipemia retinalis has been found to occur in certain types of primary familial hypertriglyceridemia and secondary acquired hypertriglyceridemia. There are 5 types of primary hypertriglyceridemia first described by Fredrickson et al., that have now been modified by the World Health Organization (see Table 2) to lipemia retinalis, occurring in 4 of the 5 types. Secondary acquired hypertriglyceridemia can occur as result of uncontrolled diabetes mellitus (DM), obesity, alcoholism, renal failure, systemic lupus erythematosus, hypothyroidism, nephrotic syndrome, biliary obstruction, and certain medications, such as estrogen and beta blockers. However, because not all people with these metabolic conditions will have associated high triglyceride levels, a subtle inherited metabolic defect may confer susceptibility and caution should be used when prescribing estrogens, thiazides, or beta blockers in patients with hypertriglyceridemia.

<table>
<thead>
<tr>
<th>Table 1</th>
<th>Grading system for the stages of lipemia retinalis</th>
</tr>
</thead>
<tbody>
<tr>
<td>Grade</td>
<td>Triglyceride level (mg/dL)</td>
</tr>
<tr>
<td>I</td>
<td>2,500-3,499 (early)</td>
</tr>
<tr>
<td>II</td>
<td>3,500-5,000 (moderate)</td>
</tr>
<tr>
<td>VIII</td>
<td>&gt; 5,000 (marked)</td>
</tr>
</tbody>
</table>

**Case Report**

A 46-year-old white man presented for a diabetic eye examination with no ocular, visual, or systemic complaints. He reported his last eye examination was 3 years prior at another Veterans Affairs facility; he had fundus photos taken on October 7, 2008, which were reviewed through the interconnected electronic medical record system and noted to be unremarkable in both eyes (OU). His ocular history was unremarkable. Medical history was significant for asthma treated with albuterol and mometasone inhalers; sleep apnea, for which he used a continuous positive airway pressure machine; type II diabetes managed with metformin, 1,000 mg twice daily; hypertension treated with amlopidine, 10 mg daily, atenolol, 100 mg daily, and lisinopril/hydrochlorothiazide (HCTZ), 10 mg daily. The patient also had hypertriglyceridemia treated with gemfibrozil, 600 mg twice daily, niacin, 1,000 mg daily, and fish oil, 1,000 mg twice daily; depressive and anxiety disorders medicated with citalopram, 40 mg daily, and clonazepam, 0.50 mg twice a day; obesity with a weight of 335.9 pounds and a body mass index of 42.8; and prior cholecystectomy. Social history consisted of nicotine dependence with a half-pack-per-day cigarette smoking habit for more than 20 years and an alcohol consumption of 6 cans of beer per day for many years. His family history included a
mother with type II diabetes; a father with hypertension, carotid artery disease, and chronic heart failure who died at age 65; and a brother and niece with Crohn's disease.

His best-corrected visual acuities were 20/20 in the right eye and the left eye, respectively. Entrance testing, slit lamp examination, and tonometry findings were unremarkable. Dilated fundus examination found dilated, creamy-colored arteries and veins nearly equal in diameter, with the veins being only slightly larger (see Figure 1). The overall fundus appeared lighter in color. Previous laboratory findings from 3 months before this eye examination were abnormal, with a triglyceride level of 4,374 mg/dL (range, 10 to 160 mg/dL), cholesterol level of 452 mg/dL (range, 150 to 200 mg/dL), and glycosylated hemoglobin of 7.7 (range, < 6.0). Medications were reviewed, and it was taken into consideration that the patient was prescribed a beta blocker and thiazide combination to treat his hypertension because beta blockers and thiazides have been found to increase triglyceride levels. However, the effects were considered a minor contributor to this patient's extreme triglyceride level. According to recent studies, combination of HCTZ and atenolol treatment can increase triglyceride levels by 33% to 46% from the patient’s baseline triglyceride level. Although our patient's baseline triglyceride level was unclear, his triglyceride levels ranged between 405 and 822 mg/dL before his severe hypertriglyceridemia incident, which already accounted for the HCTZ and atenolol effect. During the eye examination, the patient admitted to discontinuing all of his medications for the previous 2 months, recently restarted the medications as prescribed by his primary care provider, and was scheduled for follow-up with his endocrinologist in 2 weeks. External examination was unremarkable. The patient was educated on his eye findings that were directly associated with his hypertriglyceridemia and the need for compliance with all prescribed medications as well as keeping his follow-up appointments with the endocrinologist. Repeat laboratory testing was ordered, and the patient was educated on lifestyle and diet modifications.

Approximately 8 hours after his eye examination, the patient was brought to the emergency department by paramedics with a several-hour history of epigastric pain that was radiating to his right upper quadrant and substernal area. The pain was severe and had worsened over the last few hours. The patient also reported nausea and vomiting with diaphoresis. Upon arrival, the patient’s temperature was 98.1°F, and his blood pressure was 156/95 mmHg. Laboratory results were significant for a high glucose level of 386 mg/dL, high amylase levels of 10.7, high lipase levels of 38,037 U/L (range, 25 to 250 U/L), and high triglyceride levels of > 5,250 mg/dL (range, 10 to 160 mg/dL) (see Figure 2). A computerized axial tomography scan of the abdomen without contrast showed mild infiltration of the peripancreatic fat around the pancreatic head and proximal body that was highly suspicious for pancreatitis. Magnetic resonance imaging of the abdomen with and without contrast also found

Table 2 World Health Organization classification with clinical descriptive names and corresponding Fredrickson numerical types (in parentheses)

<table>
<thead>
<tr>
<th>Lipoprotein pattern</th>
<th>Major elevation in plasma</th>
<th>Ocular features</th>
</tr>
</thead>
<tbody>
<tr>
<td>Familial chylomicronemia (type I)</td>
<td>Chylomicrons, triglycerides</td>
<td>Lipemia retinalis, iris and retinal xanthoma, lipid keratopathy, adult-onset Coat’s disease</td>
</tr>
<tr>
<td>Familial combined hyperlipoproteinemia (type IIb)</td>
<td>Low-density lipoproteins/VLDL, cholesteryl, and triglycerides</td>
<td>Xanthelasma, corneal arcus, xanthomas of the retina</td>
</tr>
<tr>
<td>Familial dysbetatipoproteinemia (type III)</td>
<td>Remnants or beta-VLDL, triglycerides, and cholesteryl</td>
<td>Early-onset arcus senilis, xanthomas, rarely lipemia retinalis</td>
</tr>
<tr>
<td>Familial hypertriglyceridemia (type IV)</td>
<td>VLDL, triglycerides</td>
<td>Lipemia retinalis, xanthomas</td>
</tr>
<tr>
<td>Primary mixed hyperlipidemia (type V)</td>
<td>VLDL, chylomicrons, triglycerides, and cholesteryl</td>
<td>Lipemia retinalis, xanthomas</td>
</tr>
</tbody>
</table>

VLDL = very–low-density lipoprotein.

Figure 1 Fundus photography shows bilateral, dilated, creamy-colored arteries and veins with background lightening.
The patient also reported nausea and vomiting with diaphoresis and a several-hour history of epigastric pain that was brought to the emergency department by paramedics. Repeat laboratory examinations were significant for a high glucose level of 386 mg/dL, high lipase, and the need for compliance with all medications, but laboratory testing still showed elevated triglycerides and blood sugar. The patient was then admitted to the hospital for treatment of acute pancreatitis.

Intravenous fluids, morphine for pain control, and promethazine, an antiemetic, were started in the emergency department. When the patient was admitted to the hospital, an insulin drip was started, in addition to receiving regular units of insulin to aggressively control blood sugar. The patient was restricted from eating or drinking anything for 2 days, and his atenolol for hypertension was discontinued and replaced with lisinopril, because beta blockers can increase triglyceride levels. After 4 days of intravenous fluids and restricted diet, laboratory testing found decreased triglyceride, blood sugar, and lipase levels with the resolution of pancreatitis, and the patient was discharged from the hospital. An endocrinology appointment was scheduled 1 week later where the patient was found to be compliant with all medications, but laboratory testing still showed elevated triglyceride levels, although much improved from the time of admission. To bring these levels under control, it was recommended the DM be more strictly controlled by adding metformin and continuing insulin injections. Increases in fish oil to 1,000 mg, 2 capsules, 3 times a day and niacin to 200 mg daily were also recommended with a 3-week follow up.

At the 2-week optometry follow-up after the hospitalization for pancreatitis, the patient reported improvement in diet, cessation of smoking, and stoppage of alcohol consumption. He reported vision fluctuations OU while in the hospital, most likely caused by changes in blood sugar, but had no ocular complaints. His best-corrected visual acuities were 20/20-1 in the right eye and 20/20 in the left eye. Entrance testing, slit lamp examination, and tonometry were unremarkable. Dilated fundus examination was unremarkable OU. The patient was advised of his eye findings and the resolution of the lipemia retinalis OU. He was educated on the importance of compliance with all medications and to keep all follow-up appointments with endocrinology and his primary care provider. Follow-up in optometry was scheduled for 1 year or sooner with any visual or ocular changes.

Discussion

Two main sources of plasma triglycerides are found through the endogenous and exogenous pathway of lipoprotein/lipid metabolism. Endogenous triglycerides from the liver are carried on VLDL, and exogenous triglycerides and cholesterol from dietary fat are packaged by the intestine into chylomicrons. The lipoproteins and chylomicrons are then hydrolyzed by lipoprotein lipase into free fatty acids on the luminal side of endothelial, adipose, and muscle cells.

Hypertriglyceridemia occurs when fasting plasma triglyceride measurements are elevated above the 95th percentile (> 2,000 mg/dL) for a given age and sex and can lead to increased risk of cardiovascular disease and acute pancreatitis. As mentioned, hypertriglyceridemia can be divided into primary (including genetic defects in lipid metabolism) and secondary (including metabolic or endocrine disorders, lifestyle, environment, and medications) types. Clinically, the most significant primary and secondary types are those that cause chylomicronemia (plasma triglyceride levels exceeding 1,000 mg/dL), which result from impaired triglyceride metabolism and chylomicronemia syndrome, defined as chylomicronemia accompanied by 1 or more of the following: hepatosplenomegaly, acute pancreatitis, lipemia retinalis, and eruptive xanthomas. Primary causes include familial chylomicronemia, familial hypertriglyceridemia, and primary mixed hyperlipidemia and having a biochemically proven deficiency in lipoprotein lipase or apolipoprotein C-II activity (a cofactor for lipoprotein lipase). Even though most patients will have some form of genetic defect in lipid metabolism, it is often exacerbated by 3 common secondary causes: DM, excessive alcohol intake, and obesity (as in our patient).

Lipemia retinalis occurs when triglyceride levels exceed 2,500 mg/dL, which is a significant indicator of primary or secondary hypertriglyceridemia, as other lipids such as cholesterol do not contribute to the retinal appearance. Corneal arcus and xanthelasma are not seen. The color change of the fundus and blood vessels to salmon pink and creamy white is visible initially in the peripheral blood vessels where the red blood cells are formed into a thin column. With increasing levels of triglycerides, this color change extends to the posterior pole. There are several case reports of lipemia retinalis occurring in both primary (familial hypertriglyceridemia) and secondary types of hypertriglyceridemia, and in both cases vision was not affected. In addition, a case of lipoprotein lipase deficiency coupled with high dietary fat intake presented normal fluorescein angiography, visual fields, and visual acuity. However, another case report involving a patient with most likely primary mixed hyperlipidemia and secondary diabetes mellitus did report a decrease in visual acuity, but it was attributed to diabetic macular edema and not lipemia retinalis.

Treatment for lipemia retinalis involves treating the cause of hypertriglyceridemia with the main goal of maintaining fasting triglyceride levels at < 500 mg/dL to reduce the risk of acute pancreatitis. Because most cases of hypertriglyceridemia occur when a genetically predisposed individual is exposed to a secondary condition, treatment should target the secondary disorder. This includes weight loss, decrease in fat and refined carbohydrate intake, exercise, and decreasing or eliminating alcohol consumption. Blood sugar should be controlled tightly in people with diabetes, and hypothyroidism should be controlled with thyroid function regulating medications. The first line of treatment includes a fibric acid derivative, such as gemfibrozil,
statins, which can markedly reduce triglyceride levels and reduce the risk of coronary heart disease, especially in type II diabetics, and niacin, which can reduce triglycerides by 45% and can often be effective alone or in combination with a fibrate. Statins, however, are not considered a first-line therapy when triglyceride levels exceed 5,000 mg/dL, as in our patient. The U.S. Food and Drug Administration has also approved omega-3 fatty acid as an adjunctive treatment. 

Emerging treatments include rimonabant, a cannabinoid-1 receptor antagonist that decreases hunger and reduces food intake, and lipoprotein lipase gene therapy. Several case reports have shown that as triglyceride levels decrease, lipemia retinalis begins to resolve in reverse with reversion of the retinal blood vessels and fundus to their normal color starting at the posterior pole and then extending to the periphery.

### Conclusion

Lipemia retinalis is often an incidental fundus finding associated with primary and secondary hypertriglyceridemias. Although lipemia retinalis is an asymptomatic condition and recognized as being non–vision-threatening, it is important to realize the impact of elevated triglyceride levels on the cardiovascular system and the increased risk for potentially life-threatening acute pancreatitis. As such, upon clinical finding of lipemia retinalis, a thorough health, familial, and social history should be acquired as well as immediate referral for laboratory testing and systemic workup so treatment may be initiated.

### References


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Dry eye--is inflammation just the tip of the iceberg?

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KEYWORDS
Dry eye; Ocular surface disease; Tensegrity, Inflammation, Platelet-Rich Plasma

ABSTRACT

Background: Dry eye syndrome (DES) has been described by The International Dry Eye WorkShop as “multifactorial disease of the tears and ocular surface that results in symptoms of discomfort, visual disturbance and tear instability with potential damage to the ocular surface.” Inflammation has been recognized as a primary contributor to the disease and was a tremendous step forward in the description and treatment of DES. Although most treatments of DES treat inflammation, the signs and symptoms of DES usually return shortly after discontinuing the use of the anti-inflammatory agent.

Case: We present a case of a 70-year-old patient who presents with significant dry eye symptoms for 2 years. She reports the need to wear sunglasses during all waking hours, both indoors and out, and had discontinued her hobby of gardening. She had ocular rosacea diagnosed at a previous ophthalmology practice and did not respond to any past treatments. The patient was treated with 25% platelet-rich plasma (PRP) and experienced significant improvement and a return to a normal quality of life that has continued for more than a year after discontinuing treatment.

Conclusion: Our group is investigating the use of biologic therapeutics in the treatment of various forms of DES. We suggest that a clinically acceptable dosage of PRP provides the ocular surface with the components necessary to restore normal cellular tensegrity and provides a foundation to eliminate the recurrence of the inflammation associated with DES.

Dry eye or dry eye syndrome (DES) has been described as a disruption of the precorneal tear film caused by tears insufficient in volume or inadequate in function. Such a description may suggest replacing the tear film with artificial supplements would adequately control the disease. However, an explosion of research has led to a more precise definition of DES. The International Dry Eye WorkShop (DEWS) describes dry eye as a “multifactorial disease of the tears and ocular surface that results in symptoms of discomfort, visual disturbance and tear instability with potential damage to the ocular surface. It is accompanied by increased osmolarity of the tear film and inflammation of the ocular surface.” The recognition that the cause of DES is multifactorial, promotes inflammation, induces hyperosmolarity, and causes tissue destruction is key to this new definition and likewise must guide our therapeutic approaches to the ubiquitous ophthalmic disease. In short, any therapy that merely relieves symptoms can be only palliative in nature and never truly curative. The key to dry eye therapy is to re-establish and maintain ocular surface homeostasis.

The multifactorial nature of dry eye disease is well-established. Age is the most common factor associated with DES, affecting all aspects of tear production, secretion, and stability. Dysfunction of tear film dynamics including increased tear evaporation, decreased volume, disrupted flow, and increased tear osmolarity have been reported. Increases in ducal pathology of the lacrimal gland appear to increase with age. Changes in meibomian secretions and decreases in conjunctival goblet cell density and lid irregularities also have been reported with advancing age. Hormonal changes, more specifically, androgen insufficiency, can result in dry eye symptoms, and these also increase with age. Other factors capable of initiating the inflammatory cascade leading to DES include systemic autoimmune diseases, viral infections, and non–dry eye disease states of the ocular surface and lacrimal gland. In addition, some commonly used medications such as diuretics, antihistamines, and antidepressants, exacerbate the symptoms seen in patients with DES. Environmental assaults on the ocular surface by allergens, wind, and other irritants contribute to localized drying of the ocular surface and consequent symptoms.

The DEWS definition includes hyperosmolarity of the tears and inflammation of the ocular surface, because they are common findings seen in all forms of DES. Lipid alterations secondary to meibomian gland dysfunction, as well as increased evaporation or stagnation of tears from contact lens wear, can result in tear hyperosmolarity. Hyperosmolarity can initiate an inflammatory response via 2 major cellular stress response pathways (nuclear factor kappa B and stress-activated protein kinase). Tear testing has demonstrated the presence of pro-inflammatory cytokines (interleukin-1, interleukin-6 and tumor necrosis factor alpha) in the conjunctival epithelium and tears of dry eye patients. Hyperosmolarity has been shown to lead to altered mucin composition and subsequent increases in lid-induced shear stress. This mechanical stress stimulates sensory elements in the plasma membrane and can activate several membrane-bound intracellular signaling proteins, such as receptor tyrosine kinases and phospholipase A2, resulting in further inflammation.

Although DES has been accepted as a chronic inflammatory disease with a complex and multifactorial pathological basis, treatments have centered around quelling ocular surface inflammation alone, often with temporary results. Prednisolone, loteprednol, cyclosporin A, topical azithromycin,
oral tetracycline, and essential fatty acid supplements, have all been advocated to treat various forms of DES by temporarily decreasing the associated inflammation. However, a fair number of patients do not respond to these treatments or may respond favorably during treatment but relapse at discontinuation. This has led to a myriad of therapeutic approaches that reflect the inclinations of the individual prescriber but that lack robust evidence of efficacy. These therapies may benefit the patient, but the rigors of the treatment processes soon lead to non-compliance. Current treatment methods fail to address the loss of structural integrity of the affected tissues that is the result of the disease process and, also, a potentiator of further inflammation and disease. While each of these therapies sometimes leads to symptomatic relief, none of them has been shown to restore normal ocular surface homeostasis for prolonged periods.

**Case report**

A 70-year-old female patient presented to the office with significant dry eye symptoms after having been treated for years with current pharmacotherapy, fatty acid supplements, and artificial tears. The patient had discontinued wearing contact lenses 3 years previously and was frustrated with her care. She was referred by a medical colleague who had treated her for a shoulder anomaly with platelet rich plasma (PRP) and adult stem cell graft. At her initial visit she completed the Ocular Surface Disease Index survey and reported a score of 77.08, which indicates moderate to severe dry eye. The patient’s symptoms were significant for photophobia for more than 2 years. The photophobia was debilitating. She reported that she couldn’t tolerate the overhead lighting in her home and had to eat holiday meals with the lights turned down. She wore sunglasses during all waking hours both indoors and out and had discontinued her hobby of gardening. Past treatments included a combination of omega-3 supplements, doxycycline, and Restasis (Restasis; Inspire Pharmaceuticals, Durham, North Carolina) for a year. She then was instructed to discontinue the doxycycline and was started on Azasite (Azasite; Inspire Pharmaceuticals, Durham, North Carolina). The patient was on her current regimen of omega-3 supplements, Restasis, and Azasite for about a year before presenting to our office. At her initial visit, her clinical signs showed significant corneal and conjunctival staining with fluorescein and rose Bengal (see Table 1). Filaments were beginning to form on the corneal surface. Other initial clinical data can be found in Table 1.

From the clinical data, we diagnosed an evaporative dry eye caused by mixed mechanisms with a severity level of grade 3 on the modified International Task Force severity grading scheme as described in the DEWS Report. Treatment was initiated by discontinuing the Azasite (Azasite; Inspire Pharmaceuticals, Durham, North Carolina) and Restasis (Restasis; Allergan, Inc., Irvine, California), and she was started on the PRP treatment regimen described above. Within 2 weeks, the staining was resolving, and she reported marked improvement in her symptoms. In 1 month she was significantly better and did not require the use of her habitual sunglasses indoors. After 3 months of treatment, she discontinued the PRP regimen and demonstrated enough improvement in her ocular surface and tear volume to attempt contact lens wear. The patient’s right eye was refitted for monovision wear into a 1-day Acuvue® TruEye™ (Acuvue; Johnson & Johnson Vision Care, Jacksonville, Florida) daily disposable lens (8.5 base curve, 14.2 diameter; 13.00 diopeters). After 3 months of contact lens wear she continues to be comfortable with her current treatment of preservative-free artificial tears and omega-3 supplements.

<table>
<thead>
<tr>
<th>Table 1</th>
<th>Initial clinical data of patient</th>
</tr>
</thead>
<tbody>
<tr>
<td>Clinical procedure</td>
<td>Results</td>
</tr>
<tr>
<td>Blink rate</td>
<td>21 blinks/min</td>
</tr>
<tr>
<td>Anterior lid margin</td>
<td>Normal</td>
</tr>
<tr>
<td>Posterior lid margin</td>
<td>Trace telangiectasia OU</td>
</tr>
<tr>
<td>Tear meniscus height</td>
<td>1.0 mm OD and OS</td>
</tr>
<tr>
<td>Meibomian glands</td>
<td>Significant dropout on transillumination OD and OS</td>
</tr>
<tr>
<td>Meibomian gland expression</td>
<td>1/3 expressed normal meibum; 2/3 unexpressible Significant meibomian gland orifice stenosis OD and OS</td>
</tr>
<tr>
<td>TBUT</td>
<td>OD: 5 s OS: 6 s</td>
</tr>
<tr>
<td>Fluorescein corneal staining*</td>
<td>OD: 12/15 OS: 11/15</td>
</tr>
<tr>
<td>Schirmer test</td>
<td>OD: 3 mm OS: 4 mm</td>
</tr>
<tr>
<td>Phenol red thread test</td>
<td>OD: 16 mm OS: 14 mm</td>
</tr>
</tbody>
</table>

* National Eye Institute corneal grading system.

**Conclusion**

Our group is investigating the use of biologic therapeutics in the treatment of various forms of DES. We do not believe that current therapies address the loss of normal tissue integrity that occurs with dry eye disease. The symbiosis between structure and function being the hallmark of all normal organ function, the re-establishment of normal tissue structure is essential for true therapy. We have treated several patients with a careful formulation of a 25% PRP concentrate as described by Koffler. The dosing regimen of the PRP concentrate was changed from that of previous authors to a more clinically acceptable dosing regimen of twice a day. Most patients noticed symptomatic improvement in 3 weeks, and many have achieved long-term resolution of their clinical signs and symptoms even after discontinuation of therapy. We believe the combination of growth factors released by platelet activation is the multifactorial treatment to this multifactorial disease. Our group believes the combination of growth factors found in platelets provided the ocular surface with the tools necessary to repair the damage induced by the various causes of dry eye. We are currently in the process of clinical studies we hope will support our hypothesis and demonstrate that regenerative medicine holds the best chance for sustained relief for patients with dry eye and other ocular surface diseases.

**References**


Progressive, asymptomatic papilledema as the presenting sign of a Chiari I malformation

Wendy J. Haaland Stone, O.D., Erica A. Ittner, O.D., Bruce A. Teitelbaum, O.D., and Leonard V. Messner, O.D.

KEYWORDS
- Papilledema
- Chiari I malformation
- pseudotumor cerebri
- intracranial hypertension

ABSTRACT

**Background:** Chiari I malformation (CM1) is the caudal herniation of the cerebellar tonsils into the foramen magnum (FM) of at least 5 mm as shown on magnetic resonance imaging (MRI). This causes disruption of the flow of cerebral spinal fluid (CSF) and compression of brain tissue. Chiari I malformation is due to a congenital maldevelopment of the posterior fossa, causing it to be smaller than normal. The normal position for the cerebellar tonsils is at or above the FM. The symptomology with CM1 can vary from asymptomatic to significant neurologic disability. When symptoms are present, they are typically non-specific and non-localizing, frequently leading to misdiagnosis. Common complaints include dizziness, disequilibrium, vertigo, tinnitus, dysphagia, sleep apnea, tremors, palpitations, paresthesia, fatigue, muscle weakness, and neck pain. The most common complaint in symptomatic patients is suboccipital headaches, though many ocular complaints have also been reported, including blurred vision, photophobia, and diplopia, often due to cranial nerve VI palsy. The most common ocular clinical finding of CM1 is nystagmus, which has been reported in 27% of patients. Symptomatic papilledema has been reported only rarely with this condition, and it indicates the presence of intracranial hypertension.

Best-corrected visual acuities were 20/20 OD, OS, OU with a moderate myopic prescription. Her pupils, confrontational visual fields, color vision, and extraocular muscle movements were normal. Anterior segment evaluation was unremarkable, and applanation tonometry was measured at 15 mmHg OD, OS. Her blood pressure was measured at 122/75 mmHg. Dilated fundus exam showed normal periphery, vasculature, and maculae OU. However, both optic discs appeared to be mildly elevated without the presence of hemorrhages (see Figures 1 & 2. Note the presence of circumferential choroidal folds superior temporally OU. These folds, often called Paton's lines, are indicative of peripapillary retinal edema and may not be present with pseudopapilledema.) B-scan ultrasonography was performed and showed no evidence of optic disc drusen. Humphrey visual fields were performed and showed only a few edge point defects, likely due to the trial lens. The patient was diagnosed with early papilledema versus anomalous discs...
(pseudopapilledema). Due to her lack of symptoms, she was asked to return in one month for further evaluation.

At the one-month visit, the patient still denied headaches or other visual or neurological symptoms. However, the optic nerve appearance in both eyes had worsened and was suggestive of papilledema. The remainder of the ocular examination was normal. Evaluation of cranial nerves 3-12 was unremarkable, and there was no evidence of gait or brainstem/cerebellar dysfunction. Magnetic resonance imaging (MRI) of the brain, including angiography and venography, was obtained showing a CM1 with 9 mm of herniation of the cerebellar tonsils below the FM with near complete effacement of the surrounding subarachnoid space (see Figure 3). No intracranial mass, orbital mass, hydrocephalus, or vascular abnormality was found.

Discussion

While cases of symptomatic CM1 presenting as papilledema have been reported previously,\(^8,^9\) this is the first known case of a completely asymptomatic patient with papilledema as the sole presenting sign or symptom of CM1. However, it brings up the question of whether these 2 conditions are related or coincidental, as there seems to be some disagreement in the scientific literature on this topic.

Chiari I Malformation

Traditionally, there are 4 different classifications of Chiari malformations based upon the portion of the brain that is herniated through the FM and the abnormalities present. CM1 is the caudal herniation of the cerebellar tonsils through the FM of at least 5 mm,\(^1\) the least amount of all the classifications. However, symptomatic cases have been reported with as little as 2 mm of herniation.\(^2\) Though it does not typically present until the early teens, CM1 is the most common of the 4, and it is not associated with spina bifida.\(^10\) Type II is the herniation of not only the cerebellar tonsils, but also portions of the brainstem. It is associated with spina bifida, and it typically presents at birth. Types III and IV are very rare and present at birth with a high mortality rate. Type III involves the herniation of the entire cerebellum, in addition to portions of the
brainstem; type IV involves a hypoplastic, maldeveloped cerebellum. In addition to these classic categories of Chiari malformations, a newer classification has been defined, type 1.5. These patients fall between types I and II previously described, as they have some brainstem herniation into the FM but without spina bifida. This may represent a progression of CM1.

MRI is the preferred imaging method, as computed tomography does not image the posterior fossa well. Cases have been reported where a CM1 was missed on computed tomography leading to neurologic emergencies or even death in patients undergoing lumbar puncture, as this procedure can cause the further herniation of the cerebellar tonsils. In addition to the cerebellar herniation, patients with CM1 typically show a small, crowded posterior fossa on MRI. The etiology of CM1 is believed to be congenital maldevelopment of the posterior fossa, which leads to crowding of the cerebellum and medulla, pushing them downward into the FM. This can create other MRI findings, including syringomyelia in 40-75% of cases. Syringomyelia are chronic defects where the spinal cord has a tubular cavity or cyst present in several spinal cord segments. This represents the abnormal movement of CSF into the white matter of the spinal cord, and can cause significant neurologic defects. Hydrocephalus, an enlargement of the cerebral ventricles due to the accumulation of CSF which can lead to papilledema, has also been reported in 3-10% cases of CM1.

CM1 is estimated to occur in 0.6-0.9% of the general population, it tends to be more common in women, and symptoms tend to manifest during the second or third decade of life. CM1 can also be asymptomatic in 14-31% of cases, but, when symptomatic, suboccipital headaches are the most common symptom. In a study of 364 symptomatic CM1 patients, Milhorat et al. found 81% of patients complained of this type of headache. They also found 78% of patients reported ocular disturbances including visual phenomena, blurred vision, photophobia, and diplopia. The most common ocular finding in this study was nystagmus, which was reported in 27% of cases. This was most commonly lateral nystagmus (64% of patients with nystagmus), followed by downbeat nystagmus (24%), rotary nystagmus (10%), and periodic alternating nystagmus (2%). This group found that only 2.5% of patients had papilledema, and only 2% had papilledema without hydrocephalus.

The CSF flow has been shown to be abnormal in patients with CM1. Heiss et al. studied the CSF behavior with cine MRI of 20 CM1 patients compared to healthy controls. Cine MRI is the taking of repeated images at short timeframes that amounts to a movie where the CSF flow can be measured to see if it is disrupted by the herniated cerebellum at the FM. They found the CM1 patients had diminished anterior-posterior diameter of the CSF space at the FM, decreased CSF flow rates at the FM, and cervical CSF pressures and pulse pressures that were increased. This altered CSF flow may lead to an increase in ICP by either mechanical obstruction of outflow at the FM, the compression of draining veins in the crowded posterior fossa leading to increased brain turgor, or the scarring of ectopic cerebellar tonsils due to rubbing against the bone of FM obstructing outflow. It is thought that the disruption of normal CSF flow leads to the development of a pressure differential between the intracranial and spinal systems, which causes the development of papilledema.

Figure 4: OCT OU
the cerebellar herniation and syringomyelia. However, not all patients with CM1 have increased ICP. Treatment of CM1 is indicated if the patient is symptomatic, and the typical treatment is surgical suboccipital decompression (SOD). This procedure is intended to enlarge the posterior fossa to relieve crowding of the neurologic structures and create more room for proper CSF flow.

**Papilledema/Pseudotumor Cerebri**

Papilledema is the swelling and elevation of the optic nerve heads due to increased intracranial pressure and is typically bilateral. Its presence requires imaging to search for the etiology of the increased ICP, including intracranial mass, hydrocephalus, or vascular abnormality. If none are present on MRI, the patient may undergo lumbar puncture to evaluate the opening pressure and contents of the CSF. If ICP is elevated and there is no evidence of a cause (e.g., tumor), then PTC is the diagnosis. Once again, it is important to obtain the MRI before proceeding with lumbar puncture, as a CM1, if present, may worsen with this procedure. If CM1 is present, then the “30 degree test” is another, noninvasive, method of evaluating the presence of papilledema. This test is a modified ultrasonography where the thickness of the optic nerve is measured in primary gaze and again with the patient abducting 30 degrees. Patients with increased ICP will have lower measurements when abducting than in primary gaze, as the subarachnoid fluid is distributed along the stretched optic nerve sheath. Patients with normal ICP will have no difference in these measurements.

PTC is a diagnosis of exclusion. Most clinicians use the modified Dandy's criteria for PTC: increased ICP with normal CSF contents (250 mm of water or greater on lumbar puncture opening pressure), absence of any localizing neurological signs (except for an isolated cranial nerve VI palsy), and absence of ventriculomegaly or any mass lesion on brain imaging. More than 90% of these patients complain of headaches, and these headaches tend to be worse during Valsalva maneuver. They may also complain of nausea, vomiting, dizziness, and pulsatile tinnitus. In addition to papilledema, the eye signs may include cranial nerve VI palsy, decreased visual acuity, visual field defects, and transient visual obscurations. While most patients are symptomatic, some patients may be without symptoms entirely. The rate of asymptomatic patients has been reported at 3.7-4.8%, though one retrospective record review did find this rate to be much higher, at 24.7%.

PTC characteristically occurs in obese women of childbearing age. However, it does occur in men, children, and one study reported 10% of patients were not obese. Some theorize that the obesity causes the PTC, perhaps due to increased intra-abdominal pressure causing increased ICP. Several vitamins and medications have been implicated in the development of PTC, which would make it no longer “idiopathic.” In the absence of an exogenous cause, impaired CSF absorption causing the increased ICP is thought to be from either the blockage of outflow at the arachnoid villi, venous congestion, or diffuse brain edema.

Initial treatment in PTC includes weight loss, carbonic anhydrase inhibitors (most often acetazolamide), and even serial lumbar punctures to relieve the increased ICP. If these treatments fail, or if the papilledema and visual field defects worsen, the patient may require surgery. Optic nerve sheath decompression may be performed to relieve papilledema and save vision. However, to relieve the underlying intracranial hypertension and headaches, the patient may require a CSF shunting procedure. The most common type of shunt is the lumbo-peritoneal shunt (LPS), where a shunt is placed between the lumbar subarachnoid space and the peritoneal cavity. However, this procedure may actually cause an acquired Chiari malformation (ACM) in some patients, due to the acute pressure drop between the intracranial and spinal compartments. Ventriculo-peritoneal shunts (VPS) are less likely to cause ACM, but their placement is a more difficult procedure in patients without hydrocephalus, including those with PTC, since the ventricles are normal or small in size.

**Relationship Between Chiari I Malformation and Pseudotumor Cerebri**

There has been much discussion in the recent literature, especially the neurosurgical literature, as to whether or not these two conditions have a causal relationship. Certainly their presentations can be similar, making the differentiation difficult: both have headaches which may worsen with Valsalva maneuvers, both may present with cranial nerve VI palsies or papilledema, both present more commonly in females, and both tend to become symptomatic during childbearing years.

As previously discussed, ACM can actually occur secondary to LPS for PTC. However, a few studies have shown that at least some of these patients had cerebellar herniation before shunting, in a proportion larger than that in the general population. This is the increased ICP cerebellar herniation and syringomyelia. However, not all patients with CM1 have increased ICP. Treatment of CM1 is indicated if the patient is symptomatic, and the typical treatment is surgical suboccipital decompression (SOD). This procedure is intended to enlarge the posterior fossa to relieve crowding of the neurologic structures and create more room for proper CSF flow.

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Sinclair et al. reported a case of simultaneous PTC and CM1 in a 12-year-old girl, but felt it was coincidental, even though a subsequent record review of 156 cases found that 2.7% of PTC patients had evidence of pretreatment tonsillar displacement on MRI. Banik et al. theorized that cerebellar ectopia may be caused by the increased ICP of true primary PTC; conversely, they suggested that pre-existing cerebellar ectopia may cause increased ICP from obstruction of outflow of CSF, producing the clinical picture of PTC. Bejjani suggested that cerebellar ectopia can occur from either a small skull or increased intracranial contents. He argued that PTC causes brain engorgement that leads to an increase in the intracranial content volume, which causes the cerebellar tonsils to herniate into the cervical canal. He suggested that PTC and CM1 may occasionally be manifestations of the same cause, within the same spectrum of disease. Fagan et al. agreed with this idea of the coexistence of these conditions as a part of the spectrum of PTC and CM1 syndromes. They postulated that in patients with normal CSF flow at the FM (meaning the CM1 itself is not causing the obstruction of CSF leading to increased ICP), the CM1 may be secondary to the PTC. They named these cases “Chiari PTC,” and suggested that these patients were predisposed for the development of CM1 (e.g., due to small posterior fossa or trauma).

If one recognizes that these 2 syndromes may be interrelated, the decision of which condition to treat in symptomatic patients becomes a difficult one. Vaphiades advocated treating based on the CSF characteristics. He suggested obtaining dynamic magnetic resonance CSF flow studies, and if the flow is abnormal at the level of the FM, then the CM1 is the likely cause of the symptoms and papilledema. In another article, he suggested always attempting medical therapy for potential PTC (e.g., acetazolamide) before surgical treatment of CM1. Kandasamy et al. advocated for the treatment of whichever condition seems most likely based upon the patient’s symptoms.

Our case presents a diagnostic dilemma as to which condition, PTC or CM1, is causative of the papilledema, whether the conditions are both manifestations of the same cause, or whether their simultaneous presentation is coincidental. She was a woman in her childbearing years, which is typical of both conditions. However, our patient had no obesity, headaches, visual field loss, or other symptoms of either condition. Her papilledema is a possible manifestation of both PTC and CM1, but far less common in CM1. Lumbar puncture was contraindicated and not performed to evaluate the CSF pressure and contents due to her CM1 and a 30-degree test was not performed, so a definitive diagnosis of PTC cannot be made. We were also unable to perform any CSF flow studies due to her lack of following up for care. However, given the near-complete effacement of the subarachnoid space at the FM seen on the MRI, which is likely obstructing the CSF flow, this may be a case of the CM1 causing the intracranial hypertension and papilledema. Nevertheless, it is still unusual that the patient was asymptomatic and remained so. If the CM1 is severe enough to cause papilledema, additional neurological symptoms should be present. In fact, this is the first reported case of CM1-induced papilledema without additional neurological symptoms.

**Conclusion**

Papilledema is a condition not uncommon to present to optometrists. In an asymptomatic case like this, an eye exam is the only way the patient’s neurologic problem will be found. While the typical causes are well known, this case emphasizes the importance of obtaining neuroimaging on patients with papilledema, as the cause may be outside the typically expected list of culprits. Additionally, the actual pathogenesis of the papilledema may be elusive. The optometrist’s role is not only as the primary identifier of the problem, but to monitor the patient for resolution after referral to an appropriate specialist.

**References**


