An Inquiry-Based Approach to Teaching Sphero-Cylindrical Ametropia

MEWDS: a Teaching Case Report

When a Red Eye is More than Meets the Eye: a Teaching Case Report on the Public Health Role of the Eyecare Provider

Progressive Supranuclear Palsy: a Teaching Case Report

Is Educational Theory of Use in Optometric Education?

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An Inquiry-Based Approach to Teaching Sphero-Cylindrical Ametropia
Nicole M. Putnam, PhD | Optometric Education: Volume 45 Number 3 (Summer 2020)

PDF of Article

Background

A clear understanding of the optics of the eye remains a fundamental basis of optometric education. As technology continues to improve, modern instrumentation and corrections can utilize measurements that are more sophisticated and manufacturing standards that are more precise. Optometrists need a clear understanding of the optical basis of sphero-cylindrical ametropia and the relationship between the refractive state of the eye and its correction to be able to evaluate and utilize new and evolving technologies.

Students in the first-year program at the Midwestern University Arizona College of Optometry (AZCOPT) are required to take the OPTOG 1540 Geometrical and Physical Optics I course as part of the OD-1 curriculum. The course is in the first quarter of the optometric program and includes 30 hours of lecture and 10 three-hour laboratory periods over the course of the 10-week quarter. Lecture and laboratory materials were developed over the course of five administrations with the same course director. The entire class (approximately 55 students) attends lecture together and two laboratory sections are scheduled on the same day with approximately half the class randomly assigned to each section at the beginning of the quarter. The laboratory is equipped with eight stations with optical rails, lights, optical elements and other supplies. Most labs contain traditional demonstrations and activities designed to give students hands-on experience with the lecture content. Labs are designed to take place after the introduction to material in lecture, and each laboratory has a pre-lab reading assignment. A pre-lab assignment and a post-lab assignment are graded.

At AZCOPT, students learn the fundamental concepts of spherical and sphero-cylindrical ametropia in their first quarter of study in the Geometrical and Physical Optics I course. For many years, sphero-cylindrical ametropia was taught through a series of approximately six hours of lecture and one three-hour laboratory. Sphero-cylindrical ametropia is one of the most clinically relevant topics covered in the first-year optics curriculum and one of the most challenging. Some of the related course learning goals are summarized in Table 1. The introduction first-year optometric students receive in this course forms the basis upon which they learn advanced concepts related to spherical and sphero-cylindrical ametropia in optometric methods, contact lens and low vision courses and ultimately translate to clinical knowledge. Understanding cylindrical and sphero-cylindrical optics necessitates thinking about an optical system in three-dimensions, and students often comment that the relationship between the power of the eye/lenses and the orientation of images is difficult to visualize. Students also share that it is difficult to simultaneously consider the relationship between the power of the eye (keratometry), the power of a prescription (ophthalmic optics) and the various notations used to represent these components (transcription). Students often show a gap between the knowledge they should have obtained about sphero-cylindrical optical systems and what they display on examinations and in their methods courses.

<table>
<thead>
<tr>
<th>TABLE 1</th>
<th>Learning Goals Related to Spherical and Sphero-Cylindrical Ametropia in OPTOG 1540 Geometrical and Physical Optics I</th>
</tr>
</thead>
<tbody>
<tr>
<td>• To be able to explain the optical basis of refractive error, its measurement and its correction</td>
<td></td>
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<tr>
<td>• To understand thin lens models of ametropia</td>
<td></td>
</tr>
<tr>
<td>• To understand changes in refractive power between the spectacle and corneal planes</td>
<td></td>
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<tr>
<td>• To understand the relationship between power and axis meridians</td>
<td></td>
</tr>
<tr>
<td>• To understand the difference between plus and minus cylinder notations and convert between the two</td>
<td></td>
</tr>
<tr>
<td>• To understand the characterization of astigmatism in the human eye</td>
<td></td>
</tr>
</tbody>
</table>

There are many possible approaches to the design and execution of learning activities in the laboratory setting. There is a long history of laboratory use in science education, which is believed to provide students with valuable experiences with science concepts and the processes required in scientific fields. An inquiry-based laboratory is one way to increase the depth of conceptual and quantitative...
Inquiry-based teaching methods were used in an optometric setting by the author as a graduate student instructor in an ophthalmic aberrations laboratory. This lab was an additional lab in that course and allowed students to look at their own aberrations using a computer simulator. Then they worked in groups using a case-based approach to further investigate one condition or clinical problem. One major question remains. Are inquiry-based labs well-suited to replace traditional labs in an optometric setting, or are they better suited to be used to strengthen and widen the scope of knowledge? In addition, if used in combination with a traditional laboratory, is there an ideal order to present the labs: inquiry then traditional or traditional then inquiry?

Methods

Two innovative and interactive laboratories with an inquiry-based and a traditional component on sphero-cylindrical ametropia were implemented, replacing the one traditional lab on sphero-cylindrical lenses and the laboratory practical exam. To increase the depth of knowledge of the refracting states of the eye and their correction, inquiry-based sphero-cylindrical ametropia laboratories provide students with an opportunity to further explore and work with eye models and their own visual systems to increase the depth of their understanding of these concepts. This innovative laboratory approach can be challenging and lead to periods of questioning, but ultimately the goal is to extend the students’ fundamental understanding of a set of topics (e.g., Table 1) that form the basis of understanding in many future classes and future clinical practice. Inquiry-based laboratory activities can be challenging to students as they are more open-ended with students taking ownership over the scientific process. They often hit a point where they are stuck or do not know how to proceed, as is the case in the practice of vision science and optometry. As a result, more instructors and teaching assistants facilitated the inquiry-based lab periods to help the students design and execute their investigations compared to a traditional lab setting. (Most sessions had three to four for inquiry vs. two for traditional). An inquiry-based laboratory was added to the earlier portion of the course to introduce the concept, replacing a practice lab practical, so inquiry was not a completely new process when the sphero-cylindrical ametropia labs were administered towards the end of the quarter.

The traditional sphero-cylindrical ametropia lab was taught for five years at AZCOPT for one laboratory period for a total of three hours. The lab began with a guided set of questions in response to which the students looked through or made observations about cylindrical lenses of various powers, combinations of lenses, pinholes and slits. They were asked to make a variety of observations including blur associated with points and extended objects, image movement, image formation and other simple observations. This introduction was followed by step-by-step instruction where students set up different sets of lenses on the optical rail. This included image formation with a single positive cylindrical lens and a variety of combinations of spherical and cylindrical lenses and screens to represent different types of regular astigmatism. For the eye models, students were also walked through the process of correcting these eyes with additional trial lenses.

Two inquiry-based laboratories on cylindrical and sphero-cylindrical lenses and sphero-cylindrical ametropia were implemented in addition to shortened versions of the traditional labs by increasing the time from one laboratory period to two. The new inquiry-based laboratories each included a traditional component and an inquiry component. All students received both approaches to the same content on the same day. To determine whether the order in which the two lab sections are offered matters for the learning outcomes, one section received the traditional lab first and the other received the inquiry-based lab first. These were immediately followed by the other component. The order was then switched the following week so...
all students experienced both orders to limit differences between groups.

Figure 1. Left: This station demonstrated relationships between object and image distances, cylindrical lens orientation and image orientation. Right: This station used vases of various sizes filled with water to demonstrate relationships between index of refraction and curvature. Click to enlarge

Figure 2. Top left: This station used a model eye setup on the optical rail to demonstrate spherocylindrical ametropia and correcting lenses (glasses vs. contact lenses). Top middle, top right, top bottom: This station used eye models from Pasco to demonstrate phenomena related to spherocylindrical ametropia. A variety of options allowed the creation of various ametropic eyes: the ability to fill the eyes with water; objects on the optical rail or a computer; spherical and cylindrical lenses to act as the eye or correction; apertures to act as pupils; a moveable “retina” screen. Click to enlarge

The inquiry-based labs were similar in format. In the first 15-20 minutes, students split into two to three groups to rotate through starter stations where they observed phenomena related to cylindrical and spherocylindrical lenses (week 1) and spherocylindrical ametropia (week 2). Each station began with a facilitator introduction to the station with demonstrations followed by time for the students to explore and discuss. The stations included:

Week 1: spherical and spherocylindrical lenses (Figure 1):

1. Cylindrical and spherocylindrical lenses and image formation
2. Index of refraction and curvature
3. Looking through cylindrical and/or spherical lenses and slits/pinholes. Various +/- cylindrical and spherical lenses, pinholes and slits were provided from trial lens kits as well as near and distance eye charts and contrast charts

Week 2: spherocylindrical ametropia (Figure 2):

1. Astigmatic eye models on optical rails and their correction
3. Computer simulations of spherical and spherocylindrical imaging, e.g, sight simulator from prescription (http://www.billauer.co.il/simulator.html)

The students formed groups of three to four people in order to ask and refine a question and design and execute their own in-depth investigations into one aspect of cylindrical and spherocylindrical lenses (week one) and spherocylindrical ametropia (week 2). Members of each group presented their findings to the rest of the class in the final 20 minutes, as they had become the subject-matter experts. The total time for the inquiry-based lab component was approximately two hours.

A short quiz and survey were administered each week in the middle of the laboratory period after the first part of the lab was completed (either traditional or inquiry-based) to assess student knowledge and perception. Each lab was also followed by a quiz and survey in the next lecture for the same reasons. To ensure that any differences did not impact student assessment, the quiz scores were used exclusively as research data and did not count in the course grade. Students received full marks for
finishing the quizzes and surveys. An analysis of the grade average and distribution was performed, and students were
informed of any aggregate differences for their information.

The quizzes and surveys included several content-based questions. Students were asked to rate the lab they just completed
(inquiry-based or traditional) in the middle of the lab period, and on the following day separately rate both labs on a scale of 1-
10. Students were also asked about the impact of the lab(s) on their learning experience and about the order of the labs (only
in class following both labs). They were asked opinion-based questions (rated disagree to agree) regarding their ability to solve
basic and complex optics questions and if the lab enhanced their ability to solve problems and their conceptual knowledge.
There were additional open-ended questions. An analysis was performed to assess student performance on these quiz/surveys.

Fifty-six students were enrolled for the first time in the OPTOG 1540 Geometrical and Physical Optics I course when two new
inquiry-based laboratories were implemented in addition to traditional laboratories. All students participated in laboratories,
quizzes and examinations as a part of the normal class activities. An analysis was performed to assess student performance on
these quiz/surveys. Midwestern University IRB approval was received for this study (AZ #1075). Prior consent for disclosure of
academic records was not required for this study, consistent with the Student Handbook, Appendix 3.D.2.g. Confidentiality was
maintained and no names were used in the compiled data file. There was a risk of loss of privacy or breach of confidentiality,
but a de-identified records review and data retrieval minimized such risks.

Results

The 56 students enrolled for the first time in the OPTOG 1540 Geometrical and Physical Optics I course participated in both
new inquiry-based laboratories. They also completed traditional laboratories on the same day in a randomized order as
described in the methods section. These were part of the requirements for the OPTOG 1540 Geometrical and Physical Optics I
course. All 56 students completed the two survey/quizzes administered in the lab period, 52 students completed the in-class
survey/quiz after the first week, and 55 students completed the in-class survey/quiz after the second week.

All students successfully participated in the starter stations and recorded vocabulary and questions. Figure 3 illustrates
students engaging with the starter materials. The entire group brainstormed questions together resulting in the investigation
questions summarized in Table 3.

Students then proceeded with an investigation with the help of the instructor and facilitators. The inquiry labs culminated in
the delivery of short presentations. Pictures from the investigations are shown in Figure 4.

During the first week’s spherical and sphero-cylindrical lenses laboratories, 56 students completed a quiz and survey in lab
immediately after either the inquiry-based lab (27) or the traditional lab (29). 52 students completed the quiz and survey in the
next lecture. Results comparing performance on the four quantitative questions revealed no significant differences. Results
comparing student ratings of the labs in their usefulness in gaining knowledge about cylindrical and sphero-cylindrical lenses
on a scale of 1-10 in whole numbers where 1 is not useful and 10 is extremely useful also revealed no significant differences.
These results are in Table 4. During the second week’s sphero-cylindrical ametropia laboratories, 56 students completed a
quiz and survey in lab immediately after either the inquiry-based lab (29) or the traditional lab (27). Fifty-five students completed the quiz and survey in the next lecture. Results comparing performance on the four quantitative questions and the questions asking students to rate the labs are presented in Table 5. There was again some variability, but no significant differences.

As discussed in the methods section, the survey included some open-ended questions. The responses to these questions highlighted the value of the diverse laboratory experiences. Selected responses are in Table 6 and Table 7. In addition to the comments in the tables, numerous comments indicated the students would like more time; some felt they had to rush. This limitation of this first administration is addressed in the discussion section. Numerous comments highlighted that the students felt the traditional lab should take place before the inquiry-based lab, and many students requested more guidance.

### Discussion

Inquiry-based labs are designed such that students will learn process skills relevant to optometric practice such as problem-solving, data analysis and communication along with the content goals. The new laboratories presented here centered on a difficult topic: cylindrical and spherocylindrical lenses and spherocylindrical ametropia. The goal of participating in these interactive laboratory sessions was for students to explore and develop a deeper understanding of the individual optical concepts and how they come together. Students also had the opportunity to participate in the traditional labs, which allows assessment of whether the inquiry-based approach would be best implemented before, after, or in place of the current teaching methods.

Although there was no statistical difference in the overall final exam grades or overall course performance, a number of outside course factors should be considered in order to evaluate the impact of the inquiry-based labs. The year without the inquiry-based labs had a laboratory practical examination prior to the written final examination. This resulted in a sequence such that the traditional astigmatism laboratory was completed weeks earlier in the quarter allowing more time for review
prior to the final examination. The material was also examined prior to the final exam on the practical, hands-on examination. There were also some areas identified in need of improvement after the first implementation of the inquiry-based labs, specifically a lack of time to finish all parts of the labs that may have played a role in performance on these items. The true test will be students’ future performance in upper-level courses and on the national board exam, which would make interesting follow-up studies.

It is extremely important for optometrists to be careful consumers of new technology. An increasing number of measuring and correcting devices are utilizing the knowledge of ocular aberrations in their design. It is not feasible to adopt all new equipment or work with all companies developing correcting devices. Therefore, having a deeper understanding of the vocabulary used and the patient populations for which this may provide maximum benefit is important for future success. Astigmatism is one of the most basic aberrations and ultimately one of the easiest to understand. Although many students already know how to transpose a prescription or have heard of astigmatism through personal experiences, the 3D and complex nature of the concepts related to spherico-cylindrical ametropia and its correction remain a challenge for many students. This is a topic they will see again in many classes and in practice, and an innovative approach to extending their knowledge early in their careers could be extremely meaningful and have long-reaching impact.

Preliminary results suggest some value in the addition of inquiry-based labs in an optometric setting. Some form of traditional instruction and/or laboratory demonstration may always be needed or at least enhance the experience. Many additions and revisions to these laboratories can be considered with the goal of an improved educational experience and enhanced long-term application of knowledge. One consideration is a variation on the hybrid experience whereby one week of traditional laboratory and one inquiry-based lab occur rather than a traditional and an inquiry component both occurring twice on two separate weeks. Other changes, such as submission of questions for investigation in advance through homework or a pre-laboratory activity or assignment, may simplify parts of the experience. Another consideration could be a different approach to the presentation of results such as having students deliver their presentations the following week or in class or completing some sort of laboratory report. Inquiry-based laboratories can often be more time-consuming for reasons that include the fact that students may face situations that are more challenging in which they feel stuck. Working through these elements can lead to greater ownership of the learning process and the development of useful skills for the future, but it requires more time and many of these suggestions address that. One consideration is that inquiry-based labs are extremely well-suited for strengthening and widening the scope of knowledge rather than introducing a topic.

Conclusion

New inquiry-based labs were introduced in the Geometrical and Physical Optics I course at AZCOPT. Students were put outside their comfort zone and extended their knowledge of astigmatism. The study implemented and utilized innovative teaching strategies in AZCOPT’s optometry program. No statistical differences in exam or course performance were found with the use of traditional vs. inquiry laboratories. At least some form of traditional instruction seems to be required in order for students gain the maximum benefits from an inquiry-based laboratory. The results may inform program faculty regarding the implementation of other innovative teaching methods such as problem-based learning, a flipped classroom, etc. Finally, the results may provide additional evidence to guide educational practices for optimal outcomes on the national certification examination.

Acknowledgments

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References


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Introduction

Healthcare education is carried out primarily by instructors repeating the way in which they were taught. There is a sense that teaching is an art where only content knowledge and natural talent are needed to excel. New trends, such as the “flipped classroom,” may be applied without an understanding of the principles behind the educational strategy. If the method fails to achieve desired results, blame is placed on the method. Without an understanding of the theory that underlies newer teaching methods, instructors risk misapplying strategies, which can result in a lack of successful implementation for both teachers and learners. It is reasonable to believe that optometric education has similar challenges.

Theory is a conceptual understanding of how things work. The usefulness of theory may be more readily understood from an example from the ocular disease literature. Glaucoma is a common eye disease that can result in permanent vision loss. While the pathogenesis is incompletely understood, a number of theories exist. Theories include vascular (insufficient blood supply due to increased intraocular pressure [IOP]), biomechanical (increased IOP leading to strain of the lamina cribrosa) and biochemical (neurodegeneration from biochemical mechanisms). As well, the role of cerebrospinal fluid is seen as potentially important. These theories were built from an impressive amount of quantitative and qualitative research. The vascular and biomechanical theories are applied by treating glaucoma with IOP reduction. In addition, theories drive research and are modified based on the results of the studies. Challenging the existing paradigm is how progress is made. The same principles apply to optometric education.

When research is not based in theory, the results can be misleading. To make a point about the dangers of post hoc analysis of data, Emily Chew performed an analysis of the age-related macular degeneration data from the Age Related Eye Disease Study to show that vitamin and mineral supplementation effectiveness depended upon the zodiac sign of the subject. This was intended to be an obviously incorrect result because zodiac sign is not included in the theories of the pathogenesis of age-related macular degeneration. Similarly, if optometric education research is carried out without attention to underlying educational theory, the results may not have merit.

How people learn is also incompletely understood. There are many theories (based on extensive research) of how people learn and they may be grouped by discipline, such as bioscience, education/learning or sociocultural perspectives. Each theory in itself is only a partial explanation of how learning occurs and many have overlapping concepts. Therefore, when creating an optometric educational plan it is necessary to look to multiple disciplines and their theories to illuminate each aspect of the plan and focus on what is likely to be useful. Theory helps form a scaffold upon which good instruction can be built and allows instructors to understand why and under what circumstances certain practices are successful. The informed optometric instructor, therefore, is less likely to deviate from the model. Also, teaching based in theory, as in the ocular disease example, can be studied, which leads to modifications in the theory and enhanced educational outcomes.

This paper examines some of the theories of learning that might be useful to optometric educators and shows how they can be applied to designing a skills curriculum.

Theories of How Learning Occurs

Theories that aim to explain how learning occurs include the following.

Theories in bioscience focus on how the brain learns. Because discovery comes from science, it is sometimes easy to forget that these are theories and not truth. Neuroscience studies learning and memory formation looking at structure and molecular and cellular function of the brain and has used functional magnetic resonance imaging studies to understand cognition in humans. This research has allowed for some general principles to be developed that can be applied in optometric education. These include:

- allow for repetition of material from varying perspectives
- go deeper rather than touching superficially on many topics
- reward and reinforce (this is vital)
- actively engage the learner
• create a moderate stress level for the learner
• allow for adequate rest
• integrate multimodal (auditory and visual input) information rather than expect multitasking

Motor control theories are useful in learning skills such as automaticity and skill expertise (performance is automatized with practice allowing for attention to other factors), deliberate practice (deliberate practice is needed to achieve excellence), and challenge-point framework (learning is maximized when the task is within the ability of the learner and complexity is gradually increased with experience).

Education/learning theories can be categorized into five additional orientations: behaviorist, cognitivist, humanist, constructivist and social theories of learning. In the behaviorist view, learners are passive and a stimulus produces a reaction in the learner. Therefore, giving people feedback to allow them to change their behavior is central in this theory. In the cognitive view, learners think about the information, process it and act upon it in different ways depending on the situation. This orientation has produced theories of clinical reasoning, theories on the development of expertise and cognitive load theory. In the humanist perspective people try to achieve their maximal potential. The concepts of self-regulation, self-directed learning and self-assessment align with this orientation. Constructivist learning theory is a refinement of cognitive learning theory and is derived from constructivist epistemology where the “focus is on the mental representation of information by the learner.” The learner reconstructs long-term memory representations to be consistent with new information from the environment and experience. For this to occur, learners must process the new material and integrate it with existing understandings to form a new cognitive structure that is unique to them based upon their own process of learning.

Social cognitive theory was formed by blending the behaviorist approach, which emphasizes the importance of the environment on behavior, with the cognitive approach, which emphasizes the importance of cognition on learning and functioning. Learning is seen as a social process with reciprocal interaction between environmental, personal and behavioral determinants. Each of these determinants influences the others with varying outcomes depending on the dominant factor. In this theory humans have five capabilities:

• symbolizing capability allows us to use symbols internally to represent actions so we don’t have to carry out each action to determine the outcome
• in forethought capability we can anticipate the likely outcomes of our behavior so we can plan for those outcomes
• vicarious capability allows us to learn by observing others
• self-regulatory capability allows us to regulate our behavior by our internal standards
• self-reflective capability allows us to look critically at our experiences and think about our thought processes

In situated learning theory, learning is seen as a socio-cultural process tied to context with its social relations and practices. The term communities of practice describes the activities of a group of people with a shared interest. In legitimate peripheral participation, learners start at the periphery of the community by observing and performing simple tasks and then gradually move more centrally assuming the roles, skills, norms and values of the culture as an active participant. Learning occurs in the workplace from an individual and social perspective. In the related concept of experiential learning, learning occurs through reflection on experience in practice. Reflection is an important concept that is seen in multiple theories of learning.

Application to a Curriculum Element

How can any of these theories be applied in optometric education? As an example, optometric educators may be tasked with reviewing the psychomotor skills curriculum of their school or college. It would behoove them to look to best practices in healthcare education and understand the theoretical underpinnings when designing the program. Table 1 summarizes the design elements that could be used to develop a procedural skills stream in the curriculum renewal process. The theoretical basis supporting the design element is given as is an example of how it can be operationalized. Assessment methods are also reviewed.
Development of clear outcomes or goals enhances learning by allowing the learner to develop strategies to reach the goal and is based in social cognitive theory. In social cognitive theory, learners are active agents and learning is influenced by their personal goals, values, attitudes, knowledge and experience. Once the goals are articulated to them, learners are able to take on the goals and monitor their progress towards the goals. Humans are able to anticipate the likely outcomes of actions, and this forethought, as a capability, helps in achievement of goals under this theory. Teachers need to support learners in regulating their own learning. This is operationalized by writing learning objectives. Learning objectives are clear statements of what ability the learner will have at the end of the educational session. For example, “At the end of this course the student will be able to perform retinoscopy on a normal adult person and obtain a result within the repeatability of the technique.”

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**Longitudinal, repetitive, progressive skill development** is based on bioscience theory of motor control such as the Fitts and Posner model of automaticity in which well-learned skills become automatic with practice. Ericsson’s idea of deliberate practice to develop expertise requires lengthy, repetitive and dispersed domain-specific practice. Challenge-point framework states that an optimal challenge point exists when learning is maximized and detriment to performance is minimized. That is, the difficulty of the task is matched to the learner’s abilities. To operationalize these concepts, skill training should start in a laboratory setting where the students practice on themselves. Ideally, the sessions should be timed to one or two hours and repeated frequently. Introduction to more complex scenarios would occur during clinic rotations in primary care and then in specialty care settings.

**Guided practice with distributed feedback** is based upon the aspects of social cognitive theory that state that individuals develop positive self-efficacy through practice with positive feedback. The feedback should be constructive because without it performance achievement is reduced. Feedback is also helpful because it provides information the individual can use in reflective practice. This aspect is vital to the learning process. Indeed, the ability to self-assess is tied to the ability to reflect on practice. As described by Mann, to operationalize this the learner should be provided with immediate error correction while performing the procedure but limited verbal guidance and coaching. This is because the coaching takes the learner’s attention away from the task, overloads working memory and the learner may become reliant on the educator to complete the task. Then, at the end of the skill performance, feedback is provided. According to Nicholls et al., “Feedback is the single most influential teaching practice to promote motor learning.” Feedback should continue to occur in the clinical setting and be based on direct observation.

The elements of **transfer of skills to a clinical setting** with a cultural milieu consistent with real practice and gradual transition to independent practice with initial observation are grounded in the concepts of situated learning, communities of practice and

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**Table 1. Click to enlarge**

<table>
<thead>
<tr>
<th>Design Element</th>
<th>Theoretical Basis</th>
<th>Example of operationalization</th>
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</thead>
<tbody>
<tr>
<td>1. Development of clear required outcomes</td>
<td>Social cognitive theory (self-efficacy)</td>
<td>Form clear learning objectives</td>
</tr>
<tr>
<td>2. Demonstration of procedure and task-relevant knowledge</td>
<td>Social cognitive theory (vicarious capability and self-efficacy); constructivism; cognitive load</td>
<td>Use online learning methods to deliver the relevant knowledge ahead of the skills session. Demonstrate the skill using Peyton’s Four-Step Approach.</td>
</tr>
<tr>
<td>3. Longitudinal, repetitive and progressive skill development</td>
<td>Motor control theory; deliberate practice</td>
<td>Lab training should allow for repeated practice in episodes of short duration until the skill is automated. This is followed by progressive exposure to more difficult conditions.</td>
</tr>
<tr>
<td>4. Guided practice with distributed feedback</td>
<td>Social cognitive theory (self-efficacy, self-regulation and self-evaluation); reflection</td>
<td>Specific formative feedback during the lab training by instructors is essential. The course should continue to occur in clinical practice.</td>
</tr>
<tr>
<td>5. Transfer of skills to a real practice setting with a cultural milieu consistent with real practice</td>
<td>Situated learning; legitimate peripheral participation</td>
<td>Plan for early-clinical exposure (in the first year) self-observation progressing gradually to assisting in practice and then to direct one-on-one patient care.</td>
</tr>
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**Longitudinal, repetitive, progressive skill development** is based on bioscience theory of motor control such as the Fitts and Posner model of automaticity in which well-learned skills become automatic with practice. Ericsson’s idea of deliberate practice to develop expertise requires lengthy, repetitive and dispersed domain-specific practice. Challenge-point framework states that an optimal challenge point exists when learning is maximized and detriment to performance is minimized. That is, the difficulty of the task is matched to the learner’s abilities. To operationalize these concepts, skill training should start in a laboratory setting where the students practice on themselves. Ideally, the sessions should be timed to one or two hours and repeated frequently. Introduction to more complex scenarios would occur during clinic rotations in primary care and then in specialty care settings.

**Guided practice with distributed feedback** is based upon the aspects of social cognitive theory that state that individuals develop positive self-efficacy through practice with positive feedback. The feedback should be constructive because without it performance achievement is reduced. Feedback is also helpful because it provides information the individual can use in reflective practice. This aspect is vital to the learning process. Indeed, the ability to self-assess is tied to the ability to reflect on practice. As described by Mann, to operationalize this the learner should be provided with immediate error correction while performing the procedure but limited verbal guidance and coaching. This is because the coaching takes the learner’s attention away from the task, overloads working memory and the learner may become reliant on the educator to compete the task. Then, at the end of the skill performance, feedback is provided. According to Nicholls et al., “Feedback is the single most influential teaching practice to promote motor learning.” Feedback should continue to occur in the clinical setting and be based on direct observation.

The elements of **transfer of skills to a clinical setting** with a cultural milieu consistent with real practice and gradual transition to independent practice with initial observation are grounded in the concepts of situated learning, communities of practice and
legitimate peripheral participation. Inherent in these concepts is the idea that learning is tied to its context, social relations and practice. The community of practice is a group of people who pursue a shared enterprise. Initially novices participate peripherally (for example by observation) and then gradually move more centrally when they gain the skills, norms, values and culture of the community. To operationalize this theory optometric students should be observing in clinic during early years of optometric education. Then they may progress to assist with skills such as pre-testing and subsequently move on to assisting with examinations before they are competent enough to conduct skills independently. This can happen concurrently with laboratory practice.

**Assessment**

Assessment should be aligned with the learning objectives of the procedural skills training program. Learning objectives are what the learner is to achieve, while learning outcomes are what the learner did achieve. Miller’s framework (pyramid) for clinical assessment (Figure 1) describes four levels of ability: knows (knowledge), knows how (competence), shows how (performance), does (action). Consistent with Miller’s pyramid, a system of assessment should be in place that addresses each level in the pyramid. Written examinations such as multiple-choice questions or long-answer questions can be used to assess the concepts behind the procedures for the “knows” and “knows how” components. Direct observation of the procedures using checklists as the procedures are performed in the lab and a summative pre-clinical competency examination are at the level of “shows how.” Direct observation of optometric skills in clinic can be used for the “does” level. In addition, students should be encouraged to keep logbooks for procedures and write reflective e-portfolios to encourage self-assessment and self-directed learning.

**Conclusion**

This paper presents a variety of theories as lenses through which to view optometric education. Each theory illustrates an aspect and allows us to focus on a goal in education to address it more effectively. Some theories are more relevant to a particular educational problem than others. The theories in this paper are not exhaustive of those in the literature; they represent only a sample. Ultimately, theories are challenged through research, which allows for development and improvement over time. An example from general education is the popular theory of learning styles has been disproven through research in application of the theory to teaching and learning. A research question pertaining to the skills training example could be “Does using Peyton’s Four-Step Approach to teaching a skill improve performance on skills assessment?” This could be studied using a randomized controlled trial design in which traditional teaching is done in one lab section, Peyton’s approach is done in another, and the skills assessment results are compared.

Educational theory allows both educators and learners to develop a perspective and discourse around best practices. The discourse of a discipline provides a language for representing its work and allows for discussion and reflection. It behooves us as optometric educators to challenge ourselves to become familiar with the relevant theories of healthcare education, to apply them in our work and study the results. This will move us toward the important goal of creating a culture of using theory-informed approaches in optometric education.

**References**

SCCOMBKU Graduate is Winner of 2020 Student Award in Clinical Ethics
| Optometric Education: Volume 45 Number 3 (Summer 2020)

PDF of Article

ASCO and its Ethics Educators Special Interest Group are pleased to announce Ryan Yuan, OD, as the winner of the 2020 Student Award in Clinical Ethics. Dr. Yuan graduated this year from Southern California College of Optometry at Marshall B. Ketchum University. His winning essay, “Ethics of Ocular Pain Management,” appears below.

The Student Award in Clinical Ethics competition, sponsored by Alcon, is open to optometry students during any point in their professional program at an ASCO-affiliated school or college of optometry.

The winner receives an engraved plaque and $1,000. ASCO thanks all the students who submitted essays.

Ethics of Ocular Pain Management

By Ryan Yuan, OD

Case Description

RR is a 55-year-old Native American male with a complicated ocular history. Diagnosed at an early age with keratoconus, he underwent penetrating keratoplasty OU in 2012. He later contracted bacterial and fungal keratitis on the corneal grafts and developed secondary steroid-induced glaucoma, which was managed with tube shunts OU. In 2018, he experienced blunt force trauma OS, which caused corneal wound dehiscence, displacement of his posterior-chamber intraocular lens, and a retinal detachment. After open-globe repair, RR was prescribed scleral contact lenses OU; however, the poor fit of the lenses caused mechanical exposure of the tube shunt OD over time. Endophthalmitis caused by methicillin-resistant *Staphylococcus aureus* eventually developed OD, and the condition progressed to a blind, painful, hypertensive eye. RR had been treated most recently with diode cyclophotocoagulation and retrobulbar chlorpromazine to manage the pain.

RR’s medical history is remarkable for opioid abuse. According to his hospital records, RR had been prescribed 19 pain medications from 16 different providers over the years. He presented to our clinic asking for specific medication to alleviate the pain from his phthisical, hypertensive right eye. Given his ocular treatment history and prior opioid use, what ethical factors should be evaluated prior to initiating therapy?

Ethics Considerations

Optometrists are held to an ethical standard that requires morally responsible prescribing. For those with the relevant credentials, should opioids be prescribed to a patient with a history of misuse? Are there long-term consequences that need to be assessed? Should his request for treatment affect the clinical decision-making? Is there an optometric standard of care that can be applied?
The Opioid Epidemic

In 2018, the Centers for Disease Control and Prevention (CDC) estimated that more than 10 million people in the United States age 12 years and older reported misuse of prescription opioids. It has also been estimated that more than 47,000 deaths, or nearly 129 deaths per day, from opioid overdose occurred in 2017. Examples of opioids include, but are not limited to, natural opioids (morphine, codeine), semi-synthetic opioids (hydrocodone, oxycodone) and synthetic opioids (fentanyl, tramadol).

Few ocular conditions warrant opioid use; however, opioids may be prescribed for post-operative refractive surgery pain and can be considered in serious cases of ocular trauma, endophthalmitis, corneal hydrops or neovascular glaucoma. Credentialed optometrists who hold a Drug Enforcement Administration (DEA) license are limited in the type and quantity of opioids they can prescribe. Their role in the opioid epidemic remains to be seen. Nevertheless, any physician with the ability to prescribe potentially addictive and fatal medications must prescribe with ethical considerations in mind.

Optometric Values

Like all healthcare providers, optometrists abide by codes of ethics and values that are reminiscent of the historical Hippocratic Oath; namely, the principles of beneficence, non-maleficence, autonomy and justice.

Beneficence: The “do good” principle entails that the optometrist must take actions that are in the best interest of patients and their families. On the surface, it seems easy to apply in the everyday life of optometrists as they adhere to the optometric standard of care with refractions, ocular health examinations and the sale of ophthalmic devices with the goal of providing right and excellent care. Probing deeper, however, the optometrist must ask how to “do good” as it relates to pain management, such as in the case of RR. Utilizing the knowledge of ocular pathophysiology and the corresponding therapeutic options, the optometrist must weigh the risks and benefits of each modality. Would it be beneficial not to prescribe medication to RR? He has a legitimate reason for pain, but would his pain abate with over-the-counter oral analgesics, or does it warrant a prescription for oral opioids?

Non-maleficence: The optometrist may have good intentions in prescribing; nevertheless, a second layer of ethical consideration requires him/her to assess whether the prescribed treatment would cause harm, either immediately or in the future. It is well-known that physical dependence can form when opioids are used regularly, and the DEA ranks controlled substances (I-V) based on potential for abuse. Hydrocodone, specifically, was a widely used Schedule-III drug that was moved to Schedule II in 2014 due to higher addictive properties. In most states, DEA-licensed optometrists can prescribe Schedule III-V drugs. Specific conditions for Schedule II allow optometrists to continue prescribing hydrocodone. Though the immediate effects of opioids would bring pain relief, long-term use has been linked to increased tolerance, depression and risk of death related to overdose.

Given RR’s long history of opioid misuse, would prescribing cause him long-term harm? Do the benefits of opioid treatment today outweigh possible future risk? By not prescribing, will he become more volatile in his behavior and drug-seeking tendencies?

Autonomy: Because RR specifically requested pain medication for his ailment, the principle of patient autonomy presents. According to this doctrine, the optometrist must regard the patient’s requests when it comes to treatment options; that is to say, to a certain extent. If a patient requests medical treatment when there are no clinical findings to justify medication, the optometrist should not oblige just to satisfy the patient’s wishes. The doctor’s clinical judgment must be respected in all patient-doctor relationships. On the other hand, the optometrist ought to be morally responsible by informing and including the patient in all treatment decisions.

Is it ethical to refrain from presenting opioids as a treatment option if there is potential for drug abuse? What if the patient’s wishes differ from the optometrist’s desired treatment? Medical paternalism is an exception to patient autonomy in cases involving young children or the mentally incompetent. Could it be argued that RR is not fully competent in his decision-making due to his addictive tendencies?

Justice: Philosophically, justice typically asks who to “save” given hypothetical scenarios of scarce resources and patients suffering from differing levels of morbidities. In the context of optometry, justice can be applied instead as the duty of the optometrist to treat all patients fairly and ethically. Optometrists must adhere and apply to the relevant standards of care accordingly. In cases such as RR, there are no customary opioid-related treatment recommendations. In fact, the Wills Eye Manual only mentions opioids in the treatment of chemical burns, band keratopathy and dacryocystitis. The CDC and American Optometric Association provide loose guidelines for prescribing and propose that an opioid prescription of three days or less will be sufficient for most conditions. From this it can be inferred that opioid prescriptions should be considered on a case-by-case basis as not all situations are the same.
Would it be ethical to refer the patient to another provider who may be more qualified to handle his type of care? If RR was a personal family member, would the level and method of care be handled any differently?

Conclusion

All things considered, RR’s case was co-managed with an ophthalmologist, and it was decided that prescribing a five-day supply of Tylenol 3 (which contains acetaminophen and codeine) would be both beneficent and justified. RR, on the other hand, quickly reported that Tylenol 3 would not be strong enough for his pain. The ophthalmologist then recommended another retrobulbar injection of chlorpromazine. Chen et al. reported that retrobulbar chlorpromazine is an effective analgesic with an efficacy lasting from three months to a year. This treatment option would protect RR from potential opioid abuse, and the dialogue allowed RR to have a say in his medical care. However, the fact that RR recently had a retrobulbar procedure and still reported pain reflects the reality that 33% of patients with retrobulbar chlorpromazine treatments may need alternative interventions to manage pain. With RR’s ocular history and current condition, the last treatment option discussed was enucleation for his phthisical eye. Idowu et al. reported that long-term pain relief and discontinuation of oral analgesics was achieved in 100% of patients with enucleated or eviscerated eyes. It remains to be seen whether this treatment would provide RR with the lasting pain relief he seeks. Nevertheless, the next ethical issue at hand should be addressing his drug-seeking tendencies.

References

The Idea of Teaching as a Way to “Give Back”
Jason S. Ng, OD, PhD, FAAO | Optometric Education: Volume 45 Number 3 (Summer 2020)

As an interviewer of prospective students and prospective faculty members, I hear many mentions of “giving back” with regard to an optometric teaching career. Prospective students often cite giving back as the reason they want to eventually teach after a clinical practice career. Prospective faculty, particularly individuals transitioning from private practice or industry, often speak of how successful they’ve been and that they’re now ready to give back. It seems as if applicants expect such statements to result in a positive evaluation, but it isn’t clear to me why this should be the case.

The Way I See It

I suppose I can assume applicants who want to give back by teaching are acknowledging that teachers invest themselves in their students rather than in only themselves. They “pay it forward” and give back the time, knowledge and experience they “took from” their teachers who were giving back to them in the first place. However, in my opinion, teaching is an art, a science and a form of service. While optometric educators are constantly giving back — to our institutions, to our colleagues, and of course to the students whose lives we touch — teaching is more than that. For me, entering the teaching profession was more about making a difference than giving back. The passion and energy of my best teachers opened my mind to teaching as a profession. Like many, I’ve had great teachers and I’ve had terrible teachers. Having had both types, especially in my undergraduate studies, I experienced firsthand the difference a terrific teacher can make and this inspired me even more to want to make a difference as a teacher. I wanted to be one of the great ones because I didn’t want students to suffer through the opposite experience.

To me, working towards being the best teacher I can be is not easy. Rather, it’s an intimidating, difficult, life-devoting and never-ending process. Perhaps that’s why “I would like to give back now by starting a teaching career” sounds to me like an assumption that a great clinician and/or practice manager is automatically a great teacher. “See one, do one, teach one” comes to mind. Professionals who have done great things outside of teaching can probably “teach one.” But can they do so at an exceptional level? I don’t think the ability to give back in that way comes that easily to most, no matter how exceptional their abilities in clinical practice or elsewhere.

What I’d Rather Hear

Prospective faculty members who have never taught may be innocently ignorant of the rigors of teaching. Therefore, I don’t think they should rest their passion for embarking on a teaching career solely on a statement about giving back. I want them to say more about why they want to teach. I want them to say more about why teaching excites and inspires them. I want them to say more about how they want to be an exceptional teacher and not just a teacher. I want them to focus on how to best help students, both high and low performers. I want them to ask about teaching resources. I want them to ask about faculty development. I want to see a fire in them and come away with the impression they’ll be an active colleague in helping to shape the teaching persona of the college/university. I want them to focus on the students, the faculty, the institution and not necessarily only on their own feeling of giving back.

Maybe this is too much to expect from applicants who haven’t experienced the highs and lows of teaching or had time to develop a true passion for it. Maybe the thought of giving back is a sufficient place to start, but an open mind is crucial as well. I believe the most successful mindset is that teaching is a unique and challenging profession; success in one aspect of the field doesn’t guarantee success as a teacher; and it takes a lot of work to develop into a great teacher. Furthermore, we never really “make it” in teaching because it’s constantly evolving, all while we continue to fulfill our commitments of giving back to our students, peers, mentors, institutions and ourselves.
Call for Papers for Theme Edition: Diversity and Cultural Competence in Optometry
Optometric Education: Volume 45 Number 3 (Summer 2020)

The population continues to become more diverse, and optometry must be able to meet the cultural, ethnic, racial, gender and linguistic needs of patients. *Optometric Education* is inviting authors to submit scholarly papers addressing related themes such as diversity, cultural competency, gender issues and cultural awareness.

The deadline to submit papers for this theme edition is **March 30, 2021**

For more information, e-mail journal Associate Editor Keshia S. Elder, OD, MS, FAAO, or journal Editor Aurora Denial, OD, FAAO.
MEWDS: a Teaching Case Report
Crystal Lewandowski, OD, FAAO, and Daniel Bastian, OD, FAAO | Optometric Education: Volume 45 Number 3 (Summer 2020)

PDF of Article

Background

This case involves a 22-year-old Caucasian female diagnosed with multiple evanescent white dot syndrome (MEWDS). MEWDS is part of a group of inflammatory disorders known as white dot syndromes, which affect the retina, retinal pigment epithelium (RPE) and choroid. Often, symptoms of MEWDS are unilateral, have sudden onset, and include blurred vision, central or paracentral scotomas, enlarged blind spots, photopsia or dyschromatopsia. Observable ocular signs include multiple small, gray-white spots at the level of the RPE and outer retina. The most significant consequence of MEWDS is temporary vision changes, which usually spontaneously resolve. However visual field defects and photopsias may persist. This teaching case report highlights the role of the primary care optometrist in the evaluation and management of MEWDS. It focuses on the importance of critical-thinking skills for accurate diagnosis as well as patient education. It is appropriate for use with optometry residents and students who have had some patient care experience and knowledge in ocular disease. At most colleges, it would be appropriate for third- and fourth-year optometry students.

Student Discussion Guide

Case description

Visit 1

A 22-year-old Caucasian female was referred by her primary care physician to the vision department as a new patient for same-day evaluation. She complained of a gray spot in her vision in her left eye, associated with blur and loss of vision in part of her visual field. She denied new flashing lights, movement of the “spot” throughout her visual field, new floaters, new headache, eye pain and sun-gazing. At her last eye exam with a different doctor 10 months prior, the patient reported an unremarkable ocular history and that she didn’t wear glasses or contact lenses. Her medical history was remarkable for seasonal allergies, migraines with aura, irritable bowel syndrome and a recent history of flu a few days before onset of vision changes. She did not experience symptoms of nausea or vomiting at the time of her reported flu. Her medications included birth control pills as well as Dayquil and acetaminophen at the time of flu. There were no known medication allergies.

Uncorrected distance visual acuity (UCDVA) was 20/20 in each eye. Pupils, extraocular muscles and finger-counting confrontation fields were normal. Intraocular pressure was 16 mmHg in each eye. Undilated exam showed normal ocular health in each eye. The patient’s pupils were dilated with 1% tropicamide and 2.5% phenylephrine, and a 24-2 Sita Fast full-threshold automated Humphrey visual field (HVF) test was ordered for the patient to complete while dilating. After dilated retinal evaluation, optical coherence tomography (OCT) of the macula was ordered. Table 1 shows slit lamp findings and dilated fundus evaluation; Figure 1 shows HVF test results; and Figure 2 shows the OCT macular imaging.

The patient was diagnosed with central scotoma in the left eye secondary to MEWDS. She was shown her HVF and OCT test results and educated about MEWDS. The vision team explained that there are no treatment options for MEWDS and that symptoms should self-resolve without intervention. The patient was reassured that there were no signs of anterior or posterior segment inflammation, optic neuropathy or retinal breaks that would require treatment and that she’d be monitored closely for any changes. A two-week follow-up visit was recommended, and the patient was given an Amsler grid for monitoring her vision at home. She was offered the option of being.
evaluated by a retinal specialist at the two-week visit. She agreed and the referral was made.

Visit 2: one-day follow-up

Rather than wait for the two-week follow-up visit, the patient chose to be evaluated the next day by a retina specialist, who repeated dilated fundus evaluation with OCT and performed fluorescein angiography (FA). OCT findings were described as normal in the right eye, and ellipsoid zone disruption inferior to the fovea was observed in the left eye. On FA, there were no observable hyperfluorescent retinal lesions in either eye, confirming the absence of underlying vasculitis, leakage or optic nerve inflammation, ruling out several other potential white dot syndromes. Based on a granular macular appearance, OCT results and documented central scotoma, the ophthalmologist confirmed the diagnosis of MEWDS. The relevant patient education was delivered again: There are no proven treatments for MEWDS; it usually resolves on its own over several weeks; and, in the absence of underlying inflammation, follow-up in two weeks was an appropriate course of action.

Visit 3: two-week follow-up

The patient returned to the ophthalmology department for the two-week progress evaluation. Dilated examination showed stable ocular health and OCT findings. Continued monitoring with a follow-up examination in two months was recommended, but the patient was lost to follow-up.

Educator’s Guide

The Educator’s Guide includes the necessary information for teaching and discussing the case.

Key concepts

1. Hallmark symptoms and signs of MEWDS
2. Critical-thinking skills in diagnosis, using appropriate optometric tools when additional testing may guide to a diagnosis
3. Management and clinical course of MEWDS
4. The role of communication throughout the exam: developing patient rapport, obtaining a thorough case history, patient education

Learning objectives

Upon conclusion of this case discussion, participants should be able to:

1. Describe the ocular signs of MEWDS
2. Apply critical-thinking skills to correlate symptoms with clinical findings
3. Describe various white dot syndromes, noting similarities and differences to MEWDS
Describe additional testing that can be performed to confirm the diagnosis of MEWDS
Understand the need to investigate further if the clinical picture does not match the patient’s complaints

Discussion questions

A. Knowledge, concepts, facts, information required for critical review of the case:
1. Describe classic presentation of MEWDS and compare to other white dot syndromes
2. Determine differential diagnoses by analyzing case history, risk factors and demographics
3. Describe MEWDS etiology and patient demographics
4. Discuss how recent flu impacted this case

B. Differential diagnosis:
1. What tests were used in this case to diagnose MEWDS?
2. What are differential diagnoses based on symptoms alone?
3. How were the clinical findings analyzed to rule out or support potential differential diagnoses?
4. At this time, are there other diagnoses one should consider?

C. Patient management and the role of the optometrist:
1. What are appropriate management options in this case?
2. What is an appropriate follow-up schedule?
3. What is the prognosis for a patient with MEWDS?
4. What are patient education strategies to reassure patients when their condition is self-limiting and/or no treatment is indicated?

D. Critical-thinking concepts:
1. How does entering visual acuity impact the provider’s decision-making in this case? Would it be different if vision were worse than 20/20?
2. What inferences are made in the determination of the diagnosis?
3. If any one particular test performed during the visit was not completed, would the provider have been able to make the diagnosis?
4. What are the potential implications involving the management of this patient?
5. What if symptoms worsen or do not improve?
6. How would management have been different if an ophthalmologist had not been as easily accessible to the patient?
7. What are some effective strategies to reassure patients with self-resolving conditions?

Teaching instructions and assessment methodology

The purpose of this case report is to help clinicians review the clinical features and course of MEWDS as well as develop strategies for reassuring patients at the end of a clinical encounter. Optometry students can be guided through a discussion in a classroom or clinical setting. They should be presented with case details in a stepwise fashion (i.e., case history, dilated fundus examination, automated threshold visual fields, OCT) to think critically through the clinical presentation, devise differentials and arrive at a diagnosis. The key aspects of patient education can be discussed, including delivery of the diagnosis, management options and ocular prognosis.

The keys to diagnosis in this case were a thorough case history including systemic history and review of systems, a comprehensive dilated ocular evaluation and critical-thinking skills to incorporate and interpret additional test findings (HVF and OCT) as part of the visit. It is important that clinicians make an accurate MEWDS diagnosis and rule out other sight-threatening retinal pathology that would require treatment. MEWDS management is generally observation, and the condition has a good prognosis. In contrast, other white dot syndromes require invasive treatment and have worse prognoses.

Discussion

White dot syndromes

MEWDS was first reported in 1984, described as a transient chorioretinopathy often preceded by a viral-like illness. It is noted to be a clinical entity distinct from other inflammatory white dot syndromes such as acute macular neuroretinopathy, acute posterior multifocal placoid pigment epitheliopathy (APMPPE), acute retinal pigment epitheliitis, birdshot choroidopathy,
multifocal choroiditis (MFC) with panuveitis, and punctate inner choroidopathy. This group of inflammatory disorders produces discrete, yellow/white retinal lesions in young adults, which can be differentiated by history, appearance, laterality and FA (Table 2). MEWDS commonly presents with macular granularity and “wreath-like” hyperfluorescence on FA. MEWDS generally affects healthy, myopic women (4:1 compared to males) in their third or fourth decade, but patients in their 50s have been reported. The annual incidence is 0.22 per 100,000. There are no known genetic or racial predilections. A viral prodrome has been reported in more than 50% of cases.

Etiology

The exact pathogenesis of MEWDS is unknown, but it is thought to be caused by an underlying autoimmune (non-infectious) mechanism. Suggested explanations include viral agents entering retinal receptor cells at the edge of the optic nerve and ora serrata, triggering an autoimmune response. An association with environmental triggers and human leukocyte antigen (HLA)-B51 has been reported. Cases have also presented after vaccinations for human papillomavirus, hepatitis A, and hepatitis B. One theory holds that choriocapillaris hypoperfusion or non-perfusion results in ischemic damage to the outer retina and RPE due to a vaso-occlusive process involving small vessels, whereas other white dot syndromes such as APMPPE and MFC involve larger blood vessels. Few reports provide an explanation for differences between retinal spots and dots. A recent report suggests a shallow infiltration of inflammatory cells within the inner choroid or outside the choriocapillaris, beneath the RPE, is what causes smaller retinal dots in MEWDS.

Clinical features

Patients with MEWDS may complain of sudden, unilateral changes in vision including decreased acuity, visual field defects consistent with enlarged blind spots or scotomas, photopsias or color vision changes. Visual acuity may vary, ranging from 20/20 to 20/400 in some cases.

Clinical manifestations include small, gray-white retinal dots and spots ranging in size from 100-200 µm. Some reports have described differences between the retinal dots and spots based on their location; dots appear more anteriorly at the level of the outer retina and RPE, and spots appear deeper at the RPE and inner choroid. The macular region may also have a fine, granular appearance, which has been reported to be pathognomonic for MEWDS. A relative afferent pupilary defect (RAPD) may be present, as well as mild optic nerve inflammation, mild vitreous cells and mild anterior chamber reaction.

Retinal lesions are usually transient and tend to resolve within 3-10 weeks, with larger spots resolving quicker than smaller dots. After the acute clinical phase and resolution of retinal lesions, minimally evident macular and paramacular granularity may persist.

Differential diagnosis

Clinicians must be diligent in ruling out potentially ominous causes of a patient’s perceived visual field defect, whether it be of ocular or neurological origin. Upon chief complaint of “gray spot” associated with blur and loss of vision, a broad range of differentials could be considered including ocular inflammation, optic neuropathy, macular disease and retinal detachment. A condition commonly encountered in clinic in young, healthy patients is central serous retinopathy (CSR). However, this patient did not fit the classic demographics of being a myopic male under recent stress or with Type A personality. Other retinal conditions that may cause a patient to present with a scotoma include MEWDS and central pigment epithelial detachment.
Given this patient’s history of migraine with aura, it is reasonable to consider an atypical ocular migraine as the cause of the symptoms, but this should be a diagnosis of exclusion, which was certainly ruled out with clinical exam. Classic symptoms of ocular migraine include flashing or shimmering lights, blind spots, floating lines and zig-zag patterns, and symptoms may occur without headache. When questioned, the patient denied headache but reported recently having the flu, which was confirmed by her primary care physician and managed with over-the-counter medications.

MEWDS is diagnosed by clinical observation and can be confirmed with electrophysical and angiographic testing (Table 3). Careful case history, clinical appearance and ancillary testing help to differentiate MEWDS from other white dot syndromes. Testing may include:

- **OCT:** OCT may reveal disruption to the hyper-reflective band attributed to the photoreceptor inner segment/outer segment junction or dome-shaped subretinal hyper-reflective lesions. Increased reflectivity of the choroidal space has also been reported. OCT findings gradually resolve with resolution of the disease; however, outer nuclear layer thinning has been apparent in cases of recurrent disease, suggesting that repeated episodes may result in permanent retinal atrophy.

- **FA:** FA usually demonstrates early punctate hyperfluorescence corresponding to retinal dots and spots, with late staining of the lesions and optic nerve. Dots appear during the choroidal-filling and retinal artery perfusion phases, often in multiple clusters in a “wreath-like” pattern near the macula. Depending on duration of symptoms at presentation, variances in equipment and imaging modality, and severity of the condition, FA appearance may also include hypofluorescent lesions or even appear normal. Resolution of FA findings correlate to resolution of retinal lesions.

- **Indocyanine green angiography (ICGA):** Hypofluorescence is evident in early and late phases. Multiple hyperfluorescent dots may appear in the late phase 20 minutes after injection, characteristically more numerous and less “wreath-like” than noted on clinical examination and FA.

- **Fundus autofluorescence (FAF):** In the acute phase, retinal dots and spots show up as increased areas of autofluorescence, and pinpoint areas of decreased autofluorescence around the disc and macula may be seen. As MEWDS resolves, hyperfluorescent lesions disappear and hyperfluorescent spots may lessen in number, resolve, become smaller in size, or have a more hypofluorescent appearance (either a halo or throughout the lesion). FAF abnormalities may persist for months after diagnosis and after resolution of FA-ICGA abnormalities.

- **Electrophysiologic testing:** electro-oculogram (EOG) demonstrates reduced light-dark ratios, and electroretinogram (ERG) shows reduced a-waves and early receptor potentials.

- **Visual field testing:** Automated visual field testing may reveal an enlarged blind spot or scotomas (central, paracentral, temporal or scattered). Although not diagnostic, automated visual field testing may be useful in ruling out possible neurological causes for visual field reductions and ruling in retinal disease (especially with central 10-2 testing). Documentation of the visual field defect may be useful for patient education as well as monitoring disease course and symptoms.

- **Fundus photography:** While also not diagnostic, fundus photography may be useful in documenting the current status of the retina, which may change based on duration of disease. In early stages, photodocumentation of yellow-white retinal spots in the posterior pole and midperiphery is common. Lesions appear at the level of the outer retina, RPE and inner choroid. Foveal granularity described as small white or orange pinpoint specks may be pathognomonic for MEWDS. As the clinical course progresses, retinal dots and spots may fade or appear more reddish/brown before resolving completely.

- **There have been reports of MEWDS cases without dots or spots on clinical examination that were identified with both FA and ICGA as well as ICGA alone. There are few reports of cases without retinal spots and macular granularity alone, but this may represent differing timing in the clinical course. This case is one example of a MEWDS presentation with macular granularity as the only clinical sign on dilated examination, consistent with changes on OCT and HVF testing but not identified on FA.**

### Management

The prognosis for MEWDS is generally good with spontaneous resolution of retinal lesions within weeks to months (3-10 weeks). Symptoms generally improve shortly after; however, OCT, angiographic and visual field abnormalities may take longer to resolve. In some cases, visual field defects and photopsias may persist. Recurrence is rare, but is estimated to occur in approximately 10% of cases. An Amsler grid can be given to patients to monitor their vision at home. Treatment is not usually necessary; however, recurrent cases have been treated with cyclosporine, and rare incidents of choroidal...
neovascularization have been treated with intravitreal ranibizumab.\textsuperscript{4}

**Critical-thinking concepts**

Because the degree of visual acuity loss may vary in cases of MEWDS, ranging from 20/20 to 20/400, it should not be used as an indicator of disease. In this case, the patient’s UCDVA was 20/20 in each eye and thus did not lead to high suspicion of macular disease at the start of the examination. For this reason, color vision and Amsler grid testing were not performed at the initial visit. However, because color vision changes may be a presenting symptom, it may be helpful to perform color vision testing to document any defects as well as resolution of symptoms throughout the clinical course of MEWDS. Anterior segment slit lamp examination revealed no signs of corneal disease or anterior uveitis. Dilated fundus evaluation showed flat optic nerve heads, well-perfused with distinct margins in each eye. Thus, optic neuritis, optic atrophy and optic neuropathy were ruled out. Additionally, there were no signs of vitritis, serous detachments, pigment epithelial detachments or retinal breaks, thus CSR and retinal detachment were ruled out. The only remarkable retinal finding was a fine macular granular change outside the fovea of the left eye; there were no classic white retinal spots or dots. It is possible that retinal spots and lesions were present prior to the patient arriving for evaluation and had already self-resolved, revealing only granularity correlating to abnormal HVF and OCT test results.

This patient fits the classic profile of patients diagnosed with MEWDS. However, without a thorough case history revealing recent-onset flu and without a stepwise approach to investigating symptoms with careful dilated examination and the addition of HVF and OCT testing, the diagnosis may not have been made. In this case, the clinical picture did not include classic white spots/dots. Clinical pearls for students are to have a high level of suspicion when examination does not reveal obvious pathology and to order additional testing when clinical findings do not fully correlate with patients’ symptoms. In this case, HVF testing was performed to rule out any neurological etiology for the patient’s visual field complaints as undilated examination revealed no obvious pathology. As it turned out, the central visual field defect suggested macular pathology, and macular OCT was ordered while the patient’s pupils were dilating. Photoreceptor integrity line abnormalities on OCT correlated with fundus examination, HVF test findings and the patient’s complaint.

**Table 4.** A Six-Step Strategy for Patient Reassurance\textsuperscript{15}

| Collect Data |
|-----------------|-----------------|
| - Empathize and show concern by establishing history, question, listen, clarify. |
| - Develop rapport; collect information and examine the patient. |

| Give Accurate Information |
|--------------------------|--------------------------|
| - Inform the patient that a serious illness is not present, and condition may be temporary. |
| - Correct any misattribution of illness (“I feel bad”) by explaining to the patient the conditions that could be expected. |
| - Sometimes labeling the patient with a diagnosis may be helpful as a point of reference to the patient to better understand the condition. However, if this label creates more worry and anxiety regarding symptoms, using nontechnical descriptions or terminology may be preferred. |

| Suggest a Timeline |
|-------------------|-------------------|
| - Give patient probable timeline for resolution of symptoms and condition. |
| - Return to normal activities. |

| Consider Non-Specific Treatment (if appropriate) |
|-----------------|-----------------|
| - In some situations when organic disease is not present, recommending short-term medications and lifestyle changes such as altering diet and exercising may have a placebo effect that helps improve the patient’s symptoms. |

| Follow-Up with Patient |
|------------------------|------------------------|
| - Scheduling follow-ups with patients may give them a sense of security knowing that the provider has acknowledged and has concern about their complaint. |
| - Scheduling a follow-up decreases the likelihood of patient seeking care from other providers, and ultimately increasing healthcare costs. |

Although fundus photography may be useful in documenting the clinical course of MEWDS, and can be used as a visual aid during patient education, it was not performed at the initial visit due to minimal retinal changes observed on clinical examination as well as scheduling and billing/insurance constraints. Other tests that could have been ordered were FAF, FA, ICGA, and ERG/EOG; however, they were not available at the practice. The patient was referred to a nearby ophthalmology practice where OCT was repeated and FA was performed, ruling out underlying vasculitis or inflammation. Although FA often shows “wreath-like” hyperfluorescent dots corresponding to retinal dots observable in the acute phase of MEWDS, this patient did not present with retinal dots and thus there was no hyperfluorescence on FA. It is possible that retinal spots had already appeared and resolved in the 10 days prior to FA being performed. However, there have also been cases of MEWDS that present without retinal spots; therefore, FA should not be solely relied upon for making a MEWDS diagnosis.

Often, when health conditions are self-limiting or without clear cause, clinicians may attempt to reassure patients to decrease their concern or worry and improve outcomes.\textsuperscript{13} Clinicians may use several strategies including developing rapport with the patient, empathizing with him/her and providing accurate information and explanations about the condition.\textsuperscript{14} A six-step strategy for patient reassurance has been described (Table 4).\textsuperscript{15}

Though there are many theoretical models of patient reassurance, the effectiveness of each approach has not been extensively studied and there are no standardized methods of delivering reassurance.\textsuperscript{15} Preliminary clinical trial evidence does suggest that a more cognitive approach (information, education) may be more effective than an affective approach (rapport building, empathy) in the long run. However, a mixture is commonly employed.\textsuperscript{13} Different patients may require different strategies for reassurance, thus a patient-centered approach should be considered in each case.\textsuperscript{13-15}
Conclusions

This case serves as a reminder that young, seemingly healthy patients without chronic systemic disease can have less commonly encountered retinal pathology. This patient experienced acute-onset viral symptoms followed by changes in vision caused by MEWDS. This case report is intended to educate eyecare providers on the clinical course and management of MEWDS, highlighting the importance of thorough case history, careful ophthalmic examination, and critical thinking in analyzing information throughout the examination. While MEWDS is uncommon and generally has a good prognosis, it is important that clinicians are able to accurately diagnose and educate patients on its course.

References

When a Red Eye is More than Meets the Eye: a Teaching Case Report on the Public Health Role of the Eyecare Provider
Julia Appel, OD, FAAO | Optometric Education: Volume 45 Number 3 (Summer 2020)

PDF of Article

Background

Patients presenting for care of "red eye" can be challenging due to the numerous etiologies and potential systemic associations. A systematic evaluation of presenting signs and symptoms with development of an inclusive differential diagnosis is required, but is complicated when unknown concurrent systemic infection is present. Collaboration with the patient's primary care physician (PCP) and a working knowledge of the epidemiologic contributing factors provide a deeper understanding in this process, and add certainty to diagnostic expertise when more rare conditions present. The following clinical case endeavors to teach optometry students two critical aspects of their role as clinicians: the importance of basic knowledge about significant but relatively rare differential diagnoses of a common complaint, and that optimal care and improved outcomes result with application of epidemiologic principles. True proficiency in providing optimal care encompasses more than the understanding of diagnosis and management, and the earlier epidemiological principles are introduced, the better.

This teaching case report is for use with latter second-year optometry students (and beyond) who understand the differential diagnosis and treatment options for conjunctivitis/uveitis and have a basic understanding of epidemiological concepts.

Case Description

A 38-year-old Caucasian male presented to the eye clinic with a chief complaint of an irritated right eye of two-day duration. The patient complained of burning, tearing, mild photosensitivity and mild foreign body sensation of the right eye. There was no complaint of significant blur, pain, flashes, floaters, diplopia, headache, dryness, recent upper respiratory infection or allergy. The patient had seen his PCP one day prior when symptoms began. The PCP ruled out a corneal abrasion, recommended artificial tears, and referred the patient to the eye clinic for further evaluation if symptoms persisted or worsened.

Initial visit

Mild facial swelling near the jaw observed at the eye clinic visit was due to overnight teeth grinding according to the patient. All other systemic and medical history was negative. Best-corrected visual acuity (BCVA) was 20/25-OD and 20/20 OS. Pupillary reactions and extraocular motilities were normal.

Slit lamp biomicroscopy of the patient’s left eye was unremarkable. Slit lamp findings for the right eye (Figure 1) were:

- Mild diffuse hyperemia with limbal flush
- Trace anterior chamber reaction (cells and flare) with fine keratic precipitates diffusely
- Moderate, diffuse corneal edema with endothelial folds
- Mild central punctate staining
- Goldmann applanation tonometry: 24 mmHg OD, 14 mmHg OS at 11:50 a.m.
- Palpation of preauricular lymph nodes: no discomfort and no significant elevation on the right side relative to the left
- Gonioscopy was inconclusive OD because of poor views due to edema; angles were open 360° OS
- Pachymetry not performed at this visit

Figure 1. Slit lamp image captured at the patient’s initial visit. Click to enlarge
Fundus findings were unremarkable OD, OS and non-contributory

A working diagnosis of anterior uveitis with corneal edema and increased intraocular pressure (IOP) OD without known etiology was determined. Table 1 lists initial treatment. Initiation of intraocular pressure lowering drops would commence if intraocular pressure remained elevated at follow up visit.

One-day follow-up visit

The next day the patient reported complete adherence to the prescribed topical medication regimen, improved ocular symptoms, and mild worsening with dry mouth and testicular pain. He reported a generalized feeling of being unwell and was to return to his PCP that afternoon. See Table 1 for treatment changes. Additional findings:

- Continued mild injection OD, improving
- Moderate/severe corneal edema; pachymetry: 760 µm OD, 569 µm OS
- Improving but still elevated IOP (18 mmHg OD, 14 mmHg OS)

Further investigation of the new systemic symptoms, patient physical and ocular presentation revealed a likely diagnosis of mumps-related endotheliitis with associated trabeculitis. The provisional diagnosis of endotheliitis associated with epidemic parotids was discussed with the PCP by phone. The physician’s evaluation of the patient and subsequent immunoglobulin M titers confirmed the diagnosis. Despite vaccination as a child, the patient contracted mumps from an unknown source, which raises the questions "How can this be and how does it influence our future evaluation of vaccinated individuals?"

Table 1

<table>
<thead>
<tr>
<th>TABLE 1 Summary of Patient Visits</th>
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<tbody>
<tr>
<td></td>
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<tr>
<td>Dk corrected visual acuity</td>
</tr>
<tr>
<td>Anterior segment (all findings are OD)</td>
</tr>
<tr>
<td>Incapable of distant vision</td>
</tr>
<tr>
<td>Moderate/severe injection</td>
</tr>
</tbody>
</table>
| Further investigation of the new systemic symptoms, patient physical and ocular presentation revealed a likely diagnosis of mumps-related endotheliitis with associated trabeculitis. The provisional diagnosis of endotheliitis associated with epidemic parotids was discussed with the PCP by phone. The physician’s evaluation of the patient and subsequent immunoglobulin M titers confirmed the diagnosis. Despite vaccination as a child, the patient contracted mumps from an unknown source, which raises the questions “How can this be and how does it influence our future evaluation of vaccinated individuals?”

Table 1. Click to enlarge

Education Guidelines

Key concepts

1. Differential diagnosis of endotheliitis systemic etiologies
2. Application of epidemiologic principles and collaborative care practice in providing optimal health care
3. Vaccination against preventable infectious disease

Learning objectives (LO)

Utilization of this case to educate separately on ocular/systemic disease or epidemiological emphasis is possible, but use of this report in its entirety will enhance the student’s ability to:

1. Participate collaboratively in the diagnosis of epidemic parotids with an understanding of the ophthalmic and systemic ramifications
2. Independently diagnose and treat the secondary ocular manifestation of endotheliitis with an understanding of the underlying pathophysiology
3. Utilize and apply the epidemiological concepts determining risk of infection with this and other preventable viral etiologies
4. Gain working knowledge and use of epidemiological resources to aid in the care of patients presenting with infectious disease
**Education activities/case discussion**

Presenting the case using a grand rounds approach can help students hone their skills as they answer questions in determining additional testing and next steps for care in the evaluation and management of problem-focused exams.

- **Poll Everywhere** or **Turning Point** allow for real-time assessment of student participation, or a quiz preceding and after review and discussion of the case can assess understanding of relevant clinical care concepts. The quiz, geared toward the level of learner, can emphasize both epidemiology and ocular/systemic manifestations utilizing content provided in the discussion. (LO 1-4)

Deep learning/critical thinking opportunities exist as students break into smaller groups to independently explore and present back to the larger group or post on the course online learning management system. The discussion section and resources listed provide a starting point for these active learning exercises:

- Develop a problem representation grid (illness script) of the differential diagnosis of conjunctivitis including endotheliitis. Independent development and completion by the students or a grid provided with some data points missing are options. (LO 1, 2)
- Discuss the role of anterior chamber-associated immune deviation (ACAID) in this condition and its effect on management of ocular inflammation. (LO 2)
- Develop a basic protocol for patient medical and visual conditions that require interprofessional collaboration (include examples of which conditions need outreach and why, modes of contacting the PCP and what information should be provided. Knowledge of clinical medicine concepts is helpful or working with the instructor as facilitator are options. (LO 1)
- Access and use the Morbidity and Mortality Weekly Report (MMWR) database to research the prevalence of a viral illness in the United States and its ophthalmic manifestations (herpes zoster, Zika, chikungunya, West Nile, yellow fever, Rift Valley fever, etc.) (LO 3, 4)
- Investigate the autism vaccination controversy and debate it using two peer-reviewed resources on either side. (LO 3, 4)
- Discuss the concepts of herd (community) immunity and long-term seroprevalence/vaccination efficacy for measles, mumps and rubella given recent national outbreaks of measles and mumps. (LO 1, 3, 4)

Assessment of the above learning activities depend on the course type and facilitator goals. While rigorous grading rubrics for the activities for summative evaluations are possible, these activities lend themselves to seminar and adjunctive content and are better assessed in a formative manner for clarity and content. Access to all group activity reports or best examples of each activity posted on the learning management system for student review will maximize learning opportunities to meet all learning objectives.

**Literature review**

**Mumps (epidemic parotidis)**

Epidemic parotidis is a contagious acute paramyxovirus infection of the salivary glands presenting as swelling of one or both parotid glands and flu-like symptoms. Hence, this patient’s swelling near the jaw was not associated with teeth grinding, and his dry mouth on day two was not due to use of a cycloplegic agent but rather the parotid gland dysfunction. The incubation period of epidemic parotidis is 12-25 days and glandular involvement occurs 16-18 days after exposure. While the parotidis and viral illness are self-limiting and usually resolve within 10 days, significant systemic complications, including pelvic inflammation in males (orchitis) and females (oophoritis), can occur, especially after puberty. The patient’s complaint of testicular pain on day two was associated with orchitis, but this presentation rarely results in sterility. Neurosensory hearing loss can occur but usually returns to normal post-infection.

Before the introduction of single-dose vaccination with live attenuated virus in 1967, mumps accounted for a significant percent of cases of aseptic meningitis in the United States. Other serious systemic complications such as pancreatitis were also reported, although death from infection was exceedingly rare. Once the two-dose vaccination regimen came into common use, significant drops were seen in all systemic complications and no deaths were reported.

**Endotheliitis typical presentation**

The most common ophthalmic manifestation of epidemic parotidis is endotheliitis (sometimes associated with trabeculitis as in this presentation). The condition typically presents as:

- mild to moderate anterior chamber reaction
- unilateral involvement
fine keratic precipitates that can be confused with endothelial pigment
- sectoral or diffuse corneal edema with endothelial folds
- microcystic edema resulting from aggressive stromal edema
- elevated IOP (if trabeculitis is also present)

Pathology

Corneal endotheliitis is a clinical diagnosis based on specific characteristics (Table 2). The pathology of endotheliitis is endothelial viral proliferation resulting in an inflammatory response. There is subsequent failure of the endothelial cell pump leading to edema and loss of transparency that can lead to long-term endothelial damage. Subsequent inflammation of the trabecular meshwork is possible and leads to reduction in aqueous outflow and increased IOP.

It was not until 1985 that researchers identified the viral etiology of this presentation by isolating herpes simplex virus (HSV) in the aqueous humor and detecting HSV-1 antigen in the anterior chamber of affected eyes. Some evidence indicates that HSV originates from the trabeculum as patients with endotheliitis have shown HSV on excised trabecular tissue and this is consistent with the peripheral presentation of HSV-related endotheliitis. Later, identification of other contributing viral etiologies, including the rarely encountered epidemic parotiditis-related endotheliitis, followed.

The role of ACAID results in a relatively reduced inflammatory response despite the direct infection of ocular tissue due to the suppression of a delayed hypersensitivity reaction. However, the reduced cell-mediated immunity to viral particles shed into the anterior chamber allows for viral proliferation in the corneal endothelium leading to the presentation of endotheliitis. The pathogenesis of mumps-related endotheliitis varies from this and may be associated with the level of viremia, but the rarity of the condition makes further research difficult.

Endotheliitis is not completely benign as endothelial cell loss can occur. As this cell layer does not regenerate, future pathology or necessary procedures such as cataract extraction may result in further corneal decompensation and vision loss. Systemic treatment of recalcitrant HSV-related endotheliitis may be necessary (oral acyclovir or valacyclovir) along with topical corticosteroids. However, cytomegalovirus (CMV)-related endotheliitis often does not respond to topical corticosteroids and requires aggressive systemic management (IV ganciclovir) to protect vision. Consultation with the patient’s PCP when confronted with suspicions of systemic herpetic disease is highly recommended for best outcomes. In addition to topical corticosteroids, topical ganciclovir ophthalmic gel 0.15% five times daily supplements treatment of disciform and linear endotheliitis associated with HSV and varicella zoster virus but may show limited effectiveness for CMV etiology.

The Centers for Disease Control and Prevention (CDC) is a federal agency under the U.S. Department of Health and Human Services tasked with monitoring and prevention of disease. The agency’s mandate is to protect the nation’s health, and as part of its mission it “conducts critical science and provides health information that protects our nation against expensive and dangerous health threats, and responds when these arise.” Promoting vaccination against preventable infectious disease is a part of this mandate. Current CDC recommendations for infectious childhood diseases include vaccination against varicella, rubella, measles, mumps, polio, and diphtheria. The agency cites studies showing the safety of these recommendations as well as their impact on morbidity and mortality. Patients and their parents may ask healthcare providers for their opinions on vaccination, making a working knowledge of the subject important. Resources for practitioners and the public are available on the CDC website.

Vaccination is the most effective way to prevent the spread of epidemic parotiditis. The measles, mumps rubella (MMR) or measles, mumps, rubella, varicella (MMRV) vaccine protocols call for two doses. The first is administered at age 12-15 months; a booster is administered at age 4-6 years.

Other resources such as the CDC publication of the MMWR can provide current, evidence-based public health information as well as recommendations and their impact on public health. The MMWR provides real-time information on outbreaks occurring nationally.

Those most at risk for mumps

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**Table 2. Click to enlarge**

<table>
<thead>
<tr>
<th>Corneal endotheliitis presentation</th>
<th>Systemic etiology</th>
<th>Presentation</th>
<th>Treatment</th>
</tr>
</thead>
<tbody>
<tr>
<td>Linear</td>
<td>HSV, CMV</td>
<td>unilateral, peripheral and sectoral edema with fine keratic precipitates at sector edge (coin shaped lesions-CMV), minimal anterior chamber reaction, progressive loss of endothelial cells</td>
<td>aggressive treatment with topical and oral antiviral agents (based on specific viral etiology) and corticosteroids</td>
</tr>
<tr>
<td>Diffuse</td>
<td>epidemic parotidis</td>
<td>rare, diffuse corneal edema, fine keratic precipitates, spontaneous resolution occurs within 2 weeks, endothelial cell loss not common</td>
<td>topical corticosteroids if mumps-related with trabeculitis present, antiviral therapy considered if mumps unlikely</td>
</tr>
<tr>
<td>Disciform</td>
<td>HSV, VZV</td>
<td>most common, focal round edema, multiple keratic precipitates, melt to motte-like anterior chamber reaction, endothelial cell loss not common</td>
<td>most likely to respond to topical corticosteroids, association with latent virus from prior infectious parotiditis</td>
</tr>
</tbody>
</table>

HSV = herpes simplex virus
CMV = cytomegalovirus
VZV = varicella zoster virus
Individuals most at risk for contracting epidemic parotidis include:

- those who have not been vaccinated, including infants too young to be vaccinated
- healthcare workers in clinic/hospital settings
- those living in close quarters, such as college students or members of traditional communities
- persons traveling overseas to countries that do not require mumps vaccination
- vaccinated members of a certain U.S. population subset

Outbreaks of mumps have occurred in adult populations and in communities of all ages in the recent past. If healthcare providers are not informed, they may miss important diagnoses and patient education opportunities. The public health impact of optometrists’ contributions to care is significant as a solid understanding of epidemiologic principles is critical for evidence-based practice and can inform or guide care protocols and referrals for best patient outcomes. Treatment of a presenting condition such as endotheliitis is only the beginning. A deeper understanding of those at risk for underlying, causative systemic disease will provide for more timely identification and diagnosis with the potential to reduce the spread of infection.

**MMR vaccine efficacy**

The MMR vaccine is highly successful in providing long-term protection against measles and rubella but is not as effective in maintaining immunity to mumps over time. Mumps seropositivity shows less persistence over time with one dose resulting in 78% immunity (49-92%) and two doses resulting in 88% immunity (66-95%). While herd or community immunity to prevent spread of disease is known to be 92%, the low persistence of mumps seropositivity does not result in pervasive epidemic rates of infection. One study of a small community showed only 74% of residents with measurable immunity 15 years after a second dose of MMR vaccine, yet no members of the community developed epidemic parotidis. This is due to limited rates of exposure to the disease. Therefore, high vaccination rates paired with lack of exposure result in eradication of acute infection, but to be truly effective in this regard, both are required.

**Why did this patient get mumps?**

A survey of epidemiological resources is helpful in learning why the patient in this case contracted mumps. CDC evaluators used data from the 1999-2004 National Health and Nutrition Examination Survey (NHANES) to assess mumps antibody seroprevalence in the U.S. population. They found 90% of those surveyed were immune (borderline to prevent disease spread). However, some subsets showed lower rates of immunity. Specifically, those born between 1967 and 1976 showed only 85.7% seropositivity. These single-dose recipients were too old to have been exposed naturally as children and too young to have received the more persistent two-dose regimen. Therefore, they were more likely to be at risk of developing infection when exposed. Based on his age, this patient fell into that demographic.

Unfortunately, a reduction in vaccination rates has resulted in outbreaks in the United States in recent years. In 1998, a paper by Wakefield et al. raised concerns of autism risk in those receiving the MMR vaccine. This led to parental reluctance to immunize children against multiple infectious diseases. Ultimately, significant decreases in vaccination rates resulted in some communities. Although this theory has been debunked through retrospective and prospective research, and the original paper was found to be fraudulent and retracted, the safety concern continues to affect U.S. vaccination rates. Literature review shows that parents opting out of vaccinating their children not only puts the unvaccinated individual at risk for preventable disease but also increases the likelihood of spreading disease throughout the greater community.

**Conclusion**

A seemingly simple complaint of red eye can signal a significant threat to a patient’s health and well-being. The ability to recognize important features that indicate potentially harmful systemic etiology, such as rash, swollen glands, associated symptoms or unique ocular findings, makes optometrists effective contributors to the overall care patients receive. By collaborating with PCPs, optometrists can implement timely and appropriate ocular and systemic treatment. Recognizing a rare presentation such as epidemic parotidis-related endotheliitis is important and may become increasingly so as mumps outbreaks become more prevalent due to a reduction in vaccination rates. Understanding the relevant epidemiologic principles and being familiar with available resources enable a quick response when new infectious diseases are present in local communities and abroad.

**References**

We’ve Had Our Challenges, and Creative Solutions, During the Pandemic

Aurora Denial, OD, FAAO, DipOE | Optometric Education: Volume 45 Number 3 (Summer 2020)

While I was visiting my 5-year-old granddaughter recently, she proclaimed “I am mad at the coronavirus!” I couldn’t help but feel the same. Our lives had changed on a dime in response to the virus. Our visit was in her backyard. We were six feet away, and I was unable to hug her or play. If she or my grandson came closer, immediately a mask went on. I had not held their new baby sister for months. Much had changed professionally as well. I was teaching my course remotely and attending at least two to three Zoom-In meetings per day.

Coronaviruses are a group of RNA viruses that can cause a range of respiratory illness in humans, from colds to potentially lethal illnesses such as severe acute respiratory syndrome (SARS), Middle East Respiratory Syndrome (MERS) and the novel coronavirus disease 2019 (COVID-19). Coronaviruses are encased in a lipid “envelope” and have halo-like protrusions of protein. The word corona is Latin for halo, hence the name.

In December 2019, a pneumonia of unknown cause presented in Wuhan, China. In January 2020, the previously unknown COVID-19 was identified as the cause. The World Health Organization (WHO) classified the pneumonia outbreak as a pandemic on March 11. As of mid-June, 8,400,320 cases had been confirmed worldwide and 2,173,804 cases had been confirmed in the United States.

Perspective-changing Experiences

Although I had heard about the coronavirus in January, February and early March, I was not overly concerned nor could I have anticipated how everyday life would change. My institution, the New England College of Optometry (NECO), provided faculty with regular updates on potential plans for dealing with the outbreak.

My perspective changed drastically after two experiences. The first involved my youngest daughter, who works for a disaster relief organization, All Hands and Hearts. This organization rebuilds communities impacted by natural disasters. This is a hardy group of people, who go into the most challenging circumstances. My daughter was working on a project building schools in Sofala Province, Mozambique, which had been devastated by a cyclone in March 2019. Within 48 hours of the WHO announcement, All Hands and Hearts ceased all projects and made arrangements for employees to return to their home countries. My daughter’s expedited return to the United States occurred despite the facts that she did not have her passport on base (it was at a government office) and two people on base came down with a pneumonia-like illness (later determined not to be COVID-related). The second experience arose when I decided to stock her refrigerator and cupboards in preparation for her arrival at home after five months away. I went to the local supermarket only to find completely empty shelves. Of course, there was no toilet paper, hand sanitizer or cleaning supplies, but pasta, flour and rice were also sold out, and the supplies of meats and produce were low. Seeing a large grocery store with so many empty shelves was chilling.

I now realized the enormity of the circumstances. Going forward, we would have new norms: social distancing, PPE, frequent hand-washing, etc. The news would be filled with new cases, death toll, lack of PPE, and the sacrifices of first-line workers. Optometric education, along with all education, would also be greatly altered.

Let’s Share What We are Learning

NECO responded by moving all didactic classes online, cancelling or postponing labs, and altering clinical assignments. The college building was closed, and the clinics were open only for emergencies. All employees whose workloads were transferable to home began working remotely. I had never worked from home previously. Despite a long commute, I enjoyed coming into Boston daily and interacting with my colleagues. To my surprise, the transition to online teaching went smoothly. Technical support was readily available and course material was altered for online presentation. Initially, I was concerned about student engagement in my small group discussion courses. I was pleased to discover that students appeared even more engaged than they are in person. This may be at least partly attributed to my aggressively calling on all students to contribute. I try to
engage all students in person, but in every class there are shyer students who yield to more assertive and vocal peers. Lesson learned for me: aggressively utilize the roster for active participation by everyone. To facilitate clinical education, the college rolled out Electronic Learning Alternatives for Time not in Clinic (ELASTC).

All optometric institutions implemented similar alternative programs for teaching. The response of educators demonstrates flexibility, dedication and creativity. As much as I am still “mad at the coronavirus,” it has provided freedom to try new teaching methods that otherwise may not have been utilized. In my opinion, student learning continued to be productive and sound. Going forward if possible, it may be sensible to evaluate qualitative and quantitative data on learning during the COVID-19 crisis. For now, it would be great if all faculty share how they responded to the COVID-19 crisis, lessons learned, challenges and creative solutions, new programs, etc. We will consider publishing creative and unique approaches, in articles of 500-800 words, in the Fall (November) edition of the journal.

References

Progressive Supranuclear Palsy: a Teaching Case Report
Julie Chao, OD, and Andrea Yiasemis, OD, FAAO | Optometric Education: Volume 45 Number 3 (Summer 2020)

PDF of Article

Background

Progressive supranuclear palsy (PSP) is a neurodegenerative disorder with distinct clinical features including vertical supranuclear gaze palsy, frontal lobe cognitive decline, postural instability and progressive axial rigidity. First described in 1964 by Steele et al., PSP had been referred to historically as Steele-Richardson-Olszewski syndrome. They reported nine cases with the aforementioned findings, which veered from the typical presentation of idiopathic Parkinson’s disease.

Since its initial characterization, PSP has been further categorized into different phenotypes with notable overlap with other neurodegenerative disorders. Early in the onset of the disease, patients with PSP are often misdiagnosed as having idiopathic Parkinson’s disease. Given that the average life expectancy following diagnosis is 5-10 years, it is imperative for practitioners to identify and manage the disease appropriately.

The three cases presented here highlight common ocular features of PSP and considerations for ophthalmologic management. The intended audience is third- and fourth-year optometry students, optometry residents and practicing optometrists.

Case Descriptions

Case 1

A 78-year-old man residing at a community living center presented to the eye clinic reporting that his vision in both eyes seemed to have slowly worsened over time. The patient’s ocular history included toxic, nutritional optic neuropathy OD>OS secondary to long-term cocaine and alcohol abuse. His medical history included type 2 diabetes mellitus, osteopenia, liver steatosis, malignant prostatic tumor, hypertension, hyperlipidemia, spastic paraparesis, spinal stenosis and post-traumatic stress disorder. Medications included acetaminophen 650 mg, bicalutamide 50 mg, cholecalciferol, gabapentin 900 mg, simvastatin 40 mg, tamsulosin 0.4 mg and venlafaxine 150 mg.

Best-corrected visual acuities (BCVAs) were hand-motion OD and 20/50-1 OS. An afferent pupillary defect was noted OD. Evaluation of the patient’s extraocular muscle motility exhibited a complete restriction of inferior gaze and a moderate restriction of vertical gaze. Of particular note, the vestibulo-ocular reflex (VOR), or “doll’s head” reflex, was intact. The patient’s low phonation and mild dysphagia made it difficult to acquire a proper history to ascertain whether he had been symptomatic for his restriction of gaze.

Pre-dilated intraocular pressures (IOPs) were measured as 15 mmHg OD and 19 mmHg OS. Slit-lamp biomicroscopy revealed interpalpebral corneal punctate epithelial erosions OU, grade 2+ nuclear and anterior cortical cataracts OU with cortical spokes within the visual axis OD. Dilated fundus exam revealed cup-to-disc ratios of 0.6 OD and 0.4 OS with diffuse disc pallor OD and temporal disc pallor OS. The peripheral fundus exam was unremarkable OU. The patient had difficulty comprehending instructions for confrontational or kinetic visual fields.

The patient’s acquired oculomotor apraxia with intact VOR warranted further investigation to determine the presence or absence of a supranuclear palsy or degeneration. Magnetic resonance imaging (MRI) with and without contrast was ordered and revealed a classic “hummingbird sign” showing midbrain atrophy, as well as atrophy of the superior colliculus, which were not noted in the initial MRI study eight years prior (Figure 1). The patient’s primary care provider (PCP) was notified of the results, and a consultation to the neurology clinic for further evaluation of “possible PSP” was placed.

Figure 1. MRI T-1 weighted images, sagittal view. Left: Initial study eight years prior with greater midbrain and colliculus volume. Right: Current study demonstrating “hummingbird sign” (midbrain atrophy). Click to enlarge
Case 2

A 61-year-old man presented for an eye examination reporting vertical binocular diplopia when reading for the past few weeks. The patient’s medical history included type 2 diabetes mellitus, hyperlipidemia, psoriasis and recently diagnosed PSP. Prior to the diagnosis of PSP, the patient had experienced frequent falls and generalized muscle weakness over the course of three years. Within the course of the three years, the patient had been prescribed carbidopa-levodopa (Sinemet) and his condition was treated as atypical Parkinson’s disease. At the time of his eye exam, he had discontinued carbidopa-levodopa due to a change in diagnosis from Parkinson’s disease to PSP. The patient’s ataxia had significantly progressed, and he was wheelchair-bound shortly before his eye examination. His most recent MRI demonstrated characteristic midbrain atrophy compared to prior studies (Figure 2). Current medications included acetaminophen 650 mg, aspirin 81 mg, atorvastatin 40mg, gabapentin 300 mg qam and 600 mg qhs, lorazepam 1 mg, metformin 500 mg, sertraline 150 mg and tizanidine 2 mg.

BCVAs were 20/20 OD and 20/20 OS. Pupils were equal, round and reactive to light with no afferent pupillary defect present. Confrontational fields were full to finger counting OD and OS. Pre-dilated IOPs were 16 mmHg and 14 mmHg. Slit-lamp biomicroscopy findings revealed unremarkable eyelid and eyelash findings, small nasal pterygium minimally encroaching on the cornea, flat and avascular iridis and deep and quiet angles OU. Dilated fundus exam revealed 1+ nuclear cataract OS, cup-to-disc ratios of 0.45 round and 0.5 round OD, OS, respectively, and otherwise unremarkable posterior pole and peripheral retinal findings. Based on evaluation of the patient’s vergences, esophoria greater at near than distance and a reduced near point of convergence were noted. Given the patient’s symptom of diplopia at near, he was diagnosed with convergence insufficiency secondary to PSP. He was prescribed 2.5 prism diopters base out in single-vision near glasses for greater comfort when reading.

Four months after the initial eye examination, the patient was admitted to a nursing home and was examined at bedside at the request of the PCP and charge nurse due to a reported increase of falls. The patient reported that he preferred to use previously prescribed progressive addition lenses despite having diplopia at near. Evaluation of the extraocular muscles revealed normal lateral gaze and intact VOR but limited superior and inferior movement. Due to these concerns, the patient was educated on the importance of protecting his eyes given his unstable gait and re-prescribed single-vision distance and single-vision near glasses with Fresnel prism to adjust for changes in severity of his diplopia.

Case 3

A 75-year-old male referred for an eye exam by his neurologist presented to the eye clinic for a preoperative cataract evaluation. Doctors were unable to elicit any verbal complaints from the patient, as he had severe aphasia rooting from his diagnosis of PSP. His wife stated he had been less interactive with the family, and she was concerned he was unable to see their faces as well as he used to. The patient’s neurologist had noted that improved vision may help reduce the possibility of agitation and delirium in demented patients. The patient’s medical history included chronic cough (from pooled airway secretions), chronic microaspirations and previous cerebral vascular accident with right arm contracture. His medications included levothyroxine 0.15 mg, memantine 5 mg, ferrous sulfate 300 mg/mL, omeprazole 40 mg, albuterol 05% 2.5 mg/0.5mL, bisacodyl 10 mg, ascorbic acid 250 mg, cholecalciferol 1,000 units, acetaminophen 650 mg, levetiracetam solution 125 mg/1.25mL and glycopyrrolate 1 mg.

BCVAs were unobtainable due to the patient’s poor cognitive state. Pupils were observed to be equal, round and reactive to light with no afferent pupillary defect present. The patient was unable to understand confrontational visual field testing. Upon extraocular muscle testing, he was noted to have restricted superior and inferior vertical gaze but intact VOR movement. Slit-lamp biomicroscopy findings revealed unremarkable eyelid and eyelash findings, flat and avascular iridis and deep and quiet angles OU. The lenses showed 3-4+ milky nuclear sclerotic cataracts OU. Dilated views by fundoscopy were poor and fleeting due to the patient’s tendency to squeeze his eyelids shut with light exposure. The retina was grossly normal with optic nerve cup-to-disc ratios possibly larger than average but difficult to ascertain.

The ophthalmologists gathered from the limited data that the patient would be able to cautiously proceed with cataract extraction pending presurgical medical evaluation. Evaluation by the patient’s pulmonologist determined that rhonchi, or low-pitched rattling sounds from the lungs, were present and suggested possible aspiration or respiratory infection. Given the
patient’s demented mental status, general anesthesia would need to be administered, necessitating prolonged intubation and risking respiratory failure. Additionally, there was concern that coughing could disrupt postoperative sutures and result in a poor surgical outcome. The cataract surgery was thus postponed.

Education Guidelines

Learning objectives

1. Understand neurodegenerative diseases as they relate to ocular health
2. Become familiar with the ocular manifestations of PSP
3. Understand possible complications of general anesthesia when considering ocular procedures in patients with PSP or other neurodegenerative disease affecting motor function

Key concepts

1. Fundamental pathophysiology of PSP
2. Ocular effects of PSP and the optometrist’s role in diagnosis
3. Management of patients with PSP

Discussion points

1. What is the pathophysiology of saccades and PSP?
2. What causes PSP?
3. How is PSP diagnosed?
4. What are the ocular signs and symptoms of PSP?
5. What are some differential diagnoses that should be considered in cases of suspected PSP?
6. What is the appropriate management of patients with PSP?
7. What is the prognosis of patients with PSP?

Literature review

PSP is the most common degenerative, atypical parkinsonian disorder, with a prevalence of 6.4 per 100,000 according to Schrag et al. The incidence is reported to increase with age from 1.7 cases per 100,000 at 50-59 years to 14.7 per 100,000 per year at 80-99 years. The mean age of diagnosis is approximately 65 years, with no racial or sex predilection. No significant risk factors for developing PSP have been identified.

Discussion

Teaching instructions: Participants should read each question and consider how they would respond, then read the information provided in the text. Participants may work together in small groups or individually, either in real time or as part of a homework assignment. If working in groups, participants may split into three groups with each group focusing on one case or each group working on all three cases. Learning objectives can be assessed by comparing participants’ responses to the questions provided. This case may also be presented as a PowerPoint presentation detailing the case presentation, learning objectives, key concepts, literature review and discussion points.

What is the pathophysiology of saccades and PSP?

For horizontal and vertical saccades to be initiated, excitatory burst neurons (EBNs) in the brainstem generate a burst of neuronal discharge known as the pulse. The EBNs for horizontal saccades are located in the paramedian pontine reticular formation. The EBNs for vertical and torsional saccades are located in the rostral interstitial medial longitudinal fasciculus (RIMLF) and interstitial nucleus of Cajal. Once the pulse fires to the intended agonist muscle, antagonist muscles are relaxed.

PSP results from an aggregation of abnormally phosphorylated tau proteins. Tau proteins aid in axonal transport and support neuronal microtubules. Localized accumulation of the irregular tau proteins form what are known as neurofibrillary tangles. In addition to the tauopathy, PSP degenerates dopaminergic neurons and cholinergic neurons, lending to loss of basal ganglia, cerebral cortex and, most clinically characteristic, brainstem structures. Structures within the brainstem that atrophy are the dorsal midbrain, notably the midbrain tegmentum and pedunculopontine nucleus, which lends to postural stability, and the motor nuclei of the cranial nerves in advanced stages of the disease. Given the atrophy of the midbrain tegmentum, the RIMLF is greatly affected, which decreases the presence of vertical EBNs, and an inability to initiate vertical saccades ensues.
What causes PSP?

Although no definitive genetic factors have been identified, recent studies suggest there may be a genetic susceptibility in patients with mutations of the tau gene. PSP is currently considered a “tauopathy.”

How is PSP diagnosed?

The diagnostic criteria for PSP, which were revised in 2017 by the International Parkinson and Movement Disorder Society, must include all “basic features”: sporadic occurrence, age 40 years or older at onset of first PSP-related symptom, and gradual progression of PSP-related symptoms. PSP-related symptoms include ocular motor dysfunction, postural instability, akinesia and cognitive dysfunction. The category of ocular motor dysfunction is further stratified to include vertical supranuclear gaze palsy, slow vertical saccades and frequent macro square wave jerks or “eyelid opening apraxia.” In this delineated criterion, the highest level of certainty for each category is defined as vertical supranuclear gaze palsy, repeated unprovoked falls within three years, progressive freezing of gait within three years, and speech or language disorder that presents as some variant of primary progressive aphasia, respectively. Supportive features that may increase diagnostic confidence, but do not alone suggest a diagnosis, include levodopa resistance, dysphagia, photophobia, and hypokinetic, spastic dystartria.

Clinical forms of PSP have arisen since its original description in 1964. The original cases presented by Steele-Richardso-olszewski have been classified as Richardson Syndrome (PSP-RS), which is characterized by postural instability, vertical gaze palsy and cognitive dysfunction. Eight other clinical variants have been described based on the severity and nature of their neurological signs.

The gold standard for diagnosing PSP is a comprehensive, post-mortem neuropathological examination to identify the presence of neurofibrillary tangles. Because a definitive diagnosis requires a post-mortem exam, it is crucial for an eyecare provider to be able to identify oculomotor restrictions and make appropriate clinical recommendations for the patient’s systemic care.

What are the ocular signs and symptoms of PSP?

Patients with PSP typically present with horizontal or vertical diplopia or asthenopia while reading secondary to convergence insufficiency, horizontal or vertical gaze palsy. Patients may also exhibit impaired slow phase responses of their vertical optokinetic response, as well as slowed volitional saccades. In a few studies, abnormal acoustic-startling reflex (orbicularis oculi response to high-intensity stimulation of the median nerve) and apraxia of eyelid opening have been observed in patients diagnosed with PSP. A decrease in blink frequency may cause symptoms of dry eye, including blurred vision, foreign body sensation, burning or irritation. Appropriate evaluation of extraocular muscle motility and near point of convergence as well as slit-lamp examination to evaluate the tear film and corneal integrity are of particular importance.

What are some differential diagnoses that should be considered in cases of suspected PSP?

From an ocular standpoint, it is important to determine the etiology of a patient’s presenting symptoms of double vision when reading or limitation of gaze rather than assuming such presentations are idiopathic or, as observed in case 2 reported above, presbyopia-associated convergence insufficiency. In assessing these complaints, the clinician must elicit a thorough case history including recent falls or postural changes and memory changes. Additionally, it is critical to carefully assess extraocular muscle movement, saccades, phorias and vergences at distance and near. Forced duction evaluation is particularly valuable in differentiating a mechanical vs. a neurodegenerative or vascular cause. The posterior thalamo-subthalamic paramedian artery, which stems from the posterior cerebral artery, supplies the RIMLF. Thus, an infarction of this artery may result in a superior gaze palsy. Neoplasms, particularly pineal gland tumors, may also lead to vertical gaze palsies. Other vertical gaze palsy conditions include Neimann-Pick type C, an autosomal-recessive condition in which cholesterol and lipids accumulate, dorsal midbrain syndrome, Whipple disease, and midbrain infarction.

As previously noted, PSP is often misdiagnosed early in its course as Parkinson’s disease. It shows similar symptoms to Parkinson’s, dementia with Lewy bodies (DLB) and multiple system atrophy (MSA). Parkinson’s disease is a neurodegenerative disorder with characteristic skeletal muscle tremor, rigidity and akinesia, all of which are features observed in PSP. However, PSP typically involves greater cognitive dysfunction and speech disturbance than Parkinson’s disease. A study by Song et al. assessed patients with MSA, Parkinson’s disease and PSP. Results showed that 73% of patients with PSP had gaze abnormalities, a characteristic that was absent in Parkinson’s disease and MSA. Song also found that patients with PSP and MSA had a poorer response to levodopa, which is a staple treatment for Parkinson’s disease.

MSA is defined as an “adult-onset, sporadic, progressive neurodegenerative disease” with “parkinsonian features, cerebellar ataxia, autonomic failure, urogenital dysfunction, and corticospinal disorders” by the Second Consensus Statement on the
Diagnosis of MSA.22,23 Although difficult to definitively differentiate from PSP, any sign of autonomic dysfunction would steer the diagnosis towards MSA.

DLB is defined by the 2017 Dementia with Lewy Bodies Consortium as a disease of “progressive cognitive decline of sufficient magnitude to interfere with normal social or occupational functions, or with usual daily activities.” In DLB, patients have “recurrent visual hallucinations that are typically well-formed and detailed,” rapid eye movement (REM) sleep behavior disorder, and “features of parkinsonianism.”24 Gait abnormalities, gaze apraxia and saccadic dysfunction may be present in both PSP and DLB, but visual hallucinations are a hallmark feature of DLB.25,26

Various presenting signs and symptoms overlap between PSP and other neurodegenerative disorders, which makes PSP diagnosis reliant on the clinical findings centered around the 2017 Movement Disorder Society criteria. Neuroimaging such as MRI may be a helpful supplement for diagnosis. Various studies report abnormalities on MRI demonstrating midbrain atrophy, third ventricle dilation, T2-periquested hyperintensities and frontal and temporal atrophy. One hallmark feature of PSP on MRI is the “hummingbird sign” or “penguin sign” demonstrating midbrain atrophy. Most useful for differentiating PSP from Parkinson’s disease or other atypical Parkinson’s diseases are the midbrain-to-pontine area ratio, which tends to be lower in PSP patients, and the magnetic resonance parkinsonism index (MRPI), which is greater in PSP patients. The MRPI is a value calculated by determining atrophy of the midbrain, superior cerebellar peduncle,pons and middle cerebellar peduncle.27,28 Another promising diagnostic method is utilizing positron emission tomography scanning to track the tau protein THK5351 and determine the presence of tau aggregates specific to PSP.29 This diagnostic tool, however, is predominantly used in research and often not clinically performed due to cost constraints.

What is the appropriate management of patients with PSP?

Eyecare providers are crucial in determining the presence or absence of gaze palsies, which aids in the differentiation of PSP. Once an abnormality is determined, a referral to the patient’s PCP and neurologist should be made.

Patients with PSP often have limited benefit from levodopa therapy, as opposed to patients with Parkinson’s disease. In a study by Williams et al. investigating 91 pathologically confirmed cases of PSP, 32% of patients presented with a response to levodopa, defined as a 30% or greater improvement in symptoms.30 There are no successful pharmacological treatment options targeting the disease process, and management centers around symptomatic care. A few case reports have demonstrated some efficacy of botulinum toxin injected into the orbicularis oculi for apraxia of eyelid opening, into upper limbs to improve rigidity and into cricopharyngeal muscle for dysphagia.31-34 Additional intervention from an interdisciplinary care team consisting of speech, physical and occupational therapists is necessary to facilitate greater independent activities of daily living.

Following diagnosis of PSP, the optometrist may play a role in the patient’s care, particularly to address diplopia and dry eye symptoms. A study by Reddy et al. found that corneal sensitivity was reduced in patients with PSP, with 71% reporting they did not have dry eyes. However, evaluation of the tear film revealed reduced tear break-up time compared to the control group.35 Even without patent report of dry eye symptoms, it is important for the optometrist to manage the patient’s dry eyes with artificial tears and lubricating ointment or gel formulations for severe exposure keratopathy. Although few reports demonstrate alleviation of double vision when reading with “mirror prism,” we found that Fresnel prisms were particularly useful for alleviating the progressive double vision experienced by the patient in case 2.36,37

What is the prognosis for patients with PSP?

PSP is a rapidly neurodegenerative condition with a poor prognosis. A study by Cosseddu et al. evaluated 100 patients with PSP and found that the average disease duration following diagnosis was 8.25 years.3 Cosseddu found that patients with dementia at the time of diagnosis had a shorter survival time than those without dementia, with no other significant predictors.3 The most common causes of death for patients with PSP are respiratory-related, with the most frequent complication being aspiration pneumonia.36 Given the high frequency of respiration complications, surgery requiring general anesthesia should be given significant consideration due to the potential for respiratory failure.

Conclusion

Optometrists may play a critical role in recognizing characteristic features of PSP and managing concomitant ocular symptoms that may arise. Although no pharmacologic therapy halts progression of the disease, a multidisciplinary team can provide patients and their caretakers with the tools to maximize quality of life and minimize debilitating symptoms.

References

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