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Visibility

Education and Research
from Envision

Stargardt Disease: The View From 2011..... *Janet S. Sunness, MD*

Stargardt disease is the most common cause of macular degeneration and central visual loss in young people. Stargardt disease often develops in the teens or twenties, but may develop in younger children (the youngest child I have seen is 5) or somewhat later in life. It affects both eyes at the same time, though one may be somewhat worse than the other. In most people it affects only central vision (reading, recognizing faces and other fine tasks), but in certain more rare types, peripheral vision may be affected as well. Vision loss tends to progress rapidly at first, but for many people it tends to stabilize at about 20/200. There is also a form

of this condition that begins later in life and tends to progress more slowly.

DIAGNOSING STARGARDT DISEASE

Patients with Stargardt disease have findings that are different from patients with age-related macular degeneration, and some terminology is worth knowing. Many patients with Stargardt disease have yellow flecks in their retina. In these patients, the disease is also known as fundus flavimaculatus (the retina with the yellow spots). The macula, the center of the retina, often has a shiny appearance, which is referred to as a “beaten bronze appearance,”

and unlike age-related macular degeneration, it is often difficult to determine just by looking at someone how much of a blind spot the patient may have. The fluorescein angiogram (when dye is injected into the arm and pictures of the retina are taken) often shows an unusual appearance called a “dark choroid,” meaning that the initial flush of dye which goes through the blood vessel system beneath the retina cannot be seen. Some patients with Stargardt disease start out with a bullseye appearance to the macula, in which there is a small island of spared vision surrounded by a donut-shaped blind spot. In other patients, or later in the course of the disease,

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ABOUT ENVISION FOUNDATION

The mission of the Envision Foundation is to secure funding for the successful delivery of services offered by the Envision Vision Rehabilitation Center and the education programs of the Foundation. Envision Foundation focuses on fundraising to ensure that no patient is ever turned away—regardless of ability to pay; public education to help prevent blindness; and professional education to determine best practices in order to serve patients who are blind or low vision.

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Stargardt Disease: The View From 2011 continued from front page

there is a solid area of blind spot which does not spare the very center.

DEALING WITH BLIND SPOTS IN THE CENTRAL VISION

It is critical for any patient with blind spots in or near the center of vision to learn how to move the eye so that the object of interest is imaged onto a seeing part of the retina. Most Stargardt patients do this quite well and tend to look slightly above what they want to see. This has the effect of moving the blind spot up and out of the way. The difficulty is that as one uses retina farther from the center of the macula, larger letters are needed in order to see; magnification, using a variety of low vision devices, can be very helpful.

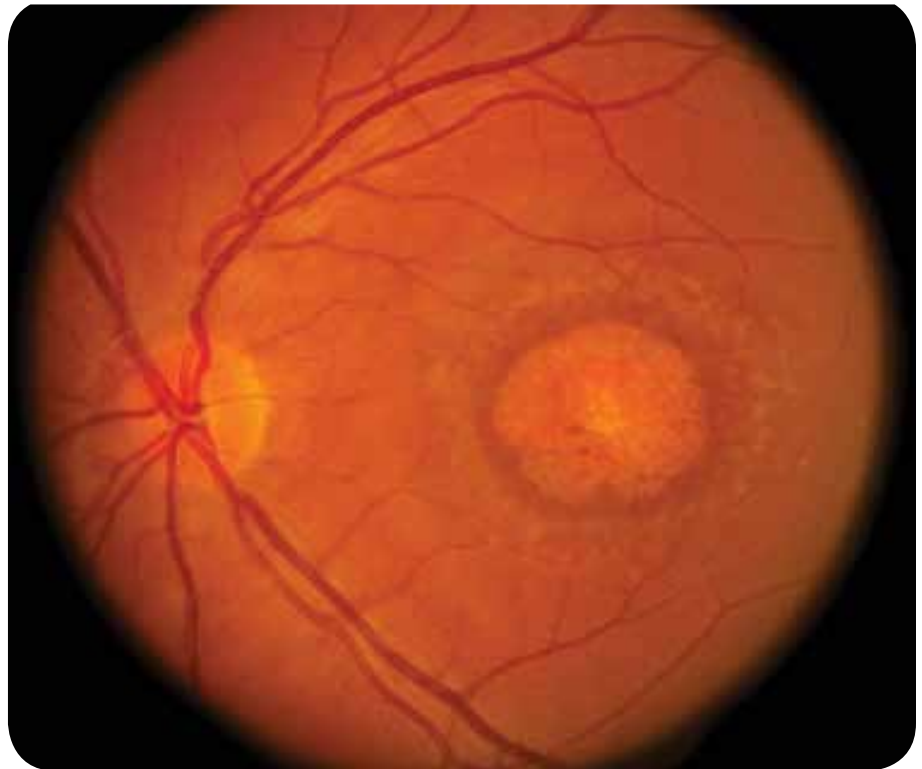
A diagnostic device called a microperimeter can allow us to plot out the exact location of the blind spot directly on a video image of the macula. This information helps us to advise patients on strategies for using the remaining seeing retina in the most effective way. Other specialized imaging, using infrared and autofluorescence imaging, allows us to determine the stage and progression of the condition more comprehensively.

LIVING WITH STARGARDT DISEASE

Most patients whom I see with Stargardt disease have adapted quite well; they lead busy and active lifestyles. Among these patients is a scientist, radio announcer, a social worker, and a tandem bike racing

competitor. They have rewarding personal and professional lives. Improved low vision interventions in the future will help patients to adapt even better to this condition. It is important to remember that a loss of vision is a real loss to the individual, and some people may react to this with anger and depression. While these are normal stages many

Some patients with Stargardt disease are able to drive. Each state has different rules in terms of the minimum visual acuity required to get a restricted license. Patients who meet this vision requirement can work with special driving instructors who can instruct them and determine whether they can drive safely. GPS systems are probably the best device for drivers



Many patients with Stargardt disease have yellow flecks in their retina known as fundus flavimaculatus.

patients go through, these emotions may interfere with the person's ability to adapt and see around the blind spot. If there is difficulty in working through these emotions, support groups and counseling may be beneficial. Support groups are very helpful in general for understanding the symptoms of central visual loss and for learning different ways of coping.

with somewhat reduced central vision. Patients can also learn how to use a bioptic telescope to read street signs and traffic signals.

THE GENETICS OF STARGARDT DISEASE

Stargardt disease is an inherited, or genetic, disease; that is, it is caused by a mutation of a gene. Most patients I see with Stargardt

are the only ones in their family to have the condition, and they are likely to have the autosomal recessive form, meaning that both copies of the gene (one from the mother and one from the father) are abnormal. The children of these patients in all likelihood will be carriers of the disease (have one abnormal gene) but will not be affected, assuming that the other parent is not a carrier. Patients may have siblings who are affected, but their parents and children are not likely to be affected. There is also a rarer autosomal



A student with Stargardt disease uses a computer with assistive technology

dominant form, meaning that only one copy of the gene has to be abnormal. In the dominant form, one parent of the patient also has the disease and other siblings or family members may have it as well.

Researchers have found that mutations in a gene called ABCA4 are responsible for at least 75 percent of the cases of Stargardt disease. This gene is responsible for helping transport vitamin A derivatives (retinoids) from the visual cycle, so that these retinoids can be regenerated and reused. When the retinoids cannot be properly trans-

ported, it leads to an accumulation of a material called lipofuscin that may be toxic to the retinal pigment epithelium, the cells under the retina needed to sustain vision. The high levels of accumulation begin at birth; it is not clear what triggers the cells to start degenerating.

ADVANCES IN POTENTIAL TREATMENTS FOR STARGARDT DISEASE

The ABCA4 gene responsible for Stargardt disease was discovered in 1997.¹ A great deal of progress has

been made since that time. Scientists have been able to develop mice that are missing the ABCA4 gene (so-called knockout mice). These mice accumulate large amounts of lipofuscin, as do people.

But since mice retinas do not have

the specific structure of a macula, they do not undergo the degeneration seen in people. Scientists have studied whether various drugs or other interventions can slow or stop the accumulation of the abnormal material in these mice. They have found that if you do not expose the mice to light, there is less lipofuscin accumulation. It is difficult to know how this applies to people, but some investigators recommend that patients wear sunglasses when they are outdoors. They also found that giving the mice Acutane (a vitamin A derivative used for severe acne that

has serious complications such as birth defects and psychiatric side effects) slowed down the accumulation of the abnormal material. Research is ongoing to find safe therapies that may help in this condition.

In the last few years, the first successful animal and human gene therapy trials have been conducted for a severe retinal degeneration that begins at birth (Lebers Congenital Amaurosis, associated with an RPE65 mutation). Normal copies of the gene are attached to virus vectors, which help to gain entry of the genetic material into the cells of the patient. This is injected underneath the retina. Similar gene therapy, using normal copies of the ABCA4 gene and a different virus vector, has been successful in the knockout mouse model. Since there is no naturally occurring Stargardt disease known in animals, scientists have used dogs that do not have a mutation to test that the gene can enter the desired cell type. These studies have gone well.² In 2011, the StarGen study of gene therapy for Stargardt disease began. The first phase is a safety study. It will involve follow up of patients for about a year to look for any evidence of side effects or complications affecting the eye or any other part of the person. For this phase, only patients with very severe vision loss will be enrolled.³

A second clinical trial that began in 2011 uses stem cells to treat Stargardt disease (NCT01345006; Advanced Cell Technology; see clinicaltrials.gov). Stem cells are transformed into retinal pigment epithelial

cells. These cells are then injected under the retina. The initial phase is to assess safety with various numbers of cells injected (beginning with 50,000). Since this has not before been performed, it is not known to what extent there will be an inflammatory reaction to or rejection of the cells, or what other side effects will occur. This study is therefore only enrolling patients with very severe vision loss at this time. While the ultimate goal would be for the injected cells to replace the degenerated cells of the patient, this will be very difficult to achieve. A more modest goal would be for the injected cells to secrete growth factors that slow down cell death.⁴


Researchers are also studying ways to slow down cell death in general. They are studying, in patients

with a severe type of retinal degeneration, whether cell death can be slowed by growth factors. They have made a small reservoir containing cells that have been genetically engineered to make a growth factor. They will implant this reservoir in the eye, and, like an internal factory, the growth factor that is made will be able to diffuse out of the reservoir and into the eye.

Investigators are working on different types of “artificial retinas” that could take advantage of the fact that the nerve connection system from the retina to the brain is still normal in patients with Stargardt disease. What are diseased are the retinal pigment epithelium and the photoreceptors, the cells that “catch” the light and send a signal to the other nerve cells in the retina. If one could

replace the photoreceptors with an electrical or other type of light sensor, and could send the signal to the nerve network in the retina, the patient would be able to see. The development of artificial retinas is still in the early stages⁵, but progress continues to be made.

CONCLUSION

Stargardt disease, for the foreseeable future, will remain the primary cause of central vision loss in young people. With appropriate low vision intervention, these patients generally do well, and they are generally open to using new high-tech options. There is great potential for future treatment of Stargardt disease in clinical trials that are beginning now. 

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Dr. Sunness is a leading international expert in macular degeneration and in methods of measuring macular function, with particular emphasis on dry age-related macular degeneration and Stargardt disease. She is an ophthalmologist, with specialties in medical retinal disease and low vision. She has pioneered the use of the scanning laser ophthalmoscope to learn more about macular disease and how patients use their peripheral retina when they have a central scotoma. She was awarded the Gass Medal for outstanding work on macular disease at the 2011 annual meeting of the Macula Society. Dr. Sunness is the Medical Director of the Richard E. Hoover Low Vision Rehabilitation Services at the Greater Baltimore Medical Center and a member of the Foundation Fighting Blindness working committee on Stargardt disease.



Utilizing Red Contact Lenses for Retinal Dystrophies

William Park, OD, FAAO



Release Date: December 2011

Expiration Date: December 1, 2014

Instruction Level: Introductory

Goal Statement: Retinal cone dystrophy affects the quality of life with debilitating photophobia and decreased visual acuity of thousands of people in the United States. No treatment for retinal dystrophies is available, but low vision aids, including red-tinted contact lenses, can be helpful in selected patients. This educational activity reviews the diagnosis, evaluation and therapeutic interventions via contact lens fitting available to patients with retinal dystrophies. Post-fitting management and patient experience will also be presented.

Faculty/Editorial Board:

William L. Park, OD

Target Audience: Optometrists

Credit Statement: This course is COPE approved for 1 hour of CE credit. COPE ID is 33197-CL. Please check with your state licensing board to see if this approval counts towards your CE requirement for relicensure.

Sponsorship Statement: This continuing education course is sponsored by Envision. Envision is a COPE-approved administrator/provider of CE courses.

Disclosure Statement: Dr. Park has no financial interest.

A retinal cone disorder is a general term used to describe a group of rare inherited ocular disorders characterized by the loss or dysfunction of cone cells, the photoreceptors responsible for both central and color vision. The most common symptoms of cone dystrophy are visual loss, sensitivity to bright lights, and poor color vision. Achromatopsia is cone dysfunction that begins at birth. A cone dystrophy is a disease that progresses gradually over time, with an age of onset ranging from the late teens into the forties. Color vision HRR testing (Hardy, Rand and Rittler, Pseudoisochromatic Plate Test Series) reveals many errors on both red-green and blue-yellow plates.¹⁻²

Patients with cone disorders often have debilitating photophobia over and above their decreased visual acuity. This prevents them from optimal visual functioning in the daytime and even in ordinary indoor illumination. Patients often need to wear a baseball cap or visor in addition to light-absorbing glasses; even then, they may still find it difficult to function.

No treatment for the disease is available, but low vision aids, including red-tinted contact lenses, can be helpful in selected patients.

In the early 1990s, this author became perplexed with two brother and sister pairs who had a high compound hyperopic astigmatic

correction in one family, compound myopia/astigmatism (CMA) in the other family; each patient was legally blind. All four were diagnosed and validated to have cone dystrophies (achromatopsia) based on electro-diagnostic testing, objective clinical findings and subjective history. All four demonstrated nystagmus, BCVA ranging from 20/200 to 20/400 and high refractive error greater than 5 diopters of hyperopia or myopia along with 3 diopters or more of astigmatism with dark 500 series CPF lenses incorporated in spectacles to counteract the incapacitating aversion to normal illumination.

The four children were 6, 9, 10 and 12 years of age; they were self-conscious because of their appearance which consisted of high prescription, brown-tinted lenses and a hat (being worn in the classroom – unlike their peers). The contact lens was originally designed with a 6 mm pupil “to hide” the red tint relative to peer pressure (all had blue irises). Upon routine CL follow-up, one of the children asked for a full tint to block out light further, stating, “she had even more boyfriends as a result of her red eye presentation.”

RESEARCH INTO RED CONTACT LENS USE

In 2004, Park and Sunness³ reviewed 23 patients with cone disorders and significant photophobia

not ameliorated with standard methods. Two patients had cone-rod degeneration, one had cone dystrophy, and the remaining 20 patients had achromatopsia.

The mean visual acuity with

glasses was 20/200 (range, 20/80–20/400). The mean age was 17 years (range, 4–55 years). Mean visual acuity with the red contact lenses improved to 20/125. Eight patients became newly eligible for

a motor vehicle license, with visual acuity of 20/100 or better.

All patients were fitted with soft contact lenses of 55 percent water content, methafilicon material (Kontur Kontakt Lens, Hercules, California, USA) using a translucent red (K21/20) 11-mm zone in a 15-mm overall diameter. These lenses allowed for approximately 30 percent light transmission. The tint density prescribed (medium, dark or very dark; percent transmission was not available from the company) depended on the degree of alleviation of photophobia indoors and outdoors.

The use of red contact lenses to alleviate photophobia capitalizes on both the tint of the lens and the advantages of a contact lens versus spectacles. Contact lenses are advantageous, in that complete pupillary coverage limits peripheral glare, reduces optical surfaces, and eliminates rear surface reflections. Furthermore, due to the nystagmus present in all the patients with achromatopsia, the visual axis is never aligned with the optical center of the spectacle lens, resulting in degradation of vision. Therefore, contact lenses are the primary mode of obtaining best-corrected visual acuity if tolerated subjectively.⁴⁻⁶

MANAGING EXPECTATIONS FOR SUCCESS

My philosophy for successful fitting and long-term wear is that I do not fit patients whom I do not think will be successful. The success is

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Cone Retinal Dystrophy & Red Contact Lenses



Before and after. After being fitted for contact lenses, this patient went on to complete her GED, obtain two undergraduate degrees and became gainfully employed with the Social Security Administration.

Case Report: How the Amber-Red Contact Lens Changed My Life

William Park, OD and Marcia Harris, CLVT, St. Louis Veterans Medical Center

Patient Ocular History

28 YO AAF

Occupation: MS, ED, CLVT at
St. Louis VA

CC: Extreme photophobia

Allergies: PCN

FOHx: AMD – Grandmother

VAsc: OD 20/300
OS 20/125-1

VAcc: OD 20/100+2
OS 20/100+1

K's: 43.50 x 44 @ 076
43.25 x 45.00 @ 092

EOMs: WNL/Full & Smooth

Color/Isihara 2/17
red/green deficiency

Anterior Segment NAP OU

IOP OD 13
OS 15

Dilated Fundus Exam

Posterior Pole Beaten Bronze
presentation with
circumscribing flecks OU
Peripheral Retina NAP OU
Optic Nerve NAP OU

Prescribed contacts

1 month later

Kontur Kontakt Lenses

Narcissus Red after 20

minutes wear time

8.6 15.0 -1.50 20/70

8.6 15.0 -1.25 20/80

OU 20/70

As a Certified Low Vision Therapist and a person with Stargardt disease, I've learned there are many myths about visual impairment: that people will eventually go blind, prescription eyewear will not help, and enhancing contrast will not improve a patient's visual acuity. Those of us in the field know most patients maintain a useful amount of vision and prescription eyewear can

improve a patient's visual functioning. Interestingly, contact lenses are rarely considered as viable options for improving the low vision patient's visual acuity. In my personal journey, I found prescription glasses without contrast enhancement offered little benefit. However, the combination of both improves my visual acuity, decreases the appearance of my blind spot and reduces light sensitivity. Fortunately, low vision specialists are now recognizing tinted contact lenses as a valuable therapeutic intervention for photophobic low vision patients like me. That's how I met William Park, OD.

Let's be honest; as a young professional, I don't want my visual impairment to be the first topic of conversation when I meet someone new. Unfortunately, wearing polarized amber sunglasses indoors

makes that difficult. I often thought to myself, "It would be great if someone could make me a pair of tinted contact lenses." I did



some investigating and learned Dr. Park and his fellow researchers were doing just that. As I read on, I discovered tinted contact lenses were ideal for patients like me who have difficulty looking directly through the center of their eyeglasses. For example, my blind spot, or central scotoma, requires me

to look off to the side to see objects when the prescription is actually in the center of the eyeglass lens. I scheduled an appointment with Dr. Park and was anxious for my contact lens evaluation.

In October 2010, Dr. Park performed a thorough evaluation, which included a scanning laser ophthalmoscope (SLO) visual field test, checking my eyeglass prescription, and a tinted contact lens evaluation. He noted a change in my prescription and my 5-8 degree central scotoma. I tried several shades of contact lenses and selected an amber-red tint. Dr. Park and his staff had me evaluate the contact lenses indoors and outdoors before making my final selection. The amber-red contact lens is the best low vision aid I've ever experienced. My blind spot is less noticeable, I am no longer affected

by indoor glare, and I can navigate social and professional situations with greater confidence.

Since the contact lenses stay on the center of my eye, that means I am always looking through the best part of the lens. When I wore prescription sunglasses, my blind spot covered a person's entire face when I looked directly at them. I had to look slightly to the right to read a person's facial expression. With my contact lenses, the blind spot appears smaller, so I can look directly at the person to whom I'm talking. The blind spot is still there, but it's so small that it only covers a small part of their face. For those of us in the field, this means my preferred retinal locus (PRL) is closer to central fixation. I no longer have to wear sunglasses to reduce the glare from my television or computer screen. I can also recognize coworkers from farther away and perform work-related tasks with more ease. As a result, I have less visual fatigue after a full day's work. The lenses are light enough that they do not significantly affect my color vision and I can typically wear a pair of store-bought sunglasses outdoors. The dramatic difference tinted contact lenses has made in my life makes me wonder how many other patients could benefit? My hope is that tinted contact lenses are considered as a potential therapeutic intervention for any low vision patient with severe light sensitivity and poor contrast sensitivity.




Pictured: 6mm vs. 14.5 mm red contact lens button on green iris eye

derived from a three- to five-hour trial in the outside environment (based on the initial low vision examination) with lenses fabricated according to the findings of that visit. It is my opinion that the contact lens trial should always be scheduled at a second visit, with subjective environmental appraisal of the contact lenses as the primary focus.

Activities encouraged are: travel in an automobile, going to a mall or shopping, eating a meal at a restaurant, attending a movie, visiting a museum or outdoor play. The sole objective is to have the support team interact with and evaluate the likelihood of the patient's successful wear based on "real ADLs." This is especially important with pediatric patients as indicated in other studies.^{7, 8}

All my patients have suffered moderate to high degree of visual impairment or worse. Substantiation of the subject's motivation (regardless of age) for potential of improved outcome measures in

visual acuity, contrast sensitivity, binocularity, stereopsis, activities of daily living (ADL) function and quality of life (short and long term) are considered.

Contact lenses should be an important consideration for best-corrected visual acuity for high refractive errors and/or anatomical ocular disease manifestations such as aniridia (prosthetic iris contact lens). They can be critical as a component of short and long-term BCVA considerations for infants due to nystagmus and their high refractive error. Contact lenses may also be instrumental in enhancing daily visual function for persons with retinal dystrophies. 

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Utilizing Red Contact Lenses for Retinal Dystrophies

Directions: To obtain 1 hour of continuing education credit, complete the exam by recording the best answer to each self-assessment question on the Examination Answer Sheet. Mail answer sheet to Envision-CE, Attn: Michael Epp, 610 N. Main, Wichita, KS 67203. A minimum score of 70% is required to obtain a certificate of completion. There is no fee for this course.

1. Cone disorders _____.

- a. Often manifest in patients as a debilitating photophobia
- b. Is a group of rare inherited ocular disorders
- c. Both a and b
- d. None of the above

2. A common symptom of retinal cone dystrophy is poor color vision revealed by testing errors with a _____.

- a. Congenital stationary night blindness test
- b. HHR series color vision test plates
- c. Distance visual acuity of count fingers test
- d. Both a and b

3. In the Park and Sunness study³ the mean visual acuity with glasses of the subjects was _____.

- a. 20/400
- b. 20/200
- c. 20/100
- d. 20/80

4. One therapeutic intervention for patients with cone dystrophy is _____.

- a. Accommodative spectacles
- b. Multifocal IOL
- c. The use of red contact lenses
- d. Video magnifiers

5. Using red contact lenses to alleviate photophobia capitalizes on _____.

- a. The tint of the lens
- b. Advantages of contact lenses versus spectacles
- c. BCVA
- d. All of the above

6. The use of red contact lenses is advantageous for _____.

- a. Hyperopia control
- b. Myopia control
- c. Changing iris color
- d. Limiting peripheral glare

7. According to the author, successful fitting and long-term wear of contact lenses is achieved by _____.

- a. A successful BCVA result on the initial low vision examination
- b. A second patient visit lens trial, with subjective environmental appraisal
- c. Pre-fabricated red contact lenses
- d. All of the above

8. During the post-fitting management phase of a red contact lens patient, which activities should be evaluated?

- a. Evaluation of a three- to five-hour trial in the outside environment
- b. ADLs
- c. Interaction with the support team
- d. All of the above

9. Which of the following is NOT a factor that would affect a patient's candidacy to be prescribed red contact lenses?

- a. Non-compliance
- b. Active eye disease, injury or any abnormal state of the cornea, conjunctiva or eyelids
- c. A diagnosis of Stargardt disease
- d. Both b and c

10. The patient in the Case Report experienced _____ as a result of being fitted with red contact lenses.

- a. Alleviation of central scotoma
- b. A preferred retinal locus (PRL) closer to central fixation
- c. Less visual fatigue
- d. Both b and c

Examination Answer Sheet
Valid for credit through December 1, 2014

Utilizing Red Contact Lenses for Retinal Dystrophies

Directions: Select one answer for each question in the exam and completely darken the appropriate circle. A minimum score of 70% is required to earn credit.

Mail to: Envision CE, Attn: Michael Epp, 610 N. Main, Wichita, KS 67203

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There is a six- to eight-week processing time for this exam.

- | | |
|--------------------|---------------------|
| 1. (A) (B) (C) (D) | 6. (A) (B) (C) (D) |
| 2. (A) (B) (C) (D) | 7. (A) (B) (C) (D) |
| 3. (A) (B) (C) (D) | 8. (A) (B) (C) (D) |
| 4. (A) (B) (C) (D) | 9. (A) (B) (C) (D) |
| 5. (A) (B) (C) (D) | 10. (A) (B) (C) (D) |

11. The goal statement was achieved:
☐ Very Well ☐ Adequately ☐ Poorly

How long did it take to complete this course

12. The information presented was:
☐ Very Useful ☐ Useful ☐ Not Very Useful

13. The difficulty of the course was
☐ Complex ☐ Appropriate ☐ Basic

Comments on this course:

14. Your knowledge of the subject was increased
☐ Greatly ☐ Somewhat ☐ Hardly

Topics you would like it he future CE articles:

15. The quality of the course was:
☐ Excellent ☐ Fair ☐ Poor

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The following is your: ☐ Home Address ☐ Business Address

Business Name

Address

City State

Zip -

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By submitting this answer sheet, I certify that I have read the lesson in its entirety and completed the self-assessment exam personally based on the material presented. I have not obtained the answers to this exam by any fraudulent or improper means.

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Lesson 111201.1

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William L. Park, OD, FAO, is in private practice in Wichita, Kan. Dr. Park is committed to outreach efforts in stemming the epidemic of diabetes.

He works exclusively with patients referred for low vision evaluation, low vision rehabilitation



and neurological vision loss. He is a past Director of Low Vision Services, Lions Research & Rehabilitation Center, Wilmer Eye Institute-Johns Hopkins University. Dr. Park can be reached at William L. Park, OD, LLC, www.parklowvision.com, 610 N. Main, Suite 201 Wichita, KS 67203, (316) 440-1690 or drpark@parklowvision.com.

Cataracts: An Overview for the Low Vision Professional

..... Dasa V. Gangadhar, MD

Low vision rehabilitation is, by nature, a collaboration of medical professionals and other low vision specialists. Not all involved with the continuum of care know the exact science of cataract formation or removal. For this reason, the basics are necessary in order to fully understand the etiology and treatment options for patients who may have vision loss due to cataracts.

NORMAL EYE ANATOMY

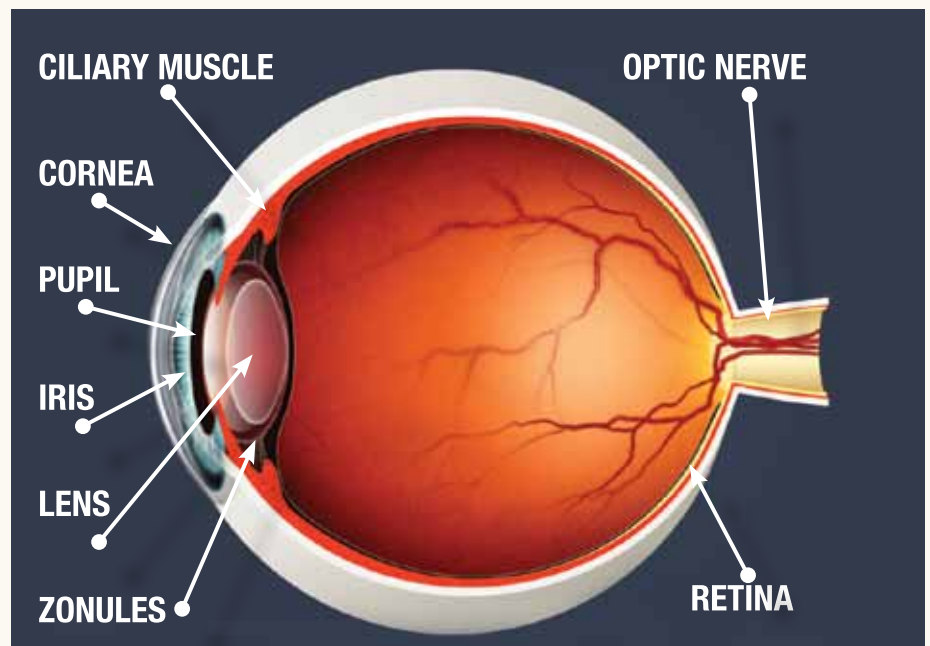
Vision is a complicated process with light rays passing through various structures of the eye (cornea, pupil and lens) before becoming focused on the retina, which serves much like film in a camera. The lens is one of the primary focusing structures of the eye, and sits just behind the colored part of the eye.

WHAT IS A CATARACT?

A common myth is that a cataract is a growth or film that forms on the eye's surface. However, a cataract is the clouding of the normally clear lens inside the eye. The normal lens is formed of neatly arranged protein fibers that allow light to pass easily and become focused clearly. In a cataract, these fibers clump together to form opaque clusters and the entire lens becomes cloudy and disrupts the passage of light into the eye. This can make images seem dull or blurry and interfere with a patient's ability to see clearly.

WHAT CAUSES CATARACTS?

For the vast majority of individuals, cataract development is part of the normal aging process. Heredity,



however, can play a role in how early or late in life a patient develops a cataract.

Risk Factors: Certain risk factors, alone or in combination with age, can increase the risk of developing cataracts as an adult.

- Smoking
- Excessive exposure to UV rays (sunlight exposure)
- Significant eye injuries
- Poorly controlled diabetes
- Long-term use of certain medications, primarily corticosteroids

SIGNS AND SYMPTOMS OF CATARACTS

The word cataract is derived from the Greek word for “waterfall.” If a cataract becomes advanced enough, a patient's vision can fail and they may feel like they are seeing everything through a sheet of rushing water. Today, it is rare for a patient's cataract to advance that far because treatment is usually sought at a much earlier stage.

Classic symptoms include:

- **Blurred vision:** Fuzzy vision is a common first symptom

of cataracts. Street signs are more difficult to see and reading becomes a challenge. Many patients even present the complaint that they can no longer see the golf ball.

- **Night vision difficulties:** Patients with cataracts may feel unsafe driving at night. Glare from oncoming headlights can become blinding and streetlights can take on a halo effect.
- **Excessive brightness and glare:** Even sunlight can be blinding and create intolerable glare for patients.
- **Prescription changes:** Frequent eyeglass prescription changes are needed. With early cataract formation, it is appropriate to change a patient's prescription in order to improve vision. However, with more advanced cataracts, eyeglass changes are no longer beneficial.
- **Dimness of colors:** Cataracts cause colors to become washed out and dull. Because cataracts usually develop slowly, patients are often unaware of this color change. It is only after cataract surgery that many patients realize the profound impact that the cataract had on color perception and brightness.

- **Increased eyestrain:** For many cataract patients, reading, working on a computer, or using their eyes for daily activities can create strain or fatigue.

- **Double vision:** Patients may see "ghost" images or an out-of-focus shadow effect.

CATARACT TREATMENT

There are no non-surgical medical treatments for cataracts. No eyedrops, exercises, medications, or glasses will cause cataracts to regress or disappear. Surgery is the only definitive treatment for cataracts. The surgical technique has progressed so much that the incision can be as small as two millimeters and a patient's return to excellent vision can often occur within a few days. Cataract surgery has become the most commonly performed surgery and one of the most successful. Surgery can have a life-changing impact for many patients.

Not everyone that has a cataract needs surgery. "If it is not broken, don't fix it." If a patient is not yet struggling or limited in their daily activities, treatment may not be necessary. Patients may be able to compensate by changing glasses, using magnifying lenses or utilizing improved lighting when reading or working.

Patients can be trained on these simple modifications through low vision rehabilitation. Through training in activities of daily living, assistive technology, adaptive aids and

"Even in individuals with low vision, cataract surgery may still be appropriate. At minimum, cataract surgery can improve peripheral vision, brightness and colors."

appropriate lighting, patients can continue to utilize their remaining functional vision.

When a patient's cataract progresses to a point where it is significantly interfering with daily living or compromising their lifestyle (driving, cooking, reading, performing their job, sewing or golfing) it may be time to recommend surgery.

Several decades ago, doctors waited until a cataract became "ripe"—when the lens became fully opaque—before surgery was performed. With modern surgical techniques and excellent surgical results, it is better to proceed earlier than was customary in the past.

SURGICAL TECHNIQUE

Modern cataract surgery is performed on an outpatient basis, either in an ambulatory surgical center or in a hospital. Surgery is performed with anesthetic eye

drops or local anesthesia. Patients do not need to undergo general anesthesia. Surgery is typically painless, both during and after surgery. Using an operating microscope, tiny surgical instruments are used to break apart and remove the cloudy cataractous lens from the eye. Ultrasonic techniques (called phacoemulsification) are the most modern techniques of cataract removal. Lasers are not used in cataract removal. The back membrane of the lens (called the posterior capsule) is usually left in

place. A man-made artificial lens is then implanted into the eye to replace the natural lens that was removed.

Visual recovery is usually very rapid and patients can resume normal daily activities almost immediately.

The lens capsule (the membrane that holds the artificial lens in place) can become cloudy several months or years after the original cataract operation. If the cloudy capsule



An intraocular lens (IOL) implant

blurs a patient's vision, a laser procedure (posterior capsulotomy) can be painlessly performed to make an opening in the capsule, thereby restoring normal vision.

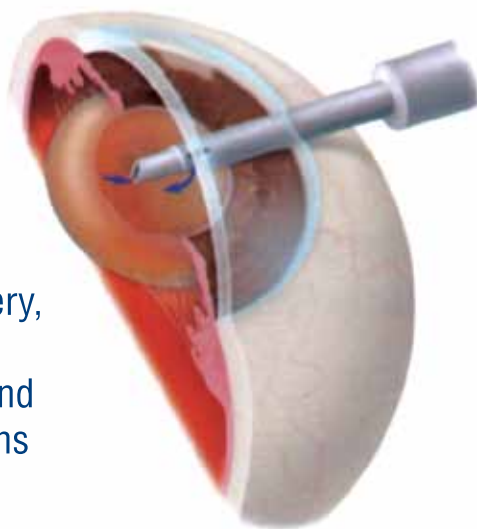
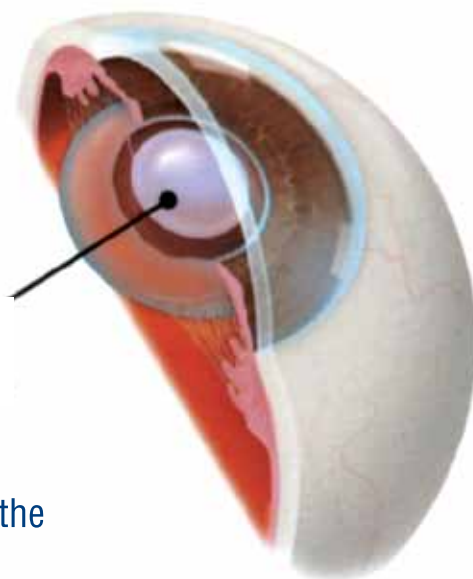
LENS IMPLANTS

Modern lens implants are usually made of a foldable material (acrylic or silicone) that can be implanted into the eye via very small incisions. The power of the lens must be individually calculated for each surgery. Lens implants today can correct near-sightedness and far-sightedness. The most modern specialized lenses can even correct astigmatism and can have multifocal powers for best uncorrected distance and near vision. Surgery can make one much less dependent on glasses, but often reading glasses or glasses with a low prescription are still needed for patients to obtain their best vision.

Intraocular lens implant

In cataract surgery, the intraocular lens replaces the eye's natural lens.

During cataract surgery, tiny instruments are used to break apart and remove the cloudy lens from the eye.




LOW VISION AND CATARACTS

There are many causes of compromised vision beyond cataracts. Macular degeneration, inherited ocular disorders, glaucoma, and other eye diseases can create uncorrectable visual compromise. Individuals with these diseases can, and will, still develop cataracts. Even

in individuals with low vision, cataract surgery may still be appropriate. At minimum, cataract surgery can improve peripheral vision, brightness and colors.

SUMMARY

Today's surgical innovations have effectively eliminated the fear of vision loss from cataracts. Modern cataract surgery is performed on millions of patients annually and is successful in over 95 percent of cases. Even patients with low vision from other causes may still benefit, to a limited extent, from cataract surgery. Regular eye exams are necessary to diagnose and treat eye diseases before they become more serious. Modern medicine continues to expand the boundaries of eye care and eye surgery. 

This article was originally published in *Visibility* 2009; 3(3):6-8

Dasa V. Gangadhar, MD, is an ophthalmologist practicing with Grene Vision Group, Wichita, Kan. Dr. Gangadhar's areas of clinical interest include cataract and implant surgery, corneal transplantation and cornea, refractive and external diseases. He is also the Medical Director for the Kansas Eye Bank and Cornea Research Center.



Envision Seeks Subjects to Participate in National Clinical Research for the BrainPort® Vision Device Clinical Study

The Envision Foundation recently announced that the Envision Vision Rehabilitation Center (EVRC) has been chosen as a site for a clinical trial research study to evaluate the safety and efficacy of the BrainPort® vision device in subjects who are blind. The EVRC is one of seven sites across the nation, selected by Wicab, Inc., the study sponsor, to participate in this clinical trial research study. This 12-month study allows subjects to use the device at home after completion of initial clinic screening and training.

The BrainPort® vision system consists of a postage-stamp-size electrode that is placed on the tongue, a base unit, a digital video camera and a hand-held controller for zoom and contrast inversion. Visual information is collected from the user-adjustable head-mounted camera and sent to the BrainPort® base unit. The base unit translates the visual information into a stimulated pattern that is "displayed" on the tongue. The tactile image is created by presenting white pixels from the camera as strong stimulation, black pixels as no stimulation, and gray levels as medium levels of stimulation, with the ability to invert contrast when appropriate. Users often report the sensation as pictures that are painted on the tongue with Champagne bubbles.

Thus far, study participants have been able to recognize high-contrast objects, their location, movement, and some aspects of perspective and depth. Trained participants use information from the tongue "display" to augment their understanding of their immediate environment.

Eligible subjects must be between the ages of 18 and 79 years of age, and have had a medical diagnosis of blindness for at least six months. Blindness may not be a result of cortical injury, such as a traumatic brain injury or stroke. Subjects must have completed rehabilitation (such as orientation and mobility training with a white cane or guide dog). Previous use of the BrainPort® vision device, pregnancy, regular tobacco use and allergies to nickel or steel exclude participation in this study. Participants should be able to easily commute to and from downtown Wichita, and are required to make four quarterly clinic visits in addition following their initial training sessions.

Potential study participants for this clinical trial should be referred to <http://vision.wicab.com/index.php>, and contact Shannon Riley, Research & Analytics Associate, Envision Foundation, at (316) 440-1528 or email shannon.riley@envisionus.com.

2012 Visibility Guide for Authors

SUBMISSION CATEGORIES

Lead Article

Theory, themed focus, or general article highlighting relevant issues facing the vision rehabilitation field.

Length: Approximately 1,500 words

Figures and Tables: May include up to 2 photos, figures or tables

Disease Etiology or Diagnosis

General article outlining new findings in the diagnosis and etiology of vision-related diseases.

Length: Approximately 1,500 words

Figures and tables: May include up to 2 photos, figures or tables

Case Study

Presentation of a patient case study (HIPAA compliant, de-identified patient information) that shows how a presenting problem was approached with vision rehabilitation interventions, follow-up and/or referrals.

Length: Approximately 1,000 words

Figures and Tables: May include up to 3 photos, figures or tables

Research

Research Review: Review of past and/or current vision research and its potential for vision rehabilitation clinical applications.

Research Abstract: An early concept, short research abstract that encompasses the exploratory stages of vision research and its potential for vision rehabilitation clinical interventions, technology applications, or surgical, pharmaceutical or gene therapy interventions. Include hypothesis, purpose or objective, research methods, results/expected results, discussion, conclusion, and future directions of research and acknowledgement.

Human Subjects Research: IRB and HIPAA compliant, de-identified patient information. If animals are used in study, the acknowledgement section should describe the animal care protocol that was followed, name the institution that sponsored the study and identify relevant IRB approval. Biomedical research involving animals must conform to generally accepted principles of animal maintenance and care, such as those of the Association for Research in Vision and Ophthalmology (ARVO).

Length: Approximately 1,000 words

Figures and Tables: May include up to 4 photos, figures or tables

Product Advertorial

Article highlighting a company, product or service of significance to the vision rehabilitation and research fields. Each issue of *Visibility* will publish only one advertorial. Advertorials are sold on a first come, first served basis. All advertorials will be marked as paid advertisements.

Length: Up to 800 words

Figures and Tables: May include up to 2 photos, figures or tables

Price: \$1,000

SUBMISSION DEADLINES

Issue Number	Submissions and Advertorial Deadline	Issue Publish Date
Volume 6, Issue 1	11/18/2011	2/3/2012
Volume 6, Issue 2	2/17/2012	5/4/2012
Volume 6, Issue 3	5/18/2012	8/3/2012
Volume 6, Issue 4	8/10/2012	11/9/2012

For more information regarding submission requirements, visit www.envisionus.com/visibility.

ENVISION CONFERENCE 2011: *Resounding Success!*

Thank You to everyone that made Envision Conference 2011 a resounding success for vision rehabilitation and research! Many hours of planning and executing went into designing the leading venue for low vision clinical education and research.

Nearly 300 low vision clinicians, researchers, educators and advocates gathered at the Hilton St. Louis at the Ballpark, St. Louis, Missouri, for the sixth annual Envision Conference September 21-24. Fifty-six clinical education sessions and 10 low vision research sessions were presented by leaders in the field. A total of 130 hours of continuing education units were offered representing the multidisciplinary nature of the conference: ACCME, ACVREP, AOTA, COPE and CRCC.

“This conference is unique in its focus on low vision. The combination of research and workshop platforms is critical to those of us in the teaching field who are required to use information and strategies that are ‘research based’.”

– Envision Conference
2011 Attendee

The focus of Envision Conference 2011 was **“Excellence in Research.”** The plenary **“Excellence in Research”** Keynote was delivered by Gary Rubin, PhD, FARVO. Dr. Rubin is a

Gold Fellow of the Association for Research in Vision and Ophthalmology and an Honorary Fellow of the College of Optometrists in the UK. In addition to publishing more than 100 scientific papers and book chapters, some of his more noted research includes reading and face recognition in people with impaired vision, a study sponsored by the National Eye Institute. He has also researched the effect of visual impairment on older people’s daily lives, a study sponsored by the National Institute on Aging. Rubin has devoted considerable time to the development and validation of new clinical vision tests used in a wide range of eye diseases including cataract, macular degeneration and diseases of the optic nerve. In addition to being this year’s keynote speaker, Dr. Rubin also moderated a vision research symposium titled “Clinical Trials for Low Vision Rehabilitation: Interventions & Methodologies.”

EXCEPTIONAL SPEAKERS

Many thanks are also due to the skilled presenters in clinical education and low vision research who



David Lewerenz, OD, presented three clinical education sessions and one hands-on workshop.

helped to make this year’s program the best to-date. Additional thanks go to the Envision peer review committees who reviewed the hundreds of clinical education and research abstracts that ultimately lead to the first-rate program. For a review of this year’s program, see the “Previous Conferences” page at www.envisionconference.org.

continued on page 20

“This was my first experience with Envision. It was a very rewarding experience and will help me in my efforts to provide the best low vision services to my patients. The team effort of disciplines was embraced by this conference and it was a great opportunity for networking and professional growth! The Envision Conference will be on my list of ‘must attend’ conferences!”

– Shirley Anderson, OTR/L, CLVT, Envision Conference 2011 Attendee

ENVISION CONFERENCE 2011 RESEARCH SESSION HIGHLIGHTS

Envision Conference 2011 took place in St. Louis, Missouri, September 21-24 with pre-conference educational workshops taking place on Wednesday, September 21. The Envision Conference is one of the largest and most respected gatherings of multi-disciplinary professionals engaged in clinical practice and research related to low vision and blind rehabilitation. The presentation of eight pre-conference workshops, 10 research panel sessions and 56 clinical education sessions ensured that there were topics of interest for all professionals present. Below are just some of the highlights from this year's research sessions.

WEDNESDAY, SEPTEMBER 21

Wednesday provided intensive workshops on a number of diverse topics for those arriving early to the conference. The first of the workshops presented was *Workshop on Prism Adaptation Therapy for Hemispatial Neglect Associated With Brain Injury or Stroke*, presented by Kevin Houston, OD, and Kia Eldred, OD. The complex relationship between diabetes and vision rehabilitation was addressed by Emilie Hagan, ARNP-CNS, MS, CDE, William Park, OD, and Karen Kendrick, OTR/L, CLVT, in *Diabetes and the Low Vision Rehabilitation Team: The Great Escape*, while *Adding Low Vision Services in Your Private Practice* was presented by Kendall Krug, OD, and Lori Grover, OD. Five other diverse workshops completed the day's activities.

THURSDAY, SEPTEMBER 22

The first day of concurrent sessions for Envision Conference 2011 got off to a great start with research *Health States of Patients Seeking Outpatient Low Vision Rehabilitation Services Within LOVRNET*, moderated by Judith Goldstein, OD. This session provided presentations on the health states of nearly 800 patients within the Low Vision Research Network (LOVRNET), including **The Relationship Between Physical Ability and Functional Ability in Patients Seeking Outpatient Services Within the Low Vision Research Network (LOVRNET)**, by Kim Schoessow, OTD, **Functional Ability and Depressed Mood Among Patients in the Low Vision Research Network (LOVRNET)** by Judith Goldstein, OD, and **The Measurement of Personality and Social Support Systems Among Patients in the Low Vision Research Network (LOVRNET)** by K. Bradley Kehler, OD. In the Afternoon, *Trouble Seeing... Trouble Sleeping: How Are They Related?*, moderated

by Olga Overbury, PhD, presented research on the timely and previously unconsidered issues related to the relationships between sleep disorder and visual impairment. A few of the presentations included **Eyes Wide Shut: Lens Properties and Sleep Disturbances**, presented by Walter Wittich, PhD, and **Prevalence of Circadian Rhythm Disorders Among Blind People With and Without Light Perception**, presented by Erin Flynn-Evans, PhD. Presentations that also generated a great deal of interest were found in *Dual Sensory Impairment*, moderated by Walter Wittich, PhD. These included **The Relevance of Providing Both Auditory and Visual Speech Cues to Older Adults With Dual Sensory Loss Impairments** by Jean-Pierre Gagne, PhD, and **Psychosocial Adjustment for Persons Aging With Hearing and Vision Loss**, presented by B.J. LeJeune, CRC, CVRT.

FRIDAY, SEPTEMBER 23

On Friday, the high quality of research sessions continued, beginning with research *Mobility Panel* moderated by Shirin Hassan, OD, PhD. Included in this session was a very interesting presentation entitled, **Enhanced Orientation and Mobility in the Visually Impaired Using Virtual Audio-Based Environments**, by Erin Connors, and a pair of presentations by Pradeep Ramulu, MD, MHS, PhD: **Restriction of Travel Outside the Home in Glaucoma and AMD: Direct Measurement Using a Cellular Network-Based Tracking Device** and **Objective Measurement of Real-world Physical Activity in Glaucoma and Macular Degeneration Using Accelerometer Devices**.

Gary Rubin, PhD, this year's winner of the Envision Award in Low Vision Research, moderated *Difficulties in Designing Clinical Trials: Finding Solutions* with

presentations by Alan Morse, JD, PhD: **The Efficacy of Clinical Trials in the Context of Public Policy**; Joan Stelmack, OD: **Recruitment and Retention in Clinical Trials**; and Graham Strong, OD, MSc: **Abandonment of Assistive Devices at the Completion of a Clinical Trial**.

In the afternoon, Ronald Schuchard, PhD, moderated *Functional and Performance Outcomes*. A few of the presentations included **Impact of Glaucoma on Activity Participation**, by Monica Perlmutter, OT, CLVT, and Anjali Bhorade, MD, **Key Life Stages and Transition Points for Young People With Sensory Impairments Between Birth and Age 25 Years**, by Carolyn Palmer, PhD, and **Increased Visual Field Variability in Newly Diagnosed Glaucoma Patients is Partly Related to Increased Depressive Symptoms or Worry About Blindness**, by Eva Bittner, OD, PhD.

Friday closed out, with an applied research session *Research on Employment*, moderated by Deborah Gold, PhD. Presentations addressed varied topics related to employment of people with visual impairments, including **Pre-employment Skills for Children Aged 5-14 With Vision Loss: An Evidence-based Model for Resource Development**, presented by Biljana Zuvela, MA; **Transition Services that Lead to Competitive Employment Outcomes for Transition-age Individuals With Blindness or Visual Impairments**, presented by B.J. LeJeune, CRC, CVRT; and **Supervising People Who Are Blind or Visually Impaired: A Qualitative Study of Employer Attitudes**, presented by Deborah Gold, PhD.

SATURDAY, SEPTEMBER 24

The excellent research sessions on the last day of Envision Conference 2011 provided plenty of reason to stay to the end, as some of the most intense interest and animated discussions took place on Saturday. *Low Vision Research: Hot Off the Press*, presented late-breaking low vision research findings. Moderated by George Timberlake, PhD, and Ronald Schuchard, PhD, this session provided an early start to the last day of the conference with presentations on **Barriers to Vision Rehabilitation: The Montreal Study**, by Olga Overbury, PhD, and Walter Wittich, PhD; **Does Improving Vision Reduce the Risk of Falls? A Review**, by

J. Vernon Odom, PhD; **Effect of Bilateral Macular Scotomas From AMD on Reach-to-Grasp Hand Movement**, by George Timberlake, PhD; and **A Test of Face Discrimination Ability in Aging and Vision Loss**, by Ronald Schuchard, PhD.

The research session *Diagnostics and Assistive Technology*, moderated by Robert Massof, PhD, was next and included presentations by Joshua Pratt, OD: **The Retinal Locus for Reading: What We Can Learn From Text Location on the Retina Just Before Word Enunciation**; Maria Enzo Vingolo, MD, PhD and Serena Salvatore, MD: **Evaluation of Fixation Stability by Bivariate Contour Ellipse Area (BCEA) Using the MP-1 Microperimeter in Mild Visually Impaired Patients**; and Lisa Mauney, MS: **Developing a Model to Predict Low Vision Users' Performance When Operating Small Visual Displays**.

Low Vision and Psychological Functioning, moderated by Laura Dreer, PhD, closed out the day and another successful Envision Conference, with research on the impact of low vision on various aspects of psychological functioning. Presentations included **The Impact of Depression on the Actual and Perceived Effects of Reading**, by Patricia Grant, MS, **Vision Impairment: The Impact on Social Cognition and Social Ability**, by Carolyn Palmer, PhD, and **On-Road Driving Performance of Moderate/Advanced Glaucoma Patients**, by Anajali Bhorade, MD.

As can be seen from these brief highlights, the quality, relevance, variety and number of this year's research sessions were the best to date, and next year promises to be even better as Envision Conference 2012 returns to St. Louis, Missouri, on September 12-15. Make your plans to attend and present at this one-of-a-kind growing, multi-disciplinary, low vision research event.

Submissions for clinical education and research sessions are open now through March 19, 2012 via the online submissions form.

Visit www.envisionconference.org to register and submit.

Envision Conference 2011: Resounding Success! continued from page 17



ONE-STOP-SHOP FOR LOW VISION RESOURCES AND SERVICES

Thanks to our exhibitors and sponsors, the Envision Conference 2011 Exhibit Hall was the place to be for special events and breaks. Leading-edge low vision technologies were on display for attendees to experience and add to their practices.

Ready Yourself for Envision Conference 2012. Registration for attendees, exhibitors and sponsors is already underway as we make plans to return to St. Louis, Missouri, **September 12-15, 2012**. See you in St. Louis for Envision Conference 2012!

“This is the only annual meeting where low vision researchers and clinicians from all disciplines can come together and spend four days discussing low vision rehabilitation as committed professionals, without time pressure, distractions or competing agendas. It is a must-attend meeting for me.”

– Robert W. Massof, PhD, Envision Conference 2011 Attendee

Important Dates

March 19, 2012

Deadline for Clinical Education and Research Submissions

June 29, 2012

Deadline for Early Bird Registration

July 6, 2012

Deadline for Advance Price Exhibitor Registration

August 17, 2012

Hotel Room Block Deadline

September 12-15, 2012

Envision Conference 2012 at the Hilton St. Louis at the Ballpark

To register, or learn more about Envision Conference, visit the website at **www.envisionconference.org** or contact Michael Epp, Director, Professional Education, at **michael.epp@envisionus.com**. For updates from Envision Conference, follow us on **Twitter (@EnvisionConf)** or find us on **Facebook**.

Envision Professional Education Calendar

January 12, 2012

Low Vision Grand Rounds – New Treatment Strategy (Bevacizumab) for Advanced Retinopathy of Prematurity. Wichita, KS. CE – ACCME, AOTA, COPE

March 15, 2012

Early Intervention and the Role of Occupational Therapy. Wichita, KS. CE – AOTA

April 12, 2012

Low Vision Grand Rounds – Update on AMD. Wichita, KS. CE – ACCME, AOTA, COPE

June 15, 2012

Neurological Vision Loss. Wichita, KS. CE – AOTA, COPE

July 12, 2012

Low Vision Grand Rounds – What's New and Exciting in Corneal Surgery. Wichita, KS. CE – ACCME, AOTA, COPE

September 12-15, 2012

Envision Conference 2012. Hilton St. Louis at the Ballpark, St. Louis, MO. CE – ACCME, ACVREP, AOTA, CRCC, COPE

October 11, 2012

Low Vision Grand Rounds – Research on Contact Lenses. Wichita, KS. CE – ACCME, AOTA, COPE

November 9, 2012

Assistive Technology. Wichita, KS. CE – ACVREP, AOTA, CRCC, COPE

For more information, visit the Education and Resources page at www.envisionus.com.