Congenital or Acquired Color Vision Defect – If Acquired What Caused it?

Terrace L. Waggoner O.D.
3730 Tiger Point Blvd.
Gulf Breeze, FL 32563
waggonert@aol.com

Course Handout AOA 2017 Conference:

I. Different types of congenital colorblindness.
   A. Approximately 8% of the population has a congenital color vision deficiency.
   B. Dichromate
      1. Protanopia is a severe type of color vision deficiency caused by the complete absence of red retinal photoreceptors (L cone absent).
      2. Deuteranopia is a type of color vision deficiency where the green photoreceptors are absent (M cone absent).
      3. Tritanopia is a very rare color vision disturbance in which there are only two cone pigments present and a total absence of blue retinal receptors (S cone absent). It is related to chromosome 7.
   C. Anomalous Trichromate
      1. Protanomaly is a mild color vision defect in which an altered spectral sensitivity of red retinal receptors (closer to green receptor response) results in poor red–green hue discrimination.
      2. Deuteranomaly, caused by a similar shift in the green retinal receptors, is by far the most common type of color vision deficiency, mildly affecting red–green hue discrimination in 5% males.
      3. Tritanomaly is a rare, hereditary color vision deficiency affecting blue–green and yellow–red/pink hue discrimination.
   D. Rates of color blindness
      1. Dichromacy Males 2.4% Females .03%
         a. Protanopia (red deficient: L cone absent) Males 1.3% Females 0.02%
         b. Deuteranopia (green deficient: M cone absent) Males 1.2% Females 0.01
         c. Tritanopia (blue deficient: S cone absent) Males 0.001% Females 0.03%
      2. Anomalous trichromacy
         a. Protanomaly (red deficient: L cone defect) Male 1.3% Female 0.02%
         b. Deuteranomaly (green deficient: M cone defect) Male 5.0% Female 0.35%
“Deuteranomaly” is the most common color vision deficiency.
c. Tritanomaly (blue deficient: S cone defect) Male 0.0001% Female 0.0001%

II. Approximately 5% of the population has an acquired colorblindness.

A. Characteristics of congenital versus acquired color vision deficiencies

<table>
<thead>
<tr>
<th>Congenital</th>
<th>Acquired</th>
</tr>
</thead>
<tbody>
<tr>
<td>1. Present at birth</td>
<td>Onset after birth – previous color vision normal</td>
</tr>
<tr>
<td>2. Type and severity remains the same</td>
<td>Type and severity can change with time</td>
</tr>
<tr>
<td>3. Both eyes equally affected</td>
<td>Frequent monocular differences in severity</td>
</tr>
<tr>
<td>4. Visual acuity and visual field normal</td>
<td>Reduced visual acuity and/or visual field defects</td>
</tr>
<tr>
<td>5. Normally a healthy individual</td>
<td>Possible associated disease process – i.e. macular degeneration, multiple sclerosis, glaucoma, etc.</td>
</tr>
</tbody>
</table>

III. Case Studies

A. A 36 year old white male helicopter pilot. He failed the visual acuity section on his annual flight physical. He was referred to optometry. Optic nerve atrophy was found in his left eye. Optical Coherence Tomography showed nerve fiber thinning and enlarged cupping in the left eye. There was a 2+ relative afferent pupil defect in his left eye. Visual field testing, using a Humphrey Visual Field Analyzer, was outside normal limits in the left eye. MRI indicated a suspected hemangioma of the left optic nerve. Color vision testing, using a combination of the Waggoner Computerized Color Vision Test and the D-15 diagnosed a severe deutan and tritan defect in his left eye. All findings, including color vision, were normal in his right eye.

1. Color vision testing was helpful and supported the diagnosis optic nerve atrophy.
2. Based on the characteristics of congenital versus acquired color vision deficiencies, the color vision deficiency in his left eye is most likely acquired.
3. CPT code 92283 describes extended color vision testing in this case.
4. Coverage/Reimbursement by Medicare and other payers for color vision testing depends on the indications/justification as well as the results of the testing and the doctor’s interpretation.
5. You must provide good documentation that supports reimbursement for color vision testing.
   a. Physician’s order
   b. Reliability of extended findings
d. Assessment, diagnosis
e. Impact on treatment prognosis
F. Doctors signature.

B. A 33 year old pilot just finished his physical and was found to be in perfect health. It was noted, using the Waggoner Computerized Color Vision Test in combination with the Richmond HRR that he had a tritan (blue) color vision defect. The defect was equal in both eyes and appeared unchanged over a 2 year period. His red/green color vision was normal.

1. Based on the characteristics of congenital versus acquired color vision deficiencies, is his tritan color vision defect most likely congenital or acquired?
2. It is recommended that vision care professionals obtain a baseline color vision test on all patients. It is important to know when counseling career choices and diagnosing acquired defects.

IV. Popular book color vision tests used to test for both congenital and acquired colorblindness: pseudoisochromatic plate color vision tests (PIP); color arrangement tests; and cone color and contrast sensitivity color vision tests.

A. Pseudoisochromatic Plate color vision tests: i.e. the Ishihara, PIP 24 Plate Color Vision Test, and Divorine.
   1. The PIP test plates are made-up of confusion colors
   2. The Ishihara with trails and Color Vision Testing Made Easy with symbols (circle, star, and square) are two pediatric PIP color vision tests.

B. Color arrangement tests: The Munsell-Hue D-15 and Munsell-Hue 100

C. Cone color and contrast sensitivity color vision tests: The Waggoner HRR and Richmond HRR indicate the type and severity of a color vision defect.

D. The Nagel Anomaloscope is currently the gold standard when testing for a red/green color vision deficiency.

V. Advances in computerized color vision testing – the new paradigm

A. Three validated standalone computerized color vision tests on the market – the Computerized Color Vision Test (CCVT) by Dr. Waggoner, Colour Assessment & Diagnosis Color Vision Test (CAD) by Dr. Barbur, and the Cone Contrast Test (CCT) by Dr. Rabin.
   1. Based on cone color and contrast sensitivity.
   2. Indicate type and severity
   3. Detect both congenital and acquired color vision deficiencies.
   5. Monitor changes in the degree of the defect
   6. Save information on your computer.
   7. Self-administered.
8. Studies indicate computerized color vision tests have slightly better sensitivity and specificity than current book color vision tests.

VI Common issues for your congenital colorblind patients and coping mechanisms recommended by my son who is colorblind.

A. Issue: Is that traffic light red or yellow? Is that a blinking yellow or red light?  
Coping Mechanism: Most people use the location of the light. You will have to memorize that the red light is at the top, the yellow light is in the middle, and the green light is at the bottom. There are also horizontal traffic lights and that means that the red light is on the left, the yellow light is in the middle, and the green light is on the right. Driving at night is a different story. More than likely you will have to approach the light cautiously until you can tell where the light is located, which you should already be doing because it is either red or yellow.

B. Issue: Identifying sunburn, rash, etc.  
Coping Mechanism: First of all, when pertaining to the sunburn, it is recommended that you apply sunscreen before going into the sun for long periods of time. Now let's be rational, not everyone wants to put sunscreen on and they want a little tan. A technique I use is applying pressure to my skin where I believe I may be getting sun with two fingers and seeing if it appears pale after removing my fingers. If it does, more than likely you are getting sunburned. Second, ask someone that is nearby. I'm sure they would be more than willing to help. Concerning rashes and other skin irritations, I recommend getting a close friend or relative to tell you.

C. Issue: A friend says to locate something by color such as, "Hey, look at that red berry on the tree."  
Coping Mechanism: Ask them to point in the direction so you get a better sense of its location. Then second, ask them to describe other characteristics about the object such as size and shape.

D. Issue: Going to buy clothes and dressing yourself.  
Coping Mechanism: Ask a close friend or relative to go shopping with you or you can ask the employees of the retail store to help you. When dressing yourself, ask friends and family if an outfit matches. Once you find an outfit that matches, I recommend wearing the same outfit in the future. Trying to match things by oneself is difficult if not impossible.

E. Issue: Cooking food that needs to be cooked throughout.  
Coping Mechanism: Review a good source on how long something should cook and then use a timer. If there is a friend around, ask them if the meat is thoroughly cooked. Using
a meat thermometer is a safe method as most tell you the temperature most meats are done.

VII Helpful advise you can give your colorblind patients who are students.
   A. First of all, do not be embarrassed to tell your teacher that you are colorblind. Once you get that out of the way, then he/she will understand if there are mistakes pertaining to color and you will not be docked points. So remember, tell your teacher immediately that you are colorblind.
   B. If You need to color something in with specific colors and instructions (E.g. maps). Purchase coloring utensils that are labeled. Don't assume you know the color because you probably don't and you will more than likely lose points.
   C. In some science classes you may be asked to document chemical reactions and other color specific tasks. You have to have an observational, color normal lab partner, otherwise, this is a very difficult, if not impossible task. Also, be sure to remind your teacher that you are colorblind if you are by yourself.
   D. When dressing in the morning, ask your parents for their opinion on what matches. Remember what outfits match each other and just continuously wear them together. When you begin mixing and matching, more than likely you are not matching just mixing. Who wants to be teased?
   E. Let the teacher know that you have difficulties with certain colors on the board and ask if she can change to something that is easier for you to see. Ask your teacher if you can sit by a friend who can help you if you can't see something because of the color contrast. I suggest asking the teacher if she will use white chalk on a chalkboard.

VIII Inform your colorblind patients about these helpful Links/apps
   B. Excellent 9 minute film on colorblindness by Laura Evans. http://nosuchthingascolor.com/
   C. A shareware program for colorblind individuals called "WhatColor". Using your cursor, you can identify any color on the screen of your PC. http://www.hikarun.com/e/
   D. You will see how a webpage looks to a color deficient person. https://www.toptal.com/designers/colorfilter
   E. See how color deficient people see colored palettes http://www.iamcal.com/misc/colors/
F. The goal of this page is to help web designers in choosing colors that are correctly visible by color blind people. http://gmazzocato.altervista.org/colorwheel/wheel.php

G. The Laxmi Eye Institute gives one the best educational slide presentations on colorblindness I have reviewed. https://www.slideshare.net/laxmiewatkins/colour-vision-and-its-clinical-aspects?next_slideshow=2


I. Color vision test you can take on-line. http://colorvisiontesting.com/online%20test.htm

J. Color Grab – simply point your smart phone at a colored object and it will tell you the color.

K. Enchroma Lens for colorblindness

IX. Ongoing color vision research and validation studies.
   A. Jay Nietz using gene therapy to cure colorblindness
      1. Injecting a virus inside two monkeys' eyes changed their genes.
   B. Department of the Navy’s research
      1. NAMRU Dayton, OH did a study called “Assessment of Color Vision Screening Tests for the U.S. Navy Special Duty Occupations“ which tested the reaction time and accuracy of CVD subjects compared to color normal subjects.
         a. PAPI and Missile Targets were used in the study
         b. The type and degree of CVD was determined using three computerized color vision tests
         c. Conclusion of the NAMRU Study - Mild CVD subjects performed nearly as well as those with normal color vision. ‘Moderate’ / ‘Severe’ CVD subjects had a slower reaction time and made significantly more mistakes.
      2. Validation Studies by the Naval Aerospace Medical Institute Pensacola, Florida
         a. Validated the Waggoner Computerized Color Vision Test
         b. Conclusion: Uncalibrated Surface Pro 3 Tablet and calibrated PC have similar high sensitivity (98%-100%) for detecting red/green CVD’s as compared to Nagel Anomaloscope

X. Performance based testing of a color vision deficient pilot
   A. The subject failed the PIP and the Farnsworth Lantern during his annual flight physical. Prior testing at the Naval Aerospace Medical Institute indicated he had a color vision deficiency (Protanomaly), but qualified for pilot training because he passed the Farnsworth Lantern. NAMI required that the subject had to pass the PIP or Farnsworth Lantern every time on “all and any” future aviation exams. Seeing he failed both CV tests, he was grounded until the condition could be accessed and his performance evaluated.
   B. The subject’s ability to recognize various colors in different flight situations and environments was tested.
C. As a control group, two pilots were selected at random to also take part in the testing/evaluation. The control group’s results were recorded for comparative purposes.

D. In order to produce an objective assessment, medical professionals and pilot instructors were required to take part in the process. Two of the medical professionals were Navy Flight Surgeons and one of the pilot instructors was a Flight Standardization Instructor.

E. The subject had to quickly identify and describe various items in the AH-1Z Simulator (SIM). The UH-1Y SIM was also used during testing because it was the type/model/series aircraft the pilot routinely flew. The test items included:

1. External:
   a. ALDIS Lamp Signals
   b. Smoke markers
   c. Ship deck lights
   d. Runway lighting

2. Internal (Cockpit):
   a. System gauges
   b. Warning/caution/advisory indications
   c. Moving maps (terrain banding, overlays, etc.)

3. Classroom:
   a. Items on paper maps
   b. Hazards/obstacles
   c. Airspace coordination area
   d. No fire area
   e. Route markings

F. UH-1Y SIM:
   1. Subject was 90% accurate on identifying red coloration on gauges. Control group was 100% accurate. Test was out of 10 items.
   2. Subject was 20% accurate on identifying red objects on the FLIR imagery. Control group was 100% accurate. Test was out of 5 items.
   3. Subject was 38% accurate on identifying red items on the moving map. Control group was 100% accurate. Test was out of 8 items.

G. AH-1Z SIM
   1. Subject was 60% accurate on identifying various colors of smoke. Control group was 100% accurate. Test was out of 5 colors.*
   2. Subject was 88% accurate on identifying various runway lights. Control group was 100% accurate. Test was out of 8 items.
   3. Subject was 33% accurate on identifying aircraft lighting. Control group was 100% accurate. Test was out of 3 items.
   4. Subject was 58% accurate on identifying various lights in the simulated boat environment. Control group was 100% accurate. Test was out of 18 items
   5. ALDIS Lamp signals: Subject was 33% on identifying various ALDIS Lamp signals. Control group was 100%. Test was out of 6 items.
6. Map markings: Subject was 82% accurate on identifying colors on a map. Control group was 100% accurate. Test was out of 17 items.

**Discussion**

1. The subject had difficulty not only accurately identifying the color red but he also had issues with greens, blues, yellows and oranges throughout the testing process. The subject would also frequently take longer to make a decision on identifying various colors, as if he was thinking about what the right answer should be. This delay was never evident with either of the control group pilots.

2. It was obvious that the subject couldn’t identify some “critical” color differences in air craft display windows or switches as well as environmental objects, markers, and map components/markings.

3. Although it is not possible to definitively state the risks based on the results, his condition may translate into slower visual identification of both internal cockpit cues and external environmental objects differentiated by color variations. Notably, he would likely experience increased difficulty with daytime target/object “talk-ons” that rely on environmental color variations. Also of concern is his ability to quickly identify hazards marked in red on a map.

4. The results of this assessment were provided to the Naval Aerospace Medical Institute for final disposition.