

## Lecture 31 – Acquired Color Anomalies

### REVIEW

Our ability to discriminate different colors is largely based on wavelength discrimination that begins with a **trichromatic** system of photoreceptors. The three cones types each contain a different photoreceptor with a different photopigment. Important characteristics include:

- Each photopigment has a different absorption spectrum, therefore each cone type has a different sensitivity spectrum.
- They are known as the S, M, and L cones, based on the location of their sensitivity spectrum peaks.
- Each photopigment absorbs light over a broad range of wavelengths.
- There is considerable overlap between the S, M and L cone sensitivity spectra.

After the photoreceptors, the signal from the different cones are combined, possibly by horizontal cells, into a complex neural network that includes two, or possibly three major pathways.

- A **red-green opponent system** that responds with inhibition at middle (green) wavelengths and excitation at long (red) wavelengths. This opponent network receives input from the M and L cones. (See Schwartz Fig. 5-14)
- A **blue-yellow opponent system** that is inhibited by short (blue) wavelengths and stimulated by middle-long (yellow) wavelengths. This system receives input from all three cones.
- A **non-opponent brightness system** that provides brightness information.

At the level of the ganglion cells and beyond, neural signals from the retina are carried along one of three major neural pathways, the **parvo, magno and konio pathways**. The parvo and konio pathways respond to chromatic (colored) stimuli in an opponent fashion, therefore they appear to be part of a system that gives us color perception. Some scientists theorize that the magnocellular system processes brightness information.

Q. How does the  $V(\lambda)$  function for a protanope differ from that of a normal trichromat?

A.

Q. How does the  $V(\lambda)$  function for a deuteranope differ from that of a normal trichromat?

A.

Q. Why are dichromats really monochromats above about 545 nm?

A.

Define the following:

- color confusion line
- copunctal point
- neutral point

### ACQUIRED COLOR ANOMALIES

Up till now we have been discussing hereditary color anomalies. Next we will consider acquired anomalies. Review Table 1, which lists important differences between hereditary and acquired color anomalies.

Acquired color vision anomalies caused by disease or toxicity can produce either red-green or blue-yellow anomalies. Among hereditary color anomalies, the red-green type are more common, so some people assume that if a person has a red-green anomaly, it is a hereditary, rather than acquired condition. It is important to understand that *not all red-green anomalies are hereditary*—in some cases patients have red-green defects that are caused by disease.

Since hereditary blue-yellow defects are so rare, you should assume that a patient with a blue-yellow anomaly has acquired it due to some disease.

In addition, anytime you detect a color vision anomaly that is asymmetric between the two eyes, you should assume it is acquired.

Q. Why should you always test color vision monocularly?

A.

Q. Which eye should you test first?

A.

**Table 1.** Comparison of hereditary and acquired color anomalies

<b>Hereditary</b>	<b>Acquired</b>
predominantly red-green	blue-yellow or red-green
predominantly males	male or female
color naming errors rare	recent color naming errors
stable with time	variable or progressive
diagnosis & classification clear-cut	may be difficult to diagnose & classify
no associated disease	associated disease
binocular	monocular or asymmetric

**Table 2. Köllner's Rule**

	<b>Blue-yellow defects</b>	<b>Red-green defects</b>
<b>Diseases of the ...</b>	media, choroid, outer retina	optic nerve, inner retina
<b>Examples</b>	cataract, diabetes, RD, macular degeneration, chorioretinitis, central serous retinopathy	optic neuritis, papillitis, Leber's optic atrophy, toxic amblyopia, visual pathway lesions
<b>Exceptions</b>	glaucoma, papilledema	dominant cystoid macular dystrophy, Strargardt's disease (fundus flavimaculatus)

**Köllner's Rule**

Traditionally Köllner's Rule has been used to help diagnose diseases based on the type of color anomaly present, assuming it is acquired. It is summarized in Table 2 above.

This rule states that diseases of the

- outer retina (outer plexiform layer, receptors, RPE) and ocular media cause blue-yellow defects, while
- diseases of inner retina (ganglion cell layer, optic nerve) and visual pathways cause red-green defects.

In practice Köllner's rule is not so useful since there are exceptions and in some conditions one kind of anomaly can change into another. For example, the blue-yellow channels seem more susceptible, so in certain diseases those defects will appear first, but later the patient can develop red-green defects. See Table 6-4 in Schwartz.

Some diseases can cause a type of anomalous color perception known as **chromatopsia**. This is a condition in which vision appears to have a colored tint, as if the person were looking through tinted lenses. Color discrimination is normal, and this is different from any of the color anomalies we discussed previously (protan, deutan, tritan defects). Examples include:

- **Xanthopsia**, or yellow vision—a side effect of some medications
- **Cyanopsia** or blue vision. Cataracts often have a yellowish tinge since they strongly absorb blue light. Patients with cataracts become accustomed to vision with a reduced blue component, so after cataract surgery they will suddenly begin receiving more blue light, causing cyanopsia. This diminishes with time. This is also reported as side effect of some medications, including Viagra.
- **Erythropsia**, or red vision. Due to snow blindness or atropine use.
- **Chloropsia**, or green vision. Due to epinephrine, or lead poisoning.
- **Ianthinopsia**, or violet vision. Due to cannabis use.

### COLOR VISION IN OCULAR DIAGNOSIS SWAP

In glaucoma, retinal neurons are gradually damaged over time, but since the visual loss proceeds slowly, usually affecting peripheral neurons first, it is essentially a symptomless disease in the early stages. Because of this, early diagnosis is the key to managing glaucoma.

Optometrists use two general strategies to diagnose glaucoma and other diseases.

- They look for signs of disease or damage in the ocular tissues; for example an enlarged C/D ratio. The problem with this approach is that you usually cannot detect pathological changes in the retinal neurons until much damage has already occurred. New instruments such as the HRT (Heidelberg Retinal Tomography), GDx (nerve fiber layer analyzer), OCT (optical coherence tomograph) or RTA (retinal thickness analyzer) are allowing doctors to evaluate subtle changes in the retina that previously were too hard to see. Adaptive optics ophthalmoscopy may also improve diagnosis in this way.
- The other approach is to measure changes in visual function caused by the disease. The problem is, Which visual function should you test? Many different psychophysical tests can be done to evaluate vision, but you prefer to find one that is sensitive to early glaucomatous damage.

In the case of glaucoma, it seems that the S-cone/blue-yellow system is more susceptible to damage than the M and L/red-green system. Short wavelength automated perimetry (SWAP) was therefore developed to isolate and test the S-cone system. This is available as an option on the Humphrey Visual Field Analyzer. In SWAP, the background is yellow and is designed to reduce sensitivity of the M and L cones. A blue light is then projected onto the yellow background and the detection threshold is measured.

### Spectral increment threshold (Schwartz p. 135-6, Fig. 6-2)

Schwartz describes a psychophysical test that can be done to test the color-opponent parvocellular system (the chromatic system). A faint monochromatic (single wavelength) light is presented on a white background and its threshold for detection is measured. The threshold for many wavelengths is tested, then sensitivity (inverse of threshold) is plotted as a function of wavelength. In normal subjects, the function shows three peaks that correspond to the S, M and L cone contributions. The peak at 440 nm closely matches the peak sensitivity of the S cones, but the peaks at 520 and 620 correspond more with the negative and positive peaks of the red-green opponent systems, rather than the M and L cones peaks (535 and 565 respectively).

In diseases that affect the S cone system, the chromatic function may have a depressed short wavelength peak. Figure 6-2 in Schwartz shows that, in the case of the deuteranope, the middle peak is missing and in a protanope, the long wavelength peak is missing.

### Clinical color vision screening for neurological disease

Several simple clinical color vision tests, known as “red cap tests” have been designed to roughly test color vision. These are sometimes used to diagnose diseases such as optic neuritis. These tests simply

indicate that a person's color vision is abnormal. They cannot validate that the perception is normal, and in the case of a color anomaly, they do not diagnose the type or severity.

**Red cap test I** (interocular comparison)

- Use a red target such as a Tropicamide bottle cap.
- Have the patient view the cap monocularly with each eye and compare redness of the cap as seen by either eye.
- In the presence of optic nerve disease, the red cap will appear to be less saturated and dimmer to the affected eye.

*Conduction defects of the optic nerve, e.g., optic neuritis, typically reduce color perception. Color defects may precede or be disproportionately more advanced than changes in visual acuity. ... In response to the above test, the patient with reduced central acuity due to an optic nerve lesion usually reports that the colored object is dimmer than when viewed by the normal eye; that is, the color is desaturated or "washed out" and appears yellow, white or gray. In the absence of pathological change in the fundus, a positive response to this alternate-eye color comparison test is evidence of an optic nerve lesion. (Duane's Ophthalmology, Ch. 2, p. 4-5)*

**Red cap test II** (monocular central-peripheral)

- Use two Tropicamide bottles with red caps or other red targets. (Make sure they are exactly the same color.)
- Hold one cap centrally and the other slightly peripherally (10 degrees).
- Have the patient view the cap monocularly and compare the redness of the central and peripheral red caps.
- Normally the central cap will appear to be more saturated, or redder.

In diseases of the optic nerve, the peripheral cap may appear to be more saturated than the central cap.

**Red cap test III** (monocular nasal-temporal)

This test is used to detect temporal visual field defects secondary to chiasmal compression.

- Use two Tropicamide bottles with red caps or other red targets.
- Hold both bottles on either side of fixation (one nasal, one temporal field), while the patient looks monocularly.
- Ask the patient to compare the redness of the two caps.
- Normally they should appear to be the same redness.
- In the presence of a large visual field defect, the red cap located in the defective field should appear less saturated.

A failure on any of these tests should be followed with more formal color vision testing, visual fields and other tests.

Q. Would these tests be useful for detecting both R-G and B-Y acquired color anomalies?

A. No, a tritan should have nearly normal color perception for long wavelengths (red), so this probably would not detect a patient with an acquired B-Y defect. It would be useful for either a protan- or deutan-type anomaly.

Q. Why would you want to test for possible optic neuritis using a red object? (hint: Köllner's rule)

A. According to Köllner's rule optic nerve diseases can cause R-G anomalies, so it makes sense to test with a red object.