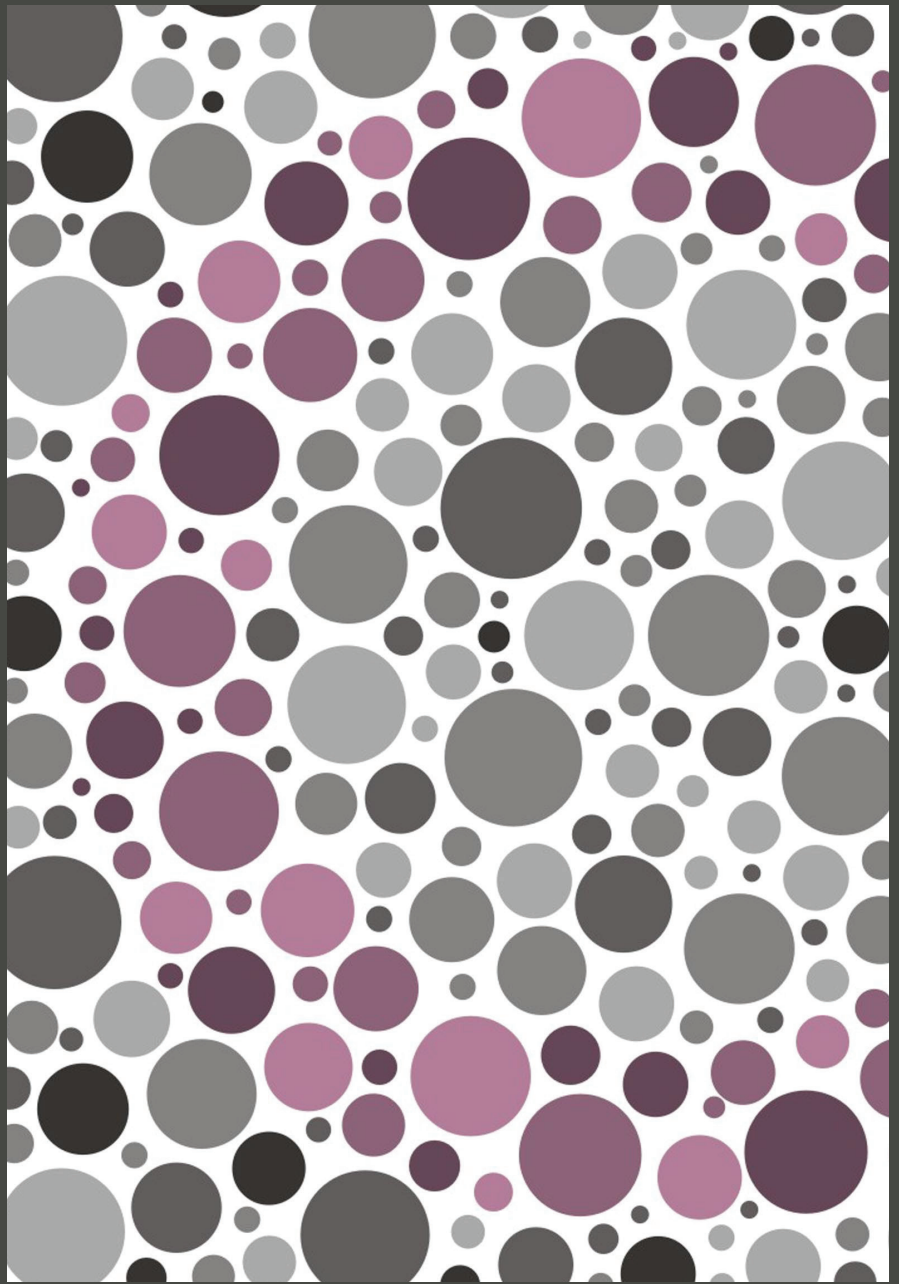


**RICHMOND PRODUCTS**



# Color Vision Deficiency

A Concise Tutorial for  
Optometry and Ophthalmology

# Color Vision Deficiency

## A Concise Tutorial for Optometry and Ophthalmology

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## INTRODUCTION

This tutorial is primarily intended for students of optometry and ophthalmology as a concise and easy-to-read pre-cursor to detailed textbooks and academic articles. The audience can include certified optometrists and ophthalmologists; pediatric and occupational MDs and RNs; allied health personnel in ophthalmology and optometry and pharmacists. The paper may be of interest to those involved in a wide range of occupations in industry, government, military and civil aviation and maritime functions. It aims to help fulfill the thirst for knowledge about color vision deficiency (CVD) and describes its causes and effects, methods of detecting and diagnosing the various forms, the effects of certain toxic substances and work (occupational) considerations. The final chapter deals with the current status of treatment and therapeutic methods.

This paper avoids using the term 'color blindness', despite the fact that it is fairly widely employed by professionals and lay persons. Among other (somewhat lengthier) definitions, CVD is, in fact, the inability to perceive certain colors in their true or 'natural' representations that results in confusion. This is explained in detail in the main part of this paper, with references from a number of eminent eye specialists who are better qualified to do so than the author. The clinical term for CVD is dyschromatopsia, which is used only by the medical profession.

The CVD Tutorial is the initiative of Richmond Products Inc. in Albuquerque, New Mexico. Richmond develops, manufactures and markets eye examination and test solutions to a broad user base throughout the world.

## FOREWORD

This tutorial helps to emphasize how sensitive and vulnerable our eyes are. Too often, as with our general health, our ability to see is taken for granted. Fortunately, we are continually reminded to take care of our eyes, as was recognized by eminent practitioners, physicists and scientists many years ago. Visual acuity and color vision have been studied as far back as the 17th century. In 1663 for example, Isaac Newton, known famously for defining the law of gravity, became fascinated with optics and started to experiment with prisms and lenses. This led, in the first instance, to discovering the color spectrum from a beam of sunlight, and in the second instance, to the development of the first telescopes. Although Newton observed that the spectrum comprises a continuous series of colors, he selected and named seven main hues – violet, indigo, blue, green, yellow, orange and red. His genius led him to postulate that the phenomenon of color vision originates from 'vibrations' set in motion at the lower area of the eye by incoming light. We know that this is not quite so, but Newton's theory kick-started a whole series of experiments, discoveries, theories and factual statements concerning the nature of color and color vision down through the ages that has helped us arrive where we are today. We now have extensive knowledge of color itself, light, and the physiology of the eye receiving color wavelength signals and how these signals are processed by the brain.

Optometry and ophthalmology are noble professions and, as also highlighted in this tutorial, there is an on-going need for research in both visual acuity and color vision, especially in more effective screening and testing. In less-developed countries (than, say, those in North America, Western Europe and others in Asia) there is a chronic shortage of trained eye physicians and nurses.

In a few years, there is the potential that my learned colleague Dr. Jay Neitz, with his team and other labs, may have success in finding a cure for congenital CVD through their research in gene therapy. This is briefly described at the end of this tutorial paper. Finally, because of the ongoing development of new prescription drugs, the importance of detection for acquired CVD will certainly increase.

I therefore wish you all, students, practitioners, nurses, and any others who choose or have chosen a career in optometry, ophthalmology, and occupational medicine, every success and encouragement. We need each and every one of you.

Dr. James D. Bailey, OD, PhD - Autumn 2010

## AUTHOR ACKNOWLEDGEMENTS

Special thanks go to Dr. Bernard Blais, MD, FACOEM, FAAO, FACS, Clinical Professor of Ophthalmology at Albany Medical College, Albany, New York. His book 'Color Vision in the Occupational Setting', along with his personal advice, have proven invaluable in preparing this paper. Dr Blais's book is recommended as further reading at the end of the tutorial. It is generally acknowledged as one of the foremost publications dealing with color vision deficiency in the workplace. In his foreword Dr. Blais says "Although there are many books on color vision, there are none that allow the user with minimal knowledge of color vision to be their 'cookbook' in the screening and ultimate diagnostic evaluation of (the) employees." His book is the exception.

Also acknowledged is the valuable help of the following in preparing this paper:

- Dr. James D. Bailey, OD, PhD, Professor, Department of Basic and Visual Sciences, Southern California College of Optometry, Fullerton, California.
- Dr. Frederick T. Fraunfelder, MD, Head of Ophthalmology at Oregon Health and Science University's Casey Eye Institute, Portland, Oregon.
- Dr. Jay Neitz, PhD and Dr. Maureen Neitz, PhD, respectively Bishop Professor and Hill Professor, Department of Ophthalmology at the University of Washington, Washington DC.

# CHAPTER 1 - Causes and Effects

## THE RETINA

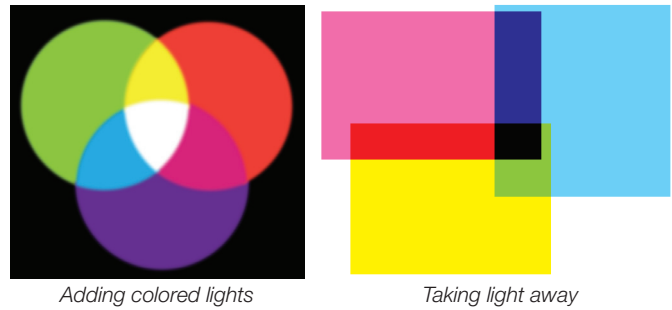
The retina of the human eye contains about 7 million cone cells and more than 100 million rod cells that enable normal vision. The majority of cone cells are located in the center of the retina. There are three sub-classes of cone cells, known as receptors I, II and III, each with photosensitivity to pigments of blue/yellow, green and red, respectively. These sensitivities result from absorption of different wavelengths of light reflecting from color pigments. Rod cells are mostly situated around the periphery of the retina and allow night vision at low light level, brightness perception and the ability to distinguish basic shape and form.

## COLOR VISION – TAKE NOTE

As with eyesight in general, color vision is taken for granted by the majority of people. The importance of correct or normal color vision should not be underestimated. Many times each day we use our color vision ability to discern and evaluate objects, signs, situations and other phenomena, often concerning matters of safety, work or pleasure, and observation in general. Use of color is considerably more prevalent in today's information-oriented environment with the advent of color computer displays and printers, for example. With age, color vision often deteriorates at a faster rate than visual acuity. Research in such matters shows that there is an increasing need for improvement in color vision standards and their acceptance internationally. Clearly, this indicates the need for further study, as well as more effective screening and test methods.

## DEFINING COLOR

Objects do not have color as a physical attribute. In fact, color is light, which is carried as specific wavelengths that the eye absorbs and the brain converts into 'messages' so that we 'see' colors. Pigments have the ability to absorb some colors and to reflect others. An object that appears blue actually absorbs all the other color wavelengths except blue. The unabsorbed wavelength is reflected back to the eye and the brain interprets the object as blue. Scientifically, 'color' refers to the sensory characteristic that different spectral dispersions produce. Somewhat surprisingly, although our color perception seems to be bright and lively, in reality we do not see the full scope of an object's spectral composition. In other words, our sense of 'color' of objects is both inaccurate and incomplete. In fact, 'natural' colors are those of an object's surface characteristics and how the surface reflects broadband (white) light and scatters it in the different parts of the spectrum. This also explains why colors cannot normally be seen at night or, at best, they are not the same as seen in daylight. Similarly, colors can vary in hue according to the degree of illumination, such as in strong sunlight or in shade. Artists are aware of the variations that influence their perception of color. In reality, we are all color deficient to a greater or lesser degree because our perception of color is somewhat limited and never 100 percent complete (see next paragraph).



<b>Primaries are red, green and blue</b>	
<b>Components</b>	<b>Result</b>
Red+Green+Blue	White
Red+Green	Yellow
Red+Blue	Magenta (bluish red)
Green+Blue	Cyan (greenish blue)

Figure 1 – Additive Color Mixing  
(Source: Blais, Page 14)

Why is this relevant to color vision deficiency? One reason is that even a person with so-called 'normal' vision does not always see or sense the detailed physical spectral differences of an object. For example, combining green light with red would produce yellow. Similarly, mixing blue with yellow will result in green (see Figure 1). In other words, our 'normal' or correct color vision is trichromatic, meaning that color can be represented in terms of quantities of three color wavelengths, known as primaries or chosen wavelengths, and can be made to appear as any spectral distribution (or any other color). The eye is 'fooled' into perceiving this. Anomalous trichromatic also refers to the ability to visualize three colors, but abnormally. This condition has three sub-categories – protanomalous, deuteranomalous and tritanomalous that refer to abnormal red-, green- and blue-sensitive colors respectively (*see Chapter 2 – Detection and Diagnosis*). Cases of anomalous trichromacy cannot sense color mixtures like those made by normal trichromats, nor, necessarily, the same matches made by other anomalous trichromats. A person who is dichromatic has difficulty discriminating between many spectral hues and lacks the ability to 'see' one of the three basic primaries of red, green and blue. They require two colors to produce a match for the full range of hues. The condition is most frequently congenital. Figure 2 illustrates the results of mixing colors together, where the resultant mixture of two colors always lies on the straight line joining them. For example, the mixture of 470 nm (blue) plus 580 nm (yellow) in the 'proper' amounts is indistinguishable from white. Following the examples above, a third category is monochromatic that refers to a person who has total color deficiency and can only see differences in light, and a single primary is sufficient to match all colors.. There are two types – typical (or rod) monochromatism applies to persons who lack functioning cone receptors; their visual acuity is poor and they have an aversion to bright light. The second type is cone monochromatism, which is extremely rare and most cases have only blue-sensitive cones. In most cases visual acuity is normal or slightly reduced.

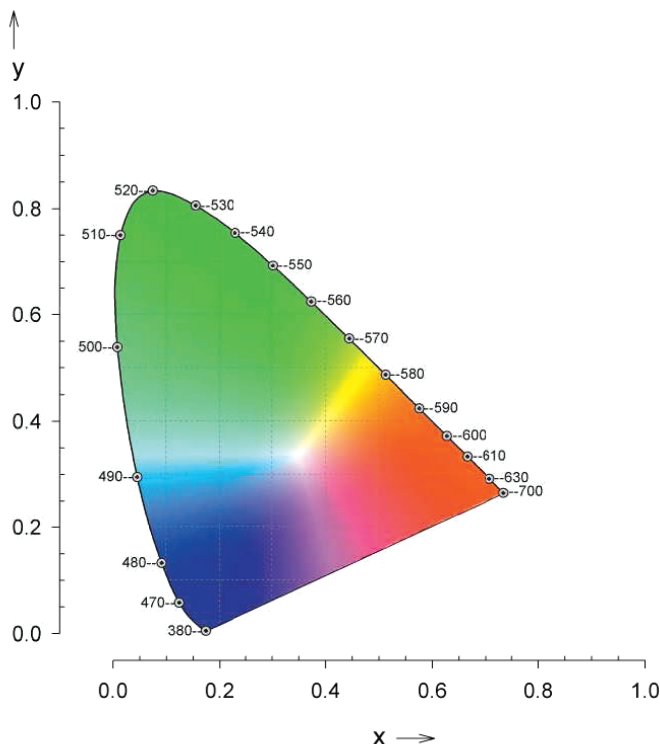


Figure 2 – Chromaticity Diagram showing the results of mixing two colors together. Pure colors, with their wavelengths (nanometers - nm), are situated around the edge of the figure. {Source: Psychophysical Measurement of Visual Function, Richmond Products, J. Bailey et al}

(See also Figure 7 – Chart of Dichromatic Confusion Lines)

## TWO TYPES OF CVD

There are two recognized types of color vision deficiency. Most cases are hereditary (congenital), while others are acquired, mainly caused by ocular or neurological disease, drug toxicity or exposure to certain solvents (see Chapter 3 - Toxicology Effects on CVD & their Detection ). In all cases, CVD in its various forms is the result of anomaly in one or more of the retinal cone color wavelengths that cause different sensitivities. Color differentiation results from comparison of activity of the cone photoreceptors by other neural processes in the retina and the brain. In normal color vision this can result in several hundreds of thousands of perceived variations in color. In congenital dichromatic CVD the number may be fewer than 100 color variations.

This information can comprise any combination of the three colors – red for long waveform sensitivity, green for medium and blue for short – resulting in up to about 17,000 perceptible variations or hues. Congenital CVD cases are almost exclusively in red/green discernment and are mostly binocular. It is important to note that persons with congenital CVD are not immune to acquired CVD. (See Figure 3 below, which shows that deutan is most prevalent).

Extent of Defect:	Type of Defect		
	Protan	Deutan	Tritan
Mild	Anomalous Protan	Anomalous Deutan	Anomalous
Medium	Anomalous Protan	Anomalous Deutan	Anomalous
Strong	Protanope	Deuteranope	Tritanope
Frequency	25%	75%	minimal

Figure 3 – Overview of Diagnoses based on CVD Test Results {Source: Richmond Products}

## CONGENITAL CVD

In the western world congenital CVD occurs in approximately eight percent of men and 0.4 percent of women. This equates to about 25 million males and 1.2 million females in the United States alone. In the rest of the world the percentages are somewhat lower, but the actual numbers are still significant and especially poignant since not many people may be aware that they suffer from CVD. Currently, there is no known 'cure' for congenital CVD (see Chapter 4 – Treatment, 'Compensation' & Cure).

The most common forms of congenital CVD are due to gender-linked X-chromosomes and hereditary characteristics. Males are mainly affected since they have only one X-chromosome and one Y-chromosome, while females have two X-chromosomes. If a male's single X-chromosome is color-defective he will be color vision deficient. For a female to be color vision deficient she must have inherited two color-defective X-chromosomes from either or both of her parents who are color vision defective, or that one or both is/are a carrier. Such situations are quite rare, which explains why females are less susceptible to CVD and that one in six females is a carrier of red/green CVD. The most common defect affects the photosensitivity of cones maximally responsive to the green region of the spectrum. Tritan CVD is neither an X-chromosome nor is it gender-linked. It is an autosomal trait whereby males and females are affected equally and the trait can appear in successive generations, whereas congenital CVD skips generations.

The frequency of congenital CVD cited above may not apply to all countries in the world.

The map in Figure 4 illustrates the different regional concentrations of congenital CVD occurrence in males. In fact, female prevalence is one 20th of male prevalence. Congenital CVD appears to be more prevalent in North America and Western Europe, as indicated by the red areas on the map. Non-Caucasian races may appear to experience lower incidence of congenital CVD but the exact causes of variations in prevalence are not known, nor yet fully understood, although they may be partly attributable to limited access to healthcare. For some areas of the world, testing and reporting of congenital CVD appears very sparse. Further, in some societies, the inheritance of congenital CVD is viewed as a social defect or stigma and is therefore underreported. Further research is warranted.

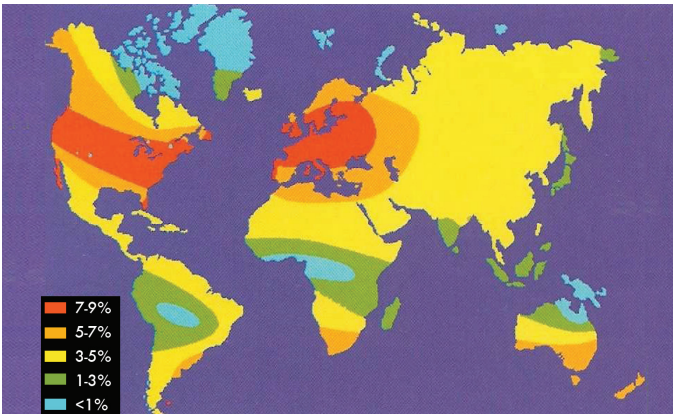


Figure 4 - Global Distribution of Congenital CVD in Males  
 (Source: Blais & Colour Blindness Causes and Effects,  
 D. McIntyre, MD, PhD)

### ACQUIRED CVD

Acquired CVD may occur at any age due to eye disease or lesions elsewhere in the visual pathways or processes. Due to greater incidence of eye disease as the population ages, acquired defects are more likely. Acquired defects occur monocularly at first and differs in this respect from congenital CVD. Some of the major causes of acquired CVD are listed below.

#### Disease

- Diabetes
- Cataract
- Macular degeneration
- Glaucoma
- Retinitis pigmentosa

#### Substance Toxicity\*

- Antibiotics
- Antidepressants
- Various other prescribed and non-prescribed medications
- Dietary supplements
- Chemical solvents

#### Trauma

- Eye or head injury

#### Neurological (optic nerve damage)

- Retinopathy
- Optic neuritis
- Neuropathy
- Lesions
- Ganglion cell

\*Chapter 3 goes into more detail on toxicology and the effects on CVD, highlighting many drugs and other substances. It is notable that substance toxicity effects appear to be on the increase. Chapter 4 discusses possible treatment of acquired CVD.

### OCCUPATIONAL (WORK-RELATED) ASPECTS

Good color vision is important in many occupations and, indeed in many everyday tasks – probably more than one would imagine. Defective color vision could be a risk factor or a serious handicap. In fact, the number of occupations relying on color is growing as more and more tasks are increasing in complexity, influenced by emerging technologies, and stricter operational regulations or work safety standards. The most obvious types of job functions where color vision is critical to the safety of personnel, or the environment, are those where color signage, warnings or coding are used on products, especially on hazardous substances, as well as road safety. Likewise, color lights are often indicators of position or situation. For example, traffic lights that are vertical, may be easily understood by a person with CVD based on the position that is 'lit', but sometimes traffic lights are set horizontally and the 'position' of each light may not be consistent. (See also 3rd paragraph on page 11, under THERAPEUTIC METHODS'). One practical example is the fire fighting service that uses color tags and codes to identify situations, fire fighting personnel, apparatus and gas and chemical detection levels.



Figure 5 – Color-Coded Control Valves on a Fire Truck  
 (Permission: City of Albuquerque Fire Department)

There are many more occupations for which color vision is critical. Most of these are shown in the table in Figure 6.

As a result, there is an increasing need for thorough color vision testing in many occupational applications and applying the results to a subject's job function, even though some functions could allow persons with some degree of color vision deficiency to carry out required tasks. In cases of congenital CVD it is usually only necessary to test once, since the condition generally does not change with age. With acquired CVD, however, the condition may develop or change over time, and therefore repeat testing is advisable. This is especially relevant if the worker is using one of the types of prescriptions listed as causing CVD or is exposed to potentially adverse/toxic environmental conditions (see Chapter 3 - Toxicology Effects on CVD & their Detection).

**Bernard R. Blais, MD, FACOEM, FAAO,  
FACS, Clinical Professor of Ophthalmology  
at Albany Medical College, Albany, New York**

### **A Further Word on Color Vision and its Relevance in the Workplace**

“In certain job functions, and depending on the nature of the business, an employee’s ability to discern certain colors can be critical. But how does a CVD employee perceive color and how safe is that person as well as his or her workplace environment? What of that employee who requires knowledge of colors and totally misinterprets the appearance?”

“No longer is it adequate to screen the patient and not interpret the findings, and apply them to a job description. Color vision happens to be one of the visual tasks that may be essential to fulfilling that job description. The perfection of the task related to required visual function in some positions might tolerate persons with severe color vision deficiency. Where congenital defects are concerned, an individual only needs to be tested once as the defect will not normally change from a congenital point of view but, perhaps, by occupation. On the other hand, acquired color vision defects may be monocular and occur at any time, especially if the cause is drug- or disease-related.”

“Outside the workplace many commuters may travel by metro and are required to read the various colored route lines at the stations. Also consider vehicle drivers who use GPS (Global Position Systems) devices. Proper color vision has no substitutions in these situations.”

“An important reminder to both employers and their employees – the American Disability Act of 1990 states that an individual must be able to perform the essential task with or without correction without significant risk or increased threat to the individual and the workplace.”

1. Aerial Photographer
2. Air Conditioner Installer/Mechanic
3. Air Traffic Controllers
4. Aircraft Navigator
5. Aircraft Pilot
6. Animal Code Enforcement
7. Applications/Quality Control Specialist
8. Art Teacher
9. Auto Body/Painting Technician
10. Biomedical Equipment Technician
11. Border Patrol Agent
12. Building Inspector
13. Building Plans Examiner
14. Bus Driver
15. Buyer for Department Store
16. CAD Operators
17. Carpet Sales
18. Civil Aviation Engineer
19. Civil Aviation Mechanic
20. Civil Service (Some positions)
21. Clinical Lab Technician
22. Clothing Sales
23. Color Photographer
24. Color Printing Press Operator
25. Commercial Printer
26. Communications Operator
27. Communications Technician
28. Computer Service Technician
29. Court Judicial Assistant
30. Court Reporter
31. Criminal Lab Technician
32. Customs Officer
33. Dairy Grader
34. DEA Special Agent
35. Dental Lab Technician
36. Dentist
37. Deputy Sheriff
38. Deputy U.S. Marshal
39. Digital Image Librarian
40. Drapery Sales
41. Dry Cleaning Spotter
42. Editor/Report Production Assistant
43. Electrician
44. Electronics Assembler
45. Electronics Technician
46. Emergency Medical Technicians
47. Engineering Technician
48. Environmental Engineer
49. Farm Products Grading Inspector
50. Firefighters
51. Freight Conductor
52. Game Warden
53. Graphic Artist
54. Graphic Designer
55. Hazardous Materials Technician
56. Highway Patrolman
57. IRS Special Agent
58. Lifeguards
59. Location Specialist (Water & Sewer)
60. Maintenance Service Worker
61. Mapmakers
62. Meat Inspector
63. Military (Many job descriptions)
64. Military Engineer
65. Military Pilot
66. Network Systems Administrator
67. Non-Destructive Inspector of Metals
68. Nurse’s Aides
69. Paint Sales
70. Painter
71. Paramedic
72. Pharmacist
73. Pharmacy Technician
74. Photographic Processor
75. Police Officer
76. Prison Corrections Officer
77. Produce Inspector
78. Professional Truck Drivers
79. Quality Control Specialist
80. Railroad Conductor
81. Railway Mechanic
82. Registered Nurse
83. Sanitation Specialist
84. School Bus Drivers
85. Shipping and Receiving Clerk
86. Software Engineer (Some types)
87. State Park Ranger
88. State Trooper
89. Street Lighting Technician
90. Teachers (Some positions)
91. Transit Department Secretary
92. Transit Vehicle Electronic Technician
93. Transportation Security Screener
94. TV Programming Director
95. Vehicle Maintenance Supervisor
96. Wall Covering Sales
97. Warehouse Worker/Driver
98. Weather Analysts
99. Youth Program Director
100. Youth Program Director

*Figure 6 - 100 Occupations where employers may require verified normal color, or, at least, verified color vision testing. (Not all require normal color vision; e.g. aviation, maritime, transport, etc.)*

*{Source: Richmond Products - Color Vision Testing and Occupational Applications}*

# CHAPTER 2 – Detection and Diagnosis

## CONDITION TERMINOLOGY

Chapter 1 explained the terms trichromatic, dichromatic and monochromatic. Because anomalous trichromatism and dichromatism indicate three or two cone pigment-photosensitive anomalies respectively, they are given classifications of color deficiency. The terms protan, deutan and tritan, correspond to characteristics derived from possible anomalies of photosensitive pigments and, therefore, on resultant anomalous color matching variables needed to produce all the spectral hues. A fourth expression, tetartan, denotes the possibility of the existence of a yellow-sensitive pigment anomaly which is theoretically possible. The table in Figure 7 gives an overview of the various classifications. Refer also to the Glossary at the back of this document for further terminology definitions. Figure 8 shows a chromaticity diagram that illustrates the relationships of perceptible colors. The lines extending across the diagram and crossing near its geometric center pass through various subsets of colors that are perceptibly indistinguishable to CVD individuals who lack one of the three normal cone photopigments. These dichromatic confusion lines of protan, deutan and tritan indicate that color vision defects are not random, meaning that we are able to test for each type of CVD and identify the specific deficiency as well as the extent of the defect as colors with greater saturation are presented. As shown, both deutan and protan CVD patients have trouble with green and red, and are just different wavelengths. Further, tritan CVD patients can have trouble with both blue and yellow.

## TESTING METHODS

The key purpose of screening and testing for CVD is to determine if the patient has normal color vision or CVD. If defective, further testing may need to be made to determine the type of deficiency (protan, deutan or tritan), whether congenital or acquired, and the extent (mild, medium or strong). This because, in many cases, CVD may have consequences for certain occupational abilities.

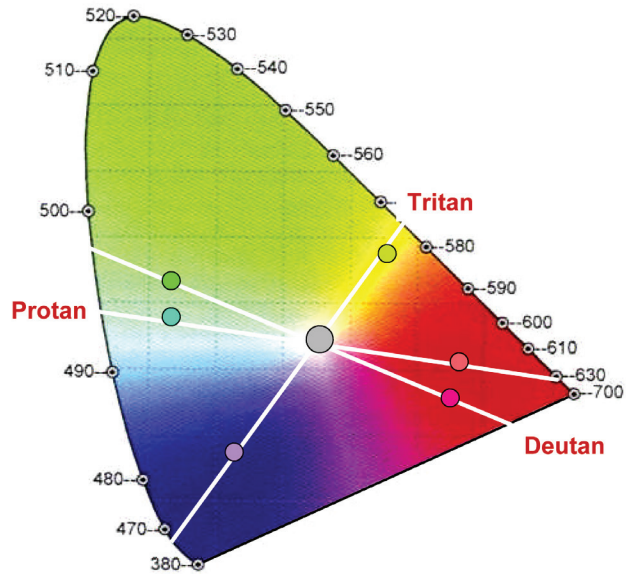


Figure 8 – Chart of Dichromatic Confusion Lines  
{Source: Richmond Products}

There are various types and brands of color vision test products available today. They fall into four main categories:

### Anomaloscope

The anomaloscope is acknowledged as the 'gold standard' for diagnosing color vision defects. These instruments are expensive, not commonly available for clinical use, and complex to use compared with pseudoisochromatic plate tests, color hue discrimination tests and lanterns (see below).



Figure 9 – The Oculus Heidelberg Multi-Color Anomaloscope (Courtesy Richmond Products Inc.)

### Classification of congenital colour deficiency

Number of variables in wavelength matching	Number of cone photopigments	Type	Denomination	Hue discrimination
1	None	Monochromat (Achromat)	Typical or Rod monochromat	Absent
1	One	Monochromat (Achromat)	Atypical, incomplete, or cone monochromat	Absent
2	Two	Dichromat	Protanope Deuteranope Tritanope	Severely impaired
3	Three (one abnormal)	Anomalous trichromat	Protanomalous Deuteranomalous Tritanomalous	Continuous range of severity from severe to mildly impaired
3	Three	Normal trichromat	Normal trichromat	Optimum

Figure 7 – Classification of Congenital Color Vision Deficiency {Source: Diagnosis of Defective Colour Vision, J. Birch – Page 30}



**Pseudoisochromatic Plates**

The most popular method, mainly because test kits are usually available in convenient 'book' form, Pseudoisochromatic plates can be easily employed in a practitioner's office and have proven to be highly successful. The principle is based on a patient's ability to recognize figures (letters, numbers or geometric shapes) within colored dots of varying sizes. Persons with normal color vision can easily see the shapes, while they are effectively hidden (or camouflaged) those with CVD cannot discern some or all shapes, the number missing depending on the type and degree of deficiency. The most frequently used pseudoisochromatic plate tests are the Ishihara and the Richmond HRR 4th Edition. Other plate tests are listed in the table in Figure 13.

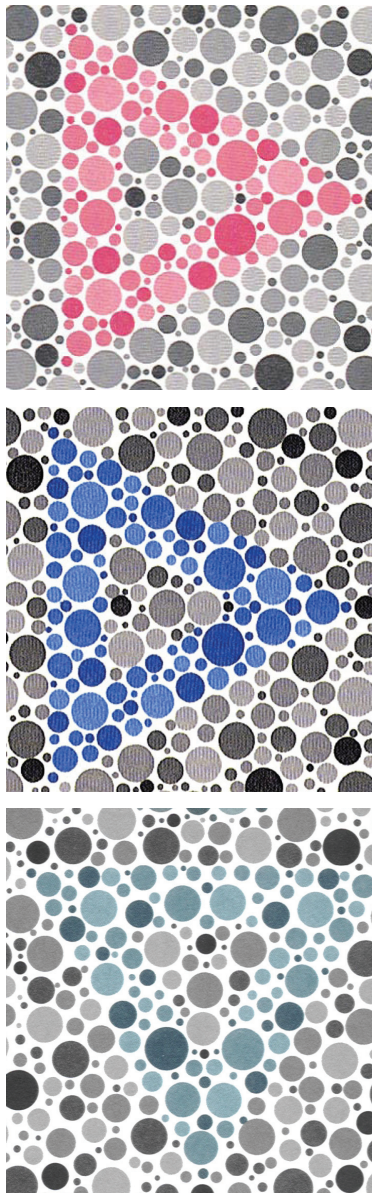


Figure 10a –Examples of Pseudoisochromatic Test Plates from the HRR 4th Edition. The symbols seen in the three plates (from top to bottom) would not be visible for, respectively, strong deutan, protan and tritan CVD cases. Other plates in the HRR 4th Edition employ symbols for screener, mild and medium defects.

{Source: Richmond Products}

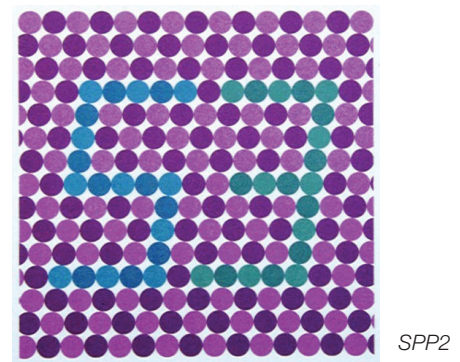
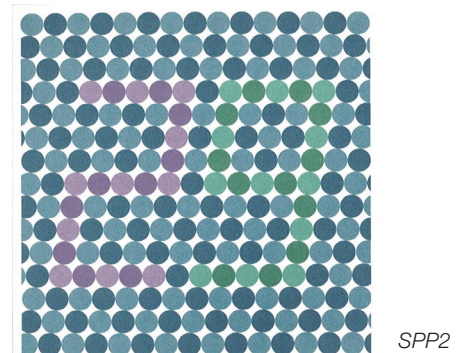
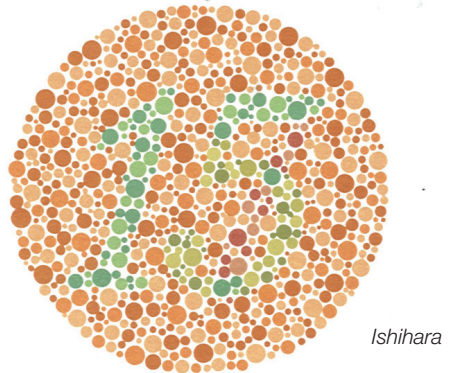
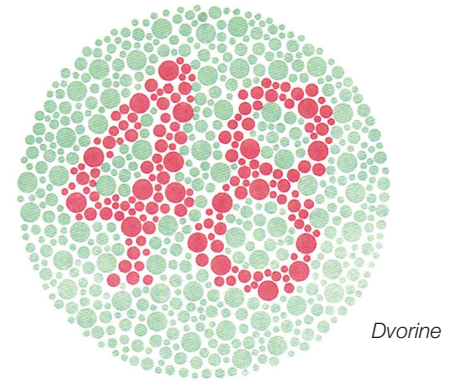


Figure 10b - Further Examples of Pseudoisochromatic Test Plates

Hue Discrimination – Also known as grading arrangement tests, the orderly progression of hue, often with respect to a reference hue. Hue discrimination tests are used to identify moderate and severe cases of both congenital and acquired CVD, mainly in adults, and may permit some classification of protan, deutan and tritan defects.

The majority of hue discrimination tests comprise a number of colored caps that a subject is required to select and identify with reference-color caps. The most widely used hue discrimination tests are the Farnsworth Panel D-15 and the expanded version, the Farnsworth-Munsell 100-Hue. Another challenge with arrangement tests is correct scoring. Scoring is especially complex with the 100 Hue Test. Some studies have shown that scoring is incorrect in 30 percent of cases.

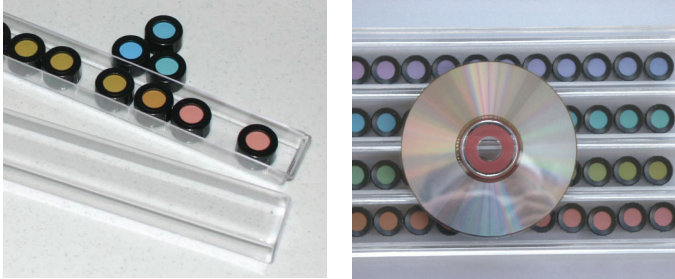


Figure 11 – The Farnsworth Panel D-15 Hue Test Kit (left) and the Farnsworth-Munsell 100 Hue Test Kit

(Courtesy Richmond Products Inc.)

### Lantern

Lantern testing is often used in occupational applications where recognition is required of small areas or pinpoints of colored lights that simulate signals. Occupation examples include mariners, airline and military pilots, locomotive engineers, etc. The method involves a lantern receptacle containing a set of color filters that produce the effect of signal colors through which the subject is required to name the colors and in a given sequence. At least 20 single, double or triple colors to be identified in rapid



Figure 12 – The Fletcher CAM Lantern

(Courtesy Evans Instruments Ltd.)

(usually about 5 seconds) succession are shown in daylight (most often), low-light and darkness conditions. Today, the Farnsworth Lantern-Falant test is the most commonly used. A recent development is the Fletcher-Evans CAM Lantern, a successor to the discontinued Holmes and Wright lantern, first manufactured in 1982. Lanterns are also relatively expensive.

### Computerized Testing

From about 2002 a number of computerized or computer-controlled color vision screening/testing products have been proposed with the aim of making color tests more readily available. Many are still in prototype stage. Since they are software-based a few are affordable and can be used at home. One product attempts to mimic three tests: Farnsworth Dichotomous D-15, Farnsworth-Munsell 100-Hue, and pseudoisochromatic plates. Until such products have been thoroughly tested, particularly for color contrast accuracy, evaluated, proven and their results published, their efficacy remains questionable. It is also essential that such products be capable of maintaining accurate calibration.

### PEDIATRIC CVD

It is extremely advisable to examine children for CVD at as young an age as possible, mainly because color vision problems can affect learning abilities and reading development. In many situations, color is innocently used in early grades as a learning aid. Children may try to hide the fact that they cannot see certain colors by watching other classmates or even copying their work. Not being able to tell the difference between colors can be a serious problem for children and can lead to poor class work and subsequent low self-esteem. It is important that parents be sensitive to look for any color deficiency or any other vision difficulties so that teachers can be notified and become better informed of the limitations or handicap that could result.

As explained in Chapter 1, congenital CVD is much less prevalent in females than in males. Despite this, it is important that children of both genders are tested. Obviously, children with severe CVD will make mistakes in color discrimination that are noticeable by parents and teachers. Color is frequently used as a teaching aid up to about the age of seven. After that, color-coding methods are used in various school curricula and color recognition is critical in many school subjects such as art, chemistry, engineering, geography, biology, political science and economics, particularly where histograms or pie charts are used. Less severe

Product	Screening	Congenital	Acquired	Ability to Classify	Determine Extent	Nr. of Plates	Pediatric Version	Time to Administer
HRR 4th Edition	Yes	Yes	Yes	Yes	Yes	24	Yes	Quickest
Ishihara – 24 Plates	Yes	Yes	No	No	No	24	Separate	Incomplete
City Univ. London	Yes	Yes	Yes	Yes	No	10	No	Moderate
Dvorine	Yes	Yes	No	No	No	23	No	Moderate
Lanthon Tritan Album	Yes (BY only)	No	Yes	No	Yes	5	No	Quick
SPP1	Yes	Yes	No	Yes	No	19	No	Slower
SPP2	Yes	Some	Yes	Yes	No	12	No	Slower
Farnsworth D15	No	Yes	Yes	Yes	Med/Strong	16 chips	Yes	Slow
Farnsworth Desat. 15	No	Yes	Yes	Yes	Mild Only	16 chips	No	Very Slow

Figure 13 – Overview of some Color Vision Testing Methods and their Evaluation

cases of CVD may not be so easily discovered until much later in life. Delaying screening may result in educational disadvantage, limitations of career choice and other employment problems when that may result in disappointment.

Traditionally, color vision testing of children was the realm of pediatric eye-care practitioners, which certainly helped to emphasize the importance. Nowadays, large groups of children are often screened for CVD at school in the context of routine health checks. If CVD is detected a child should be helped to understand that this is not a stigma so as to be treated unfairly or bullied by his or her peers. In any event, a CVD case should be discussed with the child's parent(s) and the family (primary care) practitioner. It is essential to avoid possible psychological trauma or denial. Ideally, color vision testing should be part of a child's first eye examination when formal education (In some states in the US, and possibly in some countries, such testing is mandatory). Pseudoisochromatic plate tests such as the HRR 4th Edition, that use a small set of familiar geometric shapes to be identified are particularly useful for early screening since children readily understand the concept and are most cooperative. In some cases a matching task/test may be more effective. Moreover, a suitable member of the school's health staff can easily perform the test. An abundance of attractive eye examination aids for toddlers is available from vision test suppliers. These include various age-appropriate 'toys' to make CVD testing 'fun' for the subject.

## CHAPTER 3 – Toxicology Effects on CVD & their Detection

### DRUGS & OTHER CHEMICAL SUBSTANCES

Ocular toxicology research has shown that certain prescription drugs, over-the-counter drugs, some industrial chemicals and even some herbal compounds can induce ocular side effects in humans. A change in visual acuity and/or color perception/appearance is often an early indication of these side effects. The short list in Chapter 1 (page 5) mentions a few types of substances, but is by no means complete. It should be noted that in a number of cases where substance toxicity is known to affect color vision it may be caused by exceeding the prescribed dosage, prolonged usage, or the development of hypersensitivity to the offending drug or chemical. If any of these side effects is detected, a more extensive medical history is warranted to determine what is causing the deleterious affect on the eye. In many cases, it has been reported that early detection and subsequent elimination of the source can lead to restoration of the yellow or blue sensitivity.

The reference book *Clinical Ocular Toxicology – Drugs, Chemicals and Herbs*, by Fraunfelder et al, describes a large number of drugs, industrial chemicals and herbal compounds that may induce ocular side effects in humans. Specifically, there are 96 substances listed that are 'certain' to cause color vision defects, 21 substances are listed as 'probable' and 16 more as 'possible'. These statistics are illustrated in the bar chart in Figure 14. In the past few years, this list has continued to grow due to the development of new drugs, especially those that treat conditions of the nervous system.

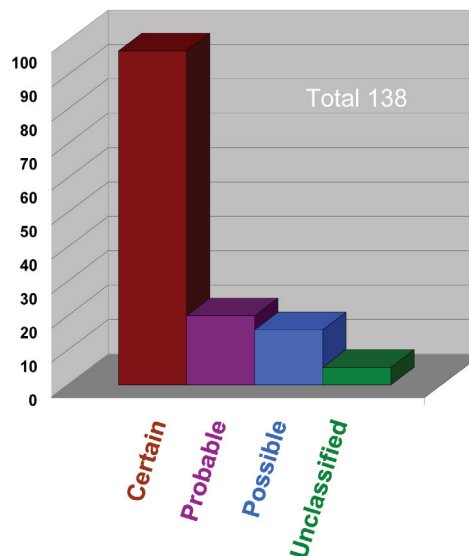


Figure 14 – Probability (x-axis) and number (y-axis) of chemical and herbal substances that may cause color vision defects

[Source: Richmond Products, adapted from *Clinical Ocular Toxicology – Fraunfelder et al*]

### DIETARY SUPPLEMENTS & VITAMIN DEFICIENCY

The worldwide dietary supplementary industry was estimated in 2007 to be valued at about USD 60 billion, and increasing. Unfortunately, in many countries, especially the US, dietary supplements are not subject to government regulations concerning pre-marketing safety, efficacy claims or adverse effects. Therefore, much care is advised before using dietary supplements, in number of types, dosage, prolonged usage and in combination with over-the-counter and prescribed medications.

Fraunfelder et al report that **Vitamin A** deficiency is often associated with alcoholism and some rare metabolic storage diseases in which lack of an enzyme affects various organs, especially the liver, and tissues. Predominant ocular anomaly symptoms of Vitamin A deficiency are night blindness and limited visual field, often resulting in acquired tritan defects. In extreme cases, this can lead to a total loss of hue discrimination and other color vision anomalies. In most such CVD cases, recovery can be effected by aural dosage of Vitamin A, as typically indicated using a hue discrimination test such as the Farnsworth-Munsell 100-Hue. However, prolonged usage of Vitamin A supplements can cause intracranial hypertension and subsequent adverse color vision effects, for example, of yellow-hued objects (known as xanthopsia which is difficult to identify since effects are not differential as in other types of CVD, including red CVD and over-enhanced red dyschromatopsia. Red dyschromatopsia is not only Red CVD but also loss of sensitivity to red light so it is not strictly protan.

Adverse ocular reactions can occur through deficiency of **Vitamin D**, but seldom include CVD. However, the treatment of any adverse vision effect should be analyzed in the context of possible drug side-effects that may lead to one or more color vision anomalies.

Frederick T. Fraunfelder, MD. Head of Ophthalmology at Oregon Health and Science University's Casey Eye Institute, Portland Oregon

### A Further Word on Toxicology Effects on Visual Acuity and Color Vision

"I am grateful that references to my work, and that of my co-author colleagues, are made in this Tutorial's chapter Toxicology Effects on CVD & Their Detection. In authoring the two books referenced in the Tutorial we intend them as a guide to help the busy clinician decide whether a visual problem is related to a chemical or is medication-induced. The clinician's experience, the knowledge and, natural cause of a disease, the adverse effects of certain compounds and other substances, and patient reports all help in making decisions."

"There are many variables and much financial investment in research leading to the assessment of a cause-and-effect relationship between very many drugs, chemicals and herbs and any particular visual anomaly. The clinician must keep in mind the strong variability of how humans metabolize or react to a drug or other substance, where the variability may affect the incidence data. A significant change in the expected course of a disease after starting treatment should heighten the physician's suspicion of a drug-related event."

"Medical case reports, reporting systems and their inherent weaknesses are still the backbone of clinical ocular toxicology. We have attempted to classify the cause or causes of a suspected adverse effect by employing 'certain', 'probable', 'possible' etc. in order to remind us that the exercise is not based on science. So, remembering that the books are only a guide for busy clinicians and will always be works-in-progress, we trust that you will be able to make use of them in your studies and eventual or present chosen careers."

## TESTING FOR TOXICOLOGY EFFECTS ON COLOR VISION

With the increasing development of prescription drugs that deal with the nervous system, the demand for color tests that detect for tritan (yellow and blue) defects has grown. The currently available tests with this capability are the HRR 4th Edition, SPP2 and Lanthony desaturated arrangement tests. Each of these also tests for protan and deutan (red/green) defects.

## CHAPTER 4 – Treatment, 'Compensation' & Cure

### 'THERAPEUTIC' METHODS

As stated in chapter 1, there is currently no known medical cure for congenital CVD. Some CVD sufferers can be helped by color filters which act to increase the contrast and reportedly make it possible to distinguish colors close to the confusion lines. Some people benefit from the use of an X-Chrom lens which is available as a contact lens. Again, these filters may serve to increase color contrast. Further, spectacles that reduce glare may also help congenital CVD sufferers.

In the absence of the development of a cure for congenital CVD, safety remains a key issue. Those with a strong or medium level of congenital CVD need to avoid activities where color confusion may jeopardize others.

In many cases, there are ways to help compensate for the inability to see or distinguish colors by the way objects or people's actions are observed. Also, relying on brightness or location, rather than color, to identify objects or situations can help. For example, by learning the order of the three colored lights on a traffic signal and knowing that if the lowermost light is illuminated, it means that the light is green (see Figure 15). In some cases of CVD (protan) the red signal has inherent low visibility/ brightness which is especially problematic with bright

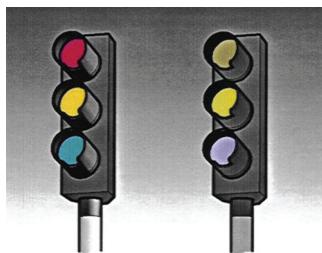


Figure 15 – Vertical Traffic Lights as seen with Normal Color Vision (left) and as they might appear with some types of CVD (right)

{Courtesy J. Neitz and M. Neitz, Medical College of Wisconsin}

daylight illumination. This also applies to vehicle brake lights. Low visibility effectively increases reaction time. However, because of the safety aspect, this is not often carried out in practice if the person with CVD is driving a vehicle, especially a heavy goods truck, or is even a pedestrian.

### POINTS ABOUT ACQUIRED CVD

If initial screening reveals an adverse color vision defect that seems to be newly acquired it is strongly advisable to check the patient's medical history to determine if the cause may be drug-induced or secondary to disease. In this way, early treatment may be effective by changing or reducing that patient's prescription. Some acquired color vision defects, caused by disease or cataracts, can be suspended by surgery, depending on the cause and not always permanently. Many acquired CVD cases caused by optic nerve diseases can be effectively treated. Other cases caused by prescription drug or exposure to solvents can often be overcome when exposure to the offending substance is eliminated.

### GENE THERAPY RESEARCH IN PRIMATES

In 2009 the Departments of Ophthalmology at the University of Washington in Seattle, the University of Florida in Gainesville and the Medical College of Wisconsin in Milwaukee published the results of a research program aimed at correcting the red-green vision of squirrel monkeys with congenital (dichromatic) CVD using gene therapy. It was shown that after applying gene therapy the monkeys were able to distinguish between patterns of gray, green and red dots.

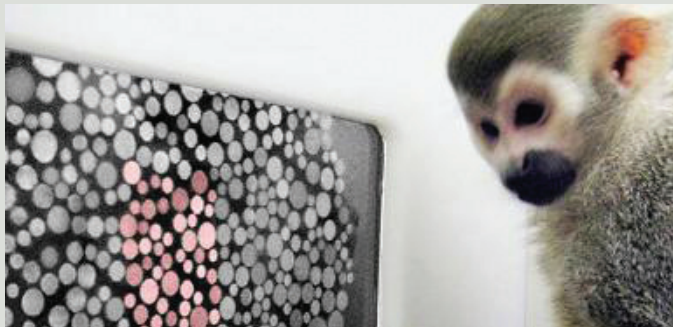
The researchers used a computerized version of a pseudoisochromatic plate method similar to the Ishihara and the HRR 4th Edition tests. As with humans who suffer from red-green CVD, the monkeys could not distinguish between these colors. Following treatment that added the missing visual pigment gene, known as the L opsin (the one missing or altered/ anomalous gene), into the retina, the monkeys were able to pass the test for all colors.

In the future it is possible that this technique could prove to be a safe and effective cure for red-green CVD and other visual disorders related to the retinal cones. Permission to perform the gene therapy on human subjects with CVD has been requested.

Jay Neitz, PhD, Bishop Professor,  
Department of Ophthalmology at the University of  
Washington, Washington DC

### A Further Word on Gene Therapy

"I certainly applaud the publication of this new Tutorial on CVD, because, in fact, my work of many years has its primary focus on the mechanisms of color vision. As briefly described in the Tutorial, scientists in the Neitz lab, along with other eminent institutions, are conducting research into gene therapy, which has so far shown some extremely interesting results. The prospect of easing the problems caused by color blindness makes it an attractive future target for human gene therapy. Because the squirrel monkey's visual system is similar to that of humans, and a human gene was used to replace the missing visual pigment of the monkeys, scientists are optimistic about the possibility of gene therapy to cure CVD in humans. In the same way that few would settle for a black-and-white television or monochrome computer monitor, it is easy to imagine that many color-blind people would want the cure if there were no risk to their vision or health. While no adverse side-effects were observed in the monkeys, the most important barrier in moving the treatment forward will be ensuring its safety for use in humans."



"While gene therapy has successfully enabled our squirrel monkeys with red-green CVD to 'see' new colors, we do not know what their physiological perceptions of those colors are. We also do not yet know if any psychological side effects might occur in humans, even though our monkey subjects have shown no such effects or other signs of distress. However, gene therapy does involve some risks associated with the viral vector and injection of the therapeutic transgene and the surgical procedure. Therefore, the first step in moving the research forward is to determine the safety of the treatment for human subjects."

Gene therapy experiments on humans must first be reviewed and approved by the US National Institute of Health (NIH) Office of Recombinant DNA Activities (ORDA/Recombinant DNA Advisory Committee (RAC) and by the Investigational New Drug Application (IND) of the Food and Drug Administration (FDA). In addition, approval must also be obtained from an Institutional Review Board (IRB) where the study will take place.

For further information on gene therapy please go to:  
[neitzvision.com /content/genetherapy.html](http://neitzvision.com/content/genetherapy.html)

## SHORT GLOSSARY

Note:

*This glossary is abbreviated to suit the conciseness of the tutorial. Comprehensive glossary publications are listed under FURTHER READING at the end of this paper.*

### Achromatism/Achromatopsia

Rare inability to distinguish colors. See also Monochromacy.

### Autosomal

Refers to dominant or recessive inheritance.

### Chromosome

One of 46 structures in the human cell nucleus which carries the genes that contain the hereditary material controlling the growth and characteristics of the body.

### Cone

Light-sensitive retinal receptor cell that provides sharp visual acuity and color discrimination. See also Rod.

### Deutan

Refers to a person who has deuteranopia, a type of dichromatism in which red and green are confused. Also deterranomaly, a type of anomalous trichromatism in which an abnormally high proportion of the green is needed when mixing red and green to produce yellow.

### Dichromatism

Moderately severe color vision defect in which one of the three basic color mechanisms is absent or not functioning.

### Dyschromatopsia

Any type or degree of defective color vision.

### HRR

Hardy-Rand-Rittler pseudoisochromatic plate test of colored dots that appear as recognizable geometric shapes. Used for identifying color vision deficiencies.

### Ishihara

Pseudoisochromatic plate test similar to the HRR test, but with certain limitations.

### Lanthony Desaturated

Refers to the Lanthony Desaturated 15-Hue Test used to detect congenital or acquired color vision deficiencies. See also Munsell Scale.

### Monochromacy

Same as achromatopsia (above)

### Munsell Scale

Standardized scale of colored materials having variations in hue and saturation.

### Nystagmus

Involuntary, rhythmic side-to-side or up-and-down (oscillating) eye movements that are faster in one direction than the other.

### Optic Nerve

Second cranial nerve. Largest sensory nerve of the eye that carries impulses for sight from the retina to the brain.

### Photophobia

Abnormal sensitivity to, and discomfort from, light.

### Photopigment

A pigment which is affected by, or unstable in the presence of, radiant energy, especially visible radiant energy, such that its chemical composition is altered. In the eye the breakdown of photopigments by light is the first stage in the visual process.

## Photoreceptor

A receptor capable of reacting when stimulated by light, such as the rods and cones of the retina.

## Protan

Refers to a person who has protanopia, a type of dichromatism in which only two hues are seen. Also protanomaly, a type of anomalous trichromatism in which an abnormally high proportion of the red primary stimulus is needed when mixing red and green to produce yellow.

## Retina

The light-receptive, innermost nervous tunic of the eye.

## Rod

A photoreceptor cell of the retina which connects with a bipolar cell. It contains rhodopsin and is involved scotopic vision. See also Scotoma.

## Scotoma

An area of partial or complete blindness surrounded by normal or relatively normal visual field.

## Sensitivity

The capability of responding to or transmitting a stimulus. Also, the extent to which a test gives results which are free from false negatives.

## Spectrum

Spatial display of a complex radiation produced by separation of its monochromatic components.

## Tetartan

Refers to a person with tetartanopia or tetartanopsia, theoretical conditions and terms for a type of blue-yellow blindness in which there are two neutral points.

## Trichromatic

Requiring the use of three color mixture primaries to match all perceived hues. Anomalous trichromatic is a form of defective color vision in which three primary colors are also required for color matching, but the proportion of primaries in the mixture-matches are significantly different from those required in normal trichromatism.

## Tritan

Refers to a person having tritanomaly or tritanopia. The former is a rare type of defective color vision in which an abnormally large proportion of blue must be mixed with green to match a standard blue-green stimulus. Tritanopia is a form of dichromatism in which all colors can be matched by suitable mixtures of only a red primary and a green or blue primary.

## Visual Acuity

Assessment of the eye's ability to distinguish object details and shape, measured by the smallest identifiable object that can be seen at a specified distance.

## X-Linked Recessive

Hereditary characteristic carried on the X (female) chromosome.

## SELECTED FURTHER READING

(note: It is advisable to consult the bibliography sections of the titles listed below for more detail)

### Specific to CVD

*Colour Blindness Causes and Effects* – Donald McIntyre, MA, PhD. Dalton Publishing, Chester, UK.

*Color Vision in the Occupational Setting* – Bernard Blais, MD, FACOEM, FAAPO, FACS. RP Press, Atlanta GA.

*Diagnosis of Defective Colour Vision\** – Jennifer Birch. Butterworth Heinemann, Oxford, UK

*Normal and Defective Colour Vision* – John Mollen et al. Oxford University Press, UK

### General Optometry/Ophthalmology

*Visual Fields – Clinical Case Presentations\** – John C. Townsend, OD, FAAO, et al. Butterworth Heinemann, Stoneham, MA.

*Neuro-Ophthalmic System Clinical Procedures* – Patricia A. Modica, OD. Butterworth Heinemann, Woburn, MA.

*The Psychophysical Measurement of Visual Function* – Thomas T. Norton, PhD, FAAO, et al. Butterworth Heinemann, Woburn, MA.

*Clinical Procedures in Optometry* – J. Boyd Eskridge, OD, PhD, et al. J.B. Lippincott Company, Philadelphia, PA.

*Diagnosis and Management in Vision Care* – John F. Amos, OD, et al. Butterworth Heinemann, Newton, MA.

### Toxicology

*Drug-Induced Ocular Side Effects* – Frederik T. Fraunfelder MD, et al. SAUNDERS Elsevier.

*Clinical Ocular Toxicology* – Drugs, Chemicals and Herbs – Frederick T. Fraunfelder MD, et al. SAUNDERS Elsevier.

### Glossaries/Dictionaries

*Dictionary of Ophthalmology* – Michel Millodot, PhD & Daniel M. Laby, MD, Butterworth Heinemann, Oxford, UK and Woburn, MA.

*Dictionary of Optometry and Visual Science* – Michel Millodot, PhD. Butterworth Heinemann, Oxford, UK and Woburn, MA.

*Dictionary of Eye Terminology* – Barbara Cassin et al, Triad Publishing Company. Gainesville, FA.

*Dictionary of Visual Science and Related Clinical Terms* (Books 1 & 2) – H.W. Hofstetter. OD, PhD, et al. Richmond Products, Albuquerque, NM.

\*out of print

N.B. It is also recommended to visit the websites of the various optometry/ophthalmology associations and organizations, which provide useful information and further links:

*American Association for Pediatric Ophthalmology and Strabismus:* [www.aapos.org/](http://www.aapos.org/)

*American Optometric Association:* [www.aoa.org](http://www.aoa.org)

*American Academy of Ophthalmology:* [www.aaao.org](http://www.aaao.org)

*College of Optometrists in Vision Development:* [www.covd.org](http://www.covd.org)

*Aerospace Medical Association:* [www.asma.org](http://www.asma.org)

*Wikipedia also offers a description of color blindness:* [www.wikipedia.org/wiki/Color\\_blindness](http://www.wikipedia.org/wiki/Color_blindness)

*Color blindness and Medicine:* <http://www.colourmed.com/authors.html>

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