

# Ocular Diseases

## Chapter 1

### Color Vision in Medicine

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#### General Introduction

#### Neurophysiology of Color Vision

The study of color vision is an interdisciplinary subject which embraces aspects of Physics, Biochemistry, Neurophysiology and Psychology. Light is absorbed by pigments in the photoreceptor layer of the eye's retina initiating a photochemical reaction. By a transducer process, which is still largely a mystery, the various attributes of light energy are coded for transmission to the brain by neural signals; here the signals are later interpreted. The perception of color is a psychophysical experience which is dependent on the physiological coding and processing that takes place in the eye and brain. The fault which gives rise to defective color vision, lies in the retina and/or visual pathway.

Color vision has attracted the attention of some of the greatest scientific minds in Newton, Maxwell, Young and von Helmholtz [27; 38; 51; 52; 53]. The original concepts that were established provided an invaluable foundation and led the way to the more recent contributions of Abney, Edridge-Green, Judd, Wright, Hurvich, Jameson, Rushton and many others [17; 37]. The last two decades have seen major and vital contributions by neurophysiologists too numerous to mention by name. There remain no single ideal method of approach to color vision testing and diagnosis; each case must be treated individually.

Light traverses the neural layers of the retina to the photoreceptors which are in the innermost region (posterior) and is absorbed by the photo-pigments contained within the

receptors; subsequently a neural discharge is initiated by which the signal is transferred to the brain. The retina has been shown to play a major role in separating out the colored elements of the stimulus from brightness characteristics and coding these two attributes. Little is known about the mechanisms converting the photochemical change in the sensory receptors to an electrical potential. It involves complex internal chemical reaction in which changes in the potassium and sodium balance play a major part. Pharmacological agents known as neurotransmitters act to carry the coded signal from one neuron to the next as they are released from neurons at the synaptic junction. One important and novel characteristic of photoreceptors is their hyperpolarizing action (production of a negative potential in the cell membrane) as a consequence of light absorption.

Nerve impulses signaling color information are relayed from the lateral geniculate nucleus via the visual radiations, to the main visual areas of the brain, the striate cortex. The principal zones were designated Area 17 by Brodmann in his classical division, and then onwards to Area 18 (the occipital or para-striate cortex) and Area 19 (the pre-occipital cortex). Some non-color fibers concerned with eye movements project to the superior colliculus and pre-tecta regions. The visual areas maintain strong foveal representation having specific locations within the region for each part of the visual field. Retinal organization or spatial mapping is thus preserved and fibers conveying signals from corresponding points of the two retinæ remain close. Foveal regions are spatially magnified in the lateral geniculate nucleus and striate cortex to ensure maximum resolution in this region [57; 58; 59].

### **Clinical Tests for Color Vision Analysis**

Color is used for decoration in modern society and to convey specific information by means of a recognized code. Accurate color specification and measurement is needed. That some people see colors differently and make mistakes identifying colors has been known for over 200 years. Congenital color deficiency (colorblindness) is found in all ethnic groups. Red-green deficiency is inherited as an X-linked trait and affects about 8% of men and 0.4% of women. There are different types of red-green color deficiency as well as differences in severity [1]. Some people have wide ranging difficulties with color and others are only minimally disadvantaged. Congenital color deficiency for blue is rare and has equal prevalence in men and women. Color vision normally remains the same throughout life but changes can occur as a result of ocular, or intracranial, pathology or the prolonged use of some prescribed drugs. Poor blue vision is the most common acquired loss. All people with color deficiency see fewer colors in the environment and confuse colors which look different to people with normal color vision. Color matching ability is impaired. There are a number of clinical color vision tests which aim to identify, classify and grade the severity of color deficiency or are designed to determine occupational suitability. The number of example of typical results obtained on a battery of color vision tests by people with different types of color deficiency have been

increased.

Color vision tests are used clinically to identify and differentiate congenital and acquired color deficiency and to select personnel for occupations which require good color vision. Clinical color vision tests are based on psychophysical methods but use pigment colors instead of spectral stimuli. Many tests aim to demonstrate isochromatic color confusions. A range of color difference are utilized to estimate the severity of color deficiency but dichromats and anomalous trichromats cannot be distinguished with pigment tests [2].

Screening tests identify people with normal or abnormal color vision. Grading tests estimate the severity of color deficiency. Some tests have both screening and grading functions. Most tests aim to classify protan, deutan and tritan color deficiency.

The efficiency of most tests in current use has been established in clinical trials with previously diagnosed normal and color deficient observers.

The terms “sensitivity” and “specificity” are in general used to indicate screening efficiency. Sensitivity is the percentage of color deficient people correctly identified as abnormal, and specificity is the percentage of color normal correctly identified as normal. An efficient color vision screening test should have a specificity of 100%. This is equivalent to no false positive results or no people with normal color vision incorrectly identified as color deficient. Sensitivity should be over 90%, preferably over 95%. This is equivalent to, respectively, 10% and 5% false negative results, or color deficient people incorrectly identified as normal. A small percentage of false negative results is acceptable as long as these are cases of very slight, or minimal color deficiency. Color vision tests can also be analyzed in terms of test/retest reliability. This measure is most applicable to grading tests which allow people with slight color deficiency to pass [2; 9].

## **1. Acquired Color Vision Deficiency**

### **Introduction**

There exist a wide group of color vision disturbances which are acquired during life, predominantly the result of ocular or general disease, the consequence of exposure to a chemical, toxin or medication, or resulting from physical injury to the head. The incidence of such disturbances is uncertain although the estimate by Smith that at least 5% of the population have an acquired defect severe as the 8% with a congenital defect is a valuable guideline [35].

Alteration to color vision which arise through life from a cause or causes other than the normal physiological processes have received far less general attention than the inherited defects. Although physicians were aware about two centuries ago of changes to color perception

resulting from disease, accounts were confined, in general, to isolated cases up until the classical description by Kollner in 1912 and renewed was shown thereafter by German ophthalmologists [18].

It is now realized that color vision changes provide a valuable means of monitoring the progress of a disease, or the toxic effects of a chemical substance, whether exposure is deliberate for therapeutic purposes or unintentional in the case of an industrial hazard. The effectiveness of treatment can be assessed by the continued monitoring of recovery of an acquired color vision disturbance. Renewed interest has thus been concentrated around the use of color vision as a diagnostic tool although there have been differing estimates of the efficiency of color vision test as the earliest index of malfunction [7; 10; 23].

Disease in this context has a wide meaning, frequently involving some inherited conditions. It embraces, dystrophies (many of which are associated with metabolic disturbances), varied consequences of trauma or of tumors and degenerative conditions (age-induced change is one example). Many other pathological conditions, involving damage to the nervous system can be included [5; 15].

Acquired disturbance of color vision can progress from normal trichromatism to anomalous trichromatism on to a dichromatic stage and even to monochromatism [10].

List below are some of the important characteristics of the acquired color vision deficiency:

- Differences in color perception between eyes
- Color loss may be accompanied by deficiencies in other visual areas
- Disturbances of blue-green-yellow vision are common
- Females are affected in the same proportion as men
- The elderly are particularly susceptible

Severity of the defect is variable according to the progression of disease and the disease's degree:

- Transient chromatopsia may be present
- Colors can often be named correctly color, perception is improved when all the external conditions are improved
- An acquired defect may be superimposed on an inherited defect
- The severity of an acquired defect depends on whether the cause is active or inactive

## 1.1. Color Vision deficiency by Multiple Sclerosis

The retinal signals travel to the brain via axons of the ganglion cells forming one million optic nerve fibers organized in a systematic manner. Macular fibers move from the fovea. Signaling color, occupy approximately one third of the optic nerve area on the temporal side, where they enter. They later move to a central location in the nerve, while towards the chiasma they shift medially.

At the optic chiasma 60% of fibers decussate in man. Fibers from the nasal part of each eye cross in the chiasma, travelling centrally in the optic tract to the opposite side of the brain; those from the temporal part of the retina remain on the same side. Within the optic tract projected fibers from the retinal areas are separated. The fibers, carrying impulses coding color, terminate at the lateral geniculate body or nucleus. Electrophysiological recordings from the tract indicate that the color patterns remain unchanged from the retinal ganglion cells; thus the two classes of non-opponent cells coding brightness and four classes coding red-green and blue-yellow stimuli are indicated at this level in primates [22; 33].

Color vision deficiency by multiple sclerosis represents the type 2 of the acquired color vision deficiency. Type 2 occurs in some lesions of the optic nerve and generally in demyelinating diseases, which cause a defective electrical conduction along the red-green axis without altering the wavelength of maximum relative luminous efficiency. Cone dystrophies and optic nerve lesions may be difficult to distinguish clinically since both abnormalities produce reduced visual acuity and central field defects. Therefore, the type of acquired red-green color deficiency may be of diagnostic importance [41].

The aim of the present work is to assess the type and degree of both red-green and blue-yellow color vision deficiency in Calabrian males affected by multiple sclerosis.

### Patients and Method

Eighty Calabrian male patients (age range 18-70 years; mean age,  $40.6 \pm 12.4$  years) showing a mean disease duration of  $10.6 \pm 8.2$  years (range 0.5-46 years) admitted to the Institute of Neurology, Magna Graecia University, Catanzaro (Calabria, Southern Italy) were enrolled in this study. 70 control subjects were matched for age and sex. An ophthalmologist examined all patients and controls in order to rule out diabetic retinopathy, cataracts, senile maculopathy, or ocular fundus anomalies. We excluded from the results analysis 7 colorblind patients and 1 patient affected by bilateral maculopathy. The analyzed sample was 72 patients. 21/72 patients reported optic neuritis episodes in their medical histories. Fixed sampling of males allowed us to avoid the genetic Lyon phenomenon [24], which is present only in the heterozygous females for X-linked diseases such as colorblindness (the inherited red-green color vision deficiency). Therefore, the exclusion of females in our sample allowed us to avoid

those heterozygous colorblind females who would be false positive for the acquired red-green color vision deficiency caused by multiple sclerosis thus altering the results analysis. The relatively high frequency of the inherited red-green colorblind females united Lyon genetic phenomenon presence by the heterozygous females for the inherited colorblindness should be the real cause to mistake discriminating between the red-green inherited colorblindness and that acquired due to multiple sclerosis. We should not comprehend if homozygous female status is really inherited or acquired; and in heterozygous status we miss all those females miming the normal color vision. The acquired color vision deficiency is real in the males cohort because they have not the compensation presence by second X chromosome and the anomaly has not hidden.

All patients and controls underwent the following test: Ishihara test [16], which is the most reliable among the pseudo-isochromatic test to identify inherited colorblind subjects. Patients who made more than five errors reading the first 17 plates were diagnosed as being colorblind. The type of colorblindness was determined by reading the last four tables. Farnsworth Dichotomous D-15 test [8] identified both the greatness of color vision deficiency by the high number of errors (maximum value, n. 15) and the type (deutan, protan, tritan). The City University test [9] identified (both binocularly and monocularly) people with different types and degrees of the acquired red-green deficiency (maximum value of errors number, n. 6) and blue-yellow color vision deficiency (maximum of errors number, n. 3).

## Results and Conclusion

37/72 of the patients showing a color vision deficiency were subdivided into two subgroups: subgroup one showed red-green deficiency (21/37); subgroup two showed a coupled red-green and blue-yellow deficiency (16/37). Furthermore, we found two distinct curves showing a groove within the first 10 years of the disease. Both monocular and binocular analyses allowed us to identify the patients showing the monocular color vision deficiency, but they were well compensated by binocular vision [33].

We think that the majority of the patients with the red-green deficiency will develop the coupled red-green and blue-yellow deficiency in the latter years of multiple sclerosis [33].

### 1.2. Color Vision Deficiency by Parkinson Disease, Parkinsonisms, Essential Tremor

The neurological literature shows highly specific visual syndromes acquired by cerebral diseases, general pathology, intracranial injury, and prolonged use of some therapeutic drugs. The abnormality can originate anywhere in the visual pathway, from the retinal receptors to the visual cortex. Dopaminergic neurons act in the outer and inner retina at multiple levels; dopamine is a chemical messenger for light adaption promoting the flow of information through cone circuits diminishing through rod circuits. Owing to the segregation of color information



at the retina into blue-yellow and red-green pathways, dopaminergic neurons can use color discrimination tasks to assess cone and retinal ganglion cells. Changes in color vision provide evidence of pathology and provide informative about color processing in the visual pathway [48].

Currently, there is no study on color vision comparing Parkinson's disease and Essential tremor; we aim to assess the color vision deficiencies of these two diseases to determine if different neurodegenerative pathways can influence color vision.

## **Patients and Method**

After informed consent, 45 male Parkinson's disease patients (age range 37-75, mean 60.0 and Hoehn-Yahr test score 3-4), 45 male Essential tremor patients (age range 38-80, mean 62.4), and 45 male controls (age range 40-77, mean 63.7, without any neurological diseases and with normal color vision) underwent the Ishihara test [16], which is the most reliable among the pseudo-isochromatic tests to identify colorblind subjects (for inherited color vision deficiency, protanopy or protanomaly; for inherited green color vision deficiency, deuteranopy or deuteranomaly). All patients who made more than five errors reading the first 17 plates were diagnosed as being colorblind. The type of colorblindness was defined by reading the last four tables of the test. We used the Farnsworth Dichotomous D-15 test [8] to identify both the greatness of color vision deficiency quantified by the number of errors (maximum value, n. 15) and the type (deutan, protan, tritan) and the City University test [9] both binocularly and monocularly, to identify the type and degree of acquired red-green color vision deficiency (maximum value of error number, n. 6) and blue-yellow color vision deficiency (maximum value of errors number, n. 3). Selecting exclusively male patients avoids Lyon genetic phenomenon [24] present in heterozygous females who mimic color vision in different degrees. All patients and controls were examined by an ophthalmologist to out rule diabetic retinopathy, cataracts, optic neuritis, senile maculopathy or ocular fundus anomalies.

## **Results and Discussion**

4 inherited colorblind patients were excluded from the results' analysis so as one who was affected by diplopy. 27/45 Parkinson's disease patients had normal color vision, 8/45 showed slight or mediocre red-green deficiency, 5/45 had blue-yellow deficiency, and 5/45 had both red-green and blue-yellow deficiencies, 45 Essential tremor patients showed normal color vision.

Reduced color vision is reported in patients with Parkinson's disease because of the abnormal phosphorylation of human alpha-synuclein localized to the inner retina cells. According to Haug [14], the influence of Parkinson's disease is most noticeable in the pathway of short wave cones because the short wave cones are widely separated. In the retina, the

small bistratified ganglion cells, which are the morphological substrate of the short wave cone pathway have much larger receptive fields than the midget ganglion cells and may be more dependent upon long range spatial interactions mediated by dopaminergic interplexiform or amacrine cells. Retinal parvocellular, koniocellular, and magnocellular damage in Parkinson's disease also confirms the impairment by long wave contrasting with the pathway typically seen in aging. Parkinson's disease affects dopaminergic substantia nigra's neurons within the brain system, and the decline in color vision may be predicted by the level of retinal dopamine deficiency [2; 3; 31].

The involvement of an olivo-cerebello-rubral-thalamic loop does not seem to influence color vision in Essential tremor [32].

The comparison in our study confirms the reliability of the simultaneous use of three tests identifying and scoring the acquired color defects in some neurodegenerative diseases, while also highlighting real brain manifestations.

### **1.3. Color Vision Deficiency by Metabolic Disease: The example of Diabetes**

According to a disordered metabolism of neural cells, diabetes may damage the nerve directly or indirectly through changes in the microcirculation. Retinal functions damaged by diabetes in the first instance by a nutritional deficit which affects the neurons and later by vascular disturbances associated with micro-aneurysms. Blood and exudative masses which may displace tissue, later damage sight and the coagulation of blood vessels used as a treatment can itself affect color and form vision. Lakowski [21] reviewed the history of color vision losses associated with diabetes and gave an analysis of different test methods in many cases; in general, blue losses are most severe. They stressed the fluctuating nature of some color vision difficulties, an the event of variation of blood sugar level. It was seen to be difficult to predict the state of vision or retinal disturbance from either normal or abnormal color vision but a useful approach was found to be a combination of data from different color test [12].

### **Patients and Method**

Eleven Calabrian male patients (mean age  $70.72 \pm 7.79$ ); mean duration's disease  $10.81 \pm 7.27$ ; mean glucose  $153.22 \pm 128.66$ ) admitted to the Istituto Nazionale Riposo e Cura per Anziani (INRCA), Cosenza (Calabria, Southern Italy) were enrolled. Twelve controls were matched for age and sex (mean age  $65.25 \pm 27.82$ ; mean glucose  $104.10 \pm 30.1$ ). Patients and controls signed their informed consent. An ophthalmologist examined all patients and controls in order to rule out cataracts, senile maculopathy, or ocular fundus anomalies. One inherited colorblindness subject was excluded by the analysis. The analyzed sample was 23 subjects. Fixed sampling of males allowed us to avoid the Lyon genetic phenomenon [24].



All patients and controls underwent Ishihara test [16], Farnsworth dichotomous D-15 test [8], the City University test [9].

## Results and Discussion

No correlation existed between duration of diabetes and number of errors by tests. The positive polarity of the normal error scores (range, 226.6-390.0) in six out of 11 patients was determined by a no any ambiguity in the axis of color confusion that was distinguished sharply by the City University test [9]. Furthermore, this test supported greatly the other two utilized tests for a better investigation during the acquired color vision screenings [34].

Diabetes also induces morphological changes in neurons, including synaptic vesicle depletion in massy fiber nerve terminals, dendritic atrophy of caspase 3 pyramidal neurons, and increases the expression of the presynaptic synaptophysin. Caspase 3 has been demonstrated to be a downstream effector of phosphorylated p38MAPK (pp38) which causes neuronal death induced by high glucose [28]. Retinal neurons may also be affected by diabetes, even before the detection of microvascular dysfunction. Diabetes increases apoptosis in neural cells in human retina early in the course of the disease. Further, there may be a primary neurodegenerative process which contributes to loss of vision in diabetic retinopathy. Neuroprotection in diabetic may be a valuable therapeutic target [13; 26].

Acquired type 3 (tritan) color deficiency, or blue-yellow deficiency, in diabetes in 1954 by Dubois-Poulsen [7] and also prior to the onset of visible retinopathy. This occurs in association with reduced sensitivity in the short-wavelength (S cone) pathway. Generally, in later stages of the disease, the red-green mechanisms are involved. This differentiation can be based on the different answers by our brain when some insult or diseases are present. The brain analyzes and thus determines the color of a surface by determining the color of every point in it by an additive mechanism, that is, by gauging the amounts of long, middle, and short wave light reflected from each point. The reason is to be found in the anatomical connections between the eye and the brain. These are organized topographically, with every point in the primary visual cortex which, until the last two decades, was considered to be the sole visual perceptive cortex and remains perhaps, even today, the more extensively studied part of the visual cortex [2; 10; 36; 49].

## 2. Congenital Color Vision Deficiency

### Introduction

Dichromatisms of the red-green type were the first conditions to be recognized. The fascinating history of the emergence of distinctive terms has been described by Helmholtz [10] with important contributions to the text by von Kries [52]. Wilson, Judd and Wasserman

extended this literature significantly [17; 52; 54; 56]. As long ago as 1811 Wardrop [10] distinguished the two red-green types of deficiency and Seebeck [38] showed how those relatively insensitive to red lights were in a minority. Von Kries proposed the name protanopes for those who *lack the first component ... of the normal*. His term deuteranopia and tritanopia followed the trichromatic approach which Maxwell supported [27].

Dalton's suggestion [10] that an unusual coloration of his ocular media had been the cause of his condition had been disproved at autopsy. However his explanation of a color vision deficiency by absorption (some wavelengths being absorbed in the ocular media before reaching a normal retina) should be noted for it explains some acquired defects. An alternative cause, proposed by von Kries [52] arose from dissatisfaction with the term red-blindness; he suggested that *a reduction of the normal retinal mechanisms could account for the visual deficiency*. The loss of one mechanism is compatible with modern concepts of dichromatism. This fact emerged as an understanding of the cone pigments developed, as will now be described.

The suggestion that variations in color vision might result from individual differences in receptor sensitivity was made by Konig and Dieterici [19]. Konig proposed the absence of one photopigment class in the fovea of the dichromat to explain defective color vision in association with the other unaltered pigments. This followed Young [53] who first propounded that the red mechanism might be lacking in Dalton's eye as an explanation of his protanopia. A century and a half passed before Rushton [37] provided objective evidence with the absence of the erythrolabe pigment in protanopia by measuring the remaining pigment, chlorolabe in situ. Photopigment characteristics were related to spectral sensitivity by Konig and Kottgen [19] who indicated how the protanopic sensitivity curve corresponded to the green sensation of normal vision.

Dichromatism is usually divided into three categories all of which involve severe difficulties with colors according with the inheritance modality, too [30].

- Protanopia. This condition is particularly associated with deficiency of appreciation of red. It is likely to arise from absence of the longwave sensitive cone pigment, erythrolabe. The inheritance is on X chromosome [28].
- Deuteranopia. While being associated with deficiency of green vision, this type of dichromatism involves characteristic confusions of colors. The inheritance is on X chromosome [28].
- Tritanopia. This is a very rare condition, where sensitive to blue is impaired and where there are confusions between blue and green at one time doubted its existence. The inheritance is on chromosome 7 [28].

## 2.1. Color Vision Inherited Deficiency in Calabria, Southern Italy

The locus of green and red color vision is on the X chromosome, on the distal side of q arm, Xq28. The color vision anomaly relative to these two colors is described as mild or great green color vision deficiency (G', deuteranomaly and G-, deuteranopy, respectively) and mild or great red color vision deficiency, described as R', protanomaly and R-, protanopy, respectively. The blue color vision deficiency is very rare, and it is inherited as an autosomal dominant character whose locus is in chromosome 7 [28; 45].

### Patients and Method

Screening procedures to identify colorblindness frequencies in continental Italy have included samples greater than 500, all male students attending secondary school.

Screening for green-red colorblindness was carried out on 14,177 males, 11-25 years of age, residing in Calabria. The sample included three different groups: 3,567 students attending secondary school in the Cosenza province, 560 students attending the University of Calabria, and 10,050 military conscripts from the three Calabrian provinces. 980 non-Calabrian and 125 foreign subjects were eliminated. Thus, the analysis was carried out on 13,072 subjects who had both maternal grandparents born within 409 Calabrian towns.

Frequencies of colorblindness in the Albanian ethnic minority in 19 towns of Cosenza and eight towns of Catanzaro, and in the Grecanonical ethnic minority in five towns of Reggio Calabria, were carried out separately.

The screening method to identify red-green colorblindness is Ishihara test [16].

All colorblindness subjects underwent an optic visit to eliminate pathologies that would produce false positive results. One colorblind patient had an acquired retinopathy and was not included.

### Results and Discussion

The frequency of colorblindness in the Calabrian indigenous population is 5.25%. The mean frequency of colorblindness in the Albanian ethnic minority of Cosenza and Catanzaro provinces is 7.40%. The small sample of subjects from the Grecanonical ethnic minority in Reggio Calabria does not permit an estimate of a valid frequency.

The three Calabrian provinces, with the exception of the ethnic minorities, show a decreasing trend in the frequency of colorblindness from North to South: Cosenza, 6.23%; Catanzaro, 4.65%; Reggio Calabria, 3.43%. The frequency, 7.40%, in the total Albanian ethnic minority is different from that, 5.43%, of the indigenous population living within the internal area where the Albanian towns are located.

The phenotypic study of colorblindness affected subjects in all of Calabria shows a frequency of 83.05% for the green color defect (deuteranomaly + deuteranopy). The red color defect has a frequency of 16.90% (protanomaly + protanopy) [45].

It is possible that the results detailed in the large territory of the Calabria region were obtainable only by studying a large number of subjects who could be subdivided by region. Calabria region is a useful example in which to make such a detailed analysis, due its population heterogeneity, geographic variation, and variability of ancient historical events. From the VIIIth to the Vth centuries BC, Calabria was colonized by Greeks who established, in a special way along the Ionian coast, some important and populous urban settlements, such as Sybaris, Kroton, and Locrys. Subsequently, the territory was occupied by other groups: Romans, Longobards, Byzantines, Normans, Arabs, Hispanics and French. Improvement of the roads during the period following World War II allowed better communication between Cosenza, Catanzaro and Reggio Calabria. The population living within the three Calabrian provinces do not have a different style of life, but geographically the three provinces are different, because both the passive geographical barrier and active barriers [45].

## **2.2. Color Vision Inherited Deficiency and the daily occupations: car driving example**

Steward and Cole reported a range of color-related difficulties experienced in everyday life. In one of the most important studies in the field they administered a questionnaire to 102 people with defective color vision and to an equal number of people with normal color vision [43].

### **Patients and Method**

Groups of subjects with either defective color vision or with normal color vision were compared in a mass screening performed to identify the frequency of colorblindness in the province of Cosenza (Calabria, Southern Italy). From 1987 to 1991, 4,194 male students (age range 11-14 years) were screened using Ishihara test [16]. All the students came from 13 of the 155 towns in the province of Cosenza, and attended secondary school. Of the 4194 students, 268 (6.0%) had defective color vision. The samples of subjects with defective color vision and subjects with normal color vision were matched for age ( $21.4 \pm 1.3$  years, and  $21.2 \pm 1.3$  years, respectively).

In 2001, after sufficient time had passed for the subjects to reach maturity, we tried to find all 268 subjects with defective color vision by telephone in order to administer a psychosocial questionnaire by telephone interview to ascertain the difficulties associated with defective color vision in terms of daily life and driving a car. We compared the answers of 151 subjects with defective color vision with those of 302 subjects with normal color vision selected as follows: for each subject with defective color vision, two subjects with normal color vision

randomly chosen from among the classmates of the subject with defective color vision at the time of screening who came from the same town. So, we reduced any bias introduced by age or socio-cultural factors.

## **Results and Conclusion**

Regular use of a car was significantly less common among colorblind subjects than among people with normal color vision. People with defective color vision stated that, in daily life, they preferred daytime driving over night-time driving compared with people with normal color vision. At night, more subjects with defective color vision had difficulty identifying the reflectors on the road compared with people with normal color vision, and in identifying the lights of the car ahead but these differences were not statistically significant. There were no differences between the two groups in identifying the colors of traffic light signals. When the relative positions of the traffic lights were changed, 4.8% of colorblind subjects had difficulty identifying the colors compared with 2.0% of subjects with normal color vision. The frequency of road accidents was also similar for both groups. No accidents had occurred while the driver was under the influence of drugs [46].

Colorblind Calabrian subjects were more psychologically subordinate in their choice of work compared with orthochromatic subjects. In fact, the former tended to opt for more subordinate work than the latter. Colorblind Calabrian subjects stated that their abnormality did not influence their choice of career. The different strategies that these colorblind subjects adopted to avoid various difficulties suggest that they did not consider their abnormality to be a real handicap [47].

### **2.3. Color Vision Inherited Deficiency and social, psycho-pedagogical features**

The individual is an unbreakable unit, each disease or anomaly, in fact, provokes behaviors involving not only the biological structure, but also the psychological and social ones. From this perspective, the colorblind subject is an individual who, within the community in which he lives, ascribes peculiar symbols to the different chromatic elements of the natural environment comparing them with emotional expressions which are different from those of orthochromatic subjects. Colorblind subject can become discouraged and can develop poor self-confidence. Some authors confirmed that colorblindness causes poor development of a color defective's personality and others confirmed that this anomaly can cause imprinting difficulties within the brain's cognitive areas which are apt for this function. Some authors emphasized the relationship between colorblindness and a defensive personality of young adults, and some studies examined some relationships between the genetic features of all the dyschromatopsies and some depression's forms [4; 29; 39; 40].



## Patients and Method

A psychological mass study was carried out in Calabria, Southern Italy between 1998 and 2000 on 63,933 colorblind schoolchildren of both sexes [11; 44].

A psychological questionnaire with 18 items was simultaneously administered to the subjects whose aim was: to verify preferences or difficulties in studying the different discipline; to compare the subjects' own skills and the obtained performance; to quantify parental satisfaction or dissatisfaction; and to gain insight on the placement of color defectives within the school contest. Further questions dealt with the ability to recognize the natural color and the possible repercussions in both the scholastic environment and in the daily life.

The pedagogical questionnaire consisting of 13 closed-open questions, was submitted to 3,082 teachers of different subjects in order to analyze the knowledge about the anomaly, their degree of consciousness about the colorblind subject, the behavior against a case of colorblindness and how frequently color is used as an element of didactics.

## Results and Discussion

This study shows that the mean frequencies of colorblindness calculated among 32,322 males extrapolated from the total sample was 4.8% in Calabria and 4.8% in Basilicata.

The psychological questionnaire showed that more than 40% of the subjects had scholastic tasks in which different colors were often present and in these cases it was necessary to be able to make a good identification of the colors. When an exact matching by age was not possible, the colorblind subjects proved to be tendentially and significantly older than the orthochromatics of the same class. Colorblind schoolchildren showed more learning difficulties than the orthochromatics. According to 44% of the colorblind subjects, school is a place made for teaching and not for learning, while among the orthochromatics this percentage decreases to 38%; the average level of satisfaction about their own school is around two tenths lower among the anopes. The colorblind subjects tend to be less satisfied about their school achievement in relation to their skills and they are significantly less satisfied about their achievement related to their actual commitment. Colorblind subjects showed a tendency toward lower perception of their skills and a significantly higher dissatisfaction with their school achievement [40; 42; 50].

The teachers' questionnaire showed that: 39% declared to possess a personal knowledge of the anomaly; 58% had some general information; the remaining 3% didn't know the exact meaning of the term colorblindness. In the same percentage, the teachers considered colorblindness to be both an "anomaly" or a "disadvantageous situation". They also felt the term signified a "biological and medical fact" of interest to the respective specialist. At least

30% of the teachers that colorblindness recognition is important both from a didactical and a formal point of view. With regard to teaching, 37% of the teachers utilized colors as basic elements of their didactic activity, while 63% of them did not.

From the results of the present it is possible to infer that a subject could develop actual social limitations as a result of dyschromatopsia turning a common impairment into a real handicap [25].

Colorblind students have difficulties in recognizing the colors with respect to the orthochromatics more learning difficulties and are lesser satisfied with their scholastic achievements when compared with the orthochromatics. In fact, they are persuaded that the school is only a teaching environment instead of a learning environment, which confirms their passive condition in the diligence required by the different disciplines. Particular importance must be attached to the perception of colorblind subjects that they are unable to satisfy the parents expectations and the discomfort provoked by the colors which are normally present in common objects [6].

We affirm that the repercussions of color vision deficit may lead to an imbalance among the three elements that comprise health; psychological and social factors, instead of reducing and compensating for the biological factors amplify the limiting consequences [20; 55].

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