

UNDERSTANDING AND COPING WITH ACHROMATOPSIA

by
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Second Edition

**A Publication for
The Achromatopsia Network
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Understanding and Coping with Achromatopsia (2nd edition)

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The information presented in this book has been gathered from many sources including input from members of the Achromatopsia Network, library research, and consultations with specialists in ophthalmology, optometry, vision research, and other fields. On page 140, there is a list of references which have been especially helpful in preparing the material included in this book. The author, Frances Futterman, serves as editor and reporter for the Achromatopsia Network Journal (formerly known as the Achromatopsia Network Newsletter).

The Achromatopsia Network is an information and support network for individuals and families who are affected by inherited, congenital achromatopsia.

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What Is Achromatopsia?

Congenital achromatopsia is a rare hereditary vision disorder which affects 1 person in 33,000 in the U. S. The incidence varies in other parts of the world.

Persons who have achromatopsia have little or no “cone vision.” In normal eyes there are 6 million “cone” photoreceptors, located mostly at the center of the retina. Achromats must rely on their “rod” photoreceptors for vision. In normal eyes there are 100 million rods, located mostly at the periphery of the retina. Rods “saturate” at higher levels of illumination. They do not provide color vision or “detail vision.” Achromats are, therefore, either totally – or almost totally – colorblind, have poor visual acuity, and are unable to adapt visually to higher levels of illumination. There are variations in the severity of these symptoms among individual achromats. There are complete rod monochromats, incomplete rod monochromats, and blue cone monochromats.

The word achromatopsia means “without color vision.” Monochromacy means to see in shades of only one color. Many people assume that rod monochromats, lacking normal color vision, see in shades of gray. However, complete rod monochromats report that the word “gray” has no more meaning for them than do any of the names for all the other colors they cannot see. They perceive many gradations (shades) from white to black; and, unless they are being required to use correct color names to communicate with normally sighted people, they tend to describe something as “darker than” or “lighter than” something else.

Since cones are concentrated in the macular (central) area of the retina, they are responsible for good central vision. A deficiency in cones results in sub-normal visual acuity which cannot be corrected with Rx lenses. Near vision is

less affected than distance vision. The average visual acuity of persons with achromatopsia is from 20/160 to 20/200, using standard vision charts. Many achromats also have refractive vision problems, ranging from extreme nearsightedness to extreme farsightedness. Many of them have a significant degree of astigmatism, more than is found in the general population. Forms of strabismus (eye turns) are also common.

In eye exams vision care specialists look for the following symptoms and signs when diagnosing achromatopsia:

- ☆ **Poor visual acuity (detail vision)**
- ☆ **Photophobia – aversion to light**
- ☆ **Little or no color perception**
- ☆ **Normal or nearly normal fundus (the back of the eye)**
- ☆ **Stable condition (present from early infancy and not progressive)**
- ☆ **Abnormal or absent photopic ERG (electrical recording from the eye under light conditions in which the cones would normally function)**
- ☆ **Normal ERG for rod vision**
- ☆ **Nystagmus – “Shaky eyes”**

The nystagmus varies with:

- 1. Distance: more nystagmus when trying to focus in the distance**
- 2. Light level: less nystagmus in low light, more in bright light**
- 3. Fixation: nystagmus is worse when trying hard to see.**
- 4. Age: usually more pronounced in infancy and childhood and tends to diminish with age.**

There is a rare medical condition, cerebral achromatopsia, which is an acquired form of total colorblindness and which can occur as a result of stroke, trauma, or some other cause. Persons with this condition do see in shades of gray, because they previously had color vision and so are able to perceive the absence of color as gray.

What Is It Like to Have Achromatopsia?

People with achromatopsia report that being colorblind is, by far, the least troublesome manifestation of this vision disorder. Far more serious to cope with are the poor visual acuity (especially for distance) and hypersensitivity to light, sometimes called photophobia.

Strictly speaking, photophobia means “fear of light.” However, anyone familiar with this term knows it does not refer to an actual fear, as do other terms incorporating “phobia” (such as claustrophobia). Achromats’ hypersensitivity to light occurs because they have only rod vision, and rods saturate at higher levels of illumination. Because of this, the visual experiences they have in higher light levels can be compared to what normally sighted persons experience momentarily when changing abruptly from a darkened environment (to which their eyes have adapted) to a high level of illumination. Examples: (1) when exiting a dark theater and facing bright sunlight, (2) when being awakened from sleep in a dark room by the turning on of bright overhead lights, and (3) when moving from a photographer’s dark-room into a brightly illuminated adjoining room. In such situations, the eyes of normally sighted persons very briefly experience rod saturation or “dazzling,” but then they quickly adapt to the bright light, through the normal interaction of rods and cones.

Not having cone vision, achromats must use tinted lenses and other strategies to adapt to higher levels of illumination. Their vision decreases as lighting increases and increases as lighting decreases. The effect of light on their eyes can cause considerable stress, but they do not experience the discomfort or pain that accompany the photophobia experienced by persons who have

certain other eye conditions. Light does not “hurt” their eyes, so to speak. However, some achromats report that they have experienced pain or discomfort in connection with having to strain to see and function in adverse conditions. But this is visual stress, not a manifestation of achromatopsia.

How well achromats are able to see can change continually (and sometimes significantly) with every change in illumination. Depending on the quantity of light, the quality of light, the direction of the light, and other factors, achromats can experience reasonably good vision, then seriously limited vision – and then can be very nearly blinded by light, as they move from place to place.

They usually do what they can to make their homes and work spaces as visually comfortable as possible. But, when the possibilities for making adaptations are restricted – as is the case with classrooms, public places, or rooms that must be shared with other persons who have entirely different visual requirements – then achromats adjust in whatever ways they can. The more involved they are in activities away from home, the more they have to cope with this variability in their visual functioning. Some choose always to carry with them several pairs of tinted lenses (from lightly tinted lenses to very dark lenses) in order to be able to maximize their vision wherever they go.

Under favorable conditions (subdued indoor lighting or outdoors at twilight or night), achromats do not experience being visually disabled. Only tasks that require color vision or good detail vision cause them to be mindful of their vision disorder in such situations. An exception to this rule is that those who become dependent on very dark lenses, whether sunglasses or tinted contacts, sometimes go on wearing tinted lenses in lower light levels and so do not have

as good vision in these settings as do those who can comfortably remove their tinted lenses whenever they are not in bright light.

When factors of illumination are changing rapidly, as when one is moving from one indoor space to another or from indoors to outdoors and back again, it is usually simpler just to accept the varying levels of visibility one experiences while wearing tinted lenses than to keep switching from one pair of lenses to another. Each individual develops his/her own system regarding when, where, and how often to switch from one tint to another and when to go without tinted lenses.

Achromats are acutely aware of the wide range of illumination levels that are encountered in this world. At the “friendliest” end of the spectrum are dimly lit restaurants, theaters, bars, and basements. At the other end of the spectrum are playgrounds, beaches, and large expanses of pavement, snow, or water during the daytime.

Some indoor spaces where achromats spend time in connection with work, school, or social activities are very bright. Rooms with large, unshaded windows, white or pastel furnishings and carpets, or ceilings and walls that are white or nearly white do not offer achromats the chance to relax and enjoy the improved vision which they are capable of having indoors.

Individual Differences

No two people with this vision disorder would give the same answer to the question, “What is it like to have achromatopsia?” This is because there are so many variables in people’s lives. One of the most significant variables is the eye condition itself. Some networkers are complete achromats, having total colorblindness, severe light sensitivity, and low visual acuity even under the most favorable lighting. Others are

incomplete achromats, with some color vision, less photophobia, and somewhat better acuity. Some achromats have very noticeable nystagmus (shaky eyes) and/or strabismus (eye turns), while others have a more normal appearance to their eyes, at least when they are in favorable lighting.

And our vision problems constitute only one aspect of who we are as individuals. All of the other aspects of who we are represent other sets of variables. How we experience the impact of achromatopsia in our lives is influenced by our aptitudes, dispositions, strengths, and weaknesses. We have different types and degrees of sensitivity. We are as diverse as any other set of people on this planet.

Also, the circumstances of people’s lives have considerable influence in determining how difficult or limiting having achromatopsia can be. For example, someone growing up under adverse conditions, with little or no access to adaptive aids or assistance in coping with vision problems will experience achromatopsia very differently from someone growing up with a caring family and in visually comfortable surroundings, provided with special services and adaptive aids. Some network members have had far greater support systems than others.

Since achromatopsia is so rare, many achromats have never had the chance to meet someone else with the same condition, while others have had a lifelong experience of knowing someone who can understand, because they have grown up with one or more siblings with achromatopsia.

Because of all of these variables in people’s lives, it makes no sense to compare people in terms of their level of accomplishments, their degree of “adjustment” to their vision disability, or anything else.

Comparing Achromatopsia with Other Vision Disorders

There are various eye conditions which involve hypersensitivity to light (doctors call it photophobia), but the light sensitivity achromats have is the most severe that can be experienced visually. Consequently, the tinted lenses that work well for achromats may not work well for other light sensitive persons – and vice versa.

There are also many eye conditions which involve defective color vision or loss of color vision. For persons with achromatopsia, it is not a matter of color vision having been “lost.” Our color-blindness has been part of the way we have seen all our lives and is due to the congenital absence of certain photo-receptors in our retinas (the cones).

The range of visual acuity reported for achromats (usually 20/160 to 20/200) is the same as that reported for many persons affected by other serious vision disorders, even though the vision of these other persons is often more seriously impaired than ours. (Some blue cone monochromats and incomplete rod monochromats have better visual acuity than 20/160, and some complete rod monochromats have worse visual acuity than 20/200.)

So it is possible for someone to be very light sensitive, have seriously defective color vision, and have 20/200 acuity, yet have an eye condition very different from achromatopsia. Hence, the need for good diagnosticians.

The way achromats see is unique. Because of this, the adaptive devices, methods, and materials needed by persons with other vision disorders are often not appropriate for achromats.

We experience severe vision impairment in high levels of illumination but remarkably good vision in favorable lighting. By contrast, many persons who are visually impaired due to other

causes experience poor vision in all settings, with no periods of being able to feel and perform much like normally sighted persons, as we are able to do in lower levels of illumination.

Most persons who are visually impaired due to other causes see worse in low lighting. Achromats see better.

Many low vision persons have experienced sudden or progressive loss of vision and need to prepare for further vision loss in the future (for example, they may need to begin learning Braille). But persons whose only vision disorder is achromatopsia do not need to adjust to progressive vision loss nor to prepare for further vision loss.

When considering what kinds of adaptive devices are appropriate for a vision impaired individual at school, at work, or in any other setting, decisions should be based on how that individual actually sees and functions and not on the assumption that all low vision persons need the same kinds of equipment, materials, etc. Achromats have a great deal of usable vision, and they tend to make very good use of it. They may or may not welcome enlarged print, for example. Many prefer to use standard reading materials held close. They may reject optical devices or use them only occasionally. Some of the adaptive devices and materials needed by other visually impaired persons – such as broad felt tip pens, bright halogen lamps, and paper with very bold lines – are not appropriate for achromats.

Some of the technology used by other low vision persons is not needed by achromats and can, in fact, restrict, rather than enhance, their visual functioning. The preferences of individuals with achromatopsia should be given the highest consideration when decisions are being made about these matters.

Vision and Coping Strategies of a Complete Achromat

Following are excerpts from chapter 8 of *Night Vision*, a vision science book that was published in 1990. This chapter, entitled "Vision in a Complete Achromat: A Personal Account," was written by Dr. Knut Nordby, who is a complete rod monochromat and a highly esteemed scientist. He was educated at the Institute of Psychology of the University of Oslo, where he studied science, music, physiology, and sensory perception. He went on to teach at this university and later became a Senior Research Scientist with the Norwegian Telecom Research Department, developing telecommunications equipment and services for disabled people. He has done extensive vision research in collaboration with scientists in England, Germany, Canada, and other countries.

In these excerpts Dr. Nordby tells about visual strategies and other methods he uses to cope with his hypersensitivity to light and low visual acuity:

.....
As far as can be determined, the retinae of my eyes do not contain any cone-receptors at all, only rod-receptors; or cones are present but in such reduced numbers that they do not contribute to the visual process in any measurable amount. Since the rods are much more sensitive to light and also saturate at lower light intensities than the cones, my visual system is well adapted for vision only under low light conditions. In fact, my vision will not function at all in very bright light (e.g. out-of-doors in full daylight) if I do not adopt specialized visual behavior and strategies.

I am easily dazzled and, in effect, blinded if exposed to bright light. If I open my eyes fully for more than one or two seconds under such conditions,

the scene I am gazing at is quickly washed out. It turns into a bright haze, and all structured vision is lost. It can be very distressing for me, and sometimes even painful, to perform demanding visual tasks in very bright light.

This hypersensitivity to light is often referred to as photophobia, but it has nothing to do with the irrational psychodynamic "phobias." In fact, I really enjoy being out in the warm sun, provided I don't have to perform exacting visual tasks. I do not like to read or write in the sun; digging the garden or mowing the lawn, on the other hand, is no problem for me, even in bright sunlight.

My main problem, then, is to restrict the intensity of the light entering my eyes. This can be achieved in several ways and I tend to use them all, alone or in combination, according to what the situation demands. In common with nearly all the other achromats I have interviewed, I have developed special visual strategies for restricting the amount of light entering my eyes.

The most obvious strategy is, of course, simply to avoid direct strong light. Staying indoors or in the shade is one way of achieving this, if there is no special reason for being in intense sunshine. Indoors, whenever possible, I try to place myself with my back towards bright windows and strong light sources and avoid having direct sunlight falling on my workplace.

Shading my eyes from direct, intense light with my hand or a visor may be necessary. Ordinarily I wear sunglasses; the lenses of my glasses are photochromatic (i.e. tinted lenses which darken in bright light). Out-of-doors I often wear an extra clip-on polarizing filter for cutting down on bright light and for dealing with

visually destructive glare and reflections from shiny surfaces. The best sun filters I have tried are the special colored glasses which are made for the retinitis pigmentosa patients. They give a very pleasant light attenuation, but they are socially less attractive because of their red appearance. If I must see small detail in bright light, I turn away from the light, putting the material in my shadow.

The most typical visual strategies I resort to are squinting – i. e. partially closing the eyes and looking through the narrow slits formed by the eyelids, and frequently blinking the eyes. This habit seems to be universally resorted to by typical, complete achromats. If light levels rise, my fully constricted pupils cannot further contract and I have to squint to deal with the higher intensities and to avoid saturating the rods. At higher light intensities even this is not enough, and I also have to start blinking my eyes to shut out excessive light. My blinking is triggered when saturation sets in. The blinking frequency is slow at first, only once every four to five seconds, but increases with increasing light intensity to three or four blinks a second.

At lower light levels, when the blinking rate is slow, the blinks themselves are also rather brief. As light levels go up, the blinking rate also goes up, and the blinks become longer. At the very brightest light levels where my visual system will function (e.g. new snow in bright sunlight) the blinks are so long that my eyes are, in effect, shut most of the time, except for brief opening blinks once every two to three seconds. Whether blinking my eyes briefly at low light levels or extending the length of the

blinks at high intensities, I still experience a visually stable world in which I can orientate myself and move about.

This squinting and blinking behavior is a strain socially. In bright light people immediately notice that something is wrong with my eyes and show this by their reactions to me. As a child I was often approached by complete strangers who demanded to know what was wrong with my eyes. Wearing dark glasses or clip-on sun filters can alleviate this social burden to some extent.

NIGHT VISION

basic, clinical and applied aspects

Edited by R.F. Hess,

L.T. Sharpe, and K. Nordby

Cambridge University Press, 1990

At higher light levels the peripheral visual field is much more affected than the central part of the field. I can still

detect movement in the far periphery, but I have much more difficulty in identifying what is moving and then reacting adequately to it. This makes me move in a rather hesitant and stiff manner, sometimes bumping into people, and to be overcautious when moving about in agitated surroundings or when encountering unmarked steps and stairs. As soon as I am in the shade or indoors, I again move in a much more relaxed and confident way.

It is very clear to me that the most debilitating, handicapping, and distressing consequence of the achromatopsia is my hypersensitivity to light. This is also the unanimous verdict of all the achromats I have interviewed so far. The practical problems of being dazzled, the narrowing of the visual fields, and the social problems of light aversion and of feeling clumsy in intense light is frequently reported by my informants as being more of a hindrance than not being able to experience colors or to discern minute detail.

In a retina where only rods function, visual acuity is drastically reduced, since it is the densely packed foveal cones that are responsible for the high acuity of central vision in the normal retina. My acuity is 6/60 (20/200) using the Snellen chart. This is 1/10th of normal acuity. This low acuity is common for all typical, complete achromats reported in the literature and all whom I have interviewed. Under optimal lighting conditions, however, my visual acuity has been measured as somewhat higher.

My visual acuity varies quite a bit, depending on the illumination. At higher luminances it quickly deteriorates, but it improves at lower light intensities, such as (1) at dusk, after the sun has set but before it gets really dark, (2) indoors with curtains drawn during the daytime, or (3) in the evening with not-too-strong incandescent, tungsten illumination.

I experience a visual world in which things appear to be well-focused, have sharp and clearly defined boundaries, and are not fuzzy or cloudy. I can easily tell the difference between what people with normal vision call a well-focused photograph and one that is not so well-focused. Once when I was giving a talk at a university, one of my slides came up slightly out-of-focus. I quickly refocused it myself, and one of the professors attending commented that, to his astonishment, I had done this as well as any projectionist.

Details that are too small for the low resolution of my coarse retinal matrix will disappear, blending into the background; but, when these details are brought closer and become large enough for me to discern, they are just as sharp and well-defined as other objects.

Together with my spectacles, a small, pocketable magnifier is the most important tool I have. I always carry one with me. I have considered other types of

visual aids, such as video systems, etc., but they all seem to fail on one or more points. Usually they are too large and heavy to carry around, too complicated to use, or too powerful, giving me too small a field of view, making it hard to read continuous text.

Typing used to be quite a bother, since I had to lean over the typewriter with my magnifier to be able to read what I had typed, but now I use a computer and a word processing program for all my typing. To overcome my low visual acuity, I get close to the screen, and I have a large (19") screen installed. My printer allows me to have all my documents printed in large type-face.

I cannot easily identify people by their facial features unless they are close. Those who do not know about my visual handicap may think that I am being aloof or downright rude. Often I fail to recognize people I know, with embarrassing consequences. Even those who know about my vision may be offended. This may be because my visual behavior is often rather normal.

I often experience negative attitudes if I expose my visual disability before people get to know me, so there are times when I avoid showing my visual handicap and simulate normal vision. If I show my handicap to people who do not know me, they tend to categorize me as a disabled person, and I may be treated in a patronizing way. If I wait to expose my visual handicap to people after they get to know me, I get fewer such reactions.

In spite of my visual handicap and all the practical and social problems I encounter, I feel that I live a very rich and interesting life. I hope that these very personal and rather private observations I have described will prove to be of value to vision researchers and people dealing with typical, complete achromats.

Inheritance Factors Associated with Rod Monochromacy

by Frances Futterman

The 2 major forms of achromatopsia – rod monochromacy and blue cone monochromacy – have quite different inheritance factors.

With rod monochromacy – the form which affects most members of our network – there is no family history of the defect, with the exception of certain unusual circumstances, such as are mentioned in the first paragraph on the following page. Males and females are equally affected. Persons with this vision disorder have inherited 2 faulty genes, 1 from each parent. This is known as autosomal recessive inheritance.

When both parents carry the faulty gene but do not themselves have rod monochromacy, the odds are 1 in 4 that a child born to them will have the vision disorder.

The birth order of affected and non-affected siblings in network families illustrate the range of possibilities in keeping with these odds. In some families 3 normally sighted children have been born before a child with achromatopsia was born. In others the second-born child or the third-born has been the one to have achromatopsia. In one family, 3 children out of 8 have achromatopsia. In several families, 3 children in succession have been born with the condition. There are twins in which one child has it and twins in which both children have it.

A rod monochromat does not risk passing on this condition to offspring unless he or she happens to mate with another carrier. If a rod monochromat were to mate with a carrier of the same faulty gene who does not have the condition, the odds would be 50-50 that offspring would be affected. If two rod

monochromats were to mate, all of their children would be expected to have the condition. If the child of a rod monochromat were to mate with a carrier of the same gene, the odds are 1 in 4 that offspring would be affected and 2 in 4 that offspring would not even inherit the faulty gene.

Blue cone monochromacy, an especially rare form of achromatopsia (BCM affects less than 5% of network members) has entirely different inheritance factors. Information regarding the genetics of blue cone monochromacy is presented on pp. 12-13 of *Understanding and Coping with Achromatopsia*.

The following information about concerns pertaining to reproduction applies only to autosomal recessive achromatopsia (rod monochromacy).

No rod monochromat in our network is known to have children or grandchildren with this condition, and none is known to have parents, grandparents, or ancestors affected by the condition. All of this is in keeping with the known inheritance factors. In fact, one of the ways diagnosticians can determine that a patient has rod monochromacy rather than blue cone monochromacy or certain other retinal conditions is by noting that the condition has not “run in the family.”

I have rod monochromacy. As a young adult, I consulted my eye doctor regarding my chances of passing on this eye condition to any children I might have. I was reassured that I need not worry about this, because it was highly unlikely that I would mate with someone carrying this rare gene.

The incidence of rod monochromacy is 1 in 33,000 births in the U.S. and in

most other parts of the world. It is rare for someone to carry the gene and even rarer for two persons carrying the gene to mate. The likelihood that two persons with the same gene will mate is higher if they are related or if they live in a tightly knit or geographically isolated community. Thus, there is a higher incidence of rod monochromacy in certain parts of the world, such as the tiny atoll of Pingelap and an enclave inhabited by Pingelapese on Pohnpei Island (as described in Oliver Sacks' book, *The Island of the Colorblind*).

Based on what is known about autosomal recessive inheritance,* the mating choices for rod monochromats which would bring risk of passing on this disorder would include the following:

1. Mating with another rod monochromat: all children would be expected to have rod monochromacy if the same abnormal gene is involved.

2. Mating with a known carrier of the same faulty gene (50% chance of passing on the defect). For example, the parents and children of rod monochromats are carriers.

3. Mating with a suspected carrier: suspected carriers include anyone closely related to someone who has rod monochromacy (e.g., a sibling or a grandchild). If the suspected carrier has the gene, chances are 50-50 that offspring will inherit the defect if he or she mates with a rod monochromat.

4. Sexual promiscuity would have to be included among the circumstances that increase the likelihood of a rod monochromat's mating with a carrier and passing on this gene to offspring.

Known carriers should be aware that the situations listed above would also

increase their risks of passing on the gene or the defect. However, the odds are lower for them than for persons who have rod monochromacy.

Apart from the circumstances listed above, rod monochromats in the U.S. and in most other parts of the world are not likely to have descendants with rod monochromacy, because (1) it is highly unlikely that they will happen to mate with a carrier of the same flawed gene, and (2) it is just as unlikely that their children will mate with a carrier.

The children of rod monochromats, although they are carriers, are most likely to mate with non-carriers, in which case their children (i.e., the grandchildren of rod monochromats) have a 50% chance of being carriers and a 50% chance of not inheriting the gene.

My son is a carrier because he has inherited from me one abnormal gene in this set of genes. He does not have rod monochromacy, because he inherited the normal gene (the dominant one) in this set from his father, who has normal vision. The only way my son would have any chance of passing on this defect to his children would be by mating with a carrier – a very unlikely possibility. If he should happen to mate with a carrier, the chances of their having a child with rod monochromacy would be 1 in 4 – i.e., the same odds that existed for the parents of all rod monochromats in this network. In mating with a non-carrier, the odds are 2 in 4 that my son's children would not even inherit the faulty gene.

My son is an artist. He has excellent vision and is a perfectionist about color and fine detail in his art work. The wonders of genetics continue to amaze me.

*More detailed information about autosomal recessive inheritance is available upon request. To receive this information, send a self-addressed stamped envelope to the address on the title page of this book. Also, see pp. 10-11 of this book.

The Genetics of Rod Monochromacy

**Alex V. Levin, M.D., FRCSC, Co-Director, Ocular Genetics Program
Joanne Sutherland, Genetic Counselor, Ocular Genetics Program
The Hospital for Sick Children, Toronto
May, 1996**

The most common form of achromatopsia, rod monochromacy, is considered an autosomal recessive disorder. Although there are other forms of achromatopsia (for example, blue cone monochromacy) which show other types of inheritance, this article focuses on current knowledge about the genetics of the “usual” form of this disorder: autosomal recessive achromatopsia, or rod monochromacy.

Genes are the pieces of hereditary material which exist in every cell of our bodies, giving directions as to what specific tasks each cell must do to make our bodies function. Many of our genes are involved with cells that have specific functions in the eye. Some of the eye genes instruct the body on how to make cone cells in the retina and how to make them function correctly.

For every gene, we have two copies: one inherited from our mother and one from our father. In autosomal recessive diseases a disorder only occurs if there is a defect in both of our copies of the gene. One abnormal copy of the gene is inherited from one parent and the other abnormal copy is inherited from our other parent. The parents are considered carriers, since they each have one abnormal copy of the gene but also a “protective” normal copy. If there is an error in one copy of a gene, the matching normal gene can usually instruct the cell properly. It acts as a back up system to cover up the error.

Carriers of achromatopsia have no visual symptoms and no abnormalities on eye examination. Therefore, they have no way of knowing that they carry the gene. Every individual in the world carries approximately 6 to 8 recessive gene abnormalities, of which only some involve the eye and vision. People only know which abnormal genes they carry if they have children with someone who happens to carry an abnormality in the same gene. Both individuals may pass their abnormal gene on to a child, who then gets a “double dose” (one from each parent), and that child shows the effect of the autosomal recessive disorder.

The likelihood that spouses carry abnormalities in the same gene is higher if the spouses are related to each other or if they come from a tightly knit or geographically isolated culture. This is why there is a higher incidence of achromatopsia in some populations, particularly if they are somewhat isolated geographically, as is the case in certain islands of the South Pacific.

If both parents are carrying one abnormal gene and one normal gene (they are carriers), the chance with each pregnancy that they would both pass on their abnormal copy (rather than their normal copy) at the same time is 25%. This would produce a child with rod monochromacy.

There is a 50% chance that one parent would pass an abnormal gene and the other would pass his or her normal copy of the gene. In this case, the offspring would be a carrier just like the parents, but would not have rod monochromacy.

There is a 25% chance that both parents would pass on their normal gene, producing a child who is not a carrier, who is unaffected, and who will not have offspring in subsequent generations who would be carriers or who would be affected. The offspring who are carriers would still have to have children with

another carrier for there to be the possibility of having a child affected by rod monochromacy.

As for rod monochromats themselves, they have inherited two abnormal copies of the responsible gene, but they too would have to have a carrier partner (or an affected partner) in order for there to be any possibility of their having affected offspring. This is because any given individual usually passes only one copy of his/her genes to each offspring. For a rod monochromat to have affected offspring, the offspring would have to also inherit a copy of the abnormal gene from the other parent.

Since the rod monochromat only has abnormal copies of the gene to pass on, all of that person's offspring become carriers.

Since it is necessary for both parents to carry the same abnormal gene in order to produce affected offspring, families may have one or more affected siblings and yet have no other family history of the disease. Autosomal recessive disorders are usually found in one generation only (i.e. among the children of two carriers).

Sometimes all children in a family will have rod monochromacy. This is possible because the chance for the parents to pass on their abnormal copies of the gene is the same for each pregnancy. (It is similar to flipping a coin. The chance that you will flip heads on any given flip is 50%. Yet you could flip heads 3 times in a row). The chance that 2 consecutive children would be affected with achromatopsia is $25\% (0.25) \times 25\% (0.25) =$ a one in 16 chance (6.2%). What are the chances for three children in a row inheriting rod monochromacy? 1.6%. Consequently, saying that there is a 25% chance for each pregnancy is not the same as saying that 25% of all children born to parents carrying the same abnormal gene would be affected.

In families from geographically isolated cultures or cultures in which intermarriage between relatives is common, the disorder will often be found in more than one generation. This pattern is sometimes called "pseudodominant inheritance," a term that refers to a different type of inheritance, autosomal dominant inheritance, which is being mimicked, due to the high frequency of abnormal genes being passed around within this tightly knit group.

But what genes are actually abnormal in rod monochromacy? This is a question scientists have been attempting to answer. It has already been discovered, as a result of studies conducted at several research centers, that more than one gene can cause the disorder. Presumably, the genes that are involved determine the way cones are made and/or the way in which cones function. Each of us has thousands of genes distributed over 46 chromosomes. This means that there is a lot of territory for scientists to explore in their efforts to uncover the genes related to rod monochromacy. Recent scientific studies have been able to link rod monochromacy in some families to an area on Chromosome 2 and in some other families to an area on Chromosome 8, but not all families studied have linked to one of these two locations. Once all of the genes are found, we will be able to better understand exactly how rod monochromacy is caused. *(Editor's note: see the information about genetic studies on pp. 118-120 and references on page 117.)*

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Author's Note: There are many subtleties involved in autosomal recessive transmission which may not have been completely covered in this article. For a better understanding of the genetics of rod monochromacy, the reader should consult a genetic counselor, an ocular geneticist (eye geneticist), or a medical geneticist.

The Genetics of Blue Cone Monochromacy

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There are 3 kinds of cone cells in the retina, each kind defined by the color of pigment it contains: blue, green, or red. This relates to the color of light detected. It is the mixing and matching of input from the combinations of these cells that allows normal color vision. In complete achromatopsia (rod monochromacy), there is a generalized defect in cone cell formation/ function, so that all 3 types of cones are affected. In blue cone monochromacy (BCM), cone cells develop normally, but the retina is unable to fill them with red or green pigment, thus leaving only blue cones.

Patients affected with BCM usually show similarities to complete achromatopsia, including reduced central vision (often better than in complete achromatopsia), abnormal color vision, nystagmus (shaky eyes), and photophobia (aversion to bright light). Unlike rod monochromats, however, some patients with BCM can also show a progressive deterioration of the part of the retina which is populated by cones (the macula). Almost always, only males are affected by BCM.

The red and green cone cell pigments are produced by genes on the X chromosome, which is a chromosome involved with determining a child's gender. Normal males have one X chromosome and one Y chromosome. Normal females have two X chromosomes and no Y chromosome. A patient who has BCM has a defect which prevents the formation of red and green pigment from a single X chromosome.

A woman who carries this defect on one of her X chromosomes usually does not have BCM, because she also has a normal X chromosome that "protects" her from the defect. This other X chromosome can produce enough red and green pigment to allow her to have a fairly normal collection of red and green cone cells in addition to her blue cone cells, so her color vision should be close to (if not completely) normal. Males have only one X chromosome, so any defect on the X chromosome has no "protection," since there is not a normal second X, as there is in females. The Y chromosome does not offer matched protective functions. As a result, if a male has the BCM defect on his X chromosome, no red or green pigment is formed. This male is affected with BCM. Rarely, a woman may show signs of having BCM, due to a failure of her normal X to adequately protect her from the deficiency of cone pigment caused by the abnormal gene on her other X.

When a person's body makes eggs or sperm, each egg or sperm contains only one member of a pair of chromosomes. Random chance determines which member of the XX (female's) or the XY (male's) pair is passed on. A woman carrier of BCM (or of any other X-linked disorder) will randomly pass on either her normal X chromosome or the X chromosome that carries the BCM defect. Each egg has a 50% chance of carrying either her good X or her defective X. If she is married to an unaffected male, her egg – which carries only one of her X chromosomes – will be fertilized by a sperm carrying either his normal X chromosome (thus creating a daughter) or his normal Y chromosome (thus creating a son). Therefore, there is a 50% chance that any child they have, male or female, will receive her abnormal X chromosome. If the child receiving the abnormal X is a girl (egg with defective X fertilized by sperm with normal X), the child will not have BCM but will be a

carrier. If the child receiving the abnormal X is a boy (egg with defective X fertilized by sperm carrying Y chromosome), he will be affected.

In summary, when the mother is a carrier of BCM and the father is unaffected, there is a 50% chance that any pregnancy which results in a daughter will produce a carrier; and, for any pregnancy which results in a son, there is a 50% chance that the son will be affected. Likewise, there is a 50% chance that offspring will be neither carriers nor affected, if the egg carrying the mother's normal X is fertilized with sperm carrying either the father's X or Y.

If a male affected with BCM has children with a normal female (she has two normal X chromosomes), then any offspring are guaranteed of receiving a normal X chromosome from their mother. The father's sperm will either contain his X, with the BCM defect, or his Y. All of his daughters will receive his X along with an X from their mother. The daughters will be carriers of BCM. All of the sons will be normal, since they will get his Y along with an X from their mother (i.e., no abnormal X). The exception to this rule would be the very unlikely event that the mother happens to be a carrier of BCM. The likelihood of this happening would be increased if the mother and father happen to be related to each other (e.g., cousins). Being without symptoms, the carrier mother would never know she was a carrier. Molecular genetic blood testing to look for the gene defect may be helpful.

Authors' note: The above information is meant to serve as a guide for understanding the basics of X-linked inheritance. Before making decisions regarding childbearing or regarding identification of potential carriers in your family, you should consult with a geneticist, an eye geneticist, or a genetic counselor for a more detailed explanation. For many patients with blue cone monochromacy, the exact genetic defect can be elucidated using a blood test. This test is available in only a few laboratories in the world. Your geneticist or eye geneticist may be able to make such a test available to you and your family, if desired.

Achromats Who See Color

Achromatopsia, strictly defined, means "without color vision," but many persons diagnosed as achromats actually have some color vision. In fact, researchers are finding that incomplete achromats outnumber complete achromats. A 1992 report on research at the University of California School of Optometry at Berkeley noted that laboratory tests revealed residual cone vision in many patients previously thought to be total rod monochromats. Some of the subjects in this study were blue cone monochromats, persons who have blue cones but no other cones.

There have been only 4 recorded histologic studies of the retinas of deceased persons who had been diagnosed as achromats. These studies all showed the presence of cones, albeit cones which were few in number, abnormal in shape, and oddly arranged on the retina.

Since research findings show great variability in the number and sensitivity of residual cones among incomplete achromats, one can assume that there are also great variations in the visual experiences of these individuals.

Some incomplete achromats have quite a bit of color vision, but most do not. More typically, they may, for instance, just be able to see some reds in certain kinds of illumination. Incomplete achromats can be very particular about what color their lenses are. Some tints (such as red) can interfere with what little color vision they have, and other tints can enhance their color vision.

Getting Diagnosed

Throughout our lives, those of us with achromatopsia have been very much aware of our unusual visual problems, and we have often had to explain about our vision to others. But few of us grew up having a name for our eye condition. In fact, many of us knew little or nothing about why we see the way we do until we were adults. For some of us, this information was eventually provided by a helpful eye care professional. For others, it came as a result of connecting with this network. And yet nearly all of us have been seen by eye doctors since an early age.

Because achromatopsia is so rare, the fact is that most vision care professionals have never had experience with achromatopsia patients. And most of those who have had such patients have not seen more than one or two in their years of practice. Among eye doctors, the ones who can be expected to have seen achromatopsia patients and to be especially knowledgeable about this disorder are the ocular geneticists and retinal specialists. Among optometrists, those who work at low vision centers are more likely to have had experience with achromatopsia patients.

A thorough ophthalmological evaluation includes discussion of symptoms, testing of visual functions, and a careful eye exam, including dilation of the pupils in order to see inside the eyes. Since eye exams are done in lighting comfortable for achromats and since achromats' eyes tend to appear normal and healthy, with no sign of disease, it is understandable that some doctors, upon first examining the eyes of an achromat, might not readily discover that the patient has a serious vision problem. Sometimes the only clues for the examining physician are (1) the patient's nystagmus (shaky eyes) – but nystagmus can accompany other eye

conditions as well, and an achromat's nystagmus might not even be very pronounced in low lighting; (2) the patient's aversion to the examining light that is directed at his/her eyes – but there are many patients who are sensitive to this light; and (3) poor visual acuity when tested using standard vision charts – but poor acuity can be a symptom of many different eye conditions.

Quite a few persons in our network have shared stories about having been suspected of faking vision impairment during vision exams, simply because their eyes had appeared to the examiner to be so normal and healthy.

So it is important to describe to a doctor what it is like to have this condition. Examples of statements that can help with diagnosis are: "I have never seen colors," "In the daytime, I cannot open my eyes and see anything except blinding light outdoors, unless I wear dark glasses," or "My child seems to see fairly well indoors and at night but behaves more like a blind child outdoors in the daytime." Even a doctor who has never before had any direct experience with achromatopsia patients should be able to recognize from reports such as these that the patient has a problem related to lack of cone vision. The doctor may then perform or schedule additional tests, consult reference books or a specialist, or go more deeply into the patient's (or the family's) medical history. The more articulate and assertive the patient is (or, in the case of children, whoever is accompanying the child) in describing what it is like to have this eye condition, the more likely one is to obtain an accurate diagnosis and appropriate help.

One network mom reported that she felt sure that the doctor examining her son's eyes in his dimly lit examining room was not taking seriously her

description of how seriously impaired his vision was. So she told the doctor she would gladly pay more if he would only go outdoors with her son and see for himself how dramatically different the boy's vision was in the sunlight.

Diagnosing infants and children is especially difficult. Two symptoms commonly indicative of achromatopsia, nystagmus and hypersensitivity to light, may be evident, but it is difficult to test for color perception, visual acuity, and other visual functions until a child is old enough to cooperate with such tests. Sometimes it is necessary for an ophthalmologist to see a child regularly over a period of several years before being certain of the diagnosis.

Color vision tests are especially important in diagnosing achromatopsia and distinguishing one form of achromatopsia from another. Rod monochromacy is the most common form (most networkers are affected by this form). A much rarer form of achromatopsia is blue cone monochromacy (fewer than 5% of networkers are affected by this form). The Berson Test is reported to be especially useful in diagnosing blue cone monochromacy.

Rarer still is a condition called cone monochromacy, in which there is total congenital colorblindness, yet no sign of other vision impairment. There are no statistics available for this condition.

It has become common for the ERG (electroretinogram) to be used routinely in diagnosing achromatopsia. While achromats and their families are well acquainted with the fact that achromats have relatively good vision in lower levels of illumination and progressively poorer vision as illumination levels are increased, it is the ERG which doctors rely on to provide the kind of objective information they need for diagnostic purposes. However, there are many different retinal disorders which can

result in an abnormal ERG, so it is important that an ERG be performed and interpreted by someone well qualified to do so. It is also important that doctors make use of all of the methods that can be used for diagnosing achromatopsia. These include careful examination of the eyes, patient history, family history, color vision testing, and testing of other visual functions. Also, it is important to inquire about other medical conditions a patient might have.

There are some rare medical syndromes that include progressive cone dystrophy or cone/rod dystrophy – eye conditions which can manifest visual symptoms similar to those associated with congenital achromatopsia. If a patient has other serious medical problems (which can include hearing impairment, obesity, cardiovascular problems, or other symptoms) and these are brought to the attention of the physician, this information may help in determining the diagnosis. The presence of other medical conditions certainly does not exclude the possibility of the vision disorder being congenital achromatopsia, but it is important to learn whether the other symptoms point to a different diagnosis.

ERGs did not play a role in the diagnosis of very many of the adult achromats in our network when they were children. Some of us have, however, voluntarily submitted to testing with ERGs for purposes such as contributing to research data. Administering an ERG is a complicated procedure, requiring considerable cooperation from the person being tested, so young children generally do not make good candidates for ERGs. Even so, ERGs are being performed on very young achromatopsia patients. Many network parents have disturbing stories to tell about when their children were subjected to this test at a very young age. From the doctor's

standpoint, the ERG can be seen as an important procedure for ruling out the possibility of medical conditions more serious than achromatopsia.

Many networkers have received little or no information from their doctors regarding their vision disorder. One can only guess as to whether the doctors simply didn't choose to volunteer information (and weren't pressed to do so) or whether the doctors may have been uncertain themselves or negligent about pursuing a diagnosis.

Many achromats have been misdiagnosed in the past or have been given vague diagnostic descriptions, such as "legally blind" or "poor eyesight." Most commonly reported misdiagnoses for networkers include ocular albinism, amblyopia, congenital nystagmus, and progressive cone dystrophy.

Different eye doctors have different ways of sharing diagnostic information with patients and their families. Some doctors may assume that exact medical terms are not desired, so they may explain a vision defect in general terms (for example: "The part of the eye that lets people see in bright light never formed properly"). When offering precise medical terms, ophthalmologists are more likely to say "achromatopsia," while vision researchers and optometrists are more likely to use the terms designating the specific form of achromatopsia – e.g., rod monochromacy (the more common form) or blue cone monochromacy (a much rarer form).

Terms can be confusing. For example, the word achromatopsia, strictly speaking, means "without color vision," and yet researchers are finding that achromats who have some color vision (incomplete achromats) outnumber those who have no color vision at all (complete achromats). And this term refers to only one of the manifestations of this vision disorder, the one that is of

greatest interest to researchers – our lack of color vision. Yet those of us who have achromatopsia know that the colorblindness is of minor importance to us. The more serious manifestations – our poor central vision and the inability of our eyes to adapt to higher levels of illumination – are considered to be "symptoms" of achromatopsia.

Doctors use the term "photophobia" in referring to our light sensitivity. Many achromats resent this term, because it translates as "fear of light" – which is not really the case – and because words ending in "phobia" imply an abnormal fear of something (as in claustrophobia). Some of us prefer to say "hypersensitivity to light" – a phrase that stresses the element of sensitivity, which truly is a part of our visual experience, rather than fear. But we learn to live with the nomenclature that is in use, inadequate though it may be.

Some people want to know precise terms more than others do. Some of us are natural born researchers and want to look up information for ourselves. My mother was content to hear the doctor she took me to as a child explain that it was as if other people had "curtains" that could be pulled to control the light and my eyes didn't have "curtains." But I was different from my mother. I needed to know much more than that.

Later on, a doctor I consulted told me that I had "aplasia of the macula." I looked this up and found out that the macula is the part of the retina where cones are concentrated and that cones are the photoreceptors responsible for (1) seeing color, (2) seeing in higher levels of illumination, and (3) seeing fine detail. Using key words like "macula" and "cones," I eventually did my own research at medical school libraries and discovered the terms achromatopsia and rod monochromacy. I was surprised at how little information I could find

about my eye condition in ophthalmology books (it wasn't even included in some of them) but was thrilled whenever I did turn up something.

I learned that, in the early 1960's, a number of people with achromatopsia had been part of a research study of patients having various vision disorders, all of them classified as having "cones dysfunction syndrome." Years later, I found that the author of a medical textbook about inherited eye diseases had included congenital achromatopsia along with various other eye conditions involving abnormal cone vision in a chapter entitled "The Cone Dystrophies." Ever since then, I have understood why some doctors use the term "cone dystrophy" when referring to achromatopsia, even though the word "dystrophy" seems to imply a progressive disease. Achromatopsia is not degenerative, but progressive cone dystrophy is. In the U.S. the term cone dystrophy is most commonly used with reference to progressive cone dystrophy. Some doctors consider achromatopsia to be stationary cone dystrophy.

For those who wish to learn about scientific studies of achromatopsia, there is a book containing abundant information on this and related topics – *Night Vision* (Cambridge University Press, 1990). Anyone interested in reading this book might want to consider obtaining it through an inter-library loan, as the book sells for over \$130.

Getting accurately diagnosed is important for many reasons. Besides satisfying the need to know what one's eye condition is and having terms to use for looking up information about it, obtaining an accurate diagnosis can lead to appropriate genetic counseling. The articles about the genetics of rod monochromacy and the genetics of blue cone monochromacy (pp. 10-13) make it clear that inheritance factors

are quite different for different forms of achromatopsia.

Also, recommendations in terms of the best tinted lenses vary with one's diagnosis. Rod monochromats are helped by certain lens colors outdoors – dark amber, dark red, and dark brown, for example – but blue cone monochromats are helped by other tints, and their color vision would be adversely affected if they were to use the tints that work best for rod monochromats.

More about terms: most achromats tend to have a visual acuity of around 20/200 and so qualify as "legally blind" when applying for special services, accommodations, benefits, etc. Some have a better visual acuity – 20/160 or even better (these tend to be incomplete achromats), and some have a visual acuity of about 20/400. These numbers have to do with whether one can see the big E or whatever else is on the top line of a vision chart and whether one can read anything below the top line.

The fact is, however, that many vision impaired persons with different eye conditions have similar visual acuity readings based on tests using standard charts, even though there is a wide range of visual functioning among these persons. Someone with albinism having 20/200 vision may have much better visual functioning than an achromat with the same acuity. And an achromat with 20/200 vision may see a lot better than someone with cataracts who has a 20/200 visual acuity reading.

These terms are part of established systems for certifying people as "legally blind" (i.e., severely visually impaired), and they play a necessary role in helping people to access many resources, such as special accommodations from the phone company (free phone with re-dial, free directory assistance, and other features), transportation benefits (Amtrak discounts, riding Greyhound

buses with a companion for the price of one fare, regional transit discounts, etc.), vocational rehabilitation services, and more. One's ophthalmologist or optometrist is often the person who provides certification of vision impairment. Patients bring to them (or mail or fax in) certification papers that need to be signed by a professional.

Thus, eye care professionals provide more than just information about diagnosis and ocular health. They also serve as important links to resources for the vision impaired. Good eye doctors are well informed about local organizations serving the vision impaired, and they make sure that their patients who qualify for these services know about them. Also, eye doctors can refer patients to low vision centers, so that they may learn about what's available in low vision devices. Ophthalmologists and optometrists in private practice have only a limited number of optical devices on hand for their patients to try out; but, at a low vision center, a patient can try out and compare many different types of devices.

Often people entering our network want most of all to find out if there is a cure for achromatopsia. It is important to help patients and their families understand that persons with achromatopsia have a congenital, inherited abnormality of the retina (see information on p. 1) and that it is not something which can be treated with surgery, medication, corrective lenses, or other kinds of treatment that are used with various other eye conditions. However, although there is no "cure" for achromatopsia, eye care professionals can help patients with achromatopsia to obtain good tinted lenses and other adaptive devices and to explore ways of controlling illumination. In other words, they can help these patients find ways to make the most of the vision they have.

Persons with achromatopsia also tend to have common refractive vision problems (nearsightedness or farsightedness and, very often, astigmatism as well), and so eye care specialists often like to concentrate on correcting these refractive errors, since providing Rx lenses is fairly easy – in contrast to the impossibility of "correcting" the vision problems caused by the absence of cone vision. However, most achromats find that the slight difference in visual clarity caused by their nearsightedness, farsightedness, or astigmatism is a relatively minor problem compared with the truly serious vision impairment caused by achromatopsia and that the Rx lenses prescribed for them make very little difference in the way they see. Therefore, after weighing both the advantages and the disadvantages of wearing corrective lenses, many achromats choose not to wear Rx lenses or to wear them only occasionally.

Such decisions usually baffle the vision care professionals, who may be inclined to believe that such patients must surely be very vain and/or very foolish. Some doctors, however, come to the same conclusion about the questionable value of Rx lenses for achromat patients after observing that the patients do not seem to enjoy noticeably improved vision when using the corrective lenses and are, in fact, restricted in terms of the sunglasses they can use because of the Rx lenses. Some well designed sunglasses that effectively protect from glare (so important for achromats) cannot be fitted with Rx lenses or worn in combination with them. Individuals who are found to need especially strong corrections for refractive errors are usually glad to wear Rx lenses. Those with only a mild or moderate degree of nearsightedness or farsightedness are generally the ones inclined to reject using Rx lenses.

Achromatopsia in Print

Before 1990, information in print about achromatopsia was limited mainly to what could be found in books and journals in the fields of vision science, ophthalmology, and optometry and a few newspaper articles which focused on individuals with achromatopsia. There was also an obscure autobiography by an English woman named June Monkhouse (*Sight in the Dark*, 1981), which included much information about the author's private life but little about achromatopsia. In 1990 the book *Night Vision*, containing much information about achromatopsia, was published by Cambridge University Press.

In 1992 and 1993 (just before this network began) many articles about achromatopsia appeared in publications for the general public, most notably the one which appeared in *The New York Times* in November, 1992 (see pp. 23-24). Since then, there have been a number of newspaper articles which focused on individual members of our network, and all of these articles included information about achromatopsia.

Since January, 1994, a substantial amount of information about achromatopsia has been published in the books and newsletters which I have produced for the network. And in 1997 a bestselling book by a popular author (*The Island of the Colorblind* by Oliver Sacks) brought this rare vision disorder to public awareness. This book may not have made achromatopsia a household word, but it has had major significance in terms of our network.

The Island of the Colorblind is rich in information about some of Dr. Sacks' favorite subjects, such as botany and islands in general. Information about achromatopsia is provided in the first half of the book – the part that focuses on the journey made in 1992 by Oliver Sacks, Knut Nordby, and others to the

islands of Pohnpei and Pingelap, where there is an unusually high incidence of achromatopsia.

Excerpts from *The Island of the Colorblind*, by Oliver Sacks

“Ordinary ‘colorblindness,’ arising from a defect in retinal cells, is almost always partial. Some forms are very common. Red-green colorblindness occurs to some degree in 1 in 20 males (it is much rarer in females). But total, congenital colorblindness, or achromatopsia, is surpassingly rare, affecting perhaps only 1 person in thirty or forty thousand. What, I wondered, would the visual world be like for those born totally colorblind? Would they perhaps, lacking any sense of something missing, have a world no less dense and vibrant than our own? Might they even have developed heightened perceptions of visual tone and texture and movement and depth, and live in a world in some ways more intense than our own, a world of heightened reality – one that we can only glimpse echoes of in the work of the great black-and-white photographers? Might they even see us as peculiar, distracted by trivial or irrelevant aspects of the visual world and insufficiently sensitive to its real visual element? I could only guess, as I had never met anyone born completely colorblind.”

“Knowing congenital achromatopsia is hereditary, I wondered whether there might be, somewhere on the planet, an island, a village, a valley of the colorblind (as in H.G. Wells' ‘The Valley of the Blind’). When I visited Guam in 1993, some impulse made me put this question to my friend John Steele, who has practiced neurology all over Micronesia. Unexpectedly I received an immediate, positive answer. There was such an isolate, he said, on the island of Pingelap. Just a few days earlier, he had seen an

achromatopic boy, who had journeyed to Guam from Pingelap with his parents. And he said the incidence on Pingelap was extraordinarily high, almost 10% of the population. I was intrigued, and I resolved that sometime I would return to the South Seas and visit Pingelap. When I returned to New York, I received a long letter from Frances Futterman, a woman in Berkeley who was born completely colorblind. She had read my essay on the colorblind painter (*The Case of the Colorblind Painter*, published in *The New York Review of Books* and in Dr. Sacks' 1995 book, *An Anthropologist on Mars*), and she was at pains to contrast her situation with his and to emphasize that she herself, never having seen color, had no sense of loss."

"But congenital achromatopsia, she pointed out, involves more than colorblindness. What was far more disabling was the hypersensitivity to light and poor visual acuity which also affect congenital achromatopes. She wondered if I knew of the book, *Night Vision* – one of its editors, she said, was an achromatope, a Norwegian scientist named Knut Nordby. He was a physiologist and psychophysicist, a vision researcher at the University of Oslo and, partly by virtue of his own condition, an expert on colorblindness. I wrote to Knut, asking how he might feel about coming with me on a 10,000-mile journey, a sort of scientific adventure to Pingelap. He replied yes, he would love to come."

"Colorblindness had existed on Pingelap for more than a century; and, though there had been extensive genetic studies, there had been no human explorations of what it is like to be an achromatope, not only to be totally colorblind oneself, but also to have, perhaps, colorblind parents, grandparents, neighbors and teachers. I had a vision, only half-fantastic, of an entire achromatopic culture, with its own singular

tastes, arts, clothing – a culture where the sensorium, the imagination, took quite different forms from our own and where 'color' was so devoid of referents or meaning that there were no color names, no color metaphors, but perhaps a heightened language for the subtlest variations of texture and tone, all that the rest of us dismiss as 'grey.'"

"When we converged in Hawaii, in the dazzling sun of Waikiki, Knut was wearing two pairs of dark glasses over his prescription lenses, a pair of polarizing tinted clip-ons and over these a large pair of wraparound sunglasses with a darkened visor. He was much more comfortable when we repaired to a quiet little cafe, where he could take off his dark glasses. I found the cafe too dark at first, and groped and blundered, knocking down a chair as we went in. But Knut, already dark adapted from wearing his double dark glasses and more adept at night vision to begin with, was perfectly at ease in the dim lighting and led us to a table."

(*After landing on Pingelap, where they were greeted by island children*)
"Beautiful!" whispered Knut, enraptured, and then, 'Look at that child, and that one!' I followed his glance and saw, here and there, clusters of children who squinted, screwing up their eyes against the bright sun, and one boy with a black cloth over his head. Knut had identified them, his achromatopic brethren – as they, clearly, spotted him, squinting, dark-glassed, beside the plane. Though Knut had met other achromatopic people, this had in no way prepared him for the impact of finding himself surrounded by strangers half a world away with whom he had an instant kinship. It was an odd sort of encounter which the rest of us were witnessing – pale, Nordic Knut in his Western clothes and the small brown achromatopic children of Pingelap. It was intensely moving."

“We visited the Edwards family. Entis Edwards is achromatopic, as are all three of his children. His wife has normal vision, though she is evidently a carrier of the gene. Entis is well educated. He is a minister in the Congregational Church and a fisherman, a man well respected in the community. But this, his wife told us, is far from the rule. Those born with ‘The Maskun’ (the Pingelapese word for achromatopsia, pronounced Mah-skoon) have less chance of marrying, partly because it is recognized that their children are likely to be affected, partly because they cannot work outdoors in the bright sunlight, as most of the islanders do.”

“Outside the dispensary, we began to give out the wraparound sunglasses we had brought, along with hats and visors, with varying results. One mother, with an infant squalling and blinking in her arms, took a pair of tiny sunglasses and put them on the baby, which led to an immediate change in his behavior. One old woman indignantly refused to try any sunglasses. She had lived eighty years as she was, she said, and was not about to start wearing sunglasses now. But many of the adults and teenagers evidently liked the sunglasses, wrinkling their noses at the unaccustomed weight of them, but manifestly less disabled by the bright light.”

(Near the end of the narrative, as he pondered whether he had found an “island of the colorblind”) **“There was an obvious kinship – not just familial, but perceptual, cognitive – among the achromatopes. Everyone on Pingelap, colorblind or color-normal, knows about the ‘maskun,’ knows that it is not only colorblindness that those affected by it must live with, but also an intolerance of light and inability to see fine detail. When a Pingelapese baby starts to squint and turn away from the light, there is at least a cultural knowledge of**

his perceptual world, his special needs and strengths. In this sense, then, Pingelap is an island of the colorblind. No one born here with the maskun finds himself isolated or misunderstood, which is almost the universal lot of people born with congenital achromatopsia elsewhere in the world.”

“Knut and I each stopped in Berkeley, separately, on our way back from the islands, to visit our correspondent, Frances Futterman. She and Knut were especially excited to meet one another. Knut told me that it was ‘an unforgettable and very stimulating experience. We had so much to share with each other that we talked incessantly, like excited children, for several hours.’ Like many achromatopes in our society, Frances grew up with a severe degree of disability, because good visual aids were not available to her. She had to avoid bright light and to contend with a great deal of misunderstanding. She had no contact with anyone who could understand her experience of the world.”

“Could there not be a sort of community of achromatopes who, even though geographically separated, are bound together through commonalities of experience, of knowledge, of sensibility, of perspective? Is it possible that, even if there is no actual ‘island of the colorblind,’ there might be a conceptual or metaphoric one? This was the vision that inspired Frances Futterman, in 1993, to start The Achromatopsia Network, publishing newsletters, so that achromatopes all over the country – and potentially all over the world – could find each other, communicate, share their thoughts and experiences. Her network and newsletter – and now a Website on the Internet – have done much to annul geographical distance and apartness. Perhaps this network, this island in cyberspace, is the true ‘island of the colorblind.’”

**Sufferers From Rare Condition
Are Blinded by Bright Light**

**Dancing
out of the
darkness**

**New Clue to Vision:
People Whose Glasses
Must Be Rose-Colored**

**Doctors learn from
rare colorblindness**

**BLINDED
by the
LIGHT**

***They were blinded by
sunlight . . . until now!***

**‘Night people’ focus of
colorblindness study**

These headlines, some of them sounding quite sensational, played a part in the development of our network. Following is a brief history of our network.

★ In 1992 and 1993, a number of newspaper and magazine articles were published about achromatopsia, focusing in part on research being conducted at that time at the University of California School of Optometry in Berkeley. Media coverage began with a story in the campus newspaper with headlines proclaiming “‘Night people’ focus of colorblindness study” (reporting on a gathering that had taken place for research subjects and their families). Then there was a segment about achromatopsia on Cable News Network (CNN), an article on achromatopsia (written by a Pulitzer prize winning reporter) in the Medical Science section of *The New York Times*, and an article in *Woman’s World Magazine*. The *Times* article was published by many newspapers throughout the U.S. Of all the media coverage during that period, the *Times* article presented the most nearly accurate information, with the least amount of sensationalism. Because of the role I played in getting the *New York Times* reporter interested in writing the article about achromatopsia and because I was featured in that article and in others, I began to receive letters and phonecalls from achromats and parents of children with achromatopsia all over the U.S. I decided to start a network.

★ January, 1994: I began publishing a newsletter for people in this network.

★ August, 1996: A site was established for our network on the World Wide Web.

★ 1996 and 1997: A bestselling book by a popular author brought achromatopsia to public awareness in many countries around the world – *The Island of the Colorblind*, by Oliver Sacks (Alfred Knopf, publisher). The information pertaining to achromatopsia was included in the first half of the book, the section focusing on the journey made by Dr. Sacks and others to the tiny atoll of Pingelap and to a district of Pohnpei Island (both in Micronesia), where achromatopsia is not rare but a common vision disorder affecting a significant number of islanders. Information about our network was included in the book, and more people connected with us.

★ Our network continues to grow, both in size and diversity. See the *Handbook of Information for Members of the Achromatopsia Network* for more information.

Excerpts from the New York Times Article on Achromatopsia, Nov. 17, 1992

Of all the articles on achromatopsia published during 1992 and 1993, the one which appeared on the Medical Science page of the New York Times on Nov. 17, 1992, presented the most complete and most nearly accurate information. On this and the following pages are some excerpts from that article, which was written by Natalie Angier.

Berkeley, Calif.: Frances Futterman's office has a cozy after-hours feel to it even first thing in the morning. The lights are turned low, and fuzzy throw-rugs cover floor and furniture alike, to absorb glare and soothe the eye. Ms. Futterman suffers from a rare genetic disorder that has stripped her eyes of the power to see unaided in daylight. If she steps out into the sun without dark glasses, she turns nearly blind, lost in an explosion of whiteness, like a winter traveler staggering through a blizzard.

Ms. Futterman, who runs a counseling service for persons with vision problems, is part of the largest study ever of a condition called rod monochromacy, a type of colorblindness so complete and so bizarre that patients with the disorder not only fail to see any colors, they also see best when the rest of us can hardly see at all: at dusk, or by the glimmer of the moon and stars.

Dr. Gunilla Haegerstrom-Portnoy, a practicing optometrist and vision researcher at the University of California at Berkeley, has assembled 65 people suffering from this and a similar type of colorblindness called blue-cone monochromacy, in an attempt to understand the nuances of color vision and the intricate genetics that leave some people partly or wholly colorblind.

Ms. Futterman and others with rod monochromacy lack cones, the photoreceptor cells in the retina that respond to color and are responsible for day vision. They can see only through the grace of their rods, photoreceptors that control night vision and are extremely sensitive to dim light.

Rods are terrific at picking up the weakest of light signals and can detect even a single photon soaring through space, but as the light brightens toward daylight, or the indoor equivalent, the rods quickly saturate into uselessness.

For the normal-sighted, the cones pick up where the rods wash out. For those without cones, they simply cannot see above a certain brightness; the screen turns to snowy blankness. And, because only the cones possess the red, blue, and green pigments that are sensitive to colors, people with rod monochromacy can, in the best of circumstances, see only varying types of brightness, a palette of silvers and diamonds.

"One thing I hate is when people say 'Oh, you see in black and white,'" said Ms. Futterman. "It's true that I can't imagine what it's like to see in color, but I see so many different 'shades.'"

Dr. Portnoy said that, by studying rod monochromacy, researchers can focus on half of the visual system without the intervening complexity of the other half.

"This is a fascinating defect for vision researchers," she said. "Cones swamp out the rods in normal vision, so this is an excellent way of studying rod function without worrying about cones." But she has also discovered that there is great variability in the number of cones that individual rod monochromats possess, from almost none to a scattering of cells around the periphery of the retina, the sheet of photoreceptors at the back of the eyeball that connects to the optic

nerve, the vision cable to the brain.

This pattern of cone distribution has led her and others to propose that the genetic defect behind the disorder disrupts, not the body's ability to generate cone cells, but its power to usher those cells to their proper position on the retina, once the cones have been created.

Dr. Portnoy and her colleagues have developed a wide range of lenses to

help people who suffer from either type of colorblindness. Rod photoreceptors are poor at detecting light falling

into the red range of the spectrum; rod monochromats not only cannot see the color red, they are relatively insensitive to red light. Thus, glasses are designed to allow in only light that is in the red range of the electromagnetic spectrum, a small fraction of the sun's overpowering glow.*

The patients experiment with how dark or light they want their lenses to be, for different lighting conditions. A fluorescent bulb throws out a lot of blue light; and so, for a rod monochromat, it appears much harsher and will require a darker protective lens than an equally bright incandescent lamp, which gleams in the redder range of the spectrum.

Yet even those who keep half a dozen different pairs of glasses with them and who switch from one to the other, as they move about from shadow to sun, are happiest when they can take their strange spectacles off and take in the grand sweep of the world unaided.

Many with the disorder are proud night owls, who love going out after dark. Even children with the disorder keep vampire hours. Debra O'Bayley of Santa Rosa, California, whose daughter Elise has rod monochromacy, said the child will do anything in order to stay up late.

"She'll be up until eleven at night, playing outside, if she can con us into it," said Ms. O'Bayley. "She'll play in her sandbox, on her bouncer, anything to be outdoors when she feels free."

Dr. Portnoy, who herself suffers from a far more common and milder type of colorblindness, red-green deficiency, estimates that about 1 in 33,000 people are rod monochromats. The disorder is

so unusual that many optometrists never encounter a case of it, and in fact the syndrome on occasion is

misdiagnosed, or undiagnosed. Because those with the trait possess retinas that look perfectly normal in an ordinary eye exam, some have been accused of malingering when they can't make out the eye chart. At centers like Berkeley, where researchers have devised sophisticated color vision charts and use electroretinographs to detect the absence of cone signals, even infants with the disorder can be diagnosed and outfitted with the proper spectacles.

For older monochromats, though, life was trial by improvisation. Ms. Futterman, who grew up in a relentlessly sunny and shade-free town in Texas, spent the first 17 years of her life without any help at all, not even a pair of sunglasses. She was terrified of going out onto the school playground during recess. She learned to make do, perpetually squinting to shield out light or blinking her eyes frequently and relying on the afterimage that remains on the retina to help her get around.

*Editor's note: Not all rod monochromats use red lenses. Some prefer to use lenses that are dark brown, dark amber, dark brown with a touch of red, or other dark tints. Magenta lenses work well for blue cone monochromats.

The New York Times
Medical Science
Tuesday, November 17, 1992

About Being Colorblind

Since not many people know about achromatopsia, some persons use the term “colorblind” to refer to common color vision deficiencies – usually the inability to distinguish red from green (a hereditary defect affecting about 8% of the population, mostly males).

Among the sighted, the only people who have actually never experienced color vision are those with congenital complete achromatopsia (rod monochromacy). There are also incomplete rod monochromats who have very limited color perception. Researchers are finding that incomplete achromatopsia is more common than complete achromatopsia. Sometimes a person diagnosed as a complete rod monochromat learns that he or she actually has incomplete achromatopsia, when tests reveal the presence of some cones in the person’s retinas (see p. 1). Incomplete rod monochromats tend to have better visual acuity and less photophobia than complete rod monochromats.

When there is more than one sibling with rod monochromacy, they often have different variations of the disorder. One incomplete RM reports that she sees some “warm” colors and her sister (an RM) sees some “cool” colors.

A very rare form of achromatopsia, blue cone monochromacy (BCM), has different inheritance factors from those connected with rod monochromacy (see pp. 12-13). BCM patients have blue cones in their retinas, enabling them to see some colors and to have somewhat better vision with less photophobia than rod monochromats.

Cone monochromacy is an even rarer form of inherited achromatopsia. Those with this condition are totally colorblind but have normal visual acuity and no unusual light sensitivity.

There are persons who have a condition called cerebral colorblindness, in

which color vision is lost as a result of trauma, stroke, or some other cause. Such cases are extremely rare and are diagnosed by neurologists, rather than eye doctors. Their eyes are not affected. Instead, the brain’s color vision center is damaged. A man with cerebral achromatopsia – an artist who had created vividly colorful paintings for decades and then suddenly, in his 60’s, lost all color vision – is described in “The Case of the Colorblind Painter” in the book, *An Anthropologist on Mars*, by Oliver Sacks (Knopf, 1995).

After reading about the depression this man had experienced (depressed at the sight of his wife’s skin, favorite foods, and other things he had formerly cherished the sight of), I began corresponding with Dr. Sacks about my experience of having lived without color vision all my life. I explained that, while being totally colorblind had frustrated me in many ways throughout my life, I had never been depressed about not seeing color. In fact, I have always found much pleasure in the beauty of the natural world. I expressed the wish that the painter could come to enjoy all the visual beauty that exists, even in the absence of color.

It is generally agreed among those of us with inherited achromatopsia that our colorblindness does not matter nearly so much as our photophobia and our low visual acuity.

But, to the painter, the absence of color was a tragic loss. He feared he would never see beauty again. In time, however, his feelings about this – and even his lifestyle and his paintings – changed dramatically. All of this is related in detail in Dr. Sacks’ book.

The painter devised a way of demonstrating to others what it was like to see as he saw. He furnished and decorated a room entirely without color. Even the

smallest objects in the room were gray, black, or white. He then invited others to spend time in this room.

It would be impossible to simulate accurately the visual experience of congenital achromatopsia, mainly because, in addition to our colorblindness, there is also our hypersensitivity to light and our lack of detail vision that would need to be part of the simulated visual experience. — Frances Futterman, editor

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Colorblindness: a Personal Account
by Knut Nordby

Following are excerpts from “Vision in a Complete Achromat: a Personal Account,” by Knut Nordby, Ph.D., which is the title of chapter 8 of the book Night Vision. Dr. Nordby is a vision scientist and an achromat.

In my early childhood, when I used coloring pencils or crayons, I always broke all the rules about the correct colors to use. I would happily color the sky green, the grass and leaves orange, the sun white, and so on. I was always corrected in these choices by those who knew better, and so eventually I gave up painting and coloring.

I would memorize the colors of my clothes and other things. I learned the rules for correct use of colors and the most probable colors of various things.

Trying to explain to someone with normal color vision what it is like to be totally colorblind is a bit like trying to explain to a normally hearing person what it is like to be completely tone deaf. My task, though, is simpler than that, because everyone has experienced achromatic (colorless or “black and white”) or monochrome pictures and has experienced the gradual disappearance of colors, as darkness sets in.

I compare my color-less world to the visual experiences normally sighted people have when viewing black and white

movies or good black and white photographs (sharply focused, high contrast with a long gray scale, such as one finds in crisp, high quality technical prints).

I have acquired a knowledge of the physics of colors and the physiology of the eye’s color receptor mechanisms; and, from art history and other studies, I have learned about the various meanings attributed to colors. But none of this can help me understand the essential nature and character of colors.

When I know the color of something, I often use its color name in referring to it to other people. Communication is made easier. This in turn leads others to use color terms when speaking with me, even those who know of my colorblindness. Sometimes this is helpful – e. g. when they refer to a red book among light colored books. But referring to a red book among dark colored books is not helpful. I cannot tell them apart.

Looking at paintings, I can appreciate form, composition, and technique, though I cannot appreciate the color aspects. I can, however, appreciate monochrome prints, graphic art, sculpture, and architecture in the same ways that those with normal color vision do.

When painting or redecorating, my wife normally selects the colors. If there is enough contrast between the old color and the new one (in tones I can see), I have no problem doing the painting.

When buying clothes on my own, I only take advice from a salesperson in whom I have great confidence. Otherwise, I ask for “safe” or neutral colors – white shirts, gray trousers, black socks and shoes, etc. For important color choices I rely on my wife or close friends who know my preferences. I often mark my socks in some way, so that I will not mix up different pairs.

Colors are often used to code or highlight information. For me, this usually

makes matters worse, because good color contrast for the normally sighted often does not transform into good contrast for me. Sometimes the contrast visible to me is so low that the information is almost lost. Black print on red price tags, yellow print on a light blue background, and dark green on a bright red background are all extremely difficult for me to perceive.

Colorblindness: a Personal Account by Frances Futterman

About seeing in shades of gray: the following paragraphs relate my own experiences as a complete achromat and may or may not reflect the experiences of other complete achromats.

As a child, I had no information whatever about my eye condition. I only knew what it was like to experience it. Whenever I had to tell others that I couldn't see colors, a common response would be "Oh, then you see everything in black and white" or "So you must see everything in shades of gray." Both statements disturbed me. Neither was true. For one thing, the only two "colors" I could really identify were black and white, and I took great pride in my ability to do this. I knew that, in the world I saw, very few things actually were black or white. In fact, I could see an amazing variety of "shades" ranging from very dark (almost black) to very light (almost white). I was able to distinguish the fine differences in things that were different shades of the same color – differences that other people seemed to have a hard time discriminating. I did not understand at the time that the term "black and white" simply meant "not in color," as in black and white movies, etc. To me "black and white" sounded like a hideous way for the world to look, and the application of this term to my way of seeing made it sound far more restricted than it actually was. So I would

adamantly declare that I did not see in black and white!

I found it equally disturbing to be expected to agree with the notion that I saw everything in shades of gray. From early on, I had developed meaningful concepts of various colors. I had a sense of what "red-ness" or "green-ness" meant, for example. I even had favorite colors, based on what they seemed to represent. But gray? I really could not get a sense of what gray was – and, to this day, I still find gray the hardest "color" to comprehend. No one could convince me that the world I knew to be so richly varied in shades of light and dark was all supposed to be called "gray"! To me, it seemed ridiculous to imagine that, for instance, a dark tree and a light colored flower would both be identified as "gray."

If I could see – or if I had ever been able to see – any colors, then I might perceive the absence of color as gray. But my visual experience has never included seeing color – and so gray, for me, has always been just one more "color name" to have to memorize and to learn how to use (in connection with cloudy skies, hair color, etc.). To those persons who try to understand colorblindness and who ask me if I see grays, I usually say something like this: "I suspect that, if it were possible for you to see through my eyes, you might find that you were seeing in shades of gray. But, since this can't happen, we'll never know for sure."

I see many different shades of light and dark. I see things as "darker than" or "lighter than" other things. Exercising my right to describe my own visual experience honestly and in terms that have real meaning for me, I have to say that I do not see gray. Perhaps someday people with achromatopsia will have a special vocabulary that can adequately describe the way we see.

INCOMPLETE ACHROMATOPSIA:

Research findings show great variability in the number and sensitivity of residual cones in the retinas of persons diagnosed as incomplete achromats, so one can assume that there is also great variability in the visual experiences of these individuals. I have received many comments from incomplete rod monochromats, describing how they see. Some can see several colors; but, more typically, they see only a little color – and sometimes can do so only under certain kinds of illumination.

Incomplete achromats can be very particular about the color of lenses they use. Certain tints enhance their ability to perceive specific colors, and other tints (such as red) can interfere with what little color vision they have. Even amber or brown lenses, which are well liked by many rod monochromats, can cause color distortion for some incomplete rod monochromats.

EFFECTS OF COLORBLINDNESS:

Achromats tend to agree that, while being colorblind matters very little to them personally, it affects their experiences significantly in connection with tasks that require color identification, color matching, color coding, etc. Being colorblind can also matter a lot in terms of the social stigmatization which is often experienced by children and teens, when others discover their inability to see color. This tends to be much less of a problem for adults.

Several networkers have expressed a mild interest in what it would be like to see in color; but achromats generally do not express feelings of deprivation, sadness, or bitterness about this aspect of their eye condition. Having poor vision and severe light sensitivity are problems that matter so much more.

However, the lack of color vision has, for many members, played a significant role in determining their choices and

level of participation in connection with various vocations and avocations.

The importance of being able to identify colors varies not only with the type of activity in which one may engage but also with the culture, the geographic location, and the times in which one lives. Persons living in modern industrialized societies face more pressures to be able to identify and to continually relate to a wide range of colors than persons anywhere else at any other time in history.

By contrast, on the little island of Pingelap, where about 6% of the population have achromatopsia, it is reported that the inability to perceive colors makes little difference in the lives of islanders with achromatopsia. This is partly because life there is so different from life in industrialized societies and partly because almost all of the colors to be seen are shades of green.

In our own society, not only do we achromats have to acquire a knowledge of countless color names that we come across as we shop, read, pursue an education, etc. – we also must make sense of numerous color related concepts which come up in daily living. How can we tell when something is “faded” or if a color is “too bold”? It can sometimes be important to know when someone looks “flushed” or “pale.” We may be advised that something we want to wear does not “go well with” something else. What a mystery! And, very often, what a bother!

Networkers have shared many suggestions for dealing with the various problems (practical and otherwise) that result from colorblindness. We have to manage one way or another to cope with this bizarre vision disorder in a world where the colors of things can matter so much, and it is helpful to be in touch with others who are dealing with the same kinds of situations.

Adapted Lifestyles and Adapted Environments

It is reasonable to assume that someone whose eyes cannot adapt normally to sunlight, who therefore experiences greatly decreased vision at the levels of illumination in which most people live, work, and move about – and who has reasonably good vision at night, at twilight, or in dimly lit settings – would choose a lifestyle that accommodates these unusual characteristics. Certainly this would have been true for persons with achromatopsia in the past, before it became possible to obtain the many kinds of tinted lenses that are now available to help achromats function in a wide range of settings. There are many options these days for those who have access to modern optical aids and good tinted lenses and who can afford them. However, serious limitations are still the reality for achromats living in various parts of the world today who do not have the opportunity to have such helpful devices.

Among those achromats who do have the sunglasses, visors, side shields, and other items which facilitate seeing in the outdoors, some nevertheless opt for adapted lifestyles, in which the use of such aids is minimized and stresses on the eyes are limited.

Many adults with achromatopsia choose lifestyles that are not so very different from those of normally sighted people, with regard to the hours they keep, the places they go, and the amount of time spent outdoors, and they make use of whatever lenses and devices help them. Others choose schedules and arrangements that maximize time spent in visually comfortable surroundings and minimize time spent in settings where they must resort to sunglasses in order to see and function reasonably well. Many make vocational and avocational choices that allow them

maximal visual comfort and minimal visual stress.

Such lifestyle options are more restricted during the years of compulsory education, when much time must be spent in brightly illuminated classrooms and on outdoor playgrounds and when standard bedtimes go along with keeping standard school hours.

Just as in the rest of the population, some achromats really are “night people,” who prefer being up till all hours of the night and sleeping late into the day, but many others are “day people,” who thrive on more conventional hours of sleeping and waking. Similarly, among achromats, the proportion of “outdoor” types and “indoor” types is much the same as in the rest of the population.

A 1992 campus newspaper article about achromatopsia research at U. C. Berkeley began: “Like subterranean moles, some people spend the daylight hours sealed in their homes with the curtains drawn, hoping to avoid the sunlight that blinds them.” This article referred to those of us in the study as “the night people.” This is an example of sensational journalism. It does not at all describe accurately the lives of persons with achromatopsia.

We do, however, learn early in life that twilight time and night time vastly expand our ability to perceive, perform, and participate in the world at large.

From the *New York Times* article (Nov. 17, 1992) which played a major role in the growth of this network: “Many (achromats) are proud night owls. Even children with this disorder keep vampire hours. A Santa Rosa mother said her child will do anything to stay up late. ‘She’ll be up until 11 at night, playing outside, if she can con us into it – in the sandbox, on her bouncer, anything to be outdoors when she feels free.’”

A *Woman's World* article about achromatopsia from June, 1993, related the experiences of another parent, whose daughter had a strange habit as a toddler: "She'd climb up on top of her toy box at night, open the curtains, and just stare out the window at the sky. She loved to look at the moon and clamored to play outside at night. Yet her days were full of hazards. She'd stumble over things when she was outside. She would step carefully around shadows, thinking they were holes. Once she fell off a patio and broke her leg."

In spite of the vast difference in what we experience outdoors in the daytime (or in brightly illuminated indoor places) and what we experience in settings where our good rod vision allows us to feel comfortable and confident – few of us (if any) would choose to live in a dark basement or other dimly lit place, shut off from the rest of the world. Physiologically and psychologically, we need sunshine in our lives. We also need to move about in the world and to socialize with others. This inevitably means being in settings where others are visually comfortable and we are not. As we mature, we make choices about how much of the brightness of "the real world" we are willing to cope with routinely and how much we want to stay in the "havens" of lower illumination.

These "havens" may be places we create in our homes, where we select window treatments, furnishings, floor coverings, wall coverings, lighting, decorations, and foliage to provide the most visually comfortable environments we can make for ourselves. They may be special places in our yards – like arbors or alcoves. Or they may be places in our communities where we like to go, because we really like the way we see and are able to function in these settings.

These might include theaters, chapels, restaurants, clubs, etc., depending on what kinds of activities we prefer.

We can choose to seek out places that are restful to our eyes. Or we can continue to adapt to whatever settings we find ourselves in as a result of the many influences in our lives. Being visually impaired is, after all, only one aspect of who we are as individuals.

Many parents have expressed great concern about how best to provide for their children's visual needs at home. Some have gone to the trouble of making their entire homes meet the special lighting needs of achromats. One mom said she was beginning to feel like a hermit, staying indoors so much. Another had begun to notice that sunlight was bothering her eyes when she went outdoors, since she was staying so dark-adapted in her home, where the lighting was kept favorable for her two children with achromatopsia.

Housing that must be shared needs to be, as much as possible, a comfortable place for all who live there. Achromats of any age mostly need at least one "haven," a place where they can retreat to and feel visually "at home" – and they need not to be subjected for long periods to highly uncomfortable illumination factors. Non-achromat parents, siblings, and various "significant others" (housemates, spouses, etc.) need to not be subjected for long periods to very low levels of illumination, unless this is really what they enjoy – and they too need at least one "haven" to go to, where the lighting is optimal for them. Compromise is essential. Some homes are not large enough to offer many choices of rooms, but there are lots of possibilities for making lighting factors adjustable. Having adjustable window coverings, having a variety of lamps in

addition to the overhead lights, and using dimmer switches, 3-way bulbs, touch control lamps, and other devices can help to make the same rooms serve the visual needs of different members of the family at different times.

One dad told me that, after several daytime trips to the beach which had been extremely uncomfortable for his daughter with achromatopsia, he decided one day to visit the beach just before dark. When he saw that not only was his daughter happy to be there at this time of day, running freely and having a wonderful time, but that she was also able to see way off into the distance, he just stood there on the beach and cried. He felt sad that he had not thought of taking her to the beach at twilight before. Since that day, he said, all of their beach outings have been planned at the time just before dark.

For another network family, outdoor

sports activities are an important life-style choice, so the parents outfitted their two young daughters who have achromatopsia with tinted wraparound “sportlenses” (with attached visors) to wear over their regular sunglasses for these occasions. The parents report that this has enabled their children to enjoy all-day outings in the snow or at the beach.

Many achromats choose to arrange their schedules so that their outdoor activities – sports, rides in the country, sightseeing, visits to amusement parks, etc. – take place at that magical time of twilight, when they are free of the photophobia and can enjoy their best distance vision. Networkers are invited to share the experiences they have had enjoying their night vision and twilight vision. Below are some of my own fondly remembered experiences.

— Frances Futterman, editor

FAVORITE NIGHT VISION EXPERIENCES So much emphasis is placed on what does not function normally in achromats’ eyes – the cone vision that isn’t there and the things we must do or wear to cope in a brightly lit world. Not enough attention is paid to the rod vision we do have and the experiences and adventures we can have in situations where rod vision is mostly what’s needed, to perceive and function adequately. In my own life I prefer to emphasize the use and enjoyment of my good rod vision. Some of my richest memories include: ★ going on an open-air tour of Yosemite Valley at midnight at the time of the full moon ★ Greyhound bus rides through New Mexico and Arizona, during which I enjoyed hour after hour of window views of the desert in the moonlight ★ swimming in a beautiful outdoor pool at night ★ attending a Japanese “Harvest Moon” ceremony in a lovely “moon garden” ★ leisurely night walks in favorite neighborhoods in the summer ★ enjoying the rides, lights, and sights of the state fair amusement park at night when I was a kid ★ standing at the waterfront and watching a fireworks display on the 4th of July ★ watching a production of “A Midsummer Night’s Dream” performed on a darkened stage decorated with twinkling lights ★ evening tours of department stores at Christmas time to view brightly decorated windows ★ being inside grand old movie theaters, for movies or stage productions ★ taking in the darkened “funhouses” or “haunted houses” at amusement parks ★ lying on the ground in a state park at night and gazing up for hours at tall redwood trees and countless bright stars ★ chasing lightning bugs, as a child, on summer nights ★ square dancing on an expansive lawn on a warm summer night ★ attending a fine Gilbert and Sullivan production performed on an outdoor stage under the stars ★

Orientation and Mobility (O & M)



This term refers to methods used to move about safely, gracefully, and confidently. O & M skills are taught as part of education and rehabilitation programs for the visually impaired. O & M programs were first developed after World War II, in response to the needs of blinded veterans. By the 1960's several training centers for O & M teachers had been established.

O & M for the visually impaired includes sensory training, orientation to surroundings, self-protection, the use of a "sighted guide," and (when appropriate) skills in using a cane. Sensory training involves learning to fully utilize all of the senses, including whatever degree of vision one may have. O & M teachers should individualize programs to the needs and abilities of their students. They should always begin with a thorough assessment of how the student functions in different settings.

Throughout the history of O & M programs, the white cane has been the primary tool used by trainers in helping blind and low vision students to travel independently. Some of these students opt for guide dogs instead. Persons who have a high degree of visual functioning, such as achromats, pose special challenges for O & M instructors. Whether using a cane outdoors is appropriate for achromats is debatable. This needs to be looked at in terms of how an individual sees and functions and also in terms of the settings in which he or she must travel. A white cane performs 2 main functions: (1) It gives the user important information about the pathway he or she is moving along, and (2) it alerts pedestrians and motorists to the fact that the person with the cane is visually impaired.

All persons with achromatopsia can learn to improve their mobility skills as pedestrians, and they can also learn

how to make the best possible use of transportation options. But what about driving? The motivations to drive are many and strong, especially in areas where public transportation is inadequate. For young people, becoming a driver can be like a rite of passage, and being a non-driver can be a social and vocational handicap.

There are many visually impaired persons who do drive, using a bioptic (telescopic aid) attached to their glasses, in order to meet visual acuity requirements for licensing. To qualify requires being able to demonstrate a high level of visual functioning. Some achromats have become drivers, but it's important to understand that there is a wide range of visual functioning among persons diagnosed with achromatopsia. Most do not have sufficient vision to become drivers. Indicators that someone has potential to be a driver include: (1) a high level of visual functioning, (2) long term facility in the use of monocular aids, and (3) a history of having successfully navigated with scooters, bikes, rollerblades, etc.

Qualified low vision drivers are generally granted restricted licenses. They should get thorough training in using a bioptic, both at a clinic or other training center and in actual driving situations. Bioptic training programs are provided at a few low vision centers and at other special training centers. In addition to using a bioptic, achromats who drive use tinted lenses that give them optimal visual acuity and that allow them to differentiate between red lights and green lights. Vision standards for drivers and laws pertaining to driving while using a bioptic vary from state to state.

For achromats, the ability to move about safely, gracefully, and confidently (which are the goals of O & M training) can change dramatically from place to

place, since how they see is so greatly affected by illumination factors. They see better outdoors in the daytime when surrounded by tall trees than in a setting where a lot of the sky is visible – and they see better on streets between tall buildings than amid 1- or 2-story structures. They see better when buildings cast dark shadows than on slightly overcast days when there are no shadows – but they see even better when the sky is heavily overcast, approximating twilight conditions. Visibility is greater when they are close to dark structures, such as brown shingle houses or red brick buildings, than it is in a neighborhood of light-colored stucco buildings. They see better in an alley-way than in a wide plaza. Certain ground covers are easy on their eyes (grass, blacktop, wood chips, etc.), and others make seeing difficult (e.g., light colored concrete). Their ability to see can change sharply just by crossing the street, turning a corner, or stepping into the shadow of a building.

Some of the O & M techniques that achromats use in moving about outdoors in the daytime are ones that they use consciously. Others are used instinctively. They employ many of the strategies used by all vision impaired persons, such as using auditory cues, choosing least stressful routes, and sometimes getting help from others. They also employ methods used by all normally sighted persons whenever visibility is temporarily diminished or fluctuating, due to weather conditions, unfavorable lighting, etc. – e.g., moving cautiously and with concentration.

One of the O & M issues that comes up most frequently for persons with achromatopsia is whether or not they should consider using a cane in outdoor travel. It is common for mobility teachers to expect persons with achromatopsia to use a white cane. These

teachers are concerned with their students' safety and with liability issues. They are trained to teach cane travel, and few of them know about achromatopsia. But achromats, because they have such a high level of visual functioning, usually reject the option of using a white cane. Unfortunately, this resistance is often interpreted as a demonstration of foolish pride or "denial" of one's visual impairment.

It is, in fact, common for persons having certain other eye conditions to deny their need for a cane, until continued vision loss forces them to accept the option of cane travel. So teachers tend to assume that all such resistance must surely be based on denial.

All factors should be considered when determining whether someone actually needs to use a cane. Achromatopsia is not a progressive eye condition. Canes are burdensome, and they signify blindness to the general public. The main function of the white cane – to give tactual information about the path just ahead – is mostly unneeded by most achromats (if they are wearing adequate tinted lenses), except sometimes at steps or curbs. What achromats usually need most in moving about outdoors is not so much to know about the next step but to know about what's farther down the street.

The cane's other main function is to alert drivers and other pedestrians to the fact that one is visually impaired. Children who must cross busy streets to and from school may very well need this symbolic mobility tool; others, who get transported to and from school or who live in neighborhoods with only light traffic, face different realities.

Some achromats carry a folding cane when traveling in unfamiliar cities or in areas where complex intersections must be crossed, situations in which a cane may offer some degree of security.

The specific mobility problems faced by individuals with achromatopsia vary greatly, depending on many factors. There is a wide range of differences with respect to how light sensitive an individual is and differences in terms of visual skills, navigational abilities, lifestyles, and geographic locations.

Another variable has to do with the lenses we use for dealing with the problem of hypersensitivity to light. As we move about outdoors in the daytime, our degree of sightedness is profoundly affected by how much light our eyes have to cope with. Light protection is provided primarily by tinted lenses, but we can also be helped by hats, caps, or visors, to shield from overhead light. For achromats who wear spectacle frames (instead of wraparound sunglasses), side shields may also play a part in light control.

Sometimes someone's mobility problems have been dramatically reduced when he/she begins using adequately tinted, well fitting sunglasses or a visor or hat with a good brim.

Other approaches to what could be called "light management" may include choosing to walk on the shady side of a street or perhaps even choosing a route which includes streets lined with trees, tall buildings, or other natural or architectural features that can affect overall brightness and thus enhance visibility for achromats. Sometimes we are able to choose the time of day for doing errands, commuting, or going on outings. Visibility for us can vary so much from morning until evening.

Improving O & M may mean finding ways to minimize the discomforts and inconveniences that are often experienced by pedestrians and users of public transportation. It may mean making certain choices in the clothing or shoes we wear. It may also mean educating ourselves – through reading, making

inquiries, and gathering handy reference materials – about how to access and utilize all kinds of transportation that are offered to us, including special taxi and paratransit services which may be available to persons with disabilities. It may mean going to a copy center and having enlarged copies made of bus or train schedules, a campus map, or specific portions of a city map.

Some persons may wish to work on improving their O & M skills with the help of a trained O & M instructor. O & M instruction is widely available to students as part of special education services, and it is also available to clients of vocational rehab programs. However, individuals who want help with O & M but who are neither students nor clients of vocational rehab programs have a harder time obtaining such services. Sometimes this is made available through organizations serving the visually impaired. And there are some O & M teachers who free lance, working with individuals on a fee-for-service basis.

As with all other professionals, O & M teachers are not all equally competent to deal with the special mobility needs of persons with achromatopsia. They may need to be educated about this rare vision disorder and the extraordinary kind of vision that achromats have.

Competent and open-minded O & M teachers carefully assess the visual functioning of a student, how well the student navigates, and what specific strengths and weaknesses the student has. The instructor will then determine what specific skills and strategies need to be learned or improved. O & M teachers who have received their education in recent times know that there is much more to orientation and mobility than simply teaching cane travel. If they start by introducing a cane when working with an achromat, you can be sure that their concept of O & M is very limited.

**O & M: a Personal Account
by Knut Nordby**

Following are excerpts from "Vision in a complete achromat, a personal account," by Knut Nordby, from the book Night Vision. Dr. Nordby, who lives in Norway, has complete achromatopsia. He is a scientist with extensive experience in vision studies, both as researcher and as subject. He travels widely in connection with his work.

At age 5 my circle of activity expanded into parts of town near my home. I developed a system for finding my way back, which I still use. I counted the streets I had crossed, keeping count of right and left turns, counting doors and shops, forming a mental topological map. When returning, I reversed directions, retracing my path, making turns in the opposite direction, and counting backwards the streets I had crossed. I learned to make use of prominent landmarks – parks, churches, underpasses, etc. Even today I find it easier to orient myself in cities having rivers, canals, overground railway lines, and other conspicuous, easily identifiable "boundaries." I have never lost my way returning from a place I had first located myself. When traveling with others, it can be more difficult to keep track of the route, especially if traveling by car.

At age 8, I learned to ride a small bicycle. In our village (in postwar Sweden) we scarcely saw more than a couple of cars a day. Much transport was carried out by horse and cart, and the road conditions did not allow speeding. So riding the bicycle in the street did not constitute a grave danger to me, despite my visual handicap.

A problem that seemed more serious to me earlier in my life than it does now is that I cannot hold a driver's license. My brother (also an achromat) and I cycle and have ridden mopeds in Oslo. He actually took some driving lessons

once. He had to give up, though, after attempting to drive down a flight of stairs in a park. Today I only cycle when I have access to reserved cycle lanes or in places where traffic is very light.

Side shields on spectacle frames shut out unwanted light, but they also prevent motion detection in the peripheral visual field, which is important for moving about safely. I don't use them.

Traffic lights sometimes pose a problem. Early in life I learned that red is at the top, amber is in the middle, and green is at the bottom. At night, in the evening, or when traffic lights are in a shadow during the day, I have no problem detecting which signal is showing and can be a law-abiding pedestrian or cyclist. In full sunlight, however, detecting signals can be quite impossible. In such situations I have to watch the traffic, following other pedestrians when they cross the street. This can lead to dangerous situations, if I should happen to follow someone who is crossing a street against a red light.

Whether blinking my eyes briefly at low light levels or extending the length of the blinks at high intensities, I experience a visually stable world in which I can orientate myself and move about. At higher light levels my peripheral visual field is much more affected than the central part of the field. I will still detect movement in the far periphery, but I have much more difficulty in identifying what is moving and reacting adequately to it. This makes me move in a rather hesitant and stiff manner, sometimes bumping into people, and to be over-cautious when moving about in agitated surroundings or when encountering unmarked steps. As soon as I am in the shade or indoors, I again move in a much more relaxed and confident way.

A special problem I have is reading the destination signs on approaching buses which only stop on signal. On

occasion, I have had to stop every bus until the right one came along, invoking the driver's wrath and being showered with abuse for my pains.

It can be difficult to find my way in unknown surroundings. I carry with me a small monocular telescope, which can also be focused up close and which I use for reading street names, flight departure indicators, and other information that must be seen from a distance. I also make preparations by studying maps and plans and by asking for directions on how to find my way.

O & M: a Personal Account by Frances Futterman

I have complete achromatopsia. For the first 17 years of my life, I had no sunglasses. During most of those years, I lived in a small Texas town with lots of sunlight, very little shade anywhere, and assorted obstacles to watch out for in roads and pathways. Needless to say, navigating outdoors in the daytime was extremely difficult. I had to develop many survival skills and strategies.

In order to see outdoors in the daytime, I had to blink rapidly and continuously. When I would open my eyes even briefly, I could not see the world around me, because I was blinded by light. It was like having strong floodlights aimed at my eyes. The squinting and blinking allowed me to obtain just enough visual information to move about in reasonably safe surroundings. For walking around town or crossing the highway on my way to and from school, I needed someone to walk with me. What my blinking made possible is rather amazing. Each time my lids opened a little, I saw only the blinding light, but each time they closed, I got a fleeting glimpse of my surroundings. With my eyes closed, I could "see" what was not possible for me to see with my eyes open. I now understand that the light entering

my pupils was saturating my rods and that, within a split second after closing my eyelids, the light level diminished sufficiently to allow my retinas, which were rich in rods but lacking cones, to experience a brief "after-image." During all those years, I made my way around outdoors by using these "after-images." My vision in full sunlight consisted mainly of seeing with my eyes closed!

Today I have various tinted lenses to use, so I am never at the mercy of such circumstances. But sometimes I choose to remove my sunglasses and experiment with this phenomenon. I open my eyes, experience the blinding light, then close my eyes and experience the fading light, which quickly brings me a picture of what I was not able to see with my eyes open. The stronger the light, the longer I am able to see the images, before they completely fade away.

Our bodies adapt in remarkable ways for survival. I believe that my rods manifest this amazing potential in order to perform as fully as possible for me, since there is no cone function to provide the vision I need in bright light. In the early 1970's, I described these "after-images" to a vision scientist who was using me as a subject in her research on rod vision. At first she refused to recognize as valid the experiences I described. It was impossible, she said, for such after-images to be so clear or to last so long. However, she was eventually convinced, and she obtained considerable data substantiating this phenomenal capability of my rod vision.

I have spoken about this with other achromats. Those who had to manage without dark glasses early in their lives, as I did, usually say they are familiar with these after-images. However, those who were provided with very dark tinted lenses early on in their lives (never having had to rely on these after-images) usually cannot relate to this experience.

Accommodations in the Workplace and Elsewhere – Our Rights under the ADA

The following comments regarding workplace accommodations are from a networker with a legal background.

“Among other things, I receive help with filing. I have to keep track of many documents; and, since ours is a small office, there is no support staff to do the filing. I cannot easily scan my desk-top or table to see what’s there. Memorizing where things are and clerical work that involves organizing, copying, typing, etc., are definitely not where my strengths are – although things are vastly improved for me, now that I have glasses with a mounted telescopic aid. These are a “must” for me for typing and computer work. Also, I have been provided with a special lamp with a dimmer switch, which I use instead of the overhead light.

“It is important for those of us with impaired vision not to be silent about what we need in the workplace and to insist that these things be provided. Most employers do not know much about what the Americans with Disabilities Act requires of them (though they may act as if they do), so I would never accept at face value an employer’s assertion that she or he has no obligation to provide a particular accommodation that is needed – as long as the accommodation is a reasonable one. It may not be reasonable, for example, to expect a small, struggling, 2-person office to hire you, if the job requires that you be on the road a lot, necessitating the additional expense of hiring a driver, but it may be quite reasonable for a large corporation to provide this type of accommodation.

“Unfortunately, while ADA regulations spell out with great specificity the dimensions of toilet stalls, the use of braille markings, and other matters that are important to the mobility impaired and the blind, they do not address issues such as lighting requirements and contrast and size of print in signage, etc. – i.e., the factors affecting visibility, which matter so much to partially sighted persons – with the same specificity. Like any piece of legislation, the ADA was drafted through compromise with various lobbyists who were putting forth their own agendas. The prime movers and shakers behind the ADA were people with mobility impairments, and the architectural changes that are now being required reflect this group’s impact, as well as the justness of their position.

“A claim for accommodations for persons with low vision is proper and legally justifiable. When a new building is constructed, the ADA does not mandate that it have dimmer switches on the lights or large print directories, for example. However, any public accommodation must make auxiliary aids and services available to persons with sensory impairments, as well as whatever “reasonable accommodations” may be required. A fast food restaurant, for example, does this by making available hand-held menus. This is not specifically mandated by the ADA, but a restaurant that offered only an above-the-counter menu would have a tough time winning a challenge brought by someone with a vision disability who had requested a hand-held menu.”

Under the ADA (Americans with Disabilities Act), it is the right of every person with a vision disability to have equal access to all public accommodations and all public educational programs, including not only kindergarten through college but also adult education centers, libraries, museums, and community centers.

Vocational Choices for Persons with Achromatopsia

On the network membership forms, new members are asked to share information about their work backgrounds, education, and training. Most networkers have provided at least a little information about their vocations. Through these forms and through my interaction with members (mostly by phone), I have been able to learn that the adults with achromatopsia in our network include computer programmers, salesmen, self-employed business people, full time homemakers, clerical workers, an investment banker, a preschool teacher, a mathematician, an attorney, writers, college professors, an economist, a political analyst, an office supervisor, and musicians. Also, an impressive number of networkers have chosen to work in the "helping professions." There are social workers, a psychiatric aide, ministers, an occupational therapist, a psychotherapist, and various kinds of counselors.

One member, a partial achromat, has worked for years for the National Parks Service and at one time led "night walks" at Yosemite National Park. Another has worked extensively in communication arts, including leading seminars and doing voice-over work for commercials. One networker is a teacher, writer, and researcher in the fields of vision science and telecommunications.

I hope that, as time goes by, networkers will share more information about their past and present experiences working in various occupations and especially about the ways they have dealt with special needs or problems related to their vision impairment, light sensitivity, and colorblindness. Sharing this kind of information even in connection with academic studies they have engaged in or avocational interests they have pursued can be helpful to us all,

as we look at ways to overcome obstacles and make adaptations in various kinds of work situations.

Employment possibilities have opened up dramatically for vision impaired people in recent years. This has occurred partly as a result of rapid advances in the development of adaptive technology (such as large monitors and enlarging software) and the availability of photo-enlarging resources. Due to these and other developments, printed information has become more easily accessible to us.

Also, there is a wide range of optical aids available to help with near vision tasks and distance vision tasks, and there are more options in tinted lenses than there have ever been before.

In addition, the Americans with Disabilities Act (ADA) protects persons with vision disabilities (and other kinds of disabilities) from discrimination by employers on the basis of the disability. As long as someone is otherwise qualified for a position and only "reasonable accommodations" are needed to enable him or her to perform the work, that person may not be turned down because of a vision impairment or treated any differently from non-disabled applicants.

There is a wide range of visual functioning among those of us with achromatopsia. The visual acuity we have for near or far varies greatly among individual achromats, hypersensitivity to light varies greatly also, and colorblindness may be total or partial.

There are also many other individual differences among us. Some networkers are drawn to highly challenging pursuits, making use of whatever technological, optical, or other devices are necessary, while others choose much less demanding paths. There is no single "right path" for everyone.

State vocational rehabilitation services and disabled student services at colleges and universities can provide much help for visually impaired adults who want to pursue educational or training programs leading to employment. The quality of these services varies a great deal from place to place, but the search for good services is time well spent. Likewise, the time spent in locating good quality career counseling (for young people or for career-changers of any age) is also time well spent. Not too long ago, career counseling, by common definition, simply meant aptitude testing. But the old conventional aptitude tests often didn't provide very useful answers, especially for someone having a vision impairment like achromatopsia. One networker related with humor the story of how her aptitude tests in college concluded that she should become either a beautician or a florist. In both of these vocations good color vision is a "must."

While it's true that we always have the option of using paid or volunteer aides in order to perform jobs requiring visual skills we don't have, not all of us are comfortable with the idea of having work arrangements in which we must rely so heavily or continuously on such assistance from others.

Workplaces and specific jobs run the gamut from "worst possible scenarios" for achromats to least stressful, least problematic situations for achromats. The former category includes many forms of outdoor labor, jobs involving considerable fine detail work, jobs in which color vision plays a large and important part, and work requiring careful, continuous visual surveying of extensive surroundings. Jobs in the "least stressful" category might include certain night jobs, work calling for interpersonal skills rather than visual skills, and jobs in darkened environments, such

as theaters. Networkers are asked to suggest examples of "worst job ideas" or "best job ideas" for achromats, both from their own personal experience and from using their imagination.

Many years ago, when I was just out of high school, I found a job as a darkroom technician in the X-ray department of a hospital. Whenever I needed to step out of the darkroom, I had to wear red goggles, in order to stay dark adapted. I didn't realize then that I had found a great job for an achromat. I only knew that I did not feel at all visually impaired in this job. I often got to serve as a "sighted guide" to normally sighted persons who had to go into the darkroom for one purpose or another (doctors, technicians, students, etc.).

It is wise, if at all possible, to check out prospective workplaces before accepting a position. Illumination factors, among other things, can vary tremendously from place to place. One networker reported having given up one job for another one which offered much better pay and benefits, only to find that the new work environment was very uncomfortable for her visually, seriously restricting her ability to do her job.

Some have opted for self-employment in their homes or in offices or other workplaces, where the decor, lighting, work surfaces, etc. can be adapted to meet their special needs, permitting a setting with optimal visibility. There are increasingly more types of work which can be done at home.

"Disability" – visual or otherwise – is a very relative concept. It can actually be defined by the situation in which one must function and the tasks one is expected to do. In some work situations, someone with achromatopsia may actually be "disabled" and be ever mindful of their sight impairment. In others, having achromatopsia can be mostly or totally irrelevant.

Social and Psychological Aspects of Achromatopsia

In order to understand the social and psychological issues faced by persons with achromatopsia, one needs to be aware that, in addition to the extreme sensitivity to light, colorblindness, and poor visual acuity which achromats experience, it is the only vision disorder which causes one to experience widely varying levels of vision impairment from infancy throughout life.

How well achromats are able to see and, thus, how easily they are able to function in the activities of daily living are greatly influenced by all factors of illumination. Consequently, their experiences in all stages of infancy, childhood, and throughout their lives range from one extreme to the other in terms of their degree of sightedness. They have remarkably good vision in some situations, significantly impaired vision in other situations, and near-blindness in full sunlight outdoors, unless they are wearing very dark lenses.

In some settings, they can freely move about and participate fully, feeling confident and “normal,” yet they can be moderately or severely restricted in other surroundings. Even in the same room, they have vastly different experiences in terms of how sighted they are, depending on the time of day, the weather, whether the curtains are drawn, the kind of lighting being used, etc.

No other eye condition brings the need to adjust to such variability in vision throughout life, including the important formative years. There are degenerative eye conditions, such as diabetic retinopathy, which can cause periodic fluctuations in visual functioning, and there are hereditary eye conditions, such as retinitis pigmentosa (often called RP), which cause those affected to function reasonably well in some surroundings but to experience reduced vision in others. (In fact,

persons with RP have the opposite problem from that faced by persons with achromatopsia – their vision is best in bright light and worst in dim light.) But none of these degenerative eye diseases is present in infancy, as is the case with achromatopsia. Persons with advanced RP or any of the disease-related or age-related eye conditions have to deal with many difficult issues due to their vision loss, but only persons with achromatopsia have the lifelong challenge of coping with vision that can change significantly from place to place and often from one minute to the next, depending on the kinds of lighting in their surroundings and the quality, quantity, and direction of the light.

There are other congenital eye conditions, most notably albinism, which cause unusual sensitivity to light, thus affecting visual functioning and visual comfort in settings where there is bright illumination. But none of these eye conditions cause extreme variability in vision resulting in the need to keep adapting and adjusting to changing levels of visual impairment such as is experienced by achromats. Nothing else even comes close.

At all stages of growing up, from early infancy on, achromats struggle with the same developmental tasks which are essential for all human beings – physically, socially, psychologically, and intellectually. As they do, they must somehow manage to integrate the changing and often bewildering and contradictory experiences that result from their extraordinary way of seeing. How they experience life in favorable lighting conditions is strikingly different from how they experience life in unfavorable lighting conditions. For an achromat, their “indoor self” is usually quite different from their “outdoor self,” at least in the daytime, and their “night

time self” is typically very different from their “daytime self.” An individual with achromatopsia who can move with grace and agility indoors or in the evening may be very tense and awkward in daytime activities outdoors.

In bright light outdoors children with achromatopsia can often feel helpless, disoriented, awkward, and insecure, but they can feel quite the reverse in low level lighting. A child who is gregarious by nature may become a “loner” on the playground. A very bright student with achromatopsia may have to endure the stigma of appearing to be a slow learner in a brightly lit classroom where visually oriented instructional methods prevail. Especially at certain stages of childhood and adolescence, it can be emotionally painful to be perceived by one’s peers as clumsy or slow-witted.

The concepts people form about themselves as they are growing up and the concepts they form about their identity among their peers can affect them profoundly. Adults with achromatopsia might look back in awe, wondering how they survived all of those stages of growing up as well as they did. Many of our networkers did so with the help and support of caring parents, siblings, friends, and teachers. Others did not have these advantages but nevertheless eventually found their way to good self-esteem and a healthy outlook.

As achromats, we have the formidable task of taking the many (and often wildly fluctuating) impressions we perceive about who we are and what our capabilities are and integrating all of these impressions into a reasonably stable and healthy self-image.

Exactly what is our place among the sighted and the blind? There is no simple answer to this question, because the place we find ourselves in keeps changing. This is especially true for those who choose to be very mobile,

live a “normal” lifestyle, and participate in many activities. We cannot adapt all of the environments in which we spend time; so, instead, we find ways to adapt ourselves to the environments, either by using various tinted lenses or by developing a variety of coping techniques (psychological as well as practical) for the times and places in which our ability to see is severely reduced.

One member of our network said, “It’s a wonder that we don’t develop dual personalities in the process of trying to adjust to our changeable visual realities. Or perhaps we do.”

When achromats participate in groups of blind and vision impaired persons, they tend to stand out as being very sighted and especially capable, so much so that sometimes questions are raised regarding whether they actually “belong” in such groups. Yet, in groups of normally sighted persons, achromats stand out as being visually impaired and so must cope with all the problems and special needs familiar to the visually impaired. Many a networker has said, “It’s like being ‘neither fish nor fowl.’”

Many achromats have chosen, at one time or another, to restrict their social lives to situations in which their vision problems are not readily apparent to other people. This most commonly happens in connection with dating. Networkers report varying experiences and varying success in these efforts to “pass” as normally sighted. Speaking from experience, some have pointed out the problems that can result from having made such a choice, such as ongoing tension, self-consciousness, and the persistent fear of being “found out.” Some, however, report having greatly enjoyed at least some of their experiences in which they kept their vision impairment a secret. Some have called it “liberating.”

Totally blind persons or those with

very little usable vision have to cope with many difficult issues related to being blind, but they do not face the kinds of problems pertaining to identity and disclosure in social groups – problems which are so significant in the lives of partially sighted persons, such as achromats. As blind persons move about and participate in various social activities, they cannot easily “pass” as sighted and so they are not continually tempted to do so, as many partially sighted persons are, especially during adolescence and young adulthood.

Partially sighted persons, on the other hand, do not have an identity that is so clearly defined and so evident to other people. This fact can greatly influence their level of “adjustment” to their vision disability. To “blend in” and be accepted as “normal” can feel so much better than having to confront the various reactions people can have to someone who has a disability. The desire to fit into social groups is such a normal, healthy human instinct. So it is understandable that the partially sighted are tempted to “pass” as normally sighted.

Also, because achromats come so close to experiencing what it is like to be normally sighted (i.e., when lighting is ideal for them), they face even greater temptation to “pass” than do other partially sighted persons. For this same reason, the task of accepting and “adjusting” to one’s vision disability is especially difficult for achromats.

The opposite of “passing” is disclosure regarding one’s vision impairment. Disclosure can lead to experiences with the full range of reactions and attitudes people have about physical “differences” and disabilities. While there are many people who are accepting and comfortable with someone who has a disability, there are many others who are not comfortable and there are those who can be over-solicitous, patronizing,

pitying, or overly inquisitive. Also, in just about any school, there are those who engage in mean-spirited teasing and name-calling. Making choices about disclosure is no easy matter. (See more about this on p. 121.)

The existence of a support network for achromatopsia (beginning in 1994) has made it possible for achromats young and old to have a dialogue about the social and psychological issues and the various ways they have coped with these issues.

As facilitator and reporter for this network, I would be seriously misrepresenting networkers’ input if I reported only their positive comments and the “good news” that they have shared with me regarding their accomplishments and strengths. A complete and honest representation of networkers’ lives would also have to include reporting their many frustrations and even the bouts with depression that have been reported by some. Many networkers have told about having struggled with poor self-esteem, anger, and anxieties related to their vision impairment. Even those of us who tend to put a positive spin on problems that arise have stories to tell about the difficult periods we have been through in our lives. Usually when networkers write in about such periods, they are referring to experiences during late childhood or adolescence. When they write in about having come to terms with various psychological issues related to their vision, it tends to be in reference to some period in adulthood.

For most achromats, the teen years have brought the most difficult experiences in terms of social and psychological issues. Peer pressure is strongest, one’s self-confidence is most vulnerable, issues related to dating emerge dramatically, one’s appearance tends to become very important, and the emotional ups and downs of adolescence have their

effect. Also, a certain “rite of passage” – that of becoming a driver – seems to be a possibility for just about every teenager except those who are visually impaired. It is not an easy period to go through, even with supportive family, teachers, and counselors.

There are, of course, exceptions. Some individuals even in early childhood have experienced the pain of being “different” as seriously as if they were in their teens. Others reportedly have made it through childhood and adolescence with few serious problems related to their vision. Clearly, there are countless individual differences among individuals with achromatopsia, not only in terms of their dispositions and sensitivities but also in terms of what they experience at home, in school, at work, and in other situations.

Fortunately, most achromats who have reported having had serious bouts with emotional problems have also shared information about the particular pathways they have discovered for working through these problems. Some have used professional counselors or

some form of therapy (there are so many different therapies from which to choose these days). Others report having benefited from reading self-help books or autobiographical writings of visually impaired persons (or persons with other kinds of disabilities). The writings of strong survivors can inspire and guide us toward healthy ways of perceiving ourselves and dealing with the challenges we face.

Others have found that studying the social and psychological factors that influence their experiences can be therapeutic. The following references are suggested: *Disability: a Psychological Approach*, by Beatrice Wright; *The Psychology of Disability*, by Carolyn Vash; *Stigma*, by Erving Goffman; and *The Making of Blind Men*, by Robert Scott. (Only one of these books pertains exclusively to the visually impaired, but all of them deal with issues that affect the visually impaired.)

In the book, *Living with Achromatopsia*, there is an entire section of networker input pertaining to social and psychological issues.

Following are some comments from persons with achromatopsia:

From a man in his 40's: “I don't want people to see me squinting, blinking rapidly, holding things close to my eyes, or in any other way having to struggle visually. I also wish I did not have to be seen wearing sunglasses indoors (when the lighting is bright) or outdoors on cloudy days. Whenever someone makes a sarcastic or embarrassing comment to me about my sunglasses, there is no way that I can quickly reply in such a way that the person will understand that my sunglasses are as necessary to me as a hearing aid or wheelchair might be to some other individual.”

From a woman in her 30's: “There was a time in my childhood when I realized that I looked different enough to make it obvious to others that there was something wrong with me. I tried to find ways to mask those differences. But is it really worth it, and does it work? Or will we always just be more self-conscious, more sensitive, and more burdened because of this? Unlike those persons who have more overt disabilities, we can make choices about how we want to appear from one situation to the next. (Do I want to read right now and draw attention to myself? Should I force my eyes open while this person is talking to me? Should I pretend that I can see what this person is pointing at?) These decisions can be stressful.”

Using Adaptive Devices

An adaptive device is any device used to help compensate for an impairment or limitation. Persons with achromatopsia make use of many of the same adaptive devices used by other visually impaired persons – magnifiers, watches with big numbers, reading stands, enlarging software, etc. They also use various devices for coping with their problem of light sensitivity; sunglasses, visors, and dimmer switches are examples of these kinds of adaptive devices.

Members of The Achromatopsia Network share information about the optical aids, sunglasses, and other adaptive devices they use. Parents in the network have to make choices about adaptive devices to obtain for their children. They look to professionals for guidance and often find there is conflicting advice from different specialists.

As with most rare disorders, not much research or writing has been done about achromatopsia. If it were an eye condition affecting the lives of a significant part of the population, then by now we would have a substantial body of knowledge to turn to for help in learning how best to cope with the special needs of achromats, and more would be known about the pluses and minuses of different options.

The choices people make can be based on (1) intuition and common sense, (2) recommendations of individual professionals, (3) experimentation, and (4) learning from the experiences of others. For many achromats, the 4th category wouldn't be a possibility without The Achromatopsia Network. Before this network existed, it was not easy for achromats to find others with whom to compare notes. Network publications report a wide range of members' experiences from which to learn.

It is important to remember, however, that what works for one person may not work for another, because of variables in visual functioning and all the other variables in people's lives.

There is no perfect solution to the problem of having to manage with only rod vision. Inevitably people must deal with trade-offs. For example, being equipped with optical aids and tinted lenses for all situations that might be encountered means dealing with the bulk, organization, and weight of many devices. "Traveling light" brings more freedom but also more limited visual functioning. Another trade-off: many devices that enhance one's vision also attract attention, so this factor must be considered. One man with achromatopsia recalls that, as a teenager, he tried wearing sunglasses with a mirrored coating; the coating made his sunglasses darker and, therefore, more helpful, but his schoolmates would stop in front of him and comb their hair, using his lenses as a mirror.

Generally, the more effective an adaptive aid is, the more conspicuous it tends to be. Concerns about appearance and about fitting in and not being "different" tend to become more important in later childhood and teenage years and to diminish in adulthood. At different ages and in different situations, one's choices in adaptive devices may vary a lot.

With optical aids for distance vision, there are trade-offs too. A 6X or 8X monocular offers many achromats the equivalent of 20/20 vision for "spot viewing," but it can be tiring to hold one to your eye for long periods. Binoculars offering comparable magnification allow for viewing with both eyes but are heavier and cumbersome. Prolonged

viewing with both eyes can be done with lightweight spectacle-mounted binoculars (sometimes called “sports glasses”), but these generally provide only 3X magnification (considerably less than a 6X or 8X monocular) and a rather limited field of vision. Students who are seated close to the chalkboard and those whose visual acuity is better than 20/200 can often manage well with the lower powered optical devices.

There are eyeglasses with built-in bioptics or with bioptics mounted on the frames. These permit distance viewing for prolonged periods without the inconvenience of having to reach repeatedly for a monocular or having to wear mounted binoculars. However, wearing bioptic devices continuously obscures one’s field of vision.

Some people are more sensitive than others to obstructions in their visual field. Some will never wear anything that restricts their peripheral vision. For this reason and others, there is a wide range of attitudes among achromats regarding the use of hats and visors. Well meaning parents and professionals, knowing that achromats see better outdoors or under bright overhead lights when their eyes are shaded, will often try to influence children and teens to wear hats, caps, or visors. This suggestion is strongly resisted by some individuals, yet others love wearing these items. Some children with achromatopsia have large collections of hats or caps and enjoy wearing them at school and elsewhere.

While it’s important to acknowledge the advantages of using something to shield from overhead light, it is also important to respect the reasons that many achromats have for not doing so. Although, in some cases, resistance is

due to self-consciousness about one’s appearance, for others it has to do with visual functioning. Achromats have little or no cone vision, but they do have rod vision (see p. 1), and rods are responsible for peripheral vision (i.e., vision from above, from below, and from the sides as one is looking straight ahead). Like all sighted people, achromats rely on their peripheral vision for safety and for accessing visual information about their surroundings, and some are especially sensitive about having their peripheral vision obstructed. Psychologically and otherwise, many achromats have a strong need to stay “connected” with the sky, even if it means having reduced visual acuity.

Dr. Knut Nordby, a vision scientist who has complete achromatopsia (see p. 5) states: “Along with my spectacles, my small folding pocket magnifier is the most important tool I have; I always carry it with me. I have considered other visual aids, but they all seem to fail on one or more points – they are too large and heavy to carry, complicated to use, or too powerful, giving too small a field of view. When traveling in unfamiliar surroundings, I always carry a small, 8-power monocular, which I can conceal in my hand and which I use for reading street names, destination signs, and other information I cannot get close to. Low visual acuity has not been an insurmountable obstacle. My sister (also an achromat) has, since her early teens, done all kinds of needlework, producing bobbin lace, embroideries, etc., to very high standards. To keep her hands free, she uses a clip-on jeweler’s loupe on her glasses. This provides high magnification but a very small field of view, so it is not very suitable for reading.”

Options for Magnification

Since persons with achromatopsia have only rod vision, they do not have clear, detailed central vision. Their distance vision is affected more than their near vision. There are a number of ways to cope with this problem.

The simplest way is to magnify what is to be viewed, and the easiest way to achieve magnification is to get closer to something or to bring it closer to us. Examples of this method include sitting in one of the front rows at a theater, taking something down from a bulletin board in order to read it, creating a seating arrangement that makes it easy to see the people in a group, getting up close to a classroom demonstration, and visiting a “petting zoo” (at any age) in order to have close-up experiences with animals.

One can choose enlarged versions of things. A 26" TV screen is easier to view than a 19" screen. Watches, clocks, calendars, remote control devices, and other items can be bought that have large numerals or other symbols. Searching for such items can be a worthwhile shopping adventure. A transit map, campus map, or section of a city map can be enlarged at a copy center, as can bus schedules and other reference materials.

Large print books and other reading materials can be found in schools, libraries, and bookstores. And copy machines with enlarging capabilities make it easy to obtain enlarged copies of reading materials, as needed.

Computer technology offers many options for enlarging, both on the computer screen and in printed form.

OPTICAL DEVICES: Sometimes it isn't possible, comfortable, safe, or socially acceptable to move close to something; and sometimes, no matter how close one gets, it is still hard to see something clearly enough. In such

cases people can make use of various kinds of optical devices.

Hand held magnifiers: For achromats, a good choice offers 4 to 6 times magnification and gives a distortion-free image (aspheric optics), with a field of view large enough to see more than one word at a time (at least 47mm lens diameter). Many achromats use the small, less expensive folding magnifiers that come with a case, which can fit in a pocket. Some prefer stand magnifiers, which can be moved about on a page. It is best to try out magnifiers of different strengths and styles to find out what works well for each individual. The best place to do this is a low vision center, where a variety of magnifiers are available. But not everyone lives near a low vision center; and, even for those who do, there can be a substantial waiting period before one can get an appointment. So it is good to know about other ways to get low vision aids.

Most eye care specialists keep some magnifiers on hand to show patients and can request samples of other magnifiers from distributors, as needed.

The National Association for the Visually Handicapped offers a catalog with pictures and descriptions of some useful and not too expensive optical devices. To find out about obtaining this catalog, see page 123 about ways to contact this organization.

A hand held monocular – half a pair of binoculars, sometimes called a miniscope – is good for reading material on chalkboards or for reading street names, signs, and house numbers, as well as other uses. A monocular can be kept in one's pocket and taken out as needed. A wrist or neck strap can be attached. A 6 to 8 power monocular can give many achromats the equivalent of 20/20 vision for spot viewing. Recent models are lightweight and can be focused up close as well as at a distance.

Learning to use a monocular can be easy or difficult, depending on one's age, manual dexterity, and other factors.

Magnifying spectacle lenses or bifocals which incorporate magnifying lenses can be useful for reading, writing, and other prolonged visual tasks.

Another useful device incorporating magnifying glasses is worn on the head; it tilts downward when one needs to look through the lenses and tilts upward when not needed. Used by hobbyists and others who must see fine detail, this is a good option for times when one needs to have both hands free for using small tools, threading a needle, pulling out a splinter, doing a manicure, etc. These devices are adjustable, have good optics, can be obtained in 3 1/2 power, and have names like Magnifocuser and Opti-VISOR.

There are also many options in spectacle lenses with built-in or attached telescopic aids. A low vision specialist can help individuals with achromatopsia find the specific devices that will work best for them.

CLOSED CIRCUIT TV SYSTEMS: In recent decades CCTVs have enabled many visually impaired persons to read small print and accomplish other fine detail tasks that they had not previously been able to do. CCTVs electronically enlarge onto a TV screen. They have controls to select positive or negative output, degree of magnification, contrast, brightness, and focus. Recent models have color monitors; users can choose either color or grayscale contrasts.

Although CCTVs have been a god-send to those who have very low vision, their usefulness is more limited for visually impaired persons who have a higher degree of visual functioning, such as most achromats.

CCTVs – a personal account by Frances Futterman: I have complete achromatopsia. I used a CCTV occasionally

several years ago, when I was enrolled in college courses and had access to them at the disabled student center. I've also tried out various models on display at professional meetings I've attended. While I've been impressed with what they can do, I have found it frustrating trying to use them for my own reading and writing. I do not need the degree of magnification that is needed by persons who have more disabling eye defects. I found that black print against a light background (dimmed) was tolerable, but reading white print on a dark background was visually uncomfortable. This is probably an individual preference and may not be true for other achromats. I found that the print quality was never as sharp as what I am accustomed to seeing on the printed page, and I was always aware of my field of vision being restricted. I value my good peripheral vision and can scan pages of printed material rather quickly. Reading with a CCTV always felt confining. Whenever I tried to read rapidly, flickering images would appear on the screen, because I was moving the page under the camera too fast.

If CCTVs had been around in my school days, I'm sure I would have gladly used one when working with maps, charts, graphs, math involving fractions, encyclopedia pages, or scientific diagrams or formulas. If I ever own one, I will turn to it for easy viewing of such things, as well as for scrutinizing photos or filling out forms which have tiny print. My choice would be one of the hand held devices that can be hooked up to a regular TV set. These devices are portable and less expensive, and they offer advantages to those of us who need sharper images with less magnification. And, with this system, I do not have to endure the bright light that most other CCTVs direct downward onto the moveable platform.

Adapting Indoor Lighting for Achromats

Persons with achromatopsia can select lamps, lamp shades, lighting fixtures, light bulbs, and various kinds of gadgets to create an environment in which their vision is enhanced and maximized and visual stress and glare are minimized. This is more likely to be possible in their living spaces than elsewhere. However, many networkers have been able to custom design office spaces to meet their special visual needs, and others have modified other environments in which they regularly work or study.

A visit to one or more stores which specialize in lighting fixtures to see the many options in lighting that are currently available is highly recommended. The purchases we make may, for the sake of economy, be at hardware, discount, or department stores, but the lighting stores are good places to get ideas. Useful items to be considered include different styles of dimmer switches, lamp shades that filter light comfortably, and multiple-setting lamps with touch controls.

For achromats, indirect lighting is far better than the concentrated light emitted from task lamps or gooseneck lamps, and light from incandescent bulbs is much easier on our eyes than light from fluorescent tubes or halogen bulbs. Incandescent bulbs that are marketed as “full spectrum” can be sampled in some stores specializing in lighting. These are appreciated by many visually impaired persons because of the vision enhancement they report experiencing while using them. Hopefully, there will be more input from network members who have experimented with full spectrum lighting, especially from incomplete achromats who have some

degree of color vision, which might be enhanced under such illumination.

Overhead lighting fixtures come in different styles and different materials. Some diffuse light beautifully, and others emit harsh light. There are many options to experiment with.

Some of the better quality lighting stores employ sales persons who are trained consultants on the subject of lighting and who are able to advise and assist customers who have special needs in terms of lighting.

Light bulb choices to experiment with include soft white (rather than clear) bulbs, lower wattage bulbs, colored bulbs, and 3-way bulbs. Several light sources placed strategically in a room can be easier on the eyes than one or two very bright light sources.

Natural light entering a room through windows and doors can be shaded from the outside by awnings, trees, or foliage and/or filtered on the inside by blinds, window shades, curtains, bamboo shades, drapes, or decorative hangings. Opaque window coverings permit more control over indoor lighting. However, not many people choose to exclude all natural light from a room.

Also, one can choose furnishings and room decor that significantly influence the quality of light in any indoor space. For example, dark work surfaces (wood-grained or dark painted surfaces) are more restful to light sensitive eyes. A work surface that is too light can be covered, partially or totally, by something dark – a desk pad, colored poster-board, painted masonite, colored felt, etc. In classrooms some teachers use black construction paper to cover a desk or table area for a student who has achromatopsia.



Making Reading Easier

What a difference it could make in the lives of visually impaired persons, if they all could have easy access (at home and at schools and libraries, etc.) to electronic scanners, high quality printers, and computers with large monitors, good enlarging software, and easy-to-read keyboards! Any printed material that wasn't easy for them to read could be scanned and transformed into pages of text with good spacing and readable typeface, meeting an individual's specific visual requirements. Whatever might be needed – whether class assignments and study notes or personal reference items, such as a list of Cable TV channel numbers or a list of commonly used telephone numbers, etc. – could be created for a vision impaired person in a large print format.

In this imagined ideal set-up, all the materials one might be expected to use in school, from grade school through college, would be provided in very readable formats. Or, if not, then high quality photo-enlarging services would be made available, at no additional cost, to remedy the situation.

Imagine the sense of freedom and ease that persons with achromatopsia would have, if such resources were readily available! To many people, what has just been described may sound like a great luxury – and yet it merely approximates the freedom and ease which most normally sighted people take for granted, as they regularly access printed material without having to resort to adaptive technology to do so.

The technology required to create this ideal situation is, of course, already in existence, but most partially sighted persons have little or no access to it. Most of them do not have the financial resources to purchase such equipment. There are sources of financial aid which are available to help visually impaired

students to obtain assistive technology, such as special education resources or help from service organizations like the Lions Club – or, at the college level, aid from vocational rehabilitation services. But adults who are not enrolled in educational or job training programs have a much harder time finding financial assistance for acquiring the adaptive technology that could make reading and writing tasks so much easier for them.

For most vision impaired students, school days are filled with visually oriented tasks that can be tiring and frustrating. And the world at large, with all its signage and printed information, is slow to progress to the standard of equal access for all, as set forth by the Americans with Disabilities Act.

While waiting for more ideal conditions to evolve, persons with achromatopsia manage the best they can in the “real world” – using their handy magnifiers and other optical devices and their various strategies for maximizing vision, such as getting up close to things and manipulating lighting factors whenever possible.

Many achromats have grown up without access to any large type materials or magnifying devices. Some feel that they have more confidence and flexibility as readers because of their lifelong experience in managing with standard size print. But some individuals seem to need enlarged print more than others do. There is great variability in visual functioning among persons diagnosed with achromatopsia.

In an ideal program of special educational resources, school materials would be made available in both standard and large type, making it possible for partially sighted students to use large type materials whenever large type is actually needed, but these students would not be required to use large type.

(Continued on the next page)

It is important to consider the preferences of individual students as to whether they actually need or want to use large type. Many do not.

One adult with achromatopsia commented: “I didn’t like large print books back when I was in school. Besides the fact that the print was inappropriately large for me, an open large print book on my desk was just another huge expanse of white – more glare for me to deal with. I can’t remember a single time when I thought, ‘Oh, good! They have this textbook in large print.’”

Using large type materials can reduce some of the stress that some visually impaired students experience in reading. But large type books are heavy and bulky, and they can be an additional stigmatizing factor in a visually impaired student’s life. Also, partially sighted persons have to get close to pages in order to read, and it can be frustrating and time-consuming to have to follow the long lines of text in large print books. The size of type used in these books is intended to meet the needs of low vision readers having various different eye defects. Such large print is just right for some students but unnecessarily large for others.

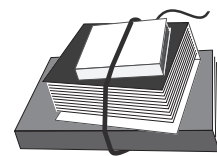
Whether large type is actually needed varies with individuals and also with age and specific situations. Storybooks for young children and school materials for the early grades usually (but, unfortunately, not always) have rather large type, in recognition of the fact that children’s eyes are still developing and need the larger size type. Thus, many vision impaired children at this age do not find it so difficult to use “regular” books. Later on, the print in school books gets progressively smaller. At some point in the middle school years, visually impaired students usually begin using large type materials. The visual comfort level in a particular classroom,

the subject being studied, various qualities of books, an individual child’s overall visual functioning, and other factors all play a part in determining whether large type is needed.

Much later in life, even those who managed to go all through school without using large type materials may find that they begin to look fondly on the option of large type – as they begin to experience the decreasing accommodative ability of their eyes which most (but not all) people experience around age 40. It is not uncommon for persons with achromatopsia to become aware of this change in their vision (which is known as presbyopia) before age 40. When this change begins to occur, they cannot choose to start holding reading material farther and farther away from their eyes, as normally sighted people tend to do at this age. So, instead, they may resort to frequent use of magnifiers and other adaptive methods and materials. This often means turning to large print materials for at least some of their reading. Fortunately, large print books have become increasingly available at libraries and bookstores and through mail order sources.

Adapted lifestyles and adapted environments: Some persons with achromatopsia, even though they have good tinted lenses, visors, and other items which make it possible for them to function adequately in the sunny outdoors, have chosen to focus on adapted lifestyles, in which the need for such aids is minimized and the demands on their vision are limited. They find or create favorable environments, and they make vocational and lifestyle choices that allow them to have maximal visual comfort and visibility and minimal visual stress. They emphasize the use and enjoyment of their good rod vision, and they like to schedule their outdoor activities at twilight or at night time.

Special Needs in School



Classroom Lighting

Unfortunately for achromats, classrooms are usually designed and illuminated to meet the visual needs of the rest of the sighted population. Bright overhead lighting, sunny windows, and light colored desks and walls tend to be the rule. Sometimes there is an exception to this rule, such as a dimly lit basement classroom, where other people may complain about the lighting but where achromats function comfortably.

Fluorescent lights, so commonly used in classrooms, are especially hard on our eyes. Some achromats wear visors or caps to shield their eyes from harsh overhead lights. This sometimes requires obtaining special permission in grade school and high school, if school regulations forbid wearing caps in class. Some classrooms are so bright that students with achromatopsia have to wear their “outdoor sunglasses.”

Many factors go into the decision about the best place to sit in a classroom for optimal vision. As with other matters, a student’s preference should be given prime consideration, even though the student may be too young to really understand and explain why a certain place works best. Only those who have achromatopsia really know what it is like to see with this vision disorder.

It is possible to darken a student’s desk by covering it with a colored desk pad, construction paper, a piece of masonite, or other dark material. It also may be possible to reduce the light at a student’s work area by covering a window, closing blinds, or turning off a section of overhead lighting. One teacher obtained a lamp that emitted soft, diffused lighting to use, as needed (in place of fluorescent lighting), near the desk of a student with achromatopsia.

For exams and other prolonged reading and writing tasks, it is often possible to arrange for a student to move to a location in the school where lighting conditions are more favorable. Sometimes a place can be found in a room adjoining the classroom, where lighting can be adjusted.

Reading Materials

Duplicated materials are often very hard to read. Photo-enlargements of required reading materials can be provided for visually impaired students, as needed. Transcribers (paid or volunteer staff) can prepare large type copies of school materials. In college, visually impaired students should be permitted to obtain enlarged copies of printed materials at campus copy centers, at the same cost as other students are charged for standard size copies. If a student cannot see what is written on a chalkboard, the teacher or a classmate can provide a copy of the material.

Adaptive Materials & Devices

Little in the way of special equipment or materials is actually needed by achromats in school. They tend to approach tasks in conventional ways. When using computers, they benefit from enlarging software, large monitors, and screen darkening controls and devices.

They can see their handwriting better if they use pens that have bold points (available in many stores), but they do not need the broad felt tip pens and bold line writing paper needed by many other vision impaired students.

Hand-held or stand magnifiers and hand-held or spectacle-mounted telescopic aids can be invaluable assistive devices in the classroom for achromats.

There are reading and writing stands which allow visually impaired students

to bring material closer to their eyes without having to bend over so much. Children commonly resist using these devices, but more mature students tend to appreciate the benefits of maintaining proper body alignment.

On the Playground

Ways for helping children with achromatopsia in outdoor play areas include (1) using the assistance of “playground buddies,” best friends, volunteers, or aides; (2) informing playground supervisors about the special problems of a student with achromatopsia and enlisting their help; (3) permission to play in shady areas (usually near buildings or trees); (4) the use of hats, caps, visors, or other devices to reduce light entering the eyes; and (5) occasional or regular use of indoor spaces during playground periods, with appropriate supervision. Integration into group activities on the playground is a desirable goal, whenever possible, and there are various ways of achieving this, depending on the specific activity.

Physical Education Classes

While each case should be assessed on an individual basis, students whose only impairment is achromatopsia should be able to participate in regular P. E. classes, if the teacher is willing to make a few minor adaptations. Elementary school P. E. teachers are usually accustomed to working with students who present a wide range of ability levels, and they usually want to include each student. They recognize that those with special needs are precisely the ones who will benefit the most from being in their classes. Occasionally, however, one encounters an elementary school P. E. teacher who is at heart a frustrated high school or college coach, who dreams of state championships and who can relate only to the athletically gifted.

Most negative attitudes from teachers stem from a lack of understanding of the student’s eye condition or from faulty assumptions about the extent of needed adaptations. The following suggestions might help parents and others who do encounter negative attitudes from teachers about the inclusion of a visually impaired student:

1. Enlist the support of teachers of the visually impaired. They should be able to offer suggestions for some simple ways to adapt P. E. activities.

2. Speak to the school administrator about offering in-service workshops for teachers on (a) awareness of differences among students and how to accommodate various special needs; (b) strategies for inclusion; and (c) the partially sighted student in the regular classroom.

3. In most states teachers must earn staff development credits to maintain their certification. P. E. teachers might be encouraged to do this by attending workshops, conferences, or courses that offer information about including students with special needs.

4. Seek out a parent group which advocates for students with special needs. They are usually very knowledgeable about students’ rights and parents’ rights, state laws regarding complaints and appeals on school issues, and where to find appropriate resources.

Suggestions for adapting physical education activities:

☆ Most P. E. activities do not require maximum lighting. Up to half the gym lights can be turned off.

☆ For ball games, if possible, use a ball that contrasts with the background. For indoors this may mean using a dark ball or a striped ball. For kickball outdoors, a light colored ball will contrast more sharply with grass and dirt.



- ☆ For students with achromatopsia, kickball or bounce ball offer a larger target and an auditory cue about the position of the ball.
- ☆ Obstacle courses, in which students crawl over, under, around, or through various things, are of particular benefit for visually impaired children and others who may have poor depth perception.
- ☆ Activities done with a partner – bag races, wheelbarrow races, etc. – are excellent for these students. They can move through space with increased confidence.
- ☆ Along with physical demonstrations, teachers should provide clear verbal descriptions of the motor activities being taught to students.
- ☆ Many P. E. activities require no adaptation at all. These include tug-of-war, jumping rope, wrestling, weight lifting, parachute play, calisthenics, martial arts, trampoline, swimming, and many forms of gymnastics.

Color in School Activities

All students with achromatopsia are either totally or partially colorblind, and teachers need to be made aware of this. When school activities involve color matching, color identification, color coding, or other uses of color, modified or alternative assignments can be arranged for these students.

Achromatopsia Awareness

Everyone benefits when the school staff and the other students at school become sensitized and educated regarding achromatopsia. A special education teacher, a parent, or the student with achromatopsia might make a presentation on this subject to a class or to a staff meeting.

Individual Differences

Some students with achromatopsia use large type books; others read books with standard type held close. Some use both. Some use magnifiers often; others use them very little or not at all. Some benefit from using CCTV's; others do not. Some find it easier, with computer or CCTV monitors, to read white text on a black background, and others prefer black on a white background. A student's preference in all these matters should be given full consideration. Individual preferences are influenced by degree of vision impairment, age, school level, and various other factors.

Disabled Student Services

Disabled student centers at colleges and universities should offer these services to vision impaired students:

- ☆ Free or low cost access to enlarged copies of class materials
- ☆ Assistance with registration, financial aid applications, and other procedures
- ☆ Help with orientation to the campus
- ☆ Loan of tape recording equipment
- ☆ Advocacy when problems arise with specific teachers and classes
- ☆ Assistance in using the library
- ☆ Reader services ("readers" can also serve as notetakers, drivers, etc.)
- ☆ Assistance in finding textbooks and supplies in the campus bookstore
- ☆ Access to adaptive equipment (large computer monitors, enlarging software, CCTV's, etc.)
- ☆ Information and referral re: other resources for the visually impaired
- ☆ Counseling regarding disability issues
- ☆ Special arrangements for exams – enlarged materials, extra time for tests, and visually comfortable settings.
- ☆ Lockers for special equipment, etc.

A substantial amount of helpful information about how networkers have dealt with the subject of "Special Needs in School" can be found in the book, *Living with Achromatopsia*. See information about this book on page 150.

Reading Formats Used by Achromats

With regard to reading, network members fall into these 3 categories:

(1) The over-40 age group: Most of us in this category grew up using standard print in school. We brought reading matter close to our eyes and did whatever else was necessary to cope. This was not easy, but we experienced the advantages of being able to access a wide range of printed materials throughout life as a result of our proficiency with standard print. Some persons in this category had access to magnifiers and large print in school. Some were even required to use large print when they did not want to.

(2) Adult achromats in their 20's and 30's: Most of these grew up having special education services and access to large print and magnifiers. Many considered large print an option, rather than a necessity – to be used only when needed (e.g., for textbooks in the upper grades or assignments involving fractions, graphs, maps, etc.).

(3) Children and teens with achromatopsia: With increased options in technological and optical devices and large type reading materials, those in this category have had considerable access to enlarged print.

Many factors can influence whether an individual actually needs large print or is able to work with standard print. First, there are the variables in visual functioning – i.e., the differences in visual acuity of complete achromats and incomplete achromats. Other factors include the visual environment (i.e., whether the lighting is favorable for one's vision) and whether one has worked with standard print from the start of school or was provided with large print early on. Some became dependent on large print, and others, out of necessity, became so accustomed

to coping with regular print that they tended to reject large print later on, when it became available to them.

And now there is another factor to be considered. In the last 10-15 years, many children with achromatopsia have been fitted early in life with dark tinted lenses to alleviate their problem of light sensitivity (such lenses were not easily available before that time), and indications are that, in many cases, this has led to a dependency on dark lenses, even in environments having only moderate lighting levels.

It is not likely that scientific studies will be conducted regarding these factors, but reports indicate that there is a significant difference in a person's ability to tolerate light if he/she became accustomed to wearing dark lenses in infancy or early childhood. Concerned network parents have been doing what they can to moderate the use of dark lenses by their children, so that the children can have maximal use and enjoyment of their rod vision, with minimal dependency on dark lenses.

Wearing dark lenses in rooms having low or moderate light levels affects the size of print one is able to read. Achromats who did not have really dark lenses until late childhood or adulthood established, early in life, the habit of removing their sunglasses promptly upon coming indoors. Doing so was natural and instinctive, as well as necessary in order to have optimal visual functioning in any room that was not brightly illuminated.

To understand the effect dark lenses can have on reading and writing, one needs only to try wearing dark lenses in indoor settings where they would normally not choose to do so. Networkers who have experimented with doing this have become aware of how much

more difficult reading, writing, and other visual tasks become.

When I started this network, one of the reassurances I wanted to offer network parents was that, even though large print is a nice option to have, we achromats are capable of managing reasonably well with standard print, just by getting close to what we are reading and, when necessary, using magnifiers. This has been true for all adult achromats I have communicated with, and it has been true for myself. But I began to hear from some parents that their children were not able to read standard reading materials in school – sometimes not even in the early grades, when school materials tend to have easy-to-read print. Upon inquiry, I would learn that these were invariably children who were fitted with very dark lenses early in life and who had become accustomed to keeping tinted lenses on most of the time, even in settings where the light was not very bright. See pp. 60-63 and 99-100 for more on this subject. Hopefully, continuing dialogue among networkers will lead to guidelines for preventing – or at least minimizing – dependency on dark lenses among children with achromatopsia.

Another factor that has entered the picture regarding options for reading has been the recent trend among some special education teachers to teach all low vision children to read and write Braille. Some proponents of this idea are very zealous in their recommendations that all children who have to read print up close or who use large print should also learn Braille.

This trend developed after a period of many years during which Braille had fallen out of favor due to the growing availability of tape recorded reading materials, optical and technological reading aids, and access to printers and copiers which could produce large

print. Then, during the 1980's, some concerned educators of the visually impaired began to speak out about the need to return to Braille as an important option not just for the blind but also for many low vision persons – specifically, those for whom the effort to read print using optical or technological aids caused stress to the eyes and fatigue. They pointed out – and rightly so – that Braille proficiency is important for those whose vision is expected to deteriorate in the future, leaving them unable to continue functioning as print readers. They also pointed out that there are many students with low vision who can benefit by using Braille for some reading tasks and print for others.

This approach makes good sense for children with progressive eye conditions, whose vision is so limited that they do at times need to read by touch. But, unfortunately, some of these promoters of Braille for low vision students have sometimes recommended this for children with non-progressive vision disorders, who are quite capable of being print readers and have no need for Braille. Achromatopsia is non-progressive, and achromats have a great deal of usable vision. (However, it is important to remember that misdiagnosis is common. Some children diagnosed with achromatopsia may actually have some other eye condition instead. Obtaining an accurate diagnosis is of the greatest importance.)

Children with achromatopsia usually resist the efforts of well meaning teachers to teach them Braille. Very few teachers know about achromatopsia, though they may be knowledgeable about other vision disorders.

Since starting this network, I have been in touch with hundreds of adult achromats – some in their 50's, 60's, and 70's. All of us are print readers. None of us has ever needed Braille.

Information about Special Education in the U.S.

Following are excerpts from *When You Have a Visually Handicapped Child in Your Classroom: Suggestions for Teachers*, by Iris Torres and Anne L. Corn, published by the American Foundation for the Blind, 1990.

When a visually handicapped child is enrolled in a regular class, the child may need to cope with academic and emotional stresses usually not encountered by non-disabled children. To ensure that the child is given the opportunity to reach full academic potential, the child will receive the services of a certified teacher of visually handicapped students.

One type of teacher of visually handicapped students is a resource teacher. Permanently based in your school, he or she is available throughout the school day for consultation, specialized instruction, and the provision of materials especially adapted for visually handicapped students. The resource room may be likened to a library in which the student spends one or two periods a day and receives small-group instruction and the necessary adaptive materials. Another type of special teacher is the itinerant teacher, also known as an educational vision consultant, who travels from school to school and is available on a scheduled basis.

The older the child is, the better he or she is able to communicate abilities and limitations. With the understanding of supportive educators, the visually handicapped student will gain self-assurance and independence.

The Education for All Handicapped Children Act, P.L. 94-142, signed into law on Nov. 29, 1975, and the 1986 amendments to the act, P.L. 99-457, guarantee that all handicapped children, including visually handicapped children, will receive a "free appropriate education in the least restrictive environment." The least restrictive environment differs from student to student and will vary

according to the student's abilities and the skills acquired by the student during his or her school career. For some students, the least restrictive environment may be the neighborhood school with regular visits from a qualified teacher. Other students may be bused to a school where they receive support services from a resource room teacher on a daily basis.

The decision to place a child in the educational setting that is the least restrictive environment is a cooperative one based on the assessed specialized needs of the child. The parent or guardian, a member of an assessment team, a representative of the local educational agency, the child's teacher or teachers, and, when appropriate, the visually handicapped student participate as a decisions-making committee. Once the appropriate educational setting is selected for a student, an Individualized Education Program (IEP) is developed by the same committee or team. Annual or long term goals are written out for use as a guide to instruction for the student's teacher.

In general, educators and professionals use the terms "visually impaired" and "visual impairment" rather than "visually handicapped" and "visual handicap." However, many of the laws governing special education stipulate that a child must be handicapped by his or her visual impairment in order to be eligible for services, and so the prevailing language must be used. In this publication, the term "visually handicapped" will be used, and it encompasses all degrees of vision loss (including total blindness) that affect a person's ability to perform the tasks of everyday life.

Options in Tinted Lenses for Persons with Achromatopsia

Tinted lenses play an exceedingly important role in the lives of persons who have achromatopsia. They reduce glare and allow them to have significantly improved vision in higher levels of illumination. But, for many networkers, the road to finally obtaining lenses that have really helped them with their special needs has included a great deal of frustration and considerable expense in connection with lenses that did not work out. The most commonly reported problem is that of having to work hard to get lenses that are dark enough to be really useful to them outdoors.

The history of the kinds of lenses achromats have managed with over the decades is as interesting as the story of what they are able to obtain now. I invite members of the network to share information about the tinted lenses they have worn in the past, as well as information about those that they presently use.

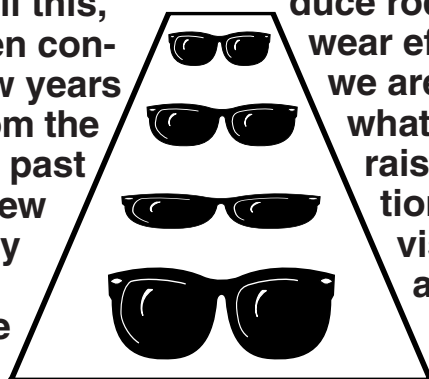
It is amazing what can be done these days for persons with abnormal sensitivity to light. Recent advances in optical dyes make possible a wide range of tints, and there are also useful options in lens treatments and lens coatings. Also, there are increasingly more ready-made sunglasses on the market that are designed to deal effectively with glare. This is, in large part, due to the growing concern that people have about protecting their eyes from harmful rays. Some of the new styles are very useful to achromats. And, in addition to all this, wearing sunglasses has been considered “cool” for quite a few years now. This is very different from the way it was when those of us past 40 were growing up. Just a few decades ago, there were very few choices in tinted lenses available to us. Basically, the choices we had consisted

of (1) relatively ineffective “drugstore sunglasses,” (2) glass lenses coated with dark gray dye – which would become nearly black by the time the lenses were made dark enough to help achromats outdoors, (3) bulky red goggles designed to help people who worked in darkrooms remain dark adapted whenever the lights were turned on, and (4) welder’s goggles or welder’s glass set in flat frames.

Achromats can’t just decide to wear sunglasses on a whim, like so many other people can. For us, they are a necessity, rather than an optional fashion accessory. Even so, some achromats have managed to work some flair, fantasy, and fashion into their wardrobe of sunglasses. Some collect lots of sunglasses, and others stick with a couple of tried and true favorites.

Although achromats have little or no cone vision, they have good rod vision (with the exception of those individuals who have some additional eye problem affecting their rods). For the normally sighted, the rods function at moderate and lower levels of illumination. But, for achromats, rods must work overtime, so to speak, helping us to function visually in all levels of illumination, since the cones are not there to become activated when lighting requires cone vision.

Our rods can serve us well, if we can keep them from being “bleached out” by higher light levels. This “bleaching out” is known as rod saturation. To reduce rod saturation, we need to wear effective tinted lenses when we are in bright light. Put simply, what we need are lenses that raise our level of visual functioning and reduce our level of visual impairment – and which also meet whatever other criteria matter a lot to us as individuals.



Factors that Affect Our Choices

Many factors influence the choices that achromats make in tinted lenses. Following are some of these factors:

1. Degree of photophobia (how light sensitive a person actually is). Complete rod monochromats tend to be considerably more photophobic than incomplete rod monochromats or blue cone monochromats. And, within each of these sub-groups of achromats, there is great variability in light sensitivity.

2. The kinds and levels of illumination that one actually must cope with on a regular basis. There are many variables in people's lifestyles, geographic locations, and home and work environments, etc. For example, the needs of a child who spends much time in brightly lighted classrooms and on playgrounds are quite different from the needs of someone whose lifestyle includes only limited outdoor excursions and little exposure to harsh indoor lighting.

3. Whether one uses Rx lenses and, if so, how strong one's prescription is. Some achromats have chosen not to wear Rx lenses, so they are able to use a variety of wraparound sunglasses, rather than being restricted to using standard spectacle frames that accommodate Rx lenses. Some prescriptions, however, can be incorporated into the lenses made to fit highly curved wrap-around frames, but this is not possible for all prescriptions. Wraparound sunglasses offer much more peripheral vision, and they minimize problems of peripheral glare, light leakage, and reflections on the inside of tinted lenses – problems which achromats commonly have when wearing dark lenses. When one must wear conventional spectacle frames for the sake of Rx lenses, the frames are sometimes fitted with opaque side shields to block light.

4. Whether one has the assistance of an optometrist or optician who is willing

to experiment with various tints, lens coatings, frames, side shields, etc. Helping achromats with their special needs in tinted lenses is not easy, because their needs do not fit the norm – to put it mildly. Lenses for achromats may have to be tinted darker repeatedly or bleached and then tinted again. And, if the optometrist or optician sets out to make a patient custom tinted lenses to mount in wraparound frames, this can be very tricky indeed. All of these procedures require patience, experience, and skill – and also, hopefully, dedication to helping patients whose lives can be so dramatically changed by successful results in achieving lenses that are tinted “just right.”

Usually the tint which will turn out to be just right for someone with achromatopsia is one which non-achromats, even the optical specialists, cannot judge accurately. Non-achromats simply are not able to see the way we see and, at best, can only make educated guesses about what will work for us.

Besides needing to get darker tints, achromats also need to try on a lot more frames than other patients do. For them, it's not just a matter of comfort and appearance but also a matter of finding frames that fit their faces snugly, blocking as much light as possible. Or it may also be a matter of finding frames that will work well in combination with clip-on tinted lenses or with wraparound (fit-over) sunglasses or whatever else a patient may use for maximum glare protection in extra bright settings.

Achromats who have caring, helpful, and resourceful vision care professionals to turn to are fortunate indeed. Those who aren't so lucky have to work a lot harder at articulating and asserting their special needs.

5. Financial resources. Networkers have reported benefiting from tinted lenses ranging from cheap (like “blue

blockers” from a discount store or curved plastic tinted lenses that slip behind conventional frames) to very expensive (such as custom designed sunglasses made with high quality lenses and designer frames). Among the most expensive lenses being used by networkers are the Corning ground glass photochromic lenses.

6. The relative importance of various other factors, such as ophthalmic lens quality, comfort, aesthetics, durability, or the amount of care required in using certain kinds of sunglasses. One person, for instance, may barely notice that a pair of glasses fits a bit too snugly, while another may become irritable or even develop a headache as a result of the pressure. Some people deeply appreciate (or may insist on) the clarity of ground glass lenses or the better quality plastic or polycarbonate lenses, while others are content with lesser quality plastic lenses. Inexpensive polycarbonate wraparound sunglasses now on the market are popular with many achromats, because of the full wrap-around tinting they provide, the built-in top shields, and their affordability. But they are fragile and must be handled carefully, so they do not make a good choice for someone who is impatient or hasty by habit or who is limited in manual dexterity.

7. Concerns about appearance and style and about not wanting to look conspicuous. Concerns such as these tend to vary considerably with age and specific situations. Image usually becomes increasingly important in the pre-teen years and tends to remain so into young adulthood. Image takes on different levels of importance in different types of work. And people vary greatly in their personalities and their sensitivities. Some individuals are highly sensitive about their appearance throughout their lives.

8. Personal preferences and reactions with regard to the specific lens colors that work well for achromats outdoors. These colors include ultra-dark shades of amber, brown, orange-red, plum, red, and reddish brown – all of which are at one end of the spectrum, so that lenses with these tints transmit light which the rods can tolerate well. Dark gray lenses are also valued by some achromats, and some report using dark blue lenses. Magenta lenses are often prescribed for blue cone monochromats.

9. Individual reactions to certain options which eye care professionals often suggest for achromats: (1) lenses that make the world appear very dark, in order to have better acuity (some achromats reject this, opting instead to have lots of sunshine in their lives); (2) side shields and visors (some persons are very uncomfortable wearing anything that restricts their peripheral vision), and (3) tinted contact lenses (some love this option, and others find it unacceptable – comments on this subject are included in the “Comments from Networkers” section).

10. The matter of what one is accustomed to and comfortable with – i.e., what kinds of lenses already play an established role in one’s way of seeing.

11. The “trade-offs” which different individuals are willing – or not willing – to accept and live with. Every option for dealing with extreme light sensitivity involves some kind of trade-off. There simply is no perfect solution that has only pluses and no minuses.

12. All the other individual differences and peculiarities that make human beings so unique and interesting.

When one considers all of these different variables in people’s lives, it is evident that we simply cannot assume that what works well for one person is necessarily right for others.

Trade-Offs – Pluses and Minuses

The choices we make in tinted lenses invariably involve trade-offs. For example, if someone who wears standard spectacle frames (because of Rx lenses) has opaque side shields attached to the frames for blocking out side glare, this will make it possible to see better through the lenses, but peripheral vision will be restricted. Another kind of trade-off: Any adaptive aid which offers maximal vision for achromats in bright light will stand out in one way or another. Those who avoid wearing anything that might be considered unattractive or that might call attention to themselves must be prepared to cope somehow with the reduced vision which inevitably occurs for them in bright light.

Some achromats choose to function with less than maximal vision in bright light – and not just for the sake of avoiding conspicuous sunglasses, because there is yet another trade-off to be considered here. Habitual (rather than occasional) use of tinted lenses that are dark enough to create twilight conditions for an achromat (thus permitting the best visual acuity for them) has been known, at least in some cases, to lower one's tolerance for light, leading to the need to wear tinted lenses even in lower levels of illumination – i.e., in the kind of lighting in which achromats are usually quite comfortable. Noticing such changes in their ability to tolerate light has prompted some persons to restrict their use of ultra-dark lenses to those times when maximum light protection is actually needed.

So much of what we experience when trying something new – and whether our reaction to it is positive or negative – has to do with what we are accustomed to. For example, someone who has grown accustomed to seeing through fine ophthalmic quality lenses will not be likely to

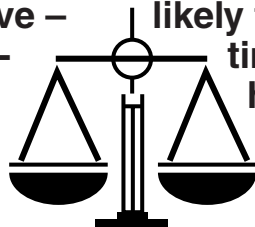
accept lenses of lesser quality, even when the lenses are part of a well designed pair of sunglasses which offer full wraparound tinting and other helpful features.

Someone who has little or no experience in using extra-dark lenses (either by choice or because such lenses were never made available to them) is likely to have developed considerable ability to adjust to a wide range of light levels and so may resist wearing anything that reduces the level of vision they are accustomed to having in moderate or lower lighting levels. By contrast, someone who has grown accustomed to using super-dark tints a lot of the time will find it hard to adjust to lenses that are not so dark. Becoming chronically dark adapted can happen so easily.

Someone who has grown up experiencing their full peripheral vision, because outdoors they have worn only non-Rx sunglasses that wrap around instead of spectacle frames fitted with tinted Rx lenses, will likely reject any glasses which have opaque side shields attached, even though the combination of good quality dark lenses and opaque side shields may allow them to have better visual acuity outdoors than they have ever had before.

Those who have grown up wearing spectacle frames fitted with Rx lenses and opaque side shields are very impressed when they try tinted contacts and experience the full visual field that contacts allow them to have. However, those who have not grown up using Rx lenses and so have not been accustomed to wearing either spectacle frames or opaque side shields are less likely to be impressed by this aspect of tinted contact lenses, since they have never had their visual field restricted in this way.

The examples just listed are based on numerous reports



received from networkers regarding tinted lens options. See this section, which begins on p. 80.

There is no perfect solution to achromats' problems with light sensitivity, no single option that fits everyone's needs, is problem-free, and works in all situations. Each option has advantages and disadvantages. The trade-off may not be something that a person using a particular option is even conscious of. Eyes are very adaptable organs and people are very adaptable creatures. We tend to assimilate the trade-offs whenever something is helping us with our special needs, especially when we are very young. The older we are when we experiment with new options, the more awareness and ability to compare and evaluate we bring to the experience and the more likely we are to have already established our "comfort zones."

Ideally, persons with achromatopsia would have access to (1) information about all of the options available to them and what's possible for optometrists and opticians to obtain or to create, using custom tinted lenses or modified lenses and frames; (2) the services of vision care specialists who are genuinely interested in helping achromatopsia patients with their special needs; and (3) the chance to try out a variety of lenses, frames, etc., in order to find out what works best for them.

For a long time, it has been standard practice at low vision centers to make optical aids available on loan for low vision patients to try out for a period of time, understanding that these patients need the chance to "live" with a specific optical aid, using it in different kinds of situations before being certain about whether it will work for them. Hopefully, someday the need of achromats to have specially tinted lenses will be recognized as just as important in terms of life-changing potential as optical aids

are, and a variety of trial lenses will be made available to them to try out in their daily activities, including several options in lenses of different colors, mounted in different kinds of frames.

Some vision care specialists I have consulted during my life have made it clear that they really knew very little about my vision disorder – and not just because of the fact that they had not encountered achromatopsia patients before or the fact that their training had offered very little information about achromatopsia. What they were willing to acknowledge was simply that they were limited – as all non-achromats are limited – in being able to imagine how I see. Achromatopsia is extremely rare, and very little about this disorder, from the standpoint of those who experience it, has been put down in print, for vision care professionals or any other interested persons to read. So how can these professionals be expected to know very much about the kinds of lenses that will work best for us, the many factors that affect our choices, and the kinds of trade-offs referred to in the preceding paragraphs?

Many years ago I spoke with an eye doctor who was very open about the lack of knowledge among vision care professionals about achromatopsia. During a routine eye exam, I raised a question regarding my vision disorder; and, to my surprise, this doctor said, "You're the expert on this subject, you know." He stated that, while he could provide information about the health of my eyes, it was I, the patient living with this rare disorder, who was qualified to educate medical professionals like himself about the non-medical aspects of it. I remember feeling so much respect for him for recognizing this.

What I had consulted him about had to do with tinted lenses. I had begun wearing some sunglasses that had

been made very dark and so was experiencing remarkably improved vision outdoors, but I was noticing that my light sensitivity indoors was increasing and that I was having to wear dark glasses more of the time. I wanted to know if a change such as this in my visual functioning was something doctors understood and asked what I should do about it. He listened carefully (one of his many fine traits as a physician), agreed with my suspicions with regard to my recently reduced tolerance for light, and then advised me to follow my own instincts about what to do. It was at that point that he made the statement about my being the real “expert” on achromatopsia.

I share this story to emphasize the point that all of us with achromatopsia are “experts” on our extraordinary way of seeing and to assure networkers that there really are professionals out there who understand and respect this fact.

If achromatopsia were not so rare – if, for example, there were as many people with achromatopsia as there are persons with cataracts, arthritis, and other common medical conditions – then persons growing up with achromatopsia and parents of children with achromatopsia would be able to access a considerable amount of information about it – not just information from medical and optometric perspectives but also a substantial amount of conventional wisdom passed on by people who have lived with this eye condition. There would be much material in print written by achromats who had experimented with different approaches to their hypersensitivity to light and had shared their advice and the results of their experiments.

If achromatopsia were a common disorder, there would be peer support groups and self-help articles in popular magazines about it. One might even

have neighbors or friends with achromatopsia who could be consulted.

But, of course, this is far from the reality. Medical knowledge about achromatopsia is barely adequate, and other information about it is limited mainly to our network books and newsletters.

Reports from networkers reveal that, in families where there is more than one child with achromatopsia, it is common for different siblings to choose different options in tinted lenses. In some families, for instance, one sibling uses tinted contacts and the other uses tinted spectacle lenses. In another family, one child requires glasses made with very dark lenses with Rx, and the other is content with some off-the-rack wrap-around sunglasses with mirrored lenses and prefers not to wear Rx lenses.

Some of these differences between siblings are clearly indicative of the differences in individual personalities and levels of vision impairment, but another contributing factor might be the different ways in which a child’s light sensitivity has been “treated” early in life. For example, one network parent reported that she used to feel guilty because she had waited so long to get really dark sunglasses for her first daughter; so, when a second daughter was born with achromatopsia, she made sure to have her fitted early on with the special dark (red) lenses that she had obtained for the older girl. However, this mother has noticed, with some concern, that now, several years later, the younger daughter almost never voluntarily removes her dark red lenses – while the older daughter, the one who did not get fitted with special dark glasses early in life, has always been able to comfortably remove her glasses when coming indoors. Other reports from networkers also seem to indicate that, the earlier that children are fitted with very dark lenses, the more they seem to develop

a dependency on wearing dark lenses, even in settings where the lighting is not particularly bright.

With diagnosis being made so much earlier these days, parents of children with achromatopsia must make difficult choices while a child is still an infant – regarding when ERG's (electroretinograms) are done, what kinds of tinted lenses to obtain, whether to introduce Rx lenses, etc. Vision care professionals vary considerably in terms of the lenses they recommend, and very few of them are truly knowledgeable regarding the needs of achromats.

In some ways, life was simpler for achromats who grew up in the past and for their parents. The challenge is to utilize the resources available these days without making choices which could lead to problems for a child in the future. Some parents buy several kinds of lenses, some darker than others, and then monitor the child's use of the darkest lenses, to prevent excessive use of dark tints, which can lead to decreased tolerance for light.

Readymade Sunglasses

Some of the least expensive options in tinted lenses are readymade (over-the-counter) sunglasses with tints dark enough to be useful outdoors. This was the main option available to achromats in the not too distant past, and it is the choice of many achromats today who choose not to wear Rx lenses or who wear Rx contact lenses. Most adult networkers in the over-40 age group had only readymade sunglasses early in their lives. They made the most of such tints and frame styles as were available in stores.

If one shops around and tries on many different styles of sunglasses, it is possible to find some that fit snugly, have an acceptably dark tint, and block peripheral light reasonably well.

Only an achromat can view a display of sunglasses and spot ones with a tint that is likely to be dark enough for them. Non-achromats simply cannot see tints the way achromats see them.

Often tints dark enough for achromats can be found among sunglasses called "blue blockers." Some brands of blue blockers are darker than others. And, within a display of the same brand, one can usually find some that have a darker tint than others.

Other readymade sunglasses useful to achromats include those designed for use in high glare situations, such as snow sports, water sports, and mountain climbing. These are found in shops specializing in outdoor sports gear. The lenses are typically a dark amber. As in the case with ordinary sunglasses, some are darker than others.

The optical quality of readymade sunglasses ranges from poor to excellent, usually reflecting the price. Some of the brands networkers report using include Bollé, REI, Revo, Vuarnet, Zeiss, and Bausch & Lomb.

Some networkers who choose to buy over-the-counter sunglasses do so in order to be in fashion, but others mention convenience and affordability. The lenses in readymade sunglasses are hardly ever as dark as custom tinted lenses made specifically for achromats, but some network members have pointed out that regularly wearing sunglasses that are not super-dark has allowed them to be more adaptable when making transitions between outdoors and indoors and between bright outdoor areas and shady outdoor areas. They don't have to stop and remove – and then replace – their sunglasses as often as when moving about in varying lighting conditions wearing super-dark sunglasses. However, for more extensive periods of time in bright sunlight

or times when safety is an issue or a visually challenging activity is involved, those who regularly wear readymade sunglasses generally have on hand some extra dark sunglasses.

Achromats who routinely wear hats, caps, or visors generally do not mind so much whether the frames of their glasses fit snugly in order to block out all peripheral light. The need to reduce peripheral light as much as possible is more important to those who choose not to wear anything that shields their eyes from overhead light. Many persons dislike wearing hats, caps, and visors. Some feel it is important to maintain their full peripheral vision, even if it means less visual acuity.

Over-the-counter sunglasses were not created with achromats in mind. Tints that are dark enough to help achromats significantly outdoors are neither desirable nor advisable for normally sighted persons, who only need extra dark lenses in situations involving extreme glare, such as mountain climbing, or when the pupils of their eyes have been dilated for eye exams. And having frames that fit snugly to one's face is not of great concern to most people. But, for achromats, light entering the frames from the sides, from above, or from below can make a big difference in how well they can see through the lenses. So achromats need to spend more time trying on sunglasses than other people.

Although few readymade sunglasses have lenses dark enough for achromats outdoors, it is not hard to find sunglasses with frames that fit closely around the face. In recent decades numerous such designs have been created in response to the growing concerns about protecting one's eyes from ultraviolet rays. Achromats have benefited from this trend in sunglass designs.

Snugly fitting frames can be fitted with dark lenses, making it possible to have wraparound protection from high levels of illumination which their retinas cannot tolerate.

There can be significant differences, however, in how well various sunglasses fit different individuals. On the rare occasions when achromats get to meet one another, they naturally check out one another's favorite sunglasses. Typically it is found that one person's favorite sunglasses do not fit the other person. Head sizes and facial structures vary so much. Also, sunglass features desirable to one person may not even be acceptable to another.

Some adults with narrow faces or small heads find they can wear sunglasses designed for children, which block out light better for them than sunglasses designed for adults.

It pays to shop around. One networker found that inexpensive "glacier glasses" (designed for use in snow sports), with soft leather side shields, sold at a sporting goods store fit her face perfectly and comfortably, while a similar pair with a famous name brand and selling for five times as much had stiff, uncomfortable side shields, did not fit her head well, and let in a great deal of light at the sides.

Lenses in readymade sunglasses can be dyed darker if the lenses are plastic (it is very difficult to add tinting to polycarbonate lenses). The charge for having this done is relatively low. If you are not sure what the lens material is, ask. Several networkers have reported success using this option at shops where lots of sunglasses are sold, such as LensCrafters. This option is not possible with lenses that have a special coating (such as a scratch resistant coating or mirror coating).

It is important to understand that

having extra tinting added to sunglass lenses is not the same as having custom tinted lenses made at an optical lab. In shops selling sunglasses, technicians who do the extra dyeing cannot be expected to bleach and then re-dye the lenses if they become too dark, so it is important to monitor carefully the progress when making the lenses darker. Also, the resulting tint and lens clarity will not be comparable in quality to tinted lenses made at an optical lab.

Custom Tinted Lenses

A major option in tinted lenses for achromats is having custom tinted lenses made to fit selected frames. This option has been around for a long time, but only in recent decades has it been possible to choose from such a wide variety of tints, and the choices in frame styles were more limited in the past. Adult achromats and parents of children with achromatopsia would do well to learn all they can about this option.

Although they may not be familiar with the procedures involved in having custom tinted lenses made for sunglasses, many achromats are familiar with having custom tinted lenses made for the glasses they use indoors when the lighting is too bright for their comfort but not so bright as to require sunglasses. Some achromats make considerable use of tinted lenses indoors; others prefer not to use them indoors unless absolutely necessary. A major determining factor is whether one regularly spends time in bright classrooms or workplaces or has the freedom to choose comfortable lighting and favorable locations in their day-to-day life.

Another determining factor can be whether a person began using very dark lenses early in life. Reports from networkers indicate that those who have worn dark lenses since infancy or early

childhood tend to wear tinted lenses indoors as well as outdoors.

For indoor tinted lenses, one finds desirable frames and then arranges for lenses to be made with the tint of their choice. If they use an Rx, the Rx is included in the lenses. If they do not use an Rx, then plano (non-Rx) tinted lenses are ordered. When the tinting is done, the patient tries them on to see if more (or less) tinting is needed.

The possibilities for indoor tints for achromats are many. They can choose from the tints which normally sighted persons select when having tinted lenses made for use outdoors. At optical dispensaries and shops where opticians' services are provided, there are tinted lens "blanks" to try out. These trial "blanks" are in various shades of gray, green, and brown. Of these standard tints, achromats find that the brown ones appear darkest to them. None of these trial lens tints is dark enough to help achromats outdoors, but some of them can work well for their indoor glasses.

Various achromats have found that rose, blue, or amber lenses work well for them in indoor lighting and have had their opticians make indoor glasses for them using one of these tints.

Some eye care professionals prescribe a red tint for achromats' indoor glasses, since red is often used as an outdoor tint for achromats. But, for indoor lighting, there is no evidence that a red tint is any more effective than brown, gray, or other lens colors. Achromats should have the chance to try out a variety of possible tints for use in bright indoor settings.

In contrast to the ease with which one can obtain custom tinted lenses useful for indoor wear, obtaining custom tinted lenses that work well for achromats outdoors tends to be a much

more complex process – but well worth the effort. It requires patience and co-operation from a competent, experienced lens technician, and ideally the procedures are supervised by an optometrist well acquainted with the special needs of achromats and techniques for achieving dark tints in the optical lab.

The first step is to find frames that fit well, minimizing the possibility of light entering around the frames.

Persons who don't use an Rx may choose from the numerous options in wraparound sunglasses on the market, as long as the lenses in the sunglasses can be removed to allow custom tinted lenses to be mounted in their place. This option cannot be used by someone who must use an Rx for nearsightedness (myopia) or a strong Rx for farsightedness (hyperopia), since lenses having such a high curvature cannot incorporate such prescriptions. But this option can be used when a weak Rx for farsightedness is needed.

Including an Rx for farsightedness is less of a problem when having lenses made to fit wraparound frames for very young children, because wraparound sunglasses made for small heads do not have as high a curvature as those made to fit large heads.

Since it may be necessary to try on many frames and sunglasses in order to find a really good frame for use in creating custom tinted sunglasses, it is wise to enlist the aid of someone working at the store or optical dispensary. Also, taking time to explain why you need to try on so many sunglasses or frames can make matters easier. Some networkers have found that it can help to bring along literature about the special needs of achromats.

Make sure that the frames are sturdily made and will hold up to considerable use, since much trouble and expense

will be involved in having tinted lenses made and mounted in them. However, the frames do not have to be expensive. Networkers often report finding very inexpensive sunglasses that have well designed frames. It has come as a shock to many network parents to learn how expensive it can be to have custom tinted lenses made for small children's glasses and then to find that these lenses can be mounted in the frames of sunglasses that cost as little as \$5-\$15. Of course, the lenses can be mounted in more expensive frames, which sometimes have some especially nice features. But, as far as achromats' requirements in frames are concerned, expensive does not necessarily mean better, and cheap does not necessarily mean inferior.

Some eye care specialists strongly oppose the idea of using the frames of inexpensive sunglasses in combination with special lenses made for children with achromatopsia, and they insist on using the spectacle frames available in optical dispensaries. But spectacle frames do not have the very important wraparound, close-fitting design which benefits achromats so greatly and which so many sunglasses offer. The frames these doctors recommend are best when it is necessary to incorporate an Rx (with the exception of a weak farsightedness Rx, which can be incorporated in wraparound lenses, as mentioned earlier). But seldom do spectacle frames fit a person's face well enough to block peripheral light as effectively as do wraparound sunglasses. That is why spectacle frames for patients with achromatopsia often get fitted with side shields.

Achromats whose Rx restricts them to wearing standard spectacle frames (rather than wraparound sunglasses) usually have two or more pairs of

glasses made, with different tints for different light levels. To deal with the problem of light entering around the edges of the frames, some arrange to have opaque side shields attached to the frames of glasses they will wear in bright light. Some buy “ski glasses” (also called “glacier goggles”), which already have side shields, and then have the lenses in these frames replaced with custom tinted lenses incorporating their Rx. Seldom are the existing lenses in ski glasses dark enough for achromats outdoors, although they have worked well for some achromats for snow sports, when used in combination with tinted contact lenses.

Many achromats who wear tinted lenses mounted in spectacle frames find side shields completely unacceptable, for reasons having to do with comfort, appearance, or their desire to have full access to their peripheral vision. Therefore, they do what they can to find close fitting frames instead.

Another option is that of sunglasses that do not have a high curvature (the curvature is more like that of spectacle frames) but which have side lenses mounted in wide temples, thus blocking light at the sides without blocking one’s side vision. The front lenses in these special frames can be replaced with custom tinted lenses, with or without Rx, and the side lenses (not dark enough for achromats) can be replaced with a darker tinted transparent material cut to fit the lens openings. This option is useful to (1) achromats whose Rx prevents them from wearing wraparound sunglasses and (2) those who do not have someone willing and able to take on the task of making custom tinted lenses to fit the frames of wraparound sunglasses.

The person desiring custom tinted sunglasses selects frames and chooses

a tint. Tints that most effectively filter light and enhance vision for achromats outdoors are at one end of the color spectrum – dark amber, dark brown, deep red, or dark brown with a touch of red. Some networkers report liking dark plum lenses, dark blue lenses, or (in the case of blue cone monochromats) magenta lenses – tints sometimes found in readymade sunglasses. Some of these tints are harder than others to achieve in an optical lab.

Another example of having custom tinted lenses made is when ordering tinted lenses for swim goggles. First, one orders swim goggles designed to be fitted with Rx lenses. (Skyline Northwest Corp., listed on p. 114, is one manufacturer of such goggles.) Then the lens technician prepares tinted lenses (with or without an Rx) to be mounted in the goggles in place of the existing lenses. For achromats, the models which have opaque material surrounding the lens openings are best. The translucent material around the lens openings in other models makes it hard for an achromat to see through the lenses. However, there are ways to darken translucent material; see one networker’s solution to this problem on p. 87.

Whether for swim goggles, ski glasses, or basic outdoor sunglasses, getting the “just right” tint for an achromat is not easy. The tinting should be done by someone who is very experienced in using optical dyes.

Typically, it is necessary for the person for whom the lenses are being made to make repeated visits to the optical dispensary or optometrist’s office to try out the lenses-in-progress before the tint is determined to be just right. If the lenses are too light, they can be dyed a bit more. If they are too dark, they must be bleached and then dyed again. Sometimes the bleaching

and re-dyeing has to be done more than once. Only achromats can accurately judge whether lenses are too light, too dark, or just right. This can be frustrating for a parent who must make decisions about lenses being tinted for a child, when the child is too young to participate in the decision-making process. Experienced optometrists and opticians at low vision clinics can sometimes make good “educated guesses.”

The person dyeing the lenses can be aided by print-outs from a lens analyzer (a spectrophotometer) which measures the transmission curve of visible light passing through lenses, making it easier to achieve a specific percentage of light transmission or duplicate the tint of lenses a patient has left as an example of what has worked well.

Whether having custom tinted lenses made at an optical lab or shopping for sunglasses in a store, there should be ample opportunity for lenses to be tried out in the kind of lighting in which they will be used. This is especially important for achromats, since their vision changes so dramatically in high levels of illumination. At low vision centers patients should arrange to walk around outdoors wearing the lenses, to see if further work needs to be done. At shops where sunglasses are sold (or where lenses are custom tinted), there may be sunny windows where customers can try out the lenses. Some networkers have reported leaving valuables with a store employee (“for security”) in order to go outside the store to try out lenses in full sunlight.

Custom tinting sunglasses for persons who have normal vision is not a particularly complicated matter. There are standard dyes and straightforward procedures which yield predictable results. Normally sighted persons who have tinted lenses made for them at

optical labs (usually for the purpose of obtaining tinted lenses incorporating an Rx) are adequately helped by the most common lens colors – brown, gray, or green – and their lenses do not need to be especially dark. In fact, if their lenses were to be left in the dye a bit too long, making them extra dark, wearing the lenses would adversely affect their vision. For this reason, opticians tend to balk at the idea of making extra dark lenses for anyone. Even when informed that the lenses will be used by someone with abnormal light sensitivity, some opticians are overly cautious about adding extra tint.

Sometimes persuasion and careful explanation about achromatopsia are needed to help them get past their tendency to believe that lenses dyed extra dark are unacceptable. Among those opticians who are not familiar with achromatopsia – and this means nearly all of them – the prevailing concern is that no lenses should leave their lab which might be so dark as to cause safety problems for the person who is to wear them. With adult patients, it is assumed that all adults drive, and opticians especially do not want to risk the possibility that extra dark lenses might impair a patient’s driving vision. Informing them about this rare vision disorder can alleviate their concerns and ensure their full cooperation.

Only optometrists and opticians at low vision clinics might be expected to have experience in making lenses for achromats. As for the others, one can assume that the special needs of achromats will probably be new to them.

If there is an optometrist or optician who is familiar with achromatopsia and available to supervise the tinting, this is ideal. When this is not the case, then the person ordering the lenses needs to understand as much as possible

about the procedures involved and be able to communicate clearly with the person who will be doing the work.

The least problematic lenses to have custom made are gray lenses. They are easy to tint and easy to make darker – repeatedly, if necessary, until the tint is right. However, these days there are so many choices in tints that not many people who are especially light sensitive opt for dark gray lenses. Unlike dark amber lenses and dark brown lenses, dark gray lenses cause reduced visual clarity and contrast. One may appreciate the glare protection dark gray lenses provide, but one does not see as well with them as with other dark tints. However, incomplete achromats often prefer dark gray lenses, since they do not interfere with such color vision as they have (as red lenses and amber lenses do), and medium gray lenses are often chosen by achromats for their indoor tinted lenses.

Achromats who choose dark red lenses for outdoors do not face special problems in having this tint made for them at optical labs. The lab simply orders a specific dye (Monochrome 600), which is available from a company called BPI (see the list on p. 114), a major supplier of optical dyes and related products. Since this dye is not one of the standard dyes used in optical labs, the cost of purchasing it might be added to the charges for having the lenses made. Labs connected with low vision clinics may already have this dye on hand, since it is commonly used when making tinted lenses for patients with abnormal sensitivity to light.

In contrast to the relative ease with which lab technicians can achieve dark gray lenses or dark red lenses, it is considerably more complicated to make very dark brown lenses or dark brown lenses with just a bit of red added – tints

which are preferred by many achromats. Achieving non-standard tints such as these can be a challenge even for very experienced opticians. But, for achromats who find that they have their best vision outdoors when using these tints or who choose not to wear red lenses (for a variety of reasons explained later in this book), obtaining sunglasses with these tints is well worth the time and effort invested.

Achieving a very dark brown tint, a dark amber tint, or a dark brown tint with just a little red in it is not simply a matter of leaving lenses in a certain dye solution for a longer period of time. It involves the use of two or more dyes, carefully timed moving of the lenses from one dye to the other, and careful observation to detect changes in the tint. So it pays to go to the best optical lab you can find and to request that the job be done by someone known to have a special “knack” for achieving non-standard tints.

When ordering custom tinted lenses, there are some things you can do to ensure the best possible results.

1. Inform the person who will be doing the tinting about achromatopsia and why such dark lenses are needed. Bring along information in print about achromatopsia.

2. Bring sample lenses or a pair of sunglasses that can be left at the lab to be used for comparison when the tinting is being done. If the lenses show the exact desired tint, that is ideal. Even if all you have to leave at the lab are lenses that are not dark enough, say, “I need lenses even darker than these.”

3. Use terms that convey clearly the tint you need. A common problem reported by networkers who have failed to get lenses tinted right for them involves the word “dark.” “Dark” means one thing to achromats but something

else to opticians. Opticians are mostly familiar with standard tints, which come in light, medium, and dark shades. But a brown lens which they would call “dark brown” is one which achromats would consider “medium brown” – i.e., it could be useful for achromats indoors but not dark enough for them outdoors. So, in order to get across clearly the kind of tint needed in custom tinted lenses for outdoors, we have to use terms like extra dark, super-dark, ultra-dark, or “dark dark dark.”

4. If you know the percentage of visible light transmission that is best for you (or for your child, if you are a parent), specify this. This is something that can be checked with a reasonable degree of accuracy using a spectrophotometer. (The quality of the equipment used at different optical labs varies, so try to have the dyeing done at a lab that has high quality, state-of-the-art equipment.) Visible light transmission does not have to do with ultraviolet (UV) rays, which all sunglasses on the market today are required to filter out. Instead, it refers to how much visible light gets through to the eyes. Outdoors achromats see best when most of the visible light is absorbed by lenses. Lenses that permit transmission of 2% to 5% of visible light are usually best for complete achromats and for many incomplete achromats in bright surroundings. Many blue cone monochromats and incomplete rod monochromats can manage with lenses passing 10% or more visible light. Some complete rod monochromats use lenses with 1% transmission in very bright outdoor settings.

The exact percentage of light absorption required varies with different lens colors. For example, amber lenses transmitting 4% visible light will appear darker to rod monochromats than green

lenses transmitting the same amount of visible light. Red lenses transmitting 4% visible light will look as dark to a rod monochromat as amber lenses transmitting only 2% visible light, yet, the 2% amber lenses offer more contrast and clarity than the 4% red lenses.

Each lens color has a different effect on how an image appears through the lenses, though some of the differences are subtle and persons using the lenses may not be aware of the differences. Many people notice how much sharper images appear through brown lenses and how much less contrast is seen through dark gray lenses. Amber lenses are good at cutting glare and are, therefore, often found in sunglasses made for use in outdoor sports. Blue cone monochromats and some incomplete rod monochromats notice how amber lenses change the colors of things. All achromats can see how certain colors are changed when looking through red lenses. Even so, many complete achromats choose red lenses for use in bright light, because they darken the world while offering them better visual acuity than is possible with dark gray lenses. Children with blue cone monochromacy and those with incomplete rod monochromacy who see some colors should not be fitted with red lenses, because red lenses interfere with such color vision as they have.

Just as lens “blanks” tinted with the standard tints used by normally sighted people are available to try out at optical dispensaries and at many of the shops where frames and sunglasses are sold (as mentioned on p. 65), a variety of tinted lens blanks are available for patients to try at low vision centers. Because many of these patients have abnormal light sensitivity, these trial lens blanks usually include a few very dark tints. It is common to find dark gray and dark

red among these trial lenses; but, unfortunately, it is not common for very dark brown lenses, dark brown lenses with some red added, or dark amber lenses to be included. There may be amber lens blanks or brown lens blanks that are slightly darker than the standard trial lenses found in regular optical dispensaries and opticians' shops, but none of the really dark tints in these colors (dark enough to pass only 2% to 5% visible light, such as is needed by many achromats). One reason these options are not included is probably because dark gray lenses and dark red lenses are so much easier to achieve in the optical lab, while there is more work involved in creating other dark tints.

Consequently, many networkers who have been to low vision clinics have been under the impression that they have had the chance to try out all possible tints when, in fact, they haven't. Some networkers have discovered these other dark tints on their own while shopping for sunglasses, thus learning that there are other dark options besides dark gray and dark red. These other options are of greater importance to some persons than to others. As explained elsewhere in this book, many achromats manage very well with red lenses, while some experience a variety of problems with them. For example, viewing the world through red lenses has had an adverse effect on the nervous systems of some especially sensitive individuals.

Achromats deserve to know what all of their options are, so it is hoped that low vision clinics will begin to offer more dark tints for them to try.

Some complete achromats manage well outdoors with lenses that allow more than 5% transmission of visible light, either because their photophobia is not as severe as is usual for complete

achromats or because they did not grow up wearing extra dark lenses and so have developed what vision care professionals call a "high tolerance for a degraded image" in bright light.

When creating custom tinted lenses for achromats, an optician will begin by using the most appropriate base color – i.e., a light, medium, or dark shade of whatever lens color has been ordered for the patient. When lenses have been ordered for a complete achromat to use outdoors, opticians should begin with the darkest base color. Then they take whatever steps are necessary to achieve the extra tinting needed for the desired results. For example, if dark gray lenses have been requested, they first dye the lenses with the darkest gray base color and then add additional tint using the gray dye. If dark reddish brown lenses have been ordered, they begin by using the darkest brown base color and then follow procedures using various dyes.

To make lenses that will fit into the frames of wraparound sunglasses, first a measurement is made of the lenses that came with the frames, to determine the lens curvature needed for the new lenses. Lens blanks having that curvature are ordered, with the appropriate Rx if the patient will be using an Rx; plano lens blanks of that curvature are ordered, if the patient will not be using an Rx. (Reminder: only certain Rx's can be put in high curvature lenses.)

Because wraparound frames are not standard ophthalmic frames, a special template must be made for cutting the new lenses to fit the lens openings in the chosen frames. Lenses are then cut from the lens blanks, duplicating the exact contour of the lenses that were in the sunglasses. The edges of the newly cut lenses are beveled to fit the grooves of the lens openings, and the lenses are dyed. When they are determined to

be the desired tint, they are mounted in the frames. The patient tries on the new sunglasses and, if all seems right, the lenses are glued in very carefully (a necessary step with high curvature lenses, so that the lenses will not pop out, but not necessary with standard spectacle frames). Some of these procedures can be very tricky, because the lenses have such high curvature. Each step must be done with precision.

For the optician, it is much easier to produce lenses having a standard curvature and a standard tint to fit into standard ophthalmic frames. Fortunately, for many achromats, there are kind, helpful optometrists and opticians who are willing to take on the work involved in creating non-standard solutions to meet special needs.

For achromats, the darker the lenses, the more important it is to deal with the problem of peripheral light. Only achromats who have worn super-dark lenses set in inadequately fitting frames can fully understand this problem. It is important that persons involved in having tinted lenses made for achromats know about this problem. Many achromats, at some point in their lives, have had the experience of being given dark lenses mounted in standard spectacle frames that allowed considerable light to come in at the sides. To achromats' eyes, the sharp contrast between the very dark lenses and the bright light entering from the sides can be overwhelming. Seeing through the lenses becomes difficult. Streaks of light, "hot spots," or reflections may appear inside the lenses, obscuring one's vision.

This problem can be prevented or minimized by using options already described – wraparound lenses, side shields, close fitting spectacle frames, the special style of sunglasses that has temples incorporating side lenses, or

wearing a hat, cap, or visor. Many achromats who grew up wearing dark spectacle-mounted lenses without protection from side glare developed the habit of holding their hands to the sides of their glasses, in order to see through their lenses. Since many eye care specialists do not understand about this problem, it has often happened that specially tinted glasses made for a child with achromatopsia have been rejected by the child. Children may not be able to explain what the problem is to a concerned adult, but they are quite capable of rejecting glasses that do not work for them.

Corning Lenses

Persons who use standard frames and who want lenses having the finest possible ophthalmic quality often choose Corning Glare Control lenses. These lenses are very expensive, but many networkers feel they are well worth the expense. In many cases, these are covered by a person's medical insurance. Made of precisely ground and polished glass, these lenses can be ordered, with or without Rx, for mounting in any standard spectacle frame, but they cannot be mounted in frames having a substantial curvature (i.e., the frames of wraparound sunglasses). The darkest tint Corning makes, 550XD, is the one most often used outdoors by rod monochromats. There are other Corning Glare Control tints useful to achromats in other lighting conditions.

Many find that these lenses not only reduce glare but also enhance contrast and depth perception. Individuals using these lenses sometimes find it hard to remove them even in settings where glare protection is not needed, because they become so accustomed to the enhanced vision these lenses provide. Some children with achromatopsia have

become overly dependent on them, wearing them even in the evenings and thus not getting to experience the improved visual acuity they should be able to have in lower levels of illumination. Parents would be wise to monitor how much time their children spend wearing tinted lenses, especially the really dark ones.

All of the Corning Glare Control lenses are photochromic; they become darker in bright illumination and lighter when the lighting is not so bright. Many networkers report that this feature is not significantly helpful to them, but some have found it helpful.

Corning glass lenses are impact resistant but not unbreakable or shatter-proof. Some readymade sunglasses have Corning lenses but not the Glare Control tints, which must be ordered by eye care specialists for individual patients. Corning lenses cannot be mounted in wraparound frames. However, some achromats have had them mounted in the special frames that have side lenses set in wide temples (the side lenses can be replaced with darker tinted plastic).

Polycarbonate Wraparounds

Another option for achromats is that of sunglasses consisting of pieces of tinted polycarbonate that fit together to provide wraparound tinting and are designed to be worn alone or over spectacle frames. Sometimes called “cover-ups” or “fit-overs,” many have narrow, built-in visors. Most networkers are familiar with those made by NoIR Medical Technologies, because NoIR makes many dark tints to meet the needs of persons with eye conditions involving abnormal sensitivity to light. NoIR wraparounds come in several sizes and are sold through agencies serving the visually impaired and

catalogs specializing in adaptive devices for low vision persons. They can also be purchased from the manufacturer (see listing on p. 114).

Other brands of polycarbonate wraparounds, some offering better construction and nicer features than NoIR lenses, can be found in various stores and optical dispensaries. But the other brands are designed for the general public, so the choices in lens colors are limited. Among these other brands, an achromat might find amber lenses dark enough to be useful or other tints to use as “fit-overs” for their spectacle-mounted tinted lenses.

NoIR offers several dark tints useful to rod monochromats outdoors. Various networkers report liking their 2% amber lenses, 4% plum lenses, 4% red lenses, and 2% gray-green lenses. The 1% gray lenses are helpful to those having the most extreme photophobia. These percentages refer to the amount of visible light transmission, as explained on p. 70. For example, 4% plum lenses transmit 4% of visible light rays.

As mentioned previously, different lens colors affect the image viewed through lenses in different ways. The 4% red lenses, for example, darken the world for an achromat as much as the 2% amber lenses, yet the 2% amber lenses create a sharper image than the red lenses. The 4% red lenses don’t appear as dark to a normally sighted observer as 2% amber lenses, because more light passes through them. But neither amber nor plum lenses distort colors the way red lenses do.

On the NoIR website and in their catalog, achromatopsia is mentioned only in connection with red lenses, even though achromats have reported to NoIR representatives that their preferences include plum lenses and amber lenses as well as red lenses. Hopefully, this

feedback from achromats will eventually be reflected in the NoIR literature.

NoIR offers sunglasses for small children made with NoIR lenses mounted in inexpensive but sturdy and well designed wraparound frames. (These are shown in their catalog but not on their website.) Any NoIR tint can be ordered. The price of these sunglasses depends on the tint. Tints useful for achromats outdoors, because they require extra dye, are the most expensive. They also offer a strap for keeping the sunglasses on a child's head, and they have devised a way to make the smallest of their regular fit-overs usable by young children. They shorten the temples (the side pieces) of the fit-overs and attach a strap that wraps behind the child's head. Rubber tubing is attached to the brow line of the front piece (as a "bumper").

In an attempt to offer more cosmetically appealing styles in recent years, NoIR has added some wraparound styles that have less of a "box-y" look. Some can be worn over spectacles and some cannot, and they fit some individuals but not others. Fortunately, some networkers report that NoIR sales representatives have been good about letting them return products if they don't fit or if a particular tint isn't right for them. NoIR offers adult sunglasses that have slightly wraparound frames, but the frames are poorly designed and poorly constructed.

Lens blanks can be ordered in any of the NoIR tints for cutting and mounting in spectacle frames. Customers can send their frames to NoIR, specifying which tint they want (an additional fee for this service is added to the cost of the lenses) or have their optician order the lens blanks, cut the lenses, and mount them in the patient's frames. Doing this avoids the hassle of trying

to get custom made lenses dark enough in an optical lab, but the cost of NoIR lens blanks is the same as the cost of NoIR wraparounds in that tint and not much less than the cost of having good quality custom tinted lenses made in an optical lab. The polycarbonate used in NoIR products does not have the fine optical quality of the plastics used in optical labs. And quality control varies greatly in the dyeing of NoIR lenses. One cannot have an Rx added to NoIR lenses, as when having custom tinted plastic lenses or Corning ground glass lenses made to fit spectacle frames.

Keep in mind that, whenever extra dark lenses, such as dark NoIR tints, are mounted in spectacle frames for achromats, there needs to be some plan for dealing with the inevitable problem of side glare (explained on p. 172). The darker the lenses, the more serious this problem can be.

NoIR lenses scratch easily, the wraparounds break easily, and their edges are often sharp. In fact, all brands of wraparounds constructed of polycarbonate pieces fitted together break easily at the hinges and so must be handled with care. Some brands are sturdier than others, some block light better than others, and some are more comfortable than others.

Other Options

Tinted contact lenses: There is much information regarding tinted contacts on pp. 102-104. Also, a substantial amount of networker input about tinted contacts is included in the "Comments from Networkers about Tinted Lenses" section on pp. 80-93. The list on p. 114 includes three contact lens labs that many networkers have used.

Sport shields: There are many wrap-around styles on the market designed especially for outdoor sports, offering

protection not only from UV rays but also from dust, wind, and water. These have tints that provide enhanced vision in specific outdoor environments – usually dark amber, dark brown, or reddish brown (tints appreciated by many achromats). Some styles of sport shields accommodate Rx lenses, and some come with a built-in or attached visor.

Clip-ons and slip-behind lenses:

Clip-on lenses (some of which can be flipped up when not in use) can be used with spectacle frames, to provide a tint for someone who wears clear Rx lenses or to provide additional tinting for someone wearing tinted lenses.

Clear clip-on lenses can be ordered for custom tinting.

A simple and inexpensive way to add tinting to lenses in spectacle frames is by using a flexible plastic lens, sometimes referred to as “instant sunglasses,” that fits behind one’s frames and curves around at the sides, providing tinting for one’s peripheral vision. The upper edge of the flexible lens fits over the upper edge of the frames. Two of these can be nested together to create a very dark tint when needed, but clarity is reduced. These are not available in tints dark enough for achromats outdoors; but, combined with spectacle-mounted lenses that are already tinted, they can be very useful. Since these lenses are made of plastic, it is possible for an optician or anyone else skilled in dyeing lenses to dye the gray ones dark brown, deep red, dark gray, or whatever darker tint is desired.

Combinations: Tinted spectacle lenses can be worn in combination with “fit-over” wraparounds, clip-on tinted lenses, or the flexible plastic tinted lens described above that slips behind

spectacle frames. Some “sport shields” are worn over spectacle frames, making it possible to experience the combined effect of the tinted shield plus the tinted lenses underneath. Achromats who wear tinted contacts typically wear sunglasses in addition to their contacts when outdoors in bright sunlight. For blue cone monochromats and incomplete rod monochromats with some color vision, it is important not to use different lens colors when wearing two kinds of tinted lenses.

A mirror coating on the outside of lenses makes them darker. Some readymade sunglasses already have this kind of coating. Some networkers have paid to have a mirror coating applied to their lenses. This is an expensive procedure which, after a period of time, must be repeated.

Polarized lenses offer an additional kind of glare protection. Some networkers benefit greatly from wearing polarized lenses and others do not.

Some achromats find welder’s glasses to be a sturdy and affordable option for use in outdoors activities.

There are visors that can be attached to spectacle frames and to the frames of some sunglasses, then removed when not needed.

One can order side shields for attaching to spectacle frames. Those available from optical supply sources are often stiff or uncomfortable. Some opticians fabricate side shields from soft leather or some other flexible material when making glasses for patients with achromatopsia to use outdoors.

For more information about these options, consult your eye care specialist, and check “Sources for Tinted Lenses and Related Items” on p. 124



Networkers' Input Regarding Tinted Lenses

Pages 80-93 are filled with comments sent in by numerous members of the Achromatopsia Network sharing their experiences, preferences, and thoughts with regard to the various options in tinted lenses. Not surprisingly, this subject has received more attention in correspondence from networkers than any other subject since this network began. Achromats have written about what has or has not worked for them, network parents have sought advice in making choices in tinted lenses for their children, and inquiries have often come in from vision care professionals wanting to know what the "best" lenses for achromats are.

Throughout my years as facilitator of the network, I have always advised persons making inquiries to consider carefully the input of many individuals with achromatopsia, and I have made such input readily available in the materials I have published. As has been pointed out in other parts of this book, there simply is no single solution that is right for everyone who has this vision disorder.

It is important for achromats to know that they do have choices. And the truth of this statement goes far beyond the fact that there are so many choices these days in terms of sunglasses, tinted contacts, etc. With all the advancements in optical dyes, technology, and materials, this is a marvelous age for people who have special visual needs. But even more fundamental than the fact that there are many choices in tinted lenses is the fact that achromats have choices in terms of how much time they actually want to spend in settings which make it necessary for them to wear tinted lenses.

There are, of course, situations in which achromats do not have many

choices in this matter. For example, having to comply with compulsory education laws while one is growing up means having a schedule that includes spending a considerable amount of time in classrooms that are illuminated to meet the needs of the normally sighted. It also means regularly having to spend time in sunny school yards for recess and P.E.

Exceptions to this rule include (1) the kinds of arrangements that some network parents have made, so that lighting in the school rooms is adapted and various accommodations are made with regard to playground periods, and (2) the adapted environments and adapted lifestyles that have been developed by parents who have chosen to home school their children. But these are the exceptions. For most children and teens with achromatopsia, the need to cope with lots of bright light from day to day is an integral part of their school years. Typically, these young networkers rely on tinted lenses in brightly lit rooms and on the playground at school and are relieved when they can retreat to a visually comfortable environment at home after school and on weekends and holidays.

Choices in terms of how much time is spent in bright environments are also very limited for persons who live in extremely sunny parts of the world, such as in desert country or on tropical islands. (You may wish to read about the harsh conditions faced by the persons with achromatopsia who live on Pingelap and Pohnpei Islands, as described on pp. 144-149.)

Also limited in terms of choices are persons with achromatopsia whose jobs require them to spend a great deal of their time in brightly illuminated environments and to navigate very

bright surroundings on their way to and from work.

However severe the circumstances, achromats everywhere search for ways to reduce their exposure to bright light. If they don't have dark lenses to alleviate their problems with light sensitivity (as is the case with achromats in many parts of the world), they find other ways. The achromat in the desert or on an island seeks some kind of shelter from the sun by day and makes the most of twilight time or night time. A member of the network in sunny California whose workplace and routes to and from work made it necessary for him to wear very dark lenses much of the time created a dimly lit haven for himself in his home, complete with wood paneling, soft lights, and beautifully shaded windows. The book *Living with Achromatopsia* is filled with other examples of achromats' methods of coping with bright light which do not involve wearing tinted lenses.

Most of us find that, once we get past the years of compulsory education, we have many choices – and especially when we get turned on to the idea of thinking in terms of choices. Just as we can question whether we really have to tolerate the light level wherever we find ourselves having to spend time, either temporarily or routinely (many networkers have become very assertive about asking that lights be lowered or turned off or curtains drawn, etc.), we can also free our minds to start dreaming about where we would like to live and how we would really like to spend our time, now and in the future.

How comfortable and sighted – or, conversely, how uncomfortable and visually impaired – do we feel in the places where we routinely spend time? How often are we truly able to feel relaxed and at ease? These questions are well worth pondering.

Achromats on the move tend to be continually evaluating elements in their surroundings – anticipating, preparing, measuring the lighting, selecting an appropriate pair of glasses or some combination of lenses for the place we're about to step into or removing tinted lenses as the lighting dims, etc. We are mindful of so many things that are not of concern to other people. Is it going to be sunny or overcast today? When I enter that room at the end of the hall, what kind of lighting will there be? Will I be expected to sit facing the windows? Should I switch glasses now or wait and see if I need to?

Because we do have access to such an impressive variety of tinted lenses these days, some people feel that this frees achromats – or even obligates them – to pursue the same kinds of schedules and lifestyles that are possible for normally sighted persons. Sometimes it is assumed that there is no reason why achromats should limit the amount of time they spend in bright surroundings or choose lifestyles that emphasize the visual comfort and reasonably good eyesight which they experience in subdued lighting, as long as dark lenses are available for them to wear in bright light.

But tinted lenses do not give us the cone vision that our retinas lack. They simply allow us to make use of our rod vision (our night vision) in order to open our eyes and see when we are in brightly illuminated settings.

Even with the best tinted lenses, our visual functioning in bright surroundings is much more limited than it is in low level lighting without tinted lenses. Eyestrain, fatigue, and other stress symptoms can result if we continually try to keep pace with normally sighted persons by spending as much time as they do in bright outdoor settings and trying to keep up with the same kinds

of activities. Our eyes need rest and relaxation.

Reports from networkers have consistently shown that, among achromats, there are “day people” and “night people,” “indoor types” and “outdoor types.” Many of us are not so easy to classify and are somewhere in the middle. Achromats who are strongly drawn to participation in sports and other outdoor activities tend to make use of whatever tinted lenses they can find to make this kind of lifestyle possible. By contrast, achromats who are “indoor types” tend to be less interested in experimenting with various kinds of tinted lenses, because their needs for such lenses are few. Sometimes the “night people” are successful at finding jobs which allow them to sleep during the daytime and to be active primarily in the evening and night time. They make little use of tinted lenses.

The world is full of “night people” and “indoor types” who have nothing whatever wrong with their vision, who are what they are simply because it is their nature to be that way. If an achromat happens to have this nature, it can be considered fortunate, because he or she can more fully enjoy the normal rod vision which is the birthright of achromats. Achromats have every right to choose to be a “night person” or an “indoor person,” if they are so inclined, and no one should try to convince them that they should do otherwise.

On the other hand, if an achromat is by nature an “outdoor type,” then he or she is fortunate to live in these times, because there are so many tinted lenses, visors, and other devices that can help them to maximize their “outdoor vision” in the daytime.

One networker wrote in to say that, while on vacation, she had experienced some especially good times with her

family and friends in some charming and dimly lit night spots – atmospheric places where the light was so low that she could forget all about being vision impaired. She enthusiastically resolved to have more such experiences in the future. She called it “reveling in the rod light” – referring to the gentle lighting that poses no challenge whatever to the rod photoreceptors of persons with achromatopsia.

I have fond memories of taking my son to Disneyland when he was ten years old and the next year taking him to the County Fair. But those happy memories are combined with strong memories of all the visual stress and eyestrain I experienced during daytime hours in those locations (in spite of wearing dark wraparound sunglasses and a broad-brimmed hat). By contrast, I have only happy memories of events that took place in darkened environments, such as the Dickens Fair, a “living history” attraction that was set up in an old theater, where subdued lighting helped to transform numerous indoor spaces into settings straight out of Dickens’ novels. I cherish all of my outdoor recreational experiences, but I recognize that my favorite experiences have always been those in which I was “reveling in the rod light.”

Each of us must find the balance which is right for us – i.e., how much time we want to spend in light levels where tinted lenses are not necessary and how much time we want or need to spend in bright surroundings. As light levels increase or fluctuate, we make choices about when we are content to squint and blink, when to make use of tinted lenses, when to accept a certain degree of reduced vision, and when we need or want to be as sighted as possible. Only achromats understand about this extraordinary way of seeing.

There are professionals who are knowledgeable about specific aspects of achromatopsia, but there are no “authorities” on the subject. One of the best eye doctors I have ever known once told me, “You are the expert on your eye condition.” Of course, what he meant was that I, as someone who has lived with this rare vision disorder, knew more about it than any doctor I could consult. The subject of how to cope with hypersensitivity to light is one about which all achromats are uniquely knowledgeable. The most well known coping method is to make use of tinted lenses. But there are other coping methods too, and all of them are worthy of consideration.

The following statement appears in various parts of this book: *Information about products and services in this publication is for your consideration only and does not imply a recommendation.* Whenever published material contains input from different sources, it is important that a disclaimer statement such as this be included. This disclaimer statement is especially important when publishing comments from different people about the various options in tinted lenses for achromats.

Many of the persons quoted on the following 14 pages are simply reporting what kinds of lenses they like or don’t like and what options have or have not worked for them. Others are recommending the products or solutions they prefer. All of this input has been gladly received and is gladly shared – and it is, as the disclaimer points out, for your consideration only. Specific networker comments should not be interpreted as representing the recommendations of our network in general or of this editor in particular.

Since this network began, one of the primary purposes of publications for

the network has been to offer a forum, so that a range of different viewpoints and options can be presented, making possible a dialogue about matters that concern individuals and families affected by achromatopsia.

Such a dialogue can be a mixed blessing, however, at least for some people. Some readers may find it confusing or unsettling to learn that there are so many different approaches to dealing with the problem of hypersensitivity to light. It is very human to wish that, instead, someone would just tell us the “right” thing to do. But there are so many factors that influence individual networkers’ experiences and their choices. (These factors are presented on pp. 58-61.)

Some of the comments on the following 14 pages are preceded by a phrase introducing the person being quoted as a blue cone monochromat or an incomplete rod monochromat. These are important distinctions to make, since there is so often, with these individuals, substantially less light sensitivity, the presence of some color vision, and better visual acuity (compared with complete rod monochromats). These factors can significantly affect which kinds of tinted lenses can work best for these persons.

The networkers quoted on the following pages have valuable information to share, and they deserve to be heard. Altogether these comments represent a unique collection of experiences and insights involving an extremely rare and extraordinary way of seeing, about which very little was known until recent years.

On page 114 you will find a list of sources of tinted lenses and related products. Included on the list are sources for many of the items mentioned by networkers on the following 14 pages.

Comments from Networkers about Tinted Lenses

From a man in his 40's: "From my late teens until about 10 years ago, some of my tinted lenses for outdoors were dark brown and some were dark gray. Then I obtained the darkest Corning lenses (red-orange). I found them comfortable both for outdoors and bright indoor spaces, although they made many things look amazingly different – especially things I knew to be red or orange. Such things would look very light to me with these lenses on. The problem was that other people's attention was drawn to the color of these lenses, and at that time this was more important to me than the benefit I derived from them. So, when my girlfriend began to seem ashamed of my appearance when I had these glasses on in public, I stopped using them. Now I look back and see that as a mistake. The spectacles I currently use outdoors are made by Revo. The lenses are very dark gray, and they have a blue mirrored coating. The plano (i.e., no Rx) tinted lenses I tried on in the shop were fine, but the lenses which were made to fit my frames turned out to be much thicker, with a different thickness in each lens because of my Rx. Moreover, perhaps because of the added thickness, they are darker than the lenses I had tried on in the shop, and the side reflections are disturbing. I have to take these sunglasses off as soon as I move from outdoors to indoors, in order to move around safely. Indoors, whenever possible, I prefer to adapt the lighting or to move to a space with more favorable lighting, rather than wearing tinted lenses. Once I tried to be fitted with contacts. I loved the full visual field they allowed me – no spectacle frames in the way – but I had trouble putting them in and taking them out. I found taking them out nearly impossible. The optician's assistant did all she could to help, but we finally gave up."

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"I have complete achromatopsia. What works best for me is to have on hand a variety of sunglasses for different lighting. However, I minimize my use of tinted lenses in general, so that I can make the best use of my good rod vision. I dearly love natural light, and I love not wearing glasses whenever possible. I love being in sunshine. When circumstances require that I see my best outdoors, I resort to my darkest sunglasses and sometimes a hat or visor. But, when there are no such visual demands, I use lenses that aren't so dark. I often go to a comfortable, shady spot in my back yard or to some safe and pleasant natural setting, such as a park, and just relax without my sunglasses on, enjoying the sunshine and accepting my vision as it is. When I am with other people outdoors, I keep sunglasses on, because other people need to be comfortable about my eyes."

.....
"I would love to know how some networkers have managed with tinted contacts. Besides getting the 'willies' about putting something in my eyes, I also get panicky whenever things get darker than my visual comfort level – for example, having my sunglasses on in a dimly lit room. I would probably find it inconvenient having to remove contacts frequently in order to be visually comfortable."

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From an incomplete achromat who drives: "My main experiences with tinted lenses have been with Corning lenses and NoIR wraparound sunglasses. When I am driving, I like using the 2% amber (very dark amber) NoIRs, because they cut down on the light enough for me to drive in most conditions without cutting out such color as I am able to see. I have found that the red NoIRs don't meet my needs, because they have an effect on the way I see traffic lights that is exactly

the opposite of the way things look through the dark amber lenses. With amber lenses I can see the green light but not necessarily the red light, and this is the way I am accustomed to seeing traffic lights without tinted lenses. Therefore, amber lenses don't take much getting used to. By contrast, red lenses darken the green light so much that I can't tell when the green light is on and make the red light so much lighter that I fear I might accidentally interpret it as a green light. Not a comforting thought. So I personally find the red lenses too confusing for use in driving, but they might come in handy at other times. For instance, I find red lenses nice at the beach – they make the water look so dark.”

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From a man with achromatopsia in his 50's who is a vision scientist: “I like to use a tinted plastic polarizing filter that slips behind my prescription glasses. This device wraps around at the sides, protecting my eyes from intense light from the periphery and giving me a full field of view. Before finding this type of filter, I used polarizing clip-on sun filters. I know that side shields on a spectacle frame can help in shutting out light, but they also prevent motion detection in the peripheral visual field, which is important for moving about confidently and safely. I do not use them. I want to see light, so I wear very dark lenses only when absolutely necessary. I enjoy being out in the warm sun, provided that I don't have to perform demanding visual tasks. I want to experience intense light whenever I can do so without jeopardizing my safety or the work tasks I am performing. I understand that this subject is a source of some difference of opinion among vision care professionals. I think that continually wearing lenses that are made dark enough to maintain a low level of illumination for achromats is not something that should be recommended, as it may deprive the person wearing them of important visual experiences. I often forsake my sunglasses now and then, to be able to experience the full light intensity in nature and not to have to be constantly ‘walking in the shadows.’”

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From a woman in her 30's: “I use the red/orange Corning lenses. I like to use the lightest tint in that series for indoors and the darkest tint for outdoors. I have my outdoor lenses set in the frames of ‘ski goggles.’ These frames have leather side shields. An advantage in using these lenses is that, as a pedestrian, I am able to see red lights and turn signals. Also, they help me distinguish between certain colors. For instance, with red lenses I never confuse red with black anymore, because red looks very light. There was a time when I wore contact lenses. The first pair I had were tinted the darkest shade of brown that was offered in the early 1970's. Those were hard contacts. I became allergic to either the plastic or the dye and had to switch to soft contact lenses. About that time I tried some red contact lenses, but that was a disaster. They were too dark for indoors, and the tinted part had not been made large enough to completely cover the corneas of my eyes, so they were not effective outdoors.”

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From a woman in her 40's: “I use brown polycarbonate wraparound sunglasses, and I love them. I do not wear Rx lenses, because I found that the difference they made in my vision was minimal. So I wear plano (i.e., without Rx) medium gray lenses indoors and dark wraparounds outdoors. I have tried various colors of NoIR sunglasses. I like the red ones OK, but I get strange looks from people when I wear them. I tried the darkest gray ones, but they made things very indistinct. I like the 4% plum NoIRs, and I want to try the 2% amber ones. Unfortunately, I

find that the NoIR products are poorly made. The edges can be so sharp. Sometimes I use a file to smooth out the edges.”

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From a woman in her 20's: “I always put in my tinted contacts first thing in the morning and take them out the last thing at night. I have become totally dependent on them. You could say that I have become addicted to them. When watching TV, I don't have to squint so much anymore. And I get great pleasure now from looking at the sky on cloudy days, because my red contacts give everything a rosy glow. Because I continually wear the contacts, I have found that now, whenever I am in a dimly lit room, I will do whatever I can to get as much light as possible in the room. I have changed some of the light bulbs to brighter ones, and in the evenings I turn on every light or lamp in the house.”

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From a woman in her 20's: “My life has been more comfortable since I began wearing my dark brown tinted contact lenses. Tinted contacts do not let light in at the sides the way spectacle lenses do. In combination with sunglasses, my tinted contacts allow me to enjoy sunny days. They allow me to wear untinted reading glasses without having to adapt the lighting in my workplace. They make it possible for my clients to see my eyes clearly – which is important, as I do social work. I have learned to ‘click’ my contact lenses in and out whenever and wherever I want to. Sometimes this must be done repeatedly in a single day. I use different combinations of lenses for different lighting conditions. There are times when the lighting is so dim that I wear no tinted lenses at all. Then there are twilight times and rainy day times when I wear lightly tinted contacts. And there are times when I wear my darkest contacts – for instance, when it is sunny but I am not in direct sunlight. Then there are the very sunny outdoor situations in which I combine my darkest contact lenses and my sunglasses. For me, there is one important fear in connection with wearing tinted contacts, and that is the fear that there might be a time when I can no longer obtain these special contact lenses.”

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From a networker in the U.K.: “I wore contacts for 20 years, but then I started having problems. At the eye hospital they said I had ‘insufficient tears’ (dry eyes) and that I should stop wearing contacts. I was quite happy to stop wearing them at the time, but now I would like to have that option again. The frequent changing from light tinted sunglasses to dark sunglasses and vice versa while going in and out of places is tiresome. I don't find bright sunshine so much of a problem since I found some very dark sunglasses. With these I have been able to participate in outdoor ballgames for the first time, which has been very exciting for me. We have lots of overcast weather here, which creates diffuse light, and this can be very bright. In winter it can be embarrassing to be seen wearing sunglasses – although, since sunglasses have become so trendy (and as I get older), I care hardly at all about this anymore. Tinted contacts were great because of less wear and tear on my face, ears, etc., and because of having just one pair of sunglasses to take on and off, as needed – and also being able to look ‘cool.’”

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From a woman in her 20's: “Finally, after 6 years of wearing tinted contacts, I have found a pair that works. They are so dark that I can now walk around outside on a dark or rainy day without needing to wear my sunglasses. Shopping is now easier, as the bright fluorescent lights don't bother me anymore. They also make a

huge difference in the bright classroom where I teach. After I got these new, darker contact lenses, I took down everything I had been using to shade the windows in my classroom, and it is a new experience to be able to look out the window at everything. Some people have commented on how ‘black’ my eyes look, but I feel that the benefits I get from my contacts outnumber any concerns about how my eyes look to others. And, because my contacts are red, I can now distinguish between red and black so easily. This is convenient when looking for a certain color pen or when playing cards, for instance.”

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From a woman in her mid-20’s: “I have a feeling that my tinted contact lens-wearing days are coming to an end. My eyes have been ‘drying out’ recently, and at one point I developed an eye infection, so I have been having to wear my tinted glasses instead of my contacts. I think there is probably an ideal time frame for wearing contacts, considering that most people I know who have used them have had to give them up after 10 years or so (or sooner) because of the problem of dry eyes. I now feel that people probably shouldn’t start wearing them until their mid-teens or late teens – i.e., the period in one’s life when one can get the most benefit from wearing tinted contacts in connection with starting a career, job interviews, starting relationships, going to college, etc.”

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From a networker who does not wear tinted contacts: “I gladly leave off my sunglasses most of the time when I am indoors (though sometimes I use them when watching TV) and I find this quite comfortable. I talked with another network member, who told me she puts in her tinted contacts first thing in the morning and wears them until bedtime. I think that, if I were to be looking through tinted lenses all my waking hours, I would feel as if I had an eye disease like retinitis pigmentosa. I do so love natural light.”

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From a woman in her 40’s: “Some years ago I acquired a two-tone pair of sunglasses. The lenses are darker at the top and lighter at the bottom. The top part is a reddish brown that works well for me. I bring these glasses along whenever I go to my optician to have new glasses made. He does what he can to duplicate this tint. I have also found that having dark frames helps.”

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From a man in his 30’s with blue cone monochromacy: “I wear several types of glasses: (1) Tinted Rx lenses for use in bright light indoors (2) Plain Rx lenses for low level lighting – I can also wear these outdoors in sunlight, but things look ‘washed out’ and monochromatic (3) Magenta sunglasses for most outdoor situations in the daytime – these help me see such color as I am able to see, but they are often not dark enough. I have tried brown wraparounds and found that they interfered with my potential (as a blue cone monochromat) to see certain colors, but brown lenses do help me to see details and contrast. I have magenta contact lenses, but I don’t wear them much. In general, I find contacts a hassle.”

.....
From a woman in her 50’s with complete achromatopsia: “I am so glad that, when I was a kid, I stubbornly refused to wear the Rx glasses that many people thought I should wear (for correcting my astigmatism and ‘farsightedness’). Over the years there have been so many great wraparound sunglasses available, and these have given me the very best kind of help for my light sensitivity problem, which is so much more serious than my astigmatism and ‘farsightedness.’ If I’d

had to wear spectacle frames (as most people who wear Rx lenses do), I wouldn't have been able to use the wraparound sunglasses all these years."

.....
From a man in his 20's: "Outdoors I use dark red Corning lenses mounted in special Bollé frames that have side lenses as well as front lenses. The plastic side lenses in these frames were custom tinted by the optician to match the tint of the Corning lenses. What bothers me about the red lenses is the way they distort the appearance of things. Also I find that red lenses seem to excite my nervous system, so I take them off whenever I enter my home or office. I try not to wear them for more than about 1 hour on most working days."

.....
"We obtained tinted contacts for our daughter when she was 7. She wears them when she's participating in choirs or theater groups, because the stage lights can be so terribly bright and because she feels that it is better for people to see her eyes when she is performing."

.....
"Our son, age 11, now has tinted contact lenses. He has become more confident and happier about his appearance. He has regular check-ups to make sure that they are not damaging his eyes in any way. For a while I was concerned that his light sensitivity had become worse since he had begun wearing contacts. Light did seem to be bothering him more. I try to get him to wear his glasses instead of the contacts on weekends, so that he gets more oxygen into his eyes. I also try to get him to take the contacts out when he gets home from school. He often forgets to do this and does not take them off until bedtime."

.....
From the mother of two young children with achromatopsia: "We found some wonderful inexpensive sunglasses for both girls at Sunglass Hut (a chain store). They wrap around completely, and we are going to have custom tinted lenses put into these frames. Our older daughter seems more comfortable with a dark plum tint than with a red tint. With the red lenses, she was continually asking why things change colors, and she would lift the glasses onto her forehead repeatedly to compare the way things looked with and without them."

.....
"My son (age 12) wears regular sunglasses that we buy off the shelf at the store. We shop on a sunny day and let him try on lots of different sunglasses, and he picks several pairs to buy. When he was 2, he was fitted with Bollé sunglasses with opaque side shields. He wore them for 10 years and then discovered that he could wear the kinds of sunglasses other people wear (less attention-getting) and that, instead of the side shields, he could enjoy wearing wraparound sunglasses that allow him full vision at the sides. He would have nothing more to do with his old glasses. He likes being able to wear different styles."

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"Our son (age 9) has been wearing red tinted contact lenses for 5 years. He has become very dependent on his contacts – so much so that, when he got 'pink eye' last year and was unable to wear his contacts, he was very much blind and just stayed at home from school until he could wear the contacts again."

.....
"My daughter, age 13, has worn dark red Rx lenses set in frames with leather side shields for most of her life. She depended on them for outdoors in the daytime and for indoors where fluorescent lights were used. She doesn't usually wear tinted glasses indoors; and, of course, she doesn't wear them outside after

sunset. For reading she has lightly tinted Rx glasses, but she often forgets to wear them or doesn't want to bother with putting them on. I don't know how much the Rx actually helps her. Recently she opted for some spiffy, sporty looking sunglasses, which she seems happy with. The lenses that have been put in these frames are reddish brown. They can just about pass as 'normal looking.' They wrap around her head nicely, so she no longer has to wear side shields (which were bulky and attention-getting and which caused her lenses to fog up), and she doesn't mind the fact that there is no Rx in them. Her frames cost \$40, and we had lenses made for them at LensCrafters for \$50. The people at LensCrafters were very helpful about tinting the lenses repeatedly until the tint was just right. The light weight of these sunglasses is a welcome change for her too, and they pass the teenage 'coolness' test. The only bad thing (for me) is that the tint of these new glasses is so dark that I can't see her eyes, whereas I could see her eyes through the red lenses she wore for so many years."

.....
"Image is becoming so important to our daughter. Recently she said she wants to try tinted contacts and to have 'normal sunglasses' to wear in addition to the contacts. But she is aware that some things will be harder with tinted contacts. For example, she frequently has to take off her tinted glasses in order to see better in certain places, and this kind of 'back and forth' would be more complicated with contacts. Also, I find it reassuring to know that her sunglasses are protecting her eyes outdoors to some extent, since she is more likely to bump into things than normally sighted people are. And I worry that she might forget to take the contacts out at night and that complications would result."

.....
"Our 5-year old daughter wears glasses with a blue/gray tint when she is in her kindergarten classroom and in certain other indoor environments. When she goes outside, she pulls her dark wraparound NoIR sunglasses, which are plum colored (4% light transmission), down over these glasses. These NoIR 'fit-overs' are large, because they were designed for adults, but they have been adapted in several ways to be usable for a child. Portions of the tinted ear pieces have been removed, and a sport strap has been attached to these ear pieces for keeping the sunglasses on her head. Also I obtained from my optician a 'T strap' that connects from the sports strap at the back of her head to the center of the top of her head and across the top of her head connecting the two earpieces of the sunglasses. We hope to come up with something better, but this system works great for now. When the lighting is good for her, she simply lifts the NoIR wraparounds up, and they stay on her head – and her hats (she loves to wear hats) actually hide them. Then, when she needs the dark wraparounds, she reaches up and pulls them down over her glasses. We also have tried out 2 brands of children's glacier goggles. One was too large, and the leather side shields did not block out enough light. The other fit better. We had these fitted with dark plum colored Rx lenses. She uses these when she is outside for long periods of time. They are not convenient at times when she is going back and forth indoors and outdoors."

.....
"My husband and I feel very strongly about wanting our daughters to be able to enjoy natural light as much as possible and to experience and use their normal rod vision fully. We have read about the dependency on red lenses that some kids with achromatopsia have developed. When we read about the mother who goes

into her son's room to remove his dark glasses (made with the Corning lenses) when he is asleep, this affected our decisions. Our children's ophthalmologist and the optician were pressuring us to get red lenses, saying they are the 'best.' So we were thinking of just going ahead with red lenses and urging our daughters to remove them whenever they didn't really need them. But the ophthalmologist wanted them to wear the red lenses all the time. He looked at me like I was nuts when I told him some of my concerns about this. But that's OK, because we truly believe we are doing the right thing. We are carefully looking for what will be 'best' for them in the long run. For now, we have ordered a dark amber tint for their sunglasses, and we are open to the possibility of trying red lenses in the future."

.....
"My 16-year old son wears trendy sunglass frames which are fitted with red lenses coated on the outside with a gold mirror coating. This coating makes the lenses even darker and hides the red color of the lenses. (It is important not to look too 'different' in high school.) He has had many compliments on these sunglasses. Unfortunately, the mirror coating is starting to wear off, and we need to have them re-coated (this is expensive). He does not wear his glasses indoors, as his brother does. His brother, age 12, wears a sport frame outdoors. It has wrap-around lenses, also mirror coated, and the frame's design allows for prescription lenses to be snapped in behind the wraparound tinted lenses."

.....
"My children used to wear Corning lenses, but for the last couple of years they have worn brown Rx contact lenses, and they really prefer these contacts. No more teasing from schoolmates about wearing dark glasses indoors and no light coming in from the sides, as it did with the spectacle frames they used to wear. Outdoors they wear ordinary sunglasses in addition to their contacts."

.....
"We obtained tinted contacts for our daughter (now in 5th grade) 2 years ago. She has had contacts in her eyes from sunup to sundown every day since then. We were concerned at first that managing the mechanics of the contacts would be difficult at her age, but the opposite has been true. She has learned to move her contacts off her corneas and under her eyelids whenever she doesn't need them. She can do this without touching her eyes! I have worn contacts for years, and I cannot figure out how she can do this. The contacts give her a lot of flexibility in adapting to various levels of light. She has settled on a pair of rose tinted wrap-around sunglasses to wear in addition to contacts when she is outside."

.....
"Our teenage son has a collection of different shades of dark tinted contacts. None of them are sufficiently dark to enable him to be completely comfortable outside in the daytime. He refuses to be seen wearing sunglasses in addition to these contacts, but he has developed strategies for functioning in bright light. For one, he wears his hair long and lets it fall over his eyes. For another he takes 'mental pictures' (after-images) during long blinks. He is able to wear the contacts for 12 hours or more without discomfort. Another strategy: he only goes outside in the daytime when he really has to. At school he wears one light and one dark contact lens, and he concentrates on looking through the most suitable lens in different light conditions. He also slips the lenses up off his corneas at times when he needs to see better in dim light. His doctor says this is OK but hastened to add that it is not a recommended practice. Our son takes his contact lenses out

as soon as he gets home and wears his glasses with Corning lenses for playing computer games and watching TV.”

.....
“Having glasses with medium tinted lenses for bright indoor settings has worked well for our daughters in school. I find, however, that the younger one tends to wear them too much of the time – for instance, at dusk or at night time, when she doesn’t really need them. I’m concerned that she has become overly dark-adapted. The lenses in her glasses have been tinted a dark amber for several years. Recently, when we ordered new lenses, the company accidentally tinted them more of a brown instead of amber. We were going to send them back to have the tint changed, but our daughter found that she preferred them that way.”

.....
“When our son (now 11) was very young, we obtained sunglasses with leather side shields for him. When he was 3 years old, the vision specialist fitted him with brown Lunelle Solaire contact lenses. With these lenses he has ‘beautiful brown eyes’ – when, in fact, his eyes are blue. In summer he wears darker contacts. He learned to handle his lenses himself a few years ago. I think it is important that he not look so different from the other kids at school. He had difficulty using the contacts at first, but he has no problems with them now. The minuses include the fact that he must have 2 new pairs each year, and that is expensive. However, here in the Netherlands we don’t have to pay for them ourselves. Another problem is that it takes him some time to adjust when going into a darkened room. The pluses are that he doesn’t have to wear spectacles, and this is especially nice when one lives in a rainy part of the world, as we do. People don’t understand when they see someone wearing dark glasses on a rainy day.”

.....
“We have obtained tinted Barracuda swimming goggles for both our children. Getting this done was not an easy project, but it has been most rewarding. With the goggles they are able to be in outdoor pools for hours at a time. Getting the right tint was difficult. Sometimes the padding on the inside of the goggles starts to wear out, but we have obtained extra pads and replace them periodically. The area around the lenses is not completely opaque and lets some light in, so we use very fine sandpaper to ‘rough up’ the frame. Then we paint it with very dark nail polish.” *(Editor’s note: Barracuda swim goggles are now available which have completely opaque “frames” around the lenses. See Skyline Northwest on p. 114.)*

.....
“Our son’s teacher says she has noticed a significant difference in his activity level and visual functioning when he is wearing his dark amber (‘blue blocker’) wraparound sunglasses, as opposed to when he is wearing his red lenses set in spectacle frames. Our optometrist also made some very dark glasses for him and dyed safety side shields to go with them – just in time for soccer season.”

.....
A few years ago I came across the large ‘solar shield’ sunglasses made to be worn over regular glasses. I discovered that, when worn over tinted glasses, these make everything twice as dark. Sometimes glare casts reflections inside the solar shield sunglasses, making it hard to see through them. Also, wearing two pairs of glasses can be hot, cumbersome, and unattractive. A few years ago I came across the darkest pair of off-the-shelf sunglasses I have ever seen. Not only were they super-dark, they also curved completely around the sides, allowing no light in at all. I love them. They are not as dark as wearing 2 pairs of sunglasses, but I prefer

them because they are not as cumbersome and they look ‘normal.’ I alternate between wearing these dark wraparounds and using the ‘two pairs’ system, depending on a variety of factors. Once I ordered a pair of the dark red sunglasses made by NoIR, but I didn’t like them. They made everything appear to be ‘glowing,’ and my eyes stung and burned when I had them on. Perhaps red lenses weren’t right for me because I am able to see the color red in some situations.”

.....
“Our 11-year old daughter recently got tinted contacts. It has been a remarkable experience for her. She is now able to participate more in soccer and basketball, wearing tinted sports goggles in addition to the contacts.”

.....
From a woman in her 30’s: “I had to give up wearing hard contacts after 20 years, because I developed a condition called ‘dry eyes.’ One of the contacts was a dark red lens, and one was clear. The red lens helped me see better in bright light, and the clear lens gave me the option of using the other eye when in a darkened room. I tried gas permeable contacts, but they were not able to dye the gas perm material dark enough to be of much use to me. The red hard contact lens had helped me with glare reduction and also enabled me to distinguish between certain colors. Now I have obtained soft contact lenses which allow me to have the dark red tint that I found so helpful in the past. These do not have any prescription, as I was told that soft contacts could not be made to correct my severe astigmatism. So I just put on my prescription glasses whenever I need the little bit of help that the Rx gives me. My eye doctor had these soft contacts made for me at a lab called Specialty Tints, which I learned about from another networker whose doctor used this lab to obtain the dark brown contacts she prefers. My new contacts are tinted to look like an iris and a pupil on the outside, with different tints in different areas of the lenses. One lens has a dark red center (covering my pupil) and a blue area around it (to match the color of my irises). The other lens has a blue tint in the center and a blue area around it. The red lens is so dark that it makes my visual acuity a bit worse, but with it I can now walk down the driveway to the mailbox without having to put on sunglasses. I can answer the door without sunglasses on, and I can enjoy the view from our front window without squinting. And it cuts down on the intensity of the fluorescent lights where I shop.” *(Editor’s note: If any networkers want their eye doctors to order tinted soft contacts, this source and others are listed on page 114.)*

.....
“We decided to order for our daughter (age 2) some of the sunglasses that NoIR offers for infants and toddlers. We ordered a pair with the darkest amber lenses NoIR makes (2% amber) mounted in the nylon wraparound frames they make available. We have been shopping around for other wraparound frames and, so far, have not found any that come close to these little frames in terms of staying on a small child’s head, efficiency of wraparound design, and comfort. She wears these 2% amber sunglasses when she is outdoors on bright days, and she wears a pair with the 4% amber lenses on very cloudy or rainy days. We limit the amount of time she wears her sunglasses, so that her eyes can enjoy a reasonable amount of natural light. She almost always removes them as soon as we enter a place where the light isn’t bright.” *(Editor’s note: If any networkers want to check out this option, be sure to read all of the information about NoIR lenses presented on pp. 73-74 of this book.)*

“Three years ago my doctor had to special order Vuarnet PX-5000 sunglasses (which were very dark and thus worked very well for me), because they were no longer available in stores. I went to LensCrafters and was told that they could darken the lenses of many of the sunglasses in their shop to match the tint of the Vuarnet PX-5000 lenses. I selected a pair of sunglasses with lenses that were already rather dark. They asked me to leave my Vuarnets with them and to come back in an hour while they dyed the new pair to match my old pair. LensCrafters is located in the mall, so it was easy to hang out at the mall for an hour. After their first attempt, the lenses were not quite dark enough, so they had to repeat the process. Then, when I came back again, the lenses were actually a bit darker than my old pair, but that was fine with me. I should mention that LensCrafters carries some rather expensive sunglasses and that they charged \$50 for adding the extra tinting, but I felt it was worth it. It was so nice that I didn’t have to stick to that same Vuarnet style I had been wearing for 15 years. I could pick out something more trendy. It’s nice to have a ‘new look.’ The employees at LensCrafters were so helpful. They seemed eager to assist me with my special needs. The lady who waited on me told the optician, ‘Make those glasses dark, dark, dark,’ after I told her about my vision problem. In the past I have gone to sunglass shops where the sales persons would be mostly surfer dudes. When I would tell them about why I needed such dark sunglasses, their responses would usually be something like, ‘Wow! Bummer! Sorry, but they don’t make sunglasses that dark, I don’t think.’” *(Editor’s note: Readers interested in having the lenses of readymade sunglasses dyed darker, as this networker did, should review a section of this book that begins near the bottom of p. 67 and also review the suggestions provided on the pages that precede and follow that section. Not all lenses in readymade sunglasses can be dyed darker, and sometimes the fee charged to darken lenses is less than what this networker paid.)*

.....
From a complete achromat: “I swear by ‘blue-blocker’ sunglasses. These vary a lot in tint, so I look for ones that are the darkest.”

.....
“I have found that welding shops in our area can produce lenses as dark as I need them. Some of the people at these shops were very interested in my vision problem. These dark glasses are pretty stylish and are inexpensive.”

.....
“Three years ago we obtained some red contact lenses for our daughter (now 10). She doesn’t choose to wear these on a daily basis, because she finds that, on a sunny day or in fluorescent lighting, she still has to wear sunglasses in addition to the contacts – so she says, ‘Why bother?’ She seems to be at ease with simply wearing sunglasses, as needed. It has been a challenge for her to put her contact lenses in and remove them, but we found that a few tutoring sessions, one-on-one, with our optometrist, were of great help in solving this problem.”

.....
From a man in his 40’s: “My sunglasses have brown lenses with a double reflective green-colored mirror coating on the outside. The green reflective coating is more subtle than the more traditional silver or gold mirrored coatings. In my opinion, this makes my dark lenses less noticeable to others, since the green coating reflects lighter images. My frames are fashionable in shape and size, so that my wearing sunglasses in all outdoor daytime situations will appear at least

somewhat fashionable, rather than indicative of a disability. I am very self-conscious about having to wear dark glasses when it is rainy or cloudy. I plan to try wearing a smaller, somewhat elliptical-shaped frame that would fit close to my eyes and be more like the shape of my eyes. My thinking is that a smaller frame such as that would be less noticeable to onlookers and, by fitting close to my eyes, might keep out just as much – or possibly more – light from leaking in around the sides and top.”

.....
“For years our son, age 9, has worn the smallest size NoIR polycarbonate wrap-arounds. They don’t come in a child’s size, but we have not found children’s sunglasses that wrap around and cut out light as much as these do. On the negative side, he has gotten hit in the face while wearing them, and his face was scratched. He has also become self-conscious about their appearance. Our younger son wore the NoIR wraparounds for awhile. He is much more physical and tumbles about often. After he fell once and got a serious cut around his eyebrow, we replaced the NoIRs with wraparound sunglasses designed for preschoolers and fitted with custom tinted lenses.” *(Editor’s note: The smallest adult size NoIR wraparounds have been adapted for young children. One adaptation is the addition of rubber tubing around the brow edge of the glasses as a safety feature.)*

.....
From a high school student: “I like to go to the swimming pool, ocean, lake, etc., when it’s getting toward evening or when it’s very cloudy. I can see so much better then. I wear rose colored sunglasses, and often I wear over them a special ‘sportlens’ that wraps around and is combined with a black foam visor, which attaches with velcro at the back of my head. Wearing these items together cuts out the most light for me in situations where I need this much protection.”

.....
“At school, when our son (age 6) is asked about his ‘red eyes’ (because of his red contacts), he simply answers that he is wearing tinted contact lenses, and then he goes on to say that he has ‘special vision.’ Some people have said things like, ‘He looks like the devil with those red eyes’ or ‘He looks scary’ or ‘It looks like he’s ready for Halloween.’ I’m tired of explaining it. What keeps me going is knowing that my reactions to people are being taken in by my son.”

.....
“When I tried wearing tinted contact lenses, I found it very frustrating. If a tint was dark enough to make a significant difference for me near bright windows or under bright lights, it would make everything appear too dark whenever I would look toward – or move into – any space that wasn’t very well lighted. For instance, a hallway where I had been accustomed to seeing quite well would become a dark area of low visibility. I felt like I was having to cope with a new and different kind of vision impairment in addition to the one I already had. It didn’t matter so much to me that I was squinting less with the contacts on. I much preferred the flexibility I was accustomed to having by carrying a pair of dark sunglasses and a pair of glasses with a lighter tint, to use as needed.”

.....
From a woman in her 50’s: “Many years ago, when I first started having tinted lenses made for me, there were only shades of gray, green, or brown to choose from. Of those 3 choices, only the dark brown lenses helped me outdoors. For years I managed fairly well with sunglasses with brown lenses. Later on, at a low vision clinic, I had the chance to try out many different filters. I found that I still

saw best through dark brown filters, but I learned that I could have lenses made a much darker brown than the ones I had been using. I also was able to find better frames, which let less light in from the sides. I have also found that some of the readymade sunglasses with dark amber lenses have been quite usable ‘as is.’”

.....
From a woman in her 40’s: “The red tinted contacts that were tried with me distorted the world as I knew it. I need a tint that does not change things so much. The people at the low vision clinic first gave me contacts that were tinted so dark I couldn’t see well indoors. Then they made a somewhat lighter tint for me, and I still found it hard to see well indoors. The contacts made it possible for me to open my eyes near my bright kitchen window, but then I would turn around and find that the other side of the room was dark and hard to see. When they were tinted lighter, they were then too light to be of any use to me. During the period when I was going around outdoors wearing the darkest contacts they made for me, I became very depressed. I even wondered if I was losing my mind. This all changed when I went back to using my regular sunglasses.”

.....
“The frames my two young children use are wraparound style sunglasses for infants and toddlers made by I-care International. These frames are great. They wrap around, and they are made of a flexible rubber-like material, which makes it easy for an optometrist or optician to pop out the lenses and replace them with custom tinted (darker) lenses. We use a light elastic strap to help keep the glasses on the children’s heads. These sunglasses can be found among the other I-care sunglasses that are sold at KidsMart, Toys-R-Us, and many other stores.”

.....
“After many years of managing with various sunglasses that provided only limited relief from glare, I found an optometrist who earnestly wanted to come up with glasses that would give me the best possible vision outdoors in the daytime. He tinted lenses darker than any I had ever had and fitted my frames with side shields and a detachable top shield. The result was remarkable. I was able to see more outdoors in the daytime than I had ever seen before. At first this was very exciting; but, before long, I began to notice that lighting conditions under which I was usually comfortable without tinted lenses were no longer comfortable for me. For example, I could no longer just take off my sunglasses upon entering my apartment. I was keeping them on more of the time and having to wear my lighter tinted lenses in places where I had previously been able to enjoy not having to wear tinted lenses. I also noticed that I was feeling depressed. I think that, on a very deep level, I missed seeing the bright sunshine. I decided to resume wearing my old sunglasses and to set aside the new ones for such times as I would need them most. It wasn’t long before I had ‘weaned’ myself off the extra dark lenses and was able to tolerate light better – and my depression went away.”

.....
From a man in his 20’s: “I’ve never tried tinted contacts, nor do I plan to do so. In any given day I take my dark glasses off and then later put them on again at least 20 to 30 times. That would be very difficult with contacts.”

.....
From a woman in her 30’s: “When I am under fluorescent lights all day, I find I am less fatigued at the end of the day if I have been wearing my tinted contacts. But some days I have to take them out (depending on where I am), because they just make things too dark. They don’t do anything for me that my sunglasses

can't do, except give me more peripheral vision. But I absolutely cannot wear them all day every day. I love going for walks, jogs, etc., with my tinted contacts on. The world looks bigger. I really feel more 'mobile' with them on."

.....
From a man in his 20's: "Having squinted so much since birth, the muscles in my eyelids have developed unusual strength, enabling them to perform their protective function in a very efficient way. They have rejected even the softest contact lenses that I have tried on in a matter of seconds. Every time I have tried this painful exercise, the doctor and I have always given up. Even if they are applied in the dark, this helps only at the beginning. As soon as there is light, my eyes begin watering, and my eyelids expel the lenses."

.....
"Today I tried the red contacts which I got at the low vision clinic. It was a bright and sunny day, and I found it relaxing to wear them at home. That was nice for awhile; but, as it got later in the day, I had to take them out, because they made things too dark for me. I haven't had the nerve to wear them out of the house yet, because they make me feel 'trapped.' Whenever I find myself in a place where the lighting is low, it's like walking around in the middle of the night. It's ironic that I now find myself wanting to avoid dark places because of the contacts."

.....
"Besides her 2 pairs of dark glasses (one for indoors, one for outdoors) and her wraparound sportlens, our 4-year old also uses a pair of swimming goggles that have been fitted with tinted lenses. She likes to wear these when she is playing outside, because they fit so tightly to her face that no light gets in."

.....
From an incomplete rod monochromat: "I don't allow such color vision as I have to sway my opinion on which kinds of tinted lenses actually help me see better. Seeing color is nice, but it's a vague and funny thing for me. It's so intermittent and undependable. I don't know that color vision is all it's cracked up to be. But I would say that red, magenta, and yellow lenses have the lowest rating on my list. They give the world a hot 'glow-y' sort of feeling and actually seem to increase the glare I experience. (Maybe this happens because I am able to see some of the red spectrum.) I don't like it. It reminds me of hot summer days, which I don't happen to like. I would list blue, gray, and brown as my favorite lens colors for different lighting conditions. Blue lenses are great, because blue has a soothing effect on my eyes (except for bright blue, which makes me feel harassed). I don't squint as much with blue lenses, and shadows have much greater contrast for me. Gray lenses are especially good for indoor environments. Brown lenses are good for me, because they give good contrast, even when they are made very dark."

.....
From a woman with complete achromatopsia: "Whenever I have experimented with wearing sunglasses that were made really dark – so dark that I would never have to squint or blink in bright sunlight – I would often feel insecure and unable to see when I was in shadowy places, like under shade trees or awnings or in shaded walkways. As I would move in and out of different indoor and outdoor spaces, I would find myself frequently wanting to take off the glasses in order to see. This meant having to switch quickly to a pair of glasses that were not so dark or else taking off the glasses and managing without tinted lenses momentarily – or keeping the super-dark glasses on and just accepting the experience of being much less sighted than I should have to be in such places. I finally decided that I

would save my darkest glasses for times when I had to be in sunshine for extended periods and, for regular everyday use outdoors, I could wear sunglasses that weren't tinted so dark. With my not-so-dark glasses on, I know that, if optometrists were to test me, they would probably find that I do not have as good visual acuity in bright places as I have when wearing my darkest ones. But the increased adaptability that I experience with the not-so-dark glasses is wonderful. I can just breeze along from place to place, going in and out of buildings, moving from shady places to sunny places. I never have those sudden, frustrating feelings of being 'blinded by darkness' that used to happen so often back when I tried to go around wearing the extremely dark glasses."

.....
From a man in his 40's: "I use different pairs of tinted glasses for different lighting conditions. Like others in the network, I have found that I do not really benefit from having more light protection than I need at a given time. Over-use of extra tint seems to create a dependency on these lenses, and it decreases my tolerance for light. So I've found, through trial and error, how to use tinting most carefully. Having to strain a bit to see some of the time can be a good thing. But, on occasion, I indulge myself, because I find there are times when having extra tint can be comfortable and soothing. In brightest sunlight I have stuck with the Bollé frames which have side shields. The tint is mostly brown with just a little red."

.....
From a woman in her 50's: "I have some well made polycarbonate sun shields that I like. They have many positives, but the negatives are that they are too big for my small face, they get loose and can't be adjusted, and they are mirrored on the outside. I feel funny about how the mirrored coating calls attention to me. Sometimes people stop, look at their reflections in my glasses, and fix their hair."

.....
"My daughter has worn tinted contacts for a few years. In recent months she has developed corneal irritation or infection more than once. I am worried about the long term health of her eyes. I wonder if other networkers who wear contacts have experienced repeated problems such as this and can make suggestions."

.....
"Our daughter, now 16 years old, began wearing very dark tinted soft contact lenses when she was 6 years old, and they had a very positive impact on her life. She wears the lenses all day in complete comfort. Often the contacts have been made too light or else they have had different degrees of tint, so that they had to be returned to be re-dyed. For several years she also wore photogray nonprescription glasses (tint changes with light) in addition to her contacts in bright situations. In school she wore the photograys over her contacts all day. When she was younger, either her father or I would put in the contacts in the morning, and she would remove them herself at bedtime. After 5 or 6 years she stopped wearing the photograys over the tinted contacts and instead started wearing some very dark sunglasses from the drugstore when going outdoors."

.....
From a man in his 40's: "I wear clear lenses in favorable lighting, and I have 5 pairs of tinted spectacle-mounted lenses and sunglasses for use in 5 different lighting conditions – one for fluorescent light or for times before dusk, one for cloudy days, one for sunny days, another for especially bright sunny days, and another for use on highway trips on sunny days (since the glare from the highway plus the brightness of the sun is especially stressful to my eyes)."

What Are the Best Lenses for Achromats?

The documentary film, *Island of the Colorblind*, which has been shown to television audiences in the U.S., Canada, and other parts of the world, offers glimpses of some of the realities faced by achromats who live on two islands where there are no resources such as members of our network take for granted – no readily available sunglasses, no optical devices, etc. This is how life is for many achromats living in isolated or developing parts of the world. But, lest we forget, not many decades ago, this was also the reality of achromats living in the U.S. and other places where there is now easy access to a variety of helpful sunglasses.

The film shows the achromats who live on Pingelap and Pohnpei Islands squinting, blinking, and struggling to function outdoors without sunglasses. It allows viewers to see achromatopsia “in the raw,” so to speak.

In sharp contrast, soon after this film was aired in the U.S., a brief news film about achromatopsia was also shown on many TV stations. The scenes in this film were worlds away from Pingelap, not just geographically but also in terms of a whole different reality – a reality that included the services of a major medical center and an optometrist prescribing red contact lenses for a boy with achromatopsia. The boy and his parents (who are members of our network) consider the red contacts to be an important addition to the options he already uses, which include various sunglasses for different situations. For the filming sessions, the family had brought along all of his sunglasses to show. But, in the resulting film, no sunglasses were seen, and it was made to appear that this boy had been blinded by light – until the red tinted contacts were provided.

In recent years an increasing number of eye care professionals have been prescribing red lenses for rod monochromats. Some even prescribe red contacts for all rod monochromats, even very young children. Yet readers of our newsletter are aware that there are tinted lens options other than red lenses and that members vary considerably in their tinted lens preferences. There are those who dearly love wearing red lenses, and there are those who do not choose to wear red lenses and who do not choose to wear tinted contacts. (See comments on pages 80-93.)

The two preceding sections of this book present information about many options in tinted lenses, considering various factors that can influence which options are best for different individuals and pointing out both the pluses and minuses to every approach for dealing with our extraordinary sensitivity to light. There are so many variables in people’s lives.

In 1994 I started this network for achromatopsia and began publishing *The Achromatopsia Network Newsletter*. In the newsletters I consistently tried to represent the full range of networker responses about preferences with regard to tinted lenses. I have included material about preferences ranging from what might be called the minimalist approach (i.e., minimal use of tinted lenses in general and the use of extra dark lenses only when really needed) to the maximalist approach (i.e., regular use of lenses that are dark enough to create “twilight vision” for achromats). I have endeavored to incorporate my own experiences in such a way that my views and experiences would not have a disproportionate influence on others in the network. By following this policy, I have been able to practice what I so

often preach in the newsletters – which is that, while we can all help by sharing our thoughts and experiences, the fact is that what works best for one person is not necessarily right for others.

MY PERSONAL ACCOUNT: I am a complete rod monochromat, with total colorblindness, extreme sensitivity to light, and 20/200 visual acuity. Because I am one of the older network members, I have been able to experience various approaches to “treating” achromatopsia for more than five decades.

For the first 18 years of my life, I had to manage without any tinted lenses. I lived in relatively shade-free areas of Texas. I grew up before the age of special education resources for vision impaired students and long before specially tinted lenses became available. My family was very poor, so there was nothing to make life at home easier for a kid with achromatopsia – no window blinds, draperies, or especially designed shady rooms. I nearly always had someone to walk with me for any outdoor travel that involved crossing streets; but, in all other outdoor situations, I had to manage on my own, using visual strategies which included squinting, blinking, and “seeing” by way of brief “after-images” that would appear whenever I closed my eyes in bright sunlight. This ability to see with my eyes closed (which other networkers have also reported) is described on p. 36.

At 18 I got my first pair of “drugstore sunglasses” – not much help, but definitely better than nothing. Soon afterward, an eye doctor helped me obtain some darker (brown tinted) lenses. Having these lenses and the survival skills and visual strategies I had developed, I was able to travel independently fairly well. After obtaining a scholarship and other financial aid, I left home to attend a state university and then later traveled to Tennessee to attend Peabody College

at Vanderbilt University, where I majored in special education for the visually impaired. In the years that followed, I taught in special education programs in Texas, Kentucky, and California, including two public school programs and two state schools for the blind. I worked with totally blind students, students with low vision, and students with high levels of visual functioning.

As a special education teacher, I was often assigned bright classrooms, because vision impaired students usually need more light. To see my best in these settings, I obtained brown “indoor sunglasses” that were tinted lighter than my brown “outdoor sunglasses.”

When I spent a summer teaching English to migrant farm workers in California and had to stay in a primitive cabin at a farm labor camp, I found that I needed to wear extra dark sunglasses on a daily basis. I continued to explore possibilities for better tinted lenses.

Although there is much diversity among us, we older achromats tend to have certain things in common. (1) We all had to get along with little or nothing in the way of tinted lenses in our early lives. (2) We all managed to adapt in amazing ways to this bizarre way of seeing. (3) We all have, throughout our lives, cherished our ability to see well in lower levels of illumination (i.e., most indoor settings and outdoors at twilight or in very shady places). Most of us have always tended to take off our sunglasses just as soon as we enter any place with favorable lighting. Thus, we have been able to experience being rather normally sighted for a substantial part of our lives (depending on how much of our lives we have spent indoors). We enjoyed this freedom long before we learned the reason for it – the fact that we achromats have good rod vision, even though we do not have cone vision (see p. 1).

About 30 years ago I began experimenting with using extra dark tints, side shields, and visors and discovered that, if I routinely wore sunglasses made dark enough to give me optimal vision outdoors, my eyes soon became more sensitive to light levels that I had previously been comfortable with. I worried about this change in the good indoor vision I had always cherished. I gladly decided to accept a reduction in acuity outdoors, in favor of reclaiming my ability to tolerate moderate levels of illumination indoors (such as in my apartment, the homes of my friends, etc.). I consulted my eye doctor about this, and he responded that those of us who live with achromatopsia are the real “experts” on it and that I should follow what seemed right to me in terms of how much tinting I should use.

From that time on, I have evaluated each new tinted lens option by comparing (1) how much it helps with my visual problems in bright light (the abnormal part of my vision) and (2) what effect it has on my visual functioning in moderate and lower levels of illumination (the more normal part of my vision). Some optometrists have considered me foolish for not regularly using lenses dark enough to give me the best acuity in full sunlight (though I do use such lenses occasionally, when really needed). But I realize that they cannot understand the calculation of trade-offs which I have to use, for my own good.

Vision care specialists tend to view achromatopsia in terms of that which is missing or abnormal – the cone vision that is not there and the serious visual problems which we have in bright light because of this. Few of them seem to recognize and appreciate that which is normal about our vision – i.e., our good rod vision, which has amazing potential, if not interfered with.

In 1992 I helped organize a gathering

of persons from Northern California who were subjects in an achromatopsia study going on at UC Berkeley. This was the largest assemblage ever of persons having this rare disorder. I enjoyed getting to meet fellow achromats but was surprised to see that some of them were wearing sunglasses indoors. This meeting took place in the evening in a basement room, where the lighting was not particularly bright.

I also noticed that some of those who were not wearing sunglasses didn't seem to see as well as I did and that there was something odd about their eye contact with me. It reminded me of being with some of my low vision students back when I was a special education teacher. Later I learned that these persons were wearing red contacts, so I was able to understand why they had shown signs of reduced vision – just as I would have, had I been wearing tinted lenses in that setting.

Being with them also reminded me of the two times when I had been in the presence of a young man with achromatopsia who wore red contact lenses. On each of these occasions, the lighting had been quite comfortable for me, but he had needed to keep wearing his red contacts, and he was clearly having to function with less vision than I had, since he was having to wear dark lenses in low level lighting.

I was glad to hear about achromats who were using red lenses successfully; but, when I experimented with red sunglasses, I found that I did not like them, for a number of reasons. An optometrist who has several times supervised the making of custom tinted lenses for me (which have been mounted in wrap-around frames) is enthusiastic about red lenses for rod monochromats but has been wonderful about helping me get the dark brown lenses, with just a touch of red, which I much prefer.

Sometimes I have been able to find readymade sunglasses with dark amber or dark brown lenses, which have been almost as helpful as the custom tinted lenses I have had made.

EFFECTS OF SPECIFIC TINTS: In bright light, lenses that are tinted deep red create “twilight vision” for rod monochromats and allow a bit more visible light to pass through than can pass through amber or brown lenses that are dyed dark enough to offer the same degree of glare protection. Red lenses allow the eyes of the person wearing the lenses to be seen. Other lens colors, when dark enough to give achromats their best vision in bright light, look darker to the observer and do not permit the eyes of the person wearing the lenses to be easily seen.

Lenses of any color, if tinted dark enough, allow us to open our eyes and have more vision, just as we are able to do without tinted lenses at twilight time outdoors. At twilight the light around us is not bright enough to saturate our rods and cause reduced vision.

Some adult achromats, when offered red lenses, readily accept this option. However, many of us have found that we manage quite well with dark tints that are not red. This has been expressed in numerous networker responses printed in the newsletters.

When children with achromatopsia are given red lenses, it is usually the first chance they have had to try any lenses dark enough to actually help them in bright light. Many have previously had only ordinary children’s sunglasses, which usually are so light that they are virtually useless to achromats. Some have previously been given custom tinted lenses, but the lenses have been either tinted too light or tinted dark gray. Dark gray lenses have, for many years, been provided for light sensitive patients, and many eye doctors are not

aware that there are other dark tints besides gray that can be used now.

Among those who do know that recent advances in optical dyes have made other dark tints possible these days, it has become standard practice for many of them to offer only red lenses to rod monochromats. It is easy to demonstrate the difference it can make for patients with rod monochromacy when they first get to try adequately dark lenses of any color. Since so many doctors and optometrists offer only red lenses to these patients and do not make comparably dark lenses of other colors available to them to try, many people have the impression that red lenses are the only option for rod monochromats. Eye care professionals need to learn about the other options that are available today.

PLUSES AND MINUSES OF RED LENSES: Red lenses attract attention and have a “startling” effect on other people. Very young children are usually not aware of – or concerned with – this fact. But teens and adults with achromatopsia are aware of it and often will not even try red lenses, because they would rather not have yet another stigmatizing factor in their lives.

It is very important, however, to point out that there are other reasons that some persons with achromatopsia do not choose to wear red lenses. The negative experiences they have had with red lenses and their reasons for choosing other options deserve to be given proper consideration. Professionals who think that all rod monochromats should wear red lenses tend to be aware of only one objection which patients could possibly have – the objection to the strange, attention-getting appearance of red lenses. So these professionals tend to assume that any resistance to using them must surely be based on vanity and what they

consider to be the folly of valuing one's appearance and social acceptance over improved visual acuity.

But there are other problems networkers have reported. Starting with the least threatening of these problems, red lenses distort the appearance of many things. Certain colors seen through red lenses appear dramatically different. For example, a red cap or jacket, which would normally look very dark to an achromat will appear nearly white when viewed through red lenses. One networker whose place of business is filled with red countertops found that, after she began wearing red contacts, she could barely concentrate on her work, because she found it very disconcerting to see all the "white" countertops surrounding her. Adults can have an especially hard time adjusting to these visual distortions. Young children tend to adjust more readily to them.

Some achromats who use red lenses report that they get a special benefit from these distortions. They are able to identify certain colors by comparing how things look with and without the lenses. For example, they can tell that certain objects are red, because red things look so different through red lenses. Some report that they like being able to detect red lights or that they enjoy seeing red flowers so much more clearly (since red flowers turn nearly white). Nevertheless, for many persons, the distortions are disturbing and hard to accept.

Moving on to one of the more serious objections, some networkers have noticed that, while deep red lenses allow them to have better vision in bright, sunny places, they seriously reduce their ability to see when moving from sunny areas to shady areas, from outdoors to indoors, or from a brightly lighted room to a slightly darkened hallway, etc. They have to adjust to being

occasionally "blinded by the dark," in contrast to being "blinded by the light" (something to which achromats have long been accustomed).

When I experimented with wearing red sunglasses outdoors, I was surprised at how seriously impaired my vision would become whenever I approached shade trees or a shaded entrance to a building. At such times I did not want to remove the sunglasses, because I was continuing to move about outdoors, but I also did not want to have my vision so restricted, even if only intermittently. So I was never very patient with these experiments. I was always anxious to return to my dark brown or dark amber sunglasses, which did not darken the world quite as much in full sunlight as the red lenses but which allowed me to have better vision in the shadowy places. That was very important to me. I concluded that those who can so readily adjust to wearing red lenses must be able to accept this trade-off better than I could.

TINTED CONTACTS: Once I tried red contacts. Luckily I was able at the time to try dark brown tinted contacts too and so could compare the two tints. But I could make this comparison only because an optometrist who likes to fit achromats with brown contacts had sent me some to try. In the optometrist's office where I was being assisted in trying the contacts, only red contacts were provided for patients with achromatopsia to try.

It did not take me long to decide that tinted contacts were not for me, although I realize that they are a highly valued choice for many achromats. The most serious problem I experienced was the way they made me feel so much more visually impaired than I am whenever I would step into an area that was not well lighted. I knew, without a doubt, that, after so many years of

wearing sunglasses and tinted spectacle lenses – which can so easily be taken off and put on again, as needed, for optimal vision in whatever kind of lighting I find myself in – I was not willing to give up my accustomed flexibility and visual comfort for whatever advantages tinted contacts had to offer.

However, I suspect that, if I had been offered tinted contacts in my teens, I might have opted for them, since they would have allowed me to look more “normal” in some situations.

Networkers who wear tinted contacts manage one way or another to adapt to the lowered level of visual functioning which they inevitably experience when wearing them in poorly lighted places. Some do this by repeatedly moving their contacts away from their corneas and back again, as lighting changes, but this is not an advisable practice.

The brown contacts I tried that day caused me fewer problems when moving into poorly lit areas, and they provided clearer images than the red lenses did (brown lenses are known to give good contrast), but the red lenses made everything darker when I was in full sunlight. However, with either the brown contacts or the red contacts, I found that I had to use sunglasses anyway, wherever the lighting was bright.

Some achromats have had contacts tinted dark enough for use outdoors without having to wear sunglasses too. They simply remove these very dark contacts when going indoors.

I was told by the optometrist who was assisting me that my negative reactions to red contacts was “unusual.” At first I assumed that this had to do with my particular sensitivities and my determination not to give up any of the vision I am accustomed to having. But then I began to wonder – just how unusual was I? And unusual in comparison with whom? Could it be that not

many older rod monochromats ask to try red contacts, as I had done, and are then willing to report their reactions, openly and honestly?

INPUT FROM OLDER ACHROMATS: Since starting this network back in 1994, I have heard from many middle-aged and older achromats who, like myself, have been told that they were “unusual” in some way by eye care professionals who were mainly experienced with younger achromat patients. From all reports, it seems that the most common way in which we are considered unusual is that we have surprisingly good tolerance for light indoors, compared with most younger achromats. This is evident in the way we take off our sunglasses as soon as we enter a room with reasonably favorable lighting. When we have been observed in the waiting room of a clinic or optometrist’s office, we have been told that other achromat patients have needed to keep wearing dark glasses in these rooms.

We have also been told that we seem to have a high tolerance for unclear or “degraded” images (helping to explain our willingness to go without dark glasses whenever we can). This is not surprising. Living with achromatopsia forces one to adapt to a vast range of differences in the way the world appears, since so much of what we can or cannot see is affected by illumination factors. Our ability to tolerate unclear images has, in fact, played a big role in our developing and fully utilizing such vision as we are capable of having.

It is becoming increasingly evident that the adult achromats who have not become habitual users of lenses tinted dark enough to create “twilight vision” for them – whether in the form of the darkest Corning lenses, extra dark wraparounds, or red contacts supplemented with sunglasses, etc. – tend to be comfortable and to function well in a

wide variety of settings without having to wear tinted lenses.

CONCERNS ABOUT THE CHILDREN: In sharp contrast, there is increasing evidence that many children with achromatopsia are wearing tinted lenses most or all of the time, even in lower levels of illumination. The ones who have become the most dependent on dark glasses almost invariably are those who were fitted with very dark (usually red) lenses very early in their lives. Several network parents have reported that they sometimes have to make sure that their children remove their dark glasses at bedtime! The children and teens who have been fitted with red contacts usually put their contacts in first thing in the morning and do not take them out until they go to bed at night. However, it is important to point out that, in these cases, the contacts are at least not ones that have been tinted dark enough to suffice as sunglasses outdoors; they are contacts that have been tinted just dark enough for use in bright indoor spaces, outside when it is overcast, or in combination with sunglasses in full sunlight. Even so, these reports bring cause for concern.

All indications are that children with achromatopsia who start wearing extra dark lenses early in life develop low tolerance for indoor lighting. The following factors seem to affect how seriously impaired a child's tolerance for light will become: (1) how dark the lenses are which the child has been given to wear, (2) how early in life the use of such lenses was begun, and (3) how much time a child has spent wearing very dark lenses (or wearing a combination of tinted lenses and tinted "fit-overs" to provide even more light protection outdoors).

There has never been – and probably never will be – controlled experiments regarding matters such as these. Any such study would have to be involved

with regulating the amount of light that reaches the eyes of a certain number of children over a period of time – which could be seen as either imposing light deprivation on some children or subjecting others to unnecessary light exposure, depending on one's viewpoint.

Adults, whether visually impaired or not, might voluntarily conduct their own experiments with dark adaptation and light adaptation, using various tinted lenses over different periods of time. But experimentation of this type with the vision of children would be considered unacceptable by most standards, especially if done in the early stages of a child's visual development.

The matter of choosing the best approach to the problem of hypersensitivity to light is an example of the many important things in life about which people must make decisions but for which there does not exist good scientific data to turn to for answers about the "right" thing to do. In fact, most of the big decisions we make in our lives are not ones we can base on good scientific research. So we gather such information as we can in whatever ways we can – intuition and insight, personal experimentation (trial and error), reading up on a subject, using common sense, learning from the experiences of others, and consulting specialists.

KNOWLEDGE IS LIMITED: How much do the vision care specialists really understand about ways to cope with achromatopsia? Few of them have had very many patients with achromatopsia, and even those who are fairly knowledgeable about this disorder and who have encountered a goodly number of patients with achromatopsia are limited in how much they actually understand about it. Some may even assume that it is normal for achromats to have to wear tinted lenses in low light levels, so that they wouldn't even recognize this as

something to be concerned about.

Parents of children diagnosed with achromatopsia cannot be expected to know the normal or expected visual behavior of persons with this eye condition, so they too may assume that it is normal for achromats to need tinted lenses in low light levels. And the children who have been given dark lenses early in their lives and have become dependent on them have simply adapted to these lenses and to whatever effects that wearing them regularly may have had on their vision in lower light levels.

Matters worthy of concern in connection with how achromatopsia is being treated by eye care professionals are not going to jump up and grab the attention of these professionals, but they do come to my attention as facilitator of this network, since I hear from numerous people all over the world who have this rare disorder. This puts me in the difficult position of having to call attention to these matters.

My objectives in this section are (1) to state concerns that need to be heard, (2) to raise questions that need to be asked of the specialists, and (3) to make suggestions that need to be considered. This report is not about criticism or judgment. The vision care specialists I am in touch with are very good people who sincerely want to help achromats with their special needs. There is so much that they can learn from achromats, especially from those of us who have lived many years with this extraordinary way of seeing. Hopefully, these professionals will welcome the information and understanding which this book has to offer.

A doctor or optometrist can easily demonstrate that, by giving a child with achromatopsia lenses that are dark enough to approximate twilight vision, whether in the form of deep red lenses or dark lenses of some other color, the

child becomes free to run about and play in the sunshine. This can be a deeply moving experience for all concerned – the child, the parents, and the professional who gets the satisfaction of having helped the child obtain significantly improved vision outdoors.

But wise management of achromatopsia needs to go beyond this dramatic discovery of new freedom outdoors, as impressive as that may be. Every method for controlling hypersensitivity to light needs to be looked at in terms of pluses, minuses, and trade-offs. Deciding what is truly best for achromats means looking at what is best for now and also what is best for later stages of life – best for dealing with special needs in bright light and best for the full range of life activities that take place in lower illumination. Wisdom, experience, foresight, and sound knowledge about the eyes, about vision, and about achromatopsia – all of these elements should play a part in deciding which lenses are best for achromats.

The only knowledge about achromatopsia which professionals have (unless they have studied our network literature) comes from whatever experience they have had with achromatopsia patients and whatever information has been included in their training or in the professional literature they have read. Their professional literature contains very little about achromatopsia, and such information as it does contain mainly deals with diagnosis, prognosis, inheritance factors, and vision testing.

Even those who have basic scientific knowledge about achromatopsia and strong opinions about how to treat hypersensitivity to light may have little or no understanding about our visual experiences, especially about how well we can see in moderate and lower levels of illumination. Some professionals may even believe that, if their methods

for “fixing” our light sensitivity problem (i.e., through regular use of lenses dark enough to keep our eyes in perpetual twilight) happen to adversely affect our vision in lower light levels, then this should be seen as an acceptable trade-off. But do they have the right to make such a decision?

With very few exceptions, vision care professionals have not accompanied achromats into the sunny outdoors in order to learn more about their light sensitivity problem, nor have they accompanied them home or to other comfortably lighted places to learn more about the other side of living with achromatopsia. They do not know how well we see and function in certain settings. This is not to suggest that they should be spending time with achromats, as interesting as that idea may be. The point is simply that there is so little real knowledge about what achromatopsia is. How then can any of them state with certainty what is “best” for us?

The story of what it means to have achromatopsia has never been written up in publications aimed at vision care professionals. Hopefully, many of these professionals will read our network literature and will also look into the vision science book *Night Vision* (Cambridge University Press, 1990), which includes historical and scientific information about achromatopsia, as well as a chapter entitled, “Vision in a complete achromat: a personal account,” written by Knut Nordby. Dr. Nordby has had a long and distinguished career as a scientist, university teacher, and vision researcher. His own preferred methods for coping with his achromatopsia include regular use of moderately tinted lenses outdoors and only occasional use of very dark tinted lenses.

In his chapter in *Night Vision*, Dr. Nordby describes his use of visual strategies that are used by achromats

everywhere, including squinting and blinking. These manifestations, which are crucial to the visual functioning of achromats, are sometimes viewed negatively by eye care professionals. In fact, some have even developed theories about how to manage achromatopsia based on the belief that our squinting and blinking should be prevented. Adult achromats would have a lot to say about that theory! Hopefully, the specialists will want to hear what older achromat patients have to say.

TINTED CONTACT LENSES: Some specialists believe strongly in fitting all achromats with tinted contacts and getting children with rod monochromacy fitted with red contacts as early as possible. The pluses of tinted contacts are appreciated by many networkers, and quite a few comments from these satisfied individuals have been included in network newsletters. But wise consideration of this option requires looking at the minuses as well as the pluses. As mentioned earlier, many achromats do not like wearing tinted contacts, and many do not choose to wear red lenses, for a variety of reasons.

Parents, understandably, want their children to look good and not to have to be seen wearing dark glasses so much of the time. They want to see their children’s eyes, and other people want to see the children’s eyes. Tinted contacts permit this. Another plus for tinted contacts is that children and teens who wear them reportedly have better self-image and more self-confidence in social groups.

And, since so many achromats get fitted with Rx lenses early in life and thus grow up having to wear spectacle frames, the option of tinted contacts typically represents their first chance to experience a full visual field. Many have had to grow up wearing opaque side shields, because their tinted lenses

have always been mounted in spectacle frames, due to the Rx. By contrast, those of us who grew up stubbornly refusing to wear Rx lenses (or deprived of optometric services and so never having gotten started wearing Rx lenses) have been accustomed to wearing wraparound sunglasses instead of spectacle frames. So the “wraparound” aspect of tinted contacts does not impress everyone in the same way.

When network parents tell me they are eager to have their children fitted with tinted contacts and want details about this option, I tell them that, while I do not wear tinted contacts myself, I can refer them to networkers who do and to professionals who can advise them. I tell them that there are both pluses and minuses to be considered, and I refer them to newsletters which have included networker comments on this subject.

I frequently send out reminders to networkers that input is needed from achromats who have experience with wearing tinted contacts, especially those who have long term experience with wearing them.

Some networkers have experienced minor corneal problems, irritations, or a problem called “dry eye” in connection with wearing contacts and have had to either stop wearing them or restrict the amount of time they wear them.

One network mom, whose school age son has been wearing red contacts since the age of 4, reported that, when he had to go without his contacts for a few days while recovering from conjunctivitis, he was virtually like a blind child. He was not able to just start wearing sunglasses instead, which would have enabled him to go to school or go out to play during this period. She said she felt that, if he ever has to manage without his tinted contacts for a longer period of time, he will probably be able to

make the adjustment. We need to hear from achromats who have used tinted contacts for a substantial period of time and who have been able to make the transition to sunglasses, when needed (either temporarily or long term) with little or no trouble. It would also be helpful to hear from anyone who has grown up wearing red lenses and who has been able to adjust to using other lens colors, when they decided to do so.

The strongest anxiety expressed by any networkers has come from users of tinted contacts who fear that, for one reason or another, they may not always be able to have the tinted contacts to which they have become accustomed.

It appears that the children who have been fitted with tinted contacts early in life tend to stay more dark adapted and thus to have especially low tolerance for light without their tinted lenses on. Some of them even need to wear lightly tinted contacts outdoors at night.

One teenager who has worn red contacts for many years reported that, in order to see her enlarged sheet music during band practice on a poorly lighted stage, she has to have a lamp attached to her music stand. Another teenager who has been wearing red contacts for several years will remove them in a darkened restaurant but will not take them off when going into the basement of his home. Instead he turns on lots of lights. Other users of tinted contacts report that they turn on many lights, wherever lighting is subdued, rather than remove their contacts. Some use a normally sighted companion in order to travel confidently in rural areas at night while wearing tinted contacts.

Some of these people, by the way, are not complaining about the reduced vision they have in lower light levels due to having tinted lenses on. To those who have grown accustomed to seeing through tinted lenses most or all of the

time, doing so seems quite natural. Some adults who begin wearing tinted contacts are so enthusiastic about the advantages that contacts offer outdoors that they willingly accept the trade-offs. But this is not true for everyone. For some, these trade-offs are not considered acceptable.

CHERISHING OUR ROD VISION: If you were to take a poll of older achromats about which visual experiences we have most cherished during our lives, most of us would probably answer that our freedom to enjoy relatively normal vision in favorable lighting is very, very precious to us. We have our tinted lenses and our various strategies for coping in the sunny outdoors and other bright places. But we find it supremely satisfying to enter that other reality – i.e., our homes and other visually comfortable places, where we can take off our tinted lenses and just forget about being visually impaired.

And, if you were to poll older achromats about whether we would like for our vision to be always at twilight level, through regular use of very dark glasses, you would find that many of us feel strongly about the value of having lots of sunshine in our lives and about the freedom to experience visually the full spectrum of day and night. Few (if any) of us would accept the option of seeing the world always at twilight. Like other human beings, we find the presence of sunshine uplifting and essential for good mental health.

Some adult achromats have tried hard to become accustomed to wearing very dark lenses but have found themselves becoming depressed as a result of seeing so little sunshine. They have chosen not to use extra dark glasses routinely but to keep them on hand for situations in which that much light protection is actually needed. Whether or not depression related to light

deprivation may also affect children who routinely wear very dark lenses is something about which I have not received any reports. It may be that, just as children can more easily adapt to the color distortions created by red lenses, they might also be less susceptible to the kind of depression reported by some adults who try wearing extra dark lenses on a regular basis.

In the general population the effects of light deprivation are well known, and light therapy is used during the darkest months of the year to treat seasonal depression related to light deprivation.

It is interesting to note that the lenses that are being prescribed for so many children with achromatopsia these days are making it possible for them to spend much more time outdoors than our generation did, and yet, in terms of their vision, these children are being “kept in the dark” in a way that our generation was not.

For the middle aged and older adult achromats in the network, there were no children’s sunglasses fitted with dark lenses and side shields and no wraparound frames with custom tinted lenses, such as are possible today. Consequently, such photoreceptors as we had were kept very active, accommodating as best they could to a wide range of illumination levels.

What we were able to accomplish with only (or mainly) our rod vision has been quite impressive. We grew up reading standard size print (getting close to reading material). We generally did well in school. We developed normal handwriting, though some of us have clearer handwriting than others, just as is true for people in general. We faced many problems because of our vision impairment, and we would have surely benefited by having magnifiers and monoculars such as are available today; but, since we didn’t have them, we

managed in whatever ways we could. Because there are many individual differences among us, we, of course, did not all do equally well in all subjects and all activities. But many of us went to college, and quite a few went on to graduate school – and all of this before it was possible to get good dark tinted lenses. Pointing this out is not meant to take away from the appreciation of all the wonderful options in tinted lenses these days but to emphasize the importance of recognizing the amazing potential that our eyes have.

THE EFFECT OF DARK LENSES ON READING: Some of the children with achromatopsia today are able to use standard school materials, just as we did when we were growing up, so that large type materials or magnification aids are seen as options, rather than necessities, at least in the early grades. But many others seem to require large print materials, even at an age when reading materials are usually in a print size that should be fairly easy for achromats to read. The children who are unable to use standard materials in the early grades are the ones who wear rather dark lenses indoors (and even in the evening at home).

I experimented with wearing lenses having a medium tint in the kinds of indoor settings where I usually do not wear tinted lenses, in order to see how this might affect my reading and writing. I found it to be difficult and frustrating. I suspect that I too would have had a much harder time using standard print school materials as a child, if I had been dependent on wearing dark lenses indoors. A light tint would have helped, at least in some classrooms, but the dark lenses that so many children with achromatopsia are wearing indoors these days would have seriously restricted my range of choices in reading materials then and would have

probably also affected my reading options and my adaptability as a reader in my adult life.

What is happening to the rod vision of so many children with achromatopsia today? Why are so many having to wear such dark lenses in the levels of illumination in which achromats of previous generations have been able to function well without tinted lenses? Are the specialists who are prescribing lenses for these children taking into account their overall needs and considering what is best for their visual functioning in all situations, or are they focusing so hard on trying to “fix” that which is abnormal about achromats’ vision (their hypersensitivity to light) that they are not even mindful of the effects that their recommendations may be having on that part of a child’s vision which is normal and healthy (their rod vision)?

There is such a thing as over-correcting a symptom. Over-corrections tend to cause other problems for a patient. There is also such a thing as under-correcting a symptom. Under-corrections are sometimes chosen by physicians in order to avoid – or at least to minimize – the possibility of causing other problems for a patient. And there are such things as “quick fixes,” which can bring impressive results in alleviating symptoms but which often cause side effects or after-effects. The history of medicine is filled with examples of “cures” which gave quick results but which created new problems for patients – in the same way that any natural ecological system can become out of balance, if tampered with.

These days, when doctors prescribe medications or other kinds of treatment, it is required that patients be informed about any possible side effects, even though those side effects may be known to affect only a small part of the population. It was not always thus. Not many

decades ago, doctors could prescribe medications with little or no knowledge about possible side effects and after-effects and no obligation to inform patients about such matters.

Is it possible that there may be “side effects” connected with the increasingly common practice of fitting achromats with lenses that are calculated to keep them from squinting and blinking, thus maintaining “twilight vision” for them? Have the specialists pondered or researched this possibility? Have they made efforts to keep track of their patients’ overall visual functioning (their dark adaptation, as well as their light adaptation) after having been fitted with such lenses? There are ways that these things can be monitored.

Unfortunately, many professionals do not have good systems for follow-up of patients, and many patients do not voluntarily provide feedback. Sometimes patients, especially children, are not even aware that some part of their visual functioning is being adversely affected. Patients tend to feel intimidated by professionals and often don’t think there is any opportunity for dialogue. And many of them just assume that the professionals know best.

When an optometrist recommended red contacts for her son, one network parent asked him if there was any possibility that using them might cause her son to become less tolerant of light and progressively dependent on the contacts and was told that there was nothing of the sort to be concerned about. But, when she reported this to me, I could easily recall two patients of this optometrist who had indeed become quite dependent on their red contacts, unable to do without them except in very dim lighting. I don’t think this optometrist was deliberately being deceitful. More likely, there just hasn’t been any feedback to him from these

patients or their parents and no curiosity on his part, since he (like so many people) has probably never stopped to wonder about such things.

PROBLEMS WITH RED LENSES:

I have spoken with a number of specialists over the years about the problems I have experienced with red lenses and why I prefer using certain other tints. I have hoped that, by doing so, I would raise their awareness regarding the differences that exist among achromats and sensitize them to the problems that some of us have had with red lenses. I wish I could report that my input has always been welcomed and taken seriously, but I cannot.

One of the problems I have had in my experiments with red lenses is the sense that my nervous system was being adversely affected. Symptoms such as irritability, restlessness, and sleep problems would occur whenever I would spend much time wearing red lenses. I suspect that reactions such as mine are not common, but, there are other adults in the network who have reported having similar problems.

Reports such as these do not tend to be well received by specialists. A patient talking about such matters can easily be dismissed as “flaky.” Nevertheless, I knew that the unsettling symptoms I had experienced when wearing red lenses did not occur when I would wear amber, brown, or gray lenses. After starting to network with other achromats, I felt relieved and validated to learn that some of them had also experienced mood disorders or nervous system related problems in connection with red lenses.

One man said that, on the days when he has to wear his red lenses for very many hours, he feels unusually irritable by the end of the day. Another said he was conscious of how red lenses stimulate his nervous system, so he limits

the amount of time he wears them each day. Another said he had to stop wearing red lenses, because he was aware of the “red cast” they gave to everything he saw through them, which seemed to continually jar his nervous system. He felt that this might be connected with the fact that he is an incomplete rod monochromat and he has some ability to perceive red. His sensitivity to seeing everything looking “reddish” might be compared with how many normally sighted persons would feel if they were continually viewing the world through red lenses. Many incomplete rod monochromats are able to perceive some reds.

One networker tried wearing red contacts for several days and reported that she began to feel as if she were losing her mind. She stopped wearing them, went back to her regular sunglasses, and was soon feeling fine.

It is important to emphasize that many achromats really love their red lenses and feel fine when wearing them. Some have worn red lenses for many years. Only occasionally does someone report symptoms such as those just described. But, even if such reports represent only a small minority of achromats, their experiences are valid and deserve to be taken seriously.

Unfortunately, most vision care specialists do not tend to be receptive to subjective information (i.e., information which cannot be verified with clinical or laboratory tests). However, practitioners of various healing arts, especially those who have studied color therapy, are aware of the effects that colors can have on human beings. Red is known to energize and stimulate, and it can reportedly have a therapeutic effect on individuals who need energizing or stimulating. On the other hand, red could conceivably have adverse effects on individuals who are very sensitive

and who do not need energizing or stimulating.

Many networkers are concerned that the overuse of “twilight vision” lenses with infants, toddlers, and young children with achromatopsia is interfering with some very important development that needs to take place in their visual functioning. We fear this may seriously affect their potential for mastering the natural visual strategies which they are going to need for a lifetime of seeing and surviving with only rod vision. As yet, science knows very little about the plasticity and adaptability of the human visual system, but it is widely recognized that the early years in a child’s life are crucial in terms of developing basic visual skills. The accumulating evidence that so many young children with achromatopsia are wearing tinted lenses from wake-up time until bedtime gives much cause for concern.

None of us would want the children to have to endure the kind of existence outdoors that has been endured by the Pingelapese achromats (mentioned on p. 94) and by those of us older achromats when we were growing up. But neither do we want to see them deprived of the good vision which is possible for them in favorable lighting.

It is not for me to suggest any course of action in the treatment of vision problems. What I can do, however, is to gather and report information on this subject for the network publications.

Eye doctors and optometrists are the ones who do the prescribing. Ideally, their style of prescribing will go something like this: “Here is my opinion (or my theory) about what is best, and here are some other options which you may want to consider.”

Responsible vision care for achromatopsia patients should include protecting their good rod vision as well as seeking to alleviate the problems caused

by their lack of cone vision. The ability to fully enjoy and fully utilize good rod vision is our natural birthright.

HOW PROFESSIONALS CAN HELP:

Professionals should be willing and able to answer questions about what they base their recommendations on. What experience with achromatopsia patients – and follow-up of these patients over time – can they report? What studies pertaining to achromatopsia can they refer to that have influenced their beliefs about what is best for achromats? Since starting this network, I have had searches made of the professional literature in order to compile a reference file on achromatopsia. I welcome any useful references and copies of any pertinent articles which networkers can obtain from their specialists in support of their recommendations.

Probably most of us would prefer to think that someone could just give us the “right” answer. Some totally reliable recommendation based on good scientific studies that have been conducted over a substantial period of time, using a broad sampling of subjects with achromatopsia – studies that had taken into consideration not just visual acuity and certain other visual measurements but all of the factors that constitute optimal quality of life and optimal visual functioning in all situations for achromats. This would be ideal. But achromats are extremely rare, and research pertaining to them has been correspondingly rare, as well as limited in scope. So, while we can make use of such research findings as do exist, as we seek answers to the question of what is best for achromats, we also need to gather information in other ways.

Our network is a valuable resource for this purpose. Members of our network exchange information and experiences with regard to various different approaches to light sensitivity. People

with achromatopsia deserve to have choices, to be informed about what their choices are, and to know what the pluses and minuses of the various options are. As already mentioned, many achromats have been very satisfied with the option of red contacts or red sunglasses, but others prefer other options for dealing with their light sensitivity. Hopefully, no professionals will seek to control the information that patients can receive about what their options are.

Surprisingly, there have been a number of reports about professionals who, when asked to explain their reasons for recommending red lenses, have responded with statements like, “I heard somewhere that this is what is best for achromats,” or “Everyone knows that achromats should wear red lenses,” or “Experts say that red contacts are best for rod monochromats.” Answers such as these are not acceptable coming from professionals. They should be willing and able to support their recommendations by giving specific and substantial information.

It is reasonable to expect that many professionals will suggest red lenses. What is objectionable is the dogmatic way in which a “one size fits all” manner of prescribing is being used with so many patients with achromatopsia, usually with no consideration at all being given to the other alternatives which many achromats prefer.

It is especially important that red lenses not be prescribed for patients who are found to be – or even suspected to be – blue cone monochromats or for incomplete rod monochromats who have usable color vision. The distortion of colors caused by red lenses may have little or no serious effect on the vision of children who have total rod monochromacy, but it is an entirely different matter when it comes to those who are incomplete achromats and who

need to use such color vision as they have. If it is possible that a child might be able to see red, it is not good to give the child red lenses, since they will make everything look “reddish.”

Some eye doctors, having heard that red lenses are best for rod monochromats but knowing little else about this, have actually advised some parents to see that their children wear dark red glasses indoors as well as outdoors. Some doctors have advised parents to obtain red lenses for children who have been diagnosed with achromatopsia, even when the children show no signs of abnormal light sensitivity (and, to any well informed person, this would raise questions about the accuracy of the diagnosis). If the child in question happened to have any degree of color vision, then red lenses would be entirely inappropriate and would seriously hinder the child’s color vision.

Infants as young as 5 months old who are thought to have achromatopsia are being fitted by some doctors with spectacles that have dark red lenses and opaque side shields, and the spectacles are strapped to the infants’ heads. What chance do these infants have to experience and develop their normal rod vision in favorable lighting? What chance do they possibly have to escape the fate of becoming chronically dark adapted very early in life? What if the infant happens to be unusually sensitive to the effects of red lenses, as some adult achromats have discovered that they are? How will the infant be able to develop important visual skills involving light adaptation or peripheral vision? What if it turns out to be a case of misdiagnosis? Some ophthalmologists say that it can sometimes take years before a diagnosis is absolutely certain. These are some examples of how this widespread message of “red lenses for all achromats” has gotten

out of hand.

There really are no experts on this rare vision disorder. But there are some eye care professionals and some vision researchers who are especially knowledgeable about many aspects of achromatopsia, and some of these people have developed theories about what is best for achromats. There may eventually be substantial research findings that will support one or more of their theories; but, until that time, it is important for persons who are concerned with achromatopsia to understand that these are, in fact, only theories.

The best theories are ones that are formed after taking into account all of the important factors related to the problem that needs to be dealt with. In the case of achromatopsia, this means looking at much more than just visual acuity and other visual functions which can be measured in doctors’ offices and in laboratory experiments.

As mentioned earlier, one of the theories that some professionals are currently being influenced by is the theory that it is important to prevent achromats from blinking and squinting. Some network parents who have had this theory impressed upon them by a vision care professional will say that they feel they must obtain lenses for their children that will prevent them from squinting and blinking – and yet these parents usually will also say that they really don’t want their children to become “addicted” to wearing dark lenses all the time. Unfortunately, it is not possible for both of these wishes to be granted at the same time. All indications are that the routine wearing of lenses that are dark enough to keep achromats from squinting and blinking (i.e., lenses that keep their eyes in virtual twilight) do tend to keep them so dark adapted that their tolerance for light is inevitably affected.

Some of us have found it helpful to use the analogy of persons who have certain orthopedic problems which impair their ability to move about easily, gracefully, and at a normal pace but which are not so severe as to require using a wheelchair. In terms of maintaining muscle tone and optimal health, it is often far more advisable for such persons to keep active physically and to move about on their own, rather than resorting to extreme measures, such as being sedentary or always using a wheelchair. Depending on the condition, it may be necessary to make occasional use of a wheelchair or to limit activities that require a lot of walking. But the general principle of continuing to exercise such abilities as one has is widely recognized.

There are many people who find it disturbing to see someone who has a mobility impairment struggling to walk – just as there are many people who find it disturbing to see achromats having to blink, squint, or otherwise look odd, as they struggle with visually demanding tasks in less than ideal lighting. But to assume that people with physical, visual, or other kinds of limitations should always use devices or methods which keep them from ever having to struggle with their limitations is to deny them whatever short term or long term benefits they might derive from discovering and exercising their own natural coping mechanisms. Keeping the vision of someone with achromatopsia always at twilight level, whether through overuse of very dark lenses or through the avoidance of all unfavorable lighting, seriously limits the versatility and the potential of their rod vision.

Among people with achromatopsia, just as with all other people, there are “outdoor types” and “indoor types,” and there are “night people” and “morning people.” There are individuals who are

drawn to outdoor sports, and there are those who prefer sedentary activities that take place in subdued lighting. The kinds of lenses that are needed by the ones who gravitate toward spending lots of time in the sunny outdoors will not be appropriate for the more stay-at-home types of people. Therefore, for some individuals, the long-term consequences of having developed, early in life, a dependency on wearing dark glasses in low level lighting are far more serious than they are for others.

One can always choose, at any point later in life, to begin wearing darker lenses to accommodate a transition to an outdoor lifestyle, but it is not so easy to do the reverse – i.e., to undo a dependency on wearing dark lenses indoors, for those individuals who may want, more than anything else, to have optimal vision for a lifestyle based on indoor or night time activities.

There are many things that eye care professionals can do to help minimize this problem. They can advise moderation in the patient’s use of extra-dark lenses and explain why this is important. And they can be sure that parents know what signs to watch out for that would indicate that a child is becoming overly dependent on dark lenses.

It is important not to coax or coerce a child with achromatopsia to accept glasses that he or she instinctively resists wearing. Remember that non-achromats simply cannot experience the visual reality that achromats live with.

If very dark glasses are provided for the child to use outdoors, be sure also to make available some lenses having a lighter tint, so that the child has a choice and, therefore, will not get into the habit of using unnecessarily dark lenses in moderate lighting, just because some degree of glare protection may be needed and there are no lighter tinted lenses for the child to use.

PRESCRIPTION LENSES: Almost all achromats, when given a thorough vision exam, are found to have some degree of refractive error (nearsightedness, farsightedness, or astigmatism). Some are found to have a significant refractive error, but this is not true for most of them. However, optometrists and ophthalmologists want to be able to provide some kind of help for these patients; and, since they cannot correct the impaired eyesight that results from lack of cone vision, they tend to concentrate on what they can do, which is to provide Rx lenses for whatever degree of nearsightedness, farsightedness, or astigmatism a patient may have. To them, this makes good sense. But this, like everything else in connection with treating persons with achromatopsia, needs to be looked at more carefully and more “holistically” – i.e., giving consideration to what actually constitutes optimal vision for achromats (indoors as well as outdoors) and what is best for them in the long run.

Consider carefully how one thing leads to another. Fitting a child with Rx lenses almost always means having the child begin to wear spectacle frames to accommodate the Rx lenses. Having to wear spectacle frames outdoors (instead of sunglasses that wrap around) means having to deal with the problem of light entering from around the edges of the frames, mostly at the sides. (This is more of a problem with some frames than with others.) This means either learning to live with the problems that this creates for achromats or else doing something to block the peripheral light. Often this requires using opaque side shields, which can be very uncomfortable, especially when the weather is hot and humid. Lenses can fog up easily, making it difficult to see. Side shields are conspicuous, some feel sharp against the skin, and they seriously

limit access to one’s peripheral vision. Hats, visors, or caps can be used instead of (or in addition to) side shields to alleviate the problem of side glare for those who must wear spectacle frames outdoors. But, like side shields, they also restrict peripheral vision. Since achromats have poor central vision, they tend to rely on peripheral vision even more than other people do.

After patients have been fitted with Rx lenses and become accustomed to wearing them, what often happens is that their eyes become so dependent on the Rx that, when they take their glasses off, everything looks blurry – something that was not true for them back before they started wearing Rx lenses. Some networkers have reported that this blur will eventually clear up, if they keep their glasses off long enough, so that their eyes get used to seeing as they did before using Rx lenses.

Having to wear spectacle frames, for the sake of Rx lenses, seriously limits the kinds of sunglasses that children will be able to use outdoors. They may use tinted “fit-over” type sun goggles or “fit-over” kinds of sport shields in combination with their tinted spectacle lenses, but they are deprived of many of the very best options for dealing with their extreme light sensitivity outdoors, such as close fitting wraparound sunglasses (either readymade ones or those that are fitted with suitably dark custom tinted lenses). Well designed wraparound sunglasses offer excellent protection from glare while providing full access to one’s peripheral vision. The options in sunglasses available today are attractive and comfortable to wear. Achromats who have grown up without wearing Rx lenses (or who, at some point in their lives, have stopped using Rx lenses) have been able to experiment with and enjoy a wide variety of sunglasses.

It just makes good sense to carefully compare the advantages of using Rx lenses with the effects that this choice can have on a child's present and future options in terms of coping with the far more serious visual problems that are caused by achromatopsia.

Some ophthalmologists have already thought about these matters and, having observed how little benefit some children with achromatopsia actually get from their Rx lenses (and taking into account the fact that using the Rx will seriously limit their choices in sunglasses) have advised parents that the Rx lenses are not really necessary. Usually, the children already know this, because, while they are acutely aware of the nuisances that are involved in wearing corrective lenses, they are not aware of any significant vision improvement when they have them on.

Many achromats report that the Rx makes things look a bit sharper and lets them hold reading material up to an inch farther from their faces but that they do not mind at all doing without these small benefits. Others, who use a strong Rx or who have been accustomed to using an Rx for many years, usually find their Rx indispensable.

Many people who have been fitted with weak Rx lenses early in life go on to need increasingly stronger Rx lenses as years go by, while others' Rx does not change much over the years.

Many ophthalmologists and optometrists respect the rights of patients to decide for themselves about whether or not to use an Rx. Others urge patients to wear them, even when there is strong resistance. In general, optometrists seem to be more inclined than ophthalmologists to urge children and their parents to see that the prescribed Rx lenses are worn, even when only mild corrections are involved. Optometrists tend to place an especially high value

on any slight improvement in visual acuity that can be achieved with lenses, using standard testing methods. But keep in mind that visual acuity testing is done in examining rooms that have low level lighting.

To my knowledge, no one has ever used standard tests to evaluate the differences in the visual functioning of achromats in full sunlight outdoors, comparing a patient's visual acuity when Rx tinted lenses are worn with the visual acuity when equally dark tinted lenses without an Rx are worn.

If an achromat patient is given tinted lenses for outdoors that have an Rx, it can be very important that the patient be given more than one pair of lenses. Ideally, those who really do need to wear Rx lenses should have (1) dark Rx lenses for outdoors, (2) lighter tinted Rx lenses for moderate lighting levels, and (3) clear Rx lenses for favorable lighting conditions. Sometimes children will be given only one pair of tinted lenses, ones that have a dark tint for outdoors and an Rx. Consequently, they tend to keep wearing their dark lenses in settings where a tint is not really needed, rather than to take them off and live with the "blur" that can result for those whose eyes have grown accustomed to wearing Rx lenses.

There are exceptions, however. Sometimes the Rx in an achromat's dark tinted lenses is virtually imperceptible to him/her and is not even missed when the sunglasses are taken off indoors. And those who like to use clear or lightly tinted Rx lenses indoors sometimes report that having the Rx makes so little difference in the way they see outdoors that they don't bother with having it included in their outdoor tinted lenses.

When a patient has a high degree of farsightedness or nearsightedness, all of these matters need to be looked at very differently, because high refractive

errors are usually regarded as significant visual problems. But, since so many achromats have only mild or moderate refractive errors, it is wise to look at the possible consequences of starting children with achromatopsia on the road to lifelong use of prescription lenses. Doing so is almost certain to deprive them of the chance to use many of the types of sunglasses that can offer achromats the greatest amount of help outdoors.

Many of us in this network fervently hope that achromatopsia will be better understood by vision care professionals. Our basic messages are as follows: Hear our story. Try to understand. Help, if you can – but, mainly, do no harm. At least, do no harm.

HOW ACHROMATS CAN HELP THE VISION CARE PROFESSIONALS:

1. Don't be quick to criticize those who aren't very knowledgeable about achromatopsia. Remember that this condition is extremely rare and is only one of numerous rare eye conditions which ophthalmologists and optometrists may or may not ever encounter. These professionals mostly deal with a wide range of common eye conditions and many others which are not common but not as rare as achromatopsia. Professionals should turn to their reference materials or to professional consultants when they encounter a rare disorder which they have never treated before, and they can be faulted if they fail to do this. But they need not be faulted for having been found lacking in knowledge and experience regarding achromatopsia at the time they encounter their very first patient having this rare disorder.

2. Be prepared to serve as one of the best sources of information that your doctor can consult. Many of them welcome information in print. Urge them to obtain the materials available through the network, or loan them your own

materials. Some networkers purchase printed materials to give to eye doctors, and some pass on information verbally during appointments.

3. Actively participate in decisions about what kinds of tinted lenses will be best for you. These decisions need to be based on (1) knowledge about the options that are possible for achromats (the material found on pp. 57-114 of this book can be very helpful) and (2) the knowledge and resources which each person involved brings to the situation – the patient, the optometrist or ophthalmologist, and the optician who will be creating special lenses in the optical lab (if custom tinted lenses are made).

4. Stay involved every step of the way, when custom tinted lenses are being made. Provide feedback right away and also later on, when you are able to report on how well some new lenses or sunglasses have worked for you. This is how professionals can learn what they need to know and how you will be able to get what you need to have. Don't be silent about something that doesn't work well for you.

5. If you have eye care professionals who take the time to listen, who try to understand, who treat you in a positive manner, and who help you to obtain good tinted lenses, be sure to let them know how much you appreciate them. Remember that meeting the unique needs of a patient with achromatopsia is not an easy task.

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As network facilitator and editor of the Achromatopsia Network Journal, I invite input about all these matters from individuals and families affected by achromatopsia and professionals interested in helping patients who have achromatopsia. Please send your suggestions and experiences regarding tinted lenses for achromats to any of the addresses listed on page 141.

Sources of Tinted Lenses and Related Products (U.S. only)

Most of the companies listed below deal only with eye care professionals, retail stores, or optical dispensaries, not with individual customers. These are starred (★). However, manufacturers and distributors are generally willing to mail information about their products to anyone who requests it.

- ★ **Bollé**, 9200 Cody, Overland Park, KS 66214 (800) 222-6553
<http://www.bolle.com/> (glacier glasses, sunglasses with side lenses, and wraparound sunglasses for children and adults)
- ★ **BPI (Brain Power, Inc.)**, 4470 S.W. 74th Ave., P.O. Box 559501, Miami, FL 33155
(800) 327-2250 <http://www.callbpi.com/> (optical dyes, equipment for dyeing lenses, and other supplies for optical laboratories)
- ★ **Corning Incorporated Ophthalmic**, HP-AB-02 Corning, NY 14831
(800) 821-2020 <http://www.corning.com/ophthalmic/>
(Corning Glare Control lenses)
- ★ **Crystal Reflections Int., Inc.**, 170 N. La Canada, Ste. 80, Green Valley, AZ 85614
<http://www.crystalreflection.com/> (custom tinted contact lenses)
- ★ **Franel Optical Supply Co.**, P.O. Box 96, Apopka, FL <http://www.franeloptical.com/>
(800) 327-2070 (side shields, clip-on lenses, and many other products)
- Hidalgo, Inc.**, 45 La Buena Vista, Wimberley, TX 78676 (800) 786-2021
(builds sunglasses to buyer's specifications – ask for their free catalog)
- ★ **Kontur Kontac Lens Co.**, 200 S. Garrard Blvd., Richmond, CA 94801
(800) 227-1320 (custom tinted soft contact lenses)
- NoIR Medical Technologies**, Medical Product Division, 6155 Pontiac Trail,
P.O. Box 159, South Lyon, MI 48178 <http://www.noir-medical.com/>
(800) 521-9746 (UV Shields and other tinted polycarbonate lenses)
- ★ **Orlux Distribution, Inc.**, 5440 McConnell Ave., Los Angeles, CA 90066
(800) 348-0388 (Vuarnet PX-5000, Baby-net sunglasses, also glacier glasses)
- REI (Recreational Equipment, Inc.)**, 1700 45th St. E., Sumner, WA 98390
(800) 426-4840 <http://www.rei.com/> (ski glasses & wraparound sunglasses)
- ★ **Specialty Tints**, 2525 Nabal, Escondido, CA 92025 (800) 748-5500
John Gibson, owner (custom tinted contact lenses)
- ★ **Skyline Northwest Corporation**, 0224 S.W. Hamilton St., Portland, OR 97201
(800) 547-8664 <http://www.skylinenw.com/> (for Barracuda Sports Products, including several models of swimming goggles that can be fitted with custom tinted Rx lenses)
- ★ **Yorktowne Optical Co.** (800) 233-1990 <http://www.yorktowneoptical.com>
(plastic “instant sunglasses” that slip behind frames)
- ★ **Zeiss Optical, Inc.**, 13017 North Kingston Ave., Chester, VA 23836
(800) 338-2984 <http://www.zeiss.com/> (custom-made sunglasses)



Information about services and products in this publication is for your consideration only and does not constitute a recommendation.



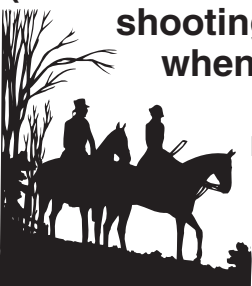
Sports Experiences of Networkers

To understand what's possible for persons with achromatopsia in terms of sports participation, it's important to be aware of the wide range of individual differences among achromats with respect to visual functioning. There are complete and incomplete achromats; and, among both the "completes" and the "incompletes," there are varying degrees of light sensitivity and visual acuity. So we should not make comparisons or assume that, if one achromat can be active in a certain sport, then others can too. Nor can we assume that, simply because one achromat has not been able to participate in a certain sport, then others cannot do so.

Normally sighted persons differ greatly in specific visual skills, and so do achromats. Some persons are able to "track" a ball or other moving object or scan their surroundings better than others. Some have better eye-hand coordination. And people vary greatly in their aptitudes for specific sports.

Having good tinted lenses naturally enhances the ability of achromats to participate in outdoor sports. Some networkers wear "glacier glasses" that have been fitted with dark lenses and opaque side shields when engaging in outdoor sports. Some wear tinted "sport shields" over their regular tinted lenses or wraparound sunglasses in addition to tinted contacts. Some have used black electrical tape to block out light from the sides of sunglasses.

Many networkers report that, when possible, they plan outdoor activities (horseback riding, jogging, biking, shooting baskets, etc.) at dusk, when their vision is at its best.



Wrestling, soccer, and martial arts are popular sports among many children and teens in the network. Strongly motivated

athletes find ways to participate in various team sports. Many networkers participate in special sports programs designed for persons with disabilities. Some of these programs are best known for serving the needs of persons with mobility impairments, but usually those who supervise these programs are trained to assist participants with any type of disability. Some adaptive sports programs have been set up strictly for the blind and visually impaired – for example, the United States Association of Blind Athletes (USABA), 33 N. Institute St., Colorado Springs, CO 80903 <<http://www.usaba.org/>>.



Not many partially sighted persons turn to these programs, either because they are unaware of them or because they don't consider themselves to be "disabled" or to have the same kinds of special needs that blind people have.

Some networkers highly recommend an organization called Disabled Sports/USA, which offers a wide range of outdoor sports experiences – skiing, canoeing, river rafting, kayaking, etc. – and which provides free instruction, free rental equipment, and other free or inexpensive accommodations. Participation in such programs usually involves explaining to those in charge of the activities how our needs differ from the needs of blind or mobility impaired participants. For info: Disabled Sports/USA, 451 Hungerford Dr., Suite 100, Rockville, MD 20850, <http://www.dsusa.org/> phone: 301-217-0960.

Horseback riding, bowling, and swimming are popular with many networkers. Some restrict their swimming to indoor pools (such as YMCA pools), some swim outdoors in the evening, and some swim outdoors in the daytime, wearing swim goggles that have been fitted with dark custom tinted lenses.

Scientific Studies of Achromatopsia

Color vision defects have long interested researchers. However, relatively few studies conducted by scientists have pertained to total colorblindness. A history of achromatopsia research is included in *Night Vision*, published by Cambridge University Press in 1990. One of the editors of this substantial book about matters pertaining to rod vision is Dr. Knut Nordby, a distinguished vision scientist who himself has complete achromatopsia. He has participated in numerous scientific studies of achromatopsia, sometimes as researcher and sometimes as subject. Readers who wish to learn more about achromatopsia research may want to obtain a copy of this book through an inter-library loan, as it is expensive. In chapter 8 of *Night Vision*, entitled "Vision in a complete achromat: a personal account," Dr. Nordby tells, both in personal and scientific terms, what it is like to have achromatopsia. See p. 151 for information on how to obtain a copy of this chapter.

When lay persons peruse scientific literature, it is easy for them to feel overwhelmed by the technical jargon. They have to struggle to find information that is meaningful to them. Some networkers have turned to medical libraries, hoping to find information about this rare vision disorder. One woman with complete achromatopsia told of her excitement when she was finally able to locate even a few paragraphs about her eye condition in an ophthalmology textbook. A network dad whose two daughters both have achromatopsia reported that he was able to move out of a period of deep sadness about his daughters' vision impairment by choosing to learn all that he could about this condition through library research and consulting vision specialists.

The question of which genes may be

abnormal in rod monochromats is one which scientists have been working to answer at research centers in various parts of the world. Presumably, the genes involved are ones which determine the way cone photoreceptors are made and/or the way in which they function. Recent studies have linked rod monochromacy to specific areas on Chromosome 2 and Chromosome 8 in many families, but this is not true for all families who have been studied (as of Sept., 2000). See the references listed on page 140 and the information about genetic research studies on pp. 118-120.

Following is a personal account by network facilitator, Frances Futterman, regarding her experience as a subject in a research study:

"In the 1970's I was the primary subject in a scientific study of rod vision. Rods are the photoreceptors which we achromats rely on, since we do not have cone photoreceptors working for us. It was presumed that the ideal way to learn about rod vision would be to study the vision of a complete rod monochromat. Tests showed me to be qualified, having normal rod vision and no cone vision. This began as a short term study by a vision scientist at the University of California at Berkeley. However, after I revealed to this researcher a certain very remarkable manifestation of my vision disorder, she obtained a grant in order to conduct further studies of my vision at Stanford University. What I demonstrated to her was my ability to 'see' after closing my eyes in levels of illumination in which I could not see anything except blinding light when my eyes were open."

To read more about this phenomenal ability of the rods to provide some persons with achromatopsia with "after images" useful for visual orientation with the eyes closed, see page 36.

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Achromatopsia Research at U.C. Berkeley – 1992

Following is a brief summary of findings in a research study of achromatopsia at the University of California School of Optometry at Berkeley, led by Dr. Gunilla Haegerstrom-Portnoy (from an unpublished report – Oct., 1992).

Variability in expression: Laboratory tests of achromats' color vision show:

1. Great variability in number and sensitivity of residual cones among subjects
2. Variability within the same family having more than one affected member
3. Variability between the two eyes of the same person
4. Variability in the same eye of a single individual, depending on where the eye test is located and depending on the light level used for testing

Tests showing residual cone function in patients previously presumed to be total rod monochromats: Spectral sensitivity tests show 25% have cones. Color matching tests show 36% have cones. Aversion index tests show 50% have cones. All measures show 60% have cones. Some achromats show evidence of having red cones, some have green cones, and some have both (most common). There is no correlation between the sensitivity of these residual cones and visual acuity. However, the more cones, the less photophobic the affected individual is.

Development of visual function in infants and children: This study includes ongoing efforts to use objective methods and behavioral methods developed for infants to assess visual acuity and contrast sensitivity development, as a function of age, to allow predictions of future visual capabilities.

Genetic Research Studies of Rod Monochromacy

A Chronological Summary

1992 – Researchers at Baylor University College of Medicine in Houston published a report linking rod monochromacy to Chromosome 14. However, soon after the publication of that report, the accuracy of its researchers' conclusions was questioned, because this study had focused on only one person – a woman who had several inherited physical defects in addition to her vision disorder, due to highly unusual genetic factors.

1997 – A research group at the University of Iowa Medical Center, led by Dr. Val Sheffield, conducted a genome-wide search for linkage of rod monochromacy, using blood samples from an inbred Jewish kindred from Iran which included a number of family members with rod monochromacy. (The term “kindred” refers to a large extended family.) The analysis presented by this research group established a significant linkage to Chromosome 2.

Spring, 1998 – Researchers at the University of Tuebingen Medical School in Germany announced that they had discovered mutations in a gene responsible for rod monochromacy. Using established genetic techniques as well as innovative molecular genetic techniques, they were able to link the locus of this disorder to a specific gene on Chromosome 2 for a number of the subjects in their study. Their attention had been directed to this locus by the 1997 findings of the University of Iowa Medical Center group. The Tuebingen researchers had recruited 8 families from the United States, Germany, Italy, and Norway. In 5 of the 8 families, the researchers were able to refine the position of the locus for rod monochromacy to a small region on Chromosome 2. The other 3 families did not show linkage to Chromosome 2. These results were published in the July, 1998 issue of *Nature Genetics* and in the August 1998 issue of *Genomics*. In a report to our network, Dr. Ted Sharpe of the Tuebingen group said that there was still much work to be done in order to locate the gene or genes causing rod monochromacy in other families. He said that it should not be surprising that more than one gene may be responsible for causing rod monochromacy, since the cone photoreceptors of the retina are very complex structures and a mutation in any one of a number of genes may cause a structural or functional defect.

Later in 1998 – Dr. Ted Sharpe reported the following: “Several research groups in different parts of the world, including ours at Tuebingen, are finding evidence that, in some families, rod monochromacy is associated with Chromosome 8. Families affected by rod monochromacy who originate from Northern Europe (Denmark, Sweden, Norway, England, Germany) tend to have the Chromosome 2 mutation, but most of the affected Italian families in our study do not. Also, there is evidence that at least one other chromosome (besides Chromosomes 2 and 8) is involved, but further studies are needed before we can know for sure.”

June, 2000 – Dr. Sharpe reported, “We have now discovered a second locus for rod monochromacy. We have found mutations causing rod monochromacy in a gene on Chromosome 8. The identification of this second gene location is a major breakthrough. We accomplished this here in Tuebingen, and a research group at Johns Hopkins Medical Center in Baltimore has also independently identified the same location. Their findings, which are based on blood samples from Pingelap and Denmark, are being published in *Nature Genetics* this summer, and our

findings are being published in *Human Molecular Genetics* this summer as well. In our study we had blood samples from Pingelap, Italy, Germany, and the United States. We know that there is another locus to be found, because some families in our study do not link to either Chromosome 2 or Chromosome 8.”

Concerning the possible results of genetic research

Dr. Ted Sharpe states, “There are several benefits that can result from our research to locate the genes that cause rod monochromacy. The first one is a long range benefit. Before any work on gene therapies can begin, the molecular genetic basis for the disorder must be identified. We have already identified 2 genes. We know there is at least one other gene to be found. This requires linkage studies and an exact phenotyping and genotyping. (*Editor’s note: phenotyping refers to a patient’s visual symptoms, and genotyping refers to the exact location of the genetic defect causing the patient’s symptoms.*) Confusion would result from grouping together individuals who seem to have the same visual symptoms but who, in fact, have different genotypes. Accurate diagnosis is of great importance. The different phenotypes may have very different genotypes. In short, before any significant advance in developing gene therapies for rod monochromacy can be made, the basic research work has to be done.

“The second benefit may be immediate. In the affected members of a family who contribute blood samples (i.e. members who have rod monochromacy), we begin by doing a gene mutational screening analysis to determine if they have a mutation occurring in one of the positions we have identified in the gene on Chromosome 2 or the gene on Chromosome 8. If a mutation is not found at one of these positions, their entire gene (at each of the 2 locations) will be sequenced to determine if any (as of yet unidentified) mutation is present. This may help us discover new mutations in the gene. If the mutational screening does reveal a mutation in the gene on Chromosome 2 or Chromosome 8 in the affected family members, the mutational screening can be extended to non-affected siblings, and they can learn whether they are carriers of the gene. If no mutation in the gene on Chromosome 2 or Chromosome 8 is found for the affected family members, we will know that the molecular genetic cause lies elsewhere. Our next move will be to perform a linkage analysis. This involves having blood or DNA samples from several families which have 2 or more instances of the disorder among siblings.

“The third benefit is another long range benefit. The more information we can obtain about the molecular genetics and pathophysiology of disorders such as rod monochromacy, the better will be our chances of finding gene therapies not only for rod monochromacy but also for other inherited retinal disorders, such as retinitis pigmentosa and Stargardt’s disease, conditions which affect many people and which, unlike achromatopsia, are progressive retinal degenerations.”

Another benefit would be that of simplifying diagnosis. Although molecular genetic analysis of blood samples or DNA samples is an expensive procedure and one which cannot be obtained in many laboratories, it would nevertheless be more straightforward and reliable than what many networkers have had to go through in their efforts to obtain accurate diagnosis.

It is important for networkers to understand that any therapies which may eventually result from genetic discoveries will take a long time to develop. Gene-based therapies will not become a reality in the near future. Each new

discovery is important, but a “cure for achromatopsia” is not “right around the corner,” as news stories about research findings often lead people to believe. The kind of research and experimentation involved is highly complex.

Many concerned individuals – networkers, professionals, and others – have expressed various misgivings with regard to genetic research. Following are comments from some members of the Achromatopsia Network on this subject:

From the husband of a woman with achromatopsia: “My wife and her brothers have chosen not to participate in genetic studies because of ethical and political concerns about genetic research in general. Among the people we know, there is an ongoing debate about genetic engineering, gene manipulation, etc. While there are cases in which the results of genetic research can help to heal certain illnesses, I suspect that one of the motivations of genetic research is to ‘enhance’ the human race, to create ‘better’ or ‘healthier’ human beings. As long as there is no real debate going on about the ethical implications of such research, I think we should be careful not to embrace some ‘exciting new procedure’ without thinking about the possible outcomes.”

A network dad, who is himself a scientist, wrote: “Considering the possibility that the time may come when the genetics of achromatopsia will be completely understood, can we then expect that some unborn babies will be aborted after DNA analysis has indicated that they have achromatopsia? Also, we will have to look closely at who will eventually be responsible for the application of the findings being made by researchers. In most cases, we scientists lose control over how new knowledge is applied.”

Regarding networkers’ concerns about the possibility that discovering the genes for rod monochromacy will lead to the abortion of fetuses found to have achromatopsia, Dr. Sharpe responded: “If someone is determined to find out whether a fetus has inherited defective genes from parents known or suspected to be carriers of a defective gene associated with rod monochromacy, this would be possible to do. However, the mutation screening for a defect on the genes we have discovered (i.e., the locations on Chromosome 2 and Chromosome 8) is not a routine screening. No regular clinic or laboratory would be able to do it. A private laboratory or university laboratory might take it on, but the procedure is very time-consuming and expensive. Of course, it is possible for this analysis to be performed here in Tuebingen, but our group has decided that we will refuse to perform such an analysis, if it is for the purpose of providing information that might lead to the abortion of a fetus. None of us working on this project considers rod monochromacy to be grounds for justifying abortion. The long range purpose of our work is not to prevent the birth of rod monochromats but rather to develop gene therapies that can be used to correct defects in the genetic code. To repair, not to destroy. These issues are, of course, also controversial. Knowledge brings responsibilities and also the danger of the abuse of that knowledge. So the answer to your question is: yes, theoretically, a fetal analysis is possible, but it would be very difficult and very expensive to have done, and we here at the Tuebingen facility would not cooperate with such a procedure, thus making it even more difficult for anyone desiring an analysis for this purpose to have it done.”

(This report by Frances Futterman was published in the Achromatopsia Network Journal in September, 2000.)

Disclosure of Visual Impairment

Like many other partially sighted persons, achromats have what can be called a “hidden disability,” because in many situations, the fact that they have seriously impaired vision is not evident to other people. Persons with hidden disabilities also include those with learning disorders, impaired hearing, or serious medical conditions which have no visible manifestations.

When one has a hidden disability, one has to make decisions about when and when not to disclose information about the disability, how to do so, and how to deal with people’s reactions to this disclosure.

Sometimes disclosure happens whether or not one chooses to disclose. Someone wearing red or orange lenses, very dark lenses, dark lenses indoors, or frames with opaque side shields may be easily recognized as being visually impaired. Also, getting close to reading matter or walking cautiously when outdoors in full sunlight can indicate poor vision. But in settings where lighting is favorable, most achromats find that their vision impairment can remain undisclosed until they need – or choose – to disclose it.

In a classroom or workplace where activities focus on reading, writing or other visually oriented tasks, someone using bioptic lenses, a magnifier, or special technological aids is easily recognized as being visually impaired. Yet the same person participating in activities involving communication skills (discussion groups, social events, etc.) may not be recognized as someone with vision problems.

Generally, the less time one spends in group situations that require color discrimination, reading and writing, or being in bright light, the less one has to stay ever mindful of being visually impaired and the less one has to be

concerned with the matter of disclosure. But, for most networkers, disclosure is an ongoing issue because of the active and integrated lifestyles they lead. They find that daily life offers ample opportunities to practice the art of disclosure. At work, at school, in dating, in job interviews, and in other situations, they make choices about when to disclose and how much to disclose about their vision. Also, there are persons they encounter from day to day who must sometimes be approached for assistance, such as the sales person, the librarian, the bus driver, the person at the fast food counter. Often achromats find they must decide whether to maintain their privacy, not ask for help or special accommodations, and accept the consequences of their decision not to disclose – or whether to explain to a stranger about their vision problem.

There are various ways to disclose one’s vision impairment. Simply saying “I am visually impaired” or “I don’t see well” can suffice in many situations. If assistance is needed, then it’s good to combine the statement about one’s vision with a statement about what specific help is needed – e.g., “I don’t see well; can you help me find a certain product?” or “I am visually impaired; would you please tell me the colors of these items?” Some networkers choose never to mention their vision when asking for some kind of help.

Many networkers report that they have chosen to “pass” as normally sighted in various situations. Sometimes these decisions have resulted in tension and unforeseen problems. However, some networkers report that their decision to “pass” has felt very liberating, at least in some situations. The temptation to “pass” is strongest among the young. Adults find that disclosure tends to become easier.

Accessing Services for the Visually Impaired

Many organizations and agencies that serve the vision impaired have names which refer only to blindness – for example, the U.S. Association for Blind Athletes, the American Foundation for the Blind, state rehab agencies for the blind, etc. In recent years, some organizations and agencies have modified their names to reflect the fact that many who use their services are not blind but partially sighted. In fact, most vision impaired persons are not blind. Some organizations formerly called “for the blind” are now “for the blind and visually impaired.” There will surely be more such changes in the future, more accurately representing all of the clients and making it easier for partially sighted persons (such as achromats) to identify with and access these services.

Because achromats, in favorable lighting, experience such a high level of visual functioning, they tend to find it especially hard to turn to services “for the blind.” Many networkers, however, have learned to make use of all of the resources that are available to them in order to get the help they need. Very often, though, partially sighted persons simply do not know what services are available to them.

In accessing these resources, one sooner or later has to deal with the term “legal blindness,” a term used by the IRS and others to determine a person’s eligibility for various benefits and services. There are two definitions of legal blindness – one having to do with lack of central vision and the other having to do with loss of peripheral vision. Someone is considered legally blind if vision in the better eye is 20/200 or less. This is the first definition, the one having to do with lack of central vision. Most achromats have a visual acuity of 20/200 or less and so are qualified for

all services, benefits, and accommodations available to the legally blind.

Even when clinicians obtain a visual acuity reading better than 20/200 for a patient by using optimal lighting in the examining room and on vision charts, they are usually glad to certify the patient’s acuity as 20/200 on application forms to obtain needed services, such as transit discount applications. Experienced clinicians realize that the carefully controlled lighting which permits an achromat to have a chart reading better than 20/200 in an examining room in no way resembles the environments in which this patient must live, work, travel, or pursue an education. To really understand the kinds of vision problems achromats have, it would be necessary to do the testing in brightly illuminated indoor and outdoor spaces.

Some persons with achromatopsia have had little or no experience with using services for the blind and visually impaired. Sometimes this is on principle, and sometimes it is because they simply haven’t known they were entitled to such services. Those who have grown up with special services in school tend to be more aware of the services that exist for visually impaired persons and to be more comfortable in using such services, having already had the experience of being identified as “legally blind” or “visually handicapped” – i.e., the terms which go along with being eligible for special services.

Many networkers have had vocational rehabilitation assistance in college or when getting established in a vocation.

Many networkers use the Talking Books available through regional libraries for the blind and visually impaired.

On the following page is a list of resources networkers should know about. There are also many services for the

visually impaired available through state, regional, and local agencies. These services vary from one region to another. They include transit discount programs, low vision optometric services, telephone accommodations

(big button phones, free directory assistance, etc.), and much more. Check with the agencies near you for information about services, discounts, special programs, and accommodations for the vision impaired.

Vision Community Services, 23A Elm Street, Watertown, MA 02472 publishes a comprehensive list of resources for low vision persons (in large print and regular print). <http://www.mablind.org/Resourcelist.htm>

National Association for Parents of the Visually Impaired, P.O. Box 317, Watertown, MA 02471, (1-800-562-6265) offers support, information, referrals, and a newsletter. Ask about regional representatives. <http://www.spedex.com/napvi/>

Council of Citizens with Low Vision International, 1155 15th St. NW, Suite 1004, Washington, DC 20005, (1-800-733-2258), a division of the American Council for the Blind (information, newsletters, meetings, advocacy). <http://www.cclvi.org/>

Vision Enhancement, a journal for persons with low vision, published by Vision Enhancement World Wide. <http://www.visionww.org/>

For information and literature regarding Social Security benefits for persons with vision disabilities, (1-800-772-1213). <http://www.ssa.gov/disability/>

The American Foundation for the Blind has a toll-free number (1-800-232-5463) which operates from 9 A.M. to 4 P.M., Eastern Time. Phone for information about agencies in your state – special education, independent living centers, low vision centers, vocational rehabilitation services, and more. <http://www.afb.org/>

National Library Service for the Blind and Physically Handicapped, The Library of Congress, 1291 Taylor St. NW, Washington, DC 20011, (1-800-424-9100) offers information about the Talking Books program. <http://lcweb.loc.gov/nls/>

The Low Vision Gateway, <http://www.lowvision.org/>, is a starting point for accessing information about low vision resources on the Internet.

Prevent Blindness publishes a directory, “Coping with Sight Loss in Northern California,” which includes information for sight-impaired persons anywhere in the U. S., not just in Northern California. To request a free copy, write to Prevent Blindness at 4200 California St., San Francisco, CA 94118. <http://www.eyefinfo.org/> for “Coping with Sight Loss” in Adobe Acrobat PDF format.

National Association for the Visually Handicapped has a San Francisco office that serves Western states, 3201 Balboa St. San Francisco, CA 94121 (1-415-221-3201) and a N.Y. office serving all other states, 22 W. 21st St., New York, NY 10010 (1-212-889-3141). NAVH offers information and referral about resources for the partially sighted, focusing on large print materials. <http://www.navh.org/>

National Federation of the Blind, 1800 Johnson St., Baltimore, MD 21230, 1-410-659-9314, a consumer and advocacy organization (publications, meetings, information lines, and more). <http://www.nfb.org/>

LEGAL BLINDNESS: There are two definitions of legal blindness. One has to do with impaired central vision, and the other with loss of peripheral vision. A person is “legally blind” if vision in the better eye is 20/200 or less. This definition has to do with central vision. Most achromats have a visual acuity of 20/200 or less.

About Not Being Blind

Many of the problems with which achromats must cope have to do with the fact that their degree of sightedness cannot be easily explained. “Neither fish nor fowl” is a phrase commonly used by partially sighted persons, referring to their being neither blind nor fully sighted. This phrase is especially applicable to achromats, since their vision can change so significantly as the factors of illumination in their surroundings change.

Early in life achromats learn that they do not have as much vision as other people have, and yet they know that they most certainly are not blind. Under favorable lighting conditions their vision is, in fact, quite good. However, when lighting conditions are unfavorable, their vision can be extremely limited.

It is often said that achromats are “blinded by the light,” referring to the saturation of the rods (the rod-shaped photoreceptors) in their retinas (see page 1), causing them to have markedly decreased vision when the light level is higher than their eyes can tolerate. Being in full sunlight without having dark lenses on is as close as they come to experiencing blindness in everyday life. However, even under such conditions, many achromats who had to manage without tinted lenses as they were growing up can attest to the fact that vision is still possible in bright light by using the “after-images” that are ever so briefly visible to them when their eyes close during rapid blinking (see pages 35 and 36). (There is also, of course, the experience of being without sight which achromats, like all sighted people, have whenever they are in total – or almost total – darkness.)

If tinted lenses had never been invented, then we achromats would be forced to adapt as fully as possible to

our lowest level of visual functioning – i.e., the near-blindness we experience outdoors in the daytime. Our ability to lead relatively “normal” lives following daytime schedules in terms of work, school, and other activities (something that many of us take for granted) is possible because we do have tinted lenses which permit us to open our eyes and see outdoors in the daytime. If there were no tinted lenses, we would have to adapt in more radical ways to the “nearly blind” end of our range of sightedness.

It should be pointed out that, even among persons for whom the term “blind” is truly applicable, there are various degrees of visual functioning. Depending on the eye condition that has caused the visual impairment, some “blind” people have light perception, some can perceive movement or color, and some can visually identify objects with sufficient magnification. Very few people have no sight at all.

In general, the term “blind” is most appropriate when referring to someone whose perception is primarily through the other senses, rather than through the sense of sight. Although achromats tend to make better use of their other senses than normally sighted persons, they function primarily through their sense of sight. In fact, achromats put a tremendous amount of effort into using their vision. Many are visually oriented to a remarkable degree. Extraordinary sensitivity to visual detail in their surroundings is frequently reported by networkers, and many of them are talented in the visual arts.

Not surprisingly then, it can be quite difficult for an achromat to accept the label “legally blind” – a term which is so important in accessing services and benefits for the visually impaired. It can

be unsettling for parents of children with achromatopsia to discover that their children get classified as “legally blind” by eye care professionals who certify patients for special services. Young persons with achromatopsia planning to enter college can find it disturbing to have to apply to organizations or agencies “for the blind” in order to obtain financial assistance, adaptive aids, etc., to meet their special needs. Sooner or later, most networkers have learned to live with these realities, as they discover that other partially sighted persons use these services and as they obtain the benefits to which they are entitled.

Networkers vary in their attitudes regarding this subject. Some easily embrace the term “blind,” acknowledging that this is simply the way things are and that “the system” is not likely to change any time soon. Some say that they feel fine about identifying with all visually impaired persons, blind and partially sighted alike. Acceptance is much easier for those who have had considerable experience with school, camp, or other programs integrating the blind and the partially sighted.

Some networkers have been influenced by a trend of thought proposing that the word “blindness” be re-defined to mean “significant visual impairment,” rather than meaning “no vision,” thus encompassing the partially sighted and persons without sight.

Public recognition of the blind and their special needs developed long before the recognition of the partially sighted and their special needs, even though there have always been far more partially sighted persons in the world than blind persons. Schools for the blind and other services for the blind were established many years before special educational or other programs

for the partially sighted came into existence. Partly this has been true because blindness has always been so much easier to recognize and identify. Also it is easier to imagine what the needs of the blind are. By contrast, partial sight is, in many cases, a “hidden disability,” and the realities and needs connected with partial sightedness vary so greatly from one person to another.

Many institutions for the blind were established during the nineteenth century. In the mid-twentieth century, as special education programs were being established in the public schools of the U.S. and elsewhere, the term “partially sighted” came into use. Some of these programs brought blind and partially sighted students together, and others provided separate classes (or different resource teachers) for blind students and for partially sighted students.

After the 1950’s, the term “low vision” gained widespread acceptance, referring to all partially sighted persons whose vision impairment could not be corrected with ordinary prescription lenses. Low vision clinics were established throughout the U.S., offering an ever growing variety of optical aids and other devices to help these patients to maximize whatever amount of vision they had.

Many professionals who serve vision impaired individuals refer to the established system of organizations and agencies serving the visually impaired as “the blindness system.” There is the American Printing House for the Blind, the American Foundation for the Blind, regional “lighthouses” for the blind, schools for the blind, state rehabilitation agencies for the blind, and so forth. Because most of these organizations and agencies for the blind were established a long time ago – long before any programs designed for the partially

sighted were created – they continue to be the recipients of funding for almost all programs and services for the visually impaired. Thus, partially sighted persons who choose to make use of services, products, and programs which are available to help them must, in most cases, apply to organizations and agencies for the blind.

In recent decades the names of some of these organizations and agencies have been modified in recognition of the fact that they serve not only the blind but also the partially sighted. For example, more than one regional “Lighthouse for the Blind” has been re-named “Lighthouse for the Blind and Visually Impaired.” However, most of these organizations and agencies have not changed their names to include “the visually impaired.” Partly this resistance to change has to do with the realities involved in competing for funding. In some cases, however, it reflects an attitude of inflexibility and failure to recognize and be sensitive to the needs of the partially sighted.

In many respects, the needs of the partially sighted are similar to the needs of the blind; but, in other ways, their needs are quite different. Very few programs have ever been established specifically to meet the special needs of the partially sighted.

In many states, network parents seeking special education and other services must apply to a school for the blind or a school for the blind and the deaf. Adults or teens with achromatopsia seeking assistance with college, job training, or employment must approach a state agency for the blind. Camps for visually impaired children and teens are usually accessible through a lighthouse for the blind or other agency for the blind.

So, while it continues to be true that

partially sighted people outnumber the blind by far, it is also true that we live in an age of ever increasing opportunities for the visually impaired, and partially sighted persons who wish to take advantage of these opportunities must come to terms with the word “blind.”

Generally the more sight one has, the harder it is to adjust to the idea of utilizing “services for the blind.” Many networkers are familiar with the terms “high partial” and “low partial.” The term “low partial” is sometimes used when referring to a partially sighted person whose degree of vision impairment is significant enough to make it relatively easy for that person to identify with the needs and adaptive methods of the blind. By contrast, the “high partials” are partially sighted persons who do not so easily identify with the blind and who have a considerable amount of usable vision – so much so that they often “pass” as normally sighted, at least in some situations. “High partials” include persons with achromatopsia, persons with albinism, and persons who are experiencing an early stage of a progressive eye condition.

There are very few eye conditions causing severe vision impairment which are non-progressive and present from infancy throughout life. Rod monochromacy (the form of achromatopsia which affects most people in our network) is one such condition. Albinism is another. Until recent years, not much was known about the visual experiences of such persons – i.e., those who live their entire lives as partially sighted. By contrast, the more common eye conditions which cause severe vision impairment have a period of onset at some point in a person’s life and are progressive. Typically, the partially sighted persons who are affected by these more common eye conditions need to come

to terms with a prognosis of eventually becoming blind or nearly blind. Consequently, it is important for these persons to (1) adjust to this reality and (2) learn methods and techniques of managing without sight. Professionals who work in services for the vision impaired are well acquainted with the understandable tendency of many of these people to be in denial about their degree of vision loss and their prognosis and to resist learning the skills that they need. Unfortunately, many of these professionals, due to their lack of knowledge about achromatopsia, presume that the needs of achromats are the same as the needs of other visually impaired persons using their services, and they interpret any resistance on the part of achromats to learning “blind skills” as indicative of being “in denial.”

Often networkers have reported that, when approaching certain organizations and agencies, they have been treated as if they were blind. Some parents have mentioned special education staff who wanted their children to use the same methods and tools as blind children. The most extreme experiences have involved the use of blindfolds.

Some network parents have been pressured to have their children learn Braille. Braille is a wonderful option for students who really need to read by touch or who are likely to need it in the future, due to progressive vision loss, but it is not appropriate for students who have been accurately diagnosed with achromatopsia. Achromatopsia is a stationary (non-progressive) vision disorder.

As mentioned on page 122, it sometimes happens that an individual with achromatopsia whose visual acuity has previously been recorded as 20/200 (“legally blind”) goes in for a vision exam, and the vision care specialist obtains a slightly improved visual

acuity after carefully adjusting the lighting on the vision chart as well as in the examining room, creating “twilight” illumination, which is ideal for achromats. The vision specialist may consider this an occasion for rejoicing, and the “improved visual acuity” (typically 20/160, instead of the previous acuity of 20/200) can look impressive on the patient’s records. But the patient still has the same vision that he or she has always had out in “the real world,” where ideal lighting conditions for achromats are seldom encountered. Since persons whose visual acuity is better than 20/200 do not qualify for certain services and benefits available to the “legally blind” (except in the case of eye conditions which cause serious loss of peripheral vision), many achromats who have had the kind of experience described above have asked to have their “old” visual acuity (20/200) recorded on any documents certifying their vision disability. Vision care professionals are generally glad to help out in this way.

Some adult achromats have chosen to affiliate with the Council of Citizens with Low Vision International, a division of the American Council for the Blind. Even though, as “high partials,” these networkers have found themselves to be considerably more sighted than almost all of the other participants, they have sent in very positive reports about their experiences at the gatherings of this organization. They have reported feeling profound relief at being present at events where having impaired vision in the norm and where all manner of special accommodations are readily available to them. The American Federation of the Blind, another consumer and advocacy organization, also has members with low vision but does not have a special division for them. Contact information for these and other organizations is provided on page 123.

Other Vision Problems Which Can Affect Achromats

Most of the information in this book pertains to the three primary vision problems which are manifestations of achromatopsia. In order of significance, these are: hypersensitivity to light, poor visual acuity, and the absence of color vision. However, there are other vision problems which can affect achromats and which need to be understood.

Normal age-related vision problems:

All parts of our body change as time goes by, and our eyes are no exception. Considering that our eyes are among the most active – and the most exposed – parts of our body, it is reasonable to expect that they will go through changes as we age.

During the first 30 to 40 years of our life, our eyes work hard to see everything they can. Changing focus from near-point activities (such as reading, writing, and arts and crafts activities) to distant viewing and then back again to near-point is relatively easy when we are young. However, as we get older, the lenses of our eyes become less flexible, so that the ability of the lenses to become rounded, as they must for looking at things up close, diminishes. Eventually most people begin to notice a blur when they are looking at something at very close range. This condition is called presbyopia (pronounced prez-bee-OH-pee-ah).

This change is experienced by most achromats, just as it is noticed by most other persons. Sometimes achromats and other vision impaired persons actually notice these changes at an earlier age – for example, in their 30's, instead of in their 40's. This is because (1) achromats, like many other vision

impaired persons have come to rely upon being able to focus up close and, therefore, will be quick to notice when close-up images no longer have the clarity they have become accustomed to, and (2) vision impaired persons cannot simply begin holding reading material farther away from their eyes in order to see it more clearly, as normally sighted persons can do. Normally sighted persons often joke about how it's not a matter of their vision getting worse around age 40 but a matter of their arms getting shorter.

Achromats experiencing these changes generally adapt by bringing reading material even closer than they have needed to do in the past, thus achieving additional magnification to compensate for the decrease in visual clarity at close range. Sooner or later, however, they resort to more frequent use of magnifiers and/or using stronger magnifiers. Many achromats who have previously made little or no use of recorded reading materials (such as the "Talking Books" that are available from libraries for the blind and visually impaired) develop greater appreciation for this option. Some begin to wear reading glasses or bifocals. Others turn to alternative approaches to vision care which have been used by many people to improve visual functions and the overall health of the eyes and the body.*

Looking at this problem in perspective can help one have a greater understanding of these normal age-related vision changes. In prehistoric times, life expectancy was only about 30 years. Back then, there were few near-point vision tasks other than preparing

* A good reference for those who would like to learn about the eyes and vision and about both traditional and alternative approaches to vision care is the book, *Healthy Eyes, Better Vision*, by Jeffrey Anshel, O.D. (published by The Body Press, a division of Price Stern Sloan, 1990), available through libraries and on the Web.

food and making weapons – neither of which represented highly detailed work for the eyes. Most visual requirements back then involved distance vision, which was necessary for hunting and for protecting oneself from danger. Even now, from birth through early childhood, human beings are basically farsighted, although they begin having to focus up close very early in life, due to the emphasis that is now placed on reading and writing. We live in an age in which near-point vision tasks, such as reading, writing, and computer work, constitute a major part of everyday activities involving our eyes.

Achromats, having only rod vision, are not able to experience the clear distance vision which cone vision makes possible for normally sighted persons (see page 1 for information about rods and cones). In order to achieve clarity in distance viewing, achromats must make use of optical aids. Near-point vision tasks are much easier for them.

Our eyes were made to last a lifetime, but the lifetime for most human beings now extends far beyond life expectancy back in prehistoric times – and even considerably beyond the life expectancy of people who lived a century ago.

Besides the hardening of the lenses of the eyes, there are other normal age-related changes which are experienced by many persons. These can include reduction in pupil size, decreased pigment in the iris, and slower adaptation of the eyes when moving from higher illumination to low illumination or vice versa. Middle-aged achromats experiencing these changes sometimes become very concerned when they begin to notice a reduction in their ability to see as well as they have been able to see in the past when stepping into darkened environments. They may find that they require a longer period of dark adaptation. And some middle-aged

networkers report having to do the unthinkable for achromats – actually resorting to using brighter light as they try to read small print or see other details more clearly.

As with all other challenges which life brings, achromats do whatever they need to do in order to adjust. But it can at least be reassuring to know that these very common age-related visual changes just described are natural and non-threatening to the eyes. However, it is important that persons experiencing these changes have regular eye exams in order to rule out the possibility of anything more serious that might be causing the vision changes.

Serious age-related vision problems:

There are a number of serious age-related eye diseases – such as macular degeneration, cataracts, and glaucoma – which, unlike presbyopia and other normal vision changes that occur in middle age, can be a real threat to one's vision. Many eye diseases can be treated, so regular ophthalmological exams are strongly recommended.

It is not known whether achromats are any more (or less) susceptible to age-related macular degeneration than the rest of the population. It has been speculated that achromats, because they tend to limit the amount of time their eyes are exposed to sunlight (through lifestyle choices and frequent use of tinted lenses that block UV rays) may actually be less inclined than other people to develop age-related macular degeneration. However, this is only speculation.

There are members of our network who have other serious eye conditions in addition to achromatopsia. Some networkers have developed age-related macular degeneration, and some have developed cataracts (these conditions are not caused by having achromatopsia). One member has had to cope with

the vision problems associated with Graves' disease (including increased sensitivity to light). Another has lived with double vision, as the result of a failed surgical procedure in childhood that had been done in an attempt to correct his strabismus (the tendency of one of his eyes to turn outward). Another has retinal damage that resulted from a stroke. And there are other examples. However, most achromats in our network have not reported having any serious vision problems other than those associated with achromatopsia.

Because achromatopsia is a stable eye condition, rather than a progressive eye condition, the manifestations of achromatopsia – hypersensitivity to light, colorblindness, and poor visual acuity – remain basically unchanged from infancy throughout life. The only known exception is that researchers have found that, for some persons who have the especially rare form of achromatopsia known as blue cone monochromacy, there is some deterioration of residual color vision and visual acuity at some point in adulthood, due to macular changes. But there has been no evidence of macular changes in persons with rod monochromacy, which is the most common form of achromatopsia, the form which affects most members of our network.

Refractive errors: Because the early signs and symptoms of achromatopsia – nystagmus (shaky eyes), aversion to light, and poor vision, especially in bright surroundings – make it obvious to parents that there is something seriously wrong with a child's eyes, children with achromatopsia usually get taken to doctors for eye exams very early in life. Consequently, these children tend to have Rx lenses prescribed for them very early in life – sometimes even in infancy. Infants and children with achromatopsia are found to have more significant

refractive errors – i.e., more significant hyperopia (farsightedness) or myopia (nearsightedness) and astigmatism – than other infants and children.

Vision care specialists differ in their beliefs with regard to the introduction of corrective lenses at an early age. Many vision care specialists subscribe to the theory that Rx lenses should be introduced very early in life. However, many eye doctors believe that it is best to let children's eyes develop naturally in the early years, postponing the consideration of Rx lenses until a child is closer to school age. Sometimes an eye doctor who had previously prescribed Rx lenses for a child with achromatopsia will later acknowledge that the Rx lenses were actually making very little difference in the degree of vision impairment experienced by the child (since the vision impairment caused by achromatopsia is so much greater than the effect of the refractive error which the Rx lenses corrected) and that having to wear spectacle frames fitted with Rx lenses was significantly limiting the child's options in terms of sunglasses. Appropriately tinted sunglasses with frames designed to provide glare protection for one's peripheral vision are the most important adaptive aids for achromats. Good sunglasses can make an enormous difference in terms of meeting the special visual needs of a child with achromatopsia.

There are adult achromats in our network who believe that they have benefited from the Rx lenses that were prescribed for them early in life, and there are other adult achromats who report being very grateful that they were not made to wear Rx lenses as children, stating that they are glad they did not develop a dependency on Rx lenses and that they have enjoyed being free to wear different kinds of wraparound sunglasses during their lives.

Rx lenses prescribed for achromats nearly always include a correction for **astigmatism**, along with their Rx for either farsightedness or nearsightedness. Astigmatism is a very common refractive error primarily caused by irregular curvature of the surface of the cornea. Many persons in the general population wear Rx lenses which include an astigmatism correction. Some achromats report that wearing lenses which correct their astigmatism has been helpful, and others report that their astigmatism correction has mattered very little to them or not at all.

More information about Rx lenses is included on page 18 and pages 111-113.

The nystagmus (shaky eyes) that is associated with achromatopsia is usually very noticeable in infants and young children, but it tends to diminish as the child matures. Nystagmus is more noticeable when there is a visual fatigue,

when the lighting is unfavorable, or when one is trying hard to see something. Nystagmus does not affect the vision of achromats.

Strabismus (the tendency of one eye to turn in or out) is common among achromats, especially in early childhood, when a child's eyes are struggling to work together. This problem, like nystagmus, usually diminishes as children mature. Sometimes doctors suggest surgery or other measures when a child's strabismus appears to be a serious problem. Some networkers report having had surgery to correct strabismus that was successful, and others report having had surgery for strabismus that was not successful or that resulted in other problems.

For further information about the vision problems that can affect achromats, be sure to see the list of reading references on page 140.

Types of Vision Care Specialists

An **OPHTHALMOLOGIST** is a medical doctor (M.D.) who has completed college, four years of medical school, a year of internship, and a minimum of three years of specialized residency training concerning diseases and surgery of the eye. Many ophthalmologists receive additional training in order to specialize in retinal diseases, pediatric ophthalmology, or some other area of specialization. Ophthalmologists test vision, diagnose and treat eye diseases and eye defects, prescribe lenses, prescribe medication, and perform surgery. Some ophthalmologists refer patients to optometrists for the prescribing of glasses and contact lenses.

An **OPTOMETRIST** has a degree in optometry (O.D.), which is awarded after college and then optometry school. An optometrist screens and diagnoses common eye problems, assesses the

efficiency and health of the eyes, and prescribes eyeglasses and contact lenses. Optometrists are trained to recognize signs of eye diseases, eye defects, and eye injuries; and, when these are detected, they make referrals to ophthalmologists. Optometrists are not physicians and do not have medical training. Some of them specialize in low vision, contact lenses, vision therapy, sports vision, or other areas.

An **OPTICIAN** is a technician who is trained to fill prescriptions for lenses written by ophthalmologists or optometrists. Opticians make eyeglasses, fit lenses into frames, and adjust frames to fit a person's face. In some states opticians are allowed to do the fitting of contact lenses. A training program (usually lasting two years) is required. Certification requirements vary a great deal from state to state.

Misdiagnosis, Uncertain Diagnosis, and Related Matters

There is far greater recognition of the term “achromatopsia” now than there was in the past, before our network began. Yet the word “achromatopsia” continues to be problematic and inadequate. Problems in connection with the use of this word have only multiplied with wider recognition and usage.

Strictly defined, “achromatopsia” means “without color vision,” and yet the fact of the matter is that persons who have achromatopsia live with vision problems that are far more challenging than the inability to perceive color.

Oliver Sacks’ 1997 book, *The Island of the Colorblind*, and the subsequent documentary film, “Island of the Colorblind,” about Dr. Sacks’ visit to the tiny atoll of Pingelap, where there is a high incidence of achromatopsia, introduced this rare vision disorder to many people around the world. (Actually the term which Dr. Sacks chose to use in his book and in the film was “achromatopia,” which is simply a variation of the term “achromatopsia.”)

As can be expected whenever a new or unfamiliar term becomes popularized, many people who began to use this term have used it incorrectly. Numerous people who have contacted me (in connection with the fact that I facilitate the Achromatopsia Network) after having come across Dr. Sacks’ book – but not having read it carefully – or after having seen only part of the film have had the impression that persons with achromatopsia are affected only by colorblindness.

Even before the book and the film were produced and distributed, I would often receive letters and e-mails from people who had learned about our network and who confused achromatopsia with the common type of color vision deficiency which affects about 7% of

males and about 1 in 200 females in the general population (this common color vision deficiency is often referred to as “colorblindness”). I have always made sure to include explanations about what it means to have achromatopsia in our network web pages and in all of the literature about our network I have distributed. Even so, these kinds of misconceptions abound.

The word “achromatopsia” has played a role in many situations which have involved misdiagnosis, tentative diagnosis, or lack of clear diagnosis on the part of doctors, as well as many cases of mistaken self-diagnosis made by visually impaired individuals.

It seems that some doctors use the term rather loosely. Many parents who have entered our network over the years have reported that their child’s eye doctor had given the diagnosis of achromatopsia after an ERG (electroretinogram) had been performed. Many of these children were infants when the ERG was done. In quite a few of these cases, when I would request further information about the child’s vision, I would learn that there were visual symptoms and/or other aspects of the situation which I recognized as being inconsistent with the diagnosis of achromatopsia. For example, sometimes I would learn that the child had abnormal rod vision, revealed either through the ERG results or through reports from the parents that the child did not show signs of having the substantially improved vision in lower levels of illumination which achromats are known to have. Or I would learn that, upon ophthalmological examination, the child’s retinas had shown signs of abnormality – unlike the retinas of achromats, which appear healthy and normal. So I began to realize that some eye doctors

were using the term “achromatopsia” in connection with any patient for whom an ERG had revealed that cone vision was either absent or very abnormal. But an ERG reading alone is not sufficient evidence on which to base such a diagnosis. Unfortunately, many doctors are not sufficiently knowledgeable about all of the factors which need to be considered when making a diagnosis of achromatopsia (see pages 14-15).

We have to make the best of the nomenclature that is in use, inadequate though it may be, and “achromatopsia” is the term that is widely accepted for designating the category of vision disorders which affect the members of our network – i.e., rod monochromacy and blue cone monochromacy.

To be absolutely accurate, one could refer to this category not just as achromatopsia but as congenital, inherited achromatopsia, thus differentiating it, beyond any doubt, from cases in which there has been loss of color vision due to some progressive eye disease.

Whenever someone interested in joining our network mentions that he or she has, at some point in their life, suffered a “loss” of color vision or an “onset” of the kinds of symptoms associated with achromatopsia, I know that this person does not have congenital, inherited achromatopsia, even though they may report that a doctor has suggested the diagnosis of achromatopsia. When the eye condition is achromatopsia, there is no period of “onset” or “vision loss,” because all of the symptoms have been present from infancy and are non-progressive.

I carefully screen prospective members of our network. The membership forms I send out ask people to provide information about symptoms, results of eye exams, the name of the eye care professional who provided the diagnosis, and the terms that were used by the

doctor when explaining the diagnosis.

Over the years, I have found that most of the people who have contacted me wanting to join the network have been persons affected by some eye condition other than achromatopsia. Most frequently, the eye condition has been either (1) some color vision deficiency that is not accompanied by poor visual acuity and hypersensitivity to light, such as achromats have, or (2) some progressive condition of the retina, such as progressive cone dystrophy or cone-rod dystrophy.

There have been a number of families who have joined our network because a child in the family was diagnosed with achromatopsia, then later reported that the child’s diagnosis had been changed. In some cases, certain other health or medical problems were detected, and it was recognized that the child’s visual symptoms, which had previously been considered symptoms of achromatopsia, were instead part of a rare medical syndrome the child had inherited. There are several rare syndromes which include achromatopsia-like symptoms (e.g., poor vision and aversion to light) along with other medical conditions. Children who have one of these rare medical syndromes have a form of cone dystrophy or cone-rod dystrophy instead of achromatopsia.

Few doctors are familiar with these rare syndromes. Eye doctors trained in retinal diseases or ocular genetics are more likely to be knowledgeable about these syndromes. I have been advised to take note whenever a parent reports that a child has a hearing disorder, a congenital cardiovascular problem, diabetes, or certain other physical abnormalities and is overweight. In such cases, I advise parents to consult a specialist to explore whether the child may have one of these rare medical syndromes.

Some Questions for the Eye Doctor

For many networkers, the road to getting an accurate diagnosis has been a long and complicated one. They have been confused – and sometimes misled – by some of the terms their eye doctors have used. One term which has frequently come up in reports from networkers is the term cone dystrophy. Some doctors think of achromatopsia as a stationary form of cone dystrophy, and so they will simply refer to the eye condition as cone dystrophy (not bothering to explain that they do not mean progressive cone dystrophy).

I asked Dr. Alex Levin about these terms. Dr. Levin, a staff ophthalmologist at The Hospital for Sick Children in Toronto and a specialist in both ocular genetics and pediatric ophthalmology, has served as a medical consultant for our network since its beginning.

Achromatopsia or Cone Dystrophy?

Question: Please explain about achromatopsia and cone dystrophy. What is the difference between them and what is cone-rod dystrophy?

Dr. Levin: The retina is the tissue which lines the inside of our eyeball. It acts like film in a camera, projecting pictures of the world around us to our brain. The retinal cells which begin the process of transforming light into images are called photoreceptors. There are two kinds of photoreceptors: rods and cones. Rods are more involved with vision in the dark, while cones give us color vision, clear central vision, and the ability to handle bright lights. Complete achromatopsia (also known as rod monochromatism) is the congenital absence of normal cone function. There are also congenital abnormalities which fall into the category of partial achromatopsia, in that not all of the cone functions are missing.

The term cone dystrophy is usually

applied to those inheritable disorders in which there is a progressive loss of only cone function (cone degeneration) over time. The cone-rod dystrophies are inheritable conditions in which both the cones and the rods undergo progressive degeneration, with the cones being relatively more affected than the rods. Whereas the congenital complete achromat is born with no normal cone function, the patient with progressive cone dystrophy or cone-rod dystrophy may, early in life, have cone function, which is then gradually lost as time goes by. In fact, after all cone function is lost in a patient with progressive cone dystrophy, the patient may eventually have medical test results and symptoms that appear to be very similar to those of patients with achromatopsia. The patient with cone-rod dystrophy will eventually experience symptoms of rod dysfunction as well, such as poor vision in darkness (night blindness) and reduced peripheral vision.

Congenital, inherited achromatopsia is not associated with a progressive decrease in cone function or decrease in vision. The problem is present at birth. Interestingly, both achromatopsia and progressive cone dystrophy may have a similar appearance on retinal examination. Therefore, at the initial consultation in examining a small child, it may be difficult to make the distinction between the two disorders. An electroretinogram (ERG), family history, observation over time, the presence or absence of nystagmus (shaky eyes, more often present in achromatopsia), and color vision testing can be helpful in distinguishing the non-progressive achromatopsia from the progressive cone dystrophy. It is the involvement of rod vision that distinguishes cone-rod dystrophy either early or late in that disease.

“Sun Staring”

Question: Some network parents have reported that their children with achromatopsia have at times, during infancy or early childhood, exhibited a strange visual behavior that is not consistent with the typical manifestations of achromatopsia. The children would sometimes open their eyes wide in bright sunlight outdoors – in some cases, even looking straight at the sun with open eyes. Some of these parents have described their children as seeming to be in a trance-like state on these occasions. As these children got older, they would no longer be seen to do this and would instead exhibit only the kinds of visual behavior which one might expect in someone with achromatopsia – e.g., avoidance of light, squinting, blinking, or closing of the eyes outdoors in the sunlight. What might explain this “sun staring”?

Dr. Levin: I have collaborated with Dr. Elise Héon, one of my colleagues here in the department of ocular genetics, regarding this question. We are not aware of any scientific discussions on this topic. We do know that, although cones develop toward the end of the first trimester of fetal development (long before the rods develop), retinal development continues for several years after birth. It is possible that persons with

achromatopsia have some abnormal cones in their retinas. Perhaps there is a delay in cone formation or maturation such that the subsequent aversion to light is delayed. The photophobia (aversion to light) which achromats have may be due to both the decreased number of cones in the retina and also the abnormal structure and function of these cones. If these theories are correct, then it may be that the child experiences what could be called an early “honeymoon period” in which the symptoms characteristic of achromatopsia are not as prominent, even though the retina is underdeveloped to such a degree that vision is still reduced during this period.

It is not uncommon to see children with low vision (due to various causes) who exhibit gazing behaviors toward lights. It has been suggested that this may be a form of visual self-stimulation, perhaps almost like a reflex, which is either performed subconsciously or even as an enjoyed stimulus.

I am hopeful that genetic research will allow us to better understand the biologic processes that are defective in retinal development which leads to achromatopsia. With such information, we can begin to understand the nature of various symptoms as they relate to anatomic and biologic correlates.

In chapter 8 of *Night Vision* (Cambridge University Press, 1990), entitled “Vision in a complete achromat: a personal account,” network member Knut Nordby writes, “According to my parents, I could control my eyes and direct my gaze even at the age of 3 weeks. Photographs taken of me a few weeks after birth show me with fully open eyes and not a trace of squinting, or partial closing of the eyes, in bright light. Nothing unusual was recorded at the time about my visual behavior (i.e., no light aversion or nystagmus). When I was about 7 or 8 months old, my parents began noticing that my eyes had started to quiver from side to side and that I had begun to blink continuously and to partially close my eyes or squint in bright light. Earlier, according to my mother, I had even looked straight into the sun with no sign of distress, and she often had to turn me around, being afraid that I would damage my eyes. This special visual behavior has also been reported to me by mothers of several other achromats.” (*Night Vision*, pp. 291, 292)

Ways to Help People Understand How Achromats See

by Frances Futterman

There are various situations in which it would be useful to be able to show a photo-simulation of how achromats see. Such a simulation would be of interest to relatives, friends, and “significant others.” Also, persons with achromatopsia are sometimes asked to explain to others how they see. Some young networkers have even been invited to make a presentation at school about their rare vision disorder. A good photo-simulation could supplement verbal or printed information. But is it possible to create a simulation that would come even close to accurately portraying the vision of achromats?

In training programs for professionals working in the field of services to the visually impaired, there has been increasing emphasis in recent years on helping people understand how the world appears to persons with different eye conditions. Brochures, posters, and other materials have been produced which display a series of pictures showing how the same scene would appear to someone with retinitis pigmentosa, someone with cataract, etc. Such photo-simulations are at best approximations – i.e., pictures that can give people a general idea of what it is like to have a particular eye condition. They do not provide exact representations, nor can they show the range of differences in visual functioning that exist among all persons affected by the same eye condition.

Sets of “simulator goggles” have also been available for many years. These have been used in teacher training programs in the fields of special education and orientation and mobility, as well as other areas of work with the visually impaired. The goggles are meant both to educate people about eye conditions and to sensitize them by providing direct visual experiences leading to greater understanding about the specific problems encountered by persons who have these eye conditions. For example, someone walking around wearing goggles designed to simulate advanced retinitis pigmentosa will gain far more insight into the orientation and mobility problems faced by someone who is left with only “tunnel vision” than can be understood by looking at a photo-simulation.

When I was in graduate school and taking a course in orientation and mobility training methods, we were required to walk around the campus and accomplish a series of specific visually oriented tasks while wearing various simulator goggles. For safety’s sake, we were always accompanied by a fellow student who was not wearing simulator goggles at the same time. It was during this period that I began to give serious thought about whether it was possible to create a simulator goggle for helping people to understand complete achromatopsia. I decided that such a simulation would be impossible to accomplish, because it would require simulating all at once the three basic visual realities which achromats experience: (1) the absence of color vision, (2) the absence of detail vision and clarity – more so in the distance than at nearpoint – and (3) extreme sensitivity to all factors of illumination. How an achromat sees in one place can be quite different from how he or she sees in a place having quite different lighting.

It is important to recognize the limitations of all devices intended to simulate the way people with various eye conditions see, whether they be simulations attempted with photography, paintings, computer graphics, or special goggles.

Over the years I have seen many badly constructed or ill-conceived simulations. A misleading simulation can do more harm than good.

An example of an ill-conceived simulation which did more harm than good was one that was used in a *Good Morning America* TV segment about achromatopsia shown on Feb. 29, 2000. Even though the producers of that segment had been provided with a substantial amount of good, accurate information about the vision of achromats, they came up with a bizarre and totally inaccurate simulation. Many of us in the network who watched that segment were disheartened to see how achromatopsia was portrayed on TV to millions of viewers that day.

By contrast, several years earlier, there had been a reasonably successful attempt by the producers of a San Francisco TV news program to simulate the kind of photophobia that achromats experience. Simply using overexposure of black and white film to light, they effectively simulated the visual experiences of a child with achromatopsia leaving subdued indoor lighting to go outdoors and play.

Several years later, a crude attempt to simulate how achromats see was published on the Web. The picture was in black and white and indicated some degree of photophobia. What disturbed many of our networkers who saw it was the way achromats' poor visual acuity was simulated. Using computer graphics, everything in the picture had been made extremely out of focus, indicating a very blurry view of the world. This is not how we see. Achromats around the world protested, and eventually this misleading simulation was removed from the Web.

In 1993 there was a brief presentation about achromatopsia on Cable News Network (CNN) which included a seriously misleading simulation. They were attempting to illustrate how the vision of a young man with achromatopsia had improved significantly after being fitted with specially tinted lenses to reduce glare. There was a very brief scene incorporating a strobe effect to represent the young man's rapid blinking while attempting to see outdoors in the sunshine without tinted lenses, followed by a scene intended to show what a difference the new lenses made. The "before" scene with the bright strobe effect was meaningful to achromats viewing the film, but the producers unfortunately went too far with the "after" scene, which showed sharp, detailed vision indicating excellent visual acuity and full color vision. Good quality lenses tinted to cut glare to a minimum can, of course, do wonders for the vision of achromats outdoors, but they cannot magically provide achromats with clear detail vision and color vision. Such vision is possible only through the function of cone photo-receptors in the retina, and achromats do not have cone vision.

In some sections of the film, *Island of the Colorblind*, which has been shown by many stations in the U.S. and Canada, as well as in various other parts of the world, viewers had the chance to see certain scenes both with and without color. The producers did not attempt also to simulate achromats' poor visual acuity or photophobia in these scenes. However, these vision problems were discussed in other parts of the film.

In his book, *An Anthropologist on Mars*, Oliver Sacks includes photographs which give readers the chance to compare the same scene with color and without color. The black-and-white photographs are not presented as being a simulation of the way persons with congenital, inherited achromatopsia see. They were included to illustrate the vision of someone who had developed cerebral achromatopsia, a neurological condition about which Dr. Sacks wrote in the first section of that

book and to which he sometimes refers in his book, *The Island of the Colorblind*. Cerebral achromatopsia is not an ophthalmological condition, as is congenital, inherited achromatopsia, and the colorblindness experienced by someone with cerebral achromatopsia is not accompanied by poor visual acuity and extreme light sensitivity, as is the colorblindness that results from the lack of cone photo-receptors in the retina.

Although we do not have photo-simulations of how achromats see, verbal descriptions contributed by achromats can go a long way toward helping people to understand about this extraordinary way of seeing. On page 2 of this book, the section entitled, “What is it like to have achromatopsia?” includes several excellent examples of visual experiences familiar to normally sighted persons which can help people to better understand the kind of light sensitivity achromats experience. Also, many of the networker comments in the section of the book *Living with Achromatopsia* entitled “Living with extreme light sensitivity” offer further insight into how achromats see.

In chapter 8 of the book *Night Vision* (Cambridge University Press, 1990), entitled “Vision in a complete achromat: a personal account,” Dr. Knut Nordby, a vision scientist who is himself an achromat, presents a substantial amount of personal and scientific information about what it is like to see with achromatopsia. Following are some excerpts from that chapter: “Trying to explain to someone with normal – or nearly normal – color vision what it is like to be totally colorblind is probably a bit like trying to describe to a normally hearing person what it is like to be completely tone-deaf, i.e. not possessing the ability to perceive tonal pitch in music. My task, though, is probably a bit simpler than the case of the tone-deaf, since practically everyone has had experiences of seeing achromatic (i.e. colorless, or black-and-white) or monochrome pictures and renderings, and also certainly must have witnessed the gradual disappearance of colors as darkness sets in. A first approximation then, in explaining what my colorless world is like, is to compare it with visual experiences that people with normal color vision have when viewing a black-and-white film in a cinema or when looking at good black-and-white photographic prints (sharply focused, high contrast prints with a long gray-scale, as in crisp, high quality, glossy technical prints). This, however, is only part of the story. To get a fuller understanding of my visual world, one must take into account not only my colorblindness but also my hypersensitivity to light and my reduced visual acuity. I experience a visual world where things appear to be well-focused and to have sharp and clearly defined boundaries, not fuzzy or cloudy. I can easily tell the difference between what people with normal vision call a well-focused photograph and a not so well-focused photograph.”

Even though Dr. Nordby correctly points out that normally sighted persons have had numerous experiences in observing black-and-white art, photographs, movies, etc., so that they can imagine a world without color, it should also be pointed out that what a normally sighted person experiences in those situations is not the same as what an achromat experiences in viewing the world without color vision. Someone with color vision perceives color-less art work or movies, etc., as being in shades of gray, but someone with complete congenital achromatopsia simply perceives a range of light, medium, and dark “shades” (densities, hues) but does not actually perceive “gray.” Only persons who are capable of perceiving colors can recognize the absence of color as being “gray.” Among persons with

achromatopsia, only the incomplete achromats who are able to see one or more colors report being able to perceive gray and, therefore, to understand the concept of gray. The rest of us (those with complete colorblindness) have simply learned to memorize the word “gray” along with all the other color names, such as red, green, etc., that refer to visual qualities we cannot see.

There is much truth to the old saying, “A picture is worth a thousand words.” So it is understandable that many people would want to see a picture showing how achromats see, in addition to the verbal descriptions we can offer about how we see. In today’s world, graphics have taken on increasing importance, with so much emphasis being placed on speed in the transferring and consuming of information. In contrast, reading and reflecting on what one has read takes time and effort, but it is well worth the time and effort for those who sincerely wish to understand achromatopsia.

A network parent wrote to me expressing the desire to understand what her young son with achromatopsia sees when he looks across the room or when he looks around outdoors in favorable lighting. As so many others have done, this parent asked “How far can he see?” My response was: “From what you have written, it seems that you have a good understanding of how bright light affects your son’s vision, and you also understand about his not being able to see colors. To understand the part about not having good ‘detail vision,’ it might help you to consider what your own vision is like when you look off into the distance, far beyond the point at which you can clearly see details. Things become progressively less distinct in appearance, less recognizable. You might use binoculars to see clearly something that is off in the distance. Well, for achromats, that indistinct way of seeing things occurs at a much closer range. The world we see is not blurred. It is just indistinct, unless we can get a close-up view of what we want to see. While you might use binoculars to look at something half a block away, we might use binoculars (or a monocular) to see clearly something across the room. There is no point in the distance at which we ‘stop seeing,’ so to speak. It’s just that the appearance of things becomes progressively more indistinct the farther away they are. We see the horizon, and we see best at twilight – the very time when normally sighted persons begin to experience diminished visibility.”

Because the way achromats see is so strongly influenced by all factors of illumination, any attempt to create a simulation of achromats’ vision would need to take into account the time of day, the location, the weather, any nearby shade or light sources, and other factors. Also, it would need to be made clear (e.g., by a caption, in the case of a photo) whether the simulation intends to show how an achromat sees without tinted lenses or while wearing tinted lenses (and, if so, what kind of tint is influencing the achromat’s vision). And the individual differences that exist among achromats should be kept in mind. There are complete achromats and incomplete achromats; and, within each of these two categories, there are those who have better visual acuity and less photophobia, and there are those who have lower visual acuity and more severe photophobia.

Who among us is qualified to produce a single picture or other form of simulation and say, “This is how persons with achromatopsia see”?

Hopefully, as our network grows, there will eventually be a collection of illustrations contributed by many achromats to show how they see. Such a collection would be far more useful than any single attempt to create a simulation.

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- Understanding Low Vision*, edited by Randall T. Jose, American Foundation for the Blind, 1983
- “Vision in a complete achromat: a personal account,” by Knut Nordby in the book *Night Vision* (see above listing), pp. 290-315

On page 117 of this book you will find a list of additional references which pertain specifically to genetic studies of achromatopsia.

★ *Understanding and Coping with Achromatopsia* provides information about diagnosis, coping techniques, inheritance factors, tinted lenses, and much more that networkers need to know about. *Living with Achromatopsia* is composed entirely of comments from networkers about their experiences and thoughts regarding living with light sensitivity, special needs in school, being colorblind, and various other subjects. For information about obtaining either of these books, contact Frances Futterman, P. O. Box 214, Berkeley, CA 94701-0214 USA.

★ *Night Vision*: For those who wish to have information about the history of achromatopsia, clinical aspects, achromatopsia research, and many other topics which are of interest to vision scientists, researchers, and vision care professionals, this book includes substantial information and lists of references.

The Achromatopsia Network

The Achromatopsia Network is a support network and information network for individuals and families affected by achromatopsia, a vision disorder affecting 1 person in 33,000 in most parts of the world. Before this network developed, very few people knew about achromatopsia. Even eye doctors and other professionals serving the visually impaired, because they rarely (if ever) encounter someone with achromatopsia, have had very little knowledge of how achromats see or how to assist them with their special needs. Before this network existed, very few individuals and families affected by achromatopsia had the opportunity to meet others similarly affected. Also before this network developed, it was very difficult to obtain any information about this rare vision disorder, and such information as could be found (in the professional literature of vision care specialists and vision scientists) was of little use to persons affected by achromatopsia. Little was known about how achromats cope with the challenge of living with very poor visual acuity, total (or nearly total) colorblindness, and the most extreme light sensitivity that can be experienced by the eyes.

The goals for this network and for network publications are:

1. To gather and share information about achromatopsia, from a variety of perspectives, and about resources for meeting the special needs of achromats
2. To promote awareness, education, and understanding about achromatopsia, especially among professionals who work with the visually impaired
3. To help individuals and families who are affected by achromatopsia to connect with one another

Books and other publications created for the network present information about many subjects – light sensitivity, tinted lenses, orientation and mobility, special needs in school, vocational options, diagnosis, adaptive methods and devices, research, social and psychological aspects, products and services for the visually impaired, and more. The network Journal also serves as a forum for networkers to share their experiences, thoughts, and suggestions. See pp. 150-151 for more information about the publications that are available.

There is no funding for this network except for what people send in when ordering books and subscriptions or when making a donation. Donations are essential in order to maintain this work.

For more information about the network and network publications, contact:

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Or visit our network's website at <http://www.achromat.org/>

Frances Futterman, who serves as facilitator for the Achromatopsia Network and editor and reporter for publications, has complete achromatopsia. Her background has been in the field of special education for visually impaired students and rehabilitation teaching and counseling for visually impaired adults.

Regarding the Book, *The Island of the Colorblind*, by Oliver Sacks and the Documentary Film, “Island of the Colorblind”

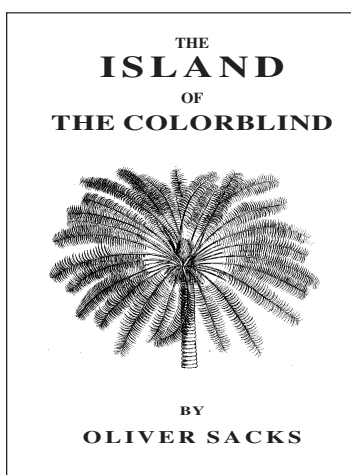
by Frances Futterman

In 1998 many PBS stations in the U.S. showed the film, “Island of the Colorblind,” which includes footage of the journey made by Oliver Sacks, network member Knut Nordby, and a BBC film crew to two Micronesian islands where there is a high incidence of achromatopsia. This film, which has also been seen in many other parts of the world, has contributed greatly to public awareness of achromatopsia, as has Dr. Sacks’ 1997 book, *The Island of the Colorblind*.

It was in August, 1994, that Dr. Sacks and those accompanying him traveled to the tiny atoll of Pingelap, where at least 6% of the population have achromatopsia, and then on to the island of Pohnpei, where there are two enclaves largely populated by Pingelapese, many of whom have achromatopsia. The trip took place 8 months after I had first contacted Dr. Sacks to introduce him to the subject of congenital, inherited achromatopsia and to our network. I knew he had been very interested in cerebral achromatopsia (an acquired neurological condition), because I had read his essay, “The Case of the Colorblind Painter” (published in *The New York Review of Books* in 1987), and felt that he might also want to know about congenital (retinal) achromatopsia. In March, 1994, when he was in the San Francisco area for speaking engagements, I had a meeting with him, we discussed achromatopsia and Pingelap, and I referred him to Knut Nordby, a vision scientist who is an achromat and who I felt might want to go along on the trip Dr. Sacks was planning. By summer’s end, “Oliver and Company” were off to Micronesia.

This journey led to the documentary film and the book, which has been translated into Italian, French, German, Spanish, Dutch, Portuguese, Norwegian, Japanese, Swedish, Turkish, Chinese, and Hebrew. Numerous newspaper articles and book reviews have been published in connection with the book, and Dr. Sacks has appeared on many radio and TV programs while publicizing the book. I could never have imagined back in 1994 all that my decision to begin corresponding with Oliver Sacks would lead to! Many of his books and essays have focused on

medical conditions that cause those affected to have very unusual ways of experiencing the world. His book *Awakenings* was the basis for a 1990 movie by the same title, in which he was portrayed by actor Robin Williams. Dr. Sacks is a popular lecturer, both in the U.S. and abroad. He usually addresses “standing room only” audiences on speaking tours, and his books tend to become bestsellers. He has a strong following in the disabled community, largely because he views people who have physical or other abnormalities with great respect and keen interest – rather than in a patronizing, sensational, or strictly clinical manner. His writings reflect gentleness, compassion, and a sense of humor. Before I contacted him in 1994, Dr. Sacks had never heard of congenital achromatopsia. What captured his interest most about our vision disorder was the



See page 151
for details about
ordering this book.

thought of what it is like never to have seen in color. So I made sure that he was well informed regarding the “rest of the story” about achromatopsia. It was important that he fully understand about our extreme sensitivity to light and our poor visual acuity – vision problems which, for achromats themselves, are far more significant than the colorblindness aspect which many people find so intriguing.

Dr. Sacks decided to travel to Pingelap to see what he began to call “the island of the colorblind.” Once there, however, he saw ample evidence of the other manifestations of achromatopsia. He could see that the islanders with achromatopsia were concerned very little with their lack of color vision as they struggled from sunrise to sunset to cope with the problems caused by the inability of their eyes to tolerate bright light. In the film, one can see that the faces of the Pingelapese achromats reflect a lifetime of having to squint hard all day long, just to be able to cope in a location where sunlight is plentiful and where there is not much shade to be found. There are scenes in which wraparound sunglasses are being distributed and tried on by the achromats. Before the trip, Dr. Sacks had contacted me for suggestions regarding sunglasses and other items that he could bring along to give to the Pingelapese achromats. He wanted to help them.

The film includes close-ups showing Knut Nordby squinting and blinking, as achromats do when the light is too bright. In these scenes it is clear that Knut is doing without the protection that sunglasses offer, yet in other scenes he is shown wearing sunglasses. While it is true that Knut, like many of us in the network, prefers to avoid over-dependence on tinted lenses (to avoid becoming habitually dark adapted), this was not the reason for the scenes without sunglasses. A major purpose of the film was to show what it is like to have achromatopsia, and the producers felt that this purpose could not be achieved if Knut’s eyes were always seen shielded (and thus hidden) by sunglasses.

Fortunately, these scenes are presented in such a way that the manifestations of Knut’s light sensitivity can be observed while also allowing other aspects of who Knut Nordby is – i.e., a highly distinguished scientist and teacher – to come across just as clearly. Many thanks to Knut, on behalf of achromats everywhere, for his willingness to be used in this very public way to help people viewing this film to understand about achromatopsia. I doubt that many of us in this network would have been willing to submit to such close scrutiny by video cameras and to have close-up views of us, without our dark glasses, seen by large audiences.

One scene is misleading. As Knut is telling about the period in his childhood when he had to attend a school for the blind and the teachers there made him use Braille instead of print, there is a close-up of someone’s fingers reading and writing Braille. Viewers may get the impression that it is Knut who is using Braille, but this is not the case. He reads and writes print, as do other achromats.

Both in his book and in the film, Oliver Sacks uses the terms “achromatopia” and “achromatope.” These terms were not in common use before Dr. Sacks began to use them, but they are acceptable variations of the more familiar terms, “achromatopsia” and “achromat.”

I make hardback copies of the book *The Island of the Colorblind* available to networkers in the U.S. (see p. 151). The “Island of the Colorblind” film is shown from time to time on PBS and other TV stations around the world. It is part of a 6-part series entitled, *Oliver Sacks: The Mind Traveller*.

Achromatopsia on the Islands of Pingelap and Pohnpei

reprinted from the Oct., 1994 Achromatopsia Network Newsletter

Following are excerpts from personal accounts shared by Knut Nordby and Oliver Sacks after their return from the two Micronesian islands where there is a high incidence of achromatopsia. (About 6% of the Pingelapese people have achromatopsia, and many more of them carry the gene).

Knut Nordby's Report

This expedition was the most exciting journey I have ever made and I feel very privileged to have been allowed to take part in it. The hardships notwithstanding, I had a most memorable experience, which gave me many important insights into my own experience as an achromat and led me to reflect on the problems facing persons with achromatopsia and to see both similarities and differences across cultural, economic, and political boundaries.

Going to what we (in Norway at least) think of as "a paradise on earth" but finding instead a third world country gave many thoughts for reflection.

My immediate impressions were of the heat, the high humidity (up to 95% relative humidity), phenomenal rainfalls (some places on Pohnpei receive over 400 inches annually!), the bright light (Pohnpei is only about 6° north of the equator), the hardness and sharpness of coral and of black volcanic basalt rock (the islands were originally volcanoes), the lushness of the dense vegetation, and the uniformity of "color" (all shades of green).

Meeting and speaking to the achromatopic people on Pingelap and in the village of Mahn on Pohnpei gave me a real jolt. I had more or less forgotten the basic problems such as they must learn to live and cope with, which are greatly compounded by living under

tropical conditions in a third world country, problems which I myself long ago resolved and learned to live with.

Coping with such extremely high levels of light in a tropical day close to the sea must be daunting to them. I had to wear double protection: i.e. both a polarizing clip-on filter on my glasses and the dark amber wraparound goggles over my glasses, to be able to see anything without intense squinting and incessant blinking. With no safe eye protection filters available, these people must resort to various strategies to avoid bright light: i.e., squinting, rapid blinking, shading the eyes, or simply staying out of the sun. This makes it hard for them to participate in work and civic activities.

Although I feel confident that the people I saw were typical, complete achromats, this is based on rather superficial observations – (1) their avoidance of bright light and the visual strategies they use; (2) their low acuity, which was, in part, confirmed by Dr. Wasserman's measurements with the Snellen chart and which matched my own visual acuity; and (3) the matches made on the Sloan Achromatopsia Test by those who understood the instructions (not all of them did, despite much prompting by interpreters). Their matches were identical to those I made under the prevailing light conditions. To be valid, the Sloan Test depends on standardized artificial illumination, a condition which could not be met there. I had to resort to this indirect validation by comparing their brightness matches to my own judgement of "correct" brightness matches, using these as comparison standards. The only proper way to diagnose achromatopsia is to use the Nagel anomaloscope, but this was not

possible there.

Seeing how these people coped – in school, at play, at work, and in social activities – was both saddening and inspiring. It was saddening, because they struggled under such adverse conditions with no optical aids or proper counseling – inspiring, because they actually made the most of their situation. Seeing two little girls in the village school in Mahn doing arithmetic lessons in class with their noses virtually touching the pages of the book reminded me of my own early school days, before I had optical aids. I had a strong desire to give them proper magnifiers and teach them how to use them. We filmed a short episode in which I lent them my own pocket magnifier.

Getting to speak to the teachers and to advise them on the management of these pupils was very worthwhile.

I had interesting and heart-warming conversations with some parents of achromatopic children, especially one mother who was concerned about her daughters' future and anxious to know if they would go blind. In Pingelapese, achromatopsia is called "Mascun" (pronounced Mah-skoon), which translates as "not see." On Pohnpei it is referred to as "the Pingelap eye disease," thus inferring that it may be progressive, even though experience and conventional wisdom should have taught them that the condition is stable. She wanted to know why her children had Mascun. Was it something she had done during her pregnancy, was it something they had contracted from others, or was it something wrong with her as a woman or as a mother?

It is not easy to explain heredity to an uneducated person who speaks little English, but I did my best to reassure her they would not go blind from their Mascun, that with optical aids and eye protection they could do as well as

others in their society, that Mascun was no barrier to education or holding a job, and that she had done nothing wrong, but that, if she had more children with the same father, they might also have Mascun. I told her I had Mascun. This seemed to reassure her.

On our second evening on Pingelap a night fishing expedition was organized. We went out at sunset in 3 open 16 ft. fiberglass boats with outboard motors (gone are the days of outrigger canoes, torches, and the gleaming muscles of rowing men). This was a very memorable experience. It is an old tradition on Pingelap to go out in canoes in the dark with torches to catch flying fish.

The setting was very pleasant – the mild breeze, the lowered temperature, cloudscapes in the horizon, the clear sky, the decreasing light and deepening darkness, the nearly luminous surf at the coral reefs, the spectacular stars and Milky Way, and the shining flying fishes soaring over the water in the light from the flashlights.

When the Pingelapese fish, they run the boat at slow speed, while searching the sea in front with a flashlight until a flying fish suddenly breaks the surface and glides across the water. They then speed up the boat and follow the fish, the man with the flashlight trying to keep it in the light beam. When they catch up with it, they fish it out of the water with a long-handled net.

I am sure that I would have no trouble in taking the position with a flashlight in the bow of the boat to track the flying fish with the beam and to net the fish, nor in taking up the position in the stern and steering.

On our last day on Pohnpei I was asked to speak to the medical students of the training program in Kolonia (under the auspices of the University of Hawaii at Manoa) about what achromatopsia is, how it is propagated, and how

achromatopic people should be treated. Dr. Sacks and Dr. Wasserman also spoke to the students.

I will have more to say as time passes about this odyssey to Micronesia. It has been the most exciting and interesting journey I have ever made, and I am still reeling from the experiences and many impressions.

Editors note: These experiences and more were discussed in an interview I had with Dr. Nordby when he stopped briefly in San Francisco after his flight from Micronesia and before his return home to Norway. Here are excerpts from that interview:

.....

When we would go out to sea near the islands, there were the very bright coral reefs – and the sunshine at sea plus the reflections from underneath were overwhelmingly bright.

On the little island of Pingelap, the only communication we had with the outside world was a citizens band radio, and sometimes it didn't work. Twice a week a small plane came to the island. The air strip was the shortest I've ever seen. When Dr. Sacks was arriving on this plane, they almost had an accident. A cross wind caught them and the plane almost skidded off the landing strip. Measuring the skid marks later, we could see that at one point it was 12 inches from a sheer drop.

We had plenty of sun goggles to give the adults, but we had nothing to give to the children.** It was heartbreaking to see them. The smallest was 9 months old. His face was severely distorted all the time in an effort to squeeze his eyes shut. I recognized it as the way my brother (who also has

** Plans are underway to send a quantity of children's sunglasses to these islanders.

achromatopsia) looked in bright sunlight when he was very small. I might have looked like that too when I was small. The eyes were like two slits. When I tried to shade him by putting my hands around his face, he would open his eyes a little to look at me. The children had no hoods, hats, or anything of the sort to shade their eyes, nor did the adults with achromatopsia. This is probably because it is too hot, humid, and sticky there to be comfortable wearing headgear.

When we first touched down in Micronesia, we saw smoke coming from the wheels. Two tires and something else on the undercarriage had been damaged. It was several hours before repairs were made. Finally we took off, but it was rocky, because by then one of the wheels was not round. This plane was shaking badly, such as I had never experienced before in all my travels. So we touched down at a military base to change two more wheels. Then they ran out of "crew time" and had to head straight for Guam. So we had to fly right over our island (Pohnpei), as we were already 9 hours late. There were people on the island waiting for us with flowers and a big feast. But our plane had to go on to Guam, where we changed planes and went back to Pohnpei via one other island. We arrived at 3 AM, having spent 24 hours getting there.

On Pingelap we couldn't take a shower or even wash our hands. They did have some water on tap, but it was not safe for us to use. The conditions under which the people live are beyond belief. Shelters are made of corrugated iron pieces put together with other "found materials." No floors – everyone slept on mats made from fibers. This was no paradise. Postcards from this area show nice beaches and people out snorkeling and scuba diving. But, if you turn that camera around 180 degrees,



you find living conditions worse than any I have ever seen in my travels. This was not the case in Pohnpei, where you can see houses. Those governing the area live on Pohnpei.

It was hard getting sleep there, because large insects kept marching across us during the night. We tried to put up mosquito nets, but it didn't work very well. And it was very hot.

When I gave the talk to those who were receiving medical education on Pohnpei, I incorporated some information taken from the network newsletters. These people will be the future doctors and nurses for the Pingelapese. They knew nothing about achromatopsia. It is known there as the "Pingelap eye disease." I tried to help them understand that it is not a disease but a disorder of the cones of the retina.

At the school we saw an achromatopic child sitting in the front row, and on the chalkboard were pictures of plants, animals, and various other things. The teacher would point to the pictures, and the whole class would say the words in Pingelapese. This little boy had to listen for the cues for what to say, so he always said the words a little later than everyone else did.

The achromatopes are referred to as "Mascun," meaning "not to see." Being blinded by sunlight so much of the time, they act very much like blind people. They are not called "colorblind," as we are called in our society. Not being able to distinguish colors there is rather insignificant, since everything is green. We tried to test some of them with various colors of yarn, but we got the impression that they didn't even have names for all the colors. However, they had many names for different shades of green – just as Eskimos have many different names for snow.

On Pohnpei Island we visited a community of Pingelapese people. We gave

away sunglasses to all the ones whose vision we tested, and we left the rest with a local nun to give away to other achromatopic people there. Before we left, we were able to see some of the people wearing the sunglasses that we had brought, and this felt good.

Oliver Sacks' Report

(From a taped phone conversation)

I believe the film crew captured some beautiful footage at Pingelap and Pohnpei. Pingelap seemed enchanting – magical – when we landed. It was like an island of children. There were about 50 beautiful, brown-skinned children, laughing and greeting us. They led us into the forest through coconut palms, with little black and white pigs scampering around. One could immediately spot the children who had achromatopsia. Their heads were tilted a certain way, and they were squinting. Bananas were everywhere. We asked one man how he could tell which bananas were ripe. He brought us two bananas which appeared to be green but which turned out to be perfectly ripe, as if to illustrate that there are other ways besides color to tell ripeness.

The next morning we went to the clinic on the island to do vision testing. There we met many achromatopic people, including a 3-month old baby and a 79-year old woman. They all seemed to have the same degree of acuity and aversion to light. We distributed sunglasses and other items.

They had various ways of dealing with their light sensitivity. One little boy had a black T-shirt which he kept pulled over his head. I saw people holding their hands up to shield their eyes. On Pohnpei, we saw some weavers who worked in a dimly lit environment – we had to open the lenses of the camera very wide in order to take pictures. We went to a little school on Pohnpei. I

think this was an overwhelming experience for Knut, reminding him of his own school days. Two children clearly didn't know what was going on in the lesson. They looked bewildered. They so obviously needed optical aids, which could make a crucial difference for them.

We met a couple who have 3 achromatopic children. The father and Knut got on very well. Knut ended up giving him his monocular and later said, "I gave him half my eyes." The man's wife gave Knut a beautiful necklace. There was a kind of deep bonding.

There didn't seem to be much isolation of the achromatopic people, but there did seem to be less going out, less play and less work outdoors in the daytime – and more of this toward sunset. I was told that they were especially good at night fishing, but I didn't get to observe this. I was, however, able to see that Knut was extremely good at catching the glint of moonlight on the outstretched fins of a flying fish. Just before dark we could see the children playing, and men would do some canoe building or cutting taro. *(Editor's note: they have only a very brief twilight period before night falls, and on Pingelap there are no electric lights to allow them to enjoy using their night vision.)*

In general, the achromatopic people were underemployed, under-married, and under-educated. We did get to meet two who were highly educated, including a young man who had taken a degree at the university on Guam.

The environment was close to monochromatic. There was a wide range of greens, which the achromatopic people weren't in the least confused by, because they discriminated by tone and leaf pattern. They were adept at identifying virtually all the vegetation and were not



deceived by color. The sunglasses we brought were new to people, and we saw varied reactions. Some seemed to preen themselves with them, and others seemed to be embarrassed by them.

It was moving to observe how Knut seemed to recognize instantly all his "brethren" – and they also instantly recognized him by his dark glasses, his posture, his blinking, and so forth. They clustered around him. On Pohnpei they even generated a myth involving him. We heard both a rational and a magical explanation of the eye condition. The rational ones said they understood that this was genetic. But we also heard from some that it was caused by a woman having walked on the beach in the bright sun while pregnant. The sun comes in, they said, and does something to the eyes of the fetus.

In general, on the islands there is a feeling that whatever is the matter, one way or another, it is due to "the white man." But Knut's advent, in particular, created a myth that achromatopic white whalers from Norway had come to Pingelap in the early 19th century and had raped or married some of the women. This island had, in fact, been visited by whalers in the early 19th century, but now it was being said that these were achromatopic whalers from the far North, who had mingled their blood with the natives. So accepting this particular myth meant having to believe that these Pingelapese were part Norwegian! I heard this being explained on Pohnpei – we have it on film – and I couldn't quite believe my ears. It was a fascinating example of instant myth-making.

We saw 2 Pingelapese communities, the original one on Pingelap and a community of those who had moved to Pohnpei, the large island, at the start of the century. They have a mountain village there, Mahn, where, in some ways, life is more pleasant. It's cooler and

they have electricity and fresh water. On the other hand, it is more ghetto-like, because the Pohnpeians granted them only a tiny bit of land and refused to grant them any more. They have had a high birth rate, resulting in a population explosion and not enough room.

The Pingelapese on Pohnpei are more inclined to better themselves through education or employment opportunities. So all the educated or professional persons with achromatopsia we met were on Pohnpei and not on Pingelap. Among them were a young man who has studied social sciences at Guam University, some school teachers, and a writer. On Pohnpei one is conscious that the achromatopic people are, to some extent, a disliked minority and confined to a ghetto.

There weren't many complaints about being colorblind. There was more concern about the photophobia and poor visual acuity. We left from 100 to 150 pairs of sunglasses with visors on the island, and I'm not sure this was enough. These sunglasses break so easily. We've got to get some more to them.

We also need to get more information to them. People came up to Dr. Wasserman, Dr. Nordby, and me and asked, "Is this progressive? Will my child go blind?" Before we left, we prepared an information sheet, which was to be translated and distributed.

It was crucial having Knut Nordby with us. To some extent, the achromatopic people are seen as sub-normal or "lower caste." I think that this ceased to be so, when they saw him. Here was this distinguished European scientist, a gentleman, who travels around the world independently, has a family, and is not disabled by his eye condition. I think it was very important for them to see this.

I think Knut found he had a lot in common with the achromatopes there. I

believe he was "at home" with them, and they were "at home" with him, in a way that wasn't true for the rest of us.

I'm very glad we went. It was truly an adventure and extremely moving. Most importantly, I think that, in a good way, things will never be the same there. There will be increased awareness and understanding for these people at every level – visual aids, improved education, less fear, and more acceptance. I certainly don't want to think of ourselves as having been there simply to film this story. The least we can do for them is to see that they have the visual aids that they need.

My last memory on Pohnpei was of a boy who was absolutely delighted with his visored sunglasses, grinning and running alongside the car as we drove away, clearly able to see much better.

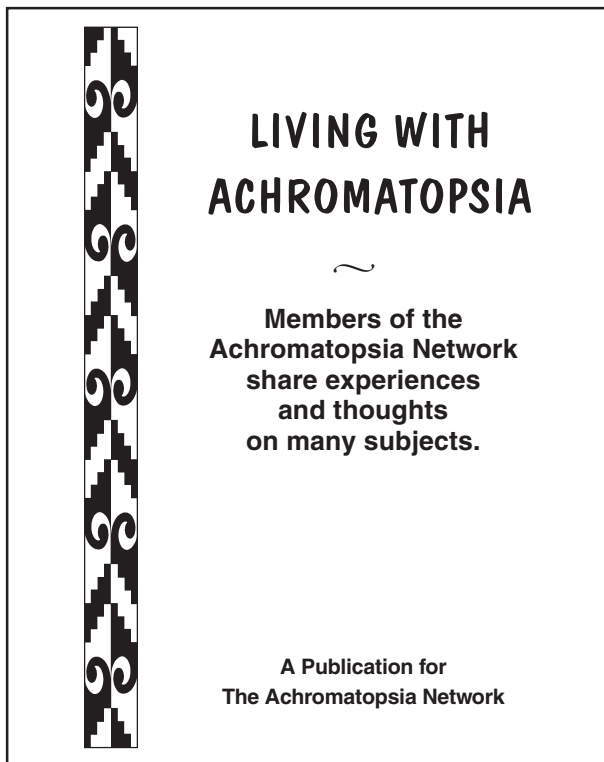
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A full account of this trip to Pingelap and Pohnpei is included in the first part of Oliver Sacks' book, *The Island of the Colorblind*, which was first published in the U.S. in 1997 (Alfred Knopf, publisher). A paperback version was published by Vintage Press in 1998. The book has also been published in the U.K. and Australia, and there are Italian, Dutch, German, Spanish, Norwegian, Swedish, Portuguese, Japanese, Turkish, Hebrew, and Chinese translations. There is also an audiobook in which Dr. Sacks reads selected excerpts from this book (Random House Audiobooks).

In a BBC-produced film series about Oliver Sacks, one segment is entitled "Island of the Colorblind." This documentary film contains considerable footage of the journey made to Pingelap and Pohnpei islands. This program was first shown in the U.S. and in certain parts of Canada in September, 1998.

Numerous articles have appeared in newspapers and magazines about both the book and the film series.

Other Publications for the Achromatopsia Network



The following is from the introduction to the book, *Living with Achromatopsia*:

What is it like to have achromatopsia? What is it like to go through life with very poor visual acuity, the inability to see color, and the most severe form of light sensitivity that can be experienced by the eyes? To have a vision disorder which causes one to experience varying levels of vision impairment, depending on the factors of illumination which vary from one place to another? Readers of this book, first published in 1999 and revised in 2004, have the opportunity to gain knowledge and insight regarding this rare disorder.

This book consists entirely of comments from persons who know first-hand about living with achromatopsia. Adults and teens with achromatopsia and parents of children with achromatopsia tell how it has affected their

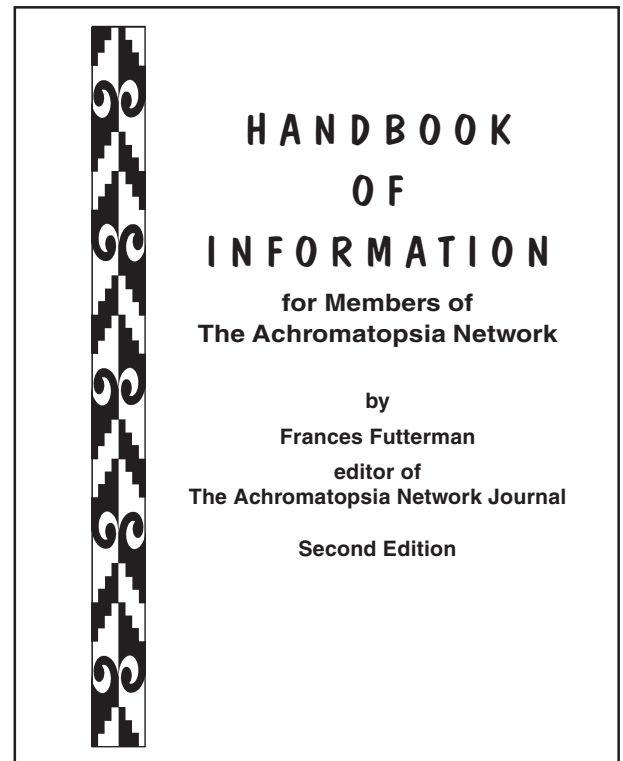
lifestyles, their relationships, and their participation in various kinds of activities. They tell about their experiences in connection with work, school, parenting, recreation and sports activities, and much more. They tell of the problems they have faced and the ways they have dealt with these problems – the coping strategies, adaptive devices, attitudes, adapted environments, support systems, and special resources that have played important roles in their lives.

The comments that fill the pages of *Living with Achromatopsia* have been gathered almost entirely through various forms of correspondence received from members of the Achromatopsia Network, a support and information network that began in 1994. Most of the comments are from networker input via postal mail and e-mail. Some of the comments have been excerpted from tape recorded letters, and some are from a published paper written by network member, Dr. Knut Nordby, entitled: "Vision in a complete achromat: a personal account." Most of these comments have appeared in the "Responses from Networkers" sections of the network newsletter (later called the Achromatopsia Network Journal) from 1994 through 2004.

Networkers' comments are categorized according to the following topics: living with extreme light sensitivity, living with colorblindness, using adaptive methods and adaptive devices, social and psychological aspects, coming to terms with terms, relationships, activities of daily living, school experiences, vocational experiences, recreation and sports, orientation and mobility, getting diagnosed, experiences with vision care specialists, using services for the visually impaired, parenting children and teens with achromatopsia, and networkers' comments on miscellaneous subjects.

For information about obtaining a copy of this book, write to Frances Futterman,
P.O. Box 214, Berkeley, CA 94701-0214 USA or editor@achromat.org

Handbook of Information for Members of the Achromatopsia Network, published in 2003, presents information about many matters that network members need to know about – information about how this network is set up, how it got started, what is offered, and the policies, procedures, and guidelines that I follow as facilitator of the network and publisher of books for the network and the Achromatopsia Network Journal. Some of this information has appeared in various newsletters, reports, and letters that were sent to subscribers from 1994 through 2003. This 40-page spiral-bound book contains sections on “Diversity in the Network,” “Quoting Networkers,” “Organizing among Networkers,” “Policies and Guidelines,” “Ways to Connect with the Network,” and other subjects.



The Achromatopsia Network Journal (previously known as The Achromatopsia Network Newsletter) presents information about many subjects of interest to our network and provides a forum for sharing networkers’ thoughts and experiences. The Journal deals with topics such as light sensitivity, colorblindness, adapted lifestyles, special needs in school, social and psychological issues, services for the visually impaired, and more. At least four mailings go out to subscribers each year. Each mailing includes two or more items. In addition to the four quarterly issues of the Journal, there are special reports and occasionally an open letter, a survey form, photo pages, or other items. Subscriptions are available only to persons who have first obtained the book, *Understanding and Coping with Achromatopsia*. The information that is in the Achromatopsia Network Journal supplements the information in the book and frequently makes reference to specific pages of the book.

The Island of the Colorblind, by Oliver Sacks (published by Alfred Knopf, 1997), is a book that has done much to increase public awareness of achromatopsia. Hardback copies of this book are no longer available in stores, but they are available (to persons in the U.S.) through this network at a very reasonable price.

Also available: Copies of “Vision in a complete achromat: a personal account,” a 28-page paper written by network member, Dr. Knut Nordby. This highly interesting personal and scientific account is a chapter from the book, *Night Vision*, published by Cambridge University Press in 1990. Dr. Nordby is a distinguished vision scientist as well as a complete achromat.

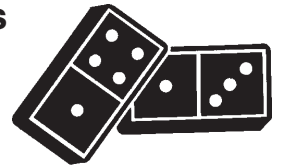
All network publications are in 14 point bold print on 8 1/2" x 11" pages.

For information about obtaining any of these publications, write to Frances Futterman, P.O. Box 214, Berkeley, CA 94701-0214 USA or editor@achromat.org.

MISCELLANEOUS SUGGESTIONS On this page and the following 4 pages, network members tell about favorite resources, devices, books, and other things.

From a man with incomplete achromatopsia: "The most valuable tool I have for distance vision is an Ocutech vision aid, which I obtained at a low vision center. It's a 4X telescope/periscope system mounted horizontally above a pair of prescription glasses. While not exactly inconspicuous, it is quite discreet, and it has made a vast difference in my life. It is especially useful for computer work, watching videos or theater, etc." *(Editor's note: Several networkers have expressed interest in the Ocutech device. It may be more useful to those who have better visual acuity, such as incomplete achromats, and it is very expensive. Contact a low vision center for information about this and other distance vision devices.)*

From a woman with achromatopsia: "As I was growing up, my family often played domino games, so I learned to play dominoes early on and loved it. Black dominoes with big white 'dots' (actually white impressions in the dominoes) were perfect for me. I could easily 'read' the dominoes anywhere on a card table, so I was not put at a disadvantage due to my vision, the way I was with most card games or board games. Later on, I found out that some domino sets are white with black dots, and these are not as easy for me to 'read' across the card table. A dark colored table top always made playing any kind of table game easier for me."



"As an alternative to safety pins for keeping socks matched, one can use 'sock locks' that are available in some stores and through some of the specialty catalogs for people with low vision. Also, there is a special checker set which consists of round pieces and square pieces, solving the problem of distinguishing between red and black checkers." *(Editor's note: Readers who would like to have a list of specialty catalogs offering items of interest to low vision people can write to Vision Community Services, 23A Elm St., Watertown, MA 02472.)*

From a woman with incomplete achromatopsia: "I've been around horses all my life, and I think horseback riding has been the best thing in the world for building my self-confidence. Horses have been my feet and my eyes in many places where I had trouble seeing. I was safe as long as I could hang on. My mom first teamed me up with her experienced mare and then gave me a small pony of my own. With them I developed more physical skills and dexterity than any gym class at school could give me. The National Therapeutic Riding Association is an organization whose members volunteer their time to assist physically or mentally challenged kids. I have donated quite a bit of time in the past to my local chapter, and it has been a very rewarding experience for me. It allows me to give others the same kinds of opportunities that I have had."



"My wife and I recently made a trip to Australia. This trip offered me a long-awaited opportunity to shop for a hat. I've always liked wearing hats, but I've had a hard time finding hats that fit me well, because I have a large head. The Stetson I bought in Texas 17 years ago no longer does the job, and the best I can do in the way of a sun shading brim is a generic ball cap. I knew I would be able to find what I wanted in Australia. For about \$60, I bought myself an Akubra, which is the

traditional Australian stockman's hat. It keeps the sun off your neck and the rain out of your face. I love it, in particular because it drastically cuts down on the reflections and light leakage that diminish the effectiveness of my wraparound sunglasses. Sure, I might look odd striding around anywhere but Australia wearing this hat and my large dark glasses, but this hat is stylish, and it has drawn many admiring comments. Whenever there is a strong wind, I have to tie it down to prevent it from being blown away. As broad-brimmed hats go, you can't do much better than an Akubra. I suppose a Mexican sombrero would be the extreme."

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From a woman with achromatopsia: "I just finished reading *The Planet of the Blind*, a memoir by Stephen Kuusisto, who grew up with a vision impairment (something other than achromatopsia). The main conflict of his life has been accepting his 'legal blindness.' For many decades he tried, with his severely limited vision, to 'pass' as normally sighted. Personally, I am all too familiar with this kind of excruciating self-consciousness and unwillingness to accept limitations – which is, at best, a self-transcendent ideal and, at worst, a self-defeating strategy. I think *Planet of the Blind* is a courageous book. I recommend it."

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"I found a book entitled, *From Homer to Helen Keller*, by Richard Slayton French, which provides a historical overview of social responses to the blind. This book was written in the 1930's, which gives it a rather dated flavor, but the author's strong and dedicated (even if opinionated) voice gives it a timeless appeal. I find that I am looking for stellar role models in terms of individuals who have succeeded in spite of having a visual handicap."

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From a woman in her 40's: "On certain occasions I absolutely love having my button to wear that says 'I HAVE LOW VISION.' I realize that some people would feel uncomfortable about broadcasting their vision disability in this way, even if only occasionally, when shopping in certain places, but for me this button is useful in so many situations. For instance, there have been several occasions when I have been suspected of being a shoplifter because of the way I seemed to take forever in fitting rooms or the way I would scrutinize labels an inch or so from my nose in brightly lit stores. I remember one occasion when I could have really used a button like this. A clerk had actually called for a security officer, because I had taken so long and because I was carrying a large handbag. Instead of allowing them to take me up to their office, I just emptied my handbag onto the counter, right there in front of God and everybody. Along with my wallet and any checkbook, out tumbled my monocular, my binocular glasses, my pocket magnifier, and six pairs of assorted glasses and sunglasses, demonstrating, beyond any doubt, that there was no room in my handbag for anything else. My husband and I were the only ones who were not embarrassed during this scene. But I think that wearing an 'I Have Low Vision' button would have been easier."



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From a man with achromatopsia in Italy: "When I hike, I use standard hiking canes made of aluminum. These have handles identical to those on sticks used for skiing. Each cane consists of 3 pieces, which can be extended or telescopically retracted, and a sturdy metal tip which is resistant to continuous contact with

rocks and stones. These are not to be confused with white canes used as mobility aids by blind people. They are found in most sports equipment shops near popular mountain towns here. They look like plain aluminum, with a colored adhesive spiral strip around them to make them more visible. People normally use them just as a support for walking along mountain paths. They fit my needs perfectly. Before I had them, I had to be much more cautious on descending trails. Sometimes I had to choose between making a good guess about how deep the next step was or sitting down on a rock and reaching out with my legs until my feet encountered a solid surface. Now all I have to do is lean slightly forward and use my canes to determine how much higher or lower the next step will be. I don't need to use them to locate obstacles in front of me, as a blind person uses a cane. These are meant to be used as a pair, but one could be used alone."

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From the mother of a 6-year old: "I found a bicycle called 'Trail-A-Bike,' made by Adams. It is a small bike that connects to the seat post of a leader bike. The trailer bike has a set of pedals and handle bars, so that a child can actively pedal and participate on a family bike ride, without the need to worry about traffic. It is designed for children from 5 to 10 years old. It does not have brakes, so there's no need to worry that the child might sit back and ride the brake while the parent tries to pedal. The child is free to pedal or coast, as desired. The item cost me around \$200 for a 1-seat version. It is also available with as many as 3 seats. It can even connect with a tandem bike, so a family of as many as 5 people can bike together." *(Editor's note: check with your local bike shop for details about this and other brands of trailer bikes and tandem bikes.)*

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One networker, a man in his 60's, writes: "I like using broad nib fountain pens with black ink." Another networker recommends bold fiber-point pens with black ink for general use and Pilot brand Precise pens with black ink for easily discernible handwriting when writing small is necessary or preferred. Many achromats routinely get up close to what they are writing and produce average size handwriting. Others like to write in large letters, and some of these use felt tip pens. Most networkers report using broad-tipped pens only when creating notes or other written material which they will need to read from a distance (such as notes for a presentation they will make to a group). Some report that they like writing paper that is some color other than white. All who have reported on the subject of writing prefer pens with black ink.

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"Those of us who are partially sighted often have concerns about our footing on hiking trails. Going uphill is fairly easy, but going downhill can be risky. Years ago I bought a handsome wooden hiking cane with a curved handle and a pointed end for getting a good grip on terra firma. Well designed canes and staffs can be found in stores that sell equipment for camping and outdoor sports and in stores that specialize in equipment for walkers. I also invested in high quality hiking boots, with soles designed for secure footing. A good hiking companion who is attuned to our special needs is great; but, most of the time, we achromats need no special assistance on hikes, especially if we go equipped with a good hat or visor and any optical aids that we're likely to need."



Hats, caps, visors, bonnets, sombreros, patio umbrellas, parasols, awnings, canopies, tents, covered backyard play structures – there are so many ways to lessen exposure to the sun for adults and children who have achromatopsia. Whenever we have a choice in colors for items such as these, the best choices for achromats are dark colors, especially black, navy, and dark brown. Red works well too, except for those who use red lenses, in which case red fabrics will appear nearly white, thus brightening one’s view instead of darkening it. Hats and other head coverings for children are so often found only in pastel colors, but a little searching can turn up darker options. Do-it-yourself sewing or construction projects make it possible to choose the color of materials.

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Zoos, etc.: Hats or caps are important for everybody in bright, expansive places like zoos and theme parks, but more so for achromats. Eye fatigue can set in rapidly. Having a monocular handy (or, for younger kids, perhaps a lightweight pair of binoculars) can make a big difference in being able to see at least some of what others are seeing. Good ideas for visually impaired kids include petting zoos, indoor aquariums, dolphin petting pools, hands-on science exhibits, tunnel rides, nocturnal animal enclosures, insect zoos, and the enclosures where you find those creatures that are really hard not to see, like elephants and giraffes.

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One of the advantages for folks who can drive is being able to go around with many items they may need when away from home – everything from tissues to rain gear. People who don’t drive have to resort to backpacks or tote bags, and some opt for clothing with lots of pockets. Visually impaired persons tend to have even more things to carry around with them than the average walker. Achromats may need to carry 2 or more kinds of tinted lenses, a magnifier, a miniscope, and a visor or foldable hat. Coats, jackets or “cargo pants” that have roomy, secure pockets can serve this purpose, but some folks choose to wear multi-pocketed vests, such as are often on display during the summer months. These may be called ranger vests, photojournalist’s vests, fishing vests, or trail vests.

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Some “walking sticks” are quite elegant. They have a long history, especially in Great Britain, and they have many uses. Partially sighted walkers have even more uses for the time-honored walking stick – for example, as occasionally needed mobility aids for negotiating steps, curbs, or irregular walking surfaces. Usually available in wood or metal, these are not to be confused with orthopedic canes (although orthopedic canes have also been used by some networkers). You might want to check out two web sites: House of Canes <<http://www.houseofcanes.com/>> and Canes & Such <<http://www.canesandsuch.com/>>.

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The parasol is back in vogue, due to the growing concern about UV protection. Parasols are usually made of light colored fabrics, but they can be obtained in dark colors, as can an ever growing variety of head coverings featuring hoods, brims, or visors that are available for persons who are seeking to protect their eyes and their skin from the sun’s rays. Achromats, with their unique need for light protection for the eyes, can benefit from these fashion trends that have developed in connection with health consciousness.

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On Greyhound Bus Lines (1-800-231-2222), two people can travel for the price of one, if one of them has a disability and states that the other person is his or her

travelling companion. Amtrak (1-800-872-7245) offers a discount to passengers with disabilities, whether or not one is travelling with a companion (you can phone Amtrak and request a free large print copy of “A Guide to Amtrak Services for Passengers with Disabilities”). And local and regional transportation systems offer substantial discounts for passengers with disabilities. Certification of vision impairment is required for all such discounts.

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On the website for the organization known as NOAH (National Organization for Albinism and Hyperpigmentation), there is a webboard for drivers who use bioptics <<http://www.albinism.org/webboard/driving/>>. Some of our network members who drive using a bioptic report that they have found this webboard very helpful, and some of them have, from time to time, contributed information to this webboard from their own experiences. If you have an interest in this subject and want to check out the information on this webboard, you need to be aware that the comments that are posted come from persons with various different levels of visual impairment. For example, many persons with albinism have considerably better vision in daylight than most persons with achromatopsia.

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Coloring with fragrance: Did you know that you can buy colored markers which are scented? A green marker might smell like a pine tree, a red marker might have a cherry scent, etc. These markers might enhance some art activities for colorblind kids. Crayola scented markers can be found in many stores.

TELEPHONE RESOURCES

Many services and accommodations for visually impaired persons vary from state to state. For example, phone companies offer special accommodations to sight impaired customers. In California these services include free directory assistance, automatic (speed) dialing, and the long-term loan of a push-button phone that has large numbers, re-dial, and other features. Check with your local agencies for the visually impaired and your local and long distance provider to learn what’s available in your area. Special services and accommodations usually require certification of vision impairment. Obtaining free directory assistance may only require a written statement by the visually impaired customer regarding the fact that the small print in telephone directories represents a hardship.

HAILSTONES AND HALIBUT BONES

Mary O’Neill’s book of poetry about all the colors of the spectrum, is a children’s classic. It makes a lovely gift for any child, but it can have special meaning for a child without color vision. Following are some excerpts from the section about the color red:

“Red is a sunburn spot on your nose. Sometimes red is a red, red rose. Red squiggles out when you cut your hand. Red is a brick and a rubber band. Red is a hotness you get inside, when you’re embarrassed and want to hide. Fire-cracker, fire engine, fire-flicker red. And, when you’re angry, red runs through your head. Red is a lipstick. Red is a shout. Red is a signal that says, “Watch out!”

This book is a Bantam paperback, costs \$7.95, and can be found – or ordered – at many bookstores.