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Albinism: Genetic Condition, NOT a Curse

Outline

Abstract

Imagine a large, clear colour photo printed on the front page of the newspaper. Now imagine that someone in the photo is wearing a golf shirt with some lettering or a logo on the shirt pocket. You look at the photo up close, trying to read the lettering printed on the shirt. To your dismay, you can't quite make it out. Overall, the picture is not blurry. Yet when you look at the small details, you just can't make them out. If you were looking at the original photo the paper used, however, you could make out those words. This is how an albino person looks at the world.

Coping and dealing with albinism has run the gamut from being militant in one's racial conviction to wearing dark make-up and hair colour or surgically altering one's appearance to pass for white. Some may argue that these may not be coping at all. Albinism is generally present since the creation, from the earliest stages of development in the uterus. Albinism is an old, old "race." It has been recorded in every part of the world, in every era, in every culture

known to humans. It has been said that Albinism may represent in fact the oldest recorded genetic condition.

What is Albinism

The word “albinism” refers to a group of inherited conditions. People with albinism have little or no pigment in their eyes, skin, or hair. They have inherited altered genes that do not make the usual amounts of a pigment called melanin. One person in 17,000 in the U.S.A. has some type of albinism. Albinism affects people from all races. Most children with albinism are born to parents who have normal hair and eye colour for their ethnic backgrounds. Sometimes people do not recognize that they have albinism. A common myth is that people with albinism have red eyes. In fact there are different types of albinism and the amount of pigment in the eyes varies. Although some individuals with albinism have reddish or violet eyes, most have blue eyes. Some have hazel or brown eyes. However, all forms of albinism are associated with vision problems.

Visual Problems: People with albinism always have problems with vision (not correctable with eyeglasses) and many have low vision. The degree of vision impairment varies with the different types of albinism and many people with albinism are “legally blind,” but most use their vision for many tasks including reading and do not use Braille. Some people with albinism have sufficient vision to drive a car. Vision problems in albinism result from abnormal development of the retina and abnormal patterns of nerve connections between the eye and the brain. It is the presence of these eye problems that defines the diagnosis of albinism. Therefore the main test for albinism is simply an eye examination.

Skin Problems: While most people with albinism are fair in complexion, skin or hair colour is not diagnostic of albinism. People with many types of albinism need to take precautions to avoid damage to the skin caused by the sun such as wearing sunscreen lotions, hats and sun-protective clothing.

Reasons for Albinism

As discussed earlier Albinism is a genetic condition. The genes for OCA are located on “autosomal” chromosomes. Autosomes are the chromosomes that contain genes for our general body characteristics, contrasted to the sex chromosomes. We normally have two copies of these chromosomes and the genes on them – one inherited from our father, the other inherited from our mother. Neither of these gene copies is functional in people with albinism. However, albinism is a “**recessive trait**”, so even if only one of the two copies of the OCA gene is functional, a person can make pigment, but will carry the albinism trait. Both parents must carry a defective OCA gene to have a child with albinism. When both parents carry the defective gene (and neither parent has albinism) there is a one in four chance at each pregnancy that the baby will be born with albinism. This type of inheritance is called “autosomal recessive” inheritance.

Types of Albinism: While most people with albinism have very light skin and hair, not all do. Oculocutaneous (pronounced ock-you-low-kew-TAIN-ee-us) albinism (OCA) involves the eyes, hair and skin. Ocular albinism (OA), which is much less common, involves primarily the eyes, while skin and hair may appear similar or slightly lighter than that of other family members.

Over the years, researchers have used various systems for classifying oculocutaneous albinism. In general, these systems contrasted types of albinism having almost no pigmentation with types having slight pigmentation. In less pigmented types of albinism, hair and skin are cream-colored and vision is often in the range of 20/200. In types with slight pigmentation, hair appears more yellow or red-tinged and vision may be better. Early descriptions of albinism called these main categories of albinism “complete” and “incomplete” albinism. Later researchers used a test that involved plucking a hair root and seeing if it would make pigment in a test tube. This test separated “ty-neg” (no pigment) from “ty-pos” (some pigment). Further research showed that this test was inconsistent and added little information to the clinical exam.

Recent research has used analysis of DNA, the chemical that encodes genetic information, to arrive at a more precise classification system for albinism. Four forms of OCA are now recognized – OCA1, OCA2, OCA3 and OCA4; some are further divided into subtypes.

Oculocutaneous albinism type 1 (OCA1 or tyrosinase-related albinism) results from a genetic defect in an enzyme called tyrosinase (hence 'ty' above). This enzyme helps the body to change the amino acid tyrosine into pigment. (An amino acid is a "building block" of protein.) There are two subtypes of OCA1. In OCA1A, the enzyme is inactive and no melanin is produced, leading to white hair and very light skin. In OCA1B, the enzyme is minimally active and a small amount of melanin is produced, leading to hair that may darken to blond, yellow/orange or even light brown, as well as slightly more pigment in the skin.

Oculocutaneous albinism type 2 (OCA2 or P gene albinism) results from a genetic defect in the P protein that helps the tyrosinase enzyme to function. Individuals with OCA2 make a minimal amount of melanin pigment and can have hair color ranging from very light blond to brown.

Oculocutaneous albinism type 3 (OCA3) is rarely described and results from a genetic defect in TYRP1, a protein related to tyrosinase. Individuals with OCA3 can have substantial pigment.

Oculocutaneous albinism type 4 (OCA4) results from a genetic defect in the SLC45A2 protein that helps the tyrosinase enzyme to function. Individuals with OCA4 make a minimal amount of melanin pigment similar to persons with OCA2.

Researchers have also identified several other genes that result in albinism with other features. One group of these includes at least eight genes leading to Hermansky-Pudlak Syndrome (HPS). In addition to albinism, HPS is associated with bleeding problems and bruising. Some forms are also associated with lung and bowel disease. HPS is a less common form of

albinism but should be suspected if a person with albinism shows unusual bruising or bleeding.

Ocular albinism (OA1) is caused by a genetic defect of the GPR143 gene that plays a signalling role that is especially important to pigmentation in the eye. OA1 follows a simpler pattern of inheritance because the gene for OA1 is on the X chromosome. Females have two copies of the X chromosome while males have only one copy (and a Y chromosome that makes them male). To have ocular albinism, a male only needs to inherit one defective copy of the gene for ocular albinism from his carrier mother. Therefore almost all of the people with OA1 are males. Indeed, parents should be suspicious if a female child is said to have ocular albinism.

For couples who have not had a child with albinism, there is no simple test to determine whether a person carries a defective gene for albinism. Researchers have analyzed the DNA of many people with albinism and found the changes that cause albinism, but these changes are not always in exactly the same place, even for a given type of albinism. Moreover, many of the tests do not find all possible changes. Therefore, the tests for the defective gene may be inconclusive.

If parents have had a child with albinism previously, and if that affected child has had a confirmed diagnosis by DNA analysis, there is a way to test in subsequent pregnancies to see if the foetus has albinism. The test uses either amniocentesis (placing a needle into the uterus to draw off fluid) or chronic villous sampling (CVS). Cells in the fluid are examined to see if they have an albinism gene from each parent.

For specific information and genetic testing, one should seek the advice of a qualified geneticist or genetic counsellor. The American College of Medical Genetics and the National Society of Genetic Counsellors maintain a referral list. Those considering prenatal testing should be made aware that people with albinism usually adapt quite well to their disabilities and lead very fulfilling lives.

Effects

In the United States, most people with albinism live normal life spans and have the same types of general medical problems as the rest of the population. The lives of people with Hermansky-Pudlak Syndrome can be shortened by lung disease or other problems. Other conditions include Chediak-Higashi and Griscelli Syndrome.

In tropical countries, those who do not use skin protection may develop life-threatening skin cancers. If they use appropriate skin protection, such as sunscreen lotions rated 20 SPF or higher and opaque clothing, people with albinism can enjoy outdoor activities even in summer.

People with albinism are at risk of isolation because the condition is often misunderstood. Social stigmatization can occur, especially within communities of colour, where the race or paternity of a person with albinism may be questioned. Families and schools must make an effort not to exclude children with albinism from group activities. Contact with others with albinism or who have albinism in their families or communities is most helpful.

Assistance for people with Albinism

There are dozens of different low vision aids to help improve the lives of people with albinism. Each aid has its own advantages and limitations. To get the most from a low vision aid, it's important to see and understand what that device can and cannot do. Since different aids help with different tasks, the person with low vision will likely choose a mix of different low vision aids to accomplish all of his or her goals.

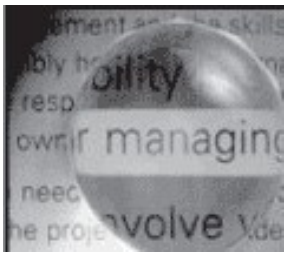
Tasks, goals, and technology change over time. Whether an individual has used low vision aids for years or just for a short time, the individual should periodically revaluate the low vision aids he or she uses.

Glasses: Glasses cannot fix low vision. Eyeglasses are not considered a low vision device unless they have a high reading “add” or contrast enhancement tint, but they deserve special mention here since glasses ensure that the person's eyes see the clearest image possible and can easily focus at close distances. It is important for babies and young children with albinism to wear glasses to correct refractive error if they are nearsighted, farsighted, or have astigmatism. If a child with albinism who needs glasses doesn't wear them, that child may tire easily and quickly give up doing certain tasks, and may even affect their development.

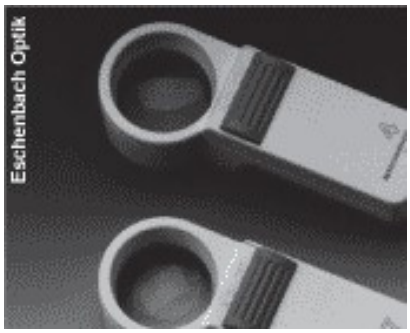
Short viewing distances: Beginning in infancy, people with albinism instinctively hold things closer to their eyes to see them better. Up until the age of five or six, this technique sufficiently compensates for low vision because books for young children already feature large print, or they can sit closer to the TV. As a result of using this desire of moving closer to what needs to be seen, children this age generally do not need low vision aids. Looking closely at objects does not hurt the individual’s eyes, and this strategy continues throughout life.

Near vision aids: Dome magnifiers, reading glasses, hand-held and stand magnifiers, and microscopes are near vision aids that help people with albinism read, look at pictures, diagrams, and maps, and accomplish other tasks that require seeing small details up close.

Dome magnifiers [Figure 1] are one of the easiest aids to use and arguably the most important for those with albinism. Despite their seemingly modest ability to magnify print (1.7 to 2.2 times), in combination with a short viewing distance many reading tasks can be easily accomplished even in the primary grades of school. These magnifiers are also known as bright field magnifiers, paper weight magnifiers, and Visolett magnifiers.



Reading glasses help the user focus on text or other objects while holding the object close to the user's eyes. Reading glasses allow the widest field of view for reading compared to other aids. Hand-held and stand magnifiers [Figure 2] enlarge close-up images, allowing the user to see small print and images at greater distances from the user's eyes. There are many different styles and sizes of magnifiers useful for people with albinism.



Microscopes help people see smaller details than magnifiers produce. Microscopes enlarge close-up objects the same way telescopes enlarge far-away objects. Some telescopes and bioptic telescopes, designed for distance vision, also allow the user to refocus the scope for up-close use.

Distance vision aids: Telescopes can help people with low vision improve distance vision. Distance vision includes seeing a chalkboard in a classroom, seeing a menu board at a fast-food restaurant, or seeing the stage at a concert or the action at a sporting event. The two specifications that differentiate telescopes from one another are the magnification power and the field of view. Magnification power indicates how much larger an image appears through a telescope compared to how large it appears to the naked eye. Typical magnification powers for low vision telescopes range from 2x to 8x. When a person with 20/100 vision uses a 4x telescope, that person

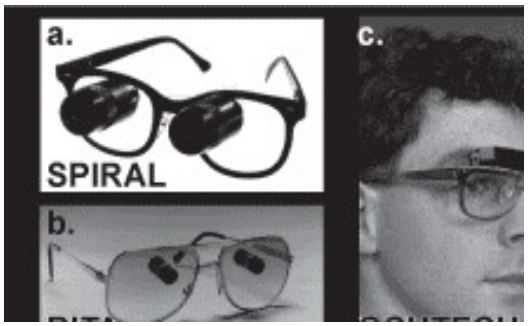
theoretically sees an image 4 times bigger than normal and, therefore, sees the same details a person with 20/25 vision sees. In general, the greater the magnification power of a telescope, the more fine detail the person with low vision will see using the telescope. However, as magnifying power increases, the field of view, or the size of the area enlarged by the telescope, generally decreases.

One of the most common low vision aids is the hand-held telescope, also called a monocular [Figure 3]. Hand-held telescopes come in a wide variety of sizes, magnification powers and prices. Hand held telescopes work best to quickly view a distant object, such as reading a sign or locating an object. Clip-on telescopes allow the user to slip the telescope over his or her glasses for hands-free use [Figure 4]. The clip-on telescope works well for the person who needs to use the telescope for extended time periods, such as for watching TV, a movie, or a live stage performance. The user can remove the clip-on telescope and use it as a hand-held telescope for quick viewing tasks.

A bioptic is a special pair of glasses with a telescope permanently mounted in the glasses' lens. While looking straight ahead, a bioptic user sees a normal, unmagnified image through the glasses. Then by dipping one's head slightly, the bioptic user instantly sees a magnified image through the telescope. This "bi-optical" system allows the user to rapidly switch between a normal view and a magnified view without ever using his or her hands. In some areas, some people with low vision can use bioptics to drive. A person with albinism can also use bioptics in the same situations he or she might use a hand-held telescope, such as seeing a classroom chalkboard. Hands-free use can help older students take notes in class more quickly and easily. Some bioptic users

prefer telescopes in both eyes, while many users with albinism choose to have only one mounted telescope for their dominant eye.

One of the most common and simplest bioptic designs is from Designs for Vision [Figure 5a]. The Designs for Vision bioptic uses a telescope that extends about 3/4 to 1-1/2 inches from the front of the glasses' lens and is 1/2 to 1 inch wide. Designs for Vision offers both fixed focus telescopes in a black metal housing and slightly larger focusable telescopes in a black plastic housing. They also make a bioptic telescope with a clear plastic housing to improve the bioptic's appearance.



To make using a bioptic more discreet, Edwards Optical created the BITA system [Figure 5b]. This bioptic uses telescopes that are only 1/2 to 3/4 inches long and about as wide as a pencil. The telescopes also extend behind the glasses lens, instead of in front of it. The BITA design is much less noticeable than traditional bioptic designs, but the small telescopes also produce a smaller field of view.

Ocutech manufactures a bioptic system that mounts the telescope across the glasses' bridge [Figure 5c]. This approach creates a wider field of view at higher magnification powers compared to conventional bioptic designs, plus offers a different appearance. Ocutech also manufactures an auto-focus bioptic, which automatically changes the focus as the user looks at different objects. The auto-focus bioptic is bulkier and more expensive than other bioptics.

Electronic aids: When traditional optical low vision aids don't help accomplish a task, electronic aids might help. Closed-Circuit Television (CCTV) systems help people who need greater magnification than reading glasses, magnifiers, and microscopes provide [Figure 6]. CCTV systems also allow the user to adjust the size, brightness, and contrast of the magnified image to best match the user's vision. The user can even read white letters on a black background to decrease glare. CCTV systems have historically been far more expensive and far less portable than other near vision aids. Newer CCTV designs, however, such as the MagniCam®, are now smaller, more versatile, more portable, and less expensive then previous CCTV systems. Other electronic aids help users see far away objects.

Computers and Software: Desktop and laptop computers have advanced the ability of those with albinism to learn, communicate, and follow their chosen careers. Monitors are available in various sizes to suit an individual's magnification needs.

Magnification can be changed in common software such as Microsoft Outlook, Excel, and Word. This can be done in a variety of ways such as directly changing it on the toolbar, or by going to the toolbar clicking on the "View" option. Another method that is to hold down the Ctrl key on the keyboard and then turn the small wheel in the middle of the mouse away from you or towards you to change the print size on many word processing and HTML screens.

Specialized software is also available such as Zoomer, ZoomText, Kurzweil, or Big Shot. These have many advanced features such as magnification of specific areas on a page, or a split screen option.

Contrast enhancement aids: For people with albinism, bigger isn't always better. Increasing the contrast of text is often more effective than increasing the size of text. Black felt-tipped pens and dark lined paper can make writing easier for some people with low vision. #1 pencils (as opposed to traditional #2 pencils) and bright-colored chalk can also help students with albinism.

Writing guides, which are templates with open areas where one writes, can help people write in straight lines, or with tasks like writing checks. Colored filters can make it easier to see certain colors. For example, a yellow filter can make light blue letters appear darker and easier to read. Lighting plays a major role in how well a person with low vision sees. Experimenting with different types and brightness of light, as well as the location of the light, can make tasks a lot easier. Keep in mind that too much light and too little light can both cause problems

Finding low vision aids: An optometrist or ophthalmologist who specializes in low vision evaluates the individual's vision, and then recommends specific aids based on the patient's needs and goals. Low vision clinics usually have a variety of aids on site to see and try as well as specialists trained to help the user get the most out of the new aid. Since everyone's vision and goals are different, it's important to try out as many aids as possible before choosing. Often, the newest or most expensive aid is not the best choice. If a low vision specialist only prescribes products from one manufacturer, the patient may want to visit another clinic featuring different products before buying an aid, especially when considering expensive aids, such as bioptics or CCTV systems.

There are catalogue companies that sell some low vision aids, such as sunglasses, magnifiers, and hand-held telescopes. If the individual knows exactly what he or she needs, buying these products through a catalogue may cost less than buying from a low vision specialist. Other aids, however, such as bioptics, require a custom fitting and prescription from an eye doctor.

Sun Protection: People with albinism can enjoy the outdoors by limiting their exposure to sunlight, wearing appropriate hats and clothing, and using sunscreens diligently. However, the task of preventing damage to the skin over a lifetime is a difficult one. Preventing sunburn is important, but not sufficient to prevent sun-damaged skin.

It is an invisible part of the light spectrum of the sun, the ultraviolet light that damages skin. The shorter wavelength ultraviolet light, UVB, has a much bigger role in causing sunburn than the longer wavelength UVA. Manufacturers made sunscreens to block UVB and not UVA, so normally pigmented people could tan without burning. Newer research suggests than UVA, since it penetrates more deeply, may cause skin cancer and premature "aging" of the skin. Now sunscreen manufacturers label sunscreens as "broad spectrum," which means they block both UVB and UVA.

Sunscreens: People with albinism should use sunscreens labeled SPF of 20 to 30. "SPF" means "sun protection factor." This number comes from a standard test in a laboratory. The test measures the time it takes people wearing a standard amount of sunscreen to sunburn under a standard ultraviolet lamp, compared to the time with no sunscreen. In theory, if a person could stay in the sun 10 minutes without burning with no sunscreen, he or she could wear an SPF 20 sunscreen and stay in the sun 20 times 10 minutes or 200 minutes before burning.

The current SPF system focuses on sun burning, and mainly measures UVB protection. Since UVA penetrates the skin deeply but has much less energy for burning, researchers have become concerned that sunscreens could create a false sense of security, and allow people to sustain damage to their skin. The FDA is considering adding a second rating system for UVA protection.

In actual use, a sunscreen may not provide as much sunburn protection as expected from its SPF rating, largely because people do not apply enough. Most people apply about half as much as the amount used for SPF tests. Full body protection for an adult requires nine portions of one-half teaspoon each, or a total of about an ounce. For sunscreens that come in four-ounce bottles, this amounts to one-fourth of the bottle per application.

People often develop blotchy sunburn from spots missed with sunscreen lotions. Since sunburn develops two to four hours after the sun exposure, and

sunburn may peak in intensity as long as 24 hours after sun exposure, it is important to apply the sunscreen systematically. Don't forget the tops of ears, and the backs of arms and legs. Bicyclists need to protect the low back, where shirts often ride up.

It helps to apply the sunscreen one-half hour before going into the sun, since some of the screening chemicals bind to the skin. Remember that rubbing with towels or friction from clothing such as between the legs can remove protection. Simple immersion in water does not remove protection of sunscreens marked "very water resistant" (80 minutes in water by FDA definition) or "water resistant" (40 minutes in water). The FDA has proposed dropping the term "waterproof." Most people, especially children, do more in the water than just immerse themselves. They may rub off some of the sunscreen, and the old warning to "reapply after swimming" may be wise.

Which sunscreen should people with albinism use? It is difficult to recommend a specific sunscreen, because formulations change. Consumer Reports publishes a review of sunscreens every couple of years, and their evaluations show that labels of SPF and water resistance are usually accurate. In the Consumer Reports cost comparisons, some products cost as much as six times more than others. Some manufacturers are now marketing sunscreen in bigger bottles at much less cost.

Should people with albinism go for the highest SPF available? Using sunscreens with SPF higher than 30 offers little benefit, and more concentrated chemicals might be more likely to irritate or cause an allergic rash. The FDA has proposed limiting the number to 30. In my view, it would seem wise to use any number of products that have an SPF of 20 to 30. Paying a high price for a few more SPF points is probably a waste. Sunscreens with SPF in this range will include oxybenzone or a similar chemical that makes them "broad spectrum." Oxybenzone blocks some UVA light, but not the longest UVA wavelengths. Another chemical, Parsol 1789 or avobenzone, provides "broader" spectrum coverage and might help people with fair skin prevent sun damage. Unlike oxybenzone, avobenzone is

patented and expensive, and therefore it is not used in many products (Shade UVA Guard is one). Titanium and zinc oxide screens provide very broad spectrum coverage and, if you have the patience to rub them in, may serve very well with little risk of allergic reactions.

Individual preferences will determine the best product, and factors such as consistency and smell may play a big part in the choice. Some products have an alcohol base that goes on easily but may sting and dry the skin. Others have a lotion base like baby lotion. Oil-based lotions are more likely to aggravate acne. Dry lotions contain plastic like polymers that make them very water resistant. Although they don't feel oily, they can make your skin feel sealed.

Spray sunscreens are very sticky and hard to apply evenly. Self-tanning lotions contain chemicals that develop into a brownish color in sunlight. While this product might seem attractive to people with albinism, in actual use the skin may look mottled or dirty because the chemicals accumulate in skin lines and pores.

Problems with Sunscreens: Sunscreens can cause troublesome allergies. One sunscreen ingredient, PABA, causes allergic reactions frequently, and now all products are "PABA free." It is possible to develop allergies to any of the usual UV-blocking chemicals or even to "inactive" ingredients of the lotion or fragrance. People who develop allergic reactions should seek advice from a dermatologist. A day before you intend to use a new sunscreen, apply it to a small area on one arm and wait to see if any itching or rash occurs. To prevent allergic reactions to sunscreen, it might help to wash sunscreens off at night.

Some sunscreens are marked for "sensitive skin." Some of these contain the usual UV-blocking chemicals and may have no special properties to prevent allergy. Others contain titanium dioxide, an inert (chemically inactive) substance often used as a white pigment in paint. To work as effectively as the active chemicals, titanium dioxide has to be thick enough to be visible;

that is, it does not "rub in" easily and leaves a whitish color. Some sunscreens combine an opaque pigment like titanium dioxide with bright colors to apply across nose, cheeks, or ears like face paint, and these might serve to augment regular sunscreens for children.

Sunscreens also include the warning "consult physician before using on babies under six months age." The concern is that babies will absorb more of the chemicals through their skin, and the chemicals may cause unforeseen problems. It is unlikely that research on the safety of sunscreens for babies will ever be done. Sunscreens containing inert blocks like titanium dioxide are probably safe, but babies should not have direct sun exposure in any case.

Avoiding Harmful Rays: Even for older children and adults with albinism, it is important not to rely on sunscreens, and to limit exposure to the sun. Most ultraviolet rays come between 10 a.m. and 2 p.m. Standard Time, or 11 a.m. and 3 p.m. Daylight Savings Time. Planning outdoor activities for morning or evening is the single most important measure for people with albinism to avoid sun damage.

The National Weather Forecasting Service gives predictions of UV risk on a scale of 1 (low) to 10 (high) for many communities. They base these predictions on the angle of the sun at noon at that place and date, the altitude, and the predicted cloud cover. These are how these factors come into play:

Latitude: A person who can tolerate one hour of sun in Florida without burning can tolerate two hours of sun in New Jersey under the same conditions.
Season: The greatest intensity of ultraviolet light occurs at the summer solstice, about June 22. May 1 has as much intensity as August 15.
Altitude: Each 1000-foot increase in altitude adds 4% to the intensity of the sunburning rays. The intensity of sunlight at 5000 feet is about 20% greater than at sea level.
Weather: A bright day with a thin cloud cover has 60 to 80% of the ultraviolet rays that are present on a clear day. Clouds can cool and give a false impression that there is little risk of sunburn.

In addition to the UV forecast, consider your surroundings. Sand reflects 25% or more of ultraviolet rays so that it is possible to get sunburned while sitting in the shade on a beach. Fresh snow reflects 70 to 90% of ultraviolet rays. Water can also reflect UV rays. Reflected light may burn areas which are usually shaded, such as those under the nose or chin. As much as 96% of ultraviolet rays can penetrate clear water.

Clothing and Hats: Clothing is important. Up to 50% of the ultraviolet rays can go through wet cotton tee shirts worn for swimming. Colored clothing and denser-woven clothing allow less light penetration. Several manufacturers are promoting densely woven "breathable" synthetic fiber clothes for sun protection. These allow protection with long sleeves in warm weather. With time this type of clothing may become less expensive and more common.

Hats are essential. Hats with brims at least 3 inches wide all the way around are best for protecting the face, ears, and neck. Hats with narrower brims, such as tennis hats, at least protect the ears.

Sun Damage:

What to Watch For:

Ultraviolet light over many years creates "actinic" changes in skin, and increases the risk of skin cancers. These problems are a cause for concern, but not alarm.

Actinic changes include fine wrinkling, yellowish discoloration, leathery thickening, and dilated capillaries branches, which appear as clusters of tiny red streaks on the skin surface.

The best treatment for these actinic changes is prevention. It is difficult to reverse these changes with chemical or surgical treatments.

One treatment for actinic changes is application of tretinoin (Retin A, Renova, other brands). Tretinoin cream or ointment can reverse some of these changes, at least temporarily. Tretinoin must be applied daily for months, and redness, irritation, and increased sun sensitivity are usual side effects. The

use of tretinoin for people with albinism has not been studied, but original studies were performed on albino hairless mice, and it seems likely that tretinoin could work for people with albinism who can bear the side effects. Tretinoin is a prescription medicine, and should be used only under a physician's supervision.

UV damage can also result in actinic keratoses (carrot-TOE-seez). These are small reddish or tan thickenings with an irregular rough or scaly surface. These develop mainly on the face and the backs of hands, and especially on bald scalps. Over years, some of these can develop into squamous cell skin cancers if left untreated. Physicians can remove small numbers of actinic keratoses by applying liquid nitrogen to freeze them. For more widespread actinic keratoses, dermatologists may prescribe an irritant ointment such as 5-fluorouracil, which creates redness and peeling.

Most skin cancers are basal cell cancers. These look like dome-shaped pink bumps, with "a raised pearly edge and visible red capillary blood vessels. They may show scaling or scabbing in the center. In most instances, basal cell skin cancers can be removed with relatively simple surgery, and they do not tend to spread through the system.

Squamous cell cancers are less common among people with albinism in the United States, but can be more troublesome. They appear as firm red bumps that go on to form an ulcer surrounded by a firm border. They usually occur on the scalp, ears, back of the hands, or the lower lip. If treated early, squamous cell skin cancers also can be cured with surgery, but techniques must be used to be sure all of the cancer is removed. Squamous cell cancers starting on the skin spread through the system in about three percent of cases. If they spread, they are life-threatening, since therapy is limited.

People with albinism can prevent serious problems with skin cancers by taking protection against the sun, by examining their own skin for long-lasting "irritated" spots, and by getting "regular physician exams" of skin as adults.

Albinism Communities: A wide range of help is available all over the world for people affected with albinism and for teachers and Parents of effected child. A few known ones are listed below:-

ABLEDATA is a federally funded project whose primary mission is to provide information on assistive technology and rehabilitation equipment available from domestic and international sources to consumers, organizations, professionals, and caregivers within the United States.

Natalie's **Way Foundation** was formed in the honour of Natalie Stasi, who, in 1997, was born with Oculocutaneous Albinism (OCA). The foundation raises money through a golf tournament. The foundation's goal is to donate money for research in albinism and for children with eye disorders.

The Nystagmus Network (UK) provides support for those with nystagmus, and the parents and teachers of children with nystagmus. This site aims to provide information about this eye condition, and some of the pages are of general interest to those concerned with visual impairment.

Positive Exposure: Rick Guidotti's Positive Exposure is a unique partnership between visual arts, genetics and education.

Bioptic driving: Information on bioptic driving with webboard.

Information for Parents and Teachers of albino children

The First Influence: A Disability or Not: Neither the general public nor those with the condition agree about whether to identify albinism as a disability. This ambiguity creates a problem in the language used to talk about albinism. It also makes it difficult for those with albinism to identify themselves as a group. In many ways, albinism is unique condition. That uniqueness, however, leads to separateness and isolation for many people. Social attitudes toward albinism are often similar to those experienced by other

disability and minority groups. These attitudes include a lack of understanding, fear of the unknown, and prejudice based on appearance.

The Americans with Disabilities Act defines disability with respect to an individual as "a physical or mental impairment that substantially limits one or more of the major life activities of such individual; a record of such an impairment; or being regarded as having such an impairment." Since albinism involves a visual impairment, some people consider it a disability. One definition of handicap is "the obstacles a person encounters in the pursuit of goals in real life, no matter what their source." Thus a person with a disability may or may not be handicapped in pursuing the life they want to live.

The identification of albinism as a disability is complicated by the concept of legal blindness. In the United States, a person is legally blind if his or her vision cannot be corrected with glasses or contacts to better than 20/200 in his or her better eye. By this standard some with albinism fit the legal category of visual impairment and some do not. Yet, in spite of varying visual acuity, many of the problems experienced by those with albinism remain similar.

The Second Influence: Physical Appearance: The first aspect of albinism which most people notice is the person's unusual appearance. The white hair and skin of oculocutaneous albinism is a powerful factor from the moment of birth. The new baby will often be much lighter in colour than any family member. In non-white races the coloring of the baby with albinism is a dramatic contrast to the family and community. Colour is a highly charged characteristic in our culture now and historically. Strangers will often make unwanted and unkind comments about the appearance of a child.

Beyond colour, a child's eyes may be moving rapidly and not focusing together. The child may have to squint, tilt his or her head, and hold things close in order to see. Children with albinism often use glasses and optical aides to enhance their vision. Therefore the child with albinism often feels isolated not only in physical appearance but also in the conduct of everyday life.

This perception of being different can lead to an immense effort to act as much like "normal" as possible. A person with albinism can feel a lot of pressure, both from themselves and from other people, to minimize the differences albinism causes. This effort can result in a great deal of stress for a person continually trying to maximize visual ability. The pressure can even lead a person with albinism to deny entirely that he or she has albinism, thereby losing touch with a very important aspect of one's identity.

Family and close friends can counter this isolation and denial. Being prized and valued as a whole person is the foundation for a lifetime of self-esteem and inner strength. This prizing must include an honest acknowledgment and acceptance of the condition of albinism. It is vitally important that families can freely discuss the impact of albinism in each of their lives.

The Third Influence: Language, Myth, And Stereotype: Language can shape ideas and create reality. The word "albino" is commonly used in many languages including English. Some people are comfortable with the word and prefer being called an albino. However, people often use the word "albino" in hurtful ways. Many feel it is dehumanizing to refer to a person in terms of a condition. Although slightly cumbersome, the terms "person with albinism" and "people with albinism" put the person first and the condition second.

Teasing and name-calling are other ways in which language can be very dehumanizing. Almost all children face teasing during their school years and they need to develop positive coping strategies. Parents, teachers and increased education about albinism can help with this problem.
Throughout the world, people have misconceptions about albinism, ranging from notions that people with albinism have magical powers to the belief they are retarded. Among African-Americans, a common myth is that babies with albinism result from a union between an African-American woman and a Caucasian man. Another common myth is that people with albinism must have red eyes. People with albinism usually have blue or gray eyes which

sometimes appear reddish in certain types of light. Sometimes, myths are so widespread even the person with albinism believes them.

The media, including literature and film, have contributed to stereotypes of albinism. The character with albinism is often portrayed as villainous, deviant, supernatural or sadistic. Also some news reports and encyclopaedia articles have included false or incomplete information about albinism. It is difficult for the public to know what is true and untrue about albinism.

The Fourth Influence: The Family -It is vital that the family have accurate information about albinism. New parents need support and time to understand the condition of their child. Parents and family members may need to face some unpleasant stereotypes they have learned about albinism. Siblings need to understand why their brother or sister looks different and why they seem to be getting so much attention. There is no single force greater than the family in helping a child understand and accept himself or herself.

The Emotional Component of Albinism: Along with the external influences of society, every person has a vital and essential emotional response to their personal experiences with albinism. These personal responses shape who we are and how we adapt to albinism. A strong emotional response is a normal part of living, growing, and intellectual development. Suppressed emotions often turn inward and cause stress, depression and physical maladies.

Emotional responses to albinism will occur throughout life because of the many challenges and frustrations the condition presents and the many societal influences already mentioned. It is very important to develop healthy ways to express and integrate these emotions. First, it is necessary to recognize feelings and determine their source. Parents can help children label the feeling the child feels, then help the child connect that feeling to a specific reason or event. For example, a parent might say, "I know you're sad because you have to put on sunscreen before you go swimming." Then the parent can help the child "do" something with the emotion such as to talk about it, play,

yell, run, cry-whatever physical outlet will release the feeling. This validation and release are essential in processing an emotion.

Adults with albinism can go through this process by having friends and family listen to their frustrating, discouraging, or proud experiences. Some ways to release an emotional charge are physical activity, taking action in the form of education or advocacy, journal and letter writing, or doing something nurturing for yourself. Sometimes professional help from a therapist or counsellor can assist a person work through the highly charged issues of albinism. Coping with albinism often isn't easy. However, working though the issues albinism causes not only leads to great personal satisfaction, but also to a greater understanding of human kind.

Coping with Teasing and Name-Calling: Many parents said that teasing, insensitivity, and ignorance about albinism were their greatest challenges. The young people reported being called names like "Whitey," "Snow White," "Casper," "Four Eyes," "Blind Eyes," "Grandma," and "Grandpa." They were asked why their heads shake, did they pour bleach all over themselves, and other embarrassing questions. They also reported being excluded from games because they weren't "good enough."

Understanding this behaviour is one of the first steps in learning to deal with these experiences yourself or to help your child cope with albinism.
What makes children (and others) tease and engage in name-calling? What causes the crude remarks, the callous behaviour and the insensitive questions?

One reason may be fear. Ancient people believed that to name something was to control it. We know today that people can be controlled by repetitious name-calling when they begin to believe the name, and act accordingly.

Other reasons for name-calling, teasing and insensitivity are lack of knowledge, curiosity, a genuine desire to learn, and an inability to express questions constructively.

Some people tease in order to get to know another person better or to express affection. This kind of teasing usually isn't hurtful. However, if the person being teased doesn't have a positive self-image, and isn't comfortable with albinism, even affectionate teasing can hurt.

First, develop a positive self image and a positive attitude towards albinism. Add to that a thorough knowledge of albinism itself and you'll find yourself becoming more comfortable when discussing it with others. The standard "dumb" questions can become opportunities for education if one has self-confidence.

Parents can help their children deal with the hurtful comments by encouraging them to share their experiences and their feelings about them. Parents can also help their children by exploring new and positive ways to respond to the teasing and name-calling in the future and by practicing through role playing.
Sometimes presenting information in class about albinism can reduce the amount of teasing and name-calling. Parents or the student can make the presentation, depending on the circumstances. Parents may need to educate the teachers first.

Schools should teach kids that hurtful behaviour towards people with any sort of difference is a form of discrimination. A series of lessons on disabilities can provide information about albinism in the context of a variety of disabilities and other differences. Videos, puppet shows such as Kids on the Block, and other materials can teach this lesson on a child's level. There are additional ways of dealing with teasing and name-calling, such as using humour.

Most students with albinism can participate in general education programs with appropriate support services. Since not all children with albinism have the same visual acuity, and since not all use their vision with the same efficiency, it is important to consider the individual abilities, needs and skill level of each child.

School systems must provide education in the "least restrictive environment" to children with disabilities. The program must meet the child's individual education needs. For most children with albinism, this environment is the "typical" local classroom setting, in which a teacher certified in the area of education of students with visual impairments instructs and facilitates approaches to allow the student to participate in all appropriate activities.

A Teacher of Visually Impaired (TVI) is a special education teacher with expertise in the education of students with visual impairment. To identify a TVI and to access special education services, parents of a child with albinism should contact their public school system's administrative office. This contact should occur well before kindergarten. In all states, the school system can provide information about preschool and early intervention supports and services. Beginning at age three, public schools must provide education and educationally related services to children whose physical, emotional, learning or sensory impairment constitutes an educational disability. For children with low vision, these services will include assessment of visual function (A Functional Visual Assessment), as well the educational and other assessments as appropriate. Suggestions to parents about ways to help their child is an important element of the assessment and evaluation process. This assessment paves the way for the child's education team to develop an Individual Education Plan (IEP). The educational placement is then determined by the team with consideration of the array of options which are available and/or need to be made available to the child

The Classroom Setting: The student, his or her parents, classroom teacher, TVI , and optometrist or ophthalmologist must work as a team. The team should consider seating, lighting, materials, social and emotional growth, the impact of visual impairment on development as well as the use of appropriate technology and optical aids.

The student, classroom teacher, and TVI should discuss classroom seating. Most students with a visual impairment will sit in the front row, but not all can see the blackboard even from there. Some students will not wish to sit up

front and should not be forced to do so, but should be provided with the information in other ways. The seating position should avoid glare from the side windows and overhead lights, and should avoid shadow. In a classroom with fluorescent lights, a seat between banks of lights is better than one directly under a bank.

The student may need to move to a different seat to see movies or filmstrips. Many students with albinism will have trouble reading from overhead projector screens because of glare. The student may copy the acetate sheets from the overhead projector as the teacher finishes using them. Or the student may ask to copy another student's notes. Or another student may take notes on carbonized paper and give the original to the student with low vision.

Use of Large Type: Students with albinism may or may not need large print. The team should look at the distance and near vision acuities from the eye examination. The TVI should evaluate the student working with various print sizes. Each spring the TVI can gather all books the class will use during the following year and review the books with the student to ascertain which need to be enlarged. Contrast is probably more important than print size in making this determination. The student's judgment as to which books need to be enlarged is one important element in the decision process.

Standard classroom materials through the third grade use primary size type, which is often sufficiently large for those with low vision. By the third or fourth grade, it may be advisable to enlarge math books because some of the symbols are quite small (fractions etc). Students may be willing to use large print in school in the fourth through sixth grades. But by junior high they may not want to use large print in the classroom because of peer pressure. They may be willing, however, to use the large print books at home. It is important for the team to think about how they might be able to assist the student in becoming more self confident about the tools that are required in the classroom. It is unwise to make the assumption that the student will be uncomfortable about using large print or other aids in the classroom.

Books for keyboarding class usually need to be enlarged and placed on a slanted reading stand because it is not possible to get close to the print while using the keyboard. Likewise, music scores for band and orchestra instruments usually need to be enlarged due to the distance from which they must be viewed.

For those students who require large print books and materials, the TVI usually can obtain large-type versions of the usual classroom textbooks either from the American Printing House for the Blind in Louisville, Kentucky, or from other sources that reproduce texts in large type. In some cases, the local education agency must photo-enlarge the textbook themselves. Some students may prefer tape recordings rather than large type.

Teacher-prepared handouts may be photo-enlarged or read to the student. By photocopying, it is often possible to increase the contrast of low contrast text materials and drawings.

Standardized achievement tests may need to be photocopied and enlarged, since the test booklets usually are to be reused and cannot be marked in, and the print is small. The machine scored answer sheets which accompany the booklets are in very small print. The task of looking from test booklet to answer sheet and back, over and over, is extremely difficult for visually impaired children. When the test is enlarged, the student can mark answers directly on the booklet. Later, the answers can be transferred to a machine scored answer sheet. ACT's, PSAT's and SAT's are available in large print and on cassette from the publisher through the school. These tests can also be taken under untimed conditions if that is the accommodation that the team has determined appropriate for educational testing situations.

Nonoptical Aids to Vision: Computers can be of great use to students with low vision. Students with albinism should begin to learn keyboarding skills with typing readiness computer games as early as kindergarten; they should begin direct instruction in typing or keyboarding as early as the third grade.

Computers with large screen monitors and software for large character display may help older students with writing projects.

A CCTV (closed circuit television) may help a student to read charts, graphs, pictures and text. When using a CCTV, try reverse polarity (white letters on black background) to reduce glare.

Optical Aids: For reading, the student with albinism may use stand magnifiers, hand-held magnifiers, or special reading glasses, such as bifocals with a strong reading lens or telescopic reading lenses clipped over glasses.

Contact lenses may help for distance vision. Some students use monocular telescopes (small hand-held telescopes). Older students may use a bioptic lens systems, which can improve reading and distance vision. Bioptics incorporate telescopic lenses into standard glasses, to allow the student to use either the standard correctional lens or a telescope for intermediate or distant vision. Some bioptics may include two different telescopes- one for distance and one for reading- mounted in the same carrier lens. Newer designs of bioptics use small lenses that are lightweight and cosmetically more acceptable, but may be more difficult to use because of a smaller field of vision.

In consultation with the TVI, an optometrist or ophthalmologist experienced in low vision should prescribe specific aids for the individual student. In the doctor's office the student should have the opportunity to try all types of aids and to select the most comfortable type of aid for a given visual task.

After an aid is prescribed, the TVI works with the student while he or she learns to use it properly. Low vision clinics should allow students to use aids on a trial basis, and to return them if the aids are not helpful. To review the usefulness of an aid, the student should return to the doctor about one month after an aid has been prescribed.

Students often are reluctant to use optical aids because the aids draw attention. To be like everyone else, students may avoid using the aids and pretend to be able to see when they cannot. Younger students may hide, lose, or break their glasses. It is always appropriate and important to help the student appreciate the benefit of the aid and feel comfortable using it in a wide variety of situations.

Physical Education: The school staff needs to find ways to include students with low vision in all activities, and to avoid leaving them sitting on the sidelines. "Small ball" games (tennis, badminton, softball, baseball) are difficult for children with albinism, due to both the size and speed of the balls. Games involving large balls work much better (soccer, kickball, basketball). Yellow balls and colored bases make ball games more visible for the child with albinism. Swimming, aerobics, gymnastics, track, wrestling, horsemanship and skiing are sports which can be enjoyed with minimal adaptations required. It is important for the Physical Education program to keep in mind that fact that learning about the sports and activities (Playing field, rules, roles of team members etc.) is as important, and sometimes more important, than being able to play the game with increasing success.

Social Support: Teachers must avoid drawing attention to the student with albinism for reasons directly related to the Albinism. Students with oculocutaneous (skin as well as ocular) albinism stand out in any case, and may require emotional support for responding to their classmates. Parents can help their children deal with teasing or other forms of discrimination by encouraging them to share their experiences and their feelings about them. Conferences including the student, teacher, vision teacher, and parent may help to support a student.

Alternatives to Mainstreaming: Though most children with albinism will be able to succeed in a typical education setting, sometimes the least restrictive environment for a child may be a half-day resource room with other visually impaired students, or a special class, or even a special school for students with visually impairments. The child may require education outside the

mainstream classroom because the child's self-esteem is very low, or the child's skill levels are below those of his classmates, or the child emotionally cannot deal with the demands of the regular grade placement.

If such problems require removing a child from the classroom, it is important that the educational team develops a plan to address these problems, such as group or individual counseling as part of the school day, individually designed remedial instruction in weak areas, or individual instruction in study skills, organization skill and social skills. It is important to place a time limit on such placements, and review goals and progress closely.

In considering alternatives it is important to consider the fact that no single approach works for all students with albinism. However, with patience, understanding, and hard work, the team can find ways for a student to function successfully in the general education program.

For Parents: An IEP for Your Child

The Individual Education Plan or IEP is an important part of the education process for the visually impaired student. The IEP is the blueprint for the coming year. It describes adaptations and modifications required by students to compete with their sighted counterparts. For parents and students, the IEP meetings sometime prove intimidating and filled with uncertainty, but it helps for parents to understand the process and their rights.

Parents should prepare for the meeting by understanding its purpose, and their rights under the Individuals with Disabilities Education Act. In addition to the federal law, many states have enacted state regulations and the local school district may have its own guidelines.

Parents can obtain copies of these laws and regulations from the school system. The School is required by law to provide parents with a copy of their rights under IDEA in a format that is accessible to them.

Parents might consider bringing a list of questions to the meeting or sending them to the special education lead person ahead of the meeting. They should remember that they have at their disposal the collective knowledge of the professionals who will be at the meeting.

It is important to have the most current report from the child's ophthalmologist or optometrist. This report is helpful, along with input from the TVI, in determining the best modifications to the classroom environment and print materials.

Parents should keep a file with past IEP reports and most recent reports by the school psychologist, social worker, and other educational evaluations necessary for meeting federal, state and local IEP regulations. Parents may request these reports from the appropriate school personnel.

The IEP team should include the parent(s), at least one regular education teacher and one special education teacher of the child, a local education agency representative who can authorize services, an individual who can interpret the instructional implications of evaluation results, and any others who have knowledge or special expertise regarding the child, including related services personnel as appropriate. It is important that both parents attend the IEP meeting-it does make a difference. The child may also attend the IEP meeting, and must attend if over the age of majority (18 in most states).

Parents should keep in mind that they know their child best. They should listen to their child, and advocate for him or her. It is helpful to take notes to remember the points that may need clarification, and ask questions after the report or at appropriate times throughout the meeting. Parents should not sign the IEP until they have read it and agree with its content. However, they may be asked to sign a form that states they were present at the meeting, though the IEP is not yet written. It is appropriate to sign that you attended the meeting.

Common ideas about low vision may not apply to the child in question. In particular, large print or pre-recorded text materials are not always appropriate, and should not be accepted as substitutes for poor visual aids. The IEP allows parents of children with visual disabilities to advocate a program tailored for their child.

Conclusion

A very basic human need is to be "seen" "by another person-to be known and to be accepted. This is poignantly true for the person with albinism who may be immediately "noticed" by many, but truly "seen" by few. This explains why it feels like a hidden condition despite its obviousness.

Being involved with others is a way to decrease isolation and share in the combined knowledge and experience of the group. It is a way to gain confidence and strength in meeting the challenges of this condition.

Albinism, often unexpected in a family, can be a catalyst for acceptance, understanding, and love that encompasses all family members and each of their individual differences. It is a physical manifestation of uniqueness, with unique joys and hardships. Through the efforts of dedicated families and individuals, albinism is becoming known and understood.

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