

Our Eyes Are Different

An Overview of Albinism and the Eye

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Claude Monet painted captivating works of art and is considered the founder of the French Impressionist movement. Franklin Delano Roosevelt raised our nation's spirits during times of war and economic hardship over four consecutive presidential terms. Harriet Tubman's courage led hundreds of African Americans from slavery to freedom. Now you might ask, "What in the world does this have to do with albinism?" The answer is they all created, led and emancipated with a visual impairment.

You are probably aware that albinism almost always has an impact on the eyes leading to subnormal vision.

However, with advancements in corrective and rehabilitative technology and guidance of an eye care specialist, the effects on daily activities do not have to be debilitating. My goal is to bring some *insight* (pun intended) into the relationship between albinism and the eyes and visual system. As this is my debut article, I will begin with an overview of how albinism affects the eyes both structurally and functionally. In upcoming columns, I will focus on each of the ocular effects in more detail and address ways to utilize decreased vision to its maximum potential.

In order to understand the influence of albinism on the eyes and vision, it is important to understand some basic anatomy of the eye and its circuitry. Think of the eyes and visual system as a camera. Like a camera, the visual system transforms visible light into recognizable images. Eyes contain lenses to focus the entering light similar to a camera's optical system. Chemical reactions take place in the retina (the back inner layer of the

eye) as chemicals aid in the development of film. As light enters the eye, it first hits the cornea, or the front lens of our hypothetical camera, which begins to initially focus the light. Then the light continues through the pupil (the black hole in the center of the iris, or colored part of the eye). The iris controls the amount of light entering the pupil as would an aperture in a camera. Finally, the light is focused by the second inner lens (the intraocular lens) before landing on the retina.

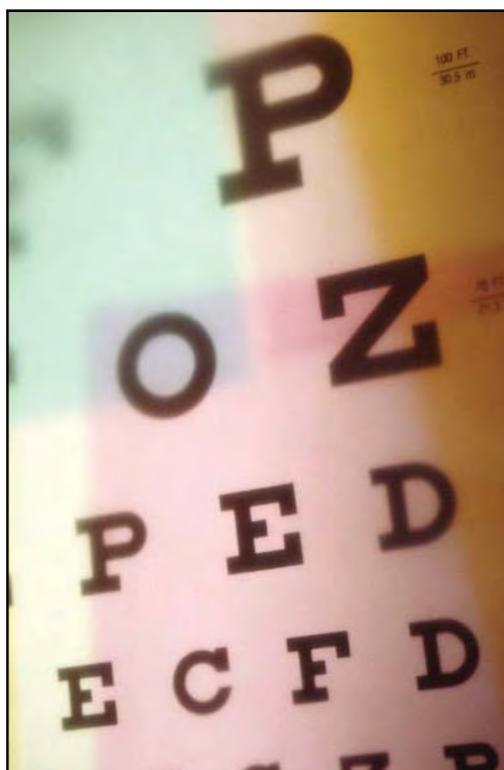
When the light comes into focus directly on the retina, no glasses or contact lenses will be needed to see well at distance. If the light comes into focus in front of the retina, the eye is considered nearsighted and would require a minus lens prescription to diverge the light and push the light back onto the retina. If the light comes into focus somewhere behind the retina, the eye is considered farsighted and a plus lens prescription is required to converge the light so the image lands on the retina.

Astigmatism is another type of refractive error in which the optical system fails to land the light directly on the retina. The center portion of the retina contains a pigmented area called the macula with a small depression in its center called the fovea. These structures are responsible for allowing us to see color and fine detail. The retina begins to process the light and triggers impulses that are sent through the optic nerve, hit the optic chiasm where some of the fibers cross, then are sent to the brain. Finally, the brain deciphers the code and allows us to perceive our surrounding world.



Albinism is classified into two broad categories: ocular albinism and oculocutaneous albinism. Ocular albinism affects the eyes with minimal to no skin involvement, while oculocutaneous albinism affects the eyes, hair and skin. People with albinism produce a low amount of a chemical called melanin resulting in decreased pigmentation of the skin, hair and eyes. Melanin helps protect the eyes from harmful ultraviolet (UV) damage and naturally decreases in the eye with age, which is why older patients have a predilection for macular degeneration and cataracts. Eyes that are lighter in color are also at a higher risk, as there is less protective melanin. For this reason, those with albinism need to be especially cautious of the sun's UV rays and wear sunglasses with UV protection whenever outside.

Decreased melanin production results in poor development of the retinal pigment epithelium (the back colored layer of the retina), choroid, fovea and iris. In the book, *The Da Vinci Code* by Dan Brown, there is a description of the character, Silus (the evil-“albino”-Catholic-monk-assassin) that states, “his irises were pink with dark red pupils.” On top of Brown's character portrayal being the typical offensive stereotype and completely unoriginal (an evil person with albinism has previously played the antagonist in *End of Days*, *The Matrix Reloaded*, *Cold Mountain* and dozens of others films), it is also inaccurate. The iris is never red or pink, but simply lacks pigment. The decreased coloring from lack of melanin allows the retina to shine through the iris so you are actually seeing the reddish-pink coloring of the layers behind the retina and retinal blood vessels.



Ocular symptoms of albinism include decreased vision, double vision, light sensitivity and difficulty with depth perception. Underdevelopment of the retinal pigment epithelium and fovea results in decreased vision. The poor development of the fovea may also result in a quick involuntary eye movement known as nystagmus, which may also further impair vision if it is severe. Double vision is due to a condition known as strabismus, or an eye turn. Strabismus, if left untreated, can lead to amblyopia (better known as a lazy eye), as the brain will choose to ignore the turned eye and rely on the vision from the aligned eye. In those with albinism, more nerve fibers cross at the optic chiasm than those without albinism resulting in decreased depth perception.

Treatment for myopia (nearsightedness,) hyperopia (farsightedness) and astigmatism include single vision spectacles and bifocal or multifocal glasses or contact lenses. If glasses or contacts do not alleviate visual symptoms, telescopes, magnifiers and other low vision aids are beneficial. Low vision devices should be prescribed by a low vision specialist following a thorough

evaluation during which the patient's daily activities and visual goals are relayed and the physician prescribes devices to help meet the patient's visual requirements. Treatment for strabismus includes spectacles made with prism, which aids in aligning the eyes, and surgery of the ocular muscles. Nystagmus may also be reduced, or dampened, with the aid of prismatic spectacles or a surgical procedure. Since many cases of albinism are diagnosed by optometrists or ophthalmologists during

routine eye examinations, early eye exams are critical in order to check for ocular signs of albinism, provide treatment for the visual conditions and secure the best visual outcome for the child. In fact, the American Optometric Association recommends a first eye exam between the ages of 6 to 12 months.

When diagnosed with albinism, there are many challenges, one of the most paramount being vision. Although the vision challenges may seem discouraging, there are a number of options available to make the previously impossible task possible.

Most argue Monet's best works were created late in his career. It is believed this praised abstractionism was a true depiction of how he began to view the world as his vision worsened. If Monet had quit painting, then his "Water Lillies" collection would simply



not exist. If Roosevelt had let his vision impairment squelch his ambition, the United States may have fallen during World War II and there would not be Social Security. If Tubman had been defeated by her decreased vision, many African Americans would never have experienced freedom. Today, with the assistance of new technology and your eye care professional, many of the visual effects of albinism can be overcome, perhaps with practice, or maybe in the blink of an eye.

If you have any questions, I would be happy to address them in upcoming articles. Please mail them to:

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