



# Fact Sheet

## Stargardt's Disease

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*Information retrieved from the Lighthouse International at [www.lighthouse.org/medical/eye-disorders/stargardts-disease/](http://www.lighthouse.org/medical/eye-disorders/stargardts-disease/) and from an article by Drs. Richard L. Windsor and Laura K. Windsor retrieved at [www.visionww.org/drswindsor-stargardt.htm](http://www.visionww.org/drswindsor-stargardt.htm).*

**What Is Stargardt's Disease?** Stargardt's disease is the most common form of inherited juvenile macular degeneration. It is characterized by a reduction of central vision with a preservation of peripheral (side) vision. Dr. Stargardt first described this condition in 1909. It is a common forms of a juvenile macular degeneration. Stargardt's disease may occur in one of every 20,000 children over the age of 6 and is usually diagnosed before the age of 20. Boys and girls are equally affected by this condition. Over 25,000 Americans have Stargardt's disease

**Clinical Description:** Stargardt's disease, also known as fundus flavimaculatus, is usually diagnosed in individuals under the age of 20 when decreased central vision is first noticed. On examination, the retina of an affected individual shows a macular lesion surrounded by yellow-white flecks, or spots, with irregular shapes. The retina consists of layers of light-sensing cells that line the inner back wall of the eye and are important in normal vision. The macula is found in the center of the retina and is responsible for the fine, detailed central vision used in reading and color vision.

The progression of visual loss is variable. One study of 95 individuals with Stargardt's disease showed that once a visual acuity of 20/40 was reached, there was often rapid progression of additional visual loss until acuity was reduced to 20/200 (legal blindness). By age 50, approximately 50 percent of all those studied had visual acuities of 20/200 or worse. Eventually, almost all individuals with Stargardt's disease are expected to have visual acuities in the range of 20/200 to 20/400. The reduced visual acuity due to Stargardt's disease cannot be corrected with prescription eyeglasses or contact lenses. In late stages of the disease, there may also be noticeable impairment of color vision.

**What Causes Stargardt's Disease:** In 1997, Foundation Fighting Blindness researchers isolated the gene for Stargardt's disease. The ABCR gene produces a protein involved in energy transport to and from photoreceptor cells in the retina. Mutations in the ABCR gene, which cause Stargardt's disease, produce a dysfunctional protein that cannot perform its transport function. As a result, photoreceptor cells degenerate and vision loss occurs.

**What are Symptoms of Stargardt's Disease?:**

- Bilateral, decreased central vision in childhood or young adulthood;
- Visual acuity may start at the 20/40 level and later decline to 20/200 or slightly worse. Stargardt's disease may first be detected by a mild loss of visual acuity (sharpness of vision). In the early stages, the person's vision may be near normal. Visual acuity measurements may also vary due to the effects of light exposure and one should not be alarmed if your visual acuity varies on each test. The majority of Stargardt's patients have visual acuities from 20/100 to 20/400.
- Central scotoma (blind spot in central vision). Stargardt's disease creates central blindspots that increase in size as the disease progresses. Individuals often learn to turn their eyes in a specific direction to work to see around the blindspots. This is called eccentric viewing.
- Blind Spots: As Stargardt's disease progresses, individuals may experience small areas of vision loss or blind spots. As images fall upon the damaged areas of the retina, objects may disappear and reappear causing a come and go effect.
- Abnormal color vision: The condition eventually damages the macular area where cone or color vision cells are. Color vision declines as the disease progresses, but individuals with Stargardt's disease usually maintain a significant amount of color vision
- Photophobia -- an abnormal visual intolerance of light
- Night blindness and Poor Light/Dark Adaptation: Children with this condition may report difficulty adapting to the dark after sunlight exposure. Light striking our retina causes chemical reactions to occur in the rods and the cones. When exposed to bright sunlight, the retina may become bleached by the light and the sharpness of vision may decrease and blind spots may become denser. These are temporary conditions, but can be prevented or lessened by use of sun filters and hats.
- Reduced Depth Perception: Depth perception requires good vision in both eyes. Anything that decreases vision in one or both eyes will affect depth perception.

**Diagnosis:** At the beginning stage of the disease, the retina may appear normal upon routine examination. As the disease progresses, clinical signs of the condition can be viewed with ophthalmoscopy. Your ophthalmologist or retinal specialist may perform additional common diagnostic procedures -- visual acuity tests, color vision testing, the Amsler grid test or an automated macular field test to assess vision. Other tests may also be ordered, such as an electrodiagnostic test to examine macular function.

As with macular degeneration patients, fluorescein angiography may be necessary to document

the pattern of blood vessels and determine whether dye leaks from the vessels. A doctor can determine which areas of the macula are damaged and whether there are abnormal blood vessels to be treated.

Stargardt's disease, an early-onset form of macular degeneration, is an inherited disease. The condition is programmed into one's cells at conception. It is not caused by injury, infection or exposure to a toxic agent.

**Educational Recommendations:** The Foundation Fighting Blindness strongly recommends that people with Stargardt's disease wear brimmed hats or visors and sunglasses when outdoors. Low vision devices and orientation mobility services are recommended. A certified teacher of students with visual impairments will be able to provide more details on an individual learner's educational needs.

### **Resources on Stargardt's Disease**

- Fact Sheet - <http://www.visionaustralia.org.au/info.aspx?page=598>
- Foundation Fighting Blindness Webpage specific to Stargardt's Disease :  
[http://www.blindness.org/index.php?option=com\\_content&view=article&id=53&Itemid=74](http://www.blindness.org/index.php?option=com_content&view=article&id=53&Itemid=74)
- FamilyConnect Stargardt's Disease Forum:  
[http://www.familyconnect.org/message\\_board\\_subjects.asp?FolderID=68](http://www.familyconnect.org/message_board_subjects.asp?FolderID=68)

**For more information about the CO Services for Children and Youth with Combined Vision and Hearing Loss Project and the project's services specific to Colorado children who are deafblind, please contact:**

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*Fact Sheets from the Colorado Services to Children and Youth with Combined Vision and Hearing Loss Project are to be used by both families and professionals serving individuals with vision and hearing loss. The information applies to children, birth through 21 years of age. The purpose of the Fact Sheet is to give general information on a specific topic. More specific information for an individual student can be provided through personalized technical assistance available from the project. For more information call (303) 866-6681 or (303) 866-6605. Updated: 10/10*