

OPHTHALMOLOGY & OPTOMETRY

Stargardt Disease



Overview

Stargardt disease is the most common form of inherited juvenile macular degeneration. It is caused by a deterioration of the eye's macula. The condition typically develops during childhood or adolescence, but sometimes does not cause vision problems until later in life. The macula is part of the retina that is responsible for color perception and central vision, which is needed for detailed tasks such as reading, writing, driving and seeing other fine details clearly.

The rate at which vision is lost varies for each person. However, eventually most people with Stargardt disease have 20/200 to 20/400 vision, which cannot be corrected with prescription eye glasses, contact lenses or refractive surgery.

In most cases, Stargardt disease is inherited as an autosomal recessive disorder. People who have one gene for the disease paired with one normal gene are unaffected. These people are called carriers. When two carriers have children, each child has a 25 percent chance of inheriting two copies of the Stargardt gene (one from each parent). Children who inherit two copies of the Stargardt gene will have the disease.

Our approach to Stargardt disease

UCSF's ophthalmologists provide comprehensive evaluations and care for all types of eye conditions, from the most common to the rare and complex. They are experts in inherited eye diseases, such as Stargardt disease.

Although Stargardt disease currently has no treatments, researchers are working to develop them. Interested patients may be able to receive experimental treatments by joining a clinical trial. Lifestyle changes, such as avoiding excessive light and not taking supplements that contain vitamin A, can slow vision loss. Low-vision aids, mobility training and other supportive care can also help patients with performing everyday tasks.



Awards & Recognition



Among the top hospitals in the nation



Rated high-performing hospital for ophthalmology

Signs & symptoms

In the early stages of Stargardt disease, vision may be near normal, which can delay diagnosis. As the condition progresses, it causes loss of central vision, which is needed for detailed tasks such as reading, writing, driving and seeing other fine details clearly.

Other symptoms include:

- * Difficulty perceiving color
- * Blind spots in the field of vision
- * Difficulty adapting to dim or dark spaces such as a darkened room or outside at dusk, or after being exposed to bright lights

Diagnosis

In many cases, your ophthalmologist may be able to detect Stargardt disease by examining your retina and macula. These parts of the eye often develop yellowish flecks that are deposits of a fatty byproduct of normal cell activity, called lipofuscin, which accumulates abnormally in people with Stargardt disease.

A test called [fluorescein angiography](#) or optical coherence tomography (OCT) may also be recommended to obtain more detailed images of your retina. In addition, it may be necessary to measure how the retina responds to light with electrodiagnostic tests such as an [electroretinogram \(ERG\)](#) or multifocal ERG.

Treatments

At this time there is no treatment available for Stargardt disease. However, researchers are studying gene and drug therapies. One promising study involves delivering a healthy version of the gene that causes Stargardt disease, ABCA4, into retinal cells to restore production of the normal protein. Your ophthalmologist may discuss the option of testing your blood for mutations in this gene. Another study aims to replace the cells that live under the retina, the retinal pigment epithelial (RPE) cells, using stem cell therapy. Other approaches aim to develop drugs that can reduce the buildup of lipofuscin, which is responsible for vision loss in people with Stargardt disease.

Also, it is essential that people with Stargardt disease

Chen et al.



Visual aids and adaptations around the home and at work, and training from social services can help those with Stargardt disease. It may also be important that people with Stargardt disease visit a genetic counselor who can discuss disease inheritance and help with family planning, career choices and other issues related to living with the condition.

UCSF Health medical specialists have reviewed this information. It is for educational purposes only and is not intended to replace the advice of your doctor or other health care provider. We encourage you to discuss any questions or concerns you may have with your provider.

Where to get care (2)

Helpful resources

[California Department of Rehabilitation Services for the Blind and Visually Impaired](#)[eyeGENE](#)[Foundation Fighting Blindness](#)[The Carver Nonprofit Genetic Testing Laboratory](#)[World Access for the Blind](#)

Related conditions

[Leber congenital amaurosis \(LCA\)](#)[Retinitis pigmentosa](#)[X-Linked retinoschisis](#)

Support services

**Patient Resource****Spiritual Care Services**

Chaplains representing many faiths are available around the clock to provide support, comfort and counsel to patients, families and caregivers.

**Patient Resource****Case Management & Social Work**

Connect with a team that can help you find resources, solve problems and advocate for you during treatment at UCSF.

**Class****Mindfulness-Based Stress Reduction Class**

This eight-week class teaches mindfulness practices that can reduce stress and improve your overall health, such as meditation and body awareness.

**Patient Resource****Patient Relations**

We welcome feedback about your experience at UCSF Health. Find out how to contact us with comments, questions or concerns.

