What is Stargardt disease?

Stargardt disease is an eye disease that causes vision loss in children and young adults. It is an inherited disease, meaning it is passed on to children from their parents.

Stargardt disease is a form of macular degeneration, and is often called juvenile macular degeneration. Macular degeneration is when part of the retina that gives you central vision, called the macula, breaks down. In people with macula, called photoreceptors, die off. Central, or detailed, vision becomes blurry or has dark areas. It may also be difficult to see colors well.

What are Stargardt disease symptoms?

Stargardt disease usually develops in children and teenagers. Someone may first notice a problem with their central vision. It can be blurry, distorted or have dark areas. Side (peripheral) vision is usually not affected.

It may take longer than usual for vision to adjust when going between bright and dark areas.

For some people, Stargardt disease progresses slowly, then speeds up and levels off. At about 20/40 vision (meaning someone sees at 20 feet what a normal-seeing person sees at 40 feet), vision loss can speed up. Vision may rapidly get worse until it reaches about 20/200. After this point, vision generally stays about the same.

Eye Words to Know

Retina: Layer of nerve cells lining the back wall inside the eye. This layer senses light and sends signals to the brain so you can see.

Macula: Small but important area in the center of the retina in the eye, which is the region of keenest vision.

Macular Degeneration: A disease that causes the macula to break down, affecting central (detailed) vision.

Photoreceptors: Special cells in the retina that allow you to see light and color.
While central vision will be lost, many people with Stargardt disease may keep good side vision for the rest of their lives.

Who is at risk for Stargardt disease?
Usually Stargardt disease is passed down from parents. To have symptoms, you must inherit the gene (called the ABCA4 gene) from both parents. Someone who inherits the gene from only one parent will be a carrier for Stargardt disease, but will not have symptoms.

How is Stargardt disease diagnosed?
Your ophthalmologist will dilate (widen) your eye’s pupils to look at your retina. People with Stargardt disease have yellowish flecks called lipofuscin in and under the macula. Sometimes these flecks extend outward in a ring. Lipofuscin are fat deposits from normal cell activity.

A test called fluorescein angiography may be used. In this test, a dye is injected into your arm. The dye is photographed as it circulates through the retina’s blood vessels. In people with Stargardt disease, the photos show a dark area within tissue of the retina. This helps the ophthalmologist diagnose Stargardt disease.

How is Stargardt disease treated?
Unfortunately, there is no cure for Stargardt disease and no treatment to slow it down. Wearing sunglasses may help with the bright light sensitivity of Stargardt disease. Wearing sunglasses can also prevent further retina damage from the sun’s harmful ultraviolet (UV) rays.

People with Stargardt disease should not smoke cigarettes or be around cigarette smoke. Also, some studies suggest that taking a lot of vitamin A could make the disease worse. While the vitamin A in foods is fine, avoid taking large doses of it as a supplement.

There are genetic treatments already for similar diseases. Doctors are hopeful that Stargardt disease can be treated soon as well. There are helpful resources for people adjusting to vision loss. These can include special devices, useful tips for daily living and training to help you get around. Ask your ophthalmologist to help you find low vision resources.

Your ophthalmologist is committed to protecting your sight.
Stargardt Disease

Summary
Stargardt disease is an inherited eye disease. It is a form of macular degeneration that develops in children and teenagers. The disease causes blurry central vision, and makes colors appear less bright.

People who have Stargardt disease may find low vision resources helpful.

Get more information about Stargardt disease from EyeSmart-provided by the American Academy of Ophthalmology-at aao.org/stargardt-link.