



Understanding Stargardt Disease

It is estimated that Stargardt disease affects about 1 in 6,500 individuals.

1. Introduction

Stargardt disease is a rare genetic eye disorder that causes progressive vision loss. This disease occurs when fatty material builds up on the macula, the area responsible for sharp, central vision, and the area is eventually surrounded by a ring of white or yellow spots. While peripheral vision is not usually affected, people with Stargardt disease typically have problems with night vision that make it difficult to navigate in low light, and some experience impaired color vision.

2. Causes

Stargardt disease is an inherited genetic condition. The body uses vitamin A to create cells in the retina, then the ABCA4 gene makes a protein to clean up the leftover fatty material. In people with this condition, the ABCA4 gene does not work, leaving the fatty material to build up in yellowing clumps on the macula. Over time, this material kills the light-sensitive cells to destroy a person's central vision.

3. Symptoms

The signs and symptoms of Stargardt disease typically begin in childhood and will continue to progress into adulthood. A slow loss of central vision is the most common symptom, but the speed of vision loss will vary from person to person. Other symptoms include:

- + Gray, black, or hazy spots in the center of your vision
- + Sensitivity to light
- + Trouble adjusting to lighting changes
- + Color blindness

4. Diagnosis

Patients can be checked for Stargardt disease as part of a dilated eye exam. Physicians will look for irregular yellowish flecks extending outward from the macula in a ring. The number, size, color and appearance will vary from person to person. Some other tests that can help diagnose this disease are:

- + Visual field testing
- + Electroretinography (ERG)
- + Optical coherence tomography (OCT)
- + Autofluorescence imaging
- + Genetic testing

Did you know?

Stargardt disease is the most common form of inherited macular degeneration, affecting about 300,000 people in the U.S.

For more information on Stargardt disease and supportive resources, please visit www.nei.nih.gov.

References:

<https://www.nei.nih.gov/learn-about-eye-health/eye-conditions-and-diseases/stargardt-disease>

<https://medlineplus.gov/genetics/condition/stargardt-macular-degeneration/#frequency>

<https://www.macularsociety.org/macular-disease/macular-conditions/stargardt-disease/>

<https://www.fightingblindness.org/diseases/stargardt-disease>

<https://www.aaopt.org/eye-health/diseases/what-is-stargardt-disease>