

Retinitis Pigmentosa

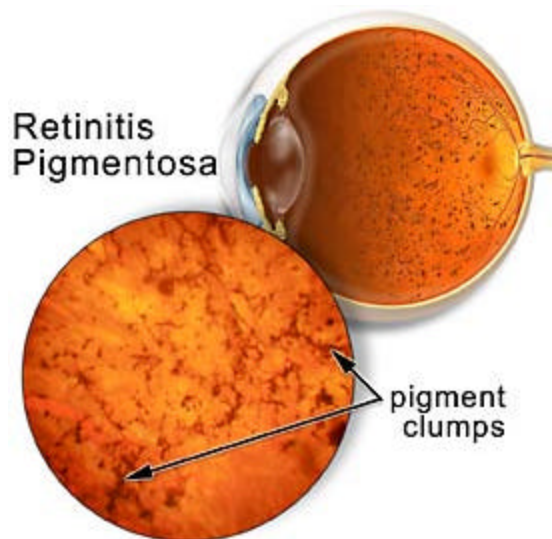
Overview

Retinitis pigmentosa (RP) is a rare, hereditary disease that causes the rod photoreceptors in the **retina** to gradually degenerate. The rods are located in the periphery of the retina and are responsible for peripheral and night vision. Cones, another type of photoreceptor, are densely concentrated in the **macula**. The cones are responsible for central visual acuity and color vision.

The disease may be X-linked (passed from a mother to her son), autosomal recessive (genes required from both parents) or autosomal dominant (gene required from one parent) trait. Since it is often a sex-linked disease, retinitis pigmentosa affects males more than females.

People with RP usually first notice difficulty seeing in dim lighting and gradually lose peripheral vision. The course of RP varies. For some, the affect on vision may be mild. Others experience a progression of the disease that leads to blindness.

In many cases, RP is diagnosed during childhood when the symptoms begin to become apparent. However, depending on the progression of the disease, it may not be detected until later in life.



Signs and Symptoms

- Difficulty seeing dim lighting
- Tendency to trip easily or bump into objects when in poor lighting
- Gradual loss of peripheral vision
- Glare
- Loss of contrast sensitivity
- Eye fatigue (from straining to see)

Detection and Diagnosis

Retinitis pigmentosa is usually diagnosed before adulthood. It is often discovered when the patient complains of difficulty with night vision. The doctor diagnoses RP by examining the retina with an **ophthalmoscope**. The classic sign of RP is clumps of pigment in the peripheral retina called "bone-spicules." A test called electroretinography (ERG) may also be ordered to study the eye's response to light stimuli. The test gives the doctor information about the function of the rods and cones in the retina.

Treatment

There is currently no standard treatment or therapy for retinitis pigmentosa; however, scientists have isolated several genes responsible for the disease. Once RP is discovered, patients and their families are encouraged to seek genetic counseling.

Current Research

Scientists at Johns Hopkins University are developing a micro-computer chip prosthesis called the Multiple-unit Artificial Retina Chipset (MARC). Once implanted in the retina, the chip transmits images to the brain that are captured from a small camera mounted on the patient's glasses. The chip is still in development and is not yet available for widespread use.

Doctors continue to search for treatments for RP but have yet to find a cure.