

Frequently Asked Questions: Retinitis Pigmentosa and Retinal Implants

What is retinitis pigmentosa (RP)?

Retinitis pigmentosa (RP) is a genetic condition that compromises vision and often leads to blindness. Signs and symptoms often first appear in childhood with severe vision problems typically developing in early adulthood.¹

What are the signs and symptoms of RP?

Initial signs and symptoms of RP include the degeneration of peripheral and night vision as well as a loss in color perception and central vision (sometimes referred to as tunnel vision). Most people with RP are legally blind by age 40, with a central visual field of less than 20 degrees in diameter.²

What causes RP?

Photoreceptor cells (rods and cones) in the retina are genetically programmed to produce the wrong amount of proteins which hinders the retina's ability to interpret images. Demise of the cells occurs gradually which is why vision deteriorates over time.³

Are there risk factors for RP?

Family history is the only risk factor as RP is an inherited condition.⁴

How prevalent is RP?

RP is one of the most common forms of genetic retinal disorders affecting approximately 200,000 people in the U.S. and Europe.⁵

What can people with RP see?

What, if anything, people with RP can see largely depends on the severity of their condition and how long they've experienced symptoms. Some people can see images but no light or color (night blindness); others can see a very small area of the visual field (tunnel vision); and then there are those who are completely blind.

¹ <http://www.nlm.nih.gov/medlineplus/ency/article/001029.htm>

² <http://www.nlm.nih.gov/medlineplus/ency/article/001029.htm>

³ http://www.blindness.org/index.php?option=com_content&view=article&id=50&Itemid=67

⁴ <http://www.nlm.nih.gov/medlineplus/ency/article/001029.htm>

⁵ Bloomberg News, 2009

(<http://www.bloomberg.com/apps/news?pid=20601124&sid=aYKW3Z05XoxE>)

How do people with RP function in their daily lives?

People with RP may use a cane and possibly a guide dog to help perform routine tasks like crossing the street and cooking dinner. While it is possible to pursue many of the same activities as a normal-sighted person, it typically takes people with RP longer to complete them, and they may require an adaptive device to do so.

Are there any available treatments for RP?

Physicians recommend sufferers wear sunglasses to protect the retina from ultraviolet light which may help preserve vision.⁶ However, there are currently no approved or commercially available treatment options that can restore vision, nor are there treatment options to retard the progression of the disease.

Are there treatments in development for RP?

Controversial studies have suggested that treatment with antioxidants (such as vitamin A palmitate) may slow the disease from getting worse, and the pharmaceutical industry is working to develop drugs for this purpose.⁷ Treatment with vitamin A-based drugs however, can not restore previously lost vision so it is only a potential option for patients in the early stage of the disease.

One of the most exciting and promising advances in the field is the development of microchip, retinal implants that can restore vision to a functional level for blind patients.

What is a retinal implant?

Retinal implants are electronic microchips that are inserted in the eye of a blind person to generate artificial vision; this is a similar idea to the use of cochlear implants returning the gift of hearing to those impaired.

What is the goal of retinal implants?

The goal is to restore vision to a point in which a person can perform normal, daily activities without other assistance.

Are there different kinds of retinal implants?

Yes, there are two main approaches—subretinal and epiretinal—scientists have taken to develop retinal implants. They differ in several ways including placement of the chip, number of electrodes produced by the chip and the mechanics involved in operating the chip.

Is there consensus among scientists on the most promising approach to retinal implants?

There is emerging consensus among researchers that the subretinal approach is better positioned to produce positive results for patients because of several factors including:

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- **Location:** subretinal implants are placed below the retina in the macular region where light-sensitive photoreceptor cells are located. It is these cells that are responsible for producing clear images in normal-sighted people.
- **Stability of implant:** scientists believe subretinal implants are unlikely to loosen over time.
- **Large quantity of electrodes:** subretinal implants omit substantially more electrodes (1,500 vs. 60) than epiretinal implants. The increased number of electrodes allows for the light and dark images to appear more vibrant, making them easier to see.
- **More direct vision:** unlike epiretinal implants, subretinal implants do not require a camera to capture and interpret images.

Have subretinal implants been tested in a human clinical trial?

Yes, the first human clinical trial of subretinal implants was conducted in 2005 by German-based Retina Implant AG; eleven people were implanted. The results were unprecedented and are expected to be published in a peer-reviewed journal in early 2010. Unlike patients who have received epiretinal implants, Retina Implant AG patients were able to see objects and shapes so clearly they could combine letters to form words, or essentially, read at a basic level as well as recognize foreign objects.

Of interest, scientists at MIT are also conducting clinical trials with their own version of a subretinal implant; however, they do not anticipate implanting in humans until 2013.

Are there plans for a second clinical trial?

Yes, the next human clinical trial will begin in 2010. Several sites across Europe will participate in the implantation of 60 patients including the U.K. In this upcoming trial, patients will have the option to keep the implant permanently, whereas in the first trial, the implant had to be removed after four months. One patient had such positive outcomes however, that he lobbied the government to allow him to keep the implant. Permission was granted, which has helped Retina Implant gather qualitative data regarding the implant's long-term effects. To date, no complications have been reported.

Are there plans for a clinical trial to take place in the U.S.?

No, at this time there are no plans for Retina Implant AG to conduct a clinical trial in the U.S.

Is there potential for retinal implants to treat other conditions that cause blindness?

Retinal implants hold much promise for the blind community. In the future, patients who suffer from blindness due to other conditions like age-related macular degeneration (AMD) may be candidates for retinal implants.