## **Retinitis Pigmentosa**

Retinitis pigmentosa (RP) is a common form of retinal degeneration which affects over four million people world wide. It is the name given to a group of inherited eye diseases that affect the retina.

In most cases the early symptoms of RP develop between the ages of 10 and 30.

### What is the normal structure of the eye?

The eye is made of three parts.

- A light focussing bit at the front (cornea and lens).
- A light sensitive film at the back of the eye (retina).
- A large collection of communication wires to the brain (optic nerve).

In the retina there is a covering of rod and cone photoreceptor cells. The rods and cones convert light into electrical impulses that transfer visual information to the brain, where 'seeing' actually takes place.

## Symptoms

R.P. causes a gradual breakdown and degeneration of the rods and cones within the retina, resulting in a loss of a person's ability to see in certain conditions. As the disease progresses, a characteristic pigment can appear in the retina – hence the term Retinitis Pigmentosa.

# Rod and cone photoreceptors are good at seeing different things

Cones are concentrated in the central retina, known as the macular and are good at 'seeing' things that are still, in daylight, in colour and in fine detail.

It is the macular that is used for reading, writing, and detailed close work and recognising faces – in other words, our central vision.

Rods are found in the peripheral retina and are good at 'seeing' things that move, are in the dark but they only see in black and white and also in less detail.

This means that peripheral vision is affected which causes difficulty with navigation and night vision and people may become a little more clumsy by bumping into things and knocking things over plus steps and kerbs become hazardous as they are difficult to see.

In most forms of Retinitis Pigmentosa it is the rod cells that are usually first affected. These forms of RP, sometimes called rod-cone dystrophy, usually begin with night blindness.

Night blindness can be described by likening it to the effect on a normally sighted person when they enter a dark building on a bright, sunny day – it takes a few moments for sight to adjust. However, people with Retinitis Pigmentosa cannot adjust well to dark and dimly lit environments and it is not uncommon for sight to be affected for up to 15-20 minutes.

Other forms of RP, sometimes called cone-rod dystrophy, affect central vision first. With the loss of cone cells also come disturbances in colour perception.

As the disease progresses and more rod cells degenerate, then peripheral vision is affected.

#### Can RP be treated?

Currently, no treatment is available to cure RP or arrest its progress. It is the result of incorrect instructions being passed to the body's chemistry by faulty inherited genes.

However, there is a lot of research being undertaken world wide and many of the genes responsible for the numerous types of RP have been located and their defects identified. Scientists are following many lines of research, including gene therapy and stem cell therapy and they are hopeful that treatments will become available within the next decade.

### **Charles Bonnet Syndrome**

Many people with RP also experience Charles Bonnet Syndrome or visual hallucinations.

For further information please contact:

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