Health

I'm 24 and going blind

Motorcycling was his passion in life. But a routine eye test showed that **David Bradford** would never be able to ride again – and worse was to come

Nothing in life prepares you for finding out that you are going blind. I had no idea there was any problem with my eyesight until I casually mentioned to my optician, at a routine sight test, that I struggle to see in the dark. She made a detailed examination of my eyes and said nothing, but I knew from the look on her face that she had seen something ominous. I was told to go back the following day for a visual fields test. This assesses peripheral vision, and involves staring at a central dot of light inside a darkened box and pressing a button each time you catch a glimpse of another dot of light, away from the centre point. I waited and waited, staring into blackness, detecting no flashes until the final stages of the test. My heart sank; I knew something was seriously wrong. The optician's verdict began with those fateful words that only ever preface truly awful news: "I'm sorry to have to tell you this, but..."

Sorry to tell me that my peripheral vision does not meet the standard required for driving. How did this happen? One minute, I'm happily reading the optician's test chart to confirm, as usual, that my mild short-sightedness hasn't got any worse, the next I'm losing my driving licence and my life is in disarray. It doesn't get a lot worse than this, I thought. I was wrong.

The optician refused to be drawn on why my peripheral vision was subnormal, but it took no more than a couple of minutes of Googling to discover the most likely diagnosis – retinitis pigmentosa (RP). I discovered, with increasing dread, that RP is the name given to a group of genetic conditions that affect the light-receptive cells in the retina at the back of the eyes. Self-diagnosis using the internet is a dangerous game that often leads to unfounded fear, but the descriptions of RP matched perfectly my own circumstances. I discovered that RP manifests itself first as night-blindness and causes the loss of peripheral sight, leading to tunnel vision. Then the hammer blow that left me sick with despair: RP is progressive and ultimately leads to blindness, and there is no treatment or cure.

My life was taking a turn over which neither I nor the medical profession had any control. This just wasn't in the script. How can I be going blind? Where were the warning signs? Sure, I struggle getting around at night, but that's been the case for as long as I can remember. I can recall night-fishing trips as a teenager when I'd stumble chaotically over bags and equipment. If by luck a fish was hooked, I'd rely on friends to help get it netted and guide me back to the tent. It was obvious that my pals could distinguish basic shapes and contrasts after dark, whereas all I could see was opaque blackness. In daylight, my vision was apparently as good as theirs.

Poor night vision had never held me back. My family lived deep in the Sussex countryside, miles from anywhere, so I was keen to get my own wheels as soon as possible. At 16, I bought a moped and, at 17, took my motorcycle test and got my first "proper bike". Motorcycling became an all-consuming passion. Despite riding almost every day, in darkness as well as daylight, my vision was caused a mishap on the road. Just as I'd adapted to poor night vision in other aspects of life, I rode accordingly after dark.

'On night-fishing trips I'd stumble over equipment. My pals could see shapes after dark, I could only see blackness'

After university, I pursued my love for bikes by taking a job on a motorcycle trade magazine. Writing about my hobby seemed a great way to earn a living. Two years later, I landed my dream job and became a staff writer at *SuperBike Magazine*, Britain's top-selling sports-bike magazine. I am 24 now and still writing for *SuperBike*, but I will never again ride a motorcycle on the public highway. The DVLA revoked my driving licence shortly before Christmas because my peripheral vision falls short of their rigid requirements. The impact has been twofold. Firstly, there's the personal desolation at losing what is not only a treasured pastime but also a means to independence and freedom. Secondly, there's the professional diminution at having lost the ability to perform a key part of my job, which is also the part with all the perks and fun – road-testing the latest machines.

In RP sufferers, peripheral vision is lost gradually, usually ebbing away from the edges and spreading inward toward the centre, causing so-called tunnel vision. My case is atypical in that my mid-peripheral vision has been affected first. I still have a wide visual field but there are missing spots of vision between the centre and the periphery. In practice, this means I can see whatever I'm directly looking at and whatever is directly to the sides, above or below, but the immediate foreground is disrupted. I'm told these blind spots will get larger and spread outward, leaving just a central area of useful sight.

Approximately one in 3,500 people in the UK are affected by RP and it is the most common cause of blindness among young people. Even so, I had never heard of it, nor had any of my colleagues, friends or family. Most cases of RP are diagnosed between the ages of 10 and 30, and many sufferers are legally blind by the age of 40, but there are many different forms of the disorder. In fact, the singular name retinitis pigmentosa, coined by Dr Franciscus Donders in 1857, is considered inappropriate by modern ophthalmology, which prefers the collective label "retinal dystrophies".

Naturally, my first question to the consultant was, how quickly will my sight deteriorate? The answer was maddeningly indefinite; the symptoms of RP generally worsen slowly, and many sufferers retain some useful vision well into old age. But the rate of loss is not always constant and periods of rapid loss may occur. It is impossible to give an accurate prognosis in cases such as mine, in which there is no family history. There is no family history because I have, in effect, inherited the condition in two halves from my parents – one faulty gene from each.

There is currently no treatment or cure for any type of RP, but exciting progress is being made in several key areas of investigation. Gene therapy and stem-cell research, in particular, are showing great promise – and creating big news splashes. The three-tiered mission for researchers is to discover which genes cause RP ("gene mapping"), identify the exact nature of these genes ("gene cloning"), and find out exactly how and why they damage the retina. This is a long, painstaking process and so far only a small proportion of the genes and genetic mutations responsible for RP have been mapped and cloned.

Nonetheless, the eye is particularly suited to gene therapy, and trials have already achieved encouraging results. In one trial, corrective genes inserted into the eyes of mice were shown to halt retinal degeneration, while another trial using dogs actually restored sight in seven of the eight animals treated. A bold new trial, focusing on the same gene but involving human patients, will begin at Moorfields Eye Hospital, in London, later this year.

"The RPE65 [name of the gene] trial is the first of its kind and it's great that Britain is leading the way. Its effects should be seen quite quickly, within some months, and, if successful, will represent a crucial step forward," says Professor John Marshall, Frost Professor of Ophthalmology at St Thomas' Hospital, London.

Despite the potential of this trial, each type of RP is different and requires a different treatment; one success opens the door for another rather than provides a universal solution. When I ask Professor Marshall whether it is realistic to hope for a treatment or cure within my lifetime, he advises cautious optimism, "Although there is still a mountain to climb in terms of understanding RP, research is slowly but surely filling in the gaps and providing the full picture that will be needed in order to defeat all retinal dystrophies."

Apart from being prohibited from driving, my life is not yet affected, and I'm doing my best to stay positive. My predicament is difficult for other people to comprehend; I have been stripped of my licence and told I'm losing my sight, yet, to all intents and purposes, I can see perfectly well. I am aware that this will become a greater problem as my sight deteriorates, and testing challenges lie ahead. My priority, however, is to make the most of my sight while it is still serviceable, without becoming angry about the fate I've been dealt. Incredibly, I feel stronger and more resolute since being diagnosed, which is a reaction I would never have anticipated. Whether I'm trying to run faster, write better, or just win an argument, I feel propelled forward by a new impetus. My motorcycling days may be over but I'm determined to invest that passion elsewhere.

A common condition with no cure

RP is the most common cause of blindness among young people in the developed world, affecting one in 3,500 people.

It is the name given to a group of inherited disorders that harm the function of the retina, the portion of the eye responsible for transmitting light messages to the brain.

The retina has two main layers, the pigment epithelium, which provides the retina's nourishment, and a thicker layer, the neural retina, which is made up of layers of photoreceptor (light-sensitive) cells. RP affects both layers.

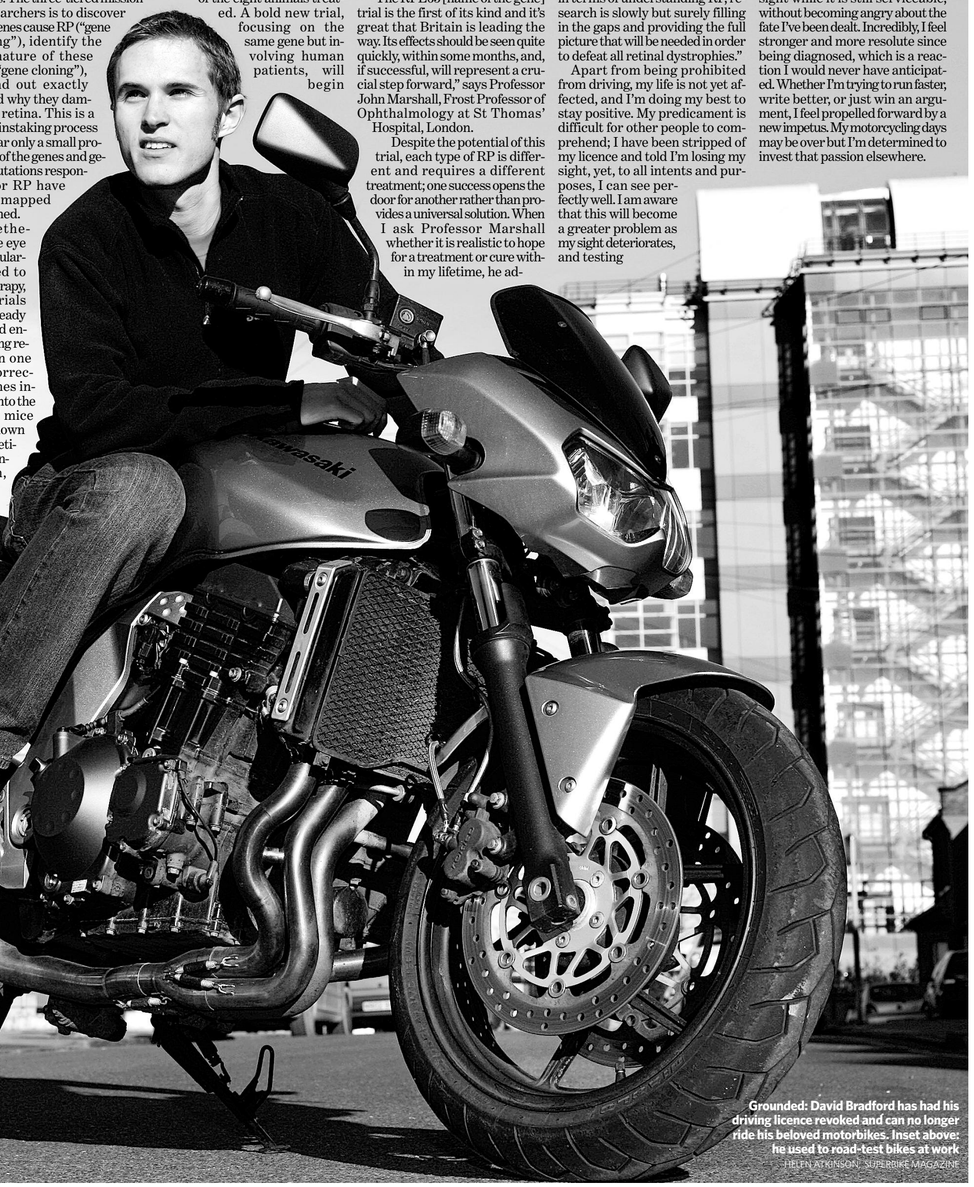
There are two types of photoreceptor cells – rods, used for peripheral vision and in poor light, and cones, responsible for seeing colour and detail. RP affects both cells.

RP is usually diagnosed between the ages of 10 and 30. The first symptom is night-blindness but it leads to the loss of peripheral sight (tunnel vision) and ultimately blindness.

There is currently no treatment or cure for RP.

There are five key methods of potential treatment under investigation: transplanting healthy cells into the retina; implanting microchips that mimic the action of photoreceptor cells; inserting healthy DNA using gene therapy; generating new retinal cells from stem cells; and protecting retinal cells using neuro-protection agents.

For more information, visit www.brps.org.uk



Grounded: David Bradford has had his driving licence revoked and can no longer ride his beloved motorbikes. Inset above: he used to road-test bikes at work

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