

# Walking hats

and other anecdotes related to retinitis



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## Introduction

Persons with Retinal Dystrophies often find themselves in unexpected and awkward situations due to their condition. Some of them are embarrassing, but most are hilarious, depending on how the patient and the people around him or her understand the reasons behind a certain unusual behaviour.

For instance, a person with tunnel vision might see only hats moving around in a crowded place. The name “Walking Hats” was therefore a suitable, yet practical name for this publication, which has already been published in Finnish and Swedish.

The majority of “Walking Hats” consists of clearly written medical explanations about different types of retinal dystrophies and how they affect the patient’s ability to see and function in daily life. In addition, all sections are accompanied by short, real life stories written by the patients themselves.

This publication aims at helping patients, be it during an early or latter stage of their eye condition, to realise why they see and act in a different way to other people. It is relieving for the patients to know that someone else has exactly the same problems as they do. Thanks to this practical approach, the information is invaluable for ophthalmologists, rehabilitation professionals, as well as social workers, with whom the patients need to communicate at some point of their lives. Nevertheless, it is much easier to explain your situation by giving this publication to your relative, friend or even a work colleague.

Variations of Retinal dystrophies, as well as their symptoms, are similar in different parts of the world. On the other hand, cultural values, communicational behaviour and expectations may differ widely from country to country as the individual tries to find an equal place in society. We hope that despite the slightly Scandinavian point of view, the publication gives a more personal perspective to anyone who reads it. As the English version of “Walking Hats” is given out in electronic format at Retina Finland’s homepage, we invite you to write your own short story to us, in order to build up “Walking Hats” to become even more globally oriented.

Helsinki, July 22<sup>nd</sup>, 2005

Pekka Rantanen  
Retina Finland

# Some Basic Facts on the Visual System

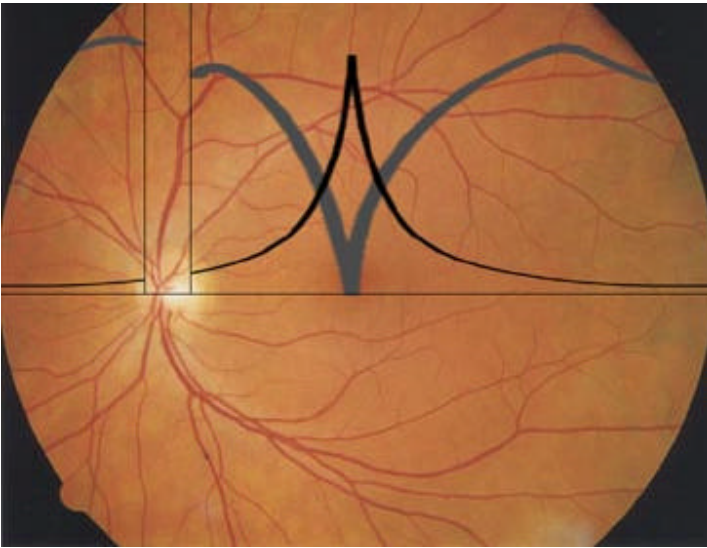
## Retina; cone and rod cells, distribution and function

Cone cells function in daylight; information from the three different types of cone cells conveys the colourful image that we see during the day. In twilight, cone cells' functioning decreases at the same time as the rod cell system increases its function. Colours first fade and then disappear when illumination decreases. As long as we see colours at all, there is some cone function present. The image of the rod cell system is in different shades of grey. Rod cells are most sensitive to the blue-green part of the spectrum, and thus blue shades are lighter than red shades in dim light.

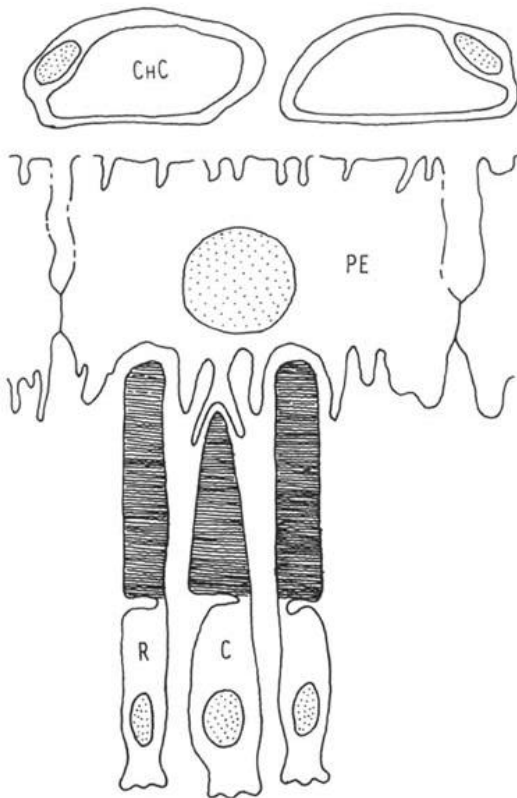
The distribution of the cone and rod cells in the retina varies, as shown in Figure 1. Cone cells, 5-6 millions in all, are numerous in the centre of the retina, called fovea, and sparse in the peripheral retina. There are some 160 million rod cells in the retina, none in the fovea and most densely packed in an area of retina that corresponds to a visual field between 20 and 30 degrees from the middle. This type of rod cell distribution causes a scotoma (= area of no-vision in the visual field) in dim light of one degree in diameter in the middle of the visual field. It influences visual functioning only when we want to see a dim star. If we know the exact location in relation to other stars, we see it best by looking just past its position.

Changes in the functions of the sensory cells, the cones and the rods, are not very well known because we can rarely take small pieces of the human retina for laboratory examinations. The present knowledge is based on investigations using animals that have progressive retinal changes similar to human retinal degenerations.

The rod cells grow in their outer part, called the outer segment, by adding new lamellae in the proximal end. From the tip of each outer segment a small part is shed every day. It is normally eaten up by the surrounding pigment epithelial cell. This usually, very regular function is disturbed in *retinitis pigmentosa*; the outer segments become longer than normal and parts of the outer segment remain undigested in the pigment epithelial cells.



**Figure 1.** Distribution of the sensory cells in the retina. In the very middle of the fovea there are some 200,000 cone cells per square millimetre. The number of cones per mm<sup>2</sup> decreases rapidly outside the fovea (black line). There are no rod cells in the fovea. Their number per mm<sup>2</sup> increases toward the periphery of the retina, is highest in the “midperiphery” and then decreases toward the far periphery (grey line).



**Figure 2.** The outermost layers of the retina are the layer of pigment epithelial cells (PE) and sensory cells, cone (C) and rod (R) cells. Pigment epithelial cells surround the outer segments of the sensory cells. In the outer segments of the sensory cells are the special pigments that absorb light energy, rhodopsin in the rod cells and three different pigments in the cone cells. The outer layers of the retina receive their nutrition from the blood flow in the inner layer of the choroid, called choriocapillaris (CHC) from where the nutrients are transported through the pigment epithelial cells.

## **Inherited disorders of the retina**

Retinitis pigmentosa, Usher Syndrome, Retinoschisis, Choroideremia, gyrate atrophy and many different macular degenerations are inherited disorders of the retina. They are usually described as retinal diseases and each of them has a specific number in the international classification of diseases. However, they do not have features that are typical of diseases: there is no pain, nor inflammation. It is important to give this information to children and young adults, healthy, active individuals, who should study and choose a suitable occupation for their usually long life.

In the Finnish edition of this publication we used the term "*retiniitikko*" for all people who have inherited retinal disorders. A similar general term does not exist in English, so we will refer, for example to a "person with RP" or an "individual with Usher Syndrome".

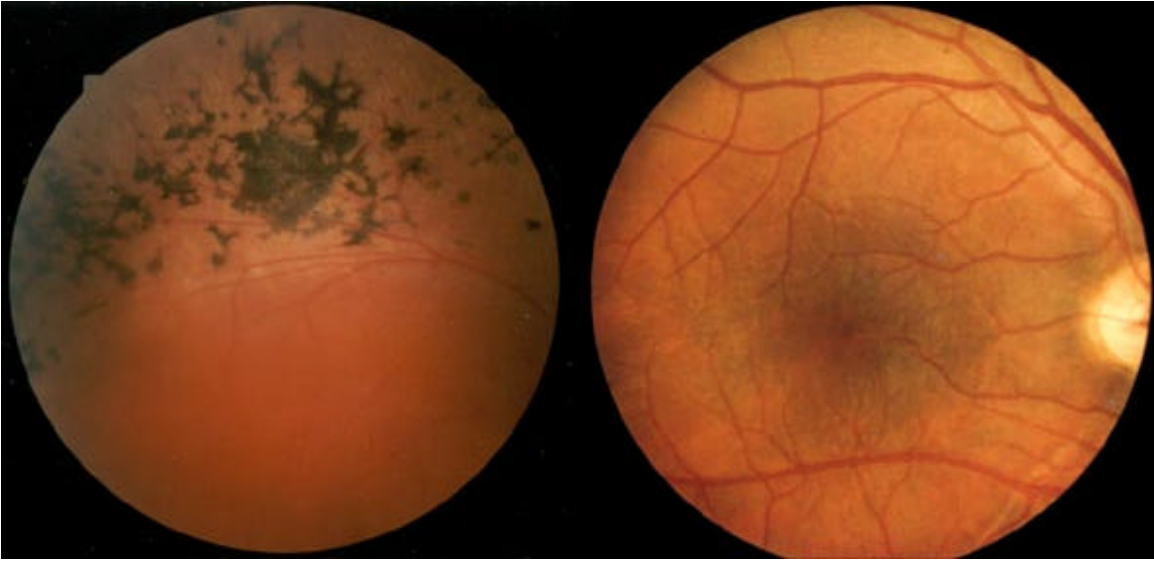
Slowly progressive retinal disorders cause changes in retinal functions. Changes begin early, at preschool age, but they usually do not start to disturb daily functions until school age. Different losses of retinal function appear slowly, one after another, and thus a young person with a retinal disorder must start to cope with a new loss just when (s)he has learned to live with the previous loss. In macular degenerations, vision may sometimes decrease very rapidly if the thin walls of new-formed vessels bleed in the middle of the retina.

Changes in the retina do not show on the eye. Therefore, many individuals with retinal disorder try to hide their condition and pretend to be normal-sighted, although their vision may be seriously damaged. These has sometimes caused dangerous accidents and strange misunderstandings. They have also happened in families and at work places where other members of the group have been aware of visual impairment, but have not reacted fast enough when abnormal visual input caused a strange response in the person with a retinal disorder.

Since changes in the retinal disorders have large individual variations, there are also large variations in symptoms, problems and misunderstandings.

### **Retinitis pigmentosa (RP)**

Retinitis pigmentosa is an umbrella name for a group of retinal disorders with a slow loss of function of sensory cells and typical dark pigment "spiculae" in the midperiphery of the retina (Figure 3).



**Figure 3.** *Left: On the retina of an older person with RP can be seen an uneven distribution of pigment and tapering of the veins. Right: The retina (both pigment epithelium and blood vessels) of his grandchild appears normal even if the typical functional changes are present. The diagnosis of children and young people is often delayed because the outward appearance of the retina remains unchanged for years.*

When Professor Donders described retinitis pigmentosa in 1857, he thought that the changes were caused by an infection. “Retinitis” means retinal infection or inflammation. The development of retinal changes was very actively investigated in the later part of the 20<sup>th</sup> century. The term “apoptosis”, meaning abnormal coding for cell death, is now used to depict the too early death of cells in typical parts of the retina. The changes start often as small changes in the rod cell functions and thus affect vision in dim light. Cone cell functions are affected later and lead to loss of function in the midperipheral parts of the visual field: first small scotomas that then become larger and slowly form the typical ring scotoma. A similar ring scotoma is present in rod cell vision long before cone cell changes, but visual fields are not measured at very low, scotopic, luminance levels and therefore our knowledge of the earliest changes is scanty. These early changes occur before school age when the measurement of visual fields is difficult.

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## **Patient's story**

I have tunnel vision, but I was not using my white cane that time because I was in a familiar area. Narrowing my eyes, I was trying to find my way around in the sunlight and felt myself bumping into the arm of another pedestrian. "I'm sorry," I said, "I didn't notice you!"

The man I had bumped into walked on, but turned around suddenly and remarked: "No need to apologize. You see, I have what's called tunnel vision!"

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## **Usher Syndrome**

Usher Syndrome is an inherited dual sensory impairment caused by changes in the inner ear and in the retina. Three functionally-different types of Usher Syndrome are known. In each type, retinitis pigmentosa is the retinal disorder. In Usher Syndrome type I, hearing impairment is profound, in type II it is moderate and in type III it is progressive at school age. Usher Syndrome type III is common in Finland. Not all people who have retinitis pigmentosa and a hearing impairment have Usher Syndrome. These two impairments are common and may occur independent from each other in a person who does not have the Usher gene.

When a hearing impaired/deaf child has retinitis pigmentosa, it is important that (s)he learns written language well and, if possible, also learns to speak. If sign language and other visual communication become impossible in adult life, reading Braille can be used as an effective way of communication. However, Braille requires knowledge of written language. There are several useful communication devices based on Braille, and thus a deaf-blind person may have sufficient communication with the surrounding society, whereas a deaf-blind person using only tactile sign language rarely has contact with enough many people who use tactile sign language. In Usher Syndrome type III, inner ear implants have restored hearing in many persons, even so well that the person can use mobile phones.

In early intervention and at school age it is very important that healthy, active children with Usher Syndrome are not stressed with the information of dual sensory loss, but are taught to live with their condition without fear. This approach is important in rehabilitation of all people with dual sensory

impairment, actually, important in all care and rehabilitation. Changes in vision are due to the retinal disorder and it is possible to learn compensatory techniques and strategies.

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### **Patient's story**

I was leaving the meeting-room of our society with our chairman. We were already at the front door when we heard rustling and rumbling sounds from the inside. Our chairman turned back to say goodbye and thanks one more time.

Since I knew we were the last persons to leave, I could not help but tell him that this time he had bid farewell to a dishwasher. For the rest of our journey, we were quite amused by the excessive friendliness of our chairman.

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### **Retinoschisis**

“Split retina”, the Finnish name for Retinoschisis, depicts the nature of a particular type of this condition: there is fluid between the retinal layers and there can be a hole in the inner layers of the retina. This separation of retinal layers from each other does not always occur; in a mild form of retinoschisis there are only thin radiating folds in the macula, the “wagon wheel” sign. This mild form is common in Finland. In it visual acuity is 0.2 - 0.6 (20/100 - 20/30) during school age and 0.2 in adult age. Contrast sensitivity is near normal and discrimination of movement normal. Investigations at the University of Helsinki, Department of Psychology, have shown that men with this disorder are good drivers. They have driven cars for 20-25 years, 700 000 to 1.5 million kilometres without traffic accidents and tickets. They also experienced themselves as normally-sighted. Men whose vision was worse had never tried to get a driver's licence.

Retinoschisis is inherited as an X-chromosomal trait, from a grandfather to his daughter's son. Female carriers have normal vision, and thus can pass the gene from mother to daughter for several generations without it affecting anybody in the family.

## **Choroideremia**

Choroideremia is a degeneration of the choroid that leads to the degeneration of the overlying retina. Since the outer layers of the retina are dependent on choroidal circulation, they cannot survive when the choroid degenerates. In the advanced cases there is only a small typical island of functioning retina in the centre of the macula. Functionally, choroideremia resembles retinitis pigmentosa causing night blindness and tunnel vision, but disturbing, illusory light phenomena are rare.

## **Gyrate atrophy**

Gyrate atrophy is a rare inherited retinal degeneration that is related to disturbance in the ornithine metabolism. This amino acid role in gyrate atrophy was first documented in Finland in the 1970s. Functionally, gyrate atrophy resembles retinitis pigmentosa.

## **Macula degenerations**

The group of inherited disorders in the centre of the retina includes conditions that cause loss of vision in children, in young adults or late in life. In children, vision loss may be the only impairment, as in Stargardt's degeneration or a part of a syndrome, as in Spielmeier-Sjögren (Spielmeier-Vogt, JNCL) syndrome, a progressive neurological disease.

In adult age, macula degeneration may be areolar degeneration, areolar meaning that there are well circumscribed round or oval areas where both the retina and the choroid have disappeared. If such degeneration occurs in the foveal area, loss of reading vision is severe. Sometimes, however, several areolar degenerations are located at the outer parts of the macular area without the person being aware of any loss of vision.

A great majority of persons with macula degeneration are elderly. This "senile" type of macula degeneration is either dry involution of the retina or "wet" degeneration with new-formed vessels growing from the choroid under and into the retina. The vessel walls are thin, so bleeding into the surrounding retinal tissue is common. Even a drop of blood causes a dark spot in the visual field. If bleeding is in the foveal area, loss of vision is nearly momentary. When blood is partially absorbed, it partly becomes scar tissue; a whitish scar is seen in the macula, known as Kuhnt-Junius degeneration. Bleeding may occur repeatedly over the years and lead to severe loss of vision.

The dry type of macula degeneration develops slowly and there is no treatment, although some dietary measures may delay the enlargement of the atrophic changes. The wet type of macula degeneration has been treated by photocoagulation, laser coagulation and, as the latest treatment, by combining a dye and laser treatment to destroy newly formed vessels. Treatment should be given at an early stage of the development of the new-formed vessels. At this early stage, subjective symptoms are vague and patients are seldom aware of their importance. Early diagnosis is most often made of the changes in the second eye when the central vision of the first eye has decreased.

When central vision decreases, visual acuity decreases, and texts must be large and well printed. Often there are several islands of functioning retina with varying visual acuity, contrast sensitivity and size. Therefore, visual acuity, measured during a clinical examination, may be close to normal but the patient requires large texts for reading. The patient uses a small island of vision during an examination and a larger area with lower visual acuity for reading. Optimal text size best depicts the function in the central retina. Magnifying optical devices and video magnifiers should be evaluated early in the course of macula degeneration to enable a person to continue reading. Talking books are another good solution to reading print.

Even today, patients hear from some ophthalmologists that “there is nothing to be done”, meaning that there is no surgical or medical cure. Rehabilitation is as important as medical treatment and can be an integral part of treatment. Since rehabilitation and special services depend on the ophthalmologic report, people with retinal degenerations and other vision losses should ask for a comprehensive evaluation of their vision: measurement of visual acuity alone is not enough.

In nearly all countries, there are associations of visually impaired where information on medical, educational and rehabilitative special services is available.

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### **Patients' stories**

The cake set on the table was far too enticing for me to resist, even though the treats of a buffet table pose serious traps for people like us. Still, this time I decided to succeed. I was happy to find a cake slicer, managed to take a piece of the cake, and lifted it to my plate.

“This seems like a really big slice,” I thought when balancing my way towards my seat. I raised my plate to take the first spoonful of the delicious cake. Then something hard hit my forehead. I could not help but feel with my fingers what it had been. Oh dear, I had managed to locate the cake slice, but the huge cake server was standing pointedly in the middle of my slice!

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I stepped into a chemist's from bright daylight – naturally without my cane, being a brave man. There seemed to be quite a lot of people inside the store. From earlier experience I knew that the waiting number dispenser was in the corner, so I started walking towards it.

“Oh, here it is. Now this was easy to find.”

I pressed the button and the woman in whose blouse it was moved away without a sound. Embarrassed, I walked away. Still, I was so upset by this incident that I thought I'd better explain that I could not see properly. I approached the woman again, but did not get far before she ran out of the door.

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## **Symptoms of vision loss**

### **Night blindness, delayed visual adaptation**

When the function of rod cells decreases, it leads to loss of vision in dim light. At the same time, there are changes in the cone cells; their adaptation to lower luminance levels becomes slower than normal. Delay in visual adaptation to lower light levels occurs early in the development of retinal degenerations, and is therefore a useful clinical test in vision screening among hearing impaired children, to diagnose Usher Syndrome or retinitis pigmentosa early.

Visual adaptation time can be measured by using a simple test where the person must perceive differences between red and blue at a low luminance level. After the ceiling lights have been switched off, the luminance level needs to be so low that a normally-sighted person starts to see colours after 4-5 seconds. Thus, with a test that lasts less than 10 seconds, we can screen for retinitis pigmentosa in deaf children.



**Figure 4.** Restaurants are often so dark that a person with RP cannot see properly. Spotlights that normally sighted find decorative may dazzle him/her so that even the image in the small tubular field disappears in a glare.

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### **Patients' stories**

Once, when a friend of mine was visiting I was on the floor on all fours looking for my shoes, among many other pairs under the coat rack. She has tunnel vision and in poor lighting she is practically blind. Waiting to be harnessed, her guide dog set himself obediently on the right side of his owner. To my surprise, I started feeling a hand stroking my head. This seemed somewhat strange in this situation, but I didn't mind. Then my friend started to laugh with tears in her eyes, and I realised the caressing was caused by her attempts to harness her guide dog.

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It was a beautiful sunny day. I was walking around town, when I sensed a smell of coffee impossible to resist. I entered the café. Pitch black. But it was too late to turn back, because someone behind the counter asked me what I would like to have. I ordered an espresso and a cream roll. I even used my ears trying to figure out where there were vacant tables in the café. I started walking cautiously towards a table and placed my tray on it. Finally, I crammed myself onto the chair. At the same time an angry lady stood up and spat out, "Why do you deliberately choose my table when there are nothing but vacant ones around!"

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I was at the cinema with my new girlfriend. The lights were switched off and the darkness was complete. I enjoyed the company, and the film was also interesting, until my need to go to the toilet grew unbearable. I had not yet told her about my night-blindness, and I did not consider this to be the right moment for it. I made a plan and left to find my way out. I counted my steps from the end of the row to the door of the theatre to be able to return to the right seat. Having finished my business in the toilet I opened the door of the cinema theatre – the darkness was even more complete. I fumbled in the darkness counting my steps, sat down, had a sigh of relief and leaning against my friend asked “Well, what have I missed?” After a deep silence an unfamiliar woman blurted that I had probably mistaken her for someone else.

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## Visual field changes

Loss function occurs first in the rod cells in the midperiphery of the retina. This causes patches of decreased function (scotomas) of the visual field in dim light. Changes in the function of the cone cells follow soon after and small scotomas appear in the visual field in daytime vision. These small scotomas cannot be perceived because our brain “fills them in” as it fills in the blind spot that we all have corresponding to the head of the optic nerve, where there are no sensory cells and therefore no response to light.



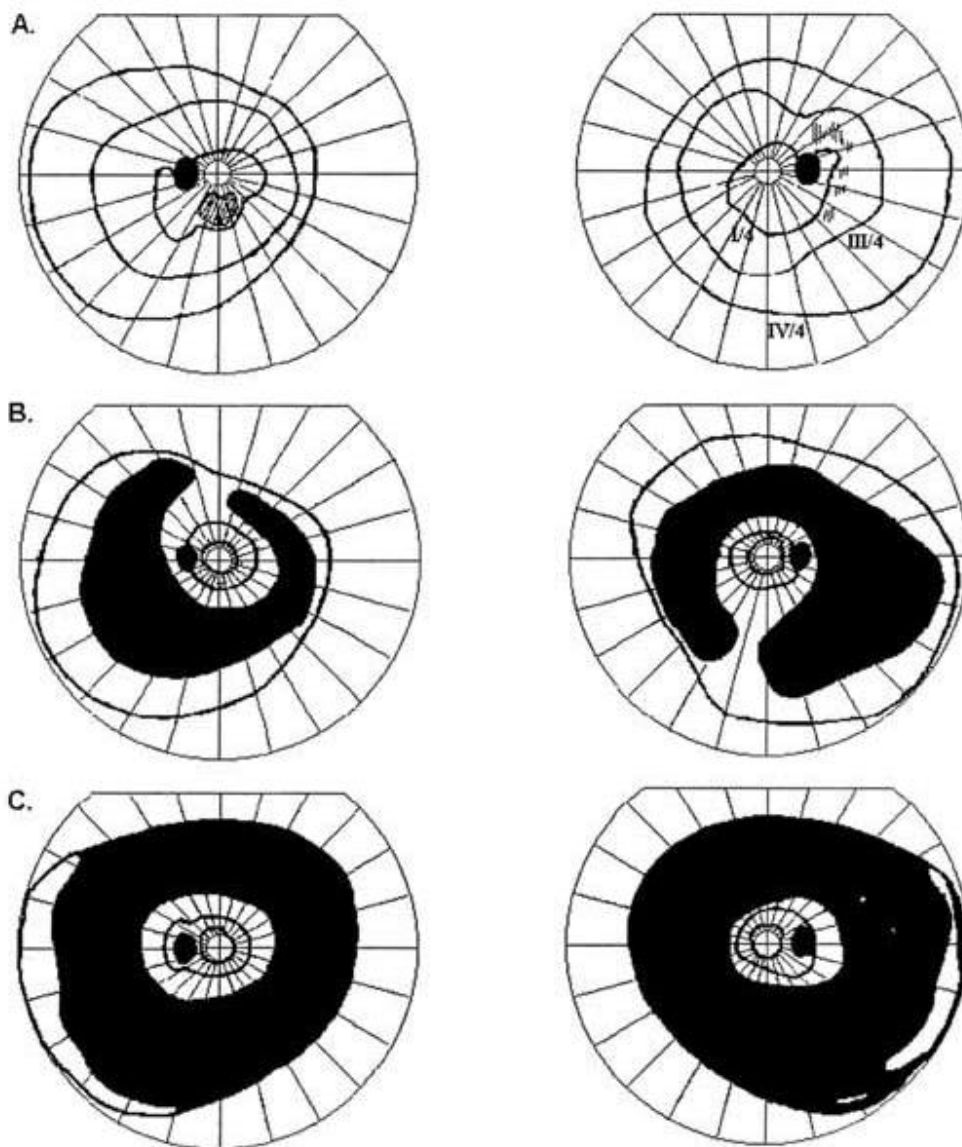
**Figure 5.** *The blind spot is about 15 degrees from the point of fixation. To see it you must test the eyes in turns. Close the left eye and direct your gaze at the cross. Move the booklet from 30cm (one foot) slowly, farther away. If you keep looking at the cross, you will notice that the circle first disappears and then reappears. When the circle is invisible it falls within the blind spot. You can find the blind spot of the left visual field by fixating your gaze on the circle and by watching the cross disappear. A person with RP may have such a limited visual field, a tubular field, that the blind spot falls outside it and cannot be demonstrated.*

When the poorly-functioning patches enlarge and become confluent, a *ring scotoma* is formed (Figure 6). A beginning ring scotoma does not disturb daily functions because the eyes move and the poorly-seeing area in the visual field falls on different details in the environment. Short-term memory summaries the image and compares it with the images in the memory and thus, subjectively, there is no scotoma. Children and young adults are often confused by the information that that are “blind areas” in their visual field when no such areas can be perceived. Especially automated visual fields often give the impression that there is a sizable loss of function when there actually is no loss of function in daylight. Visual fields are measured at low luminance levels and therefore do not depict vision in daylight. If the visual field demonstrates a ring scotoma, it is important to assess the size of the scotoma in room light (see: [www.lea-test.fi](http://www.lea-test.fi) >> Assessment of Vision>>Manual>>Part II, 3.1. Vision for Communication) and outside, in daylight.

When the ring scotoma is large enough, small objects may disappear in the visual field when they happen to be within it. This may puzzle a child when small colouring pencils or rubbers disappear and reappear on the desk. It causes unpleasant surprises also for adults with RP.



**Figure 6.** This is a photographic demonstration of the visual field that corresponds to Figure 6. The left picture corresponds to 7.A, the right to 7.B. Because the eyes move, the ring scotoma is filled by the surrounding visual information. If the person is easily dazzled, the situation would be experienced as in Figure 4.



**Figure 7.** Development of changes in the Goldmann visual field of a child/young person with RP.

A. First, the area where the small stimuli are seen decreases in size. The line marked with I/4, that corresponds to the smallest brightest stimulus, is normally as large as the area depicted by the larger spot III/4. Sensitivity in the midperiphery has decreased but the size of the visual field is still normal.

B. Later, sensitivity in midperiphery decreases more and a ring scotoma can be measured.

C. The ring scotoma of a young person has become so large that only narrow peripheral remnants of the visual field are present in the Goldmann visual fields. The area marked with black, and called the “absolute scotoma”, is absolute only in the Goldmann perimetry. There may be a good perception of larger objects and motion, movement in daylight.

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## Patients' stories

I was taking part in the Winter Seminar of the Finnish Retinitis Society in the north of the country. The programme included downhill and cross-country skiing. At the ski rental place I spotted a pair of skiing boots lying in the middle of the floor. I said that boots should not be left around like that, because we all know how easily anyone of us could stumble over them. However, the boots were not only boots; to my great astonishment they were also attached to a pair of legs which started moving rapidly.

I was shopping for food in my regular supermarket. I was cautiously walking along the aisles trying to dodge other customers. I bumped into something soft at the dairy product shelf, and realised that there was a woman crouching over to reach the milk cartons.

"I'm sorry, I didn't see you", I said confusedly.

"Are you trying to say that you could not see my enormous backside?"

As the woman began to walk away, I really could not understand how it had been possible for me not to see her.

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I was in a shopping mall with my husband. When we had taken care of our errands, we went our separate ways. There seemed to be a crowd of people at the mall, and I decided, quite impolitely, to walk from between two men talking with each other. Why won't they choose a little less crowded place for their chatting, I thought. Back home my husband described how I had crammed myself through the tiny space between those two men even though they were the only persons in the whole hall.

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I was on a holiday with my family in a rented cottage. One night I decided to watch my favourite programme on television. I was perplexed by the fact that there was no picture on the TV screen, even though the sound was clear.

"Mum, you are looking in a completely wrong direction", said our son. Then I realised that instead of the television, I had been staring at a light-coloured towel which the boy had placed on a chair after swimming.

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## Disturbing illusory light phenomena

Activation of the sensory cells causes a change in the electric potential of the cell that travels from cell to cell through the retina toward the brain. In its journey, it is affected by the function of neighbouring cells as if going through several filters. When the retinal networks are damaged an, image may spread within the retina and cause activation of groups of cells that send this activation toward the brain at the same time as it travels sideways within the retina.

The earliest illusory light phenomena are small spark like lights, “fire flies” visible in winter. Many children talk about them before school age.



**Figure 8.** Light phenomena in the visual field often have the form of a thicker or slenderer sausage. Sometimes two sausages are attached to each other. In this picture, the sausages are depicted as seen by one person with RP when talking to a person in front of her.

When the ring scotoma has become rather deep, activation may spread along its edge in the damaged network of retinal cells. People with RP describe the resulting illusory lights surprisingly similar. Their form resembles a sausage or banana or a rod. They appear one after another at a point in the visual field, move slowly around the central visual field along the edge of the ring scotoma

keeping their orientation unchanged in relation to the meridian of the visual field (Figure 8). After a few circles they disappear at another point of the ring scotoma, one after another. Coughing, laughing or a sudden movement may trigger the light phenomenon, but often they appear without any noticeable cause. The speed of movement varies from person to person and also in the visual field of a given person.

“Sausages” are not the only disturbing light phenomenon in RP. Flickering, trembling nets in front of the image or “worms” wiggling through the central visual field are rather common. Some people describe the change in image quality as a general disturbance, much like the picture on TV when there is poor quality transmission. Sometimes the image disappears in a glaring mist, when the person is dazzled. Dazzling light sources can be quite small, sometimes one candle.

Macula degenerations also cause illusory images but they are different from those caused by RP. These sometimes very dramatic illusions are called Charles Bonnet illusions, because they were first described by the French doctor Bonnet. When there is a central scotoma with distortion of the image, small losses of information and changes in colour perception the brain functions try to make sense of the incoming information and combine it with the picture in the visual memory. The resulting image may look so real that it frightens the person. There is great variation in these perceived illusions; flowers, celebrities, friends and family members appear from nowhere. Changes that clinically look much the same in siblings in a family may cause very different illusions. A patient complains that his two sisters see butterflies and flowers and he sees only streetcars.

Charles Bonnet illusions should be kept in mind if an elderly person with macula degeneration says that a deceased family member came through the wall into the room. It could well be that the disturbed image happened to become projected on a wall and it looked so real that the person could not see the difference between reality and illusion. The person is not mentally disturbed; the experience is not a true hallucination but an illusory image caused by distorted visual information.

## **Glare and dazzle**

One of the most common complaints among people with RP is glare. It has several causes. If there is a small central cataract in the posterior pole of the lens, it diffracts, spreads the light and causes “light mist” on the image. When

the number of cone cells in the central retina decreases, they cannot keep the rod pathway quiet in daylight and the activity of the very sensitive rod cells causes a very disturbing, sometimes painful glare.

Even normally-sighted people become dazzled when they walk toward a setting sun. The image resembles a black-and-white photograph developed on coarse paper with some colour details appearing now and then.

There are many special filter lenses that effectively decrease glare. A filter lens is a lens that absorbs a certain part of the light spectrum. Lenses that are often helpful when there is a glare caused by RP, absorb light in blue green. A blue green light activates rod cells. When it is nearly completely absorbed, the amount of light is at the level of twilight and the remaining rod cells function normally. As I explain to my patients, “the rod cells believe that it is night and function normally, the cone cells know it is daylight and function normally; thus there are millions of rod cells functioning at the same time as the cone cells”. When we place these filter lenses in front of our eyes, we experience a subjective increase of light, although a sizeable part of the light is absorbed in the lenses.



**Figure 9.** When a person with RP is looking toward a dazzling surface, like the white sand in this case, the details and contrast of the image may disappear. The boy resting on the grass in the shadow of the tree is likely to disappear in the shadow so that the person with RP may stumble over him or his bicycle.

The first filter lenses for RP were developed at Corning in the late 60s. These orange coloured and red lenses are called Corning CPF 511, 527 and 550. The numbers refer to the wavelength, until which the lens absorbs blue green light. The higher the number the more light is absorbed. These lenses are also photochromatic; they become darker outside when activated by UV light. They are glass lenses. There are also plastic lenses with similar absorption curves, but they are not photochromatic. Multilens filters (Multilens, Sweden) are also produced with a coating that absorbs polarised light reflected from shiny metal surfaces, white sand and water. These lenses are nicely brown.

Filter lenses are chosen outside by the person with RP, by him- or herself. It is often advisable to lend the set of filter lenses over a weekend so that the person can try them at different luminance levels.

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### **Patients' stories**

I was looking for a new shirt for the summer in the menswear department of a large store. There were dazzling spotlights on the dummies. I looked at the shirts the dummies were wearing and felt the fabric with my fingers. One of the dummies moved slightly. I thought it was only a trick of my eyes and tried again with more determination – the dummy started walking away in a swift manner.

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I was returning from a boat trip with a friend of mine on a sunny summer morning. We took a taxi in the harbour and my friend, acting as my guide, started packing our luggage in the trunk, assisted by the driver. The sun was shimmering from the metallic surface of the car so that my eyes were hurting. I thought I had better get inside the car as quickly as I could and began to fumble my way inside the vehicle. Then the taxi driver grabbed my arm and gently directed me from the driver's seat to the backseat. The driver probably thought that the girls had had quite a boat trip.

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## How does the brain see?

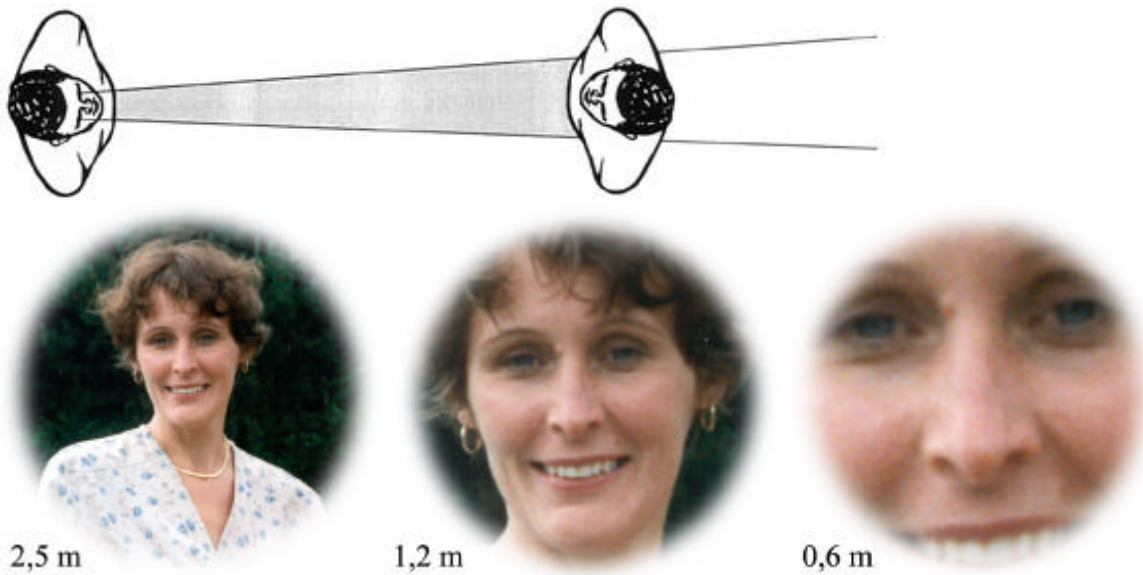
When a person with RP has ring scotoma in his/her visual field, it may cause a misinterpretation of the surrounding world because of how our brain uses visual information. Most of the inflowing visual information is not allowed into the brain functions, only the part of the image that our attention is directed at is “analyzed”. The image surrounding the area of interest is only refreshed if there is a change in it, and not always even then. Therefore, we experience that we see the room around us even if we are only actively looking at a small central part of the visual field.

The way in which our brain uses visual information makes it easier to understand why a person with RP “sees” better than the limited visual field should allow. His/her eyes scan the environment and the images are summed up into a large picture in the short-term memory. Therefore, the person may have a feeling that (s)he sees the whole street in front of him/her. When there is no function in the peripheral visual field, changes in the surrounding environment are not registered. Therefore, sometimes surrealistic images are experienced: a stroller with a baby moves by itself on the sidewalk, a person holding his hand strangely in front of him is searching to be found walking after a dog, a hat may seem to float in the air if it fills the entire visual field of the person with RP.

Tubular field causes problematic situations in moving and in communication. Therefore, the effect of a tubular field on the functioning of a person with RP should be well understood by other people (which is not the case at present). The farther away a person with tubular field is looking, the more (s)he sees. If the visual field is 10 degrees in diameter, the person sees an area 10cm in diameter at a distance of 57 cm (two feet), 20cm in diameter at 115 cm (four feet), 40cm at 2.3 metres, 4 metres in diameter at 23 metres and 16 metres in diameter at 100 metres (Figure 11).



**Figure 10.** *If a person with RP is familiar with a place, (s)he may have a feeling that (s)he sees it well, although only a part of it is visible. In this picture the clear details in the middle of the picture depict the area that the person sees within his/her visual field, the defocused part of the picture depicts the image in his/her short-term memory. If a big hat appears close by, it may fill the whole tubular field and seems to float in the air.*



**Figure 11.** A small tubular visual field may contain a large space if it is far away. However, details may then be too small to discern. This is good to keep in mind in communication.

*If a person has a tubular visual field of 10 degrees in diameter he sees a person as shown in the three photographs: the closer the person comes, the less is seen of her. A tubular field is rarely as round as in these pictures. Usually it is slightly oval.*

## Functional capability and retinal disorders

Because retinal disorders affect vision in varying ways, it is difficult, actually impossible, to choose a few visual functions to characterize the functional capability of people with different retinal disorders.

Tunnel vision in retinitis pigmentosa (RP) is disturbing in group communication, especially when sign language is used; macula degeneration (MD) makes the seeing of facial expressions and recognition of faces difficult. A person with tunnel vision may need to use long cane techniques to walk but may read without glasses; macula degeneration may not affect walking but the person may prefer talking books to reading print.

If we place the strategies used by a person with RP and another person with MD in a table, as in Table 1, we see that no single clinical measurement depicts the function or loss of function caused by retinal disorders. The common practice of saying that the loss of vision is a particular percentage is misleading. In different countries it means different things. In some countries where decimal visual acuity values are used, visual acuity 0.5 is taken as equal to 50% of vision. In other countries, the “percentage of vision loss” means what percentage of the maximum tax deduction the person is eligible to. In most countries, a low visual acuity value at full contrast means severe visual impairment. This is understandable because those who write the law are academically-schooled people with normal sight. For them, reading is an essential part of life. However, visual acuity as low as 0.2 (20/100, 6/30) may not cause a noticeable hinder in occupational life, which includes driving, computer work and demanding negotiations every day.

Visual functions and functional vision should both be assessed. Measurements of individual visual functions help us to understand the strategies a person uses in the four main functional areas: communication, moving and orientation, activities of daily life and sustained near vision tasks, like reading and writing. The use of visual acuity and the size of the visual field as the only measures of visual functioning were chosen for population based studies, and is too coarse an evaluation for decisions on functional ability. We need to know visual function at low contrast, the structure of the visual field, adaptation problems and motion perception, and assess the use of different strategies in the four main functional areas.

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## **Patients' stories**

I was an enthusiastic motorcyclist in my youth. Once I was riding fast through a wide sandy depot. All of a sudden I lost my balance, somersaulted over the handlebars and landed on the soft sandy surface. I cursed my suddenly poor bike-riding skills and my inability to maintain balance even on straight and even sand. I got up and walked back to see if the motorcycle was damaged. Dear God! Somebody had left a used car battery standing alone in the middle of the otherwise deserted depot area – and of course I had to crash with the only possible obstacle there. I had not seen it in the bright sunshine. After this incident I did not dare to mount the bike more than a few times, because I realised that even the smallest surprise could prove to be fatal.

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I walked out of the Department of Economics at my university. My intention was to go to Liisankatu street and towards the railway station. There was quite a snowfall, and after walking for a while I started to wonder why I had not yet reached Liisankatu. Finally, I had to admit to myself that I had got ten lost in a place that I was quite familiar with. I tried to find hints about my location by listening and using the little bit of remaining sight that I had. Eventually, two men who appeared from a nearby building asked if I was looking for something. I said that I would like to know where I was so that I could call a taxi with my mobile phone. The men advised me to order the taxi to pick me up at the Department of Pathology. So a half-blind person was rescued by a taxi, even from the door of the Department of Pathology.

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I arrived at a seminar a bit late. I tried to move around quietly, but coming inside from the bright outdoor light I could not see very much. Already at the coat rack I made a noise by dropping a coat hanger on the floor. Then I had to enter the seminar room, and I thought that this time I would do it smartly. I said "Good morning" at the door and started walking towards the back of the room only to bump into a hand that was extended to greet me and that I had not noticed at all. However, the ophthalmologist supervising the seminar was ready to acknowledge the situation by using the incident as a live example of problems that visuallyimpaired face in everyday life.

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I was shopping with my husband. In spite of my poor eyesight, I had come along to provide a second opinion. When he tried on a felt hat, I said "That doesn't suit you at all!" Obediently he removed the hat. Once the hat was off his head, it revealed a man that was a total stranger to me.

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## **Communication field**

When a hearing-impaired person uses sign language and/or lip reading, it is important to assess the quality of the communication field. Its size for signing and reading finger spelling is one variable, the other is the ability to read lips, if that is used by the person, and to see facial expressions.

Signing space is usually half a meter in diameter, at a distance of slightly more than one metre. Half a metre at a distance of slightly more than a metre corresponds to 25 degrees of a visual field. If a person with tunnel vision has a visual field of 10 degrees in diameter and motion perception, seeing movements is affected, signs need to be smaller and slower than normal, and the pauses between the signs slightly longer than in standard sign language. This is not taught in the training of sign language interpreters and leads to inexact interpreting, especially during the assessment of vision: the luminance level in an office varies during the assessment and there is rarely a spotlight illuminating the interpreter.

Retinal degenerations may decrease motion perception at a time when visual acuity is close to normal. The communication field should always be assessed in detail and it should be described clearly enough so that the information can be given to the interpreters.

The card is designed to give detailed information on the needs of a visually-impaired deaf person:

Communication prerequisites of a deaf-blind person:

**Name:** \_\_\_\_\_

**Size of visual field:** \_\_\_\_ degrees, meaning:

\_\_\_\_\_ cm at a distance of 1.2 metres

\_\_\_\_\_ cm at a distance of 2.3 metres.

**I use / do not use lip reading.**

**Signing with usual speed / slowly.**

**Finger spelling with usual speed / slowly.**

**I am / am not sensitive to glare.**

**Other details in interpreting:**

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## **Patient's story**

I came across a woman on the street. She greeted me cheerfully and asked how I was doing. I gave her some general answers, after which she had to leave because she was in a hurry. For a long time I thought who she might have been. She had obviously heard of my visual impairment, because she brought her face very close to mine thinking this would help me see her better – however, this meant that my field of vision could only include the gold tooth in her mouth. Try recognizing someone by that!

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## **Functional changes in retinal disorders**

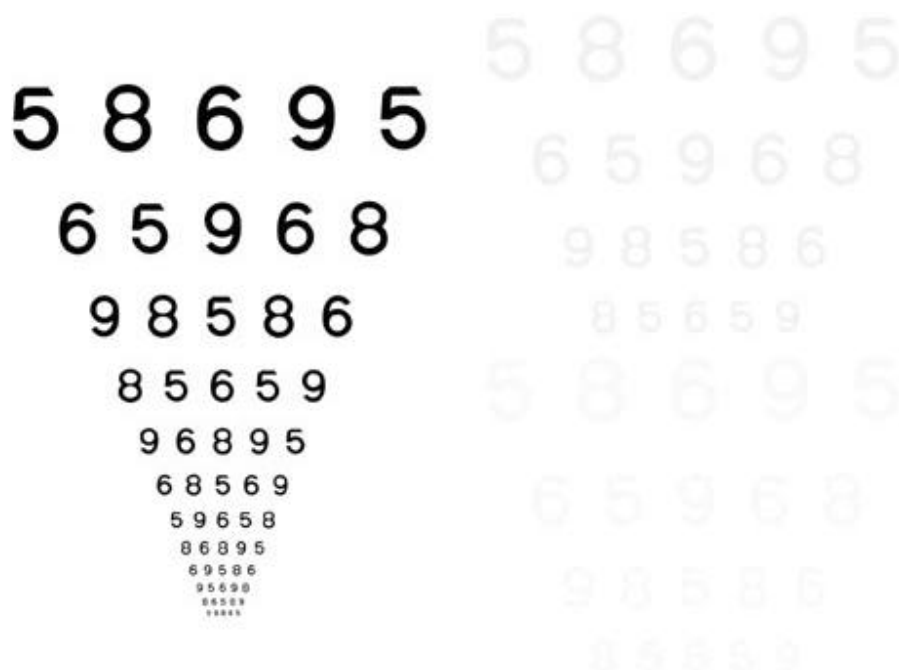
Changes in visual functioning vary depending on the location of the lesion. Macular degenerations affect more the central vision that is needed in reading and tasks demanding near vision and less or not at all the peripheral visual field that is important in orientation and moving. In retinitis pigmentosa, functional changes usually begin with delays in dark adaptation. Later loss of visual field and contrast sensitivity are typical changes. Vision at high contrast may remain good for years. It is good to know the most common tests measuring visual functions in these disorders.

## **Visual acuity**

Visual acuity is measured in vision screening before and at school age, so we have all experienced it. The tests are printed at full contrast, i.e. with dark letters, numbers or paediatric symbols. They measure function in the small area used for the fixation of gaze on details that we want to see, they tell nothing about retinal function around this area. In an extreme case, there can be normal visual acuity within less than half a degree at the centre of the large central scotoma. With such a tiny central visual field, one can only see a few letters. Also, in retinitis pigmentosa, visual acuity may remain normal, although changes in other visual functions cause a severe decrease in visual functioning.

Visual acuity is usually measured as the first function during an examination. It is important to keep in mind that refractive error may have changed after the

previous examination and thus spectacle correction needs to be fitted before the measurement of visual acuity.



**Figure 12.** Changes in visual acuity should be followed up at high and at low contrast levels. The test on the left is the usual high contrast test, on the right it is printed at approximately 5% and 2.5% contrasts. Very small details are only visible at high contrast. Changes in RP and Macula degeneration often first occur at lower contrast levels.

## Contrast sensitivity

Contrast sensitivity means the ability to perceive low contrast information, faint shadows. It can be measured similar to visual acuity, at high contrast at 2.5% or 5% contrast. To make it possible for each reader to follow up the changes and variation of contrast sensitivity and visual acuity at home, there are three different prints of the number chart at full, at 2.5% and 5% contrast, in Figure 12. Follow-up measurements are easiest to make if you measure at what distance you can barely read (= threshold value) the uppermost line and the fourth line on the chart, and write these values down. If later the distances at which these two lines on the low contrast charts are visible have become shorter, the likely explanations are either loss of contrast sensitivity or change in the refractive power of the eye. Both changes may occur simultaneously, independent of each other.

Sometimes contrast sensitivity may improve when a cataractous lens is removed or vitreous floaters sink to the lower part of the vitreous and disappear from the visual field. It may also happen that contrast sensitivity improves when macular oedema decreases.

In retinitis pigmentosa, contrast sensitivity may start to decrease quite early, but sometimes is still close to normal values in the middle of a tunnel field. When contrast sensitivity decreases, faint shadows, facial features and pastel colours are difficult to perceive. Going down stairs, noticing the curb on the street and recognising familiar people across a street may become more difficult than earlier.

When contrast sensitivity decreases, reading newspapers becomes difficult because magnification does not improve the text as before. A higher luminance level increases contrast sensitivity but in RP tolerance a higher luminance level may dazzle. Therefore, often the best way to magnify a text is to use reversal image, white or yellow text on a dark surface. The luminance of the text can be very high and yet the total luminance is so low that the person is not dazzled. The amount of light that is experienced as comfortable and the colour that gives the best contrast vary from person to person, so it is always worthwhile to experiment with lamps, filters, TV reading aids and computer screens.



**Figure 13.** *This kind of table may cause a panic reaction in a person with poor vision. Contrasts between the tablecloth and the china are poor and the candle may dazzle. Equally unpleasant would it be to cope with glass plates and reflecting crystal glasses.*



**Figure 14.** *Snow is a difficult element for people with retinal disorders, as well as poor contrast, faint shadows on cloudy days and dazzle on sunny days. If there is no sand on a path, it is impossible to see it.*



**Figure 15.** *When the tubular field is only 1-2 degrees in diameter, it is impossible to navigate on a path with no sand on it, even if the remaining central island is clear. One either follows a person (hoping that he is going in the right direction) or uses a white cane.*

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### **Patients' stories**

I was on a holiday visiting a friend who lives in France. We spent one day shopping in the lovely small boutiques of the town. When we got back to her apartment, we began trying on the new clothes we had bought. She was in

the bedroom and I was in the livingroom where I had spread my new clothes on the sofa. After some time I decided to gather my things from the sofa. My eyes picked up a clear high-contrast pattern among the pillows. I decided to start my sofa-clearing from that and grabbed it firmly – only to hear a loud scream! I was frightened and could not instantly understand what had happened. Then I blushed with shame, until laughter won over the embarrassment. The laughter spread also to my friend who had came back to the livingroom to sit on the sofa without my noticing it and whose blouse pocket with its clear pattern I had grabbed.

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I came to the bus stop with two large plastic bags in my hands. There was a little boy sitting on the bench. I lifted the bags in my lap and started to set myself, sitting next to him on the bench when I felt something soft underneath me. First I thought that the child had placed his school bag there, but to my horror I noticed that there was another child sitting next to him. I stuttered something about my sight problems. Standing up and still embarrassed, I looked at the child on whose lap I had sat. In the bright sunshine the colour of his clothes was completely similar to the colour of the bench.

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## **Visual field**

There are numerous different instruments for the measurement of a visual field. They are either called perimetres or campimetres. The old gold standard was the Goldmann perimetre, a large white cupola on which small light spots can be moved toward the centre to record where the person can detect a spot of a certain size. The modern automated perimetres most often use minute spots of light that are presented very briefly, and therefore look much like the “sparks” or “fireflies” that spontaneously appear in the visual field in RP. It may be difficult to decide when a small light is a test light and when it is an illusory light.

Automatic perimetry may result in a recording that shows an “absolute” ring scotoma at a time when the person has no ring scotoma in a Goldmann perimetry. Automated perimetry often gives the wrong idea about the functional visual field. Recent studies have shown that also a Goldmann perimetry may show “absolute” scotomas that do not affect a person’s function. The luminance level at which the Goldmann field is measured is low, so in daylight visual

function may be much better. Scotomas should be evaluated using flickering stimuli, whenever possible to document the function of the magnocellular pathway that transfers low contrast visual information in different shades of grey and motion information, but no colour information. A good example of difficulties in the assessment of a visual field is the case of one of the best young Finnish ice hockey players who has a large “absolute” scotoma in the lower part of both visual fields in the Goldmann perimetry. Never the less, he responds to flicker perimetry within the scotomas at every point measured with a 4 degree stimulus.

#### Measurement of flicker sensitivity and motion perception

These two measurements have been used in experimental clinical investigations for nearly 30 years, but simple clinical measurement techniques are still waiting for their designers. It is selfevident that the perception of motion, movement, is essential when moving. It may be affected early in the course of RP, when the visual field is still large and visual acuity good. Typically, such a person bumps into objects as if (s)he had tunnel vision. Moving in a crowd on streets or stores becomes nearly impossible because one cannot estimate the speed of movement of other people, and bumps into people. Some people with RP describe that they cannot walk outside in places where branches of bushes and trees move in the wind, and other people with RP must wait for a while when they turn to allow the image to “catch up”. The image seems to stay behind and reaches the correct direction with a delay. Then it is impossible to use the tubular field as a long cane.

Sensitivity to flicker may increase in certain retinal conditions. If a person becomes dizzy and nauseated when working at a computer screen, it is wise to test whether the symptom disappears when the person uses a screen that does not flicker at all. In monkeys, it is possible to measure responses in the midbrain to flicker stimulation up to 120 Hz flicker.

## **Colour vision**

Colour vision may remain close to normal quite late in RP. Since the number of blue sensitive cone cells is smaller than that of red or green sensitive cone cells, shades of blue and blue green are often the first one to become problematic. Very few persons with RP have liked to pick blueberries in their childhood. The blue end of the spectrum becomes increasingly difficult with advancing age, and at the same time confusions appear between other colours: light pink and yellow look the same, while moss green, snuff brown, dark burgundy and dark grey all look the same.

Even a normally sighted person confuses the colour of dark socks in a bedroom with warm Rosalux lights. Daylight lamps are useful in colour discrimination. It would be a great help if they were used in shops, especially in clothing shops.



**Figure 16.** *The recognition of colours is difficult for many people with RP. Problematic pairs are for example blue and green, yellow and pink, brown and violet. All dark shades seem black regardless of colour.*

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### **Patient's story**

There was still time before the departure of my train, so I decided to have a look at the sale in a nearby store. After looking at the clothes for some time, I decided to try on a burgundy-coloured trouser suit. It fit like a glove. I decided to keep the top of the suit on and went to the counter to pay. At the platform I glanced into the bag in which the trousers of the suit were. I was shocked to discover that they were actually mustard-coloured. Well, that was not a very good bargain, I thought: a burgundy jacket and mustard-coloured trousers. I was upset by this for the whole of the train journey.

Next morning I went outside to look properly at the colour of the clothes. Oh my God: the top and the bottom were of the same colour, but the colour was regular brown. In the evening my husband came home from work and I asked him what the colour of the suit was.

"It is really beautiful burgundy." "You mean, both the top and the bottom?" I asked confused. "Yes" answered my husband and so I became a very happy owner of a new trouser suit.

## Who is the RP Personality of the Year?

If you meet the following requirements, you can be a candidate for the Survivor of the Year Award!

1. You have not made any lampposts sway in a year.
2. You have not tried to enter a taxi through the boot.
3. You have not been kicking tomatoes or mandarins around on the floor mistaking them for children's toy balls.
4. You have not wiped any dishes from the table on to the floor.
5. You have not been picking up meatballs from the stove on the weekend, the ones you cooked earlier in the week.
6. You have not put butter milk instead of milk nor salt instead of sugar in your coffee.
7. You have not called your dear husband to come in when the caretaker was walking in the yard.
8. You never sat on a black cat sitting on a dark-coloured sofa.
9. You have not kicked over the cleaner's water bucket in the dark hallway of your block of flats.
10. Nothing out of the ordinary has happened to you during the whole year.

