

A photograph of a young man with short blonde hair, seen from the back and side, looking forward. He is wearing a dark jacket. He is walking on a paved path that runs alongside a body of water. In the background, there is a modern building with large windows and a bridge. The trees have yellow autumn foliage.

# **LOOK >> FORWARD**

*Methods, techniques, comfort and possibilities related to LHON and similar conditions*

# ***LOOK >> FORWARD!***

*Facts, methods, techniques, comfort and possibilities related to LHON and similar conditions*

## **LOOK FORWARD!**

*Texts: Gunnar von Sydow, Krister Inde, Helena Lindemark, Hampus Wännerdahl, Jens Pettersson, Ola Carlsson-Fredén, Christian Fabienne, Johan Wingestad, Bertil Sköld, Jan Eidvall.*

*Editorial staff and editing: Gunnar von Sydow and Krister Inde*

*English translation: LanguageWire AB/Sheila Widingsjö Söderberg*

*Photos: Maria Annas pages 7, 24, 26, 29, 32, 34, 36-38, 41, 45, 46, 54, 58, 82, 86, 104, 108, 112; Helena Lindemark pages 62, 63; Stefan Wettainen page 68; Ola Carlsson-Fredén pages 88-99*

*Illustrations/drawings: Rune Johansson page 17*

*Graphic design, cover: Bulldozer Kommunikationsbyrå AB, Karlstad*

*Graphic design, content: Maritha Frykelid, CityTryck i Karlstad AB*

*Printing: CityTryck i Karlstad AB*

*Paper: The cover has been printed on Ensocoat, 250 g, and the content has been printed on Tom&Otto silk, 150 g.*

*Funding: The book is part of the Leber X Project, which is funded by Allmänna Arvsfonden [Swedish Inheritance Fund]. The project is run in partnership between the non-profit organization LHON Eye Society and the adult education centre Hagabergs folkhögskola in Södertälje, Sweden, and is headed by Bertil Sköld. The translation into English was funded by Santhera Pharmaceuticals.*

*This book has been produced as an audio book with ABF and the Swedish Association of the Visually Impaired (SRF).*

*Look Forward! is also available in a web version at [www.lhon.se](http://www.lhon.se), along with supplementary materials, references, videos and updates.*

*The book's first edition of 800 copies will be available in January 2017.*

*Additional copies can be ordered from [kontorsservice@srf.nu](mailto:kontorsservice@srf.nu) or Swedish Association of the Visually Impaired (SRF),*

*122 88 Enskede, Sweden. Phone 08-39 90 00.*

*© Copyright LHON Eye Society*

*ISBN 978-91-639-2643-3*

# CONTENTS

About this book	4	<b>TRAINING TO LIVE WITH A LHON PERSPECTIVE</b>	
Preface	6	Magnification - the solution for impaired vision	68
		Build up your eccentric viewing ability!	76
<b>MEDICAL PERSPECTIVE</b>		Keep your body in shape	83
Learn more about LHON	8	Improve your lighting environment	86
		IT aids in everyday life	100
<b>PSYCHOSOCIAL PERSPECTIVE</b>		10 tips for getting your dream job	104
A battle to be won every day	25	Higher-level studies - better job	108
Félicia and Patrick - a Cinderella story	32	Conclusions from the Leber X Project	114
Björn Carlsson - You learn to be an optimist	39	Resources at the national and regional level	117
Malin Wixner - Nothing's impossible	42		
Anton Blomberg - Attitude is everything	44		
Susanne Boström - Show them what you've got	47		
With new conditions	50		
Arbetsförmedlingen, Försäkringskassan	55		
Family of someone with LHON	59		
My son developed LHON	61		
Many different ways to react	64		

# HOW THIS BOOK CAN HELP YOU LOOK FORWARD

Thank you for choosing to browse through this book. It has been published by the LHON Eye Society with much-needed funding from Allmänna Arvsfonden [Swedish Inheritance Fund]. It is part of the Leber X Project, which focuses on gathering knowledge and experiences about the rare disease LHON.

The Leber X Project was launched in 2015 with a number of meetings at different locations in Sweden to discuss the opportunities and challenges of living a good life with a severe visual impairment. It was at that point that material for this book began being collected.

The physical book you are reading right now has its limitations. It is in printed form and therefore will most certainly become outdated. Hopefully, this will happen quickly. Who knows? Researcher could make a crucial breakthrough at any moment!

Everything found in this book has also been pub-

lished on the Leber X section of the society's website, which is found at [www.lhon.se](http://www.lhon.se). A number of videos created for the project can also be found there. In these videos, a number of people with LHON talk about what it is like to adapt to the challenges caused by a visual impairment. Some of the videos explain how to best utilize one's remaining vision, particularly how to build up abilities in eccentric viewing, which is the art of focusing on peripheral vision.

Look Forward! reflects the ambitions of the Leber X project. The book begins with a thorough review of the medical aspects - heredity and symptoms, the state of research, and the scientific view of the positive and negative effects of various medicines and other products.

This is followed by a few chapters on the psychosocial aspects, where a number of individuals with LHON talk about how they reacted to the diagnosis

and how they manage to live a good and meaningful life with vision disciplinary action.

A lot depends on finding a job that one is suited to, enjoys and can handle. One chapter discusses how to get a job and what assistance is offered by Arbetsförmedlingen [Swedish Public Employment Service] and Försäkringskassan [Swedish Social Insurance Agency].

Because LHON impacts the entire family, we have devoted a chapter to how family and other loved ones are affected. How should you treat someone who is suffering from an often unexpected visual impairment? This is a question that is also relevant for social workers and others that individuals with LHON come in contact with. The role of the social worker is addressed in its own chapter.

One section of the book discusses how best to handle your visual impairment. It contains concrete descriptions of how to use magnification as effectively as possible. As previously mentioned, it also talks about how to build up your eccentric viewing ability.

Many with a visual impairment easily develop a habit of not being very mobile and sitting in the same position for hours on end. So, we have provided some tips on simple physical exercises that are effective.

There is a lot that can be done with lighting to better orient yourself at home and at work, and quickly switch between work lighting and comfort lighting – and avoid dazzling, which is a major annoyance to many with LHON.

To a large extent, being able to cope well is a matter of knowing about and managing various aids. This book focuses on IT aids, but over time other aids will also be described on the website (IT aids also develop at a rapid pace – the website will keep a watchful eye out for new technologies).

The book concludes with a few chapters on the art of seeking a job and earning a degree, which is often a must-have in order for a visually impaired individual to get a good job.

At the end of the book, you will find contact details to organisations, authorities and doctors who can provide additional advice and information.

Pleasant – and hopefully helpful – reading.

And do not forget – [www.lhon.se](http://www.lhon.se).

Go to the Leber X tab.

# YOUR NEW LIFE NEEDS A GOOD START

*Look Forward! is a kind of starter kit for those of us with a visual impairment. Although we may not even be able to discern the face of the person we are standing and talking to, we can nonetheless look forward to a good future.*

My life changed practically overnight when I was diagnosed with LHON in 2009. I felt lost as I tried to work my way through new challenges and possibilities. When the LHON Eye Society was formed, I came into contact with others in the same situation. We soon realized that a starter kit like this would be invaluable. We all felt like we could have really used a guide through this unfamiliar landscape. And now the starter kit stands ready - another sign that things are moving forward and that we with LHON and other causes of visual impairment will be better equipped to live a good life.

## **SORROW, ANGER AND ACCEPTANCE**

Developing a visual impairment means having to go through a process similar to the grieving process when a loved one passes away. Sorrow and anger are needed before acceptance and optimism can settle in. This book provides an introduction to these important psychosocial powers that are put in motion. It will guide you through what you need to be able to pursue an education or work while having a visual im-

pairment. The book also talks about the current state of medicine and research. Thus, it may also be helpful to healthcare professionals, employers, Försäkringskassan, Arbetsförmedlingen, vision centres, schools, universities, and many others.

Look Forward! also presents the ever-growing array of aids, particularly the new IT-based ones, and provides concrete tips on how to improve your home and your workplace by installing the right lighting.

The content of the book is also compiled on a separate website, which includes additional materials and even videos. The website is also intended to serve as a forum for continued dialogue with the readers - all stakeholders and concerned parties. We also hope to be able to use the website to help you follow the development of aids, and new research findings.

## **LEBER X AND ALMÄNNA ARVSFONDEN**

Look Forward! was produced as part of a project called Leber X on the initiative of the non-profit support organization LHON Eye Society. I would like to thank Allmänna Arvsfonden [Swedish Inheritance Fund] for

the generous support that made the project possible. I would also like to thank the adult education centre Hagabergs folkhögskola in Södertälje, Sweden - particularly everyone who worked so hard, arranged meetings and seminars, conducted interviews, wrote pieces, took photos and videos, and contributed their knowledge and experiences!

The LHON Eye Society is an organization for individuals diagnosed with LHON, individuals who are carriers of one of the three mutations, and family and others who do not have the trait themselves but live in an LHON world. We want to support everyone living with a visual impairment, including their family, and contribute to active research that makes real progress.

Helena Lindemark started the association in Stockholm in March 2012, when her son's vision deteriorated at age 14.

#### **WE NUMBER IN THE HUNDREDS**

We estimate that in Sweden there are about 200 individuals in the same situation and some 60 families that have mutated mitochondria that could lead to LHON and thus impaired vision.

A lot of progress has been made over the past 40 years. But, we expect even greater progress over the coming 40 years. By 2060, maybe we will all be able to LOOK FORWARD!

*Hampus Wännerdahl,  
Chairman of the LHON Eye Society*





# LEARN MORE ABOUT LHON

## CAUSES OF LHON

LHON stands for Leber's Hereditary Optic Neuropathy. LHON is a rare disease caused by a mutation in a specific section of the mitochondrial DNA (mtDNA). It is point mutation, which means that only one single building block of the mitochondrial DNA has been changed or replaced.

Mitochondria are small entities that are found in all the cells of the body and act as the "power plants" of the cells. Through chemical reactions in the mitochondria, the energy we take in as food is converted into forms that can be utilized by the various organs of the body. With LHON and other mitochondrial diseases, the mitochondria do not work properly.

LHON was first discovered in 1858, and was named after Professor Karl Gustaf Theodor von Leber, who 13 years later (1871) described the disease in 15 patients - mostly young boys and men - in four different families.

It was not until 100 years later, at the end of the 1980s, that it was determined that the disease is due to a change in the mitochondrial DNA. Theodor Leber also lent his name to another eye disease, Leber Congenital Amaurosis (LCA), however this has no link to LHON beyond the person who first described the disease.

The point mutations that can cause LHON usually

affect vision only. It is primarily the central visual field, where vision is its sharpest, that disappears completely or partly and becomes blurred. Most who develop the disease develop a severe visual impairment, which usually makes its appearance when young or at midlife. Although the visual impairment reverses itself in some cases, for most it is life long and they have to learn to live with the level of vision they have left. There is currently a great deal of research under way, which will be described later in this chapter.

## LHON IS A RARE DISEASE

At present, there is no definitive knowledge of how many have the disease in Sweden, but the idea of creating a registry in Sweden is being discussed. Based on the prevalence of the disease in Denmark and Finland, it is estimated that 180-200 individuals in Sweden have developed the disease. The incidence in Finland is estimated as 2 per 100,000 inhabitants, while in Denmark it is 1 per 54,000 inhabitants. Since it has only been possible to diagnose LHON with certainty through DNA analysis since 1988, there are probably a number of people who have not yet been correctly diagnosed. It is also possible to be a carrier without developing the disease. In northern England, estimates show that about 12 out of 100,000 inhab-

itants are carriers of some type of LHON mutation. An Australian study of how many individuals with a severe visual impairment have LOHN determined that the figure is about two percent.

Four times more men develop LHON compared to women. In Denmark, 88 men and 22 women were registered in 2015.

#### **HEREDITY AND THE RISK OF DEVELOPING THE DISEASE**

Because LHON is caused by a mutation in the mitochondria, the disease is passed down from the mother since only the woman's mitochondria are inherited. Although it affects more men than women, men will not pass the disease along.

Almost everyone with LHON (over 90 percent) carries one of three different LHON mutations, known as m.11778G>A, m.3460G>A and m.14484T>C. To keep

things simpler, going forward we only use the numbers, which indicate where in the mitochondrion the mutation is located.

The risk of developing LHON and the visual impairment becoming permanent depends on factors such as what type of mutation the person has. Of the three most common LHON mutations, the one with the best prognosis - that is to say the best chance of the disease reversing - is mutation 14484. Research has shown that about half of those who carry this mutation can regain some or all of their sight. For 3460, the proportion is around 25 percent, while the chance of regaining vision is lower than this for the most common mutation in Sweden, namely 11778.

In addition to the more common mutations, there are additional point mutations that are located at a different spot in the mitochondria and are labelled



Normal vision



Vision with LHON

*LHON varies greatly from person to person, but can lead to vision like that depicted in the image on the right.*

with other numbers. They have also been shown to be capable of producing LHON with the same or similar symptoms. It may take longer to arrive at a reliable diagnosis in these cases, particularly if the family history of LHON is unknown.

This is because it is possible to be a carrier of a LHON mutation without developing the disease. So far, research has been unable to determine why some people develop the disease and others do not. One important aspect is the proportion of mutated or altered mitochondria. But, it is possible to have 100 percent mutated mitochondria (called "homoplasmy") without developing the disease. Although most with the trait have homoplasmy, in 10-15 percent of carriers only part of the mtDNA is mutated (called "heteroplasmy"). Generally, a lower proportion of mutated mitochondria reduces the risk of developing the disease. But, there are rare cases where persons develop the disease even with heteroplasmy of 50 percent or lower. Thus, in order to develop the disease there must be other factors beyond carrying the mutation.

There is a great deal of research about what could trigger the disease. Among other things, researchers have studied whether toxins found in smoke, some chemicals, and some type of medicines and drugs could increase the risk of the disease being triggered. If you know you are carrying the LHON mutation, it is a good idea to be careful and ask for advice on what to avoid. Plus, you should completely refrain from smoking.

LHON is four times more common in men than in wom-

en. On average, about 50 percent of men who carry the mutation develop LHON, while only 10 percent of women develop the disease. The reason for this is not known. Some researchers believe the X chromosome may have a protective effect. Since women have two X chromosomes and men only have one, this could partly explain why the disease is so much more common in men. There is also research studying whether the female sex hormone, oestrogen, could have a protective effect, but no clear link to this has been established as yet.

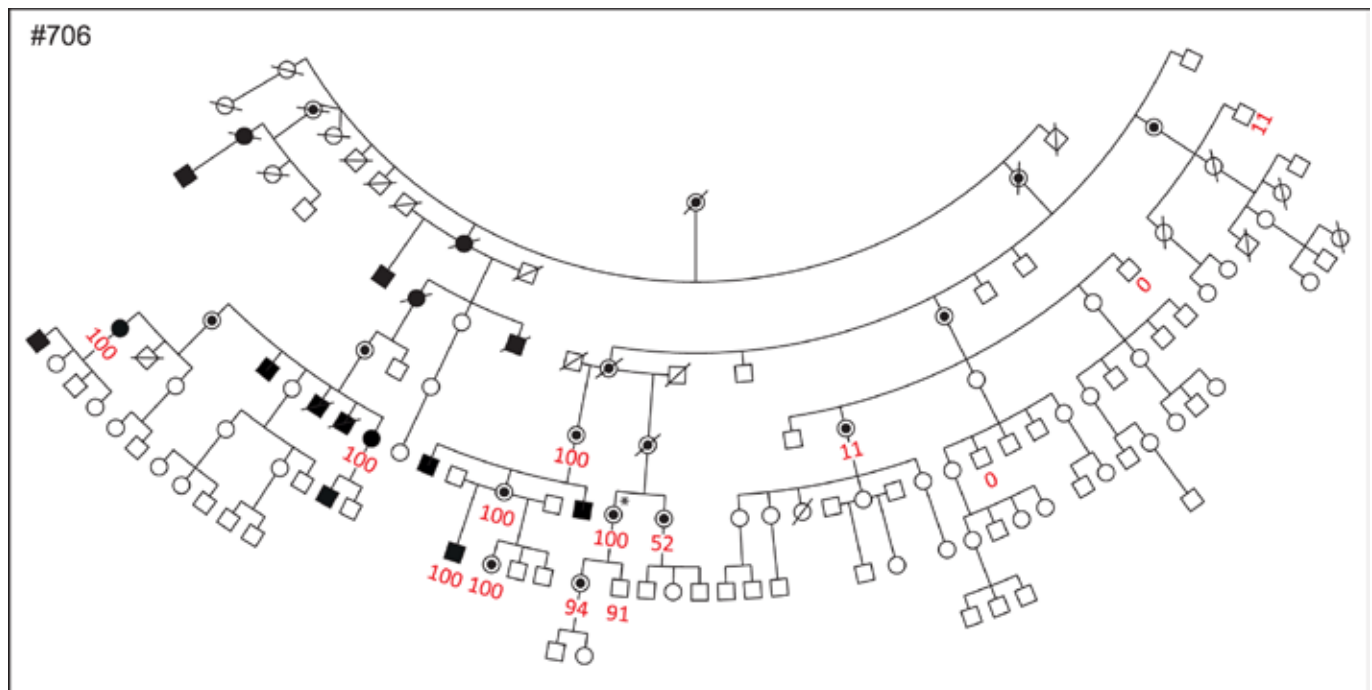
Researchers have also found other hereditary factors in mtDNA that affect the risk of developing LHON. Through the course of evolution, humans have developed various haplogroups. All individuals belong to a specific haplogroup, each of which is defined by inherited changes in the genome (called polymorphisms) that are not pathogenic. The combination of belonging to a specific haplogroup and being a carrier of one of the LHON mutations could put someone at a greater or a lesser risk of developing the disease. Researchers are also looking into whether there can be a certain haplotype on the X chromosome that increases the risk of developing a visual impairment, and some correlations have been found. But, the haplotypes contain a number of different genes, so the research is complicated. So far, it has not been possible to identify which gene or genes increase the risk of the disease.

#### **AN INTERESTING DANISH STUDY**

In Denmark, there is good knowledge of the number

of cases of LHON. There is also knowledge of how many and within which families the LHON mutations are found. A Danish study published in March 2016 studied the incidence and heredity related to LHON in Denmark. The study focused on how the proportion of mutated mitochondria can be distributed in different ways within families/relations. The figure below shows a family carrying mutation 11778 that was able to trace the disease back through seven generations on the mother's side. The first woman in the family that is known to carry the trait was born in 1842. They were then able to follow who developed the disease and to some degree who was a carrier in later generations.

Two different branches of the family tree developed in totally different directions. In the figure, a circle symbolizes a woman and a square a man. A solid circle or square indicates that the person has/had LHON. A circle that is partly filled in means that the woman was a carrier, while the red numbers indicate the proportion of mutated mitochondria of the person in question. Squares and circles that are left blank and do not have a red number indicate that the person in question did not develop LHON, but it is not known whether that person is a carrier or not. Figures with a line through them indicate that the person in question is deceased.



## **OTHER FACTORS THAT CAN AFFECT RISK**

In addition to hereditary factors, there are a number of substances and factors that are believed to be possible triggers or to increase the risk of a person developing the disease. For example, researchers have found a substance in smoke (both tobacco smoke and other smoke) that they think may have a triggering effect. They also looked at substances in certain medicines, drugs and toxins that are suspected of affecting mitochondrial function, which could cause them to have a triggering effect. Since the substances are believed to adversely affect mitochondrial function, they could - purely hypothetically - also adversely affect individuals who have already developed the disease. There is no evidence that there is anything that worsens the disease in individuals who already have a visual impairment. But, to be on the safe side - whether a carrier or diagnosed with the disease - it is a good idea to have an idea of things that could have a negative effect and - naturally - those that could have a positive effect.

## **PHYSICAL EXERCISE = POSITIVE!**

To start with something positive, research has shown that when you move your body - cardio workouts in particular - the body forms more mitochondria. It is not completely certain whether this is positive or negative for LHON, especially if the person in question has 100 percent mutated mitochondria. On the other hand, physical activity is good for the whole body, so overall it is definitely positive to get your body moving. It is also be-

lieved that certain vitamins and the coenzyme Q10 could have a positive effect. Since LHON relates to a problem with the body's energy production, getting plenty of rest and a healthy diet should also have a positive effect.

## **TRIGGERING FACTORS - HARD TO KNOW FOR SURE**

Since LHON is a rare disease, it is hard to compile statistically reliable data about what can trigger the disease. There is much research on this, and new reports are continuously being published. Researchers are not in agreement about what applies, but it is a good idea for people who carry the trait or have developed the disease to know as much as possible - partly because knowledge among healthcare professionals is limited and partly to be able to apply the "precautionary principle" themselves. Here are some factors that various researchers believe may have a negative effect as of 2016 (For more complete and updated information, please see the references to various websites on [www.lhon.se](http://www.lhon.se)).

## **SUBSTANCES THAT MAY HAVE A NEGATIVE IMPACT**

Smoking is known to have a negative effect on mitochondrial function and increase the risk of triggering LHON. Researchers have also studied different types of substances that are suspected to adversely affect mitochondrial function. Such substances include certain bacterial toxins, mycotoxins, snake venoms and the environmental toxin "rotenone", which was previously used to treat lakes prior to the planting of salmonid fish. There is also research investigating certain medi-

cines that are believed to have negative effects.

The Swedish National Board of Health and Welfare's website provides information on mitochondrial diseases. It also indicates medicines that are believed to inhibit the mitochondrial respiratory chain and could thereby have negative effects for individuals with a mitochondrial disease like LHON. Such medicines include propofol, valproate, tetracyclines, chloramphenicol, linezolid, and barbiturates. Researchers are not in agreement on this, but caution and discussion with your doctor is a good idea if you carry any of the LHON mutations. More information is available in Swedish for both patients and healthcare professionals at FASS as well as the Swedish Medical Product Agency's Läkemedelsboken [Pharmaceutical Handbook].

Large amounts of alcohol, mental stress, and physical injury/head trauma are other factors that some researchers think might be related to whether a person develops LHON or not. A combination of several different factors could further increase the risk.

There are many cases where no possible triggering factor was found beyond heredity and that the person in question is male. Examples of this include children who developed LHON before the age of ten without being exposed to any known triggering factors. In such cases, the person's haplogroup and other hereditary factors may be involved. In a similar manner, some carriers may have been born with or developed some form of protective factor that reduces the risk of developing the disease.

The list below briefly describes a number of fac-

tors, including medicines, that are believed to have a negative effect:

- **Smoking/smoke** - Cigarette smoke as well as other types of smoke contain toxins that could affect and reduce the number of copies of mtDNA in the cells. Smoke can also negatively impact mitochondrial function and energy production.
- **Industrial toxins, environmental toxins and toxic chemicals** - Rotenone is one environmental toxin known to adversely affect mitochondrial function. Some industrial toxins and chemicals can also have a harmful effect.
- **Drugs, narcotics** - It is important for individuals with LHON mutation to know that the risks associated with drugs are greater for them than for others. Because of the clear link between toxins in smoke and mitochondrial effect, it is especially important to stay away from drugs that are smoked. Cocaine and ecstasy are other drugs that have been shown to have a particularly negative effect on individuals with LHON mutation. With the link between LHON and the intake/effect of toxins and chemicals, other types of drugs should definitely be avoided as well.
- **Alcohol** - This link is less clear, but researchers recommend that you avoid consuming large amounts of alcohol if you carry a LHON mutation or have LHON. However, no clear link to LHON has been seen with moderate consumption. A combination of multiple factors, such as smoking plus a high level of alcohol consumption, increases the risk.

- **Head injury** - Some researchers have investigated cases where the individual suffered some form of severe trauma or blow to the head. However, they could not determine whether it was a coincidence or there is a causal relationship.
- **Mental stress and/or serious illness** - Some researchers have identified a link between mental stress and development of LHON. Others have also found cases where the individual developed LHON shortly after a serious illness.
- **Vitamin B12 deficiency** - Some patients with LHON have been found to have a vitamin B12 deficiency. However, it is not known whether this triggered LHON or there are other underlying causes of the B12 deficiency. B12 deficiency can cause poor vision in and of itself without LHON being diagnosed.
- **Starvation/low nutrition intake** - This can also have a negative impact and is believed to be a triggering factor in some cases.
- **Medicines** - Knowledge about LHON is limited among healthcare professionals, and there are quite a number of medicines that could negatively affect individuals with LHON or could trigger the disease. LHON is a rare disease, and as a result there is a lack of evidence-based research with real-life experiments. Knowledge about which medicines could cause negative effects is therefore primarily based on theoretical studies. For individuals who carry the trait or have been diagnosed with the disease, it is good to know which medicines these are, even

if negative side effects are only suspected. This makes it easier to have a discussion with your doctor, and use these medicines with caution or choose other options if possible.

In many cases where there were side effects such as impaired vision, the vision returned to approximately the same level as previously once treatment ceased. In general, it is good to be careful with medicines and vaccinations that FASS or the patient leaflet indicate have a high risk of side effects that affect metabolism and/or the central nervous system. A list of some medicines that researchers suspect could be harmful in connection with LHON is found below. Note that the list is not exhaustive since it is difficult to assess side effects for individuals with such an uncommon diagnosis as LHON.

#### **MEDICINES THAT MAY BE GOOD TO AVOID**

Some types of antibiotics are believed to have negative effects. At the 2015 LHON Conference organized by the United Mitochondrial Disease Foundation (UMDF) in the United States, participating researchers agreed that a standard antibiotic treatment would probably not have negative effects on LHON mutation carriers. The types of antibiotics that researchers have found to have adverse side effects in connection with LHON mutation include the following (for more information and references, see [www.lhon.se](http://www.lhon.se)):

- Erythromycin
- Streptomycin

- Tetracycline - There is research both about possible negative effects and a possible protective effect.
- Minocycline - For this medicine as well, research shows contradictory effects on mitochondria.
- Ethambutol
- Linezolid
- Isoniazid - The research on this medicine's possible negative effects is outdated (1994) and it is doubtful that the conclusions still apply.
- Telithromycin
- Chloramphenicol - A type of antibiotic that was de-registered as a medicine in Sweden, but is available in other countries and as eye drops and ointment in Sweden.
- **Cyanocobalamin** - En form of vitamin B12 that could have a negative effect. However, studies investigating this are outdated and it is doubtful that the conclusions still apply. When treating B12 deficiency, it may be useful to consider choosing a drug with another active substance in connection with LHON, suspected LHON, or trait carriers, for example Hydroxocobalamin.
- **Propofol**
- **Tadalafil**
- **Barbiturates**
- **AZT**
- **Lactated Ringer's Solution**

Please visit [www.lhon.se](http://www.lhon.se) for references to current research on the medicines in this chapter.

## MORE ABOUT MITOCHONDRIA AND CAUSES

Every cell of the body contains a number of tiny structures that have important functions. The primary role of these structures, known as mitochondria, is ensuring the body converts enough energy to enable us to perform actions like moving about, growing, thinking and seeing. They also play a major role in controlling cell development, rejuvenation and death. Each cell is made up of a large number of mitochondria. They are believed to have originated from bacteria contained in primitive cells more than 1,000 million years ago that enabled the cells to utilize oxygen. Within the mitochondria, a large number of chemical reactions occur to convert energy in the various organs of the body. The LHON mutations in the mitochondrial DNA affect three different genes that serve as templates for producing proteins that are part of "enzyme complex I" within the mitochondrial electron transport chain. The change impairs the mitochondrion's capacity to produce energy.

In total, there are about 200 known mitochondrial mutations that can produce completely different effects. It is not fully known why the typical symptoms of LHON mutations involve visual impairment. The location of the mutations within the mitochondrion is an important aspect. Another is believed to be that a great deal of energy is required for the eye to be able to see. Although it is more uncommon, there are even some cases where other parts of the central nervous system are affected in connection with LHON. Such cases are usually referred to as "LHON Plus".



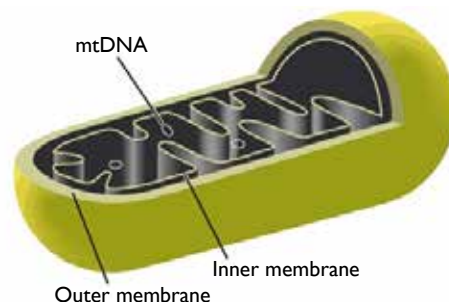
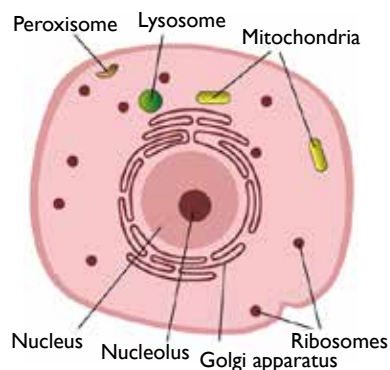
## WHAT HAPPENS WITH LHON?

LHON generally involves damage to the ganglion cells of the retina. The retina has more than one million ganglion cells, which receive electrical signals from the photoreceptors (rods and cones). Each ganglion cell has a nerve fibre (axon). All of these ganglion cell axons join together to form the optic nerve, which transports the signals to the brain's rear optic tracts and visual cortexes. It is not totally clear why the retinal ganglion cells are affected, but these cells contain a large number of mitochondria and are therefore

and optic radiation to the visual cortex.

A person with a LHON-based visual impairment first develops swelling in the front part of the optic nerve. During a fundusoscopic examination, swelling and redness are seen in the optic nerve head (optic disc or "blind spot"). Magnetic resonance imaging (MRI) often shows a thickening of the optic nerve and an increased signal in the acute stage.

After a few months, many of the ganglion cells have died. The swelling and redness of the optic disc has disappeared and is replaced by optic disc pallor. The op-



*The illustrations depict images of a cell (on the left) and a mitochondrion (on the right)*

presumed to have a high energy requirement.

The photoreceptors themselves (the rods and cones) are not affected. Damage to the ganglion cells affects the entire optic nerve from the retina in to the switching station for the optic tracts in the brain. The figure shows a simplified image of the optic nerve from the eye's retina to the brain's visual cortexes, via the optic chiasma

tic nerve appears thinner in an MRI. The course of the disease may be documented and followed with fundus photography and with optical coherence tomography (OCT). OCT is imaging of the various layers of the retina that can be used to measure the ganglion cell layer. OCT is often a better measure of retinal changes than visual acuity as measured with eye charts.

### WHEN VISION STARTS DETERIORATING DUE TO LHON

Vision loss from LHON usually appears between the ages of 15 and 35 through rapid deterioration of the central vision. It is unusual for onset of the disease to be seen after age 50, but there are cases of onset between the ages of 50 and 75. There are also cases of children developing the disease as early as age three, but this is also very rare.

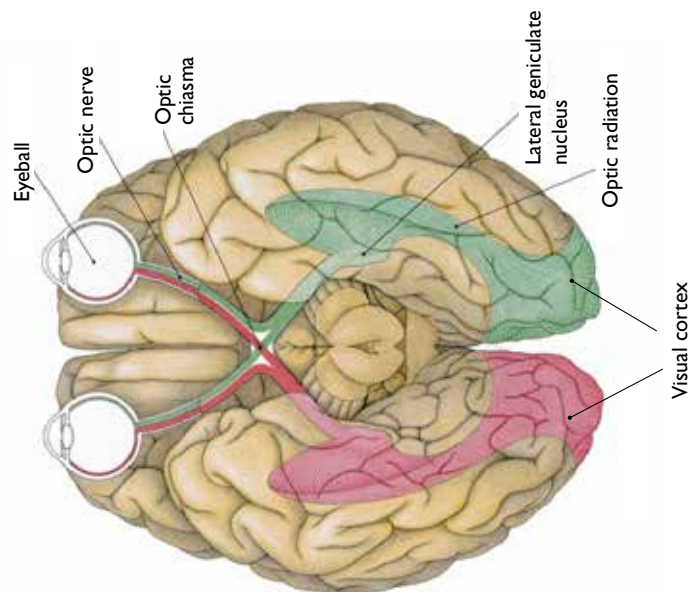
The disease usually progresses quickly, and vision in the central visual field is often lost almost completely within a few days or a few weeks - all without pain. With most, one eye is affected first and then the other eye is usually affected a few weeks to a few months later. About one-fourth experience vision loss in both eyes

right from the start, while in other cases it can take up to one year before the other eye is affected. There are also cases where the vision loss only affects one eye.

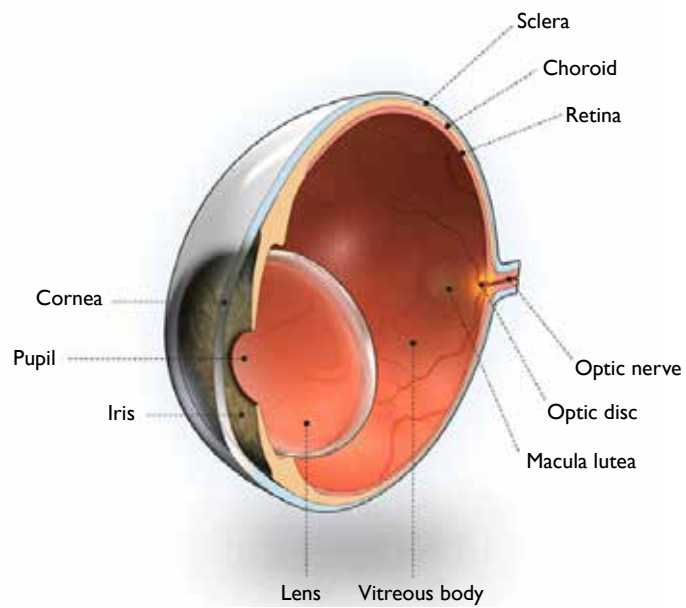
During the acute phase of the disease, vision can continue deteriorating for some time and then stabilize later.

There are also individuals who regain all or part of the vision. The chance of vision improving depends on factors such as which mutation is involved.

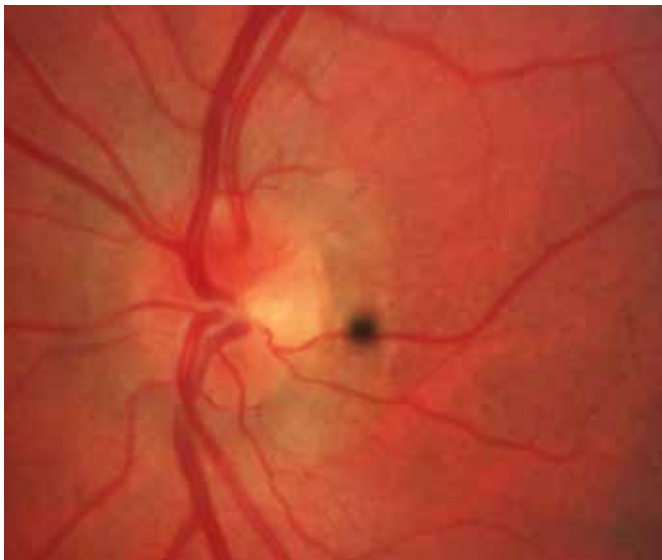
- Mutation 11778 (the most common type in Sweden) is associated with the smallest chance (less than 25 percent) of vision improving.
- Mutation 3460 is associated with a 25 percent chance of vision improving.



*The image's path via the eye, optic nerve and optic chiasma to the visual cortex*



*Parts of the eye*



*Fundus in acute phase*



*Ten months later - chronic phase*

- Mutation 14484 is associated with a 50 percent chance of regaining vision.

It is usually the central visual field that is affected. Most retain a large proportion of their vision in the peripheral visual field. Although visual acuity is significantly worse in the peripheral visual field, there are methods available to learn how to use it optimally through eccentric viewing, which is described in a separate chapter.

## DIAGNOSTICS

The characteristic symptom of LHON is rapid, pain-free loss of vision that cannot be corrected with glasses. It usually occurs first in one eye and then in the other within the space of a few weeks or months. Once the progression is known, LHON may be suspected. This is especially true if it occurs in a young person (under age 30 and male) and if siblings or other relatives have a visual impairment.

But, since as many as 40 percent of the cases have no known family history of LHON, the disease may be suspected even if the latter criterion is not met.

## EXAMINATIONS PERFORMED FOR DIAGNOSIS:

- Assessment of best corrected visual acuity (possible refractive error and need of glasses for distance vision).
- Visual field test - maps out the visual field and indicates and can estimate any damage to the optic nerve or optic tract.
- Eye examinations with ophthalmoscopy and fundus photography.

- Optical Coherence Tomography (OCT) - a relatively new technique for examining the eye with high magnification and precision, which also enables direct measurement of the eye's tissues. OCT is similar to ultrasound, but uses reflected light instead of sound. The properties of the light enable the scan to provide very good image resolution with great detail.
- Contrast photography (fluorescein angiography) - maps out blood circulation and blood vessels in the retina and the choroid.
- Electroretinography (ERG) - measures the function of the photoreceptors and is performed if a hereditary retinal disease is suspected.
- Visually Evoked Potential (VEP) - measures the electrical responses induced in the cerebral cortex during visual stimulation or stimulation of the neural pathways of the visual system.
- DNA analysis - Diagnosis is confirmed through analysis of mtDNA in the blood. The analysis focuses on the three most common LHON mutations. If the test is negative, further genetic or other testing could be used.
- Magnetic resonance imaging (MRI) is often performed on individuals who have a vision loss and swelling of the optic disc. The scan is primarily performed to rule out brain tumour or MS as a cause of the symptoms, and with LHON it can show swelling and increased signalling in the optic nerve behind the eye.

A spinal tap (lumbar puncture) is often performed to rule out other causes of vision damage, such as multiple sclerosis (MS). If polyneuropathy is suspected, the function of the peripheral nerves is investigated by means of electroneurography (ENeG).

### **PRENATAL DIAGNOSIS**

Prenatal diagnosis is possible if the mother's mutation is known, but all children of a mother who is a carrier will also be a carrier. If the mutation is found in the foetus, then it is only possible to perform an approximate risk assessment for potential visual impairment later in life.

### **RISK ASSESSMENTS IN CARRIERS**

It is possible to perform carrier testing on asymptomatic individuals with a sibling, mother or other relative on the mother's side who has LHON with a known mutation. If a mutation is found, then it is only possible to perform an approximate risk assessment for visual impairment later in life. With all testing, genetic guidance should be given both before sample collection and when the results are given.

### **OTHER POSSIBLE SYMPTOMS - LHON PLUS**

While loss of vision is the dominant symptom of LHON, there are rare cases when other parts of the nervous system are also affected. Examples of such are rapid, involuntary eye movements (nystagmus), coordination disorders (ataxia), involuntary muscle contractions (dystonia), shaking (tremors) and/or epi-

lepsy. Muscle weakness and sensory disturbances due to impaired function in peripheral nerves (polyneuropathy) may also occur. There is also research indicating that some heart defects may be linked to LHON Plus.

### **LHON AND MS?**

Older articles indicate that women with LHON are at increased risk of developing MS or a clinical picture similar to MS. In cases of MS, an MRI shows changes in the white substance of the brain or spinal cord and signs of inflammation in the spinal fluid (cerebrospinal fluid) can be seen. However, it is currently unclear whether there is any association with LHON or it relates to individuals suffering from two different diseases.

### **TREATMENT/SUPPORT**

There is currently no cure for LHON, but a great deal of research is being conducted around the world. Efforts therefore mainly consist of compensating for the visual impairment. More information about rehabilitation and support can be found in other chapters of this book.

A number of vitamins and other factors (B2, B3, B12, C, E, folic acid, alpha-lipoic acid, carnitine, creatine, arginine) have been tested without any effect being shown.

**Coenzyme Q10** has long been used to treat various diseases involving impaired mitochondrial function. One reason for studying it is that Q10 is known to be an important part of the mitochondria's energy production. Q10 in its natural state, which can be bought in health food stores and over the counter at

some pharmacies, does not seem to have any major effect.

**Idebenone** is an artificial form of Q10, where the molecules are smaller than in the natural state and are therefore believed to be more easily absorbed by the cells and mitochondria. Idebenone was originally developed by a Japanese company to treat Alzheimer's disease. However, it could not be shown to have any clearly positive effect on dementia diseases. However, positive effects were seen in the treatment of Friedreich's Ataxia (FA), which is a rare hereditary neurological disease. According to the National Board of Health and Welfare's information about FA, the medicine protects certain proteins (enzymes) in the mitochondria from being harmed by excess iron in the body, thus slightly improving mitochondrial function. Idebenone does not cure FA, but has been used for several years and is given to most individuals with FA in Sweden. Mnesis, which contains 45 mg idebenone per tablet as active ingredient, is usually prescribed for FA.

**Research studies of idebenone with LHON** have indicated a positive effect on vision in some individuals, particularly if treatment is initiated at an early stage. It has also been noted that the chance of improvement increases if the patient's eyes have different degrees of vision loss. The research studies were conducted as randomized, evidence-based studies and the conclusion basically indicates that 900 mg idebenone per day can have some positive effect on the eyesight of some people, but not everyone. The reason why vision

does not improve for everyone is believed to depend to some extent on how long the person has had the visual impairment. The effect is felt to be the greatest during the first year, especially if the medicine is initiated before the second eye is affected.

According to the research studies, it may take some time before any positive effect can be seen from idebenone. The recommendation is therefore to use the medicine for at least six months - preferably one year - to determine whether it is effective or not in a specific case.

Check of visual acuity, visual field testing and OCT before and after treatment can provide important information in determining whether the medicine is working or not. In addition to positive effects on vision, a number of individuals using idebenone have described a positive impact on their energy level and general state of health. However, this is not medically or scientifically proven and could theoretically relate to a placebo effect. Idebenone is a powerful antioxidant. Antioxidants have several important functions, including neutralizing free radicals that could otherwise have a damaging effect on the body and contribute to the development of cancer, cardiovascular disease and more. In recent years, excessive antioxidants have been shown to have a harmful effect and, for example, have speed up the process for people with cancer. At present, it is not known what effects prolonged treatment with idebenone at a dose as high as 900 mg per day could produce. The advice is therefore to reduce the dose after a period of time.

Raxone, a medicine containing 150 mg idebenone per tablet, was approved by the European Medicines Agency (EMA) for use as an anti-LHON medicine within the EU in 2015. Despite this approval, it had to undergo social economic investigation by the Dental and Pharmaceutical Benefits Agency (TLV) before it could be prescribed as a subsidized medicine in Sweden. TLV decided in favour of this in October 2016.

**EPI-743** is another form of artificial “quinone” that has even smaller molecules than idebenone and is believed may have a positive effect on LHON. Research is very limited thus far, but the results have been positive for several participants of the studies. More extensive studies with EPI-743 have been conducted for the very rare and serious mitochondrial disease Leigh’s Syndrome, which usually affects small children and results in, among other things, changes in the brain. Based on research results, EPI-743 has been approved as a so-called orphan drug for Leigh’s Syndrome in the United States.

#### **ACTIVE RESEARCH**

Research on mitochondrial diseases is very active around the world. In Sweden, it is conducted by expert teams at Sahlgrenska Academy, University of Gothenburg, and Karolinska Institutet Stockholm (Huddinge and Solna), as well as other research institutes such as the Royal Institute of Technology and Uppsala University’s Department of Immunology, Genetics and Pathology, Biomedical Centre, and Department of Neuroscience.

Mitochondrial diseases constitute a large area where much is not yet known. New diseases are being discovered, along with new mutations in the mitochondrial DNA that cause them. Various treatment trials are also in progress. The fact that LHON stems from a point mutation in a specific part of the mitochondria that most often affects only vision has caused many researchers to take an interest in the disease even though it is so uncommon. If – or more accurately when – a cure for LHON is found, this could bring researchers closer to a cure for other, more common diseases that may be difficult to research.

For LHON, treatment studies are currently underway with idebenone and other forms of artificial quinones (EPI-743) as well as with various vitamins. Although none of these medicines has been shown to cure the disease, they could possibly increase the chance of keeping the ganglion cells alive and/or hypothetically help them be repaired if/when a cure is found in the future.

Studies are also taking place with gene therapy in the hope of correcting the lack of enzyme complex I activity that the mutation leads to. Gene therapy studies are being conducted around the world, including the United States and France.

The American database [www.ClinicalTrials.gov](http://www.ClinicalTrials.gov) contains information on ongoing, completed and planned clinical trials in USA and other parts of the world, including Europe. The information is updated continuously and both ongoing and planned trials can



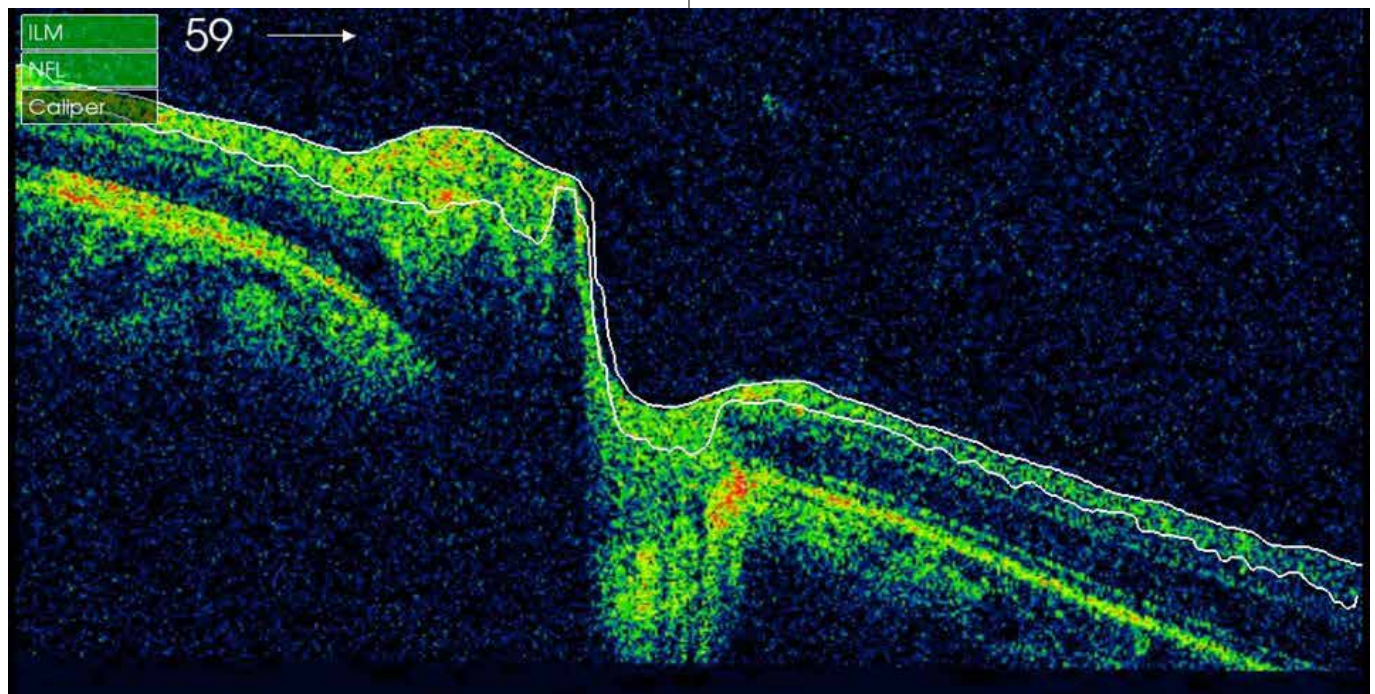
be found using the search terms “LHON” and/or “leber hereditary optic neuropathy”.

The European database Orphanet collects information on research concerning rare diseases, [www.orpha.net](http://www.orpha.net). Use the same search terms there for information on research related to LHON.

### **IN VITRO FERTILIZATION (IVF) OF EGGS WITH REPLACED MITOCHONDRIA**

The possibility of using IVF after first moving the mother’s cell nucleus into a donor egg where the donor’s cell

nucleus has been removed has been studied in animal models. Use of this method prevents the mutated mtDNA from being transferred to the fertilized egg. However, the method is not yet available for humans in Sweden. Both medical research and legal and ethical aspects must be sorted out before this will be possible. Authorities in England, however, have decided to approve this type of treatment in humans. It will most likely (at least initially) be used primarily for other mitochondrial diseases, like those that can lead to a child becoming severely disabled or unable to survive childhood.



*With an OCT scan, you can see how the nerve fibre layer of the retina has changed. It is a more precise measure of the condition compared to the standard visual acuity measurement using an eye chart.*





*Living with a visual impairment and dealing with all of life's challenges - a conversation between Agneta Löwenring Beck, visually impaired at age 19 (now age 65), social worker, licensed psychotherapist and mentor and Krister Inde, visually impaired at age 19 (now age 69), behavioural scientist, author, educational specialist for the visually impaired, and honorary doctor.*

# A BATTLE TO BE WON EVERY DAY

*With a combined 80 years of experience living with LHON, Agneta and Krister talk about how you learn to master life with a visual impairment. It is a challenge made easier by accepting you have poor vision. It's your eyes that are bad. Not you!*

## CHANGES ARE A CONSTANT CHALLENGE

"I travel a lot and lecture in different places," says Krister. "Yesterday was Copenhagen, tomorrow is Södertälje and today I'm sitting in Stockholm. With travel, you must constantly know where you are, use your aids, and ask people so you can find your way. You have to believe in yourself to figure out where you're going without panicking or feeling defeated if you don't know where you are. I used to be able to find my way around Malmö really well, but there's been a lot of construction. I find change frustrating. You become very conservative when you don't see well..."

"Exactly!" says Agneta, jumping in.

"... you don't want much of anything to change. Do you have an example of some place where you could find your way around and then they changed something?" asks Krister.

"It's mostly in shops. Suddenly the milk has been moved somewhere else. But the whole world is constantly changing, so it's important to keep up."

## MORE THAN THE VISUAL IMPAIRMENT

Having poor vision - especially when the impairment

first occurs - makes you become very aware of yourself and worried about being seen as different. It feels frustrating, even scary. It is important not to let your visual impairment define who you are. Krister explains:

"In the past, I didn't want to let on that I have poor vision. I thought people would have a poor opinion of me if they knew. That I would be thought of as less of a person. It took several years to overcome that feeling. What about you?"

"It's important to make a conscious decision that you are not less of a person and not to let others see you that way," says Agneta. "When my opinion of myself is based on other people, I'm actually seeing what I think I should see. If people want to think of me as less of a person, so be it. I know that's not the case."

"When I lecture, I often say that I'm a heterosexual white man with poor vision who must also come out sometimes. It's almost easier for homosexuals to come out than it is for the visually impaired."

"That's a big part of life - daring to be yourself. I am a whole made of many parts. Sure, I have a visual impairment - but there's more to me than just that," Agneta points out.

"So, if I ask whether you are visually impaired, your answer is...?"

"Yes, I am. Also. But, I am also an employee, a home owner, a mother and a wife."

"That's really interesting," says Krister. "Back through history, many visually impaired people chose a partner who was also visually impaired. They viewed it as a type of bond between each other. I feel a bond with you because you have poor vision, but that may be the only thing we have in common - even though it is an important bond. You sometimes need to meet others in the same situation."

"Yes, I have much the same view of meeting others with poor vision. When I was young, I was at a rehabilitation centre where most of the other people were completely blind. So, we were in somewhat different situations. It was hard for me to talk about what I had lost when the others had lost so much more. So, I think it is frustrating when people confuse those who have lost all their vision with those who have some sight left."

"I was once part of a TV show called *Jag tycker blinda människor är äckliga* [I think blind people are disgusting]. I found them threatening at the time. Was I also going to go blind? At the time, I didn't know why I was losing my vision."

"I've actually only known my diagnosis for four years," says Agneta. "I got sick when I was a 19-year-old au pair in Germany and was studying at the university in Freiburg. So, my first experience when I developed my visual impairment was at a German

hospital, where I had to stay for a while."

"I found out mine after 30 years, in 1996," Krister responds. "That knowledge is very important. I usually say there are two types of ophthalmologists - bad and good. I met a bad one because he said 'It could be a brain tumour. We'll have to wait and see.' To a young boy! Nowadays, they just take a blood sample to make the diagnosis. And even though there's no cure today, you know that research is being conducted to find a cure. How would you react if you got your sight back today?" Krister asks Agneta.

"That's a tough question. I probably wouldn't dare to drive a car... Well, maybe eventually. I drove for one year. And it gave me such a sense of freedom. I've missed that."

#### OPEN WITH YOUR AIDS?

There are some aids you can use out in public, like very strong glasses. There is a company in Gothenburg that sells these types of glass for upwards of SEK 50 million a year. Yet, you very rarely see people using them and reading with the text right up at their nose. "But, that's



what we do," state Krister and Agneta.

"I've decided that I can do anything, any time, and whatever way it takes," says Krister. "Including sitting with my phone or my newspaper up to my nose. Otherwise I wouldn't be able to read."

"Then you've taken control, which is exactly what you should do."

"But, it's not easy."

"No. It's a long struggle."

"Do you put on your strong glasses anywhere?"

"Not really," says Agneta. "But, most places, like on the bus."

"What's stopping you?"

"It's mostly at work. I don't always put on the glasses in front of my patients because I don't want to draw attention away from them. But, when I'm with my colleagues - absolutely. It's a way of adjusting to the situation. Sometimes it brings about too many questions and puts too much focus on me."

"And I try to learn a lot by heart," says Krister. "If I'm giving a lecture I've done before, I generally know what's coming and don't have to have as detailed notes. This lets me write a little larger and use weaker glasses. Then I don't have to have things as close to my eyes. I don't want to disturb my story by behaving differently."

Agneta agrees:

"Exactly. It disturbs those listening. When I think about the fact that you can feel good even though your vision is poor, a lot is about being part of a context. I think that's important."

## FEELING A CONTEXT

Job, family, friends, hobbies. Everything that helps to create a sense of context also helps to create a good, meaningful life. An example Agneta gives is that the thought of retirement and not working any more makes her a little nervous.

"But, you can find new contexts," she says. "It doesn't have to be work. It could be the gym. But, you have to be a part of things somewhere."

"Yes, that's what happens when you don't have a job or you're not happy with what you have," says Krister. "You feel like an outsider, and you might blame this on your poor eyesight."

"It's a pitfall since you've lost the sense that you're worth something despite your vision problem," continues Agneta. "You let the disease define you."

Krister and Agneta agree that with all of today's aids, TV systems, strong optics, and so on, people who develop a visual impairment have more freedom to choose their path in life. But, that also means you have to take some responsibility for your own life. It takes a strong person to be able to live without defining yourself through your disability.

"That strength you have as a person, I think I got mine from my family and from meeting role models who have poor vision and do well in life," says Krister. "When I meet someone like you, you become a role model."

"It's the same for me. My family, my husband and my children. And a relative with the same diagnosis. He's no longer alive, but he pushed me to continue my

studies. And I did. He was a lawyer and managed well thanks to his determination, curiosity and energy. You need to have these kind of characteristics.”

“When I developed my visual impairment, I was doing my mandatory military service,” explains Krister. “I was sent home and worked as a teacher for as long as I could see out of one eye. Then I lost vision there as well. I was supposed to go to the School of Business, Economics and Law in Gothenburg. I had my life planned out. I was going to be some kind of economist. But, I ended up in Växjö at a school with rehabilitation for the visually impaired. Then my twin brother called and told me to drop out there and start at university. You need someone like that in your life. Someone who tells you to not let your impairment keep you from living your life.”

“It was the same for me. My visually impaired relative thought I should continue my studies. And I realized it was important to me to have a good job and a good education.”

“So, he became your mentor?”

“Yes, and not just in practical matters. We talked a lot with each other. I also knew that he had overcome the same types of challenges. But, I did an internship once as a nurse’s aid and things didn’t turn out so well. Like once at meal time. I thought we got ice cream, but it turned out to be mashed potatoes. It didn’t feel good. I then realized

that with a better education I could get a job where I would be respected. It has a lot to do with self-respect. You have to have a job you’re good at. For example, cleaning does not work well for a visually impaired person. I might have done that if I hadn’t had such poor vision.”

“Did you get really tired of having poor vision when you first developed the impairment?”

“Yes, of course. It’s a major upheaval. When you

first develop the impairment, you must allow yourself to go through different phases - denial, losing your desire to do things, and so on - until you finally reach a point where you can deal with the situation, even though you may

not have accepted it completely. There is movement in that, and you have to allow yourself to stop for a moment as you reach the different steps.”

#### **DIFFERENT DAYS, DIFFERENT ENERGY**

“When it comes to energy...” Krister contemplates. “I’ve noticed that on days where I don’t have as much energy, or feel sick, or have a headache, that I find it more difficult to cope and function as usual. I therefore have to think about staying healthy, including exercising.”

“Yes, really,” agrees Agneta. “Exercise, eat right, get enough sleep. It’s a lot like with burnout. You have to set limits for yourself, say no to the right things, and take care of yourself.”

*“It’s your eyes  
that are bad.  
Not you!”*



"Do you feel like your vision is different on different days?" asks Krister.

"No, I don't think so. I may feel really tired in my whole body... But, no my vision doesn't feel different."

"I feel like I do. I think it's my energy level that changes, not my actual vision. The energy to compensate and use the vision you have left. I've trained a lot to direct my gaze in the right direction and position the image correctly in my retina and then use the macula - the best spot in the retina. But, if I do that for a long time and sit and hold the image really close, my neck starts to hurt."

"Yes, in that way my vision seems worse on certain days, when I'm tired. Or in certain situations. Like when I've been sitting at the computer too long," states Agneta.

"What days do you find it a bigger struggle to deal



with the impairment? And what days is it OK?"

"Generally, it can be a struggle on sunny days, even though I like to be in the sun. And when it's pouring down rain. How about you?"

Krister thinks about it and then says: "I'd have to say it's days when I have to go somewhere I've never been before."

### THE ART OF DOING IT YOURSELF

The conversation turns to the limitations encountered by a person with a visual impairment, no matter how determined they are to live a normal life. Krister talks about how difficult it is to travel by public transport and to find your way if you need to switch trains or buses.

"It's nice to be able to do it. It's a matter of independence. But, imagine just being able to just get into a car and drive. I remember when I was in Uppsala and had been dealing with the poor vision for a year. I suddenly grabbed the car keys, hopped into the car and began driving because I had repressed the fact that I had trouble seeing. Suddenly I was out in traffic driving a car. And I realized - shit, I can't see!"

"It sounds like something you dreamed," comments Agneta.

"But, it really happened! I parked the car on the street, locked it and walked home, where I told my girlfriend that the car was parked on St. Persgatan. I forgot I can't see well. I'm not sure if I was in denial or if I was out of habit."

"Maybe a bit of both. At the same time, there are

many things you think won't work but actually do. I grew up in Norrland, so we went skiing a lot. I thought that downhill skiing was out of the question due to my visual impairment. But, then I realized that everyone has to go down, so it would have to work. And it did for a number of years. But, now it's more difficult with all the snowboarders who travel across the slope, making it more difficult to navigate. Now, I'm very pleased with the fact that I've been downhill skiing a little here and there around the world." Agneta beams with pride.

Krister responds with a strange situation. "I was once skiing with a bunch of friends in France. It was a large system and I suddenly lost track of the others. I didn't know where I was. So, I decided to stay put until someone I know found me. And they did after a few hours. I was stranded. I didn't want to say I was visually impaired and ask for help because I was worried that others would think I couldn't take care of myself and shouldn't even be on the slope. Back then, at the start of things, I wouldn't even take the bus between Linköping and Motala. I had a lot of anxiety that I would get lost. Today, I fly to the Middle East as if it were Södertälje. And you get very good help on the flight! The same applies with all of the yellow-vested passenger monitors of the SJ train system. But, you have to be willing to ask." He says the following almost to himself.

"Yes, you have to."

"You don't have to tell them your life history. It's enough to say 'Excuse me. I don't see very well. What does that sign say?'"

"And then you get help 99 percent of the time."

"I've come up with a little way of doing things.

When I climb into the car, I say to someone standing there: 'Excuse me. Do you see well?' 'Yes,' they respond. 'Unfortunately, I don't,' I say. 'Do you know where seat 67 is? That's my seat.' It works every time. It's a less evasive way to express yourself." That is Krister's opinion at any rate.

"It's important to find ways to get around so you can avoid the victim role." Agneta sounds like a true therapist now.

### **SOMETIMES IT'S A STRUGGLE**

Agneta and Krister agree that it is OK to feel sorry for yourself sometimes. You do not have to be strong and super-capable all the time. Because it is undeniable that it is often a struggle not to be able to see properly. Missing out on things people around you are talking about. Taking a walk and not knowing what things look like. Not even getting a glimpse of a bird the people you are with are talking about. You could pull out the binoculars, but that takes a little time. The bird may have flown away by then.

"You have to try to hang out with people who are good at visualizing," emphasizes Krister. "Who are good at describing what they see. I myself am very irritating to be out walking with because I want to know about everything we pass by. What does it say there? What was that? And so on. I don't care if people get irritated. I won't stop being curious."

Grieving all of these small day-to-day losses is both natural and inevitable according to Agneta and Krister.

"You have to allow yourself to be sad sometimes. You can't just swallow everything and have it sit like a lump in your stomach," as Agneta puts it.

"On the other hand, it is also possible to take advantage of the impairment," says Krister and then tells us about a fellow student who told him that as a child he tried to convince his mum to buy him toys by saying "But, mum, I only have one eye."

A nail had gone through his other eye. But, there are different ways to request benefits.

"It's easy to ask for extra help or extra compassion because you have a vision problem."

"And it can be a shock when you realize that you don't always get it."

### **INTERACTING WITH OTHERS**

An understanding and benevolent world can be of great help when you struggle with all of the big and small challenges of day-to-day life as someone with a visual impairment. It is wonderful to meet people with natural empathy. Krister tells how when his children were young they understood that "daddy has broken eyes" and brought their things really close when they wanted to show them and talk about them. And now the grandchildren do the same thing.

"I also had a neighbour who worked at a tyre company.

One time, we took a trip to the west coast together. And every time we came to a new place, he would say something like 'Now we're in Mellerud'. 'Why are you saying that?' I wondered. 'Because you can't read the signs' was the response. It's great when you meet people who are so immediately empathetic. It makes you happy."

"It's great," agrees Agneta. "Not everyone has that gift, so it's great when you meet someone who does."

"In contrast, I once had a good friend who always

stood still when he met up with me in town. He then waited to see whether I saw him or not. And that it was 100% certain that I didn't know who it was. I finally ended up telling him that I didn't like his games and asked why he couldn't just say something like 'Hi Krister,

it's Göran'."

Agneta smiles and agrees. She has been through the exact same thing a number of times.

"When things like that happen, it makes you sad for a little bit. You make a note of it, but you have to let it go to some extent," she says. "It can't make you avoid going out as a means of avoiding meeting people who don't greet you or whatever the issue is."

Agneta and Krister agree that the benefits of daring to venture out in the world and take risks outweigh the disadvantages. Each and every person has the right to be a little different. Everyone has their own way of interacting with others in society.

*Focus on  
what you're  
good at.*





# FÉLICIA AND PATRICK - A REAL CINDERELLA STORY

*At a time when Félicia Adlersfeld had first developed LHON and was very sad and angry, she had an errand to run at a Clas Ohlson shop. She stepped into the store and curtly asked for extra assistance. The person who helped her was Patrick Castillo. He could not forget that demanding girl. Seven years later, he read about Félicia in the newspaper. Today, they are married and have a little daughter.*

The family lives in Hallonbergen, a northern suburb of Stockholm. Their apartment is on the ground floor and they have a large, lush garden. A quiet idyll in the big city.

"It's a really great place to live," says Félicia as she walks around barefoot on the lawn, where Patrick is playing with their daughter Patricia, who is 11 months old, is found of laughing, and is learning to walk.

"It's great to be able to just let Patricia go out into the garden, where she can play and have fun with our dog Simba. We're close to nature, with a nice lake. Yet the town centre with shops and the metro is just a few hundred metres away.

Félicia and Patrick usually compete about who makes the best coffee. Today, it is Patrick's turn. He fiddles around with the coffee machine for awhile and then offers a drink that would make any barista jealous. Both he and Félicia beam with pride when I and photographer Maria give them praise. They are proud - proud of each other.

"But, we wouldn't be sitting here, would never have met, if I hadn't developed LHON," says Félicia. Every cloud has a silver lining.

Even though it has been ten years since their first encounter, they have really only known each other just over three years.

At their first meeting, Félicia was 21 years old and had just developed her visual impairment.

"I was still in some sort of shock," she says. "Angry and bitter. I stormed into that Clas Ohlson in Solna and was aggressive and demanding. I didn't give much thought to the guy who was helping me. More than he seemed nice and was patient with me."

Once she got what she needed, Félicia stormed back out.

Then seven years passed.

"Then I happened to see that girl in the newspaper," explains Patrick. "I remembered her and thought about her often. She made a deep impression and I felt like I

understood her and the difficulties she faced. I myself have ADHD and know how it feels to have difficulties you need to overcome.”

The newspaper Patrick was reading in the autumn of 2013 had an article about how the visually impaired Félicia hiked from Stockholm to Västervik together with her faithful pit bull Simba.

“Now, I finally knew her name. And I found out she had a blog, which I naturally began reading right away. I then got a more balanced picture of who she was as a person. Félicia was more than just a tough girl who could stand up for herself. She also wrote about her feelings, her moments of weakness and doubt. And her sense of humour was a lot like mine!”

And then one day in December 2013, Félicia was suddenly standing there in the shop again. Not at all as angry or demanding. Patrick made sure he was the one who got to help her. Among other things, she wanted to join the customer club.

“It felt good to be recognized after so many years,” says Félicia. “At this point, I was used to my visual impairment, and felt confident in being seen.”

A few weeks later, in February, it was time again. Félicia visited the shop together with her sister. That time, Patrick was manning one of the cash registers. Félicia’s sister offered to pay for her items so the purchase could be registered on her club card.

“But, Félicia has her own club card,” said Patrick.

The sister was amazed, Félicia was embarrassed, and the two left the shop laughing hysterically.



"A few days later, I contacted Félicia via Messenger," explains Patrick. "I wanted to apologize for making her feel uncomfortable."

And with that, they began talking via their mobile phones. About everything under the sun. About life. About what they did and where they lived, what they liked and disliked. After awhile, Patrick offered to go out with Simba. Even though he did not usually go for walks otherwise. Félicia thought that he and the dog should get to know each other first, so they started taking the dog for walks together. Long walks. Often lasting several hours.

Things went quickly after that. They soon both realized that they considered the other their soul mate. They had fun together. They felt a sense of harmony and security with each other. After two months, they decided to become a couple. After another two, Patrick proposed.

That was 7 June 2014.

"It was so sweet," explains Félicia. We were down at Lötsjön Lake in Sundbyberg, where Patrick lived. In a meadow, there was a little travelling funfair that was packing up. I talked to the guy in charge of the ferris wheel, and he let us take a ride. Without my noticing, Patrick made arrangements for us to stop when we reached the very top. And there, he pulled out a ring and proposed. It was both fun and romantic."

Six months later, they married on a rocky beach in Hawaii, where Félicia's father lives. Nine months after that, Patricia arrived, a true child of love with a name

that is a combination of her parents'.

And now? Everyday life, a sense of belonging. Mutual support. Félicia is on parental leave from her job as a personal assistant, while Patrick is studying to become a sound engineer.

"We've only known each other two and a half years, and we've built up all of this," says Félicia. "It's been a fantastic journey. When I first developed my visual impairment, I thought there was no way I could live a normal life. A life like this," she says as she stretches her hand out across the living room, where Patricia is playing on the floor, and towards the garden.

Being visually impaired does not mean that a relationship has to be unequal. That is something she and Patrick agree on.

"We simply help each other out with everything," says Patrick. "I really want to help Félicia with everything she has trouble managing. But, it turns out that she can manage just about anything."

At the core, they feel their relationship works because they complement each other. At the same time, they have a deep bond through their similarities.

"Yes, for example, we have the same type of humour," says Patrick. "We're not just a couple. We're friends as well."

He feels like he has gotten more help from Félicia than he has been able to give her.

"She helps me build structure into my life. With my ADHD, I used to have trouble finishing what I started. But Félicia has helped me, for example she coached



me into getting my driving licence.”

Since Patrick spends long days at his school, Félicia manages most of the practical aspects at home. Like shopping and cleaning. Things that are more difficult with a visual impairment.

“You develop little tricks,” she says. “I often ask the staff for help. I then listen for heavily loaded trolleys because then it’s more likely to be someone putting out products. It’s easier in the winter, since people without coats are more likely to be employees. And, naturally, I do a lot of shopping online. Cleaning is a little so-so. I usually walk around the apartment bare-foot, so I can feel if there is something on the floor.”

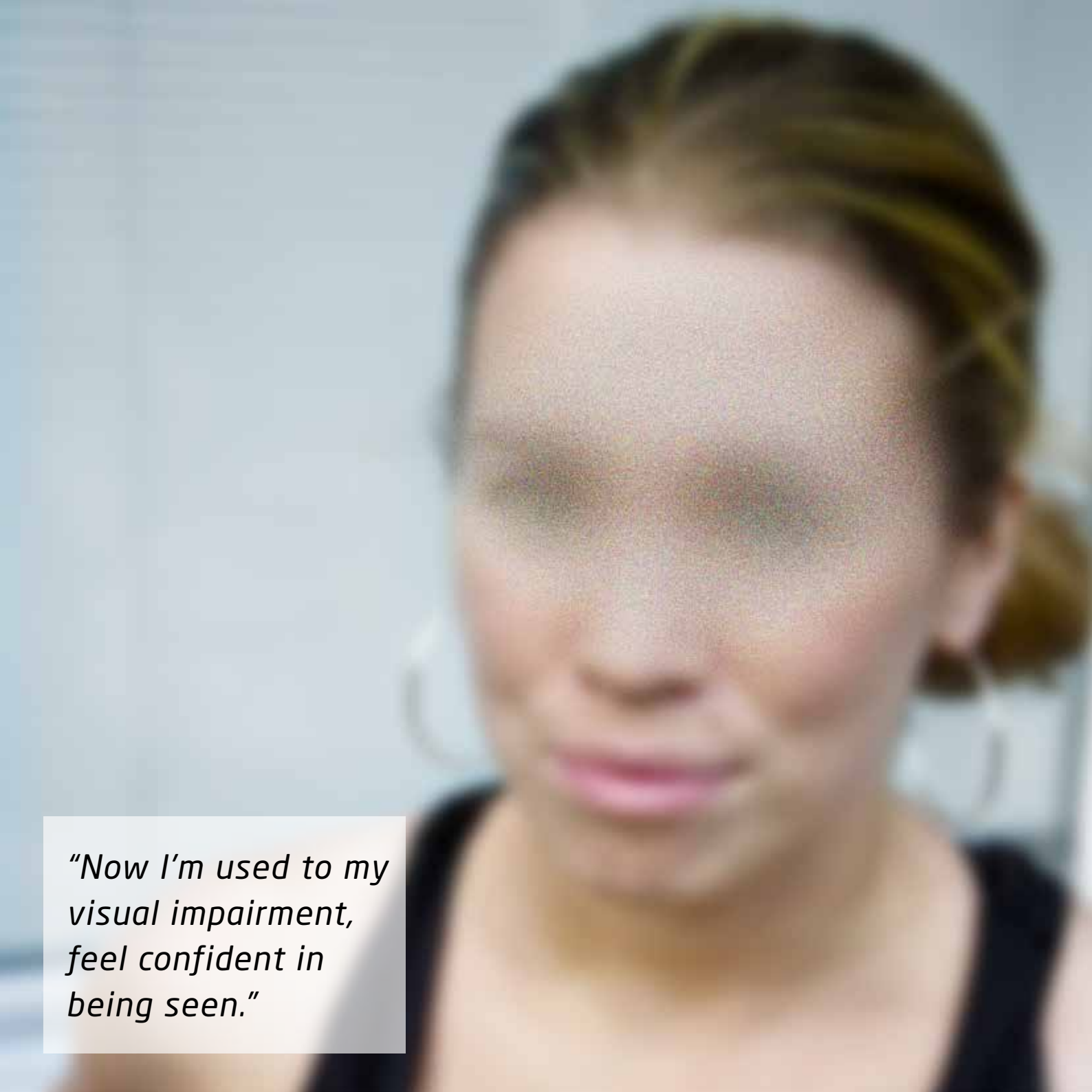
Are there any day-to-day tasks that you need Patrick’s help with?

“Yes, things like cutting Patricia’s nails. And, he’s good at letting me know where he is so I don’t have to look around.”

“And when we’re out somewhere eating at a buffet, I usually fill Félicia’s plate as well,” adds Patrick. “Different buffet dishes may look the same. And I know what Félicia likes.”

According to Félicia and Patrick, such details in everyday life are important building blocks for developing a stable and well-functioning relationship. If you meet the right person at the right time in life, an impairment is pretty much a non-issue. And when Félicia met Patrick, she had a few years’ experience in living with her visual impairment and had come to the realization that life could actually work well.





*"Now I'm used to my  
visual impairment,  
feel confident in  
being seen."*



## MEET FOUR PERSONALITIES WITH LHON



## BJÖRN CARLSSON

### “YOU LEARN TO BE AN OPTIMIST”

*On New Year's Eve 1987, 28 years ago, Björn Carlsson suffered a blow to the eye. After a few days, his vision began to deteriorate. The macula had been damaged and was restored using laser technology. But, his vision loss continued and also affected the other eye. The doctors thought it was a tumour, possibly MS. Nine years later, Björn was diagnosed with LHON.*

This spring, Björn Carlsson turned 50. Together with his brother, he runs a company that sells machinery, primarily to the construction industry - lifting platforms, mini-dumpers, loaders and the like. Things have gone well, even through all the economic crises. In April, Carlsson & Co moved to new, larger premises in Falkenberg.

“One of the hardest things about developing such a severe visual impairment is that I couldn't drive any more,” says Björn. “I love cars and machines, and even had a truck driving licence. And I sometimes still help customers reverse with their trailer...”

When Björn developed his visual impairment, he was 22 years old, had studied automotive engineering, and worked at his father's company, which also sold machinery. Among other things, he drove a truck and delivered machines to customers.

“My dad was a real entrepreneur who started a new company every five years. I inherited that spirit

and that's what made me and my brother start our own company, in the midst of the 1993 recession,” he says.

We find ourselves at the company's stand at the Nordbygg trade fair in Älvsjö outside of Stockholm when Björn talks about his life with LHON. He is keen to show that his visual impairment does not stop him from talking with customers and demonstrating how the machines he sells work. Even though he cannot see the faces of the people he is talking to. According to Björn, most people he meets have no idea he has poor vision. You cannot tell by looking at him. He moves between the machines unhindered and is quick with a joke and a laugh.

“I only have 5-8 percent of my vision, no locomotor vision,” he explains. “And to top it off, I'm near-sighted. I've done quite a bit of training to be able to utilize the peripheral vision I have left. So, with good magnification tools, I can read and send emails and things



like that. And I take my electric bike to work. That alone gives me a great sense of freedom.”

### **TOUGH CONDITIONS**

The initial period of the visual impairment was tough, Björn recalls. Since he primarily worked as a driver, his work was greatly affected right away. He had to climb out of the cab and start working in the spare parts warehouse. The first year, as the disease progressed and his vision worsened, was a period of anxiety and concern. How bad would his vision get? Would he lose his sight completely?

When Björn came home from a holiday trip to Gran Canaria in October 1988, his sight was so poor that he ended up spending two weeks in hospital in Halmstad. He was examined thoroughly, including a CAT scan, but the doctors could not find any tumour, could not make a certain diagnosis, and ended up calling it an undefined inflammation of the optic nerve. It took nine years before Björn learned what kind of disease he has. It was all thanks to a professor in Finland who made the right diagnosis, which Björn had confirmed through a test in Lund that he had to request himself.

It was these years of uncertainty that were the toughest according to Björn.

“At that time, I had to come to grips with my new situation,” he explains.

Through Arbetsförmedlingen, he took a number of courses, including typing, and could get the needed aids that were available at the time, like a large read-

ing machine with camera and a huge black and white television. He worked for a while as a telemarketer for a company. He soon understood that they employed him to receive the 100 percent wage subsidy and thereby free labour. Björn quit and returned to his father’s company.

“It was a valuable experience,” he says. “Encountering a company that wanted to exploit the system.”

### **LIKES TO WORK**

“Although it takes longer and I often get very tired, I think it’s fun to work,” says Björn. “In preparation for the company’s move to new premises, I’ve worked up to twelve hours a day, seven days a week. I participate in all the work tasks, but selling and making deals are what I enjoy most. Although it can be a bit strange at times when I don’t recognize people I’ve met. Many have no idea about my visual impairment. It is, of course, a constant problem, but you are constantly learning new little tricks to overcome all big and small obstacles. It’s important to not give up and to look for possibilities. If nothing else, you learn to become an optimist.”

For many years, Björn found it tough, even embarrassing, to tell people about his poor vision.

“But today, I can be much more open, which makes my life much easier. Especially since I got in touch with LHON Eye Society and finally got to meet others in the same situation. Knowing I’m not alone and being able to share experiences – that was a huge boost for me.”

Björn's main advice for those who develop a visual impairment today is to make contact with others with the same problem.

"Find a network. Don't isolate yourself. It's really important," he says. "It gives you comfort and practical tips, and can also be good support for your family and other loved ones."

Björn also thinks you need to read up on things and not give up when it comes to getting aids.

"In the beginning, it was easy. Arbetsförmedlingen and Försäkringskassan asked what I needed and I got it. But nowadays... For example, I had to fight hard to get an adapted cash register system for the company, and once I got it there were parts missing. Now, I need a camera. But, it seems to be something within me. I am a salesman and have no trouble presenting my case. But, not everyone can do that."



MALIN WIXNER

## "NOTHING'S IMPOSSIBLE"

*Losing almost all your sight is a great loss according to Malin Wixner. But, she feels she has a good and normal life thanks to a great deal of support from her partner and her employer - and, naturally, her uncompromising willpower. But battles must be continuously won to live this normal life. "When you have an impairment, you are forced to fight harder," she says.*



One summer day in 2003, Malin Wixner, age 27, was putting on make-up and discovered that her vision was blurry when she closed one eye. At first she thought there must be something in her eye, or she was stressed. But it quickly got worse. She went to an optician, who sent her to a doctor straight away.

"The doctor who examined me was not sure what the problem was. 'It could be MS, a brain tumour, or Lyme disease,' he said. It was awful to hear. I went home full of anxiety," explains Malin.

The next day, she had to come back for an MRI. The doctor prescribed cortisone. It took a month before Malin was given a diagnosis. About six months later, the sight in the other eye began to deteriorate. It got worse quickly over the course of a few weeks.

"I understood that this would occur, but I wasn't ready for it at all," she explains. "I really hoped it would take a long time."

Today, Malin is 40 years old and lives in Falun. All she has is somewhat blurry peripheral vision, but she can move about quite freely in familiar environments. But on sunny winter days, for example, when

the snow covers all contrasts, it is a challenge everywhere.

### **STRONG SUPPORT**

Malin had been working as a teacher for about a year when she developed her visual impairment. At that time, she taught fourth grade.

She remembers it being a difficult time. She got a great deal of support from her employer. The head teacher did everything possible, including arranging for counselling. Malin went on sick leave from April to August. After a year of work training, she was invited back to the class she had previously worked with, where she knew the students. But, it soon became to difficult to work with a regular class. Malin was then given the opportunity to undergo professional development in Örebro from 2009 and 2013. After that, she began working as a special needs teacher for students requiring special support.

"I handle groups of no more than five students," explains Malin. "I often have lessons with one student at a time. It works really well and I'm very happy."

Malin also received invaluable support from her partner, who is now her husband.

"He really helped me keep my self-esteem and not become my diagnosis," she explains. "We also have a son we became parents to in Colombia."

### **IMPORTANT TO BE OPEN**

Despite her severe visual impairment, Malin feels she

lives a rather normal life. She makes great use of her aids – large screen, magnifying program, speech features on her computer and mobile phone. She also has CCTV, which she can use to scan documents and have them read aloud or have the text enlarged. Malin had a great deal of contact with Försäkringskassan when performing her work training. Today, she receives disability allowance. She receives her aids through Arbetsförmedlingen, which has a partnership with a supplier.

"Once you get used to everything taking a little more time, things work really well," summarizes Malin regarding her experience of the aids.

In addition to the shock and sorrow she felt when diagnosed, Malin thinks the most difficult experience was the uncertainty she felt after work training as to whether she would be able to find a job at all. "That would be the deciding factor in whether I had a role in society or not," she feels.

"But, everything worked out really well, so my advice to those who develop a disability is to apply the mindset that nothing is impossible. You just have to find new ways of doing things."

"I also think you need to be open about your impairment and be confident enough in yourself to talk about it and not try to hide it. Everything is also so much easier if you get in touch with others with poor vision as early as possible. They are proof that it is possible to live a good life and have a job. You need to be involved and not let yourself fall into us vs. them thinking. Do not use your visual impairment to define who you are."

## ANTON BLOMBERG

# "ATTITUDE IS EVERYTHING"

*Anton Blomberg is 24 years old and has been living with LHON for the past 14 years. He earned his degree in economics in 2015 and is doing his best to navigate the job market. At the time of writing, he is filling a temporary role at Karlstad-Hammarö Chief Guardian Administrative Services Department to help the municipality take care of the many unaccompanied refugee children.*

"Naturally, I hope it will turn into a permanent position," he says. "It's a fun job that requires great accuracy, and it suits me."

Anton has central vision loss in both eyes, but can see well enough with his right eye to be able to continue playing floorball. His golf handicap is 36, and he played quite a bit of football in his adolescent years. He can make out some letters on the ophthalmologist's eye chart and has trained his ability to find the best retina position.

The visual impairment seemed to come out of nowhere. When he was ten, he noticed that he had trouble seeing the numbers in his maths book. He then figured out that he had more trouble seeing out of his left eye. The deterioration came and went. When it began to make him clumsy, he visited an

ophthalmologist before heading off to an ice hockey camp in Gotland. A few worrying weeks followed – one of the doctors' first suspicions was that Anton could have a brain tumour.

After a few months of repeated examinations, Anton was diagnosed with LHON. "At the time, we had no idea that we had this disease in our family. But, a few years later my uncle also developed it."

### PERSONAL ASSISTANT IN SCHOOL

Anton feels like he got a lot of help in completing his studies. The school and the municipality arranged for a coordinator. Anton got help learning how to master the keyboard and practice using various computer-based aids. From fourth to ninth grade, he had an assistant almost every day in school. At high school,

he had an assistant by his side the whole time who helped him with tasks such as taking notes.

After graduation, Anton began studying economics at Karlstad University. At that point, he no longer needed assistance. "The only assistance I needed was a little extra time to take my exams. But, I had developed good study habits and was very ambitious," he says. "I worked hard with my studies, and handled the other aspects of my life well. I could go for walks and bike rides, and could play some football. Still - it was really difficult to study. It's then I noticed how big a role your attitude plays in things. You can achieve whatever you set your mind to."

Anton completed his degree in autumn 2015. It was then time to search for a job.

While he was a student, he had a part-time job at ICA and gained work experience. He was able to continue there a while. But, he now wanted to work as an economist.

In January 2016, he was hired by the municipality as a Chief Guardian Assistant. This involved reviewing the reports submitted by individuals serving as appointed guardians.

"It was a job with a lot of small details to keep track of. There were tons of documents to analyse. I got a number of aids, like a camera and magnifying program for the computer, from Arbetsförmedlingen."

Last winter, Anton took a course offered by Arbetsförmedlingen regarding regulations for the finance sector, including the new regulations related to mon-



ey laundering. He has now applied for a job here and there, and while waiting for a job offer has accepted a temporary position helping the municipality manage their social obligations to all refugees who arrived in a short period of time.

"Naturally, it takes me a little longer to handle all the documents," says Anton. "But I can handle it all. Well, except one thing. I have to ask someone else to put the paper documents into binders. With my eyesight, I could easily get things disorganized."

#### **LESS STRESS WITH WAGE SUBSIDY**

Anton never intended to request a wage subsidy. However, in consultation with his employer, he now receives a low subsidy, mostly to keep him from feeling stressed. He also receives disability allowance from Försäkringskassan to cover extra expenses.

"I found out about that possibility by chance. It





wasn't anything Försäkringskassan told me about. Sometimes, an administrator from Arbetsförmedlingen comes in to see how things are going. But, I handle all my day-to-day issues myself."

Anton does not mention his visual impairment on his CV. "Not since I was on a job interview at a bank. Everything felt good, so I told them about it towards the end of the interview. Right then, they seemed to lose interest. The interviewer promised to be in touch, but she never did. When I contacted her, she told me the position was filled. When I asked what she thought my shortcomings were, she replied 'stiff competition'. But, though a friend who works at that particular bank, I found out I was more over-qualified than anything else."

Handling the outside world's negative expectations of someone with a visual impairment is perhaps the biggest difficulty Anton experiences.

"It's also the case that most things take a bit more time," he says. "You have to have patience and work harder than others. And that has an upside as well, since it helps you grow and learn to always be on the ball. You become a good listener, and can get a sense of other people's energy in a way most never learn."

Anton's advice to others who end up in the situation is: anything is possible. "And it's very hard to prove otherwise," he states. "In daily life, you should also establish routines as early as possible. That makes it easier to get things done. But - that's something that really applies to people in general."

## SUSANNE BOSTRÖM

### “YOU HAVE TO SHOW THEM WHAT YOU’VE GOT”

*When Susanne Boström visited the optician at age 18 for an eye exam and to get a certificate of eligibility for a driving licence, she discovered that she could barely see anything at all out of her right eye. A few hours later, she had been examined by five different doctors at the eye clinic. “It could be a brain tumour,” they said.*



Susanne had just finished the two-year healthcare programme at high school and got a job in elderly home help services in Umeå. She needed the driving licence to be able to get around to all the patients she needed to visit. Now, everything was turned on its head.

“I was beside myself when the doctor told me I might have cancer,” explains Susanne. “I went through a very anxious period. But a professor who came up from Uppsala calmed me down when he thought I might have a sports injury. I played a lot of football at the time. But, my eyesight got worse and worse, and it soon began affecting my other eye. It was a relief when I was diagnosed with LHON a few weeks later. At least it wasn’t cancer!”

That was 1991. Today, Susanne is 43 years old

and has been working in the administrative office of Sikeå Handball Club since 2007. Sikeå is a small town in Robertsfors Municipality. She works part-time 75 percent. The club receives wage subsidy. She also receives 25 percent sickness compensation.

Susanne has a large vision loss in both eyes. She has no locomotor vision, but with her right eye can count fingers at a distance of half a metre. She has trouble seeing posts, pavements and other obstacles when she is out walking. It is hardest in the winter, when the snow eliminates all contours. Her job can be stressful at times, especially before major tournaments. When Susanne thinks things are stressful, she often gets a terrible headache and has more trouble seeing than usual. That is why she has applied for a new job within the municipality a few times.

“But, it’s really hard when you have a visual impairment.”



## **IMPORTANT TO TALK TO OTHERS IN THE SAME SITUATION**

When Susanne was diagnosed, it was clear she would not be able to continue in home help services since she could not make home visits any more. A social worker at the vision rehabilitation centre suggested that she apply for disability pension.

“That made me mad. I was just about to turn 19. I couldn’t imagine spending the rest of my life as a pensioner! I took the initiative to travel around to different workplaces and meet people with visual impairment to find out how they managed things. And I met with Arbetsförmedlingen’s vision team, which taught me that you can work and live a normal life even with a visual impairment.”

While in high school, Susanne interned at a group home for elderly with dementia. She contacted them and asked whether they had a job for her - and they welcomed her with open arms. She was diagnosed in March and started working in August.

In between, she took a seven-week employment rehabilitation course in Skellefteå.

“I learned a little Braille, how to wash clothes, and a bit about working in an office. It was useful, mainly because the course participants spurred each other on. There should be more courses like this, where you can learn more about how to better manage your daily life.”

Susanne spent seven years working at the dementia home. She performed all work tasks except handing out medicine, since she felt there was a risk of her mixing up tablets, which often looked quite similar.

But, the residents just got older, and sicker, and more helpless. Eventually, Susanne felt like she could no longer guarantee their safety.

## **EXPERIENCE IN HOW AN ASSOCIATION WORKS**

After project employment at the municipality’s Development and Field Research Unit, which needed help analysing operations, Susanne was offered a job as an administrator at the Swedish Association of the Visually Impaired (SRF) in the summer of 1998. There, she handled member issues until she was laid off in connection with the association’s reorganization in 2005. She was pregnant at that time and gave birth to a son in May.

“When I lost my job, I began studying part-time at the adult education centre Komvux - social studies, information technology, layout, and more. At first, I received money from my unemployment fund, but then a new government took over and it was stopped. So, I had to take student loans instead.”

Once she finished her courses, Susanne did not really know what she wanted to do. She was unemployed for six months. But then in October 2007, she heard that the handball club needed a new clerk.

“And I got the job. I had plenty of experience in how an association works after all my years with SRF. I knew how the board worked, I have an interest in sports, and the work as a clerk was very similar to what I did at SRF.”

Another contributing factor was that Susanne could be employed with wage subsidy.

"Unfortunately, not everyone understands that you get wage subsidy for a reason," says Susanne. "Some tasks simply take me longer to complete. Comparatively, things were a little more sheltered at SRF - at the same time, it's fun when things just roll along. Even if it can be a bit stressful at times."

At all of her jobs, Susanne received help from Arbetsförmedlingen's vision team to get the most crucial technical aids she needs. Particularly, a 24-inch widescreen monitor, where she can magnify all of her computer-based documents. For her home, she has received aids such as a reading camera, magnifying program, as well as a number of tips on things like apps for her phone.

The only aid she has received at the administrative office is the large monitor. No adjustment to the lighting or anything like that.

"No, the environment hasn't been adapted at all," she says. "But, it doesn't bother me so much. I know it so well."

### **IMPORTANT TO KEEP YOUR SELF-ESTEEM**

Even though Susanne may lament not being able to drive a car, she knows you can live with a visual impairment and have a good life.

"You may have to struggle a bit more than you would have otherwise," she says. "I may not have had the same freedom when choosing what type of job I wanted to look for. But being forced to fight for what you want helps you grow as a person."

Susanne's main advice for those who develop a visual impairment is to contact others with poor vision as soon as possible.

"For me, it was really helpful and comforting to get living proof that life wasn't over, and I could live a normal life with family and job," says Susanne, who has been married to her high school sweetheart for a number of years - a man she met before her LHON diagnosis.

"Then, you have to show them what you've got," she says. "Part of that is acquiring skills, knowing how to use the computer, being good at keyboard finger-ing, and so on."

"Try to develop your social competence." It may sometimes be needed as compensation for when you need more time to complete a task. For example, make sure you are always on time and keep deadlines."

One of the most important things, according to Susanne, is making sure you keep your self-esteem. Remember that you still have your personality and your value as a human being. "Because it's really easy to lose self-esteem when you get a diagnosis like this," she says. "No one wins by you becoming a puddle of grief."



# WITH NEW CONDITIONS

A visual impairment can come quickly and unexpectedly. It turns your life upside down and makes you change the way you live.

Whether you have had a visual impairment all your life or it sneaked up on you over a long period of time, it creates conditions you need to adjust to. But, if it happens quickly - a person with LHON can lose most of their vision in just a few months - it is a very traumatic and shocking experience. It is then important to find contexts in which you feel secure and energized. It can be through your family and loved ones, good friends and co-workers, knowing how to handle practical tasks in daily life, and having good contacts, like your local vision centre, Arbetsförmedlingen and organizations like LHON Eye Society.

## **AM I REALLY BLIND?**

All of the terms used in relation to vision loss can be confusing. A common definition of visual impairment is "when it is difficult or impossible to read printed text or if it is not possible to orientate oneself using vision". A more medical definition is that you are visually impaired if your vision is worse than 0.3 on a standard eye chart with the best possible correction.

At the same time, vision is made up of a number of components. That makes it hard to provide clear

measures of what visual impairment really is. In addition, different people experience very different problems despite having the same level of vision loss. It is therefore essential not to compare yourself with others, but instead base things on your personal needs and your own life situation.

## **CAN I DO THE SAME THINGS I DID BEFORE MY VISION IMPAIRMENT?**

It is fully possible to continue doing most activities, and even try new activities. Naturally, there are some limitations, like driving a car or doing a job that is dependent on being able to see well. But overall it is fully possible to continue doing most of the things you did previously.

If you take advantage of the rehabilitation offered, you can handle almost anything with the right aids and training. Of course, it is also a matter of your own ability and motivation. You have to be aware that whatever you do will take more time than it did before. But you can do it. Having poor vision is impractical. You have to be more disciplined and patient than before. Talking to and comparing experiences with others who are dealing with new conditions can be psychologically beneficial and give you a number of practical tips.

## **I'M REALLY WORRIED ABOUT MY VISION LOSS.**

### **HOW WILL I MANAGE NOW?**

It is scary to end up in a situation where the conditions have been so radically changed. But, good help is available, particularly through the rehabilitation activities offered by the various vision centres. To visit a vision centre, you need a referral from your ophthalmologist. Through the national special-interest organization Swedish Association of the Visually Impaired (SRF) and other support organizations like LHON Eye Society, you can get in touch with others in the same situation, find information about the subject matter, or participate in the activities open to members.

Although it may seem hopeless at first, over time you will once again feel a desire to move on and return to your "old" life, although with new conditions.

### **HOW CAN I RETAIN MY INDEPENDENCE AND SELF-RELIANCE?**

Everyone who suffers a vision loss goes through an initial period where situations that used to be easy to master are suddenly full of challenges that must be overcome.

As you "learn" and become more comfortable with your new situation, you realize the importance of being able to manage things yourself as much as possible. It gives you both self-confidence and self-esteem. Avoid making things easy on yourself by asking for help in situations that you really can handle yourself. Tell your family and friends what you can handle and what kind of help you actually need. Your ability to

live independently will then gradually increase.

Remember that your family's desire to help often lies in their frustration over their inability to make things better for you. In other words, reach an agreement with the people in your close circle that you will ask for help when you need it. That eliminates the guess work and avoids a lot of irritation.

### **HOW CAN I TAKE CARE OF MY HOME AND HOUSEHOLD?**

The first and most important thing to think about is to make your home safer. A visual impairment puts you at greater risk of hurting yourself in your home. For example, the visual impairment increases your risk of falling – a big problem for the individual and for society.

- Make sure that there are no dangling cords at the coffee maker, toaster, electric kettle or any other device.
- Check the carpets so you do not trip or slip on them.
- Get good lighting throughout your home. See this book's section on lighting.
- Never leave doors half-open. If there are others living in the home, make sure everyone understands the risk of leaving a door half-open. Doors must be completely open or closed.
- Get an anti-slip mat for your bath or shower.
- If you have glass doors, mark them clearly.

Another thing to remember is that keeping things tidy and organized is essential. To make it easier for you to move about freely and safely, careful thought should be put in how you arrange the furniture. Allowing a lot of free space for walking helps you to stay oriented.

A proven tip is to follow the right-left principle. This means that you (and other family members) place your items to the far left, for example. This can apply to towels, toothbrushes in the bathroom, shoes in the hall, clothes on hangers, and so on.

In general, decide where things belong and always put them back in this spot once you are done using them.

### **CAN I GROCERY SHOP AND COOK FOOD?**

Being able to go shopping on your own once you have developed your visual impairment will give you a sense of freedom and independence, but may feel practically impossible. There are ways to get around some of the difficulties.

Give careful consideration to where you want to shop. Good service and positive treatment are important. Plan your purchases carefully. If you can, choose a time when there are fewer people in the store. That makes it easier to get help if you need it. If necessary, you can label the products right in the store. This makes things easier for products like milk and spices that come in different versions that have similar-looking packages. There are different labelling systems to choose from. Check with your vision centre or Iris Hjälpmedel.

There are a number of ways to make cooking easier.

- Use large pots and large bowls to reduce the risk of something boiling over or spilling.
- Turn the handles of pots and pans inwards so you don't accidentally get caught on them and pull something down.

- Utensils with long handles reduce the risk of you burning yourself.
- Make sure things sit steadily and do not slide around easily.
- Frying is difficult. Bear in mind that a lot of items can be cooked in the oven just as easily.
- Be careful with oil, which could catch fire. There is no noise or bad smell when the oil gets too hot.

For more cooking tips, see "Vardagstips för synskadade" [Everyday tips for the visually impaired], which is found on SRF's website. It also contains other practical tips on everything between heaven and earth.

### **WHAT TYPES OF WORK CAN I HAVE WITH A VISUAL IMPAIRMENT?**

In general, a person with a visual impairment can handle almost any occupation. There are a number of examples, both in Sweden and internationally, listing well over 100 different jobs held by a person with a visual impairment. When choosing a field, you should think about what skills and interests you have.

If you had a job when you developed your visual impairment, you should always try to continue working at your current workplace. Do not quit and do not listen to anyone who may tell you that you have to stop working. With adaptation of the workplace, good technical aids, and maybe even altered work tasks, there are great chances of you being able to fully manage a job situation.

In many cases, one difficulty is convincing the em-

ployer that you are still capable of handling your work tasks despite the vision loss.

With the vocational rehabilitation offered by Arbetsförmedlingen, advice and support, and the aids and initiatives directed to both the employer and yourself, there is a good likelihood of you continuing your work or starting a new job. If you are unable to continue in your present job, or you have not come out into the workforce yet, please refer to this book's section on searching for a job.

Experience has taught us some things about a successful career. You must be good at your job. For you to be able to work full time with a visual impairment, the work must suit you and the working environment must be good. Make demands. Work should not tire you out so much that you have no energy for a social life.

Getting good at using your aids is essential. Take the time you need to thoroughly learn all the options available through modern IT technology, like computer adaptations and smartphones with a large and growing range of apps.

It is also important to be mobile and good at travelling and finding your way on your own. In many jobs, this is a must.

Finally, the elusive skill commonly referred to as "social competence" is very important. As a person with a visual impairment, it is important to be able to explain your level of sight in a good and undramatic way. There are many situations where you will need to ask for help, and in such cases it is much easier if

you are flexible and keep things light.

If you have a severe visual impairment, you can get tips from the article "Bra jobb för gravt synskadade" [Good jobs for the severely visually impaired] on SRF's website.

## **ARE THERE AIDS THAT HELP WITH READING AND WRITING?**

There are a number of different aids to help with reading. These include optical and electron-optical magnification, but they require that you have some sight left. In addition, there are many types of reading machines, synthetic speech, Braille via a screen reader, and so on.

The ability to take in text has increased significantly at pace with technological development. No one has to lose out on communication nowadays. Having access to societal information as an individual is a right.

Anyone who cannot write in the traditional way can communicate via spoken information or computerized assistance. In many cases, it is a matter of motivation and personal choices, depending on how important you think it is to continue your previous ways of reading or writing.

Most of these aids are available via the county council vision centres. Their rehabilitation services investigate your needs by working with you directly or through Arbetsförmedlingen and Försäkringskassan. They then help you choose the aids best suited to you, your needs and your preferences.





# ARBETSFÖRMEDLINGEN [Swedish Public Employment Service] AND FÖRSÄKRINGSKASSAN [Swedish Social Insurance Agency]

## **VISUAL FUNCTION INVESTIGATIONS AND AIDS**

Do you have a visual impairment that limits your opportunities in working life? Then, a vision specialist may be one member of the team you meet at Arbetsförmedlingen. The vision specialist focuses on aspects such as investigating the consequences of your visual impairment and how your job situation can be adapted.

Arbetsförmedlingen has six specialized units in the country that can provide support to individuals with a visual impairment. In addition to vision specialists, there are social consultants, psychologists and SIUS consultants (SIUS - Special introduction and follow-up support). They also have contact with companies which supply work aids, the county council vision rehabilitation/vision centres, eye care professionals, the National Agency for Special Needs Education and Schools, and others.

Individuals with poor sight often need personal guidance and technology to be able to use a com-

puter, read a text, or write. Assistance is often in the form of magnification programs and synthetic speech, and learning how to handle help functions in the phone or tablet.

Or it could relate to optics, lighting, ergonomic advice or other special solutions for those with very special needs.

If you seek and get a job, you can get funding for your aids from Arbetsförmedlingen during your first year of employment. After that, Försäkringskassan and/or your employer takes over responsibility for paying for aids at work. If you have a job with wage subsidy, Arbetsförmedlingen is responsible for the adaptations and work aids that are needed for as long as the support is provided.

## **ROUTES TO WORK**

People with impaired vision who use optics or different types of magnification systems sometimes need answers to questions like: How do I read instructions?



How can a read off a measurement? How do I inform people so they understand my needs? How can I read off a script and at the same time see what I'm typing in my computer?

Arbetsförmedlingen's vision specialists work with both those entering the labour market and those who work with a wage subsidy. They are happy to answer any questions from employment officers, employers, job seekers, Försäkringskassan offices, and others.

### **SUPPORT FOR PERSONAL ASSISTANCE**

This support is intended to financially compensate the employer for additional costs stemming from an employee with disability requiring an assistant.

A personal assistant can be a co-worker who handles tasks the person cannot do themselves. A person with limited sight cannot drive a car. Or may have difficulty going through the post quickly, or reading and finding the needed information in a large amount of text. In such cases, they may need a driver, a reader, an extra secretarial resource, or someone who guides them to places they cannot find or have never been before.

### **WAGE SUBSIDY**

Wage subsidy is financial support to an employer who hires someone with a disability, that is to say an impaired work capacity. This makes it possible to utilize the person's competence and skills in the regular labour market.

### **INTERNSHIP**

The main purpose of an internship is to strengthen the individual's ability to get a job. An internship at a workplace is used to provide professional orientation, professional practice, work experience, or to maintain and strengthen professional competence. A supervised internship can also be used as part of preparation for the person to start their own business.

### **FIXED DEVICES FOR BETTER ACCESSIBILITY**

Arbetsförmedlingen can also provide financial support to the employer to make adaptations to the premises and environment that make it easier for the employee to find things or move about the workplace. This can include better lighting, blocking strong sunlight, accessibility for people with a physical disability, or other environmental measures that are positive and make it easier for a person to get or keep their job.

### **SPECIAL INTRODUCTION AND FOLLOW-UP SUPPORT**

This is individual support provided before and during employment for up to one year. It involves an SIUS consultant with competence in introduction methods helping job seekers who need individual support to find, obtain, keep or return to a job.

The SIUS consultant helps them find a suitable internship location that could lead to employment, and follows along to the workplace to ensure that the employee gets help getting to the job and making

their way around the premises, performing the work tasks, and gradually increasing their work tempo.

#### **LABOUR MARKET TRAINING PROGRAMME PROVIDES NEW KNOWLEDGE**

For some people, it may be necessary to supplement their knowledge or change profession to enter the labour market. A labour market training programme could then be a good opportunity. During the programme, you can receive support from Arbetsförmedlingen in the form of aids and materials.

#### **STARTING A BUSINESS - A DREAM FOR MANY**

A person with a business concept can receive support to start their own business. They can receive both advice and special financial support during start-up to help the business become competitive and adapted to both the ideas and the business owner.

## *REACH OUT*

**YOUR LOCAL ARBETSFÖRMEDLINGEN** can help you contact the vision specialists you need to meet for a visual function investigation and a discussion on aids. They will also help you if you need to meet any of the other specialists at the unit.

[www.arbetsformedlingen.se](http://www.arbetsformedlingen.se).

Phone: 0771- 416 416



**FÖRSÄKRINGSKASSAN** provides financial assistance for consultant investigations and aids to individuals who have been employed for more than one year and are not receiving wage subsidy.

[www.forsakringskassan.se](http://www.forsakringskassan.se).

Phone: 0771-52 45 24



**AID SUPPLIERS** are organized in an industry association called Svensk Syn. You can reach the various companies most easily via their website [www.svensksyn.se](http://www.svensksyn.se).

Leverantörsföreningen  
**SVENSK SYN**



# FAMILY OF SOMEONE WITH LHON

**THE WHOLE FAMILY IS AFFECTED** - *When someone in the family develops LHON or another visual impairment, it affects the whole family, particularly in the form of stress and depression. What can and should be done?*

Since LHON is passed down through the mother's side of the family, mothers often feel guilty about passing the disease to their child/children. There are often no previously known cases in the family, and you are unaware that you are carrying the mutation. The shock can be particularly severe the first few years, especially right at the start. Since LHON affects about half of the men who carry the trait and about 10 percent of the women, it is a bit like a lottery.

Other family members are also affected, and relationships within the family can easily become strained. Siblings may worry that they will also develop the disease. It is both common and natural that the person and their family members feel that life is unfair, and psychological stresses can cause not only depression but also feelings of irritation and anger within the family.

## **WHAT IS THE RIGHT AMOUNT OF SUPPORT?**

Everyone may feel really unsure about what is required of them - and how much they should help the person in both small and large tasks. It can be both difficult and emotionally-charged to give the person

the help that they are requesting. Many who develop LHON want to take care of themselves as much as they can. Getting "too much" care in day-to-day life may make the person feel more helpless than they really are. Giving too much care and help can often amplify a disability rather than being of real help.

Many practical aspects of daily life are affected. Common household chores like cooking, driving, washing clothes, and going shopping suddenly become difficult to share. Providing psychological support and perhaps taking on greater financial and practical responsibility for the household may be quite stressful for a family member. Especially, as said, before aids are in place and the person has learned new techniques for handling practical household chores. It takes a bit of sensitivity to, for example, discuss how the home should be decorated - adapted lighting, where everything should be situated, and so on.

## **HARD TO KNOW WHAT SOCIETAL SUPPORT IS AVAILABLE**

It is not easy to find out what support possibilities the community offers or what rights you have as a



person with a visual impairment. In the beginning especially, the person who developed LHON needs help to figure out what applies, along with numerous and sometimes complicated contacts with the various healthcare centres, the Försäkringskassan office, and so on.

Family members who want to help, also need to read up on everything related to social contacts - not least because LHON is a rare disease that healthcare professionals often lack knowledge about.

Even if you get advice from others in a similar situation, it can be complicated by the fact that the support society provides can take different forms in different parts of the country - even if the same laws and regulations apply. In principle, the vision centres have social workers and psychologists available to talk to, and some vision centres offer study groups and group meetings for both individuals with visual impairment and their family members.

There is a risk that some medicines may make the disease worse or trigger it in other family members. This is another matter where healthcare professionals often lack sufficient knowledge. This makes it especially important to be well read and/or have the support of e.g. a diagnosis association in your contact with healthcare.

The National Board of Health and Welfare has published a useful article about supporting family members of people with disabilities. There is also the Swedish Family Care Competence Centre.



*Doctor Martin Engvall of Karolinska University Hospital wrote the majority of the LHON information available on the National Board of Health and Welfare's website.*

**THE NATIONAL BOARD OF HEALTH AND WELFARE** has published a great deal of information on its website [www.socialstyrelsen.se](http://www.socialstyrelsen.se). You can find a lot of information by searching for LHON on their website. You will then find a wealth of informational material with links to more detailed information. The information is intended for the public and for professionals.

**NATIONAL BOARD OF HEALTH AND WELFARE**

# MY SON DEVELOPED LHON

In autumn 2009, my son Fredrik began to complain that he had trouble seeing. He wanted to get his vision checked. But, since just a few years earlier we were told he had extremely good vision (he 'should become a pilot' according to his doctor), I thought for quite awhile that it was migraines. I gave him paracetamol tablets, but it did not help. The day before Fredrik's 15th birthday, we finally went to an optician. It's hard to describe the shock, fear and the feeling that this can't be real when we discovered that he not even see the top line of the eye chart with one eye. And glasses didn't help.

We spent his 15th birthday at the eye hospital with doctors and nurses who seemed just as surprised as I was. They performed a lot of different tests, consulted with each other, checked things in books, and finally said they didn't know what it was, but they suspected a brain tumour. They didn't tell Fredrik that last bit, but he realized that it was something they suspected. I think you develop a sort of superpower as a parent in a situation like this. Most family members would probably agree with that. No matter how worried or upset I was, I did everything in my power not to show that to Fredrik, and it worked.

I met with both a social worker and a psychologist and tried to be encouraging and comforting and talk

about how good the medical care is and that it will probably get better soon.

Like many others with LHON, no one in our family had the disease, which is why it took a while before the doctors could make a diagnosis. Instead of a moped and other fun things that 15-year-old guys usually do, there were a number of hospital visits to ophthalmologists, neurologists, and others. Just before Christmas, I got a phone call I'll never forget. I learned that the doctors suspected a rare hereditary disease that can only be inherited from the mother, and they wanted to perform a DNA analysis before the holiday.

Over Christmas, the whole family went away on a trip. We thought about whether we should really go,



and I'm very glad we decided to do it instead of sitting at home, crying and worrying about the future. Doing activities together helps both the person with LHON and the family members think about something else and let life go on. It was easier to enjoy the moment and make the best of the situation at the time.

Fredrik still had good sight in one eye, and it made my heart feel really good to see him see and experience so many new things on the trip. Best of all was when Fredrik and I went on a hike in the jungle.

When we got home, the doctors performed another brain scan, but didn't find anything wrong. It was reassuring to know it wasn't a brain tumour.

About three months after the DNA analysis, we got the results. Fredrik had mutation 3460 and a large proportion of mutated mitochondria. We also learned that even if you have 100 percent mutated mitochondria, there is a chance that all or part of your sight will come back and the other eye won't be affected.

Spring and summer were a tough time, and we oscillated between hope and despair. Fredrik began taking Q10 before Christmas and began with Idebenone soon after he was diagnosed. We hoped that it would cause the disease to reverse itself.

Landing in the new situation and thinking about the future are important in order to be able to move on in life - both for the person with LHON and for their family.

In Fredrik's case, it took almost a whole year until the other eye was affected. By the summer, I knew

the doctors had seen signs that something was going on with the right eye as well, but I hoped that they were wrong.

When LHON strikes, it can progress very quickly. Fredrik went from vision of 0.8 in the right eye just before the start of school to just 0.01 by the end of the first week of high school. There were many visits to the vision centre and visits to the ophthalmologist and neurologist. Fredrik didn't want to talk to a psychologist, but thankfully I could. As a mother and carrier (I have 50 percent mutated mitochondria), it's easy to feel a sense of guilt and it took a long time before I was able to get over that feeling.

I used to have a job that involved a lot of trips abroad. When Fredrik became severely visually impaired in his other eye, I told my manager that I did not want to have to travel for the next few years. Three months later, I was terminated due to work shortage.

Losing your job in and of itself can be a minor tragedy. Combined with everything else, it made things really tough for a while. I wasn't doing well myself and was not the best mum in the world every day, but thankfully Fredrik had come a little closer to accepting the situation and then slowly began learning to live with the disease.

As the saying goes, "every cloud has a silver lining" - thanks to losing my job, I had some time on my hands and took the initiative to start an association for LHON. It helped me greatly to be able to talk to

another mother whose son had the same mutation and developed LHON just before Fredrik. I therefore thought how great it would be if there was an association where we could support each other and even be able to support research. After a little digging, I managed to find a number of people with Swedish-sounding names within an international LHON group on Facebook. I also had some relatives on my mother's side who each were or could be a carrier. On 31 March 2012, we had our inaugural meeting and the LHON Eye Society was formed.

Another benefit of losing my job was that I had time to read a lot of research reports on LHON and quickly became an "amateur-expert" on the medical aspects. I, Fredrik and the healthcare professionals all benefited from this. A dilemma with a rare disease like LHON is that most healthcare professionals do not know about the disease. In the beginning, Fredrik had a lot of questions for the ophthalmologist that I was often able to provide better answers to. We therefore decided that another important task for our association could be informing and educating healthcare professionals about LHON - through this book, for example.

*Helena Lindemark*

---

*Helena and her son Fredrik on a visit to the jungle when he was 15 years old - before his second eye had begun deteriorating.*





# MANY DIFFERENT WAYS TO REACT

A social worker at a vision centre will meet many different reactions to the same vision problem, the same visual impairment, and the same family situation.

Bodil Jönsson, Professor Emerita in Lund, where she has spent quite a few years working in rehabilitation technology, has expressed this as follows in a lecture: "The perceived disability is not the same as that diagnosed or treated."

In other words, it is practically impossible to know how a person will react to a LHON diagnosis. LHON is characterized by rapid vision loss, often when the person is relatively young, that leads to severe visual impairment. This means that their entire living situation changes quickly. Rehabilitation must therefore start as soon as possible.

Rehabilitation can vary, from a comprehensive plan for how to get on with things and function through acceptance and then training new behaviours to quickly implementing measures to enable the person to continue where they were and gradually address how new needs can be met.

Past experience is not always enough to master the completely new and unfamiliar situation. Existential questions are common: "Who am I now that I can no longer drive a car, moped or motorcycle?" "Who am I now that I can no longer work in my old profession?" "Can I be a good parent/partner?"

There are no special rehabilitation programmes or templates to follow. An individual's attitude towards the visual impairment and rehabilitation is affected by many different factors, such as age, whether they are studying or working, and previous experience with disability, whether their own or someone else's.

It is often easier to relate to the situation if the individual already has positive role models of others who have been in a similar situation. At the same time, family support is crucial, by showing "we love you just as much now as before".

Another important factor is the general view of people with visual impairment as it is likely that this view will also be held by the person affected. Different cultures can have completely different views of

people who develop a disability. In some cultures or families, the shame may be so great that people with disabilities are hidden away. This type of attitude makes rehabilitation more difficult.

### **A SHARED RESPONSIBILITY**

Nowadays, a disability is not considered to be a matter an individual must face alone. According to the World Health Organization, the degree of disability is determined by how the person's environment is designed. Naturally, this is a matter of integration, that is to say the external environment supporting the individual's ability to handle the situation and continue working from a new starting point. Of course, without the drama and abyss described in older literature about what happens when a person loses most of their eyesight.

Attitude and reaction can differ greatly depending on age. Children and adolescents have less opportunity to make use of life experience. At the same time, such experience can make it more difficult for an older person to create new living patterns. Children and adolescents often have closer ties to their family. The risk of being overprotected by parents is great.

### **FROM SHAME TO RECONCILIATION**

It is very important that the individual gets help to deal with the grief and the crisis and to talk about

the feelings awoken by the visual impairment. It is common to feel ashamed. Since the visual impairment is not visible, it is easy to hide and many try to keep it a secret. This makes it harder to ask for help. It may take time before the person comes to grips with the fact that this is the way things are.

Reconciling oneself to the poor vision is a process. The radio personality Tappas Fogelberg summarizes it as follows: "It's my eyes that are bad, not me."

*"I don't accept  
my visual  
impairment, but  
I have to learn  
to live with it."*

Rehabilitation is about helping the person gain insight into and the ability to overcome the limitations related to the visual impairment with the help of training and aids. Over time, it also involves the person reorienting themselves in terms of work and other activities.

It is natural for people to resist being identified with their visual impairment. They may therefore have trouble accepting the rehabilitation and the aids offered, such as optical aids that involve being very close when you read or using binoculars when looking at something farther away.

Even though the aids could make it easier for a person to get around on their own, there is often a great deal of psychological resistance. You want to be like everyone else and blend in, not feel different or vulnerable and in need of help.

It is important not to try to persuade the person, as this will only make the resistance stronger. You must

instead be patient and wait until they develop their own motivation. Because learning to think “I have the right to be different” takes time. A great help may be to meet others with a similar visual impairment. Joining an association for individuals with a visual impairment may be an important step to take. Positive role models play a huge role in an individual’s attitude towards their visual impairment: “If they can, I can...” It is a matter of trying to help the person go from feeling sorry for themselves (victim mentality) to seeing the visual impairment as a challenge.

During rehabilitation, you also need to help people see adversities as learning experiences rather than mistakes. Both winning and losing are part of life. Not everyone may have learned that life lesson yet. This is especially true of young teenagers, who have not left the security of their family and been exposed to the idea that you do not always “win”. The question is: how does each person relate to their strengths and weaknesses?

### **THE WHOLE FAMILY IS AFFECTED**

When a person develops a visual impairment, it affects the whole family and the person’s loved ones. Understandably, a lot of focus is on the person who developed the impairment, for example with doctor visits, sick leave, the person’s grief over the matter, uncertainty about the future, etc.

For family members, it is often a time of trying to give their loved one everything they need. You be-

come dependent on each other in a new way. It may involve driving them to various doctor visits, taking care of new tasks in the home, providing comfort and looking for possibilities, and staying positive.

They often give little time and energy to dealing with their own grief and their own reactions. Getting this opportunity is important. It is important to be seen for who you are and not just as a relative or loved one. Questions like “And how are you?” and “How are things going for you?” are important to give the family member a chance to stop and think about how they are doing. It may also be very helpful for family members to have individual counselling sessions as a complement to sessions with their partner, with other family members and loved ones.

Naturally, it is important for those personally involved to get information about their loved one’s vision loss and how they can best provide support and help. In many cases, a family meeting can be a good way to inform family members, especially children, who often have questions of both a practical and an emotional nature. However, you must always remember how different things can be from one case to another.

At an information meeting, the family members can, for example, test out simulation optics, which are glasses that give them an idea of what the visual impairment is like and thereby better understanding of what it could be like to live with it. Getting family and loved ones involved in rehabilitation is of great value.

## **SOCIAL WORKER'S ROLE**

The most important tool for all counselling sessions is your own intuition and your ability to be perceptive. There is no blueprint for how to approach things. It is our own inherent ability that helps us understand each person's needs and find an approach that is right for that particular individual.

The social worker's approach involves being inquisitive, perceptive, and responsive, and meeting the person where they are at the moment. It means being careful when bringing up issues the person has not touched on themselves.

The starting point for all sessions is how the person is feeling, what questions they want to address, and generally what limitations they perceive, how they can be overcome, and what kind of help they would like.

Working together to set up an agenda is an important part of making the person feel involved. The aim is to reduce anxiety, create a sense of context and trust, and build bridges that invite dialogue. Another important role for the social worker is to provide vicarious hope – in other words, to communicate the message that there is hope.

Asking open questions gives the individual a greater opportunity to choose their answer.

## **EXAMPLES OF QUESTIONS FOR THE FIRST SESSION INCLUDE:**

- What does the visual impairment mean to you?
- How do you perceive your situation?
- Is there anything in particular that you've been thinking about and want help with?
- How do you feel those around you are responding?
- What limitations and opportunities do you see based on your visual impairment?
- What information has your ophthalmologist given you about your eye disease?

## **THREE TIPS FOR PEOPLE WHO HAVE DEVELOPED LHON**

1. Keep in mind that even if the situation is tough, the body needs to be kept in shape. Physical activity and a varied diet are important. Activities fight against depression, so try to be active in different ways.
2. Try to accept the rehabilitation offered. Ask for a rehabilitation plan so that you get involved in the rehabilitation. Think about what limitations you want to overcome and set clear, measurable goals so you can recognize when you make progress.
3. Contact others in a similar situation as soon as possible to learn from their knowledge and experiences. This can be through group activities offered by the vision centre as well as through an association.

**10  
CM**

**2.5X**  
MAGNIFICATION



**8,5  
CM**

**3.0X**  
MAGNIFICATION



**6,2  
CM**

**4.0X**  
MAGNIFICATION



**4  
CM**

**6.0X**  
MAGNIFICATION



**3,1  
CM**

**8.0X**  
MAGNIFICATION



**2,5  
CM**

**10X**  
MAGNIFICATION



# MAGNIFICATION - THE SOLUTION FOR IMPAIRED VISION

*Learning to use magnification correctly can be a great strategy for mastering the small and large challenges of day-to-day life for everyone living with LHON. With LHON, the symptoms are the same: a loss of vision straight ahead. This is called a central scotoma. Visual acuity is greatly reduced. One guideline is that if visual acuity is worse than 10 percent (i.e. 0.1), you have a total central scotoma or vision loss. When visual acuity is better, you can still use your central vision. This is referred to as a relative scotoma.*

## IT MAY SEEM A BIT COMPLICATED

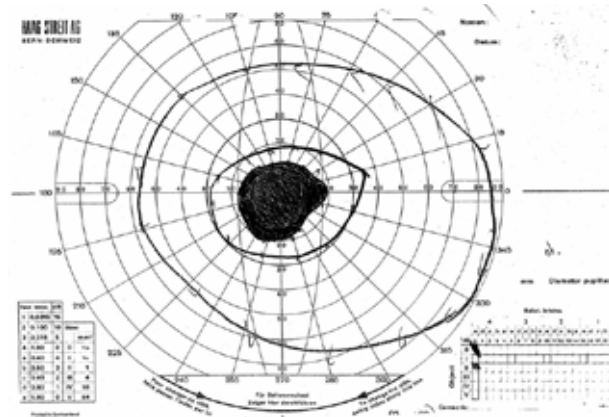
In reality, full vision does not mean 100 percent. Full vision is usually said to be having vision better than 0.7. There are people who have visual acuity of 2.5 or even 3.0 - without binoculars! So, we are all individually different.

In each and every case. Visual acuity below 0.7 is usually called “reduced vision”. If it is below 0.5, you are no longer permitted to drive a car. If it falls below 0.4, you are referred to as partially-sighted.

When visual acuity drops below 0.1, you are said to have a severe visual impairment and you sometimes (in unknown environments) have limited or no locomotor vision. If you cannot see anything at all, you are blind. People diagnosed with LHON, Leber’s Hereditary Optic Neuropathy, often have residual sight and a visual acuity of less than 0.1 - although there are exceptions where people with both 0.2 and 0.3 have been diagnosed with LHON. The consequence is usu-

ally a visual acuity of 0.02-0.08, which means no or almost no function in the macula.

This means that the area in and around the macula (about 5-10 degrees of the central visual field) is basically gone. The retina and visual field are divided into central vision and peripheral vision. A full peripheral visual field is 180 degrees with both eyes. It decreases



*A central scotoma reduces visual acuity to below 0.1.*



to about 130 degrees when you look out of just one eye. The macula is just a few millimetres in diameter and makes up just 5-10 degrees of the central visual field. Some people are able to see through the fog (the cloudy central scotoma) and use the few remaining vision cells in the macula, but a great deal of light and movement by the object are usually required in order to sense an object centrally. In basic terms, you cannot see forward. You can then use eccentric viewing - in other words, finding another spot on the retina to position the image. Refer to the next chapter on eccentric viewing!

### 0.5 IS REQUIRED TO READ

It takes a visual acuity of at least 0.5 to be able to read a regular newspaper text (in 8-point font). This means that someone with 0.2 must enlarge the text 2.5 times to be able to see to read. The text must then be held closer to the eyes to make it bigger on the retina.

The publication KOM NÄRMARE [Come closer] describes the entire scale, from 2X to 10X magnification. See a brief version at the beginning of the chapter and on [www.lhon.se](http://www.lhon.se).

From a 35-40 centimetre reading distance for the sighted to a 2.5 centimetre reading distance with the strongest magnification in glasses frames - 10X magnification (or 40 dioptres optic power in optician-speak. A dioptre is a measurement of the optical power of the lenses, plus and minus). The reading distance is obtained by dividing the dioptre figure by 100:  $100:40=2.5$  cm. There are even stronger magnification

systems, but the nose could then get in the way...

If you want to know what magnification you need, you can test it in KOM NÄRMARE at the vision centre or at [www.exfix.se](http://www.exfix.se). Using your best eye, try to read the large text at a distance of 25 centimetres. Start from the back and then move towards the front until it no longer works. You can then see what magnification is needed in your case and whether your visual acuity multiplied by the magnification equals 0.5, which is the limit for being able to read.

### EXAMPLES:

- If you have a visual acuity of 0.1, you should use 5X magnification to obtain a visual acuity of 0.5. 5X magnification is optically 20 dioptres and makes the reading distance five centimetres. Add an additional 1X magnification for every four dioptres. 24 dioptres equals 6X magnification and so on.
- If your vision is 0.05, 10X magnification is required and you must hold the text 2.5 centimetres in front of your eyes and the optical aid.  $10 \times 0.05$  equals 0.5. However, if you need 10X magnification, you must learn to direct your gaze above or below the text since you do not see anything straight ahead. The central scotoma must be positioned above or below the text to give the text access to an area that is not damaged by LHON (or another cause). However, the area outside of the macula has a poorer resolution capability than within the macula area, so you need to find your Preferred Retinal Locus (PRL).

## OPTICS FOR MAGNIFICATION

It is not the optics themselves that magnify. Magnification occurs when you move closer. Optics enable you to hold something close and at the same time get a sharp image on the retina.

It is the same phenomenon that most experience when they reach their 50s - you need reading glasses to be able to hold the text at a normal reading distance. In the same manner, you need strong reading glasses or magnification correction to be able to see larger text sharply.

Your specialist optician at the vision centre (or a private optician with specialist expertise) can test out what magnification you need for normal text or to be able to read from a screen with larger text. The optics must be adapted according to your visual acuity and what you want to view or be able to see.

## OPTICS UP CLOSE

Quality is important. This is usually said about everything from cars to food. When it comes to optics, the optical errors - referred to as aberrations - must be minimal when you need to use strong magnification.

One solution is to combine aplanatic lenses. This means there are two lenses with the convex (outwardly extending surface) facing each other that together provide the required magnification. It has also been found that the quality is better if you combine plastic lenses with mineral glass and all four surfaces undergo anti-reflective treatment, rather than just



*Two lenses in combination with anti-reflection treatment on all four surfaces produces the least amount of optical errors.*

a strong optic lens made of plastic. In addition, the visual field is larger if you use two lenses together.

There are several different variants of combined lenses to provide close-up magnification. A photo of a system called A2 (A for aplanatic and 2 for two lenses) from Multilens in Mölnlycke is shown above. The A2 system can produce 14X magnification, but 8X, 10X or 12X is most common because otherwise the reading distance is too small. However, it is need that is the governing factor.

## IT IS NATURAL TO READ CLOSE UP

The next step is to use the strong lens any time, anywhere, and in any way necessary. Naturally, there is

great resistance when you cannot read the same way as everyone else. Sitting on a train or bus and reading with the text all the way up to your nose attracts attention. But, you are allowed to be who you are and read the way you need to read. You are just like everyone else, you just use a different method when reading.

### QUICK READS

When you read a text in newspaper or document, mag-



*If you use a short reading distance, you must hold your head up so you do not develop neck pain.*

nifying correction in your glasses is preferable. However, when you need to read something quickly - like a price tag, the temperature on the washing machine, or portion instructions on the oatmeal package - a magnifying glass can be used. You can then look at the magnified text in the glass while holding it close. This differs from a loupe, where you help the eye see close like in the strong aplanatic lenses on the previous page.

### SEEING OR JUST DETECTING OBJECTS AT A DISTANCE

When you want to see and read a sign, you must also magnify it, but it may be hard to get up close to a street sign to see what it says. An alternative for such cases is to have binoculars in your pocket that provide 4X, 6X or 8X magnification. This gives you improved visual acuity and you can figure out where you are. Another alternative is to use the camera feature on your smartphone to zoom in on what you want to read.

Even at a distance, it is important to use eccentric viewing.

When you are out walking or biking, it is important to not look straight ahead all the time. Use eccentric viewing to avoid walking into or colliding with objects.

With LHON, the central scotoma is often concentric, meaning there is a hole in the middle of the visual field straight ahead. About 5 to 15 degrees of the central vision may be gone. The recommendation is to look up or down, either all the time or occasionally as you move forward. You will then detect objects in front of you in time so you can avoid them. If neces-

sary, you can use your binoculars or phone to check out what the object is.

Many individuals with LHON or another severe visual impairment say that they find it hardest to orient themselves on sunny days because the strong shadows can be perceived as “black objects”. When running or biking, adjust your speed based on how well you can detect objects by looking up, about 15-20 degrees. Even if you can read with eccentric viewing of 5 degrees up or down, a larger adjustment is required to see farther and more of a side view for orientation. Most direct their gaze 15-20 degrees upwards, which gives them a larger panorama.

It can sometimes be a good idea to find a new Preferred Retinal Locus (PRL) just to the left or the right of the scotoma if you want to detect things with more detail and at the same time use binoculars or your phone’s camera feature. Once you have become



*Binoculars get you closer and make things larger.*

more advanced in your use of eccentric viewing, you can have up to three different PRLs – for reading, for a quick read, and for orientation.

## **MAGNIFICATION AND FILTERS**

The publication “*Kontrast och bländning*” [Contrast and glare] on [www.multilens.se](http://www.multilens.se) describes different filters that can be used alone or together with magnification to increase contrast sensitivity and improve visual comfort. It is glare in particular that you want to avoid by combining different filters.

The cut-off filters remove certain parts of the visible light. The human eye can see between 380 and 780 nanometres (Nm), from ultraviolet to infrared.

Experience has shown that blocking the energy-rich blue light (from 380 to 450 Nm) can often produce better contrast. Adding a polarizing layer on top of the light yellow lens also reduces the incident light (transmission). This increases contrast while reducing

glare from the sun and haze.

The effect of filter lenses is very subjective. Test different filters in a variety of light environments to find the best “outdoor glasses” with or without distance correction. Sometimes, adding a filter to reading glasses can provide a better viewing experience, especially if you are light sensitive.

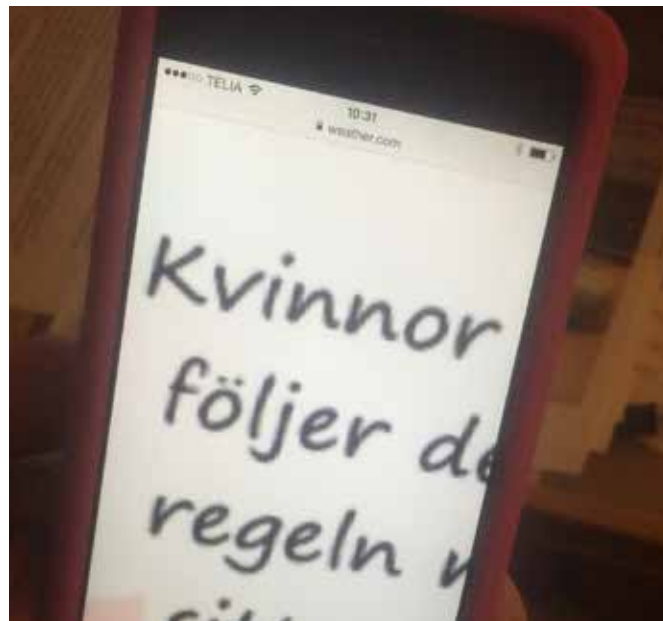
You won’t know until you’ve tried it out. What is good for one person might work for another. But, it’s far from sure. We are all different, despite having the same cause of our impaired vision. There are a lot of contributing factors:

- How far you have come in the process of accepting things as they are
- How well the other parts of the eye are working; floaters in the eye, for example, can cause glare
- How things are at work
- How well you have adjusted to changing optics in relation to the problem that needs to be solved
- If you’ve developed a habit of using your optics at the theatre, when looking for a specific address, or just reading a sign.

### **ELECTRONIC MAGNIFICATION**

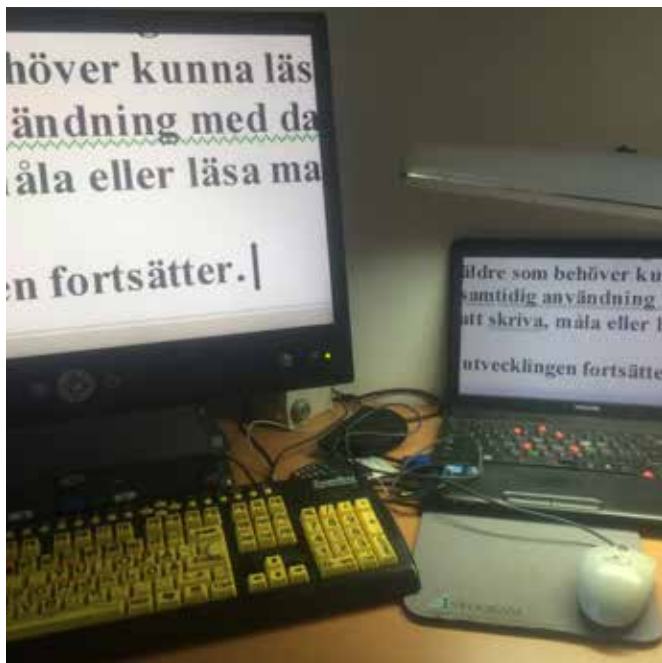
Apple entrepreneur Steve Jobs should be given a lot of credit for developing the iPhone, the world’s first smartphone, with the express idea of making it available to as many people as possible. In the basic version, if you go to “Settings”, “General” and then “Accessibility”, there are a number of features to help

both the visually and the hearing impaired. The visual aid features include Zoom and VoiceOver. Another part of the book describes the most common and simplest features for zooming or listening to text [GV7] in smartphone of various makes and models.



### **CCTV SYSTEM OR VIDEO MAGNIFIER**

The first CCTV systems (Close Circuit Television System) came out in the early 1970s, initially through a home-made design by the American mathematician Samuel Genensky. Today, there are a large number of products on the market that are designed to suit different uses. Video magnifiers can product magnification on the retina of up to 50-60 times, and provide a more comfortable working distance compared to optics in glasses.



*A video magnifier connected to a computer can be used to read a document under the camera while writing it.*

### **CCTVS IN MANY VERSIONS**

- For students with moving cameras that can view at different distances
- For the elderly who want to read their mail or solve the crossword puzzle
- For simultaneous use with a computer
- For writing, painting or drawing
- For reading text and typing simultaneously - with split vision, two images on one screen

And the development continues.

### **ELECTRONIC MAGNIFIERS**

You probably know that the camera feature on your phone is an electronic magnifier. But there are also other products that are designed exclusively for use as electronic magnifiers. They come in sizes ranging from 5 to 15 inches. And while they may feel too large for your pocket, they can prove very valuable when studying or working at your desk. You can place one over a document you need to see, or use it to read off a phone number or the number on your credit card. Let's call them advanced reading systems.

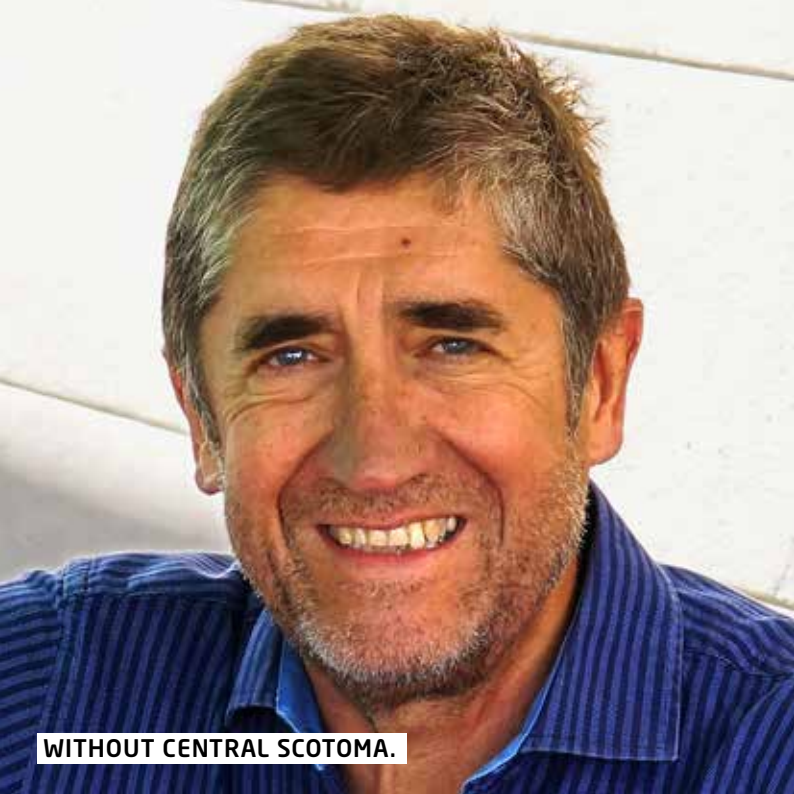
An iPad is another electronic magnifier with a larger screen when you use its camera feature like you would your phone's. You may then need to supplement it with magnification correction - strong positive lens - to obtain a sharp and enlarged image.

The development has just begun. We can already see systems that are as thin as paper and can photograph texts you need to read later, and even convert them to speech.

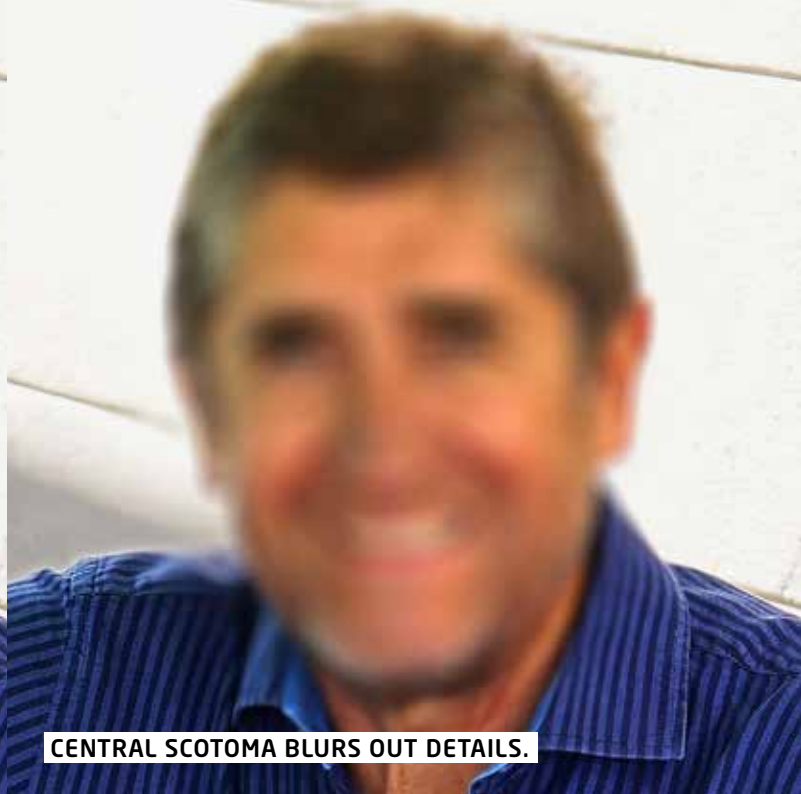


*You place an electronic magnifier directly on the text.*





WITHOUT CENTRAL SCOTOMA.



CENTRAL SCOTOMA BLURS OUT DETAILS.



ECCENTRIC VIEWING DOWNWARDS.



ECCENTRIC VIEWING UPWARDS.



# BUILD UP YOUR ECCENTRIC VIEWING ABILITY!

*Eccentric viewing is using your peripheral vision when your central vision is not working. Building up this ability helps you to manage day-to-day life better. Reading, orienting yourself and working practically! It improves your quality of life. This is how it works.*

There are two options available if you want to read but have no central vision. You either enlarge the image *a lot*. If you make a letter large enough, you can perceive it by all cones in the entire retina participating in resolution of the character. On the next page, you can see an enormous letter. Individuals with really poor eyesight can perceive this because it covers a large part of the retina when you view it close up.

But, it is hard to read such large letters, even when they are projected onto a screen using a CCTV system. Reading this way goes very slowly, one letter at a time - that is to say just one character in each fixation and fixation field.

A better option is to try to find a spot that still works outside of the damaged central parts of the retina. Vision research refers to this location in the retina as the Preferred Retinal Locus (PRL).

The PRL for reading can be found at a number of different locations for different individuals depending

on the shape of the scotoma: above or below, to the right or the left of the scotoma in the retina. By using a PRL test to try out these different gaze directions, you can find the one that is best for you.

## **WHEN SHOULD ECCENTRIC VIEWING BE USED?**

It is often said that when the central vision is completely gone, there is no alternative. Visual acuity is then 0.1 or lower. It can also be higher than 0.1 if you have multiple small scotomas centrally and do not have room for the text in "Gloria in the garden". Even then, it may work well to position the image and the text to the side of the impaired central vision.

By checking your visual field diagram, you can see whether the Preferred Retinal Locus you choose is really the best based on the measured visual field. When interpreting the diagram, it may be a good idea to consult an educational specialist for the visually impaired, an optician, or an ophthalmologist who

**K**

knows how the retina works

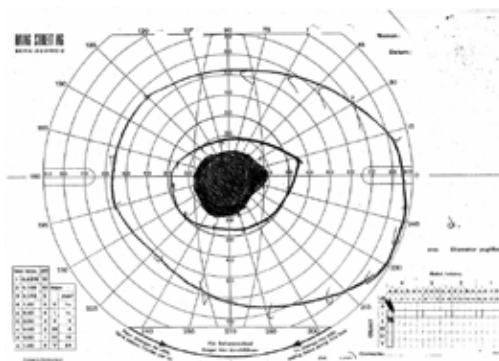
There are 180,000 to 200,000 cones per mm<sup>2</sup> in the macula, but there are only 8,000 to 10,000 in the periphery of the retina. Thus, it is not possible for vision to be as sharp. You must therefore compensate for the lower cone density in the periphery with high magnification. When positioning your PRL for reading, it is important to ensure it has a large horizontal position, in other words that the fixation field is as large as possible. We know that the fixations and the size of the fixation field determine how quick and efficient you are at reading.

### FIXATION LINES

Once you have chosen a PRL for reading, you need training to actually use eccentric viewing when reading. You can practice keeping the scotoma (the area of vision loss) away from the text by using text with fixation lines.

Such exercises are available on the website created as part of the Leber X project. You can find it via the link [www.exfix.se](http://www.exfix.se)

The fixation lines act like a “fence” that keeps the scotoma to the side of what you are trying to read.



*With the Goldman visual field test, you can see what central vision loss looks like.*

### SCROLLED TEXT

Another method is called MoviText. You then use a magnification program called Zoomtext 7.1 in your computer. With the screen set for magnification and the Zoomtext program open, you open Word and choose the document you want to read. You then use the command Control-Shift-r. This accesses a program module called DocReader with the Ticker Mode application. There, the text will appear at the top of the screen and you can set the speed and magnification that suits you best. When you press Enter on the keyboard, the text begins scrolling

from right to left and runs from the top of the text to the end. This is called scrolling text. While the text is moving, you can (without using optics) find the PRL you think works best and choose the magnification you need.

You can also try reading scrolled text at [www.exfix.se](http://www.exfix.se) to see how it feels. Training without optics in the beginning can make it easier to learn to use strong magnification glasses at a short reading distance later. It makes a big difference in quality of life.

In general, more than 8X magnification is almost always needed, and up to 10-12X magnification in reading glasses. You can get this using aplanatic lenses

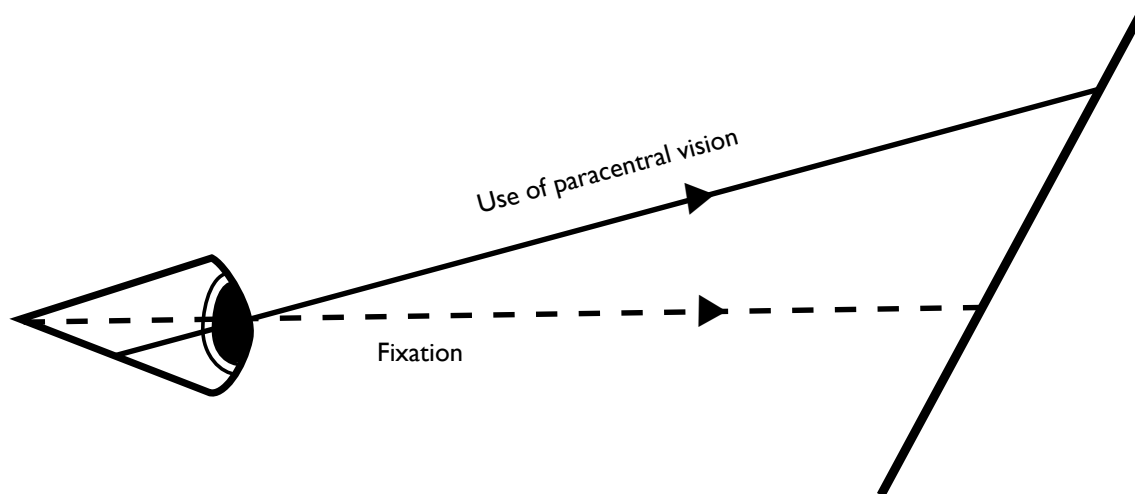
with the designation A2 from Multilens ([www.multilens.se](http://www.multilens.se)). If you train your eye movements, eccentric viewing, before starting to use powerful magnification optics, it will be much easier to learn the technique of using the peripheral vision outside of the macula when you read.

### **THERE ARE MANY DIFFERENT PRLS**

You have several different PRLs. One for reading, one for using binoculars, one when you ride your bicycle, and one when you are looking at the computer or watching TV. But, if you start by finding your PRL for reading, you are well on the road to looking at things a whole new way. The next step is to develop a PRL for recognizing people and finding signs.

A person can read, for example, by positioning the central scotoma above the text. The gaze is directed about ten degrees above the text so that the letters end up below the scotoma. A German method of training involves keeping the eye still and moving the text downwards or upwards. This is called "Xcentric Fixation" and is described in more detail at [www.exfix.se](http://www.exfix.se).

Another PRL could be to look to the left of a person's face when you want to recognize someone at a distance of half a metre. When riding bike or walking, you need to direct your gaze higher, upwards of 20 degrees from what you want to detect, to be sure nothing is in the way. Naturally, you should also adjust your speed.



*This simple drawing shows that you must direct your gaze above (or next to) the text for it to fall in the PRL. There, you can identify heavily magnified characters and learn to read eccentrically or recognize a face.*

## THE LATEST RESEARCH - DOES IT PAY TO TRAIN A PRL?

We asked Doctor Tina Plank of Regensburg, Germany, to comment on this text. She is a brain researcher investigating what happens in the brain when you train eccentric viewing and the ability to keep your gaze still in the chosen direction, known as fixation stability. What does she consider the benefit of eccentric viewing, and what happens in the brain when you have found and use a PRL?

She writes the following:

"When eccentric viewing works as intended, the person has chosen and uses an area outside of the central vision loss. Training can influence and help stabilize fixation so that you consistently use the selected area outside of the central scotoma, thereby facilitating daily vision tasks, like reading and seeing by directing a steady gaze towards an object of interest - with or without an aid. This enables people to regain their independence, and often has positive effects on their quality of life.

Eccentric viewing training and training to improve fixation stability are also closely connected to an improved ability to process visual impressions in the brain. Training in eccentric viewing can be used to prove that improving fixation stability (keeping the eye/eyes steady in the same direction) is positively linked to increased brain activity. (Rosengarth et al.,

2013). This can be seen in both areas of early visual cortexes and in areas significant to the vision process for recognizing objects and faces. It has also been found that when scanning using vision, a greater fixation stability ability is important for improved performance. Then as well you can see increased activity in the visual cortexes of the brain." (Plank et al., 2013).

In other words, something happens in the brain when you learn to see with another part of the retina. The areas of the visual cortexes in the brain that previously received signals from the macula develop increased activity after training. This can be seen with MRI (magnetic resonance imaging).

It is very interesting because it shows that the brain changes and

adapts!

It is not really known exactly how it does this, but research has determined that the vision area of the brain is activated - even more if you can develop good fixation stability and thereby maintain a steady gaze in a single direction for a prolonged period of time.

## PRACTICAL TRAINING OF VIEWING FIXATION

The next page illustrates how much of the visual field is lost with a central scotoma in both eyes.

*"Preferred Retinal Locus is best when central vision is gone".*



CENTRAL SCOTOMA

*The photo is taken from the video with the same name as this book - LOOK FORWARD.*

*TRAIN ECCENTRIC VIEWING USING TESTS AND EXERCISES ON THE WEBSITE*

WWW.EXFIX.SE

# DON'T FORGET TO KEEP YOUR BODY IN SHAPE

*Those of us with a visual impairment tend to have problems related to too little exercise and too much time sitting. Over time, this increases the risk of stiffness and pain. It is easy to end up sitting in the same position all the time when looking at our mobile screen.*

Those of us with a visual impairment in particular tend to draw our shoulders up towards our ears when we should really be sitting relaxed and lift the book or newspaper with our hands. We also sit for long periods looking at our mobile phones, which we hold in a particular position. This is something most people do – sit hunched over with the screen at their chest. It is really not a restful or good posture. Those of us with vision problems also tend to spend a lot of time in front of the computer. This is often our tool, our entertainment, and our way of making contact with the outside world.

Sitting in a forward-hunched position creates tension in the back, neck and shoulders. After a while, the muscles become stiff and tired. This makes it particularly important for us to train strength, mobility and balance, and to stretch properly.

The first tip is to stand up, shake your upper body, roll your shoulders a few times, and stretch your neck. All it takes is a few minutes every now and then.

Do not sit in the same position for too long.

Take a brisk walk of at least 30 minutes each day. This

can be divided into several shorter walks.

- Work on your posture – straight back, relaxed shoulders.
- Stretch and do mobility exercises three times a week.
- If you can, do strength training two-three times a week.
- If you find it hard to fit in this exercise yourself, find a physiotherapist or hire a personal trainer to create a programme specifically for you.

## EXERCISE TIPS

Examples of physical activity that meet these recommendations are:

A 30-minute brisk walk five times a week, or a 20-30-minute run 3 days a week, or a combination of these.

Further health effects can be achieved by increasing the amount of physical activity. This can be done by increasing the intensity, increasing the number of minutes a week, or both.

Physical activity that strengthens muscles should be done at least twice a week for several of the body's major muscle groups.



# EXERCISE TIPS FROM JENS -



## *POOR POSTURE*

*Because we sit so much, the muscles in our back can be weak. The chest muscles then take over and we end up with our shoulders rotated forwards. This results in a slouched posture and a forward head posture.*



## *GOOD POSTURE*

*A good posture that produces better flexibility looks like this:  
Feet pointed straight ahead.  
Hips pressed forwards and back flat (no inward curve) by tensing your inner abdominal muscles.  
Chest pushed forwards, shoulders straight.  
Head is kept straight above the body.  
Gaze is directed forwards.*



## *PRACTICE GOOD POSTURE*

*As easily as rolling your shoulders rearwards every now and then, you can hold your hands out at your side and then turn them outwards. Take a deep breath and imagine extending your neck up through your head. This is how you guide your body into a good posture.*



## *BACK EXERCISE - 1*

*Attach a rubber resistance band (available in sporting goods shops) to a door handle, pull your arms back close to your body and pull your shoulder blades together.*

# KEEP YOUR BODY IN SHAPE



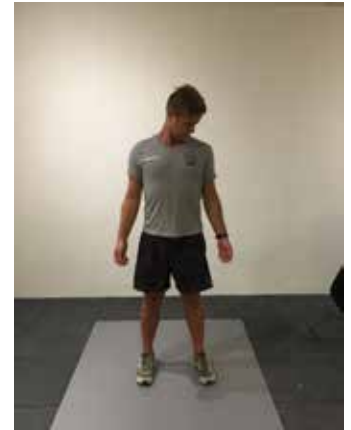
*BACK EXERCISE - 2*  
*Repeat 3x10. At the last repetition, keep your arms pulled back for ten seconds.*



*CHEST STRETCH*  
*An extremely simple and effective exercise is to put your hands behind your back and press your chest muscles forwards.*



*NECK STRETCH - 1*  
*Stretching your neck is particularly important when you have been sitting in front of a screen for a long time. Relax your shoulders, bend one ear towards your shoulder and hold for about 20 seconds. Repeat this on the other side. Repeat daily.*



*NECK STRETCH - 2*  
*Finish the neck stretch by guiding your nose toward your armpit. Hold for 20 seconds. Repeat this on the other side. Repeat this exercise daily.*



# SOME SIMPLE TRICKS TO IMPROVE YOUR LIGHTING ENVIRONMENT

*We all depend on good lighting to feel good. With simple means, we can create a good lighting environment that makes it easier to find our things and that works naturally - inside and out, in the kitchen or at the workplace, whether at the stairs or in the wardrobe.*

What is good lighting? Basically, it is a matter of investigating what needs you have at both the general level and the detail level, and then making sure the right light ends up at the right place.

This creates balanced light conditions that foster both function and well-being.

Here, we focus on three main principles that together create a good whole in a room, namely task lighting, ambient lighting and orientation lighting.

**Task lighting** is concentrated light that produces a relatively high lighting level in a limited area. This can include a desk lamp, a reading lamp next to an armchair, spotlights directed at bookshelves, or under-counter lighting in the kitchen.

**Ambient lighting** supplements task lighting to create good contrasts. Examples include ceiling lamps,

floor lamps with uplighting and spotlights directed towards walls.

**Orientation lighting** guides us when we move about. It can be achieved by lighting target spots like doors and illuminating stairs and other objects. It tells me where I am and where I'm going.

## A PERCEIVED PROBLEM IS AN ACTUAL PROBLEM

As part of the Leber X Project, we reviewed a number of authentic home environments of people with LHON and were able to improve functions and increase well-being using very simple means. The task required us to first and foremost listen to the problems the people in question perceived and then work together to find solutions.

We almost exclusively used products available on

the consumer market, without costing a lot of money. They proved to work surprisingly well. But, it requires you to make careful choices when buying - many lamps in the shops are substandard, particularly in relation to glare.

For individuals with LHON in particular, it is important to:

- Create clear contrasts through a balanced relationship between task and ambient lighting.
- Due to a high sensitivity to glare - make sure the lights are screened, they light where they should, and connect a dimmer.
- A short reading distance requires a lamp that can be positioned close without being in the way. Modern LED technology makes this both possible and simple.

- Find out where problems exist. The problem can often be solved by moving the lamps or making a small change - like screening, another light source, or using a dimmer.

Last, but not least - each individual has their own unique lighting needs, regardless of their level of sight. Planning lighting requires a great deal of sensitivity to and respect for how an environment is perceived. This is especially true in the home, where well-being and style are as important as function.

*Here are a number of solutions to different problems. Examples that can spark your own ideas!*



*Problem: Finding a wall switch when the contrast is low.*



*Solution: Contrasting switch/dimmer.*



*Problem: The living room seems dark, making it difficult to read. Both ambient lighting and task lighting are needed.*



*Solution: Floor lamp with large textile shade produces non-glaring ambient lighting while the floor lamp at the sofa provides targeted reading light. Both lamps can be dimmed between mood lighting and full lighting.*





*Problem: The living room where the family spends much of its time is dark, does not have good contrast, and feels unclear.*



*Solution: A spotlight with clamp attachment provides targeted light in the bookcase. Lighting a vertical surface in the direction of gaze also makes the room feel generally brighter and clearer.*



*Problem: One single, strong, circular spotlight in the bookcase. It only lights itself; the surroundings are perceived as comparatively dark.*



*Solution: A worktop light along the bottom edge of the top cabinet. It produces raking light along the shelf and all the way down to the floor, where Hampus usually plays with the children.*



*Problem: Anton has a lot of trouble finding clothes in his wardrobe. He blocks the light from the ceiling lamp when he is standing in front of his clothes.*



*Solution: Wardrobe lighting in the form of an LED strip has been installed. The clothes are now easy to see. The light is switched on and off when Anton opens and closes the wardrobe door.*





*Problem: Too much contrast between the computer screen and the dark surroundings.*



*Solution: Table lamp that can be directed as needed. Here towards the wall behind the monitor for good contrast...*



*... and here in front of the monitor as working light.*



*Anton did not use this lamp because he felt it caused too much glare. With a simple plug-in dimmer, he can now adjust the brightness to a comfortable level.*



*Problem: Lars has trouble doing kitchen work due to uneven lighting (dark work surface in the centre). Also, the light cannot be dimmed and must be switched off completely during the meal.*



*New under-cabinet fixture that can be dimmed so the light can be adjusted between full working lighting and mood lighting.*





*Problem: No working lighting. There is a spotlight in the ceiling, but it does not provide good working lighting since Anton casts a shadow on the counter.*



*Solution: An LED strip provides good working lighting. Mounted on the wall because there is no cabinet.*



*Problem: This lamp type is basically good. It provides ambient lighting up to towards the ceiling and reading light downwards, creating a good light balance. However, Agneta felt that the illuminating spot in the top caused glare.*



*Solution: A piece of regular aluminium foil was placed in the fixture to reduce light penetration through the lower lens.*



*Problem: The lamp type and position are generally well thought-out - a camp fire to gather around for chats. However, the light source found in it is too strong and not dimmable.*

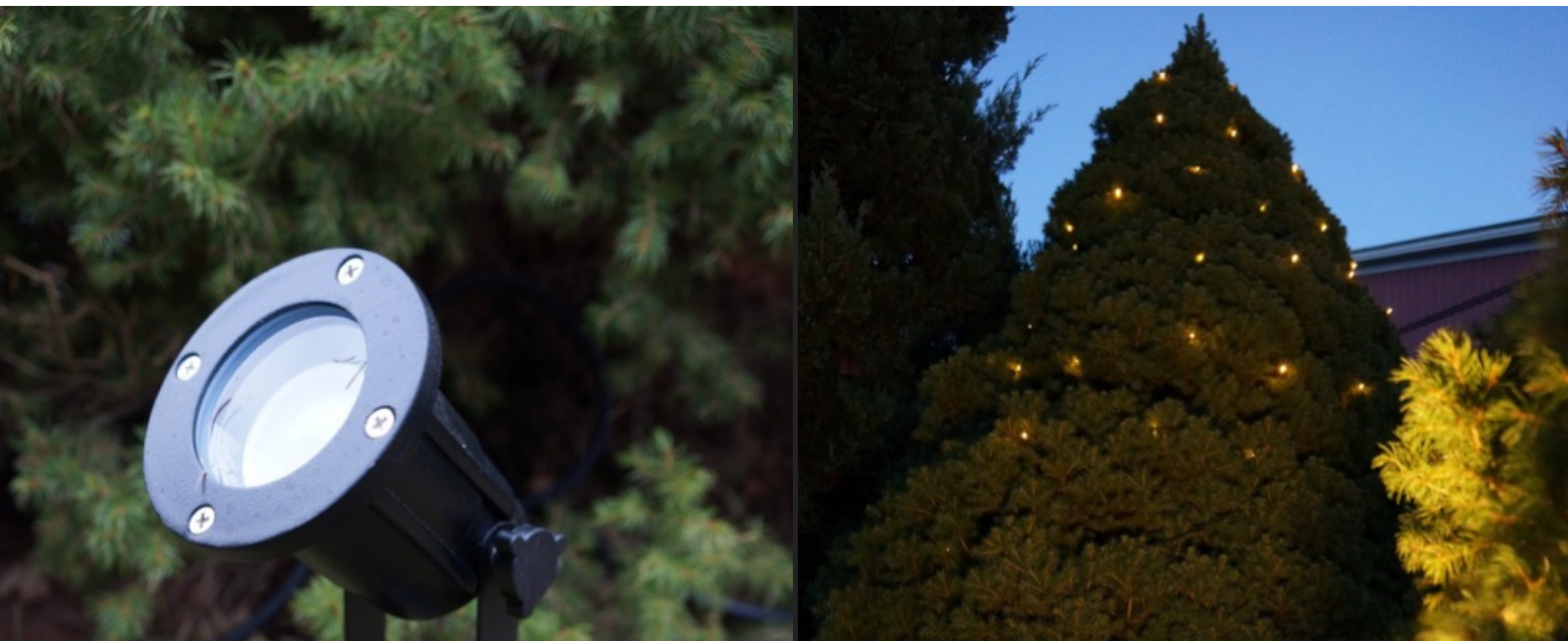


*Solution: A dimmable halogen light source and plug-in dimmer make it possible to adjust the light based on need.*





*Problem: The staircase lacks lighting; the steps are hard to distinguish. Solution: Small spotlights are mounted hidden behind the handrail. They shine down on the steps and provide clear contrast between the vertical and horizontal surfaces.*



*A dark and unclear outdoor environment becomes clearly distinguishable through spotlights that illuminate the bushes and guide visitors to the entrance of the house.*



Kalender



Bilder

Kamera



Videor



Väder



Kartor



Game Center



Klocka



Aktier



Påminnelser



Anteckningar



App Store

1



iB

# IT AIDS – A NATURAL PART OF DAY-TO-DAY LIFE

*When used correctly, IT aids are like an extension of your senses. They help many visually impaired orient themselves and communicate better with those around them. They have become a natural part of things in most situations, from taking a walk or going to the cinema to handling studies, trips abroad or qualified work.*

This is one way to summarize a workshop organized in September 2016 by the association Unga Synskadade in Stockholm. A panel of young IT enthusiasts with visual impairment discussed the benefits of IT aids and ranked some of the most useful apps.

The overall key word of the discussion was independence. Thanks to IT technology, even people with severe visual impairment can live a more independent life.

The smartphone has become almost an extension of the nervous system that can be adapted to suit your individual needs. There is now an overwhelming number of apps that interpret and explain what the visually impaired cannot see themselves.

For example, the internet and social media have become more accessible thanks to audio description of images. Other apps make it possible to easily navigate a web shop's assortment, and upon delivery of food products read off their EAN codes and use voice messages to be able to sort them and put them in the right spot in the fridge, freezer and kitchen cabinets. There are apps that provide audio description of a

film at the cinema or a TV show through the mobile phone. There are also DVDs with tracks for audio description. Others make it easy to perform tasks like booking a laundry time, paying pills, and keeping track of finances. And even though their features can be very advanced, the panel felt that mobile phones are amazingly easy to use.

There are now also computer games, for both PC and mobile phone, that do not require any sight. There is a plethora of programs for recording and editing music and audio files.

Of course, much of this can be done through a computer, like internet banking. But, what is really happening now is that so many services and features are moving to the mobile phone. It can be used for almost everything. And you always have it with you.

## **THE DEVELOPMENT HAS JUST BEGUN**

There are many apps that use GPS technology. They make it easier to orient oneself when moving about and travelling. And there also apps that make it easy



to search for and book travel in real time, see what platform the train will arrive on, what gate the flight leaves from, and so on.

Studies are made easier by the growing availability of audio books and speech synthesis - like reading PDF files aloud. The fact that libraries and entire education programmes are now digitalized makes them more accessible. There are still differences between cities and regions, but development is under way.

It has become commonplace for schools and institutions of higher learning to have digital learning platforms. They can be used, for example, for organizing distance courses, gathering course literature (both full texts and literature lists), turning in assignments via a website, attendance reports, schedules, video conferences and audio recordings from conferences and discussions, and chatting with other course participants, for example during a live video lecture. The learning platform gathers everything for studies into one place, usually a website but in some cases also optimized for smartphones and tablets.

The increasingly advanced technology also opens up more job opportunities for individuals with visual impairment. Even blind people can handle a growing number of jobs with the help of speech synthesis and Braille display in mobile phones and computers. It is becoming increasingly common for internal business systems to be adapted for the visually impaired, although they still can be perceived as fairly inaccessible. Here too, development is in progress.

The workshop participants were very excited about what the future will offer. In particular, they expected a growing number of special solutions for different types of disabilities to be integrated into society. The same way that Apple incorporated many help features right from the start in their first iPhone, accessibility is being planned in more and more in architecture, traffic systems, residential areas, workplaces, public transport systems, and more.

Incorporating accessibility right from the drawing board is called universal design. It can involve alternatives to stairs for wheelchair users, audio and light signals in all types of environments for people with impaired vision or hearing, tracks in pavements and platforms for those who use a white cane, and so on.

Demand for this is increasing in a general sense as well since even people without any impairment appreciate simpler and clearer features in all environments. For example, many sighted people use voice control in their mobile phones. Individuals without a driving licence are eagerly awaiting self-driving cars, and so on.

#### **WHERE CAN YOU FIND GOOD APPS AND AIDS?**

The discussion participants also shared tips on good apps and where to find useful information on what is available.

The most basic tip in regard to apps was to first and foremost explore your smartphone thoroughly. Modern phones often have more built-in help features than people think. Like enlarging text or inverting

colours. Other features can be used as aids, like using the camera to take a picture of an objecting and then enlarging it by zooming.

Information on the latest apps and aids can be found at places such as:

- [www.rnib.org.uk](http://www.rnib.org.uk) A British radio station with about the same focus as Radio SRF.
- [www.coolblindtech.com](http://www.coolblindtech.com) presents a wealth of technology news and is also a global community. It works actively to develop universal design.
- [www.applevis.com](http://www.applevis.com) is a similar community for users of Apple products.
- [www.tbteknik.se](http://www.tbteknik.se) is a Swedish website about technology and IT for the visually impaired.

The workshop participants also ranked a number of apps they particularly liked. Among the general apps, they considered Sweden's Radio Play, Facebook, Swish and Tågtaflan (which keeps tracks of all train departures throughout Sweden and which platform the trains leave from) to be the best. They are the most useful and the easiest to use. Thereafter came Messenger and Spotify, followed by the SL app, Res i Stockholm, and BankID.

Among the special apps for visually impaired users, the following were considered most useful:

- **BlindSquare**, orientate yourself on a map.
- **TapTapSee**, photograph an object and the app will then identify it and present it using speech synthesis.

- **KNFB Reader**, converts printed text to speech - documents, receipts, notes.
- **Movietalk**, audio description of films.
- **Be My Eyes**, live video with sighted volunteers who help the visually impaired with everything possible, like orientating themselves or reading instructions.
- **Legimus**, reads audio magazines and audio books; created by Myndigheten för tillgängliga medier [Authority for Available Media].
- **Seeing Assistant**, helps the visually impaired identify colours and light sources and interpret EAN or QR codes; can be used as an electronic loupe, etc.
- **Blind Bargains**, discounted goods and services for the visually impaired (including aids), news, magazines, etc.
- **Light Detector**, translates light to sound so that the blind can orientate themselves, know where a window is, etc.
- **A Blind Legend**, mobile action game based on sound instead of images.
- **Blindfold**, a series of games for the visually impaired: racing, bowling, hockey, basketball, bingo, farm, blackjack, etc.

## KNOWLEDGE AND PRACTICE

There are many opportunities, but it is important to become acquainted with them and practice using them whenever they are needed. A whole new technology - or IT rehabilitator - and an institution that can do all this and more needs to be developed as soon as possible.



YORS

HYREX  
KEDJEN

BARON

Dansk Design  
Produceret i EU

BARON - Den rigtige blander til den rigtige pris



# 10 TIPS FOR GETTING YOUR DREAM JOB

*Are you ready to start looking for a job? Great! Here is how you can increase your chances of getting a job that you enjoy. Everyone faces obstacles and difficulties when it comes to job hunting, and a visual impairment adds to this specific challenges that others do not have. For example - when should you let them know about your visual impairment and how should you handle the employer's concerns and questions during an interview?*

## **SUCCEEDING IN WORKING LIFE**

There are many things to consider when looking for a job. In interviews with individuals with visual impairment, we found four areas we think are important to be good at:

**KEYBOARD TECHNIQUE:** Learn to type on different keyboards using the touch method, where you do not look at the keys.

**IT/TECHNOLOGY:** Be aware of and master the special programs and adaptations for computers, notation aids and mobile phones that are available and constantly evolving.

**MOBILITY:** Develop a good ability to orientate yourself and move about independently, as well as a good ability to master the practical situations that arise in daily work.

**SOCIAL COMPETENCE:** Develop your ability to create relationships and work well with others. This includes being able to (and daring to) describe your situation

and suggest solutions that affect the working environment of both you and your co-workers.

## **TEN STEPS TO START YOUR JOB SEARCH**

Before you start, you should know that it is not easy for anyone to find a job, but when you achieve your goal it is well worth the effort!

How long it takes to find a job depends on factors such as:

- where you live and if you are able to move
- your knowledge and qualifications
- the availability of jobs in the labour market
- how much time you invest in your job search.

Ten good steps you can take to prepare are found on the next page.

### **1. DECIDE WHAT YOU WANT TO DO**

SRF's website ([www.srf.nu](http://www.srf.nu)) contains information about jobs that we know work for individuals with

visual impairment and what you need to be able to handle to work in the profession in question. You should also register with Arbetsförmedlingen to gain access to the support available there.

## **2. LEARN TO USE THE INTERNET**

There is a lot of information online, especially if you can speak English. You miss out on a world of opportunities if you cannot work online or even submit applications via the web. We recommend visiting [www.rnib.org](http://www.rnib.org) in the UK and [www.afb.org](http://www.afb.org) and [www.nfb.org](http://www.nfb.org) in the USA.

## **3. TRY NOT TO DO ALL YOUR JOB SEEKING ALONE**

Finding that first job can be tough and may take longer than you expected. So, to keep your spirits up and increase your chances, seek support in your job search. Take advantage of the experience, skills and networking capabilities found all around you. People you know - as well as Arbetsförmedlingen - can help you find resources, make contacts, stay optimistic, give you ideas, and encourage you to explore your options. Develop your network of contacts. Statistics show that who you know makes a huge difference in how long it takes to find the job you want. See page 55 about Arbetsförmedlingen's initiatives.

## **4. BE WILLING TO ACCEPT ANY JOB**

All job experience will help you find the job you want. If you can afford it, accept an internship or work as

a volunteer for a while to gain relevant work experience. This will also help you expand your network. The journey to your dream job can be long. That's how it is for most of us. Try to find a job that suits your skills and, if possible, your interests.

## **5. INTERVIEW SOMEONE WHO HAS THE TYPE OF JOB YOU WANT**

This can give you more knowledge about what it takes and what you should say to a potential employer when you come for an interview. Check around to see if there are persons with visual impairment who have a similar job. You can get some valuable tips from them.

## **6. CONTINUE DEVELOPING YOUR SKILLS**

Above all, good communication skills can make the difference between a job offer and no job offer. If there are courses available that you can take to increase your employability, take them. And work on your attitude - be as enthusiastic as you can. It really helps!

## **7. DO NOT SPEND TOO LONG IN YOUR STUDIES**

If you are going to school just to gain time because you cannot make a decision, this could be a complete waste of time, energy and money. Studies should be used to create a good foundation to stand on. You can always go back to school if you discover you need an additional degree or additional skills.

## **8. DON'T PUT ALL YOUR EGGS IN ONE BASKET**

Instead of searching for a job just one particular way, make use of all possibilities you can think of. Remember - the amount of time you invest in your job search affects how quickly and how successfully you find the job you want. Set aside a specific time each day for your job search.

## **9. ORGANIZE, ORGANIZE, ORGANIZE**

Make notes while considering what strengths you can offer an employer and what your dream job requires. You may want to use a folder or an electronic file to keep track of who you talk to, write down good advice and possible leads, etc. This will help you remember what you have done and will give you confidence in your job search. And will help you stay focused.

## **10. EXPLAIN YOUR VISUAL IMPAIRMENT USING POSITIVE TERMS**

It is important to find a good way to describe how you see and what consequences it has. Here are some things to think about:

- A. Write down your account. Go through it with family members, friends and people in the workplace. Repeat your account until you feel comfortable with it. Remember to also ask for feedback about your body language.
- B. Avoid going into too much detail when describing your visual impairment. You might find it highly

interesting, but the interviewer really only wants to know four things: Can you do the job safely and reliably? Will you be there? Can you do the job as well as - or better than - someone else? How will you do things a person with normal vision does visually, like reading, writing, using the computer, and so on? If you prepare good answers to these questions, you can significantly increase your chances of getting the job.

- C. Be positive about your knowledge and abilities. Give examples of assignments or jobs you have done. The more positive you are and the clearer you can convey your competence and prove that you can handle the job, the better. Even though you "happen to have a disability". The reverse is also true. The more you talk about your disability and its consequences, the greater the risk that you will give the potential employer negative associations.
- D. Be prepared for questions like: "How will you handle this task or this other task if you cannot see well?" If you have prepared good answers to standard questions, you can handle the interviews better and more easily.
- E. Bring your aids along to the interview. This way, you can show your potential employer that the computer will help you do the job. For example, you can demonstrate how you work with a computer.





# HIGHER-LEVEL STUDIES - BETTER JOB

*Getting an education from a college or university generally gives an individual with visual impairment the best chance of getting a good job and having a good life. Naturally, such individuals face more challenges and difficulties than others. Just like in all other situations. But you can do it. You have to make it work, and there are a number of aids available. Higher education institutions are also becoming more aware of what visually impaired students need.*

The following review of problems and solutions - of the responsibility of the school, the teachers and the students - is based on seminars conducted as part of the Leber X project with study-experienced individuals with LHON, questionnaire surveys conducted by Unga Synskadade, and experiences reported at Cambridge University in England.

Earning a degree for a knowledge-based profession is good training for working life. Society expects colleges and universities to offer equal conditions for people with and without disabilities. This means that visually impaired students can make clear demands for adapted study conditions. Not all higher education institutions offer the degree of adaptation expected. A visually impaired student may therefore need a hefty

dose of energy and willingness to take the initiative.

Study conditions can be divided into the following categories: the school's physical environment, how it treats its students, how instruction and exams are carried out, and to what degree the school accepts the student's aids. And, of course, there is the student's own level of ambition and social competence.

This is important. When you decide on an education programme, make sure you choose something you find interesting and that is realistic based on all of your conditions.

## **THE BEST ADVICE - BE OPEN ABOUT YOUR VISUAL IMPAIRMENT**

To be able to complete your studies in a somewhat normal amount of time, you must communicate the

fact that you have a visual impairment. Many make the mistake of trying to manage everything themselves. Others choose to use their aids more in their free time than in the classroom. They may be afraid of being labelled “handicapped”, don’t want to feel like a burden, or want to test their own capabilities.

Even if you can make it work, it’s at the price of a huge effort and almost guaranteed poorer results than if you had been open about what you deal with.

### **BETTER PHYSICAL ENVIRONMENT - BENEFITS EVERYONE**

You do not have to have a visual impairment to appreciate a clear physical environment that is easy to navigate. It should be easy to find your way to the right classrooms, which may be spread out over a number of floors, levels and buildings.

For a visually impaired student, it is particularly important to have, for example, some form of orientation lighting or guide lighting between buildings, contrast marking on the floor, ramps instead of stairs, hand-rails to follow, voice information in lifts, and so on. Many also call for tactile maps of the area - something offered by some colleges. If not, the vision centre can help with such.

Details in the physical environment can also play a big part in daily life. Paying the coffee vending machine by text message can be a daily source of irritation. Or finding your mail slot or checking out information on bulletin boards. All disruptions, even ones that are small and seemingly trivial, accumulate and steal a lot of energy.

### **RIGHT SUPPORT IN INSTRUCTION**

A visually impaired student needs more time to handle just about everything. It takes planning. One huge need of visually impaired students is for the course literature to be finalized early on and for them to be given enough time to have it recorded or in some other way adapted for use with aids.

Here, the teacher plays a central role. To provide proper instruction to everyone, the teacher needs some special training to be able to understand what a visually impaired person needs. More on the teacher’s role is found later in the text. But, a basic need is for the literature to be decided on early - so there is time to convert it to audio books, Braille books, tactile picture books, and e-textbooks before the start of the course. If this is not possible, it could severely delay the student’s entire course of studies. This could have financial consequences with a need for higher student loans and social consequences by the student being forced to change class multiple times.

More and more universities and colleges are now offering a mentor who can help visually impaired students with practically everything, such as navigating the campus, getting in touch with the school’s coordinator, and getting the support and aids they need, including planning their studies and thinking about what is most important to focus on in a course. Many mentors also arrange regular discussion meetings where students with some form of disability can exchange ideas and experiences.

Examples of pedagogical support and aids that universities and colleges are expected to offer nowadays include:

- Adapted exams, such as individual exams on the computer, oral presentation instead of written exam and vice versa, extended time to take an exam, and being able to sit in a separate room.
- Adapted schedule, such as part-time studies.
- Literature on adapted medium.
- Note-taking support. Someone who sits in on lessons and takes notes. This can often be a paid extra job for a fellow student.
- Some computer support. Many have good experiences with digital whiteboards.
- Extra guidance and, as mentioned, mentors and mentor groups.
- Language support: advice related to writing, particularly layout.
- Sign language interpretation.
- Speech-to-text.

### **TEACHER HAS GREAT RESPONSIBILITY**

It is of great importance that each teacher provides instruction in such a way that everyone can keep up. Teachers can therefore take courses to improve their understanding of the needs and conditions of students with different types of disabilities.

For visually impaired students, it may involve simple things like the teacher verbalizing what they are writing on the board (particularly when introducing new

terms - including how they are spelled) and saying the name of the person they want to answer a question instead of just pointing.

Some general advice to teachers who have one or more students with a visual impairment:

Especially at the start of the course, it may be difficult for these students to find their way around and get to the classroom on time. You must therefore be very clear with all practical information, especially when switching classrooms.

Keep in mind that these students generally need more time with everything, from keeping up with things during lessons to taking the actual examination.

Planning and looking ahead are key. Among other things, it involves reaching an agreement on how the teacher and student will communicate with one another. Bulletin boards and mail slots might not always work.

Sometimes an email or text message will work. In other cases, a person needs to seek out the student and let them know what is going on, for example if the classroom is changed at the last minute.

Students with special needs will probably contact the teacher before the start of the course to get a better understanding of the scope and requirements of the course. If they do not, then it makes it easier for everyone if the teacher reaches out to the student. Find out what needs each student feels they have.



Teachers need to familiarize themselves with the various computer aids the students may wish to use or that the school provides, and incorporate their use in the teaching as much as possible. More time may need to be given for note-taking. Or it may be necessary for the student to make notes using a computer or have an assistant take notes – or you could even consider recording the lecture.

It may take longer for visually impaired students to begin writing due to the extra time needed to read questions or instructions. The same applies to proof-reading written work and compiling a bibliography. Presentation requirements might not be met if the student is not given support in this.

Since time is a key issue, forward planning is required even for individual elements of the course. In addition to the course literature and a clear review of the syllabus, it is very helpful if the student can be given some lecture notes or any images that will be used during the lecture in advance. Most prefer to receive material in digital form.

Keep in mind that it is always important for a student to know if the level of errors in their work is unacceptable and to get help in finding alternative ways of improving accuracy.

During group work and seminars, other students may express concern that having a visually impaired member in their group may hinder them or create more work for everyone else. Such concern is often unfounded – but if it occurs it is an issue. It is then im-



portant to sit down with everyone involved to discuss how to handle the situation.

### **THE STUDENT ALSO HAS A RESPONSIBILITY**

As previously mentioned, it is important for an individual student with visual impairment to overcome any shyness or pride, and to as openly as possible tell their students and classmates exactly where things stand. It is only then that those around you can understand why you may need extra time and aid, especially when taking exams since other students could otherwise feel it is unfair. And they can understand why you sometimes have trouble recognizing people or finding a free seat in the lecture hall.

Colleges and universities have made varying progress when it comes to offering the aids and special considerations required. Even today, there are visually impaired students who are denied aids, accessible literature, or more time to take exams.

Many students have said that they were shown so little understanding that they were forced to change their study programme. Thus, the student sometimes has to struggle to get this help. Most of the time, you can get help from a guidance counsellor or a vision centre. You can also contact Myndigheten för tillgängliga medier (MTM), which can provide both literature and apps for download. One example is the free app Legimus for reading audio books aloud in your phone.

### **UNIVERSAL DESIGN**

An increasingly stable trend right now is the development of universal design. This means that the entire environment is made more accessible to all. That it can be used in several different ways. Not just schools, but also workplaces, modes of transportation, the entire urban environment.

For a visually impaired student, this can be anything from text being available through speech synthesis or the university environment being equipped with orientation lighting and different markings that make it easier to find your way around. Universal design is also about the availability of knowledge transfer, for example in the form of special introduction courses or distance learning opportunities.

Everyone benefits from this. Even individuals without any form of disability can benefit from a context that is easier to absorb and navigate through in a way that suits the individual.

*“Universal design involves making an environment accessible to all.”*



**LEBER**X  
ett synprojekt

# CONCLUSIONS FROM THE LEBER X PROJECT

During the first year of the Leber X Project, we collected information from individuals with LHON and their family members in a number of different ways. Among other things, we discussed how family members can support the individual with LHON - and what support family members need in the new situation since it is the entire family that is living with the disease.

Some participants with LHON said that you can live a really good life with a visual impairment. Not all normal-sighted people are happy. Whether you develop LHON or not is a lottery, just like life itself. If we learn to appreciate the good things in our lives and put our qualities and abilities to good use, we can all choose to feel like we have won the lottery.

One conclusion was that mothers and grandmothers need to let go of all the feelings of guilt so common in this group - regardless of the "child's" age. These feelings of guilt are psychologically stressful and hardly help the matter. On the contrary, they are more likely to complicate the situation. The family does not need another "victim".

Being the father of a child with LHON can also be psychologically stressful. This is particularly true if

the mother is wrestling with feelings of guilt because the father then has two people to comfort and support, which can be very stressful.

As the partner of someone who develops LHON as an adult, you also face a new set of circumstances. Life is turned upside down, especially in the beginning when your partner needs both comfort and help with daily routines like household chores or driving.

It is important to remember that a lot can be gained from listening to the experiences of others - and sharing your own. Many of the participants in the workshops indicated that they would be happy to serve as a contact or support person to other families.

## **SOME DIFFICULTIES AND SOLUTIONS**

Although they came from different parts of Sweden, most of the participants had come to the same conclusions in relation to difficulties and solutions.

*Not all normal-sighted  
people are happy...*

#### FOR EXAMPLE:

- Clear and open communication within the family solves a lot of problems. No one should feel reluctant to talk about how they are feeling. You cannot provide support if you do not know what the other person needs.
- Try to do things together and spend time together as usual to give yourself something else to think about.
- Reach out to other families in a similar situation. It provides both comfort and inspiration, as well as practical tips on everything from daily chores to contacts with various authorities.
- Family members should not try to guess the needs and wishes of the person with the visual impairment. Ask! And listen!
- Do not be afraid to seek outside help - from support persons, associations, organizations, or the municipality and county council. For example, SRF can help if you feel you have experienced discrimination at school or in the workplace.
- Take the strain off each other and try to find a new normal as early as possible.
- Humour can have a healing power. It can sometimes help to laugh a little at the new situation.
- Do not hover. Let the person who developed the visual impairment build their skills and learn to accept and live with their new circumstances. Practice new techniques!



# RESOURCES AT THE NATIONAL AND REGIONAL LEVEL

*In addition to LHON Eye Society, the organization responsible for this book, there are several bodies in Sweden that provide support and investigate diseases, and provide support in the school, rehabilitation, aids, and more.*

There are two centres in Sweden that investigate mitochondrial diseases – one at Karolinska University Hospital in Stockholm and the other at Sahlgrenska University Hospital in Gothenburg. Each has a team of paediatric and adult neurologists specialized in diagnosing and treating mitochondrial diseases, chemists and geneticists for biochemical and molecular-biological diagnostics, and pathologists with knowledge of morphologic diagnosis (how organisms are structured).

Persons suspected of having a mitochondrial disease should be referred to one of these teams for investigation, which includes biochemical diagnosis of the muscle sample immediately upon collection. With LHON, a blood sample is sometimes all it takes. The team can also provide treatment recommendations.

The eye clinics cooperate with vision centres or the like, where various visual aids can be tested.

The National Agency for Special Needs Education and Schools ([www.spsm.se](http://www.spsm.se)) is a state-owned nationwide authority. The authority's role is to provide special

education support to municipalities and other principals responsible for preschools, schools, care of school-aged children, independent schools under state supervision, and adult education. Via national resource centres with specific orientations, investigations and visits are conducted for individual children and adolescents, and information and training are provided to parents, teachers and other staff. One such centre is Resurscenter Syn, which is located in Stockholm and in Örebro.

Resurscenter Syn Stockholm, Rålambsvägen 32 B (visiting address), Box 19, 102 26 Stockholm, phone: 010-473 50 00, email: [rc.syn.stockholm@spsm.se](mailto:rc.syn.stockholm@spsm.se).

Resurscenter Syn Örebro, Eriksbergsgatan 3 (visiting address), 702 30 Örebro, Box 9024, 700 09 Örebro, phone: 010-473 50 00, email: [rc.syn.orebro@spsm.se](mailto:rc.syn.orebro@spsm.se).

A centre for rare diseases is under construction and development at the university hospitals. The aim is to increase knowledge in this area and improve the care of persons with rare diseases.

## RESOURCE PERSONS

Expert team at Karolinska University Hospital in Stockholm: Paediatric Neurology Karin Naess, Paediatric Clinic, Docent Göran Solders, Neurology Clinic, Docent Inger Nennesmo, Pathology and Cytology Laboratory, Karolinska University Hospital, Huddinge, 141 86 Stockholm, phone 08-585 800 00 plus Docent Ulrika Von Döbeln, Professor Nils-Göran Larsson and Doctor Martin Engvall, Centrum för Medfödda Metabola Sjukdomar [Centre for Congenital Metabolic Disorders], Karolinska University Hospital, Solna, 171 76 Stockholm, phone 08-517 700 00, Doctor Liu Ying, Eye Clinic, Södersjukhuset AB, 118 83 Stockholm, phone 08-616 10 00.

Expert team at Sahlgrenska University Hospital in Gothenburg: Professor Már Tulinius and Docent Niklas Darin, Drottning Silvias barn- och ungdomssjukhus, 416 85 Gothenburg, phone 031-343 40 00, Professor Anders Oldfors, Pathology Laboratory, Sahlgrenska University Hospital/Sahlgrenska, 413 45 41 Gothenburg, phone 031-342 10 00.

Link to website for resource persons, etc. within rare diseases: <http://www.nfsd.se>

LHON Eye Society has closest contact with Dr Martin Engvall, [martin.engvall@karolinska.se](mailto:martin.engvall@karolinska.se)

## SPECIAL-INTEREST ORGANIZATIONS

LHON Eye Society, email: [info@lhon.se](mailto:info@lhon.se), [www.lhon.se](http://www.lhon.se)  
Facebook page: [facebook.com/LHONEyeSociety/Discussion](https://facebook.com/LHONEyeSociety/Discussion) group on Facebook: [facebook.com/groups/LHON](https://facebook.com/groups/LHON).  
Sverige/ MitoSverige, email: [info@mitosverige.org](mailto:info@mitosverige.org),

[www.mitosverige.org](http://www.mitosverige.org)

Rare Diseases Sweden, Sturegatan 4 A (visiting address), Box 1386, 172 27 Sundbyberg, phone: 08-764 49 99, email: [info@sallsyntadiagnoser.se](mailto:info@sallsyntadiagnoser.se), [www.sallsyntadiagnoser.se](http://www.sallsyntadiagnoser.se)

The organization works for people living with rare diseases and various disabilities.

Swedish Association of the Visually Impaired (SRF), Sandsborgsvägen 52, (visiting address) 122 88 Enskede, phone: 08-39 90 00, e-mail [info@srf.nu](mailto:info@srf.nu), [www.srf.nu](http://www.srf.nu)

Riksorganisationen Unga Synskadade, Sandsborgsvägen 44, 122 33 Enskede, phone 08-39 92 85, email [info@ungasynskadade.se](mailto:info@ungasynskadade.se), [www.ungasynskadade.se](http://www.ungasynskadade.se).

In the United States, there is the United Mitochondrial Disease Foundation (UMDF), email [info@umdf.org](mailto:info@umdf.org), [www.umdf.org](http://www.umdf.org).

The Orphanet database collects information about special-interest organizations, particularly in Europe, [www.orpha.net](http://www.orpha.net), keywords leber hereditary optic neuropathy.

The Swedish National Agency for Rare Diseases (NFSO), which provides information on rare diseases on behalf of the Swedish National Board of Health and Welfare, has a calendar on its website with current courses, seminars and conferences related to unusual/rare diagnoses, [www.nfsd.se](http://www.nfsd.se).

LHON.org is an American international network.

# **FIVE GOOD THOUGHTS**

- 1. DON'T FEEL ASHAMED - EVER!**
- 2. DON'T WORRY ABOUT MAKING MISTAKES - EVERYONE DOES!**
- 3. ASK FOR HELP WHERE AND WHEN YOU NEED IT!**
- 4. CHALLENGE YOURSELF!**
- 5. BE POSITIVE AND OPTIMISTIC!**



*Don't forget to visit*

**www.lhon.se**

*where you can read the book with aids. There are also videos and other supplementary information.*

*For example, one of the videos shows how you can train your eccentric viewing ability, which is also explained at*

**www.exfix.se**

*The information on the website is updated continually as the society's operations grow and medical knowledge increases.*

## LOOK FORWARD - LIVING A GOOD LIFE WITH A VISUAL IMPAIRMENT

LHON is a rare hereditary disease that primarily affects the optic nerve. The disease creates a central vision loss. In simple terms, you have poor vision straight ahead. Most who are diagnosed with the disease ask themselves: What do I do now to look forward? That is exactly what this book is about.

LOOK FORWARD is a guide for individuals with a visual impairment – and their family and other loved ones. The book came into being on the initiative of the support organization LHON Eye Society. For this reason, it focuses on LHON. The book describes the causes and heredity of the disease. One section examines the state of research and theories about triggering factors. A number of individuals with LHON and their family members describe how they reacted to the diagnosis and what happened next.

The book contains concrete tips to help you live a rich and good life with a visual impairment, including tips on:

- rehabilitation
- vision training and the art of using peripheral vision
- different ways to master optical enlargement
- adapted lighting
- the art of effect organization of your home
- aids, with a focus on IT and apps
- physical exercise and training
- possibilities and rights in relation to studying and working

The book is part of the Leber X Project, which is funded with support from the Allmänna Arvsfonden [Swedish Inheritance Fund]. The material is also available and continuously updated on the internet at [www.lhon.se](http://www.lhon.se). The aim is to help individuals with LHON or other causes of visual impairment – and their family members – get back to living a life that, despite being different, is just as good as before. In Swedish, we like to define LHON as "Lev Här Och Nu", which means living in the here and now.

Medically, LHON stands for Leber's Hereditary Optic Neuropathy. It is a rare disease, which is precisely why this information is needed. Even healthcare professionals have very little knowledge about LHON.

Additional examples can be requested from [kontorservice@srf.nu](mailto:kontorservice@srf.nu).  
The Swedish version of the book is also available as an audio book.

**LOOK >> FORWARD** **LHON**  
EYE SOCIETY



ISBN 978-91-639-2643-3

