

# Uveitis (Engelse vertaling)

Uveitis is the collective term for all forms of inflammation of tissues in the eye. This umbrella term therefore includes all kinds of specific forms of inflammation in the eye.

In addition, other tissues of the eye and around the eye can also become inflamed. Examples include inflammation of the hard sclera (the white part of the eye), the cornea (the transparent part), the eye muscles, the optic nerve and the other structures in the eye socket. These types of inflammation do not fall into the category of uveitis, but uveitis specialists are often involved in the treatment of these diseases.

## Many different forms of uveitis

Uveitis comes in many varieties.

- Part of the eye infections is caused by a pathogen such as a virus, bacteria or parasite. In such cases it is often necessary to use antiviral drugs agents, antibiotics or antiparasitic agents as a treatment.
- In the majority of uveitides, we think that your own immune system is the main driver of the inflammatory process. In those cases immune-suppressing drugs and eyedrops are the main basis of therapy.
- Some forms of uveitis are very typical and manifest in a narrowly defined way. Examples of such very typical forms of uveitis are birdshot uveitis and VKH uveitis.
- In other patients with uveitis, we find evidence that the eye disease is associated with abnormalities elsewhere in the body or with a specific genetic susceptibility. In such cases, other organs are sometimes also affected. In these forms of uveitis, the inflammatory disease in the eye can vary greatly from patient to patient. Examples of this type of uveitis are uveitis in sarcoidosis and uveitis associated with the genetic profile called HLA-B27.
- In about half of the patients, however, we do not find any abnormalities that fit exactly with one of the specific syndromes. In such cases, we use the place within the eye where the inflammatory activity is greatest as a guideline in classifying the form of uveitis.

To classify the non-specific variants of uveitis, ophthalmologists use the following broad regions of the eye:

- The front (anterior side). This part of the eye contains the inner lining of the cornea (the endothelium); the anterior chamber (the space between the cornea and iris); the iris (the iris, which gives the eye color) and the ciliary body (the tissue behind the iris that makes eye fluid

that keeps the eye pressurized). Inflammation in the anterior part is collectively referred to as 'anterior uveitis'.

- The middle part (intermediate part). In this part, the inner lining of the eyeball is called the pars plana. Inflammation in this part is called 'uveitis intermedia' or 'intermediate uveitis'.
- The rear part. In the back of the eyeball are the retina, the optic nerve (nervus opticus) and the vitreous body (corpus vitreum). Inflammation in this part is called posterior uveitis.
- The entire eye. If the inflammation is active in both the front and the back, this is called 'panuveitis'.

## **Additional examination in uveitis**

In addition to the examination in the office, the ophthalmologist will in some cases also order additional examinations. In the event of repeated eye infections or very serious eye infections, it is often necessary to order blood tests, take a lung X-ray and possibly even have a little fluid examined from the eye.

## **No diagnosis is also a diagnosis**

In about half of the cases of uveitis, the ophthalmologist does not find any abnormalities that could explain why this person develops an eye infection, despite doing blood tests and other additional tests. This can sometimes help to better predict how the eye disease will respond to treatment and how the eye disease will behave in the longer term.

## **Often a chronic disease**

Many of the more severe forms of uveitis tend to flare up years after the initial episode. In some patients, the uveitis even remains dormant despite treatment. Because of this nature of the disease, people with uveitis often require long-term ophthalmic care.

## **Cornerstone of treatment: prednisone**

Because in many cases of uveitis the immune system is highly activated and thus impairs the function of the eye, it is often necessary to inhibit the immune cells. The most commonly used and most effective immune suppressant we have is prednisone. Ophthalmologists use prednisone not only in tablet form, but also as eye drops and as a liquid that is injected under the eye mucosa and is slowly released from there and inhibits eye inflammation.

## **Tapering off prednisone, replacing with another immunosuppressant**

Prednisone is a drug that has been used for many years and is therefore very safe. Nevertheless, prednisone has a number of adverse side effects, particularly with long-term use. Therefore, it is often necessary to gradually reduce the dose of prednisone and start another immune suppressant at the same time.

These are drugs known as DMARDs: disease-modifying anti-rheumatic drugs. These drugs are less able to suppress the active disease, but they can prevent the immune system from becoming overactive again and causing the disease to flare up.

Examples of DMARDs commonly used by ophthalmologists are methotrexate, azathioprine and cellcept. These are usually tablets or capsules.

In addition to the DMARDs, some immunosuppressant drugs are also available that must be administered by infusion or injection. The best-known example of such a drug called a 'biological' is adalimumab.

## **Limiting collateral damage/consequential damage**

The main goals of the treatment are to calm down the inflammatory process as quickly as possible to prevent permanent damage to the eye and to prevent the inflammatory disease from repeating. When treating uveitis, it is always necessary to find a balance between the intensity of the treatment and the expected benefit for you as a patient. Because many of the medicines used can also have side effects, your ophthalmologist will always make this assessment with you. It is also important to consider any negative effects that the medications used may have in the long term. For example, prednisone-based drugs can lead to a higher eye pressure and other immunosuppressive drugs usually lead to an increased susceptibility to infections and a slightly increased risk of developing precancerous conditions.

## **Relationship with complaints in other parts of the body**

Because eye infections are sometimes associated with changes in the immune system that can also be expressed in other parts of the body, your ophthalmologist will often ask additional questions about complaints in other parts of your body. Examples include non-specific complaints of the lungs, intestines and skin, but also specific autoimmune diseases such as rheumatoid arthritis, vasculitis and Crohn's disease.

If patients have such physical complaints, the ophthalmologist may refer them for further examination by, for example, a rheumatologist, gastroenterologist or dermatologist.

In people who are already known to have an expression of an autoimmune disease elsewhere in the body, the ophthalmologist will contact the practitioner in question to coordinate the immune-suppressing treatment.

## **Long-term**

For a large proportion of people who experience an attack of uveitis at some point in their life, this is a one-off event. Depending on the type of eye infection and the way in which the disease responds to treatment, parts of the eye may or may not be damaged.

Because the course of this type of inflammation differs so much from person to person, it is not possible to make general statements about the long-term consequences of uveitis. In one person extensive damage occurs and vision is permanently disrupted, in another person the eye recovers completely in a short time.

In practice, the ophthalmologist therefore sees people who ultimately do not experience any discomfort from the inflammation, but there are also patients whose uveitis causes permanent damage to their eyesight despite maximum treatment. Permanent damage can occur, for example, due to damage to the retina due to the inflammation or because the inflammation or the treatment leads to high eye pressure, which then affects the optic nerve.

## **Cataract**

Long-term use of high doses of prednisone or drugs derived from it can accelerate the formation of opacities in the lens. This process of cataract formation is a natural process that usually leads to an annoying haze over the course of decades. In people who have to use many prednisone-like agents for uveitis, cataract formation can sometimes accelerate so much that it is necessary to perform cataract surgery at a relatively young age, in which the cloudy lens is replaced by a clear plastic lens.

## **New treatment methods and research**

Research into new treatment methods for uveitis has benefited in recent years from the new drugs that have entered the market for the treatment of more common autoimmune diseases, such as rheumatoid arthritis. In addition, fundamental immunological research provides a better understanding of these types of diseases. As an academic ophthalmology department, we are regularly involved in both fundamental and clinically applied research and in this context sometimes ask patients if they would like to participate in this, for example by donating a blood sample. For all medical research that goes beyond normal clinical care, patients must give permission in advance for that specific case.

## **Practical information**

### **Alarm symptoms**

Uveitis is a disease that can flare up suddenly. We advise patients to always contact a doctor in case of acute changes in vision (for example, reduced vision, seeing dots, distorted vision); in case of acute pain in the eyes (behind, above or on the eye or pain when moving); with sudden redness of the whites of the eyes and with other sudden changes for which there is no good explanation.

We advise patients who are not under the care of an ophthalmologist or who have not been to an ophthalmologist for more than a year to consult with their general practitioner. If the GP deems it necessary, those people can urgently refer to an ophthalmologist.

We advise patients who are already under the supervision of one of our ophthalmologists to contact the Ophthalmology outpatient clinic for consultation. In the event of acute complaints during the weekend, you can also consult the ophthalmologist on duty by telephone.

### **Flare up on vacation**

Patients who have previously experienced a flare of uveitis often recognize the symptoms of new disease activity. The ophthalmologist often already makes agreements with these patients about how to act in the unlikely event of a flare-up of uveitis, for example during a holiday. In general, we advise patients to use the immune-suppressing drops twice as often per day and to consult an ophthalmologist within a few days.

### **Eye pressure checks**

Because prednisone-derived drugs can increase the eye pressure, it is sometimes necessary to monitor the eye pressure. Sometimes this is done at the outpatient clinic, but it is often also possible to make an appointment with the ophthalmologist to have the eye pressure checked at the local optician and to contact your ophthalmologist in the event that an agreed value is exceeded.

### **Side effects of the medication**

All medication used can cause side effects, but fortunately only a small proportion of people suffer from this. Side effects vary by drug. In general, we advise patients who experience a side effect of a medication to talk to their doctor. It is important to estimate how serious the side effect is. In the event of severe symptoms such as acute redness of the skin, shortness of breath, fainting or vomiting, it is advisable to immediately stop taking the medication and immediately consult by telephone.

In case of mild side effects such as a bloated feeling or mild dysregulation of bowel movements, people can continue to use the medicines and consultation does not have to take place immediately.

### **Pregnancy, breastfeeding and fertility**

We advise patients with uveitis who are pregnant, breastfeeding or want to become pregnant (including men!) to always discuss this with their doctor. Some medicines may not be used in those cases because they can cause damage to the unborn fetus and/or the child.

### **Heredity**

In principle, uveitis is not a directly hereditary disease. However, genes can play a role in the activity of the immune system. In some families, several family members have an immune system that is more prone to developing autoimmune diseases. That is why the ophthalmologist sometimes asks whether you, as a uveitis patient, have parents or other family members with conditions such as rheumatism.

### **Mood**

The mood of patients with uveitis can be disrupted by both the disease and the medications used. On the one hand, the visual impairment, the need to visit the hospital and the often chronic nature of the disease can lead to stress, gloom and uncertainty. On the other hand, medications, especially prednisone, can lead to chemical dysregulation of mood. We advise patients to always report these kinds of mood changes to their doctor. If necessary, the GP can check whether psychological help is useful.

### **Work and education and help with visual impairment**

Because good vision is a requirement for almost all professions and education, patients with uveitis often experience problems in these areas. For specific questions in the field of work, we advise people to contact your occupational health doctor. In some cases it may be useful to try to maximize the use of visual aids. The ophthalmologist can help you make an appointment at one of the institutions that deal with visual rehabilitation, such as Low Vision/Ergra and Bartimeus.

### **Finally**

If you have any questions after reading this leaflet or after talking to your doctor, write them down and bring them with you to your next visit. If you have questions that cannot wait, please contact the outpatient clinic.

## Contact details ophthalmology outpatient clinic

<u>Location</u>		<u>Telephone (during office hours)</u>	<u>Telephone (outside office hours)</u>	<u>E-mail address</u>
VUmc	Front desk R, 2nd floor	020 444 1170	020 444 4444	oogheelkunde@amsterdamumc.nl (non-urgent matters)
AMC	Front desk A2, 2nd floor	020 566 3100	020 566 9111	idem
Surgery planning Amsterdam UMC		020 444 1170	n/a	oogoperatie@amsterdamumc.nl (non-urgent matters)
<a href="http://www.vumc.nl/oogheelkunde">www.vumc.nl/oogheelkunde</a>				