# Coats disease

## What is Coats disease?

Coats disease, also known as exudative retinitis, is a very rare eye condition that affects the smaller blood vessels (capillaries) found in the retina. The retina is the light sensitive layer that lines the inside of the back of your eye. Coats disease can make these blood vessels weak and grow incorrectly, causing them to leak fluid and blood under the retina. This means that the cells of the retina can’t work properly, and this can cause sight to be affected.

Coats disease affects males more than females. It’s usually diagnosed by the time a person is 20, but in most cases, it’s first detected in childhood before the age of 10. Adults can also be affected, and some people, often middle-aged males, can have a milder form of the condition.

Coats disease only affects the health of the eye so people with the condition are otherwise generally healthy.

## How does the eye work?

Light enters our eyes and is focused onto the retina lining the back of our eye. The retina is made up of light-sensitive cells that convert light into electrical impulses which then travel along the optic nerve to our brain. Our brain processes these signals so that we can “see” the world around us. When you are looking directly at something such as words, photos or the television, light is focused onto a tiny, highly specialised area of your retina called the macula, which enables you to see fine detail and gives you your central vision. The macula also gives you much of your ability to see colours. The other areas of your retina give you peripheral vision. This is your sight above, below and to the sides of your central vision when looking straight ahead.

For the retina to work properly, it requires a good blood supply. This is provided by a network of blood vessels on the retinal surface and by a layer of blood vessels underneath the retina.

The following diagram shows the cross-section of an eye. From the front to the back of the eye, it is labelled:cornea, pupil, iris, lens, optic nerve, macula, retina and choroid.



## What causes Coats disease?

The cause of Coats disease isn't fully known (the medical term for an unknown cause is idiopathic) and there are no known risk factors that make it more likely to develop.

It has been suggested that for some people, Coats disease may be related to a mutation (fault) in the NDP gene which develops after conception, meaning that the condition has happened by chance and was not inherited from their parents.

## Is Coats disease inherited within families?

Coats disease is not an inherited condition,meaning that it is not passed on within families. As the condition is rare, it’s extremely unlikely for it to affect more than one child or person in the same family. This means that brothers and sisters of a child who has Coats disease are not likely to have the condition too.

## How does Coats disease affect the eye?

Coats disease affects the smaller blood vessels (capillaries) in the retina. Retinal capillaries are important in supplying the retina with blood which carries nutrients and oxygen to its cells, so that they work correctly. The cells of the retina need to remain healthy for you to be able to see clearly.

Coats disease causes retinal capillaries to develop incorrectly. They become wider (dilated) and twisted, which makes them more noticeable when the inside of the eye is examined. The medical term for these blood vessel changes is telangiectasia.

As well as becoming dilated, the retinal capillaries also become weak and leaky. This causes some of the fluid from the blood to leak out of the blood vessels and into the retina. This fluid builds up in the retina and causes it to become waterlogged (swollen). The medical term for this swelling is oedema. Eventually, proteins and lipids (fats) leak out of the blood vessels and build up underneath the retina. These are known medically as exudates, and they cause a yellow appearance in the affected area of the retina. Retinal exudates can be seen by an optometrist (optician) or ophthalmologist (hospital eye doctor) when they examine the eye.

Where there are areas of exudates and telangiectasia, the retina won’t be able to work properly. This in turn, may affect your sight. There are five different stages that describe how Coats disease can progress. These stages relate to how much the retinal capillaries and retinal tissue have been affected.

### The five stages of Coats disease:

**Stage 1**: Telangiectasia (dilated and twisted capillaries) which cause minimal change to the retina and to vision.

**Stage 2**: Telangiectasia and exudates which cause increased changes to the retina and, if uncontrolled, may lead to changes in vision.

**Stage 3**: Large areas of swelling with retinal detachment (the retina comes away from the back of the eye). Vision is likely to be poor at this stage.

**Stage 4**: Complete retinal detachment and glaucoma (raised pressure in the eye). These complications can be treated, but sight is still likely to be very poor.

**Stage 5**: The eye has no sight, and there is no treatment that can improve sight. In most cases the eye isn’t painful, but if pain develops, treatment may be needed for this, and to try to preserve the eye.

## How does Coats disease affect sight?

For most people, Coats disease only affects one eye, so many with the condition will have good sight in their other eye. It is a condition which can get worse, affecting more of the retina over time, but it can also stop getting worse on its own, so that not everyone progresses to stage 5. This means that the effect the condition has on someone’s sight will vary from person to person. It’s not known why the condition can affect some people more than others, making it very difficult to predict how it may progress over time.

How much a person’s vision is affected in the long term will depend on the age at which Coats disease is first diagnosed and how severe the condition is at that point. Generally, the outcomes for sight are poorer in younger children who are at a more advanced stage of the condition when they are first diagnosed. The retinal changes caused by Coats disease tend to progress more quickly in younger children, especially those under three years old. Those changes are more difficult to control and are more likely to lead to a greater long-term reduction in a younger child’s vision. When the condition develops in older children and young adults, it can cause much milder retinal changes which progress more slowly, so that their vision is less affected over time.

In the early stages of Coats disease, a person’s peripheral vision is most likely to be affected. This means that their awareness of things above, below or to the sides when looking straight ahead will not be as good in their affected eye.

If the condition progresses, and more of the retina is damaged, there will be a greater loss of vision in the affected eye. If Coats disease affects the macula, then a person’s central, detailed vision will be reduced.

In the advanced stages of Coats disease, more vision will have been lost as more of the retina has been affected. When a large area of retina has become swollen due to leaking fluid, it can cause the retina to detach from the back of the eye, which can cause further loss of sight.

## Will I notice any signs of Coats disease?

### What’s my child’s vision like?

Often, an eye examination is the only way to tell if your child has an eye condition. Children can adapt very quickly to a change in their vision and there can be very few signs that they have any problems, especially if only one eye is affected. Most children with Coats disease don’t have any symptoms – their eye doesn’t look unusual, it isn’t painful or red, and young children won’t usually be aware that there is a problem with their vision at all. Many children have their vision screened when they start school at the age of four or five. However, this does not happen in all areas of the country. Your child may be diagnosed after having difficulties with the vision screening or following an appointment with an optometrist. Older children may be able to tell you that the vision in one eye has got worse.

If you’re concerned about how well your child can see, or their vision is not screened at school, taking them to an optometrist will mean their eyes can be fully examined and their sight checked.

### What does my child’s eye look like?

Sometimes, you may notice that your child’s eye has an odd appearance in photographs, particularly where flash photography is used. Normally, flash photography causes a red-looking pupil but an eye with Coats disease may have a pupil that looks white or pale yellow instead, which some people describe as a glow in the eye. A white pupil is known medically as leukocoria. If you notice that your child has the appearance of leukocoria at any time, it’s important that their eyes are examined urgently by an optometrist or ophthalmologist, because as well as Coats disease, there are other serious eye conditions that can give a similar appearance which need to be ruled out.

### Does my child have a squint (strabismus)?

Some children who have Coats disease may develop a squint (medically known as strabismus). This is sometimes described as having a ‘turn in the eye’. Having a squint means that their affected eye doesn’t look in the same direction as the other eye. You can find more information about strabismus on our website **rnib.org.uk/eyehealth** or by calling our Helpline on **0303 123 9999**.

## How is Coats disease diagnosed?

You may detect that your child has a problem with their eye, or you may be alerted by vision screening at school. However, the only way to diagnose Coats disease is by examination of the back of the eye. If an optometrist has examined the back of your child’s eye during a routine sight test and believes they may have Coats disease, they will refer them to the eye hospital to be seen by an ophthalmologist.

### What happens in the hospital eye clinic?

At the hospital eye clinic, drops will be put into your child’s eyes to make their pupils larger. This allows the ophthalmologist to get a better view of the retina at the back of their eye. A bright light will be shone into their eyes so that a full examination can be carried out. Specialised photographs and scans of the back of the eye may also be taken.

Sometimes the ophthalmologist may want to perform another test known as a fluorescein angiogram. This is where a dye is injected into a blood vessel in the arm, which travels through the bloodstream to the eye. Alternatively, the dye may be given with fruit juice as a drink. When the dye reaches the eye, a series of photographs are taken which will show the blood vessels of the retina filling with the dye. From these photographs, the ophthalmologist will be able to see if any of the smaller capillaries are leaking and be able to diagnose Coats disease. In younger children, the fluorescein angiogram is usually done with the child asleep in the operating theatre.

## How is Coats disease treated?

The aim of treatment is to stabilise the changes that are already happening within the retina. In very occasional cases, treatment can improve vision, but in most cases the aim of treatment is to prevent vision from getting any worse.

In the early stages of Coats disease, vision may not be affected, and for some people, Coats disease won't develop any further than stage 1. If the condition is very mild, treatment may not be needed straight away, and the ophthalmologist may decide to monitor the eyes at regular appointments instead.

However, where there are a lot of changes to the retina, even when the vision remains good, the ophthalmologist may decide to carry out some treatment to help prevent a deterioration in sight.

The treatment offered will depend on what areas of the retina have been affected and by how much. Coats disease can be treated using a laser treatment, known as laser photocoagulation, and cryotherapy (freezing treatment). The aim of both these treatments is to help seal up the retinal capillaries to stop them from leaking further. Laser treatment may also be given to areas of the retina that are starved of oxygen because their normal blood supply has been affected. Treatment then aims to prevent further new abnormal blood vessels from developing in response to this lack of oxygen. Most children would have these treatments carried out under a general anaesthetic.

In more advanced stages of Coats disease, where the retinal capillaries have leaked a lot and large areas of the retina have already been affected, laser photocoagulation on its own may not be sufficient, and it may be necessary for retinal surgery to be considered. If the swelling has caused the retina to detach from the back of the eye, the treatment will be aimed at re-attaching the retina. The type of treatment required may vary depending upon how large the detachment is, and how long the retina has been detached for, and it may include surgery to re-attach the retina.

You can find more information about retinal detachment on our website **rnib.org.uk/eyehealth** or by calling our Helpline on **0303 123 9999**.

Occasionally, in certain circumstances, injections may be given into the eye. These treatments may include either anti-VEGF medication or steroids.

Anti-VEGF stands for anti-vascular endothelial growth factor. The long-term effects of anti-VEGF in childhood is still not fully known, however as part of the treatment of severe Coats disease, anti-VEGF may be offered to some people, as it can reduce the growth of new, leaky blood vessels and the bleeding they cause. Anti-VEGF treatment can reduce the risk of damage to the retina caused by these new vessels, which in turn can help to avoid further deterioration in sight.

Steroids can be given either around the eye or as an implant in the eye and are typically given in Stage 2 Coats disease, as they seem to be more effective in reducing the fluid leaking into the retina. They also last longer inside the eye than anti-VEGF drugs.

Anti-VEGF medication or steroid implants that are injected into the vitreous gel of the eye are known as intravitreal injections. The injection is given in a clean sterile room or an operating theatre to reduce the risk of infection. Before the injection, anaesthetic eye drops are instilled to make the eye numb, so the injection is not painful.

You can find more information about anti-VEGF treatment on our website **rnib.org.uk/eyehealth** or by calling our Helpline **0303 123 9999**.

## Does Coats disease lead to any other complications?

As well as retinal detachment, there may be other complications for someone with Coats disease which also require treatment, such as uveitis (inflammation inside the eye), the development of a cataract (clouding of the lens inside the eye) or glaucoma (raised pressure inside the eye which can damage the optic nerve). Cataract and glaucoma are also side effects that can develop from having steroid treatment. Although uveitis, cataract and glaucoma are treatable, this treatment will not improve the effect that retinal exudates have already had on central vision, as unfortunately, these changes cannot be reversed.

You can find more information about the treatment of uveitis, cataract and glaucoma on our website **rnib.org.uk/eyehealth** or by calling our Helpline on **0303 123 9999** to ask for our information on these eye conditions.

There are, unfortunately, some people for whom treatment is not successful because their eye doesn’t respond to any of the treatments they receive. In these cases, it’s possible that they may lose all the sight in their eye. Usually, their eye won’t become uncomfortable. However, very rarely, glaucoma develops which cannot be controlled, and this can cause the eye to become very painful. If the eye pressure remains too high and causes continual and intense pain, the ophthalmologist may suggest that removing the eye and replacing it with an artificial one is the best next step to take, but this is always a last resort and is not suggested very often.

**Most people with Coats disease do not experience this type of complication and so they will not need to have their eye removed.**

## How does someone manage with sight in only one eye?

Coats disease usually only affects one eye while the other eye may have good sight. When someone has good sight in one eye, and poor or no sight in the other, they have what is known as monocular vision.

Having monocular vision can make depth perception and hand-eye coordination more difficult. With some tasks, your child may appear clumsy and uncoordinated at first, for example when they throw and catch a ball. However, this generally improves as they get older and adapt to being monocular.

Children tend to adapt very well to using their better eye. This doesn’t mean that they will be overusing or damaging their better eye, as this adaptation is a natural process. It’s unusual for children with good vision in one eye to need additional support in their education. People with monocular vision are not considered partially sighted.

Even with good vision in their better eye, someone with monocular vision doesn't have as much peripheral vision as someone with two eyes. With time they will adapt to this without realising, by turning their head more to see things around them. Other people can help by sitting or approaching someone with monocular vision on the same side as their good eye, to make it easier for them to be seen.

It’s important for people who have good sight in only one eye to have regular eye tests with an optometrist so that the health of their good eye can be monitored. Your optometrist should let you know how often you need to have your eyes examined.

For people who have sight in one eye, it is a good idea to consider the use of protective eyewear or sports goggles for certain sports or activities to help prevent injury to the good eye. You can speak to a dispensing optician at your local optician’s practice for more advice about this. Dispensing opticians are qualified in the dispensing and fitting of spectacles and can give professional advice about suitable frames and lenses.

People with monocular vision are still able to drive a private vehicle (Group 1) so long as the vision in their better eye is unaffected by any other eye conditions and it meets the visual requirements for driving. However, monocular vision would mean you couldn't hold a heavy goods vehicle (HGV) or public service vehicle (PSV) licence (Group 2). Some professions, such as being a pilot, police officer or certain roles within the Armed Forces require a specific level of vision to be reached in both eyes. Keeping this in mind can help with planning a career choice for the future.

You can find more information about monocular vision on our website **rnib.org.uk/eyehealth** or by calling our Helpline on **0303 123 9999**.

## Coping

It’s completely natural to be upset when you or your child has been diagnosed with Coats disease and it’s normal to find yourself worrying about the future and how you or your child will manage with a change in vision. For a child, it may be difficult to predict to what extent their vision will be affected by Coats disease in the long term. Therefore, it’s important for them to attend their hospital eye clinic appointments as advised by their ophthalmologist, and to have regular eye examinations as advised by their optometrist, to ensure they have the greatest chance of developing the best vision they can. It’s also important as adults to have regular eye examinations too, with either an ophthalmologist (if still under the care of the hospital) or an optometrist at least every two years, or as recommended, to ensure that your eye health is monitored.

### Support from RNIB

It can sometimes be helpful to talk about how you’re feeling with someone outside your circle of friends or family. By calling our RNIB Helpline, you are no longer alone. We can support you at every step, putting you in touch with the advisors you need from any of our supportive teams. From support with education to advice on employment, from using assistive technology to understanding more about your eye condition, we are here to help. Our Counselling and Wellbeing team is also available to provide the emotional support you may need. Your GP or social worker may also find a counsellor for you if you feel this might help.

### The Eye Care Liaison Officer (ECLO)

You may think of further questions about Coats disease on your way home from a hospital appointment or in the days and weeks that follow a diagnosis. There is someone to turn to with these questions. Your eye clinic may have a sight loss advisor working alongside the doctors and nursing staff. This advisor may be known as either the Eye Care Liaison Officer (ECLO), the Vision Support Officer or the Early Intervention Support Officer, and they are on hand within your hospital to provide you with further practical and emotional support about your eye health. To find out if your hospital eye clinic has an ECLO, you can search within the RNIB Sightline Directory: **sightlinedirectory.org.uk**

Alternatively, you can call our Helpline on **0303 123 9999** to speak to our advisors within our **Eye Health Information Service**, as they would be happy to discuss any questions you may have.

## Sources of support

### Helpline

If you have questions about anything you’ve read in this factsheet, or just want to speak further to someone about Coats disease, please get in touch with us. It doesn’t matter if you or your child has just been diagnosed with Coats disease or you’ve known about it for a while; we’re here to support you at every step.

Whether you want to know more about your eye condition, buy a product from our shop, join our library, find out about possible benefit entitlements, be put in touch with a trained counsellor, or make a general enquiry, we’re only a call away.

Call our Helpline on **0303 123 9999**, we’re ready to answer your call Monday to Friday 8am – 8pm and Saturday 9am – 1pm. You can also email us at **helpline@rnib.org.uk**. You can also say, “**Alexa, call RNIB Helpline**” to an Alexa-enabled device.

You can also get in touch by post or by visiting our website:

**RNIB**

105 Judd Street

London WC1H 9NE

**rnib.org.uk**

### The Sight Advice FAQ

The Sight Advice FAQ answers questions about living with sight loss, eye health or being newly diagnosed with a sight condition. It is produced by RNIB in partnership with other sight loss organisations. **sightadvicefaq.org.uk**

**Connect with others**

You can meet or connect with others who are blind or partially sighted online, by phone or in your community to share interests, experiences and support for each other. From book clubs and social groups to sport and volunteering, our friendly, helpful and knowledgeable team can link you up with opportunities to suit you. Visit **rnib.org.uk/connect** or call **0303 123 9999**.

### Other useful organisations

**Contact** supports families with disabled children, providing guidance and information as well as bringing families together to support each other.

Helpline: **0808 808 3555**

Website: **contact.org.uk**

**Email: info@contact.org.uk**

**LOOK UK** supports young people (up to age 29) and families of children living with a visual impairment.

Telephone:**07464 351958**

Message via form at **look-uk.org/contact/**

Write toFred Bulmer Centre, Wall Street, Hereford, HR4 9HP

Web: **look-uk.org**

**Royal Society for Blind Children** provide a range of services in London and across England and Wales for blind and partially sighted children and young people, their families, and the professionals who work alongside them.

Tel: **020 3198 0225** (9am-5pm, Monday to Friday)

Website: **rsbc.org.uk**

Email: **connections@rsbc.org.uk**

**VICTA** supports children and young people (up to age 29) who are blind or partially sighted and their families across the UK.

Tel: **01908 240831**

Email: **admin@victa.org.uk**

Website: **victa.org.uk**

**Guide Dogs** supports children and adults with sight loss with many services, both with and without dogs.

Helpline: **0800 781 1444** (9am-5pm, Monday to Friday)

Email:**information@guidedogs.org.uk**

Web: **guidedogs.org.uk**

## We value your feedback

You can help us improve our information by letting us know what you think about it. Is this factsheet useful, easy to read and detailed enough – or could we improve it?

Send your comments to us by emailing us at **eyehealth@rnib.org.uk** or by writing to the Eye Health Information Service, RNIB, 105 Judd Street, London, WC1H 9NE.

## Information sources

This factsheet has been written by the RNIB Eye Health Information service. Our factsheets have been produced with the assistance of patient and carer input and up-to-date reliable sources of evidence. The accuracy of medical information has been checked by medical specialists. If you would like a list of references for any of our factsheets, please contact us at **eyehealth@rnib.org.uk**.

All our factsheets are available in a range of formats including print, audio and braille.

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RNIB is a member of the Patient Information Forum (PIF) and have been certified under the PIF TICK quality mark scheme.

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