

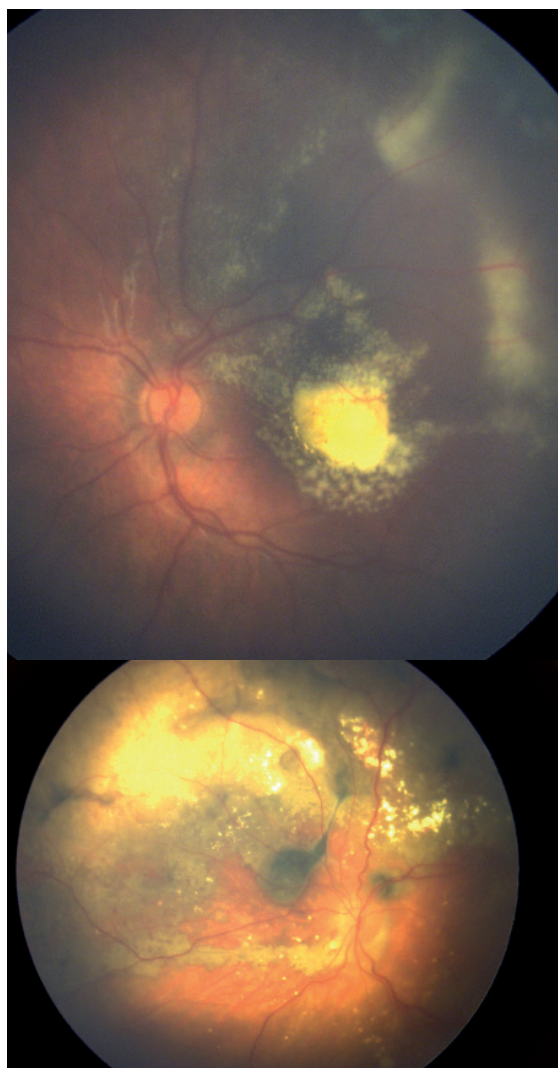
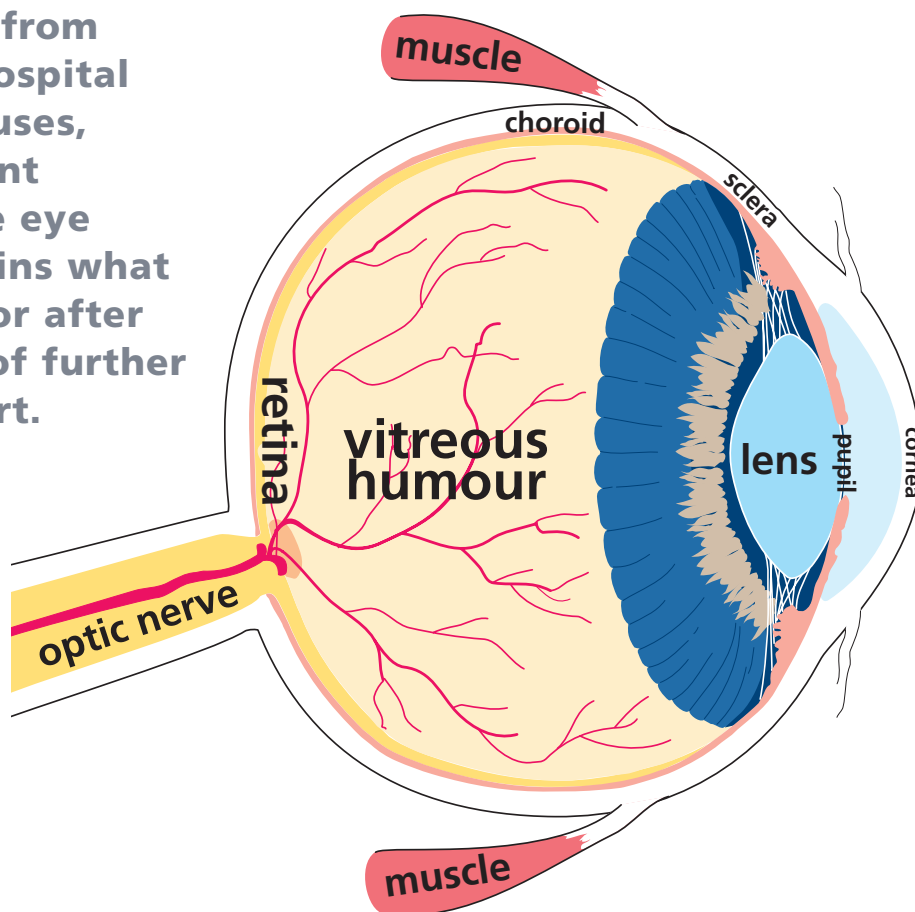
Coats' disease

NHS

Information for families

Great Ormond Street Hospital
for Children NHS Foundation Trust

This information sheet from Great Ormond Street Hospital (GOSH) explains the causes, symptoms and treatment of Coats Disease, a rare eye condition. It also explains what problems to look out for after diagnosis and sources of further information and support.



What is Coats' Disease?

Coats' disease is an abnormality of the blood vessels in the back of the eye – the retina. The retina is the layer on the inner surface of the eye that receives light and, together with the brain, turns that light into a picture.

The abnormal blood vessels may be leaky, which means that fluid can leave the vessels, becoming trapped within, and sometimes underneath, the retina causing it to lift up – this is called a retinal detachment. Fluid and detachment, when involving the central retina (macula), damage the ability of the retina to produce a clear picture.

How is Coats' Disease diagnosed?

A child with Coats' disease may comment on their vision becoming poorer in one eye, or be unaware of the change, since it is painless and slowly progressive. In this case poorer vision in the affected eye may be picked up on a routine eye test.

Sometimes others will notice the first signs, which can include a yellow or white pupil. When seen by an ophthalmologist, Coats' disease is diagnosed by clinical examination, when the back of the eye is looked at through a pupil dilated with eye drops.

This in itself may be sufficient, but many will go on to have an examination under anaesthetic and a retinal angiogram (known as a fundus fluorescein angiogram) to visualise the leaky vessels. Specialised photographs and scans of the back of the eye may also be taken. Two photographs of eyes with Coats' disease are shown below. The yellow areas on the retina are where leakage is taking place.

Who gets Coats' Disease?

Coats' disease is much more common in males than females. It is most commonly discovered in children before the age of 10 years, but can present in adulthood too. It is a rare condition, affecting 1 in 100,000 people.

What causes Coats' Disease?

We do not yet know the cause of Coats' disease and there are no known risk factors. There is one gene that may be implicated, the NDP gene, in which mutations (changes) may be found, although its role in developing Coats' disease remains uncertain. Genetic testing is something that you may wish to discuss with the Ophthalmologist.

Will my other children get Coats' disease too?

Coats' disease is not an inherited condition. This means it is not passed on through families. Brothers and sisters of an affected child are not likely to develop Coats' disease – it would have to occur again by chance. Since Coats' disease is so rare, the risk of this disease occurring again in a single family would be very unlikely indeed.

How will it affect my child's vision?

It is important to know that while it is possible for Coats disease to affect both eyes, in over 90 per cent of cases it is unilateral; that is, it only affects one eye. Children with Coats' disease therefore usually have one eye that is structurally normal and should see well for their lifetime.

If Coats' disease affects the centre of the retina (macula) or leads to total retinal detachment in the affected eye, vision in this eye can be permanently lost. The amount of vision lost is dependent on the location and extent of the retinal detachment and this is something your ophthalmologist will be able to discuss with you.

Is it associated with other health problems?

No. Coats' disease is a condition that is limited to the eye.

Will it get worse?

Coats' disease is known in most instances to be a progressive condition. It can however, stop progressing on its own, but it is not known when or why this occurs, or in whom. Poorer outcomes are usually in those children who are at a more severe stage of the disease at the point of diagnosis.

Is there a cure for Coats' disease?

Unfortunately there is no absolute cure. There are treatments that can be offered.

What are the treatments for Coats' disease?

Your child may be offered laser treatment. The laser is targeted at the abnormal blood vessels, at the point where they are leaking, which causes them to seal up. In addition, any areas of the retina that are shown on fundus fluorescein angiogram to have lost their normal blood supply will also be treated with laser. This is to prevent further leakage and also the growth of abnormal vessels that can occur in areas of retina starved of its oxygen supply.

Other treatments offered may include injection of a therapeutic substance called bevacizumab (Avastin®) or ranibizumab (Lucentis®), which can also reduce abnormal leakage and blood vessel growth.

If the retina becomes detached, laser treatment alone may not be sufficient and a more invasive surgical procedure may be required to reattach the retina.

What are the aims of treatment?

The primary aim of treatment is to prevent further deterioration in vision as a result of fluid reaching the central retina (macula). When the macula is already involved, treatment can, in some instances, improve central vision if the fluid resolves.

In the most advanced form of disease, where an extensive retinal detachment is present, the aim is to re-attach the retina where possible, and to manage the development of any secondary complications including elevated pressure within the eye, a chronically uncomfortable eye, and shrinkage of the eye.

Unfortunately Coats' disease can still worsen despite treatment.

What are the complications of Coats' disease?

Coats disease can lead to blindness or uncontrolled pressure inside the eye, which can sometimes cause pain. Rarely this can mean the eye needs to be removed.

It is very important that if your child's vision declines, or their eye becomes red, painful or they develop a headache that does not settle, that you contact your treating Ophthalmologist as soon as possible, or your local A&E if you have cause for concern over the weekend or out of office hours. These symptoms may be an indication of one of the above complications and require prompt treatment.

Contact Ophthalmology at GOSH for urgent review

- Clinical Nurse Specialist – ext 0487 (Monday to Friday 8am to 6m)
- Ophthalmology Doctor on Call – bleep 2226 (Monday to Friday 9am to 5pm)

Out of hours

- Contact Moorfields – Upper House Paediatric Eye Doctor on call for GOSH (usually based in the Accident and Emergency department at Moorfields)

Further information and support

Royal National Institute of Blind People (RNIB) support anyone affected by sight loss. Call their helpline on 0303 123 9999 or visit their website at www.rnib.org.uk

There is no UK based support organisation for Coats' disease but the umbrella organisation Contact a Family may be able to put you in touch with another family affected by the condition. Call their helpline on 0808 808 3555 or visit their website at www.cafamily.org.uk

There are two organisations in the United States that offer support and advice to anyone affected by Coats' disease: the Coats' Disease Foundation website at www.coatsdiseasefoundation.org and Know the Glow which aims to raise awareness of several eye conditions that can become apparent when a flash photograph is taken – visit their website at www.knowtheglow.org

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Compiled by the Ophthalmology department
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