



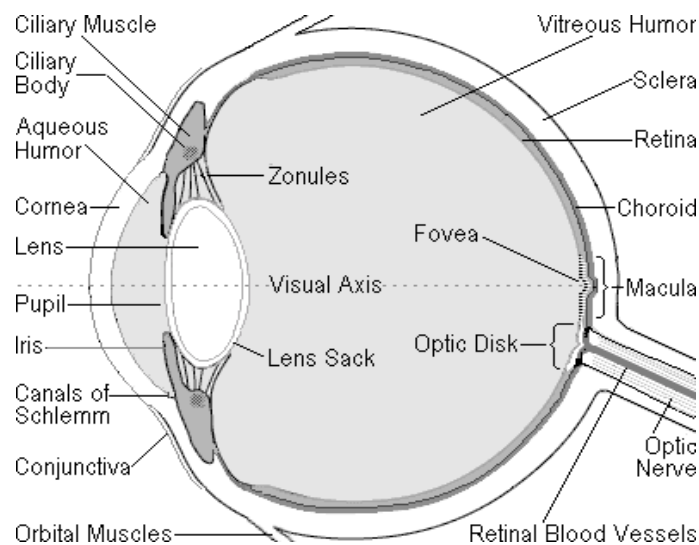
Birdshot Uveitis Society Factsheet

About Birdshot Chorioretinopathy

Please use this fact sheet as background information to help discussion with your doctors. Individual cases may vary enormously and so the best information will always come from your doctors. The information in this sheet has been checked for accuracy by leading uveitis specialists.

Birdshot Chorioretinopathy (shortened to Birdshot Uveitis or Birdshot) is a rare and potentially blinding chronic posterior uveitis. Uveitis means inflammation of the uvea, the part of the eye that is made up of the iris, the choroid layer and the ciliary body. The uvea provides most of the blood supply to the retina, which is the light sensitive tissue lining the inner surface of the eye.

In the picture of the eye below, you can see the position of the iris, the choroid and the ciliary body.



What are the symptoms of Birdshot?

Usually, the initial symptoms of Birdshot (and many other forms of posterior uveitis) will be floaters and/or blurred vision caused by inflammatory cells in the vitreous (these inflammatory cells in the vitreous are called vitritis). You can see where the vitreous (vitreous humour) is in the diagram above.

In the initial stages, you are likely to have a gradual, painless loss of vision involving one or both eyes. However, you may be able to continue to see well (retain good visual acuity).

There are sometimes other symptoms which may include:

- ⤴ Night blindness or Impaired vision in dim light (called nyctalopia)
- ⤴ Defective colour vision and difficulty distinguishing between certain colours (called dyschromatopsia)
- ⤴ Sensitivity to bright lights or glare (called photophobia)
- ⤴ A perception of flickering or flashing lights (called photopsia)
- ⤴ Fluctuating vision
- ⤴ Pain in the eyes
- ⤴ Decreased ability to judge depth
- ⤴ Shimmering vision (this is difficult to describe, but some people describe it as being like looking through dirty glass or through water)
- ⤴ Distorted images (called metamorphopsia)
- ⤴ Decreased peripheral vision (peripheral vision is the ability to see on either side, whilst looking forward)
- ⤴ Ceiling Fan effect – when you close your eyes you are left with an image of a ceiling fan whirring around.

If you have had Birdshot for some time, an ophthalmologist is likely to be able to see oval shaped spots, which are orange to cream in colour, at the back of your eyes on your retina. The reason this disease is called Birdshot is because these spots resemble the pattern seen when you fire birdshot pellets from a shotgun.

Because Birdshot is a rare disease, it can often be misdiagnosed in the initial stages, particularly as the 'birdshot spots' are often not clearly visible to begin with.

What causes Birdshot Chorioretinopathy?

It is not known what causes Birdshot. It is believed to be an autoimmune disease. An autoimmune disease is an illness that occurs when the body tissues are attacked by its own immune system.

The majority of people who are diagnosed with Birdshot carry an antigen called HLA- A29. An antigen is a substance that enters the body and stimulates the production of an antibody to fight what the immune system perceives as an invader. Because most people who are diagnosed with Birdshot carry this antigen, it may mean that they have an inherited immune dis-regulation, and research is currently being undertaken to try and clarify this.

However, many people carry the HLA-A29 antigen and never get Birdshot. It is therefore assumed that other factors must initiate the onset of the disease (a 'trigger'). This 'trigger' may be a virus, a bacteria or an environmental factor.

Background

The first case of Birdshot was described in 1949 by two ophthalmologists, Franceschetti and Bable, but it was not until 1980 when two other ophthalmologists, Ryan and Maumenee coined the term 'Birdshot' because of the resemblance of the lesions to the scatter pattern from a birdshot gun.

There is a general view amongst many researchers and ophthalmologists that Birdshot is a relatively new disease which had not occurred prior to the first description in 1949. This is because the symptoms (including the lesions on the retina) are so striking, they would have been recognised had they occurred prior to 1949.

Frequency of Birdshot

Birdshot is a rare disease. A rare disease is defined in Europe as: a disease affecting less than 5 per 10,000 inhabitants and is fatal or severely debilitating. Because Birdshot is a relatively new disease, it is difficult to identify how many people have been diagnosed with it. Recent research (carried out by Rea Mattocks and Annie Folkard, founders of BUS) suggests that whilst Birdshot remains a rare disease under this definition, it is either increasing in incidence or being more readily diagnosed.

What is the progress of the disease (prognosis)?

Birdshot seems to come in a 'spectrum' from mild to severe, from easy to treat to difficult to treat. For those people with 'mild' Birdshot, they can often maintain good eyesight (visual acuity) with little or no medical intervention. However, Birdshot is a potentially blinding disease, particularly if it is severe and untreated.

The typical course of Birdshot, like other autoimmune diseases, is characterised by flare-ups of inflammation inside the eye (exacerbations which cause loss of visual acuity if left untreated) and remissions. Because of this, many ophthalmologists believe in quite an aggressive approach to ensure that inflammation is kept to a minimum and the maximum amount of visual acuity can be preserved.

The most concerning issue relating to Birdshot is that if it is severe and is left untreated, or if there are many flare-ups, it can lead to macular oedema (this is a swelling of the macula layer in the eye) which can cause blindness.

Who gets Birdshot?

Because of the link to the HLA-A29 antigen, which is found more frequently in Caucasians, most people with Birdshot are Caucasian. There does not seem to be a gender bias, although some ophthalmologists report diagnosing more women than men.

In the past, it was believed that the average age of onset of Birdshot was 50 to 55 years (i.e. that it usually starts in people who are aged approximately 50 to 55) and that it is a disease unlikely to affect people under the age of 35. However, we now know that people in their 20s and 30s are being diagnosed, and it may be that children, too, can suffer from Birdshot. It is important to remember this, as some specialists who do not often see Birdshot, may still be under the assumption that it only happens to people in their 50s upwards.

Is there a cure for Birdshot?

At this moment in time, there is no known cure for Birdshot. Most research being undertaken is looking at finding alternative and more effective medications that have fewer side effects. However, there is some research being undertaken to establish the genetic links and the 'triggers' for Birdshot and this may lead to a cure or a prevention regime in the future.

Currently, the aim is to ensure that people with Birdshot reach remission or a stabilisation of their condition, using medication which can sometimes be quite toxic and needs careful monitoring. However, getting diagnosed and receiving appropriate medication can sometimes feel like a bit of a lottery. This is because:

Firstly, the condition is difficult to diagnose, due to its rarity (many doctors and opticians will never have seen a case of Birdshot).

Secondly, there is still a belief amongst certain ophthalmologists that Birdshot is a self-limiting disease and will eventually 'go away' or 'burn out'.

Thirdly, even if you are diagnosed, because this disease is a rare disease, newer forms of medication may be difficult to access on the NHS, as there is not a large body of evidence to support their efficacy.

Fourthly, it is our experience that each individual with Birdshot responds differently to different medication regimes and has a virtually unique course that their disease follows.

Much of the treatment for Birdshot, therefore, has to rely on a very strong relationship between the patient and their ophthalmologist with frequent contact and a willingness on the part of the ophthalmologist to listen to what the patient believes is going on in his/her eyes and a willingness to use newer forms of medication if the more traditional ones appear not to work.

How is Birdshot diagnosed?

Because Birdshot is difficult to diagnose, and is not widely known about, it may be diagnosed through 'default' – that is, all other, more common conditions that cause similar symptoms will be tested for, before Birdshot is considered.

Usually, the definitive diagnosis for Birdshot will be made through a blood test to establish whether you test positive for the HLA-A29 antigen. You may also receive one or more of the following tests:

Fluorescein angiography (fluorescein – the type of dye that is used; angiogram – a study of the blood vessels) is an extremely valuable test that provides information about the circulatory system and the condition of the back of the eye. The test is performed by injecting a special dye, called fluorescein, into a vein in the arm. In just seconds, the dye travels to the blood vessels inside the eye. A camera equipped with special filters that highlight the dye is used to photograph the fluorescein as it circulates through the blood vessels in the back of the eye. If there are any circulation problems, swelling, leaking or abnormal blood vessels, the dye and its patterns will reveal these in the photographs. Please note that you will have yellow skin and yellow eyes for a while and your urine will be bright yellow. This is normal.

Indocyanine green angiography (ICG). This is a similar procedure to fluorescein angiography. ICG angiography uses IndoCyanine Green dye which fluoresces in the infra-red (non-visible) light. The infra-red wavelengths have the ability to penetrate the retinal layers, making the circulation in deeper layers visible when photographed with an infra-red sensitive camera.

Optical Coherence Tomography (OCT). This is a non-invasive technology used for imaging the retina, the multi-layered sensory tissue lining the back of the eye. This can determine whether you have any optic nerve damage or macular swelling.

Electroretinogram (ERG). This is a series of non invasive tests where small wires are placed alongside your eyelids and receptors are placed on your forehead and the back of your head. You will then look at a range of flickering lights and patterns. This will assess how well you have maintained your eyesight – particularly in your retina and choroid and your rods and cones. This test is painless, although it may be slightly uncomfortable.

Visual Field Test. This is a method of measuring an individual's entire scope of vision, (central and peripheral [side] vision). Visual field testing maps the visual fields of each eye individually

What happens once you have been diagnosed?

If you have had a full range of tests prior to diagnosis, you will have a record of how your eyes are functioning and you and your ophthalmologist will be in a good position to judge the efficacy of any medication regime.

Currently, the right treatment has to be tailored to suit the individual concerned, and will depend on the results of any of the above tests.

There are two specific areas that your ophthalmologist will want to address: firstly trying to reduce the inflammation and secondly trying to control your autoimmune system so it stops attacking your eyes.

The current protocol is to treat with a combination of steroids (to reduce the inflammation) and immuno-suppressants (to stop your immune system attacking your eyes). These are strong drugs and do have side-effects, so it is important that there is proper monitoring and good access to your GP, ophthalmologist and rheumatologist. The reason you are likely to need to see a rheumatologist as well as an ophthalmologist is that the rheumatologist has expertise in using immunosuppressant medications. The goal of the treatment is to find the lowest dosage of medication that will allow you to maintain your eyesight and stabilise your condition, or allow you to reach remission.

If you are put on steroids, you will also be given medication to counteract any damage to your bones that high doses of steroids may cause. These are usually alendronic acid and calcium with vitamin D tablets. If you are not given these automatically, you should ask your consultant about this.

You may also be given gastric medication to counteract any effects on your stomach.

If this first line of treatment proves unsuccessful and inflammation returns, there are other medications which can be used and which may suit you better.

Unfortunately, at the moment there is no one definitive medication regime that works with everybody who has Birdshot.

Monitoring

If you are taking immunosuppressants and steroids, you will be (or should be) monitored for any unwanted side effects. Your rheumatologist or ophthalmologist will advise what the monitoring should be, but they will usually include blood tests and DEXA scans. It is important to follow your specialist's instructions and get these tests done when you are told to, as they are designed to safeguard your health.

Blood Tests and blood pressure monitoring

The Blood tests check that your kidney and liver are functioning well, measure your cholesterol levels and the levels of the immunosuppressants in your system to make sure you are not having too much or too little. These routine blood tests are done very regularly to begin with and then between every 6 – 12 weeks depending on your consultant's advice and your test results. The blood tests can be done at the specialist centre, or by your GP, or local renal unit. Because the medication can increase your blood pressure, this too, should be monitored regularly.

You are also likely to have regular visual field tests and/or OCTs and/or ERGs (described above). You may also be sent for further Indocyanine Green or Fluorescein angiograms.

Dexa scan. If you have been taking oral steroids for some months, your GP should arrange for you to receive a bone density (DEXA) scan periodically (usually once every other year) to check that your bones have not thinned as a result of the steroids that you have been taking. The information below explains what this involves and gives a bit more information about how it is done. You may have to ask your doctor to arrange this for you. In our experience it does not necessarily happen automatically.

What is a Bone Density scan (DEXA)?

A DEXA scan is used to measure the density of bones. It is mainly used to diagnose osteoporosis (loss of bone material) and to assess your risk of having a fracture.

It is an enhanced form of x-ray technology that is used to measure bone loss. DEXA scanning is today's established standard for measuring bone mineral density (BMD).

DEXA is most often performed on the lower spine and hips. In children and some adults, the whole body is sometimes scanned. Peripheral devices that use x-ray or ultrasound are sometimes used to screen for low bone mass. You should wear loose, comfortable clothing, avoiding garments that have zippers, belts or buttons made of metal. Objects such as keys or wallets that would be in the area being scanned should be removed. This procedure is completely painless.

Further information and Support

There is a large amount of material about Birdshot which can be accessed simply by typing Birdshot into your web browser. One of the most acclaimed explanations of Birdshot, written by a leading American ophthalmologist, Dr C Michael Samson, can be accessed on

<http://emedicine.medscape.com/article/1223257-overview>

If you live in the **US**, you can access an on-line discussion forum for Birdshot, run by Dagmar, who has Birdshot.

<http://bsrc.lefora.com/forum/category/bsrc-forum-topics/page1/>

BUS (the Birdshot Uveitis Society) provides information and support to anyone with Birdshot. It also collects information to establish how many people have Birdshot and to stimulate research. Joining BUS as a member is free of charge, and allows you access to Birdshot Days held in the UK where professionals and patients come together to exchange information. If you have been diagnosed with Birdshot and want to contact us (Rea and Annie, who both have Birdshot and have set up and run BUS, the Birdshot Uveitis Society) you can email us directly on: info@birdshot.org.uk

If you would like to access our website, it can be found at:

www.birdshot.org.uk

If you would like to write to us, our address is:

Birdshot Uveitis Society, PO Box 64996, London SW20 2BL

Tel: 0845 605 1195

The information contained in this Factsheet has been written for patients by Rea Mattocks and Annie Folkard (both patients) in conjunction with Mr Nigel Hall, Consultant Ophthalmologist at University Hospital Southampton Healthcare Trust

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