

13. Behçet's - In Children

How are children affected by Behçet's disease?

Behçet's disease (also called Behçet's syndrome) is a disease causing **inflammation** in the body, including the blood vessels (vasculitis). It is most common in people from the Mediterranean, the Middle East and the Far East ('the Silk Route'), but it does also occur in Caucasians of European origin and other ethnic groups. Although the usual onset is in adulthood, the disease often does begin early in childhood but may not be diagnosed for many years since the symptoms can be very non-specific. There is a spectrum of disease, ranging from children with **recurrent** mild mouth ulceration to those with severe complications affecting other organs in the body (see below). We do not know what causes Behçet's disease, or how to predict how severe a child's illness will be, but we believe that a combination of genetic traits (such as a marker called HLA B51), which are found more commonly in certain ethnic groups (particularly in patients from Mediterranean areas and the Far East), in combination with environmental factors, are likely to contribute to the onset and severity of the disease. This is an ongoing area of research.

There is no specific diagnostic test for Behçet's disease, and it is important that your doctor excludes other conditions that could mimic Behçet's disease in children. These include inflammatory bowel disease, immunodeficiency, periodic fever syndromes and autoimmune diseases such as systemic lupus erythematosus. Your doctor may wish to undertake genetic tests, and other investigations to exclude these alternative diagnoses.

Complications and how they are treated

Mouth and genital ulcers

Mouth ulcers and genital ulcers are common, and can be painful for children. They rarely cause long-term disabling or disfiguring complications. The treatment of these therefore includes simple measures such as **topical** corticosteroids, or steroid creams, sprays or mouth washes. Other treatments may include medicines such as colchicine or other immunosuppressive agents (see below).

Skin rash

This can take different forms, including red nodules, acne-like spots, folliculitis, or unusually severe reactions to stings or other trauma (a reaction called pathergy).

Sometimes this pathergy reaction is helpful in diagnosing Behçet's disease, although the test is currently rarely performed in the UK paediatric practice. This mainly applies to patients from Turkey or the Far East, because this type of reaction is seen much less commonly in patients from Northern Europe. Treatment is with steroid creams, colchicine or, for more resistant or severe disease, oral steroids and/or [immunosuppressants](#) under expert advice (see below).

Eye disease

This is a more severe complication of Behçet's disease and requires careful assessment by an [ophthalmologist](#), and specialist treatment. [Uveitis](#) and [vasculitis](#) affecting the eye can occur. **(See separate medical factsheet: Behçet's disease and the eyes)**. Typical treatments will include corticosteroids, given either by mouth or as eye drops, and medicines to help prevent spasm of the iris muscle in the eye and/or immunosuppressants taken by mouth or sometimes [intravenously](#). Sometimes the eye can be injected directly with corticosteroids (under general anaesthetic), but this is rarely required for eye disease in children.

Arthritis

This is fairly common in children with Behçet's disease. It may affect one or more joints. This may be helped by simple treatments such as paracetamol or anti-inflammatory medicines such as ibuprofen, but colchicine or immunosuppressants may sometimes be required to control more severe arthritis.

Blood vessel disease

Any blood vessel in the body can be affected by inflammation in Behçet's disease, including blood vessels of the heart, lungs, intestine and brain. [Aneurysms](#) of the main lung arteries can occur – a severe but thankfully very rare complication of Behçet's disease in children. These require urgent specialist assessment and treatment.

Gastrointestinal involvement

Again, although severe, this is a relatively rare complication of Behçet's disease in children. Ulcers can occur in the wall of the intestine causing tummy pain, weight loss, fever and sometimes the passage of blood. Investigation of this would include an endoscopy (intestine telescope test), or video capsule test and tissue [biopsy](#), which is routine but would be performed by a paediatric [gastroenterologist](#) with experience in doing this. Again, treatment would depend on the severity, but it would include colchicine, immunosuppressants and corticosteroids.

Thrombosis

This means formation of blood clots, usually in a vein, and is recognised in some children with Behçet's disease, albeit rarely. It is not possible for doctors to predict who will be at risk of this. The usual treatment will include reducing blood vessel inflammation (see preceding section above), low doses of aspirin and/or [anticoagulants](#), depending on the site and severity of the thrombosis.

Involvement of the brain and nerves

Although headaches are relatively common in children with Behçet's disease, serious neurological complications are thankfully rare in children, although they can occur. These can cause severe headaches, strokes, thrombosis in the main vein of the brain causing very severe headache, and sometimes involvement of the nerves to the peripheries. Sudden onset of new headache, severe pins and needles or other neurological symptoms requires urgent specialist assessment, an MRI brain scan and appropriate treatment, usually with corticosteroids combined with immunosuppressive agents under specialist advice.

Kidney involvement

Although recognised, kidney involvement is rare in children with Behçet's disease. Inflammation of the kidneys (glomerulonephritis) can occur and would require a kidney biopsy for diagnosis. Sometimes, inflammatory proteins made by the body in response to poorly controlled inflammation caused by Behçet's disease can accumulate in the kidney. This rare long-term complication is called amyloidosis and can be prevented by ensuring that the Behçet's disease is well controlled generally. It is very rare in children but can occur in adults.

Effect on growth, puberty and school

Severe uncontrolled Behçet's disease can impair growth and delay puberty, and can cause osteoporosis (brittle bones) in children. This is due to the underlying disease and sometimes its treatment with high doses of systemic steroids for prolonged periods of time. This is why it is important that the overall treatment of your child's disease is monitored carefully by a specialist. Many children miss a lot of school because of undiagnosed and/or poorly controlled Behçet's disease. Once diagnosed and treated properly, however, virtually all children can attend school normally and participate fully in physical activities such as PE.

Treatment

Anti-inflammatory, immunosuppressive and anticoagulant treatments

A variety of anti-inflammatory or immunosuppressant medicines are used in the treatment of Behçet's disease in children. These are also used in adults. Treatment is tailored to the individual patient, and your doctor may try different medicines over time to find which is best for your child. This generally involves starting with simple treatments that have the least side-effects, working up to stronger treatments for those who require it. We call this approach the Behçet's disease 'therapeutic ladder'.

Generally speaking, mouth and genital ulcers are treated where possible with topical agents including steroid pastes, mouthwashes, creams and/or sprays. Mild disease affecting other organs (such as more severe mouth ulcers, arthritis and skin rashes) can be treated with anti-inflammatory medicines (including colchicine, or medicines like ibuprofen). More severe disease requires immunosuppressant medicines such as azathioprine, mycophenolate mofetil, ciclosporin, tacrolimus or cyclophosphamide, often used in combination with

corticosteroids depending on the severity of the disease. Increasingly we are using newer treatments such as anti-TNF-alpha medicines (etanercept, adalimumab and infliximab). Your doctor will provide specific information regarding the potential side-effects of each of these medicines. Another medicine that may be considered is called thalidomide, although this is less commonly used since anti-TNF-alpha treatments have become available. Again, the use of this treatment is tailored for specific individuals, and your doctor will discuss this option with you if necessary. If major thrombosis has occurred, your doctor may recommend anticoagulant medicines such as heparin and warfarin. Your doctor will advise you regarding the specific side-effects and monitoring required for these types of medicines.

Newer treatments

This is an area of ongoing research, and there are new treatments that have been used in small numbers of children with Behçet's disease. These include anti-TNF-alpha medicines (including etanercept, adalimumab and infliximab), interferon-alpha, anakinra (which blocks interleukin-1, another inflammatory hormone) and a medicine called lenalidomide (a newer and potentially safer version of thalidomide). Experience with these newer therapies in Behçet's disease in children is limited, but there have been reports of potential benefit in children. In particular, anti-TNF-alpha treatments are routinely used to treat children with more severe Behçet's disease, and have largely replaced thalidomide.

Explanation of words highlighted in factsheet in the order they appear

Inflammation:	the way the body responds to irritation, infection or injury. Blood collects in infected areas causing reddening, swelling and pain
Recurrent:	something that happens again from time to time, or more frequently
Topical:	topical medicine is medicine usually put directly onto the skin in the area affected
Immunosuppressants:	drugs that reduce inflammation over long periods of time (generally years rather than months)
Ophthalmologist:	doctor who specialises in studying and treating diseases of the eye
Uveitis:	inflammation of the front of the eye around the iris which may cause redness and pain
Vascular:	anything to do with blood vessels: vasculitis is inflammation in veins and arteries
Intravenously:	when a drug or other fluids given directly into a vein
Aneurysm:	when the wall of a blood vessel becomes weaker and bulges or swells at the point of weakness
Gastrointestinal:	about the stomach and the intestines (guts)
Gastroenterologist:	a doctor who is specially trained in the diagnosis and treatment of intestinal disorders

Biopsy: removing a small amount of tissue for examination under a microscope.

Anticoagulant: drug used to stop the blood clotting

Systemic: something which has an effect on the whole body, not just a part of it. Systemic therapy means taking medicines by mouth (as tablets), or as injections or infusions