Behçet's Syndrome Society

Registered Charity No: 326679
Caring for those with a rare, complex and lifelong illness
www.behcets.org.uk

Behçet's Syndrome in children: Information sheet for parents.

Background

Behçet's Disease (also called Behçet's Syndrome) is a disease causing inflammation in the body, including the blood vessels (vasculitis). It's most common in people from the Mediterranean, the Middle East, and the Far East. It does also occur in Caucasians of European origin and other ethnic groups.

Although the usual onset is in adulthood, the disease can and does occur in children. 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, or how to predict how severe your 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 disease onset and its severity. 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 such as inflammatory bowel disease, immunodeficiency, periodic fever syndromes or autoimmune diseases such as systemic lupus erythematosus.

Symptoms your child may have and their treatment

1. **Mouth and genital ulcers**. Mouth ulcers and genital ulcers are common, and can be painful for children. It is rare that these cause long-term disabling or disfiguring complications. The treatment of these therefore includes simple measures such as topical corticosteroids (adcortyl in orabase), or steroid creams or sprays (different options are provided at http://www.behcets.org.uk). Other treatments may include medicines such as colchicine or other immunosuppressive agents (see below).
2. **Skin rash**. These 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 to diagnose Behçet's disease. 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).
3. **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. Typical treatments will include

corticosteroids either given 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. Sometime the eye can be injected directly with corticosteroids (under general anaesthetic).

4. **Arthritis.** This is fairly common in children with Behcet's disease. It may affect one or more joints. This may be helped by simple treatments such as paracetamol or anti-inflammatory medicines like ibuprofen; but may ultimately require immunosuppressants to control more severe arthritis.
5. **Blood vessel disease.** Any blood vessel in the body can be affected 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.
6. **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) and tissue biopsy which is routine but would be performed by a paediatric gastroenterologist with experience in doing this. Again the treatment of this would be dependent on the severity, but would include immunosuppressants and corticosteroids.
7. **Thrombosis.** This means formation of blood clots usually in a vein, and this is recognized in some children with Behçet's disease. It is not possible for doctors to predict who will be at risk of this. The usual treatment of this will include anti-coagulants and/or low doses of aspirin depending on the site and severity of the thrombosis.
8. **Involvement of the brain and nerves.** Although headaches are relatively common in children with Behçet's, serious neurological complications are thankfully rare in children, although can occur. This 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 or other neurological symptoms requires urgent specialist assessment, MRI brain scan and appropriate treatment, usually with corticosteroids combined with immunosuppressive agents under specialist advice.
9. **Kidney involvement.** Although recognized, kidney involvement is rare in children with Behçet's disease. Inflammation of the kidneys can occur (glomerulonephritis) and would require a kidney biopsy to diagnose. 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.
10. **Effect of Behçet's disease on growth and puberty in children**
    Severe uncontrolled Behçet's disease can impair growth, 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 disease be monitored carefully by a specialist.

Factsheet provided by Dr Paul Brogan on behalf of the Behçet's Syndrome Society, 2009

Anti-inflammatory, immunosuppressive, and anticoagulant treatments used for the treatment of Behçet's disease

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.

Generally speaking, mouth and genital ulcers are treated where possible with topical agents including steroid pastes, creams and/or sprays. Mild disease affecting other organs (such as arthritis) 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. 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. Again the use of this treatment tailored for specific individuals and your doctor will discuss that as an option with you if necessary.

If major thrombosis has occurred, your doctor will 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 for Behçet's disease in children

This is an area of ongoing research, and there are new treatments which have been used in small numbers of children with Behçet's disease. These include anti-TNF-α medicines, interferon-α, 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 reports of potential benefit in children have been described.