

Highlighted words (in blue) are explained in Common Terms and Definitions sheet

Behçet's **syndrome** (or Behçet's **disease**) but more simply Behçet's, is a multisystem condition, in which potentially any organ can be affected. Neuro-Behçet's is one of the more severe manifestations of Behçet's, and affects 10% of people with Behçet's'.

How does Neuro-Behçet's present?

Behçet's disease is characterised by the, inflammation of blood vessel-walls, usually at multiple sites in the body. If the blood vessels supplying the nervous system are involved Neuro-Behçet's disease occurs.

The inflammation may remain restricted to the blood vessels, and superimposed clots (thereby thrombosis), blood vessel wall tear (thereby haemorrhage), and reduction of blood circulation (thereby stroke) may appear. If however, inflammation spills into brain and spine tissue, various neuro-inflammatory symptoms appear.

These include headaches, double vision, loss of balance, speech and swallowing problems, weakness, numbness, bladder symptoms, visual problems or seizures. Intracranial hypertension which means increase in the pressure of the fluid in brain and brain haemorrhage may occur occasionally.

Usually people will have systemic features of Behçet's like genital ulcers and eye symptoms etc. though they may or may not have had a prior diagnosis of Behçet's.

Neurological symptoms may affect people either as on-off neurological illness, or can remit and reoccur, or can grumble along over long time especially if left un-treated.

Very rarely psychiatric symptoms like hallucinations and paranoia have been reported in people with Behçet's disease. This is akin to delirium and psychosis seen due to other neurological causes.

Symptom overlap with other neurological conditions, and therefore, diagnostic process may take long. Your local neurologist may need to link up with a neurologist linked to Behçet's Centre of Excellence.

Common conditions like migraines and tension headache can affect any one including people with Behçet's disease, and therefore headaches and related symptoms need through evaluation as it may not mean it is Neuro-Behçet's. But if in doubt contact your doctor.

Anxiety and depression are commonly associated with long term conditions. This is similarly the case with people with Behçet's disease.

What are the tests and treatments required?

Specialist neurological tests such as a brain and spine MRI scans, and lumbar puncture are needed.

Diagnosis requires establishing diagnosis of Behçet's; and confirming if neurological presentation fits in with identifiable/recognised neurological presentation.

Diagnosis of Behçet's is established by ISG criteria 1990, and now International Criteria for Behçet's Disease (ICBD) 2013 proposes the criteria provide diagnosis and classification of Behçet's.

Diagnosis of Neuro-Behçet's is established by International Consensus Recommendations (ICR) criteria. This was the work of an international collaboration of neurologists, non-neurological Behçet's experts, as well as people living with Behçet's disease. These are published in the *Journal of Neurology*. The full article can be accessed online under *Journal of Neurology* (2014) volume 261 Sept 2014 – ([J Neurol. 2014 Sep; 261\(9\):1662-76](#)). Your local neurologist and neurologist linked to Behçet's Centre of Excellence would be able to work together to confirm the diagnosis.

Treatment depends on the pattern of presentation – that is, whether the brain tissue is inflamed or blood vessels are affected.

Brain tissue inflammation is more difficult to treat, can recur and might result in disability. An early immune therapy may be needed for a longer time. Your neurologist will give a course of steroids immediately to control inflammation. Steroids are likely to be continued for over few months. Immune suppressive treatments like Azathioprine are also used to control the disease and prevent further relapse. These take a few months to be effective.

More recently, a number of biological treatments (Infliximab, Adalimumab etc.) have become available. These may be used for severe presentations brain and spine inflammatory presentations straight away alongside the steroids. These are also prescribed in instances of worsening or recurrences. These may be needed to be continued for a reasonable length of time. Your local neurologist and neurologist linked to Behçet's Centre of Excellence would be able to supervise the treatment.

Thrombosis requires shorter-term immune treatment usually. But in case of recurrence treatment with immune suppressive may need to continued long term. This is also the case for combination presentations. Other simultaneous body system involvement like eye, gut, and major blood vessels will also be an important factor to be considered whilst treating neurological symptoms. Your neurologist may find the above article useful towards the treatment of Neuro-Behçet's.

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