

## 16. Neuro-Behçet's Disease

Most people reading this need no introduction to Behçet's disease (BD). This readership is either affected by this condition or is involved in care of those who are. We as neurologists are involved in the care.

Involvement of the neurological system is called neuro-Behçet's and is among the most disabling complications of BD. Thankfully, it is not that common and affects about 10% of people with BD. Neuro-Behçet's usually occurs in combination with **inflammation** of the other organs systems such as eyes, joints, skin etc. and only rarely separately.

In this article we intend to discuss neuro-Behçet's disease and how it can affect people. What symptoms people have and how we can investigate, diagnose and treat this condition?

### **How does neuro-Behçet's disease present?**

Neuro-Behçet's disease can cause inflammation of the brain/spinal tissue directly, or can cause inflammation and blockage of the blood supplying them (usually veins rather than arteries).

If there is direct inflammation of the brain tissue, people can present with one feature or a combination of features including weakness, pins and needles, loss of co-ordination, double vision, confusion, fits and meningitis. On the other hand, if the blood vessels (veins) are inflamed and blocked, patients present with symptoms of raised **intracranial** pressure such as headache or blurred vision. They usually have some other background Behçet's disease symptoms such as mouth ulcers, genital ulcers or eye symptoms. Arteries are very rarely affected.

Patients with Behçet's disease commonly have headaches. Headaches can occur because of neuro-Behçet's disease per se or more often because of other reasons such as migraines. It is a neurologists' job to look into these symptoms and decide what further tests or treatments are needed.

### **What are the tests and treatments required?**

Specialist tests such as a brain scan or lumbar puncture might be required. 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.

In general, an early and strong immune therapy is needed for a longer time. Neuro-Behçet's with blood vessel involvement requires shorter-term treatment once the initial crisis is over.

Behçet's disease experts differ in their approach to treatment based on their experience. There were no clear guidelines on the main issues in diagnosis and treatment of neuro-Behçet's disease, so we set out to do this project.

### **International Neuro-Behçet's Recommendations Project**

We developed clinical and practical guidelines on the key issues in the diagnosis and treatment of neuro-Behçet's disease to assist clinicians involved in care of these patients. As Behçet's disease can affect different organs at the same time, it was imperative that we involved specialists from different fields to cover the treatment in a [holistic](#) way.

It took us almost 3 years to complete the above project. An advisory group made of 52 international Behçet's disease experts from 20 countries and 11 different medical specialities, including 23 neurologists and voluntary patient representatives, worked together on this project. Jan Mather was the patient group representative from the Behçet's Syndrome Society. Most of the UK specialists involved are also BSS members. A full list of group members is given at the end of this article.

We identified important clinical issues that needed addressing under the umbrella of this project. These covered diagnosis and treatment of neuro-Behçet's disease. This was presented at the [ICBD](#) meeting in 2010 in London. We looked at the previously published studies thoroughly and systematically. Conclusions were drawn from the literature and were discussed and voted on. Only high scoring recommendations that passed the consensus criteria were given out as International Consensus Recommendations. These were published in the *Journal of Neurology*. The full article can be accessed online under Journal of Neurology (2014) volume 261 Sept 2014. **It might be useful if you mention this article to your treating clinician.**

This project brought up clear and easily usable diagnostic criteria for neuro-Behçet's disease. We also made 16 practical clinical recommendations for diagnosis and treatment of neuro-Behçet's disease. We hope that these will be a useful resource to the Behçet's disease community.

We would like to thank all members of the advisory and consensus group members who helped towards this project. Special thanks go to Jan Mather for representing the patients on the Advisory Group. Without their help this project would not have been possible.

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### **List of all advisory and consensus group members in alphabetical order**

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**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

Intracranial: within the skull

Recur: something that happens again from time to time

Holistic: a holistic approach is one which concentrates on the whole body or person not just one part of it

ICBD: the International Conference on Behcet's Disease