

## 6. Behçet's – The Brain

### **How is the brain affected by Behçet's disease?**

Neurological ([nervous system](#)) complications in Behçet's disease arise in 5–20% of cases. There are two separate mechanisms by which this may occur, and it is most uncommon for a patient inflicted by one also to develop a neurological problem due to the other. The most common type is known as the parenchymal type, in which inflammation of the brain or spinal cord develops. This is called meningoencephalitis. The other type of neurological problem relates to development of thrombosis ([when blood clots form](#)) within the veins that drain blood from the brain. Psychiatric conditions may also occur, although this is rare.

### **Complications**

#### ***Meningoencephalitis*** ([swelling of the brain or spinal cord](#))

Patients with meningoencephalitis develop headache and drowsiness, which worsen over several days, followed by symptoms of the inflamed brain itself. What these are depends on the part of the brain affected and how intense the inflammation is. The most common site is the brainstem, and 30–50% of affected patients will have symptoms from this problem. Patients often develop double vision, loss of balance, slurred speech and difficulty swallowing. This can be mild and get better on its own within days or can be more severe, needing hospitalisation, and even so severe as to require treatment in an intensive care unit. If other parts of the brain are affected, weakness and numbness, or visual or thinking problems arise and doctors may diagnose a stroke or a brain tumour before the correct diagnosis is reached after tests. Sometimes, epileptic seizures occur. If the spinal cord is involved, patients complain of numbness, weakness, and bladder and sexual problems. Rarely, the optic nerves ([of the eyes](#)), other nerves and muscles can be affected.

#### ***Thrombosis***

Thrombosis causes pressure to develop within the brain, leading to headaches and visual symptoms. Occasionally, weakness and seizures may also occur, and brain haemorrhage ([blood escaping from a burst blood vessel](#)) may develop if the condition is not treated quickly. Occasionally, patients can develop similar symptoms without any sign of thrombosis within the brain. This is known as intracranial hypertension, and tends not to be as serious as venous sinus thrombosis. ([Intracranial hypertension means an increase in blood pressure within the brain Venous sinus thrombosis means a disturbance in blood draining from the brain because of a blockage which leads to increased pressure within the brain.](#))

## **Psychiatric conditions**

Psychiatric conditions occur only very rarely in Behçet's disease, when parts of the brain that look after emotion and thought are affected by the meningoencephalitis noted above. Occasionally, patients may present with hallucinations and abnormal thoughts such as paranoia, and difficulty thinking and remembering. This is most uncommon and normally settles down well with the correct treatment. Separate from this is the syndrome of fatigue, anxiety and depression, which can also cause thinking and memory problems but which is not related to a problem within the brain. This, in contrast, is very much more common, not just in Behçet's disease but in most chronic (long term) and difficult conditions. This is not surprising, but some doctors, even GPs, fail to recognise this, which can be frustrating to patients. It has been shown that patients with Behçet's disease have higher ratings on depression and anxiety scores, and that these scores vary with the severity of the underlying illness. So-called fibromyalgia symptoms (aches and pains with tiredness) also correlate with how the Behçet's disease is behaving, but it is also true that the symptoms of anxiety and depression can make the Behçet's disease feel worse when it is not actually in relapse. (a return of an illness after a period when things had got better). So it is a very complicated problem.

## **Investigations**

Neurological investigations include blood tests to assess the severity of the Behçet's disease, and to ensure that there is not a separate disease causing the whole problem, or just the neurological one – for example, an infection such as meningitis, a stroke or brain haemorrhage.

An MRI scan is a very helpful test and will disclose the cause of the neurological problem in around 80% of cases. In others, the problem may be too small to be visible on the scan, or too diffuse within the brain to be picked up. A spinal fluid examination is often helpful and very important when infections such as meningitis need to be ruled out urgently. The spinal fluid is taken from the back through a lumbar puncture, and protein, sugar and blood cell levels, bacteria and viruses, and signs of inflammation are assessed in a laboratory.

## **Treatment**

### ***Meningoencephalitis***

If the parenchymal (brain or spinal cord) problem is not severe and is improving on its own, then it is safe with careful monitoring to allow it to get better on its own without treatment. Most cases, however, are sufficiently severe to warrant hospitalisation, and under these circumstances patients usually require treatment with intravenous injections of steroids. This has the effect of reducing the inflammation and swelling within the nervous system and prevents it from coming back at least in the short term. Some particularly severely affected patients require more complex treatments, and we sometimes give chemotherapy drugs to suppress the immune system, such as cyclophosphamide. Nowadays, we sometimes also give modern 'biologic' drugs such as infliximab or Adalimumab.

In around 30% of cases, the condition returns after a time, and a relapse occurs. This requires more steroids; however, as it is likely that more than one relapse will occur, it is best in most cases to add in an immunosuppressant drug to suppress the inflammation in the long term, and we use drugs such as azathioprine or mycophenolate to do this. These drugs need to be monitored carefully within a specialist Behçet's disease clinic.

Patients with seizures sometimes take antiepileptic tablets. Physiotherapy, occupational therapy and speech therapy are very important as the patient recovers, and sometimes treatment in a neurorehabilitation unit is recommended.

### **Thrombosis**

Patients who have a venous sinus thrombosis ([disturbance in the drainage of blood from the brain because of a blockage](#)) are treated with heparin in hospital and are then transferred to warfarin for a time. We do not usually recommend long-term warfarin therapy in patients with Behçet's disease. It is not common for this condition to relapse.

Patients with intracranial hypertension ([a rise of blood pressure within the brain](#)) are usually treated with a diuretic (water) tablet, but occasionally more severely affected patients need to have a special spinal fluid drain called a shunt implanted. This requires a small operation, but it is often done under local anaesthetic.

We now always keep patients on azathioprine to prevent relapse.

### **Fatigue**

Fatigue management and a positive outlook to the disease are best. Avoidance of overtiredness and planning of the day, to allow rest before and after an activity, work well, and most find that fatigue improves and memory becomes more efficient. It's easy for doctors to prescribe and hard for the patients to do!