

## Headache disorders in Sjögren's Syndrome

**Dr Richard Davey, Specialist Registrar in Neurology at the Leeds General Infirmary.**

A variety of brain and nervous system problems have been associated with Sjögren's Syndrome. However, perhaps because of the relative rarity of Sjögren's Syndrome, the precise nature of these and the way in which they interact with the underlying condition remains poorly understood. Headaches are a good example of this uncertain relationship.

Headaches are very common in the general population. The majority of people have experienced some form of headache at some time in their lives, to the extent that patients

However, even severe and potentially disabling forms of headache such as migraine are surprisingly common, with a recent study of the UK adult population showing that almost 20% of females have migraine (and 10% of males). Because of this, it can be very difficult to work out whether headaches are, in fact, more common in patients with disorders such as Sjögren's Syndrome or SLE.

The current best evidence suggests that they are probably not more common, and that the forms of headache seen in patients with Sjögren's Syndrome are almost always of the same type as those seen in the general population. On this basis, most experts recommend that headaches in patients with Sjögren's Syndrome are diagnosed and treated in exactly the same way as those affecting the general population.

Most headache disorders can be confidently diagnosed through careful questioning, with tests such as brain scans having quite a limited role. The most common types of headache are tension-type headache (TTH) and migraine. The term TTH is somewhat misleading, in that it suggests muscle tension in the scalp or neck is the underlying problem, or that tension and stress cause this disorder. In fact, both of these features are equally common to many forms of headache. In terms of the differences between TTH and migraine, TTH tends to affect both sides of the head, be crushing or tight in nature and rarely causes other symptoms such as sickness or sensitivity to light. In contrast, migraine is often more severe, often affects one side of the head only, is throbbing in nature and is associated with sickness, sensitivity to light and noise, and dizziness. Some unlucky patients have both these types of headaches occurring at different times!

The term 'chronic headache' is used to describe headaches that occur on more than 15 days per month. These usually develop in people with a background of TTH or migraine, where headaches gradually become more frequent and can end up being continuous. The two important factors that tend to lead to this process are psychological disorders (depression and anxiety) and overuse of painkillers. The latter problem is particularly relevant to those with painful rheumatological conditions involving arthritis.

Occasionally, more dramatic forms of headache are seen in conditions such as Sjögren's Syndrome. These can include

blood clots in the veins inside the head and meningitis, which involves inflammation of the protective covering layers surrounding the brain.

These, thankfully, are very rare and generally cause symptoms that are quite distinct from common types of headache such as migraine. Common misconceptions include the worry that strokes often present as a headache (they very rarely do) and that brain tumours often present with headache in the absence of other, associated symptoms (actually, they almost always present with other problems, such as epilepsy, weakness or loss of balance).

Occasionally, tests including brain scans are used to aid the diagnosis of headache. Modern scans, especially MRI, have become almost too good at picking up minor abnormalities. This is especially relevant in conditions such as Sjögren's Syndrome and SLE, where small white dots or patches on MRI scans are very commonly seen. However, the underlying cause for these is not well understood, but it may involve 'ageing' of blood vessels (similar scan appearances are seen in many entirely healthy people who are over the age of 60 years). These dots have no known relevance to headaches, indeed migraine and TIH are disorders of the function of the brain, and therefore are not associated with visible changes in the structure of the brain that would be visible on a scan. Therefore, in some cases having an unnecessary brain scan can show up non-specific or irrelevant abnormalities which cause lots of needless worry.

Although there have been a number of exciting developments in the treatment of headache in recent years, often treatment relies on simple, common-sense measures. Headaches, particularly migraine, can often be triggered by certain everyday things, including a number of foods, alcohol, stress, bright light and lack of sleep. Trigger factors are often specific to each different person, so that each headache sufferer should consider what environmental factors could be involved in their own headaches. Writing some form of headache 'diary' or record is surprisingly helpful for this purpose.

Another important aspect of headache treatment is taking painkilling treatment as near to the start of the headache attack as possible. Drinking lots of fluid with a tablet will help it to be absorbed into the system quickly (or, instead, take it in soluble form), and if vomiting is a problem then anti-sickness treatment taken alongside a painkiller will often avoid the problem of vomiting the painkiller back up.

The treatment of migraine can be divided up into painkilling drugs (taken when necessary at the time of an attack) and preventative treatment (taken on a daily basis to reduce the number of headaches occurring in the first place). Simple safe painkillers are usually the first treatment to try. A single large dose at the start of the attack is the most effective treatment, for example ibuprofen at a dose of 800mg. In general,

*Continued next page*

codeine-containing drugs are not recommended. If simple drugs don't work well, you should speak to a doctor about a group of drugs called 'triptans'.

Triptans have been a revolution in migraine treatment, and typically lead to complete or near-relief from most migraine headaches within 1-2 hours of taking them. A number of different triptans are now on the market-if the first one you try doesn't work well then try another type.

Preventative treatment is usually recommended if headaches occur more than 4 times a month, or are particularly long-lasting or severe. A number of different drugs can be effective for this purpose. These include Propranolol, Amitriptyline, Pizotifen, Topiramate and Valproate. There is also some evidence suggesting that the herbal remedies Feverfew and Butterburr are useful alternatives. Generally, these preventative drugs are started at a low dose and increased gradually depending on their effect. They often take a few weeks to work, so that you should expect to need to give them a good long trial period before concluding that they have not helped.

Regarding chronic headaches, treatment of any underlying depression is often helpful. This may involve drug or behavioural therapy. Painkiller overuse is usually a key factor that needs to be addressed.

As a rule of thumb, if you are taking more than 10-15 doses of any painkiller per month then this is likely to make headache attacks occur more frequently. Many people take many more doses than this, leading to a state of nearaddiction to painkillers.

We generally find that the gradual withdrawal of all (or nearly all) painkillers results in a reduction in the number of headaches. In addition, treatments such as the triptans and preventers described above do not tend to work if painkiller overuse is present.

In summary, it is recognised that headaches are a common and troublesome complaint in patients with connective tissue disorders, including Sjögren's Syndrome. An expert assessment, via referral to a neurologist if necessary, will often mean a specific diagnosis can be made and a range of effective treatments can be recommended. A number of charitable organisations such as 'The Migraine Trust' now exist to advise you. Most of these can be accessed using the internet (try a search using 'Google').