

SJÖGREN'S SYNDROME

There are more than half a million sufferers of Sjögren's Syndrome in the UK, yet the condition remains under recognised and frequently under treated. It is the UK's second most common autoimmune rheumatic disease, but because it isn't a life threatening condition it does not command a high profile within the medical profession, and the general public is largely unaware of the problems faced by sufferers. In reality, Sjögren's Syndrome is a debilitating, distressing and miserable condition.

In Sjögren's Syndrome the secretory glands that produce saliva, tears, vaginal, skin, bowel and other secretions develop a form of inflammation, similar to that seen in the joints in patients with rheumatoid arthritis. As a result of this, these glands stop working leading to dry eyes, dry mouth, dry skin and also dryness of the gastrointestinal tract. These features, as well as being very uncomfortable and unpleasant, means that sufferers may have difficulty swallowing dry foods and also dryness of the large bowel can cause symptoms similar to irritable bowel Syndrome (IBS).

Sjögren's Syndrome is an autoimmune disease, in other words it is a condition where the body's immune system starts attacking parts of the body instead of fighting infection, this is mainly the white cells, which travel in the blood stream. In the case of Sjögren's Syndrome, they attack the glands which produce tears, saliva, skin secretions etc.

Sjögren's Syndrome can occur later in the course of other rheumatic conditions, such as rheumatoid arthritis, lupus or scleroderma, this is known as Secondary Sjögren's Syndrome. Most patients with these conditions will already be under the care of a rheumatologist who will usually make the diagnosis of Sjögren's Syndrome as soon as the dryness symptoms are mentioned.

It can also occur, however, as a condition in its own right (Primary Sjögren's Syndrome). It typically develops in women in their 30's to 60's, although it can occur at any age and affects both men and women. Features such as generalized muscle or joint aching and generalized fatigue can be the most prominent symptoms. The fatigue isn't an ordinary tiredness where you can have a good nights sleep and wake up feeling better, it's a really quite overwhelming fatigue. In the situation where a person's main symptom is tiredness and particularly if the dryness is not reported, the diagnosis may not be made for a long time, if at all. As a result of this, although the evidence suggests that Sjögren's Syndrome is as common as rheumatoid arthritis, there are a significant proportion of people in whom it is not being correctly diagnosed and symptoms are put down to "old" age for example, and people are suffering in silence.

- ◆ It affects approximately 3-4% of adults in the UK¹, with a mean age of 50 years.
- ◆ 90% of patients are women.

The clinical presentation of Sjögren's Syndrome is variable, but typical symptoms include;

- ◆ dry itchy, irritated eyes
- ◆ dry mouth and difficulty swallowing
- ◆ joint pain and muscle aches
- ◆ low mood, irritability and impaired concentration
- ◆ disabling fatigue

A number of studies have shown that the health-related quality of life of patients with primary Sjögren's Syndrome is significantly decreased and is comparable with that of patients with a condition like Systemic Lupus Erythamatosus (SLE).

Despite being a common disorder, Sjögren's Syndrome has not commanded a high profile and has been under-diagnosed and many patients have gone untreated, causing unnecessary pain and suffering. There are serious complications for some patients such as neuropathies, loss of teeth and lymphoma, and permanent damage or ulceration to the eye, which can lead to sight loss. Prompt diagnosis and treatment can prevent these complications from happening.

MAJOR SYSTEMIC COMPLICATIONS

These are some of the complications that can affect a sufferer:

- ◆ Arthritis, Raynaud's syndrome and fatigue
- ◆ Lung involvement
- ◆ Kidney involvement
- ◆ Neurological involvement
- ◆ MALT Lymphoma (44 times increased risk)

BARRIERS TO DIAGNOSIS & THERAPY

These are some of the problems a sufferer can come across:

- ◆ Patients present to different specialties, due to their varied symptoms
- ◆ Often non-specific symptoms
- ◆ Long lead time before diagnosis
- ◆ Often partial diagnosis only
- ◆ Undiagnosed patients not receiving appropriate therapy

GENERAL ADVICE

- ◆ Encourage regular dental review (6 monthly)
- ◆ Good dental hygiene (brushing, flossing, fluoride)
- ◆ Avoid excess sweet foods
- ◆ Avoid strong soaps (use aqueous creams and emollients)
- ◆ Avoid dry environments
- ◆ Avoid cigarette smoking and excess alcohol
- ◆ Receive information about self-help groups (e.g. BSSA)

REFERENCES

- ¹ Jonsson R, Haga H-J, Gordon T: Sjögren's syndrome. In: Arthritis and Allied Conditions – A Textbook of Rheumatology, Koopman WJ, ed. 14th Edition. Lippincott Williams & Wilkins, Philadelphia, 2001, pp1736-1759