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What is stiff person syndrome?

Singer Céline Dion was forced to cancel her upcoming world tour due to complications from stiff person syndrome. We explore how this extremely rare disorder can cause daily pain and discomfort for those affected, and how triggers are often hard to avoid.

What is stiff person syndrome?

Stiff person syndrome (SPS) is a very rare, long-term condition that causes painful muscle spasms and debilitating muscle stiffness. Scientists don't yet fully understand stiff person syndrome, but it's thought to be an autoimmune disorder - where [your immune system](#) attacks healthy cells - involving the messages sent between your brain and spinal cord¹.

Who does stiff person syndrome affect?

Those most likely to have stiff person syndrome are:

- Born female.
- Between 30 and 60 years of age - although it can also affect children and the elderly.
- Those with another autoimmune disease - such as [type 1 diabetes](#), [vitiligo](#), or [coeliac disease](#).

This condition is so rare that it is hard to know how many people suffer from it. It is thought that it can affect around one in one million people².

Stiff person syndrome was originally called stiff man syndrome, until the name was updated to reflect that this disorder can affect people of any age or biological sex. Although, women are twice as likely to develop this disorder as men¹.

Is stiff person syndrome painful?

When Céline Dion first took to social media to share her diagnosis, she explained that her muscle spasms affected every aspect of her daily life, making normal activities like walking a struggle. Céline has since cancelled her 'Courage World Tour' in order to build back her strength.

The severity of stiff person syndrome varies from person to person, but the two main symptoms are the same:

- **Muscle stiffness** - also called muscle rigidity, usually affects your back, chest, and abdomen and sometimes your arms and legs.
- **Muscle spasms** - can occur in one specific area or your entire body, and may last anywhere between a few seconds and, occasionally, a few hours.

Muscle stiffness causes aches and pains that can feel better or worse at different times, without a clear trigger. They may also cause pain through associated complications, such as developing an abnormal posture.

Muscle spasms cause a sharp pain. Unlike with the stiffness, these can have predictable triggers - some may be avoided to a certain extent, but not all of them can. For this reason, some people with stiff person syndrome develop [anxiety](#).

Muscle spasm triggers:

- Unexpected or loud noises - as a result, some develop [agoraphobia](#), a fear of crowded places or of leaving home.
- Stress.
- Light physical contact.
- Changes in temperature - especially if it becomes cold.

Is stiff person syndrome life threatening?

Death associated with this neurological disorder is rare. When it does happen, it's not a direct result of stiff person syndrome, but rather its complications. These complications could include severe injury, wound infections, or blood clots caused by immobility issues.

Can stiff person syndrome be treated?

Stiff person syndrome is a chronic condition, meaning you have it for the rest of your life. While there is no cure, treatment can help to ease symptoms, improve quality of life, slow its progression, and reduce the risk of other health complications.

The first step is getting a diagnosis, but as the condition is so uncommon that it's easy to mistake for other conditions with similar symptoms, such as [multiple sclerosis \(MS\)](#). However, a doctor can confirm stiff person syndrome through a [blood test](#) that checks for associated antibodies, electromyography (EMG) that tests muscle activity, and a [spinal tap](#) that looks for antibodies in your spine.

The two main treatment strategies are:

Symptom management

A person's life may be made easier with medications that help to soothe painful muscle spasms and relax muscle stiffness. [Diazepam](#) is often the first medication offered for stiff person syndrome because it can help alter awry signals sent between the brain and muscles.

Certain pain medications can also affect these signals and effectively ease pain in some people. [Muscle relaxants](#), as the name suggests, work by loosening your muscles, which can help treat both muscle stiffness and spasms.

Several therapies may also help people continue daily activities as much as possible. These include [physiotherapy](#), [heat therapy](#), acupuncture, [massage](#), and hydrotherapy.

Disease-modifying treatment - recent progress

There's some evidence that a type of immunotherapy called intravenous immunoglobulin (IVIg) treatment can improve symptoms, although this isn't the case for everyone. This involves people with healthy immune systems donating antibodies which are then transferred via a needle or catheter to someone with stiff person syndrome.

In 2022, the [National Institute of Neurological Disorders and Stroke \(NINDS\)](#) conducted a study that showed promising results for this technique³. Many participants felt less stiff, became less sensitive to triggers like noise, touch, and stress, and found their balance improved¹.

Another treatment underwent a small, preliminary study in 2022. Although research is ongoing, it gave hope that a therapeutic plasma exchange could be a safe new treatment method in the future. Here, a person's blood plasma is replaced with albumin, a manmade substance that can treat certain diseases⁴.

Stiff person syndrome may not be curable, but with ongoing research there is hope that this disorder can become more manageable and less debilitating in the future.

Further reading

1. [National Institute of Neurological Disorders and Stroke \(NINDS\): Stiff person syndrome.](#)
2. [Rare Diseases: Stiff person syndrome.](#)
3. [NINDS: Intravenous immunoglobulin \(IVIg\) for the treatment of stiff man syndrome \(SMS\).](#)
4. [American Academy of Neurology: Preliminary study: bloodyndrome.](#)

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