



STEVENS-JOHNSON SYNDROME (SJS) AND TOXIC EPIDERMAL NECROLYSIS (TEN) IN CHILDREN AND YOUNG PEOPLE

Written for young people (ages 7 - 16) and their carers

What is this leaflet for?

This leaflet has been written to help young people understand SJS (Stevens-Johnson syndrome) and TEN (toxic epidermal necrolysis). SJS and TEN are two names that describe a reactive skin condition – whilst both involve the skin, SJS describes cases with less skin involvement whereas TEN is used in cases with more skin involvement. The two names can be used together to describe the condition.

What is SJS/TEN?

SJS/TEN is a rare and serious skin condition. It occurs from an over reaction of the body's immune system which then affects the skin as well as other parts of the body, including the eyes and the inside of the mouth, nose, throat and genitals (vulva or penis).

Why have I got this condition?

SJS/TEN can be caused by your body reacting severely to an infection, such as a virus or bacteria from a chest infection or cold, or reacting severely to a particular medicine you have taken. Sometimes, it isn't clear what has caused SJS/TEN. SJS/TEN is not infectious, meaning you cannot catch or pass on the skin reaction by touching other people.

What are the symptoms?

The first signs of SJS/TEN often include a sore throat, fever and red, sore eyes. Over the next few days, a painful red, purple or dark rash appears on the skin, often in the shape of circles – the colour of the rash will vary depending on the colour of your skin. The skin, mouth, lips, eyes and genitals may also become sore and ulcers may form. Ulcers in the mouth can make swallowing painful and ulcers in the genitals may make it difficult and painful to pass urine. The eyes may also become red and sore, and this may also affect your vision. The skin may start to blister and peel, and this might spread across the skin over a few days.

How is SJS/TEN treated?

SJS/TEN can be serious and people with this condition will generally stay in hospital for treatment. If a drug is suspected to have caused SJS/TEN, doctors will stop the medication. If an infection is suspected to have caused SJS/TEN, doctors may give medicines such as antibiotics to help treat the infection. There is no single cure for SJS/TEN and often it means waiting for the body to heal. The skin normally takes one or two weeks to heal, but new sore areas could form for several weeks afterwards.

What tests do I need?

Doctors will do tests to try to understand what has caused SJS/TEN. This can include blood tests, a swab inside the mouth, swabs of the skin and sometimes taking a very small sample of the skin. If a medicine is suspected to have caused SJS/TEN, some people may need more tests after they have recovered to confirm they have reacted to the medicine.

What might happen in hospital?

Whilst in hospital, people with SJS/TEN will be cared for by several teams of doctors, nurses and other health-workers. The team will explain anything that needs to be done with you and your carers. The goal of treatment is to support the body as it recovers by making sure people have enough water, have good nutrition, are breathing freely and have as little pain as possible. Sore areas of skin may be covered with a dressing or creams to protect them, and eyedrops may be given to protect the eyes. Medicines to relieve any pain will also be given. A tube may be passed into the bladder to help pee (catheter) and a tube may be inserted into the stomach to help with nutrition (nasogastric/ NG tube). In severe cases, people may need more intensive care including support with breathing.

What can be expected after SJS/TEN?

Most people will find that their skin will slowly heal and grow back over a week or two – it may be a different colour to the rest of the skin as it heals. The skin and eyes will be sensitive to sunlight as they are healing so it is important to put on sun cream and wear sunglasses when outside.

Some people might find that they have symptoms from their SJS/TEN that last a long time. This can include skin scarring or colour changes to the skin and this can be upsetting. Inflammation in the eyes, mouth and genitals might also cause symptoms from scarring such as changes to eyesight, changes to breathing such as a long-lasting cough, or difficulty passing urine. It is also normal to feel tired for weeks and even months after having SJS/TEN. If you have had SJS/TEN and feel you are not getting better, it is good to talk about it with your parents or carers.

Will SJS/TEN happen again?

Most young people will recover from SJS/TEN and will only ever have one episode.

However, occasionally, some people may have more than one episode of SJS/TEN. Any specific medicines that have caused SJS/TEN should be avoided in the future so they do not cause another reaction. This may involve being given a bracelet to wear that alerts people to which medicines to avoid. SJS/TEN can reoccur with a repeat infection or, rarely, a different medication. Recognising the symptoms of SJS/TEN will help to seek treatment again quickly if it does happen for another time.

Having SJS/TEN has upset me, is this normal?

Being in hospital can be very upsetting, and it is not unusual to feel low or anxious after having SJS/TEN. If you feel upset about SJS/TEN, it is good to talk to your parents or carers about it.

A NOTE FOR PARENTS OR CARERS OF CHILDREN WITH SJS/TEN

About SJS/TEN:

As SJS/TEN is rare, there is still a lot about the disease process and treatment that is not fully understood. There is ongoing research to try and understand whether environmental and/or genetic factors predispose SJS/TEN and why it reoccurs in some children and not others. There is also ongoing research to understand how we can treat it early and effectively, and work to prevent long term symptoms.

In addition to SJS/TEN, there are also some other words that you might hear used which refer to the same conditions but refer to the likely cause of the reaction. DEN (drug induced necrolysis) syndrome describes severe TEN-like reactions triggered specifically by drugs. RIME (reactive infective mucocutaneous eruption) describes SJS-like reaction particularly involving the surface of the mouth and seen as a reaction to infections including a respiratory infection *Mycoplasma pneumoniae*. These terms may be used interchangeably with SJS/TEN.

During your child's admission:

Having a child in hospital is difficult for both carers and child.. Whilst in hospital, your child is likely to be managed by several different teams and at times this can feel overwhelming with lots of information. The uncertainty around what causes SJS/TEN and how it is managed can also be difficult. If you have questions or uncertainties about your child's treatment plan, you should share this with the clinical team taking care of your child.

After your child's admission:

The recovery for every child will be different, both in terms of recovery time as well as the degree of any lasting symptoms. Being vigilant to ongoing symptoms, both physical and psychological, can help identify any complications from SJS/TEN early. Long-lasting symptoms most commonly involve the skin and the eyes but can also involve the lungs and the genitals as outlined above. There is also increasing awareness that SJS/TEN can have a psychological impact on your child, affecting

their mood or anxiety levels. If you are worried your child might be experiencing any on-going symptoms, you should raise these at follow-up clinics or with your GP.

It is natural to worry about your child having SJS/TEN a second time. However, it is estimated that 4 out of 5 children only ever have SJS/TEN once. It is not currently feasible to predict which children will go on to have a second episode. That said, fear of recurrence should not be a barrier to your child enjoying a full and active life. If you are worried, it is important to familiarise yourself with the symptoms and signs of SJS/TEN (as outlined above) to identify a reoccurrence of the condition early on. If you would like further information, the doctor arranging follow-up care should be able to help with any questions you might have and advise you of reliable online sources or accessible research studies

WHERE CAN I FIND OUT MORE?

Further information for kids and parents:

- Great Ormond Street Hospital for Children – Stevens-Johnson syndrome (this page is written for SJS specifically but it is also useful for TEN as well): <https://www.gosh.nhs.uk/conditions-and-treatments/conditions-we-treat/stevens-johnson-syndrome/>

Further information for parents:

- NHS UK – Stevens-Johnson syndrome: <https://www.nhs.uk/conditions/stevens-johnson-syndrome/>
- The British Association of Dermatology's SJS/TEN patient information for adults: <https://www.skinhealthinfo.org.uk/condition/sjs-ten/>
- The British Association of Dermatology's national guidelines for SJS/TEN in children and young people: <https://onlinelibrary.wiley.com/doi/epdf/10.1111/bjd.17841>

For details of source materials used please contact the Clinical Standards Unit (clinicalstandards@bad.org.uk).

This leaflet aims to provide accurate information about the subject and is a consensus of the views held by representatives of the British Society for Paediatric Dermatology which is a part of the British Association of Dermatologists. Individual patient circumstances may differ, which may alter both the advice and treatment given by your doctor

This leaflet has been assessed for readability by the British Association of Dermatologists' Patient Information Lay Review Panel

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PATIENT INFORMATION LEAFLET**

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