

Stevens-Johnson syndrome – Toxic Epidermal Necrolysis **Patient information leaflet**

What are the aims of this leaflet?

This leaflet has been written to help you understand SJS (Stevens-Johnson syndrome) and TEN (toxic epidermal necrolysis, also known as Lyell syndrome). These two names are used to describe the milder form, SJS, and the severe form, TEN, of the same condition. The two names are often used together, SJS-TEN, to describe all cases. The first part of the leaflet tells you what SJS-TEN is, what causes it, and how it is managed while you are in hospital. The second part of the leaflet explains what to expect in the period of recovery once you leave hospital, both in the medium and the long-term.

FIRST PART

What is SJS-TEN?

SJS-TEN is a rare, severe skin reaction which comes on suddenly. It is characterised by a blistering rash in combination with inflammation of the eyes, mouth, nose and genitalia. The outermost layer of the skin (called the epidermis) can become detached leaving the layer beneath (the dermis) exposed. Internal structures such as your upper airways, lungs and digestive tract may also be affected.

What causes SJS-TEN?

In adults this is usually a reaction to medication. However, in children both infection and medication are important triggers.

Almost any medication can cause SJS-TEN including over-the-counter medications and herbal/natural preparations. Medications that are often implicated in SJS-TEN are anti-epileptic drugs, antibiotics and allopurinol. SJS-TEN may occur 4 to 28 days after starting the medication.

Infections that can commonly cause SJS-TEN in children include the herpes simplex (cold sore) virus, *Mycoplasma pneumoniae*, and other respiratory tract infections. Sometimes there is no clear trigger identified. It is not clear why some people react in this intense manner, but we believe it may be due to an extreme allergic response.

Is SJS-TEN an infectious disease and can it be transmitted?

No, it cannot be transmitted. Even in cases caused by an infection, the infection is often over by the time SJS-TEN occurs as it is a delayed reaction to the infection. Sometimes, visitors of SJS-TEN patients may be asked to wear gloves and gowns. This is to reduce the patient's risk of acquiring an infection.

Is SJS-TEN hereditary?

For some medicines such as carbamazepine and allopurinol, the risk of developing SJS-TEN is higher in some people (especially from Asia) who have susceptibility genes. It is possible that more susceptibility genes may be discovered for other drugs in the future.

What are the symptoms of SJS-TEN?

The earliest features of SJS-TEN include flu-like symptoms: sore throat, fever, and red, sore eyes. Over the next few days a painful red or purple rash appears, accompanied by inflammation and erosions of the mouth, lips, eyes and genitalia. The rash usually starts to blister at this stage. The outermost layer of skin (epidermis) may detach spontaneously or with friction, leaving the layer beneath of skin (dermis) exposed and appearing bright red, bleeding easily and feeling painful. The amount of skin involvement and degree of blistering extends over the following days.

What does SJS-TEN look like?

Blistering in SJS-TEN occurs on skin which is red or purple. In SJS, the blistering involves less than 10% of the body surface area (BSA); in TEN the blistering is much more extensive and will involve more than 30% of the BSA. The blistering reaches its maximum extent around one week after its onset. Healing of the skin may take 10-15 days. The texture of the skin gradually returns to normal but discolouration of the skin is common and may be persistent.

Involvement of the mouth can result in extensive ulceration which is usually associated with crusted and bleeding lips. The affected mucous membranes may take a longer time than skin to improve and heal. Involvement of the eyes can cause redness, weeping and crusting of the eyelids. The surface of the eye may also be affected causing redness, dryness and in some cases, scarring, that can affect temporarily or more sustainably visual acuity. Hair and nails may be shed but they

often grow back. However, regrowth may take some time, often after the skin has healed.

What is the outlook (prognosis) in SJS/TEN?

SJS/TEN is a serious illness, and while most patients do recover, the most severe cases can lead to death. The severity may not be fully apparent when you first come to hospital, but may evolve over a number of days. Specialists will monitor your skin, mouth, eyes and genital surfaces regularly to check for involvement. They will also monitor your blood tests and functioning of internal organs such as the liver, lungs and kidneys to check for progression of the disease. A number of factors, including age, pre-existing diseases such as cancer, the extent of skin loss, and the results of certain blood tests are known to be significant in the evaluation of prognosis.

How will SJS-TEN be diagnosed?

There are no specific blood tests for SJS-TEN. A definitive diagnosis of SJS-TEN is made from the doctors' assessment of the skin, usually along with a skin biopsy to confirm the diagnosis and to exclude other skin diseases that may present in a similar way. A biopsy involves taking a very small sample of skin and examining it with a microscope, a process which may take some days.

How can SJS-TEN be treated?

Early treatment is important. If a drug is suspected to be the cause, this must be stopped immediately. You will be admitted to hospital, and if your skin loss is extensive, you will be transferred to a burns or intensive care unit.

Your care will require the input from a number of specialties, which may include dermatology, intensive care, burns or plastics surgery, ophthalmology, ear, nose and throat (ENT), psychiatry, respiratory medicine and renal medicine. Other clinicians involved may include dietitians, physiotherapists and psychologists. You will need intensive skin, ocular and mouth care from specialist nurses.

Skin care involves the application of ointments or creams and application of dressings. Mouthwashes that contain antiseptics and pain-relieving agents may be used several times per day. Ointment-based moisturisers may be applied to the lips frequently. Ophthalmologists may recommend the application of eye ointments or drops.

In the initial stage of your admission, particularly if your skin loss is extensive, the room in which you are nursed may be at a higher temperature and humidity than normal until your skin heals. You may have an intravenous line inserted for hydration, pain relief and other medicines. There is no clear current evidence to show the benefit of any specific medicines for SJS-TEN. A feeding tube may be inserted through the nose into the stomach to ensure adequate nutrition. A urinary catheter and faecal tube may be inserted to help with care of the skin in the genital area and to measure urine output. Machines may be used to monitor your heart rate, blood pressure and blood oxygen saturations.

SECOND PART

After discharge from hospital, what should you expect?

Your skin, mouth and eyes may continue to bother you after discharge from hospital. As your skin and eyes may be more sensitive in general and to the sun for many months, we would recommend the daily use of moisturisers, covering up and/or using sunscreen on areas of skin that may be exposed to bright sunlight, and wearing sunglasses when outdoors. Discolouration of your skin may persist for a long time. The genital skin may be affected in both men and women, causing discomfort and/or pain during sexual intercourse. Topical treatments (creams, ointments, etc) may help in the first instance but more specialist interventions from specialists in gynaecology or urology may be considered. These issues may be addressed after your discharge from hospital in the dermatology clinic or other specialist services.

Loss of hair may occur during or after the skin disease and it may take months before there is hair regrowth. Loss of nails may also occur and it may take many months for them to grow back (longer time for regrowth of toenails compared to fingernails). The nails that regrow may be brittle and not as they were before. It is important to avoid injury to your nails by protecting your hands with gloves when doing housework or other work that may injure your nails.

Dry mouth and sensitivity to certain foods and toothbrushing is common and may persist for some time. More severe problems of the mouth may require specialist care after discharge from hospital. Dry eyes and sensitivity to light is also common. If there is scarring of the surface of the eyes, or regrowth of eyelashes inwards, more specialist care may be needed. Please ensure that you attend these specialist clinic appointments after discharge from hospital.

You may feel tired for weeks or months after discharge from hospital. It is important to rest but also to resume as much gentle activity as possible. It is normal to feel low in mood when recovering from a serious illness. Some may experience flashbacks, nightmares, fear of medicines and have difficulty resuming normal life. If these feelings persist or becomes worse, please let your healthcare providers know so that they may help you get help.

Problems with breathing due to issues with airways or lungs after discharge from hospital may persist in a small number who have had SJS-TEN and may require further monitoring or interventions from specialist services such as ear, nose and throat or respiratory medicine doctors.

Please keep a record of the medicine that caused SJS-TEN and inform your healthcare providers. We advise that you wear a medical alert device such as a bracelet, allergy card or pendant with this information. Please do not introduce any new medicines, including over-the-counter medicines, without a discussion with your healthcare provider. We would advise avoidance of herbal or natural medicinal preparations as they are not formally regulated and it is difficult to know exactly what they contain. In children where infection was the cause of SJS-TEN, there is a small chance that it may recur therefore it is important to seek medical attention promptly if this happens.

For details of source materials used please contact your dermatologist.

This leaflet aims to provide accurate information about the subject and is a consensus of the views held by representatives of the Toxic Bullous Dermatoses ERN-skin subgroup. Individual patient circumstances may differ, which might alter both the advice and course of therapy given to you by your doctor.

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