NHS (Link: www.nhs.uk/)

Stevens-Johnson syndrome

Stevens-Johnson syndrome is a rare but serious disorder that affects the skin, mucous membrane, genitals and eyes.

The mucous membrane is the soft layer of tissue that lines the digestive system from the mouth to the anus, as well as the genital tract (reproductive organs) and eyeballs.

Stevens-Johnson syndrome is usually caused by an unpredictable adverse reaction to certain medications. It can also sometimes be caused by an infection.

The syndrome often begins with flu-like symptoms (Link: www.nhs.uk/conditions/flu/symptoms/), followed by a red or purple rash that spreads and forms blisters. The affected skin eventually dies and peels off.

Stevens-Johnson syndrome is a medical emergency that requires treatment in hospital, often in intensive care (Link: www.nhs.uk/conditions/intensive-care/) or a burns unit.

Treatment aims to identify the underlying cause, control the symptoms and prevent complications.

Erythema multiforme (Link: www.nhs.uk/conditions/erythema-multiforme/) is a similar, but less severe, skin reaction that's usually caused by infection, particularly herpes viral infections, and chest infections (Link: www.nhs.uk/conditions/chest-infection/).

Symptoms of Stevens-Johnson syndrome

Skin pain is the most common symptom of Stevens-Johnson syndrome.

Flu-like symptoms (Link: www.nhs.uk/conditions/flu/symptoms/) are also usually present during the initial stages, and may include:

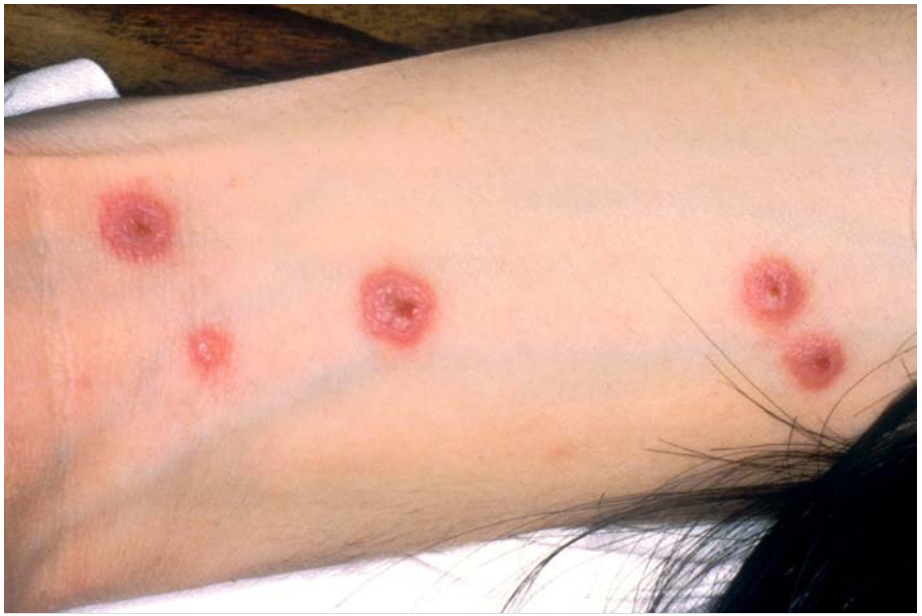
- feeling generally unwell
- a high temperature (fever) of 38C (100.4F) or above
- a headache (Link: www.nhs.uk/conditions/headaches/)
- joint pain

- a cough (Link: www.nhs.uk/conditions/cough/)

After a few days a rash appears, which consists of individual blemishes that may look like a target – darker in the middle and lighter around the outside.

The rash isn't usually itchy, and spreads over a number of hours or days.

Large blisters then develop on the skin, which leave painful sores after bursting.



Facial swelling and swollen lips covered in crusty sores are common features of Stevens-Johnson syndrome.

The mucous membranes inside your mouth, throat, eyes and genital tract may also become blistered and ulcerated.

This can make swallowing painful and lead to serious problems such as dehydration (Link: www.nhs.uk/conditions/dehydration/).

The surface of the eyes can also sometimes be affected, which can cause corneal ulcers and vision problems if not treated quickly.

Causes of Stevens-Johnson syndrome

In children, Stevens-Johnson syndrome is usually triggered by a viral infection, such as:

- mumps (Link: www.nhs.uk/conditions/mumps/)
- flu (Link: www.nhs.uk/conditions/flu/)
- herpes-simplex virus, which causes cold sores (Link: www.nhs.uk/conditions/cold-sores/)
- Coxsackie virus, which causes Bornholm disease (Link: www.nhs.uk/conditions/bornholm-disease/)
- Epstein-Barr virus, which causes glandular fever (Link: www.nhs.uk/conditions/glandular-fever/)

Less commonly, bacterial infections can also trigger the syndrome.

In adults, Stevens-Johnson syndrome is often caused by an adverse reaction to medicine.

The medicines that most commonly cause Stevens-Johnson syndrome are:

- allopurinol
- carbamazepine
- lamotrigine
- nevirapine
- the "oxicam" class of anti-inflammatory drugs (including meloxicam and piroxicam)
- phenobarbital
- phenytoin
- sulfamethocazole and other sulfa antibiotics
- sertraline
- sulfasalazine

It's important to emphasise that Stevens-Johnson syndrome is rare and the overall risk of getting the syndrome is low, even for people using these medicines.

Risk factors

Risk factors for Stevens-Johnson syndrome may include:

- **viral infections** – such as herpes, hepatitis, viral pneumonia (Link: www.nhs.uk/conditions/pneumonia/) or HIV
- **a weakened immune system** – as a result of HIV or AIDS (Link: www.nhs.uk/conditions/hiv-and-aids/), autoimmune conditions, such as lupus (Link: www.nhs.uk/conditions/lupus/), or certain treatments, such as chemotherapy (Link: www.nhs.uk/conditions/chemotherapy/) and organ transplants

- **a previous history of Stevens-Johnson syndrome** – if the syndrome was previously caused by medication, you're at risk of it reoccurring if you take the same medication again, or medications from the same family of medications
- **a family history of Stevens-Johnson syndrome** – if a close family member has had the syndrome, your risk of getting it may be increased

Specific genes have also been identified that increase the risk of Stevens-Johnson syndrome among certain groups of people.

For example, Chinese people with the HLA B1502 gene have experienced Stevens-Johnson syndrome after taking carbamazepine, and allopurinol has also triggered the syndrome in Chinese people with the HLA B1508 gene.

Diagnosing Stevens-Johnson syndrome

Stevens-Johnson syndrome should be diagnosed by a dermatologist (skin specialist).

A diagnosis is often based on a combination of your:

- symptoms
- physical examination
- medical history (including any medication you have taken recently)

To confirm the diagnosis, a small sample of skin may be removed (biopsy) (Link: www.nhs.uk/conditions/biopsy/) so it can be tested in a laboratory.

Treating Stevens-Johnson syndrome

If Stevens-Johnson syndrome is suspected, you or your child will be immediately referred to hospital for treatment.

Without treatment, the symptoms can become very severe and be life threatening.

Severe cases of Stevens-Johnson syndrome may need to be treated in an intensive care unit (ICU) (Link: www.nhs.uk/conditions/intensive-care/) or burns unit.

The first step is to stop taking any medications that may be causing Stevens-Johnson syndrome.

But it can sometimes be difficult to determine which medication is causing it, so stopping all non-essential medications may be recommended.

Treatment for relief of symptoms while in hospital may include:

- **strong painkillers** – to help ease the pain of any raw areas of skin

- **cool, moist compresses held against the skin** – dead skin may be gently removed and a sterile dressing placed over the affected area
- **regularly applying a plain (unscented) moisturiser to the skin**
- **replacement fluids** – you may receive fluids and nutrition through a tube that's passed through your nose and into your stomach (a nasogastric tube) (Link: www.nhs.uk/conditions/swallowing-problems-dysphagia/treatment/#feeding tubes)
- **mouthwashes containing anaesthetic (Link: www.nhs.uk/conditions/anaesthesia/) or antiseptic** – to temporarily numb your mouth and make swallowing easier
- **a short course of corticosteroid tablets (topical corticosteroids) (Link: www.nhs.uk/conditions/topical-steroids/)** to control skin inflammation (only on specialist advice)
- **antibiotics** (Link: www.nhs.uk/conditions/antibiotics/) – if blood poisoning (sepsis) (Link: www.nhs.uk/conditions/sepsis/) is suspected
- **eyedrops or eye ointment** – for eye-related symptoms

Once the cause of Stevens-Johnson syndrome has been identified and successfully treated (in the case of an infection), or stopped (in the case of medication), the skin reaction will stop. New skin may start to grow after a few days.

But the length of time it takes to recover from Stevens-Johnson syndrome will depend on how severe it is, and it can sometimes take many weeks or months to fully recover. It's common to feel tired and lack energy for several weeks after being discharged.

If the cause was an adverse reaction to medication, you'll need to avoid that medication and possibly other similar medications for the rest of your life.

The doctor treating you will be able to advise you further about this.

Complications of Stevens-Johnson syndrome

As Stevens-Johnson syndrome severely affects the skin and mucous membranes, it can cause a number of complications.

These include:

- **skin changes** – when your skin grows back it may be uneven in colour; less commonly, scarring (Link: www.nhs.uk/conditions/scars/) may occur
- **secondary skin infection (cellulitis) (Link: www.nhs.uk/conditions/cellulitis/)** – which can lead to further serious problems, such as blood poisoning (sepsis) (Link: www.nhs.uk/conditions/sepsis/)
- **problems with internal organs** – organs can become inflamed; for example, the lungs (pneumonia) (Link: www.nhs.uk/conditions/pneumonia/), heart (myocarditis), kidneys (nephritis) or liver (hepatitis) (Link:

www.nhs.uk/conditions/hepatitis/), and the oesophagus may also become narrowed and scarred (oesophageal stricture)

- **eye problems** – the rash can cause problems with your eyes which, in mild cases, may be irritation and dry eyes (Link: www.nhs.uk/conditions/dry-eyes/), or in severe cases may result in corneal ulceration, uveitis (Link: www.nhs.uk/conditions/uveitis/) (inflammation of the uvea, which is the middle layer of the eye) and possibly blindness (Link: www.nhs.uk/conditions/vision-loss/)

Problems with the sexual organs, such as vaginal stenosis (narrowing of the vagina caused by a build-up of scar tissue) and scarring of the penis, is also a possible complication of Stevens-Johnson syndrome.

Preventing Stevens-Johnson syndrome

If Stevens-Johnson syndrome has been caused by an adverse reaction to a medication, you'll need to avoid taking this medication and other similar medications.

Other family members may also want to avoid using the medication in case there's a genetic susceptibility within your family.

If you have had Stevens-Johnson syndrome in the past and your doctor thinks you're at risk of getting it again in the future, you'll be warned to look out for the symptoms.

If you're of Chinese, southeast Asian or Indian descent, genetic testing may be recommended before taking medications known to have an associated risk of causing Stevens-Johnson syndrome, such as carbamazepine and allopurinol.

Testing will help determine whether you carry the genes (HLA B1502 and HLA B1508) that have been associated with the syndrome when taking these medications.

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