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Patient Guide to: Stevens-Johnson Syndrome (SJS) and Toxic Epidermal Necrolysis (TEN)

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PATIENT GUIDE TO:

STEVENS-JOHNSON SYNDROME &
TOXIC EPIDERMAL NECROLYSIS
(SJS-TEN)



INTRODUCTION

This patient guide was designed to help survivors and their families affected by SJS/TEN. Throughout this guide, you will find questions to ask your doctor or healthcare professional along with space at the end of the booklet to write down their answers. The content of this booklet is for informational purposes only and is not intended to diagnose, treat, cure, or prevent any condition or disease. No material in this guide is intended to be a substitute for professional medical advice, diagnosis or treatment. Please consult with your own physician or healthcare specialist regarding the suggestions and recommendations made in this guide. The use of this guide implies your acceptance of this disclaimer.

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THE FACTS ABOUT

STEVEN JOHNSON SYNDROME & TOXIC EPIDERMAL NECROLYSIS (SJS-TEN)

WHAT IS SJS & TEN?

Stevens-Johnson Syndrome (SJS) and Toxic Epidermal Necrolysis (TEN) are two severe and life-threatening reactions, commonly triggered by medications, that cause the skin tissue to develop painful blistering rashes, die and detach from the body. SJS and TEN affect the external surface of the body, as well as the mucous membranes of the mouth, nose, eyes, genitalia, gastrointestinal, and lower respiratory tracts. They are not common, so they are treated in specific Burn Centers and need a very specialized and multidisciplinary team.

DIFFERENCE BETWEEN SJS & TEN

TEN is a more severe form of SJS. Differences between the two are based on the total body surface area (TBSA) affected. Patients can be classified into three groups:

SJS covers < 10% of TBSA

SJS-TEN covers between 10-30% of TBSA

TEN covers > 30% of TBSA

CAUSES OF SJS & TEN



The most frequent cause of SJS/TEN is an immune reaction to a medication. In children, viruses can cause similar symptoms. Almost any drug can cause this reaction but the most common are:

- Antibiotics
- Anti-epilepsy or seizure drugs
- Allopurinol
- Non-steroidal anti-inflammatory drugs (NSAIDs), such as Ibuprofen



1-9 cases per million develop SJS & TEN



5-30% risk of death with SJS & TEN, respectively

SIGNS & SYMPTOMS

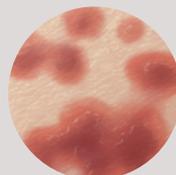
Symptoms can begin 1-3 weeks after a medication exposure. Initial signs may include a fever and flu-like symptoms followed by a red to purple rash, skin tenderness, skin blistering, and peeling.

Early Symptoms:

- Fever
- Fatigue
- Headache
- Joint pain
- Cough
- Burning red eyes

Late Symptoms:

- Blistering of the skin, mouth, eyes, & genitals
- Red rash on any part of the body
- Skin peeling
- Burning eyes



WHAT TO DO?

MANAGEMENT AND FOLLOW UP RECOMMENDATIONS



- Recognizing the early symptoms of SJS/TEN and seeking medical attention are the most important steps to minimize the disease.
- Stop the culprit drug **IMMEDIATELY**.



- Depending on the severity of your SJS/TEN, you may be hospitalized in an Intensive Care Unit or a specialized Burn Unit/Center.
- Treatment may include antibiotics to prevent infection, IV fluids to bring back fluids lost through the skin that has sloughed off, and medications for pain relief.



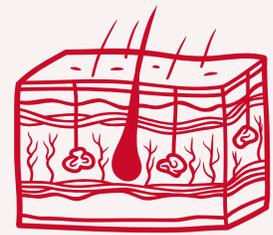
- After discharge, keep up with all appointment visits, including specialty visits.
- Try not to be afraid of all new medications. The immune response is highly specific only to the drug that caused your SJS/TEN. Talk to your doctor before starting a new medication, even over-the-counter, and be sure they know your history.



- Wear sunscreen (**SPF > 30**) reapplying every 2 hours, and protective eyewear as the skin and eyes are sensitive to sunlight and may take months to heal.
- Follow all instructions, including use of moisturizers, eye and mouth care.
- For more information, visit the specialty care and support group section.

SPECIALTY CARE

SKIN



► SJS/TEN & SKIN

Patients with SJS/TEN require immediate hospitalization as symptoms can be life-threatening. When possible, patients are treated in a burn center or intensive care (ICU) unit along with a team of specialists.

Possible complications of SJS/TEN include infection, difficulty breathing due to fluid in the lungs, and organ failure. In addition, patients may be unable to open their eyes, have a hard time eating, and suffer large amounts of fluid loss or dehydration because water escapes through the areas of lost skin. Areas of the skin may become scarred or discolored. Lifelong hair loss can happen along with loss or scarring of the fingernails or toenails. Due to the severe complications of SJS/TEN and healing time, hospitalization may last weeks to months.

► TREATMENT

Treatment of SJS/TEN will vary depending on how much of the body is affected and any complications that happen. Treatment may include:

- **Wound Care:** Hospital staff will help keep your skin clean and moist. Dead skin is sometimes removed, and bare patches are covered with a special dressing, topical antibiotics, and moisturizers to prevent infection and promote healing.
- **IV Fluids:** To prevent dehydration, multiple fluids are given into the vein.
- **Nutrition:** The skin needs protein to rebuild and heal. Wounds in the mouth or throat may bleed and cause difficulties with swallowing. A tube that goes into the stomach through the nose may be used to ensure a high calorie diet and recovery.
- **Airway Management:** A breathing tube may be required for patients with difficulty breathing along with giving oxygen.
- **Pain Management:** SJS/TEN can be very painful, and painkillers are given to help ease any pain when needed.

► FOLLOW-UP CARE

- Avoid sun exposure for several months and use sunblock anytime there is sun, the higher the number of SPF the better!
- Follow all instructions for changing your bandages at home, including putting on prescription ointments or creams. Keep wounds moist and covered.
- If your wound or bandage develops a bad smell or pus, seek immediate help as this may be a sign of infection.
- You may be instructed to use a medicated shampoo when showering. For areas of skin with hair, use baby shampoo.
- If you have mouth sores, a prescribed mouthwash and special toothbrush or sponge may help.
- Scarring such as on the face, may require special treatment once healed.
- A physical therapist can help you regain your strength to walk and get back to your normal activities.

SPECIALTY CARE

GYNECOLOGY



► SJS/TEN & FEMALE GENITAL ORGANS

SJS/TENS can affect the vulva and vagina. In the beginning, the vulva can appear red and form larger blisters while the outermost surface of the vagina can peel off. Resolution of these lesions may range from weeks to months if not treated.

Long term complications of SJS/TEN could lead to the labia sticking together and the vaginal entrance tightening. Scarring inside the vagina could lead to narrowing and rarely, closure of the vagina. Common secondary symptoms patients can develop due to long term complications include:

- Genital dryness
- Itching
- Burning or bleeding after intercourse
- Sexual dysfunction
- Painful sexual intercourse or even inability to have intercourse can occur
- Problems with urination

► TREATMENT

If female organs are present, a Gynecologist should be consulted soon after admission to the hospital for assessment of vulvovaginal involvement and counseling. During the hospital stay, treatment may include:

- Suppressing your menstrual cycles (we don't want you to have your period)
- Placing a vaginal dilator with steroid ointment or a barrier cream to prevent labial adhesions
- Placing a urinary catheter to prevent adhesions near the urethra
- Anti-virals to suppress genital herpes in affected patients
- Yeast treatment

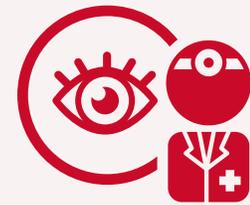
► FOLLOW-UP CARE

After discharge, it is important to follow up with a Gynecologist when the vulva or vagina is involved. Follow up recommendations may include:

- Continuation or start using a dilator with a steroid ointment such as clobetasol or betamethasone. You can also use a dilator with lidocaine jelly later on for pain control.
- If the vulva is not yet completely healed, consider application of a steroid or barrier cream around the labia minora and majora to prevent adhesions along with taking sitz baths.
- Until healing of the vulvovaginal area is complete, continue menstrual suppression.
- Your Gynecologist can do a vaginal examination for any vaginal narrowing or adenosis (patches inside the vagina that could rarely lead to cancerous changes).
- Referral for pelvic floor physical therapy may also be considered.

SPECIALTY CARE

OPHTHALMOLOGY



SJS/TEN & EYES

Eyes with ocular involvement from SJS/TEN, require acute and chronic care. Most SJS/TEN survivors may develop eye symptoms including vision changes. The common effects of SJS/TEN on the eye include:

- Corneal Scarring
- Dry Eyes
- Ingrowing Eyelashes
- Photosensitivity as well as other complications

The decrease in vision for some may get worse over time (months or years), while others can have severe visual problems right away. The more severe vision loss can be described as a “functional blindness” because most survivors may have some residual vision (about 1-15% of normal vision). However, the quality of vision is very distorted and can be affected by environmental conditions such as heat, bright lighting, and sunlight. The inflammatory process in the eyes can keep going long after patients recover from the initial acute phase of SJS/TEN.

TREATMENT

If there is eye involvement, determined by an eye exam by an Ophthalmologist, sometimes topical medication is used, and if the eye disease is severe, sometimes an Amniotic Membrane Grafting Surgery is needed.

WHAT IS AMNIOTIC MEMBRANE GRAFTING SURGERY?

- The procedure involves applying a human amniotic membrane made of a combination of tissue and cells as a biological dressing to help reduce the risk of vision loss, scarring of the eyes, and promote better wound healing of the eye.
- The procedure is performed by a cornea specialist and should be done within 7 days of the start of the skin rash. The membrane is less effective if performed later.

FOLLOW-UP CARE

It is especially important for every patient to schedule a follow up exam with an ophthalmologist (not optometrist).

- A cornea specialist will perform a comprehensive baseline eye exam to monitor for any changes of the eye disease over time.
- Management of eye inflammation may include medical or surgical treatment such as topical steroid or other anti-inflammation drops, antibiotics, serum blood tears or therapeutic contact lenses to help protect the eye and relieve any pain from dry eye syndrome.
- Surgical management might include eyelid repair which when pointed inward, can lead to the lashes rubbing on the surface of the eye. Adhesions between the eyelid and eyeball may also require surgical repair.



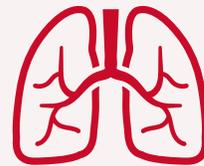
SPECIALTY CARE

PSYCHIATRY/PSYCHOLOGY

Coping with SJS/TEN can involve intense emotions for both the patient as well as family members. What you have gone through is traumatic and will take time to heal. Professionals may be helpful during the hospital admission and following discharge. A psychiatrist or psychologist can provide treatment for mental health concerns, such as post-traumatic stress disorder, related to your SJS diagnosis.

SPECIALTY CARE

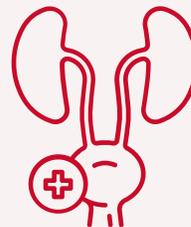
PULMONOLOGY



After SJS/TEN, more than half of patients have abnormal oxygen flow while breathing. However, severe long-term lung complications are rare. A pulmonologist will assess your breathing using a test that involves breathing through a mouthpiece. It is recommended that SJS patients follow up with a pulmonologist for 1 year after the hospital.

SPECIALTY CARE

UROLOGY



The penis and vagina can be affected by SJS/TEN and there can be pain with urination. Blood in the urine and difficulty emptying the bladder when urinating are also possible. While in the hospital, these issues may require a catheter to be placed in addition to evaluation by a urologist.

FAQs

01. WHAT ARE THE LONG-TERM COMPLICATIONS OF SJS/TEN?

- SJS and TEN can leave victims with reduced vision (mild to severe vision loss including blindness), damaged lungs or other internal organs, chronic skin pain, and scarring. Patients can suffer from anxiety, depression, or even post-traumatic stress disorder after this highly painful experience.
-

02. HOW LONG WILL IT TAKE FOR MY SKIN TO HEAL?

- The amount of time your skin will take to heal depends on the severity of the reaction. Recovery can take anywhere from weeks to months, sometimes years.
-

03. WILL I HAVE PERMANENT VISIBLE DAMAGE TO MY SKIN?

- Scarring happens if the reaction was deep enough, and follow-up care will help let you know what you expect. Not all patients will have long-term skin damage, but possible changes include darkening or lightening of the skin, scars, nail changes, hair changes, and eczema or dry skin. In some cases, scarring can be treated with laser therapy.
-

FAQs

04. IS SJS/TEN GENETIC?

- Currently, genetic tendency for developing SJS/TEN has been reported. However, this has only been seen with some specific ethnicities and associated with specific medications. Most prescribers are aware of these risk factors and should discuss genetic testing with patients at high risk if necessary. Otherwise in most cases, there is no definite genetic link, but SJS/TEN running in families has been reported.
-

05. WHAT TREATMENT OPTIONS ARE AVAILABLE?

- At this time, there is no definitive cure or treatment for SJS/TEN. The most important thing is to never take the offending drug which caused your disease again. Your disease may be treated with a combination of ointments that go on your skin or eyes, oral pills, IV medications, steroids, and antibiotic eye drops. For more severe disease amniotic membrane grafting may be done early on. It is important to keep the skin moist and continue that at home and be sure to follow up with multiple specialists following discharge.
-

FAQs

06. WHO CAN GET SJS/TEN?

- SJS/TEN can affect patients of all ages, races, ethnicities, and genders. More cases have been reported in women compared to men. Patients with a previous or family history of SJS/TEN, and immunodeficiency might be at an increased risk.
-

07. SHOULD I BE CONCERNED ABOUT THE COVID-19 VACCINE IF I'VE HAD SJS/TEN IN THE PAST OR MORE RECENTLY?

- It is understandable that survivors of SJS/TEN would be hesitant to take any medication and vaccines even outside the drug that was implicated in their reaction given the severity of the disease. However, those who have recently experienced SJS/TEN and survivors of SJS/TEN can be reassured that receiving the SARS-CoV-2 vaccine is a safer option than natural infection.
-

08. CAN I QUALIFY FOR DISABILITY AFTER SUFFERING THROUGH SJS/TEN?

- Many SJS/TEN patients are unable to work due to lingering side effects. The sooner you contact apply for disability, the better as approval can take months to be approved. You may also need to contact Social Services to receive Medicaid, or your employer for short or long-term disability benefits. Most hospitals have a patient advocate and social work on staff to help.



SUPPORT GROUPS

Be assured, you are not alone!

The **SJS Foundation** is a non-profit organization that provides support and resources to patients and families affected by SJS/TEN. More information can be found at <http://sjsupport.org/>

STEVENS-JOHNSON SYNDROME FOUNDATION

Website: www.sjsupport.org

Email: sjsupport@gmail.com

SJS KIDS SUPPORT

Website: sjskidssupport.webs.com

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