CONDITION

Ectodermosis erosiva pluriorificialis

A severe inflammatory reaction causing painful erosions around several body openings, often involving the mouth, eyes, and genitals.

WHAT IT IS
Severe inflammation affecting several mucosal body openings

BODY SYSTEM
Skin, eyes, mouth, genitals, and airways

MAIN SYMPTOMS
Painful mouth sores, red eyes, genital erosions, skin rash

WHAT CAUSES IT
Immune reaction, often to medicine or infection

HOW IT IS DIAGNOSED
Clinical examination, medicine history, sometimes skin biopsy

MAIN TREATMENT
Trigger removal, hospital supportive care, specialist treatment

CAN IT BE CURED?
Sometimes - most recover after the trigger resolves

IS IT AN EMERGENCY?
Sometimes - painful blistering or skin peeling needs urgent care

LONG-TERM OUTLOOK
Recovery common, but eye or genital scarring can persist

Overview

Ectodermosis erosiva pluriorificialis is an older term for a severe inflammatory reaction affecting the moist linings of several body openings. These mucous membranes include the mouth, eyes, nose, genitals, and anus.

The condition can cause painful blisters and raw, eroded areas. Some people also develop target-shaped spots, blisters, or peeling skin. Fever and flu-like symptoms may begin before the sores appear.

Doctors rarely use this term as a stand-alone diagnosis today. Similar presentations are usually classified as Stevens-Johnson syndrome, erythema multiforme major, or reactive infectious mucocutaneous eruption. The last describes marked inflammation of the mouth, eyes, or genitals after an infection, sometimes with little skin involvement.

Correct classification matters because triggers, treatment, and future precautions differ. A medicine-related reaction may require permanent avoidance of the suspected medicine, while an infection-related eruption may require testing and treatment for that infection.

New painful sores affecting several mucous membranes need prompt assessment, particularly with fever, eye pain, blistering, or peeling skin.

Symptoms

Symptoms may begin with fever, tiredness, headache, sore throat, cough, or aching. Painful mucosal inflammation can follow over several days.

Possible symptoms include:

Severe mouth pain may cause dehydration. Eye inflammation can worsen quickly and damage the eye surface if specialist care is delayed.

Some infection-related cases mainly affect mucous membranes, with few skin spots. They may initially resemble a routine mouth infection, conjunctivitis, or genital infection.

- Blisters, crusting, or raw areas on the lips and inside the mouth
- Severe mouth pain, drooling, or difficulty eating and drinking
- Red, gritty, painful, or light-sensitive eyes
- Swollen eyelids, eye discharge, or blurred vision
- Painful urination or sores around the genitals
- Anal pain or erosions
- A red, purple, blistered, or target-shaped rash
- Tender or peeling skin
- Cough, breathing discomfort, or airway inflammation

Causes and risk factors

Symptoms result from an immune reaction that damages cells in the skin and mucous membranes. The older term does not identify a single cause.

Medicines are important possible triggers when the illness falls within Stevens-Johnson syndrome or toxic epidermal necrolysis. The timing varies, so clinicians need a complete list of recent prescription medicines, non-prescription products, and supplements.

Infections can also trigger severe mucosal inflammation. Mycoplasma pneumoniae , which can cause respiratory infection, is a well-described trigger. Other bacterial and viral infections have also been associated with reactive infectious mucocutaneous eruption.

Cough or fever does not prove that infection caused the reaction because these symptoms can also accompany a medicine reaction. Infection test results must therefore be interpreted with the examination and medicine history.

Specific genetic variants can increase the risk of severe reactions to certain medicines, particularly in some ancestral groups. However, the condition does not usually follow a simple inherited pattern. Sometimes no trigger is found.

Diagnosis

There is no single test that confirms every case. Diagnosis depends on the appearance and distribution of sores, the amount of skin affected, recent illnesses, and medicine exposure.

Assessment may include:

A biopsy removes a small tissue sample for examination under a microscope. It can support a diagnosis within the Stevens-Johnson syndrome spectrum and help exclude autoimmune blistering disorders, infection, and other causes. Results must still be considered alongside the clinical findings.

Urgent specialist eye examination is important for eye redness, pain, discharge, blurred vision, or light sensitivity because early damage may not be visible without specialist equipment.

The diagnostic label may change as test results and the illness pattern become clearer.

- Examination of the skin, mouth, eyes, genitals, and other affected areas
- A timeline of recent medicines and infections
- Blood tests for inflammation, blood counts, kidney and liver function, and hydration
- Respiratory samples, swabs, or other infection tests
- A skin or mucosal biopsy

Treatment and management

Treatment involves removing or treating the suspected trigger, protecting damaged surfaces, controlling pain, and preventing complications. Significant cases are usually treated in hospital. Extensive skin loss may require burns-unit or intensive care.

If a medicine is suspected, clinicians normally stop it promptly and review related medicines. If painful mucosal blistering develops after starting a medicine, seek urgent medical advice before taking another dose. Do not restart a medicine blamed for a serious reaction unless an appropriate specialist clearly says it is safe.

Supportive care may include fluids, nutrition, pain relief, wound care, temperature control, and monitoring of salts and organ function. Mouth care can reduce discomfort and help with drinking. A feeding tube may be needed temporarily.

Eye treatment should begin early and may include frequent lubrication, prescribed eye medicines, removal of inflammatory material, or procedures to prevent the eyelids from sticking to the eye surface.

Genital care aims to control pain and prevent nearby surfaces from joining through scar tissue. It may involve gentle local care, prescribed anti-inflammatory treatment, and specialist follow-up.

Clinicians may treat an identified infection. Antibiotics like amoxicillin or doxycycline do not directly reverse the immune injury and are not routinely prescribed simply because skin or mucosal surfaces are open.

Corticosteroids like prednisone or hydrocortisone , ciclosporin, intravenous immunoglobulin, or other immune-modifying treatments may be considered in selected severe cases. Their use depends on the diagnosis, illness stage, and specialist team.

Complications

Early concerns include dehydration, poor nutrition, infection of damaged tissue, severe pain, and disruption of fluid balance or temperature control.

Eye inflammation can cause chronic dryness, light sensitivity, scarring, inward-turning eyelashes, or reduced vision. Complications may appear during recovery as well as during the initial illness.

Genital or urinary scarring can cause pain, narrowing, altered urine flow, or difficulty with sexual activity. Airway involvement may lead to ongoing cough or breathing problems and, rarely, deeper lung scarring.

Accidental re-exposure to a causative medicine can trigger another serious reaction. The suspected medicine and potentially related medicines should be documented and reviewed by a specialist.

Pain, altered appearance, prolonged symptoms, and fear of recurrence may also contribute to

anxiety or low mood.

Prevention

Not every case can be prevented, and a first medicine reaction cannot always be predicted.

After a suspected medicine-related episode, prevention focuses on avoiding unsafe re-exposure. Keep the medicine name in medical and pharmacy records. A medicine-allergy card or medical alert item may be useful after a confirmed severe reaction.

Do not assume every medicine taken near the illness is unsafe. Specialist review can identify the most likely trigger and any related medicines that should also be avoided.

Genetic testing can estimate the risk of certain medicine reactions in specific populations. It is not a general test for this condition and cannot rule out future reactions.

Living with it and what to expect

Active inflammation often settles after the trigger is removed or the associated infection passes. Healing may take several weeks, and energy, appetite, and comfort can recover more slowly than the skin.

Mouth and skin surfaces may heal without lasting damage, but the eyes, genitals, and airways are more vulnerable to scarring. Follow-up should reflect which areas were affected, even after the rash disappears.

Eye dryness, irritation, or light sensitivity may need treatment for months or longer. Regular eye review is important after significant involvement, even if vision initially seems normal.

Discuss genital discomfort, altered urine flow, pain during sexual activity, cough, or breathing difficulty during follow-up. These problems may be treatable.

Many people have only one episode. Recurrence is a particular concern after re-exposure to a causative medicine, although infection-triggered eruptions can occasionally recur with later infections.

When to get medical help

Arrange a routine appointment if affected areas remain dry, painful, or sensitive after the main illness settles. Ongoing eye irritation, urinary discomfort, genital pain, breathing symptoms, or emotional distress also need follow-up.

Seek urgent same-day assessment for:

Call emergency services or go to an emergency department for skin peeling, breathing difficulty, faintness, confusion, severe weakness, or inability to swallow fluids. These can indicate extensive disease or a serious complication.

- New painful blisters or raw areas affecting the mouth, eyes, or genitals
- A spreading rash after starting a medicine
- Eye pain, light sensitivity, discharge, or blurred vision
- Mouth pain that makes drinking difficult
- Fever with tender or blistering skin
- Reduced urination or other signs of dehydration

Common questions

Is ectodermosis erosiva pluriorificialis the same as Stevens-Johnson syndrome?

The name has historically described a pattern now often classified as Stevens-Johnson syndrome. Similar inflammation after infection may instead be called reactive infectious mucocutaneous eruption. Clinicians use the trigger, skin findings, biopsy, and course to determine the most accurate diagnosis.

Is it contagious?

The inflammatory reaction is not contagious. An infection that triggered it may spread, depending on the organism, but another person who catches that infection would not necessarily develop the same reaction.

Can it happen without a skin rash?

Yes. Infection-related cases can cause severe mouth, eye, and genital inflammation with few or no widespread skin lesions. A limited rash does not make painful eye or mucosal symptoms less urgent.

How long does recovery take?

Active sores may improve over several weeks, although fatigue and sensitivity can last longer. Eye, genital, or airway complications may require follow-up for months or years.

Will it come back?

Many people have one episode. Recurrence is possible after re-exposure to a causative medicine or, less commonly, after another infection. Clear documentation of suspected

medicine triggers reduces avoidable risk.

Can it cause permanent eye problems?

Yes. Scarring may cause dry eyes, discomfort, light sensitivity, or reduced vision. Prompt eye assessment during the acute illness and continued follow-up help identify complications requiring treatment.

Sources

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