CONDITION

Zambusch's disease

Zambusch's disease is a severe form of pustular psoriasis that causes widespread pus-filled spots and inflamed skin.

WHAT IT IS
Severe, widespread form of pustular psoriasis

BODY SYSTEM
Skin and immune system

MAIN SYMPTOMS
Sterile pustules, red painful skin, fever, fatigue

WHAT CAUSES IT
Immune dysregulation, sometimes involving inherited gene changes

HOW IT IS DIAGNOSED
Skin examination, blood tests, sometimes skin biopsy

MAIN TREATMENT
Urgent specialist care and systemic anti-inflammatory medicines

CAN IT BE CURED?
No - treatment controls but cannot cure

IS IT AN EMERGENCY?
Sometimes - widespread painful rash with fever

LONG-TERM OUTLOOK
Flares may recur; treatment often clears the skin

Overview

Zambusch's disease is a name sometimes used for generalized pustular psoriasis , or GPP. The established medical term is von Zumbusch generalized pustular psoriasis . It is a rare and potentially serious inflammatory skin condition.

GPP causes large areas of red, tender skin covered with small pus-filled spots called pustules. The pus is usually sterile , meaning it is caused by inflammation rather than a bacterial infection. The condition is not contagious.

Unlike the more common plaque form of psoriasis, GPP can affect the whole body and make you feel very unwell. A flare may disturb temperature control, fluid balance and the body's normal chemical balance. Some people need treatment in hospital.

GPP can occur in someone who has plaque psoriasis, but it can also develop without a previous psoriasis diagnosis. It is different from localized pustular conditions that mainly affect the palms, soles, fingers or toes.

Symptoms

A flare often develops quickly, sometimes over hours or days. The skin may first become hot, red and painful. Numerous small pustules then appear and may join into larger areas.

The pustules can dry, peel and be replaced by new ones in waves. Large areas of skin may become sore or itchy.

Symptoms can include:

Redness may be less obvious on brown or black skin. The affected skin may look purple, grey or darker than the surrounding area.

Not every pustular rash is GPP. Infections, medicine reactions and several other skin conditions can look similar. A rapidly spreading pustular rash needs prompt medical assessment rather than self-diagnosis.

- Widespread red or darkened, inflamed skin
- Many small, pus-filled spots
- Skin pain, tenderness, itching or burning
- Fever, chills, tiredness or muscle weakness
- Dry, peeling or crusted skin
- Reduced appetite, nausea or joint pain

Causes and risk factors

GPP results from abnormal immune activity in the skin. Immune signals that normally coordinate inflammation become overactive. One important pathway involves a protein called interleukin-36 , or IL-36.

Some people have a change in a gene involved in skin inflammation, such as `IL36RN`. Other genes may also contribute. However, many people with GPP do not have an identified gene change, and having a gene change does not always predict how the condition will behave.

Possible flare triggers include:

Sometimes no trigger can be found.

Having plaque psoriasis or a previous GPP flare increases the chance of another episode. The condition can also begin in someone without either history.

GPP is not caused by poor hygiene, and the sterile pustules cannot spread the condition to another person. An infection can sometimes trigger a flare or occur alongside it, so testing may still be needed.

- Suddenly stopping a systemic corticosteroid, such as steroid tablets
- Certain medicines or a reaction to a medicine
- An infection
- Pregnancy or the period shortly after birth
- Severe physical stress, such as surgery or major illness
- Stopping or changing psoriasis treatment

Diagnosis

There is no single test that confirms every case. Diagnosis is based on the appearance and distribution of the rash, how quickly it developed, other symptoms and your medical history.

A clinician may ask about psoriasis, recent infections, pregnancy and all medicines you have recently started, stopped or changed. This includes prescription medicines, over-the-counter products and supplements.

Tests may include:

Important alternative diagnoses include a severe medicine reaction called acute generalized exanthematous pustulosis , bacterial or fungal infection, and other forms of pustular psoriasis. These conditions may require different treatment.

- Blood tests for inflammation, infection, blood-cell counts and organ function
- A sample from a pustule to check for bacteria or other infection
- A skin biopsy, in which a small skin sample is examined under a microscope
- Genetic testing in selected cases, especially with early or repeated episodes

Treatment and management

A widespread flare generally needs urgent assessment by a dermatologist. Hospital care may be needed if you have fever, extensive skin involvement, dehydration, abnormal blood results

or signs that other organs are affected.

Supportive treatment may include fluids, temperature control, gentle skin care and monitoring of the heart, kidneys, liver and blood chemistry. Any infection found alongside the flare is treated separately.

Medicines that act throughout the body are called systemic treatments . Options for GPP include medicines that block specific immune signals, particularly the IL-36 pathway. Spesolimab is one such medicine used for GPP flares in adults.

Other systemic treatments may include:

The choice depends on the severity of the flare, other health conditions, pregnancy, previous treatment and how quickly treatment needs to work. Some medicines require blood tests or other monitoring.

Moisturizers, wet dressings and topical medicines applied to the skin may improve comfort, but they are usually not enough on their own for a severe generalized flare.

Do not suddenly stop prescribed corticosteroids or other psoriasis medicines without medical guidance. Abrupt withdrawal can trigger or worsen GPP in some people. If a medicine may have caused the flare, a clinician can decide how it should be changed safely.

- Biologic medicines that target parts of the immune system
- Retinoids, which are related to vitamin A and regulate skin-cell growth
- Ciclosporin, which suppresses immune activity
- Methotrexate, which reduces inflammation and immune activity

Complications

Inflamed, damaged skin can lose fluid and heat and may not protect the body as effectively as usual. Possible complications include dehydration, abnormal salt levels in the blood and difficulty maintaining a stable body temperature.

Severe inflammation may place stress on the heart, lungs, kidneys or liver. A secondary skin or bloodstream infection can also occur, even though the original pustules are usually sterile.

These complications are not inevitable. Prompt treatment and monitoring reduce risk. After the active flare settles, peeling skin, temporary color changes and tiredness may continue for a while.

Prevention

GPP cannot always be prevented. Inherited susceptibility cannot be changed, and some flares have no identifiable trigger.

If you have had GPP, an ongoing treatment plan may reduce the chance or severity of future flares. Take long-term medicines as agreed and discuss side effects before stopping them.

Keep an up-to-date list of medicines and tell healthcare professionals about your history before starting or stopping treatment. In particular, do not abruptly stop systemic corticosteroids unless you have been given a safe plan.

Prompt care for infections may also be helpful because infections can trigger flares. If pregnancy is possible or planned, discuss psoriasis medicines in advance because some are unsafe during pregnancy and treatment may need to change.

Living with it and what to expect

GPP often follows a relapsing course. This means a flare may clear and then return months or years later. Some people have only occasional episodes, while others have more frequent flares or continuing inflammation between major episodes.

Skin clearance is possible with treatment, but it does not mean the underlying tendency has been cured. Future flares may not look or feel exactly like the first one.

Regular follow-up usually focuses on controlling inflammation, watching for treatment side effects and planning what to do if symptoms return. Blood tests or other monitoring may be needed with some systemic medicines.

GPP can affect sleep, work, movement and emotional well-being, especially when skin is painful or unpredictable. Practical support, clear flare instructions and access to specialist dermatology care can make the condition easier to manage.

The condition is not contagious. You can work, attend school, exercise and have close contact with other people when you feel able. Recovery from a severe flare may take time, so activity may need to increase gradually.

When to get medical help

Seek urgent same-day medical care if you develop a new or rapidly spreading pustular rash, particularly if you have had GPP before. You should also seek urgent advice if symptoms return after a medicine has been started, stopped or changed.

Go to an emergency department or call emergency services if a widespread rash is accompanied by:

A widespread pustular rash during pregnancy also needs urgent assessment.

Routine follow-up is appropriate when your condition is stable but flares are becoming more frequent, treatment is not controlling symptoms, or side effects are affecting you. New joint pain, persistent tiredness or changes in your nails or usual plaque psoriasis should also be discussed at follow-up.

- Fever, shaking chills or severe weakness
- Trouble breathing, chest pain or a very fast heartbeat
- Confusion, fainting or difficulty staying awake
- Very little urine, severe dizziness or inability to drink
- Rapidly worsening pain, swelling or skin peeling

Common questions

Is Zambusch's disease the same as psoriasis?

It is a severe form of psoriasis called generalized pustular psoriasis. It differs from common plaque psoriasis because it causes widespread pustules and can affect the whole body. Some people have both forms.

Is it contagious?

No. GPP is caused by inflammation, not by catching an infection from another person. The pus in the pustules is usually sterile. An infection may sometimes trigger a flare or develop in damaged skin, but the psoriasis itself cannot spread between people.

Can generalized pustular psoriasis be fatal?

Severe untreated flares can cause serious complications, but this is not the outcome for most people receiving prompt care. Fever, extensive painful skin changes or signs of dehydration require urgent assessment.

Will the pustules leave scars?

The pustules do not usually cause permanent scars. Temporary peeling or lighter or darker skin color may remain after a flare. Scratching, deep skin damage or a secondary infection can increase the chance of lasting marks.

Why did I have a flare when nothing changed?

Not every trigger can be identified. Immune activity may increase without an obvious infection,

medicine change or stressful event. This does not mean you caused the flare or failed to prevent it.

Can it be cured permanently?

There is currently no permanent cure. Treatment can clear a flare, control ongoing inflammation and reduce future episodes. The result and length of remission are different for each person.

Sources

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