CONDITION

Waterhouse-friderichsen syndrome

Waterhouse-Friderichsen syndrome is sudden adrenal gland failure caused by bleeding during an overwhelming infection.

WHAT IT IS
Acute adrenal failure with severe bloodstream infection

BODY SYSTEM
Adrenal glands, blood, heart and circulation

MAIN SYMPTOMS
Fever, purple rash, confusion, very low blood pressure

WHAT CAUSES IT
Infection causing bleeding into both adrenal glands

HOW IT IS DIAGNOSED
Blood cultures or PCR, hormone tests, adrenal imaging

MAIN TREATMENT
Emergency antibiotics, steroids, fluids, and intensive care

CAN IT BE CURED?
Sometimes - immediate treatment can reverse the crisis

IS IT AN EMERGENCY?
Yes - suspected cases need immediate hospital treatment

LONG-TERM OUTLOOK
Survivors may recover or have lasting adrenal damage

Overview

Waterhouse-Friderichsen syndrome is a rare, life-threatening condition in which severe infection leads to bleeding in the adrenal glands. These small glands sit above the kidneys and make hormones that help control blood pressure, salt balance, blood sugar, and the body's response to stress.

The condition is classically linked to meningococcal disease, an infection caused by Neisseria meningitidis . Other serious bacterial infections can also cause it.

The infection may trigger widespread inflammation, damage to blood vessels, and abnormal clotting. This can reduce blood flow to major organs and cause bleeding into both adrenal glands. The glands may then stop producing enough cortisol, an essential stress hormone. This sudden hormone shortage is called acute adrenal insufficiency or an adrenal crisis.

Waterhouse-Friderichsen syndrome can progress over hours. It causes shock, meaning that blood pressure and circulation become too poor to supply the body properly. Treatment must begin as soon as the condition is suspected. Doctors do not wait for every test result before giving antibiotics and other emergency care.

Symptoms

Symptoms often begin like a serious infection and then worsen quickly. Not everyone has every symptom, and a rash may be absent early on.

Possible symptoms include:

The rash associated with meningococcal infection results from bleeding under the skin. It may begin as small red or purple spots and develop into larger bruise-like areas. On darker skin, it may be easier to see inside the eyelids or mouth, or on the palms and soles.

A rash that does not fade under pressure is an emergency sign, but its absence does not rule out the condition. Do not wait for a rash to appear if someone with a fever is becoming confused, difficult to wake, breathless, or very unwell.

Babies may show less specific signs, such as poor feeding, irritability, a weak or unusual cry, a bulging soft spot on the head, or being unusually floppy or hard to wake.

- Sudden fever, chills, severe weakness, or muscle pain
- Severe headache, vomiting, neck stiffness, or sensitivity to light
- Confusion, unusual sleepiness, agitation, or reduced responsiveness
- Fast breathing, rapid heartbeat, cold hands and feet, or very low blood pressure
- Red, purple, or bruise-like spots that do not fade when pressed
- Abdominal, side, back, or leg pain

Causes and risk factors

Waterhouse-Friderichsen syndrome usually develops during an overwhelming bloodstream infection called sepsis. Meningococcal infection is the best-known cause. Severe infections caused by pneumococcal, streptococcal, or staphylococcal bacteria have also been reported.

During severe sepsis, the immune response can injure blood vessels and disrupt normal clotting. Numerous tiny clots may form while the body's supply of platelets and clotting proteins is used up. Bleeding can then occur elsewhere, including inside the adrenal glands.

This process is called disseminated intravascular coagulation, or DIC.

The condition can affect a person who was previously healthy. Factors that may raise the risk of severe meningococcal or pneumococcal infection include:

These factors do not mean that Waterhouse-Friderichsen syndrome will occur. Many people who develop meningococcal disease do not have a known risk factor.

Bleeding into the adrenal glands can also occur for reasons unrelated to infection, such as major trauma or anticoagulant treatment. This may cause adrenal failure, but doctors may not describe every case of adrenal bleeding as Waterhouse-Friderichsen syndrome.

- A missing or poorly functioning spleen
- Certain immune system disorders, including complement deficiencies
- Medicines that block part of the complement immune system
- Some forms of immune suppression
- Close exposure to someone with meningococcal disease

Diagnosis

This is primarily an emergency clinical diagnosis. Doctors consider it when a person has signs of severe infection, shock, abnormal bleeding, and possible adrenal failure. Treatment starts while tests are being arranged.

Blood tests may assess:

Hormone samples may be collected before steroid treatment if this can be done without delaying care. A single cortisol result can be difficult to interpret during critical illness, so it is considered alongside the person's condition and other findings.

A CT scan may show enlarged adrenal glands or bleeding into them. However, moving an unstable person for imaging may not be safe, and imaging must not delay emergency treatment. In some cases, adrenal bleeding is confirmed only after the immediate crisis has passed.

Tests may also include a lumbar puncture to examine the fluid around the brain and spinal cord if meningitis is suspected. This is performed only when the clinical situation makes it safe.

- Blood cultures or molecular PCR testing for the responsible infection
- Blood counts, including platelets
- Clotting function

- Kidney and liver function
- Blood sugar, sodium, and potassium levels
- Lactate, which may rise when tissues receive too little blood flow
- Cortisol and ACTH, hormones involved in adrenal function

Treatment and management

Waterhouse-Friderichsen syndrome requires immediate treatment in hospital, usually in an intensive care unit. Care focuses on controlling the infection, supporting circulation, correcting adrenal hormone deficiency, and treating abnormal clotting or organ failure.

Treatment may include:

Antibiotics like amoxicillin or doxycycline are usually started before the specific bacterium is known. The treatment can later be adjusted using blood culture or PCR results. The exact medicines and length of treatment are based on the infection and the person's response.

If meningococcal disease is suspected, hospital staff use infection-control precautions. Public health teams may arrange preventive antibiotics for close contacts. These are meant to stop meningococcal disease in people with significant exposure, not to treat the person who is already ill.

After the emergency, doctors reassess adrenal function. If the glands recover, steroid like prednisone or hydrocortisone replacement may eventually be reduced under specialist supervision. If adrenal damage is permanent, long-term cortisol replacement may be needed. Some people also need replacement of aldosterone, a hormone that helps control salt, fluid, and blood pressure.

- Intravenous antibiotics that cover likely bacterial causes
- Intravenous hydrocortisone to replace cortisol
- Fluids given through a vein
- Medicines called vasopressors to raise dangerously low blood pressure
- Oxygen or mechanical breathing support
- Blood products when serious bleeding or clotting problems require them
- Support for affected kidneys, heart, lungs, or other organs

Complications

The combination of severe infection, shock, abnormal clotting, and adrenal failure can affect many parts of the body.

Possible complications include:

These outcomes are not inevitable. The outlook is strongly influenced by how quickly the illness is recognised and treated, how severe the shock becomes, and which organs are affected.

- Kidney, liver, heart, or lung failure
- Brain injury from infection or reduced blood flow
- Bleeding or tissue damage caused by DIC
- Loss of fingers, toes, or parts of a limb after severe circulation damage
- Hearing loss or neurological problems after meningitis
- Permanent adrenal insufficiency
- Death despite intensive treatment

Prevention

Waterhouse-Friderichsen syndrome cannot always be prevented. It develops unexpectedly in some people, and vaccines do not protect against every bacterium or every strain that can cause severe infection.

Vaccination can reduce the risk of several infections associated with the syndrome. Meningococcal and pneumococcal vaccines are offered according to age, medical risk, local immunisation schedules, travel, or outbreak guidance.

People without a functioning spleen and those with certain immune conditions may need additional vaccines and an infection plan. Their usual medical team can explain which measures apply to them.

Close contacts of someone with meningococcal disease may be offered preventive antibiotics. In some situations, vaccination is also recommended. Public health teams identify who counts as a close contact; casual contact usually does not require treatment.

Prompt assessment of possible meningococcal disease or sepsis may reduce the chance of severe complications. A person should not wait for a purple rash if other emergency symptoms are present.

Living with it and what to expect

Waterhouse-Friderichsen syndrome is an acute crisis rather than a condition that usually develops slowly over years. The first hours and days focus on survival, controlling infection, and supporting affected organs.

Recovery after intensive care can take weeks or months. Tiredness, weakness, poor concentration, sleep problems, anxiety, and reduced stamina may continue after discharge. Rehabilitation may be needed after prolonged bed rest, neurological injury, tissue damage, or limb surgery.

Some survivors regain normal adrenal function. Others are left with permanent adrenal insufficiency and need daily hormone replacement. Anyone taking long-term steroid replacement needs clear instructions about increasing treatment during illness or injury, carrying emergency identification, and using emergency steroid medicine when prescribed.

Follow-up may involve endocrinology, infectious disease, rehabilitation, hearing assessment, mental health support, or other services. The exact follow-up reflects the organs affected during the illness.

Routine follow-up is important for reviewing adrenal recovery, medication, rehabilitation, and emotional health. New fever, vomiting, severe weakness, fainting, confusion, or very low blood pressure is not a routine follow-up issue in someone with adrenal insufficiency. These symptoms may indicate another adrenal crisis and need urgent assessment.

When to get medical help

Waterhouse-Friderichsen syndrome is always a medical emergency.

Call emergency services now if someone has a fever or suspected infection together with any of the following:

Do not wait for all symptoms to appear. Do not delay while repeatedly checking a rash or trying to obtain a routine appointment.

Someone with known adrenal insufficiency should follow their emergency steroid plan during significant illness. Persistent vomiting, collapse, confusion, or inability to keep replacement medicine down needs emergency help.

Routine appointments are appropriate after recovery for monitoring adrenal function, reviewing medicines, updating vaccinations, and addressing lasting physical or emotional effects.

- A red, purple, or bruise-like rash that does not fade under pressure
- Confusion, extreme sleepiness, collapse, or difficulty waking
- Trouble breathing, very fast breathing, or blue, grey, pale, or blotchy skin
- Cold hands and feet with severe weakness or worsening illness
- A seizure
- Signs of shock, such as fainting or very low blood pressure

Common questions

Is Waterhouse-Friderichsen syndrome the same as meningitis?

No. Meningitis is inflammation of the lining around the brain and spinal cord. Waterhouse-Friderichsen syndrome involves severe infection, shock, and bleeding into the adrenal glands. Meningococcal bacteria can cause either condition, and they may occur together.

Can you have it without a purple rash?

Yes. The rash may be absent, subtle, or appear late. A rapidly worsening fever with confusion, breathing difficulty, extreme sleepiness, or signs of shock needs emergency assessment even when there is no rash.

Can the adrenal glands recover?

Sometimes. Recovery depends on the extent of bleeding and tissue damage. Doctors usually reassess adrenal hormone production after the acute illness. Some survivors need lifelong hormone replacement, while others may eventually stop it under specialist supervision.

Is Waterhouse-Friderichsen syndrome contagious?

The syndrome itself is not contagious. However, an infection that caused it may spread between people. Meningococcal bacteria usually require close or prolonged contact with respiratory or throat secretions. Public health teams arrange preventive treatment when exposure is significant.

How quickly does the condition develop?

It can develop over several hours during a severe infection. Early symptoms may resemble flu or another common illness, but the person often becomes much more unwell quickly. Rapid deterioration is one reason emergency assessment is essential.

Can vaccination prevent it?

Vaccination can lower the risk of meningococcal and pneumococcal infections that may lead to the syndrome. It cannot prevent every case because several organisms can be responsible, and available vaccines do not cover every strain.

Sources

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