

WATERHOUSE – FRIDERICHSEN SYNDROME

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ABSTRACT

There is an increasing number of cases of the waterhouse – Friderichsen syndrome in the literature in recent years, probably due to its more accurate recognition and to the increased incidence of meningococcal infections during the past two years. The condition is believed to be the result of fulminating meningococemia and shock, and the pathologic findings can be explained on the toxic results of the organisms. Present usage of the term, waterhouse – Friderichsen syndrome, should be confined to those cases exhibiting the fairly typical clinical course as described, and undoubtedly to those patients whose symptomology was confirmed by autopsy. Care should be taken in the early recognition and differentiation of waterhouse – Friderichsen syndrome from acute meningococemia. The present outline of treatment centers on control of the bacteremia and measures aimed to combat shock, with secondary symptomatic and supportive treatment.

KEYWORDS: Adult, meningococcal meningitis, Waterhouse-Friderichsen syndrome.**DEFINITION**

Waterhouse–Friderichsen syndrome (WFS) is defined as adrenal gland failure due to bleeding into the adrenal glands, commonly caused by severe bacterial infection. Typically, it is caused by *Neisseria meningitidis*. The bacterial infection leads to massive bleeding into one or (usually) both adrenal glands.

**Causes**

WFS is caused by severe infection with meningococcus bacteria or other severe infection from bacteria, such as:

- Group B streptococcus
- *Pseudomonas aeruginosa*
- *Streptococcus pneumoniae*
- *Staphylococcus aureus*

Symptoms

Symptoms and signs usually come on very suddenly. They are due to the bacteria growing (multiplying) inside the body. Symptoms include:

- Fever and chills
- Joint and muscle pain
- Headache
- Vomiting

Infection with bacteria causes bleeding throughout the body, which causes:

- Body wide rash
- Disseminated intravascular coagulation in which small blood clots cut off blood supply to the organs
- Septic shock

Bleeding into the adrenal glands causes adrenal crisis, in which not enough adrenal hormones are produced. This leads to symptoms such as:

- Dizziness, weakness
- Very low blood pressure
- Very fast heart rate
- Confusion or coma

Exams and Tests

The health care provider will perform a physical examination and ask about the person's symptoms.

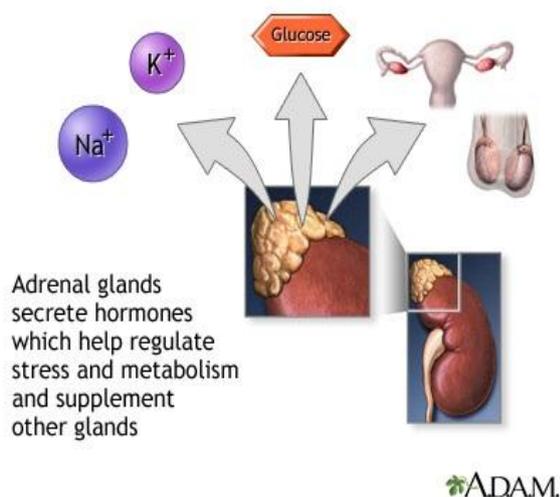
Blood tests will be done to help confirm if the infection is caused by bacteria. Tests may include:

- Blood culture
- Complete blood count with differential
- Lumbar puncture to get a sample of spinal fluid for culture
- Skin biopsy and Gram stain
- Urine analysis

Tests that may be ordered to help diagnose acute adrenal crisis include:

- ACTH (cosyntropin) stimulation test
- Cortisol blood test
- Blood sugar
- Potassium blood test
- Sodium blood test
- Blood pH test

Treatment



- The treatment is as that for meningococcal infection, fulminant meningococemia is a medical emergency and needs to be treated with adequate antibiotics as fast as possible.
- Ceftriaxone is an antibiotics commonly employed today. Ceftriaxone is a third – generation cephalosporin antibiotic. Like other third- generation cephalosporins, it has broad spectrum activity against gram –positive and gram negative bacteria.in most cases, it is considered to be equivalent to cefotaxime in terms of safety and efficacy.
- Benzyl penicillin was once the drug of choice with chloramphenicol as a good alternative in allergic patients
- Addition of adrenal support with hydrocortisone, given intravenously in a dose of 200mg per square meter body surface per four hours.
- Hydrocortisone can sometimes reversal the hypo adrenal shock.
- Hypervolemia is treated with colloids, dopamine and coagulation factors
- Sometimes plastic surgery and grafting is needed to deal with tissue necrosis.

Complication

1. Shock, extensive haemorrhage within the skin and fall into coma.
2. Death usually after a few hours, adrenal insufficiency being the immediate cause.
3. Patients who recover may suffer from extensive sloughing of the skin and loss of digits due to gangrene.
4. Meningitis generally does not occur.

Prevention

ROUTINE vaccination against meningococcus is recommended by the centres for disease control for:

1. All 11 – 18yrs olds
2. People who have poor splenic function (splenic removed or who have sickle – cell disease which damages the spleen.
3. Who have certain immune disorders, such as complement deficiency.

CONCLUSION

Waterhouse-Friderichsen syndrome, presenting with purpura fulminans, resulting from an invasive pneumococcal infection. In the emergency department, it is important to be aware of adrenal insufficiency as well as impaired splenic function whenever patients with sepsis are admitted.

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