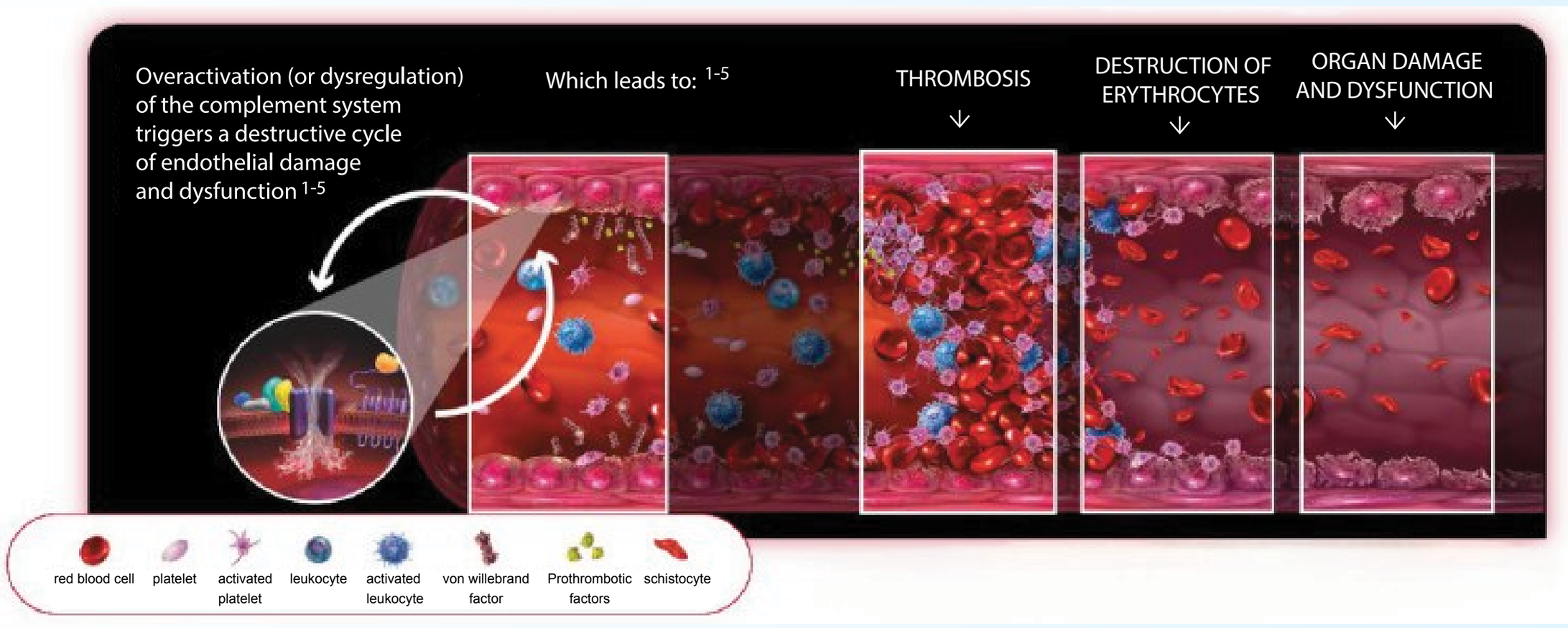
Atypical Hemolytic Uremic Syndrome (aHUS)

What is aHUS ?

Atypical hemolytic uremic syndrome (aHUS) is a rare, life-threatening disease affecting both adults and children. It is considered a form of thrombotic microangiopathy (TMA) which is characterised by destruction of red blood cells (haemolysis) resulting in anaemia, formation of blood clots (thromboses), inflammation and damage to blood vessel walls.
This leads to organ damage and reduced organ function that can be potentially life-threatening.

aHUS primarily affects the kidneys although other organs, such as the brain, lungs and gastrointestinal tract, can also be involved.



Prevalence

aHUS is a rare condition, with an estimated prevalence of 2 to 9 per million people worldwide. In Saudi Arabia, the exact prevalence is unknown due to the rarity of the disease and the difficulty in diagnosis.

Symptoms

aHUS has a wide range of symptoms depending on which organs are affected. Kidney failure is one of the most common symptoms.

Some of the symptoms of aHUS are:

General:

-Fatigue or tiredness.
-Pale skin.
-Sleepiness or drowsiness.

Digestive system:

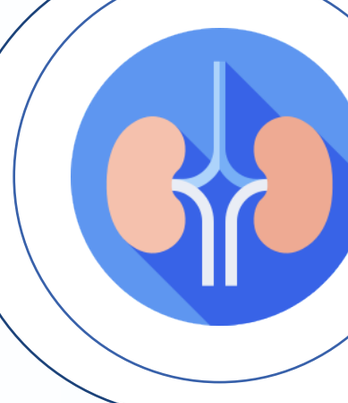
-Diarrhoea.
-Vomiting.
-Abdominal pain.

Brain:

-Headache.
-Confusion.
-Seizures.
-Stroke.

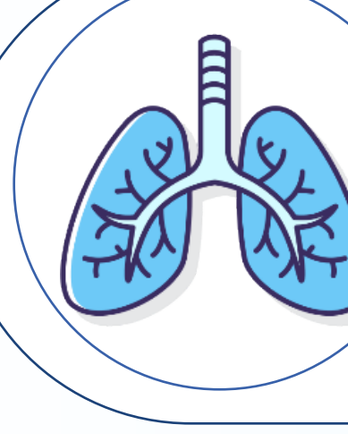
Heart:

-High blood pressure (hypertension).
-Fast heart rate (tachycardia).
-Heart attack.



Kidneys:

-Blood in the urine (haematuria).
-Low urine production (oliguria).
-Reduced kidney function.
-Kidney failure.



Lungs:

-Fluid build-up (pulmonary oedema).
-Bleeding (pulmonary haemorrhage).
-Shortness of breath (dyspnoea).

Causes and Risk Factors

aHUS is caused by a defect in a specific part of the body's immune system called the complement system, which results in its activity becoming uncontrolled causing blood platelets to become overactive.
The platelets stick together in small blood vessels and cause the formation of blood clots (thromboses) and inflammation in the blood vessel walls.

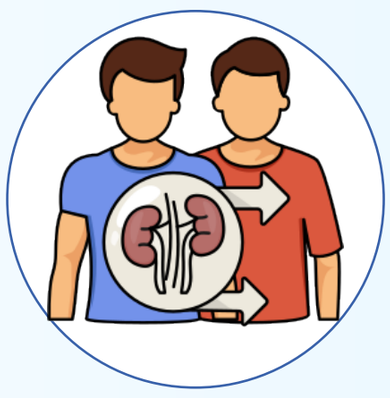
1 Genetics

In approximately half of people with aHUS, the defect in the complement system has a genetic cause.

2 Trigger event

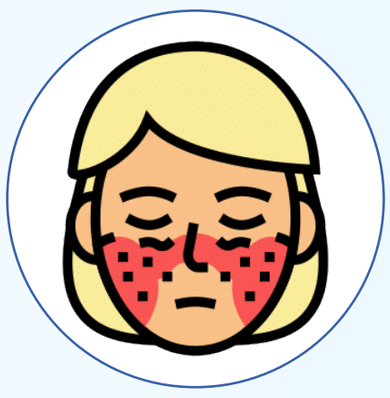
A triggering event or other clinical condition might be associated with aHUS. The trigger event can be:

Infection



Kidney transplant

Pregnancy



Autoimmune disease such as systemic lupus erythematosus

Certain Medications

Malignancy

Diagnosis

aHUS can be difficult to diagnose because it is a rare disease and has a wide range of symptoms that are similar to those of other diseases.

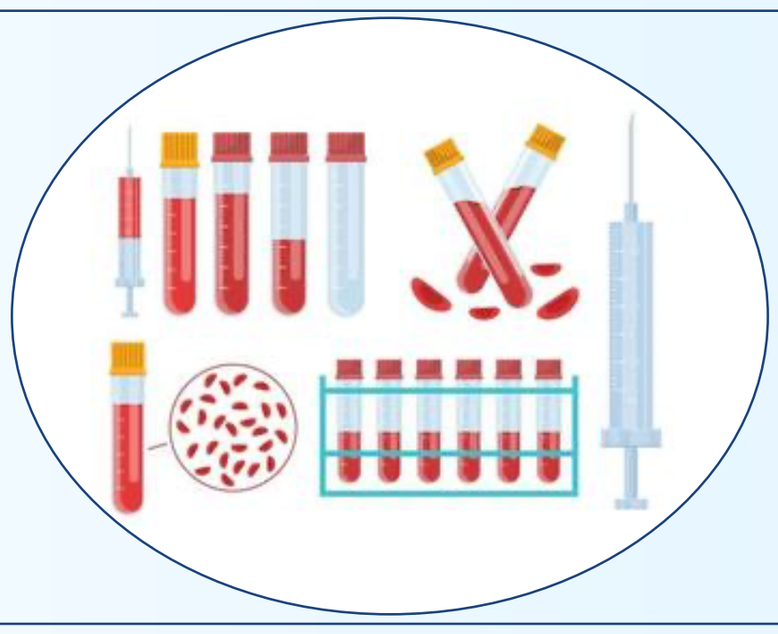
Impaired kidney function can be one of the early signs that help diagnose aHUS. If left unrecognised or inappropriately treated, aHUS has a high degree of morbidity and mortality.

Approximately 1 in 2 patients will require dialysis, suffer permanent kidney damage, or die within 1 year of first occurrence despite plasma therapy.

Some diagnostic tests that maybe performed are:

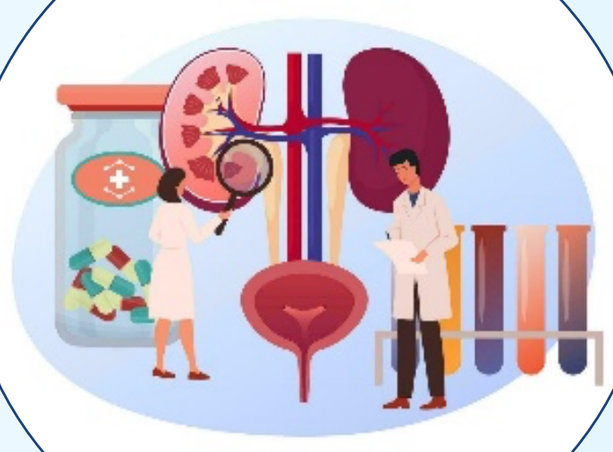
Blood counts

Blood tests



Kidney function

- creatinine and the estimated glomerular filtration rate (eGFR) which indicate how well the kidneys are working.



Genetic tests

to look for gene mutations

Exclusion tests

of (TTP and STEC-HUS).

Treatment

aHUS is a serious condition that requires immediate medical attention. While there is no cure, early diagnosis and treatment can improve outcomes.

Treatments include:

Monoclonal Antibodies

- Block complement system activation
- Intravenous infusion
- Long (Ravulizumab) or short duration of action (Eculizumab)

Antibiotics

To treat or prevent risk of meningococcal infection associated with administering Ravulizumab and Eculizumab.

Plasma exchange or infusion

Used to remove harmful substances from the blood and provide normal complement proteins.

Dialysis

May be required if the kidneys are severely affected.

Kidney transplant

In some cases, a kidney transplant may be needed if there is irreversible damage.

General Guidelines for aHUS Management

Follow treatment plans and regularly monitor kidney function.

Stay hydrated to support kidney health.

Get vaccinated for common infections to reduce the risk of triggers.

Avoid medications or triggers known to exacerbate aHUS.

Maintain regular follow-ups with healthcare providers.