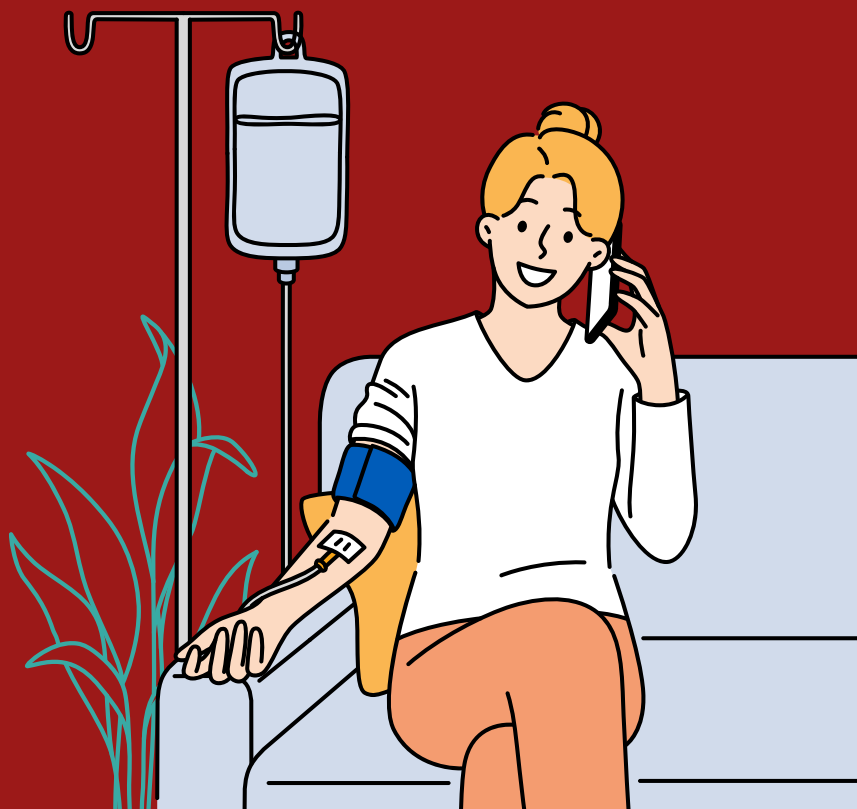
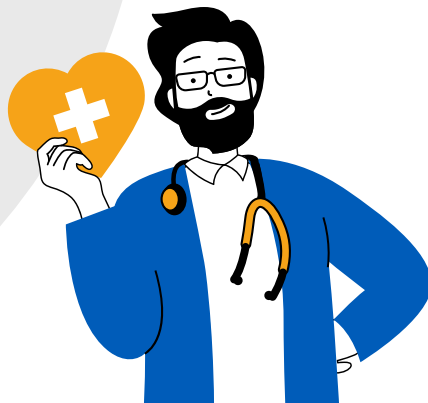


Starting Eculizumab: What you need to know



**A guide for patients
from the National aHUS Service**






You (or your child) have been referred to our service by your local doctors following a recent admission to hospital.



The aims of this leaflet are to:

-  Introduce ourselves and our role in your care
-  Help you understand why you were referred to our service

-  Explain a little about atypical Haemolytic Uraemic Syndrome (aHUS)
-  Tell you about Eculizumab therapy
-  Explain what happens next

Introducing the National aHUS Service

The National aHUS Service is commissioned by NHS England and NHS Scotland. We provide advice on the care and treatment of patients with suspected or confirmed atypical Haemolytic Uraemic Syndrome (aHUS). We work in partnership with your local medical team.

We are involved in your care and treatment decisions because your local medical team referred you to our expert service.

Our experts include doctors, nurses and scientists. We are part of the National Renal Complement Therapeutics Centre (NRCTC), within the Newcastle upon Tyne Hospitals NHS Foundation Trust and Newcastle University.

You may hear us referred to as 'The aHUS service', 'The NRCTC' or just 'Newcastle' by your local team.

Why you were referred to our service

Your local medical team referred you to our service because they wondered if you might have a condition called aHUS.

aHUS is a very rare disease that can be difficult to diagnose because there is no easy test to check whether you have aHUS or not.

Your doctor will have performed tests to rule out other illnesses and because there was a suspicion of aHUS they contacted our team in Newcastle.

All referrals to our service are discussed between our kidney doctors (who have expertise in looking after children and adults with aHUS) and your local medical team providing direct care to you.

We will advise about treatment, including starting eculizumab, and also undertake specialist tests that will help us decide whether or not aHUS is the right diagnosis. These results can take several weeks to come back.



What is aHUS?

aHUS is short for **atypical Haemolytic Uraemic Syndrome**.

Atypical – means this is a less common form of the disease

Uraemic – tells us that the kidneys are involved

Haemolytic – tells us that red blood cells are being destroyed

Syndrome – more than one thing is going on at the same time

i aHUS is an ultra-rare disease caused by a fault in the complement system. The complement system is part of your body's immune response that fights infection.

i Your body has in-built protector proteins to stop complement from attacking your own cells. In aHUS, the protector proteins are not working properly. When this happens, complement starts to attack your body's own cells, especially those that line the blood vessels, which can lead to clots forming.

i The most commonly affected organ is the kidney where the clots can block the tiny filters in the kidney. These filters normally clean toxins from the blood. When blocked, they cannot do their job.



There's a short video explaining aHUS on our website.



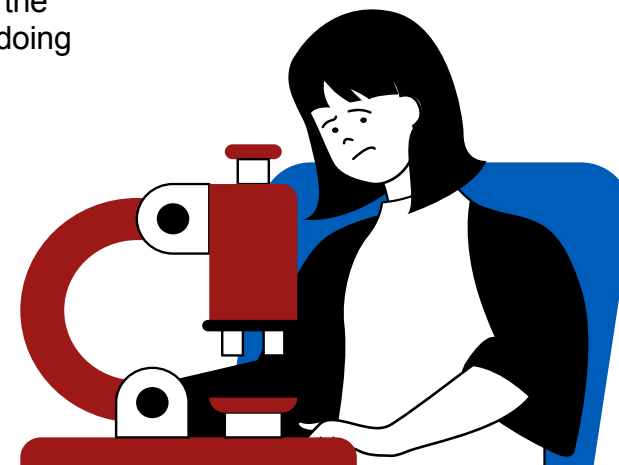
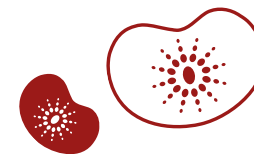
What this means for you

At this point we have not confirmed that you definitely have aHUS. But from the information shared between your local medical team and the National aHUS Service, it is a possibility.

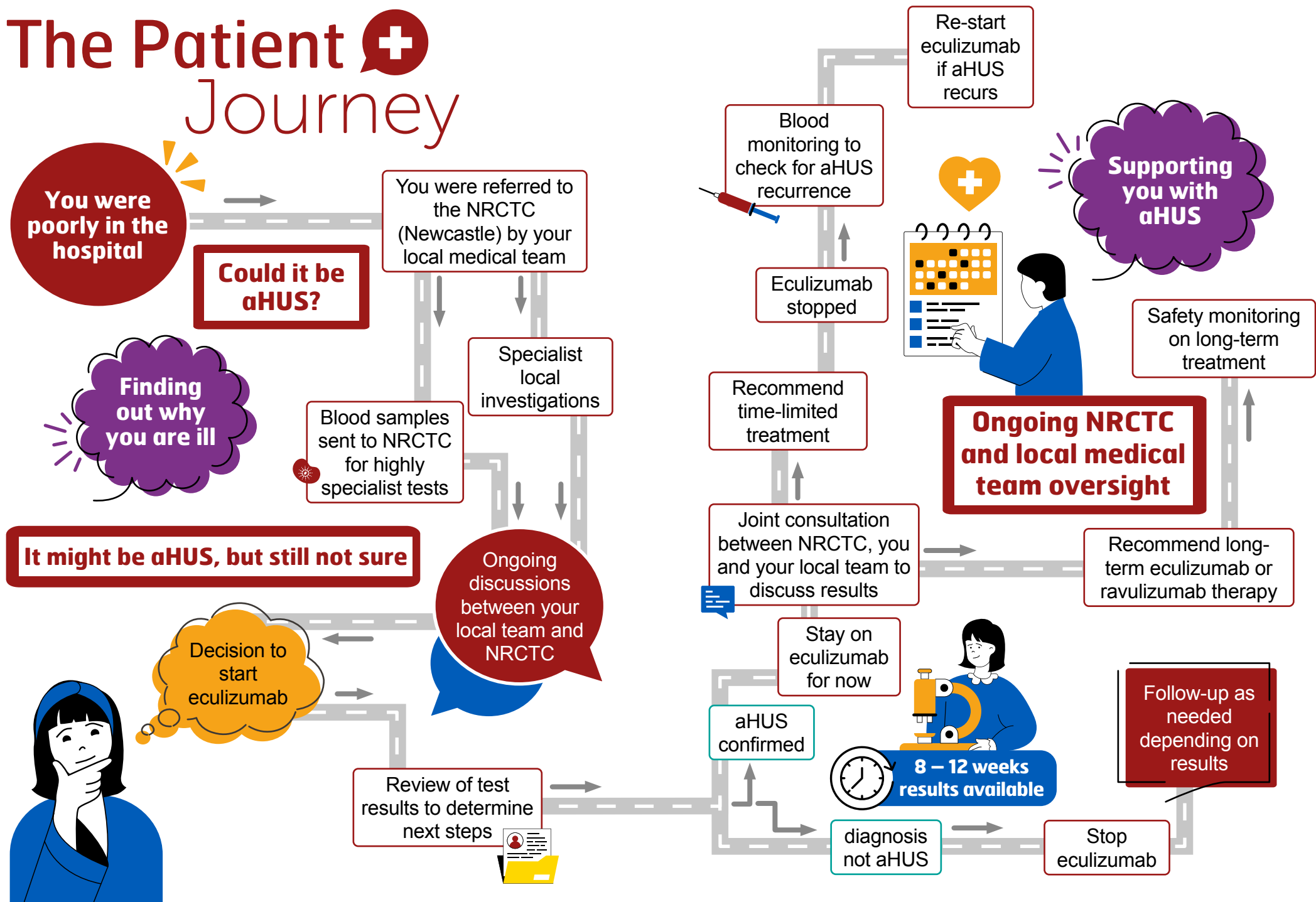
We recommended that you start eculizumab while we wait for the results of the specialist blood tests because we would not want any more damage to your kidneys as a result of a delay in treatment.

The test results can take a few weeks to come back because we look for genetic causes of aHUS, such as changes in the complement system, as well as for autoantibodies that might have developed to stop the protector proteins from doing their job.

Our team recommended that you should start on a treatment called eculizumab. Eculizumab is an approved treatment for aHUS.



The Patient Journey



Treatment for aHUS

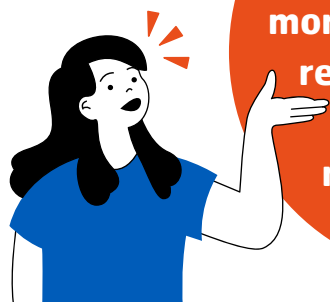
The primary treatment for aHUS is called Eculizumab. You may hear it called different names depending on the drug manufacturer. The other trade names are: **Bekemv®**; **Epysqli®**; and **Soliris®**.

- Eculizumab is an approved and effective treatment for aHUS.
- It works by blocking the action of complement from damaging your body's own cells.
- Eculizumab is given through your veins (intravenously) as an infusion. It usually takes up to an hour for most patients. This can be longer if the infusion is for a child.
- At first, infusions may be weekly then the gaps between infusions will increase depending on the weight of the patient.
- Some patients may later be switched to Ravulizumab, which is an equally effective drug for aHUS. It has longer gaps between infusions. This only happens when patients are stable on Eculizumab and they are going to stay on treatment for a long period of time.

Possible side-effects of treatment

The main side-effect to be aware of is meningococcal infection (which can cause meningitis and/or sepsis).

We recommend meningococcal vaccines and oral antibiotics to reduce your risk of getting this infection.



It is important to know about this in more detail so please read our separate leaflet about meningococcal infection.

What happens next as we assess you for aHUS?

When we have all the results back from your blood tests, we will discuss them with your local doctors. We will also discuss the results of any other tests they are carrying out to look for other possible causes of your symptoms.

If we do not think you have aHUS, we will recommend that the eculizumab is stopped.

If we do think you have aHUS - we will contact you to make an appointment with our doctors to discuss what this means. Longer-term follow-up includes clinics with our aHUS specialist nurses.

In the past, patients with aHUS were given eculizumab and ravulizumab for their lifetime.

This is changing, and most patients will have a time-limited course of treatment. This means the condition will be treated when it is active, but treatment will be stopped while the condition is under control. Patients whose treatment is stopped will be monitored through blood tests, to watch for another episode happening. Treatment can be re-started if needed.

Shared Care

For patients living in England with aHUS, your care is formally shared between the National aHUS Service and your local medical team. Your local medical team are responsible for your kidney care on a day-to-day basis. They will see you in clinic and monitor your kidney function.

Your local medical team and the National aHUS Service share information about you, including your progress on treatment, and together make decisions about your future aHUS management. The National aHUS Service

provides oversight and support to your local medical team in the specialist areas of aHUS care.

For patients living in Scotland, there are no formal shared care arrangements in place between the National aHUS Service and

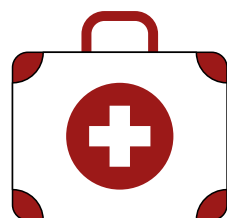
your local medical team. We do still however work closely with your local medical team in relation to your aHUS, and still offer you consultations with the National aHUS Service.

For patients with confirmed aHUS living in either England or Scotland, our service offers

long-term involvement and we run regular specialist nurse consultations with patients and families. These are undertaken either via the telephone or via a video platform (like Teams, Zoom etc).

Does aHUS run in families?

Not all patients with aHUS have a genetic change that has caused their aHUS, but some do.



If we find that you have a genetic change associated with aHUS, we will discuss this with you. We can then also offer testing to your family members.

Where to get more information

We understand this can be a very uncertain time and that you may have more questions. Access to treatments for aHUS varies widely across the world, so some information available online and via social media may not reflect the care available in this country.

For more information about aHUS and support available to patients with rare kidney diseases we recommend the following websites:

- **National aHUS Website** - contains a video and useful information about aHUS, and our service, and can be found at: <https://www.atypicalhus.co.uk>

- **Kidney Care UK** is the UK's leading kidney patient support charity. You can search for "aHUS" and it links to some patient stories as well as other useful information including emotional support: <https://kidneycareuk.org>

- **Kidney Research UK** is the leading kidney research charity in the UK. As well as giving a lot of information about kidney conditions and research, it also contains some useful information for patients about living with kidney disease: <https://www.kidneyresearchuk.org>

How to contact us:



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(aHUS National Service)



Email:

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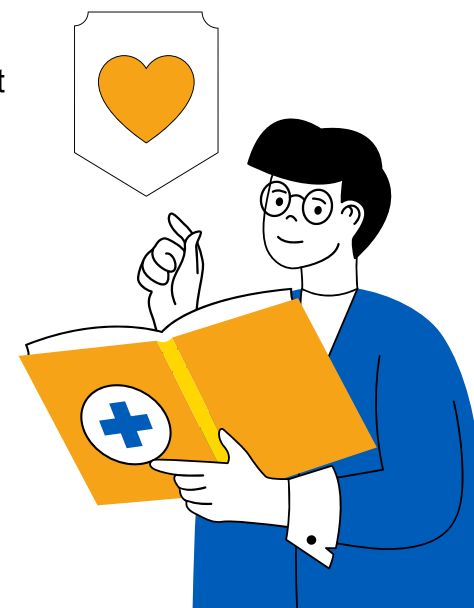


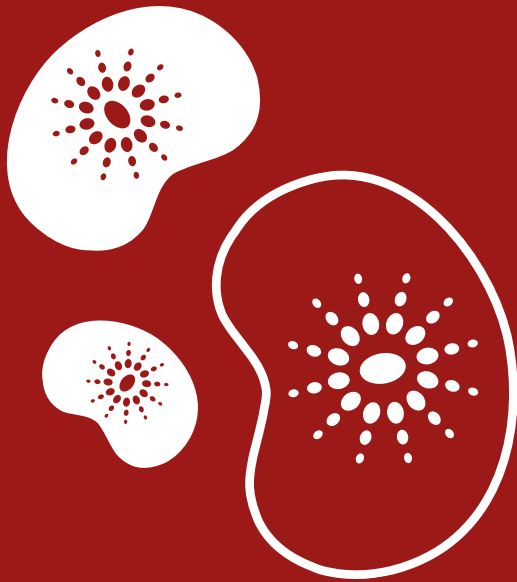
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Our Website



www.atypicalhus.co.uk