



Atypical Haemolytic Uraemic Syndrome (aHUS) Shared Care Protocol

Contact Details

Name:

Location:

Date:

Phone No:

Patient ID Label:

Surname:

Forenames:

NHS Number:

Date of Birth:

Introduction	<p>Eculizumab is available for treatment for patients with aHUS in England only if all the following arrangements are in place</p> <ul style="list-style-type: none"> • coordination of Eculizumab use through an expert centre • monitoring systems to record the number of people with a diagnosis of atypical haemolytic uraemic syndrome and the number who have Eculizumab, the dose and duration of treatment • a national protocol for starting and stopping eculizumab for clinical reasons • a research programme with robust methods to evaluate when stopping treatment or dose adjustment might occur. <p>The expert centre is based at The National Renal Complement Therapeutics Centre, Building number 26, Royal Victoria Infirmary, Newcastle upon Tyne Hospitals NHS Foundation Trust and is known as the National aHUS Service. This protocol outlines the responsibilities of the referring consultant and the National aHUS Service.</p>
Dose & Administration	<p>The dose of Eculizumab at initiation and maintenance is fixed for adults. For children and adolescents dose is adjusted according to body weight. Details of dosing schedules are available at:</p> <p>http://www.atypicalhus.co.uk/ahus/eculizumab-in-the-treatment-of-ahus/dosing-schedules-for-eculizumab/</p>
Responsibilities of the National aHUS Service	<ol style="list-style-type: none"> 1. Consider all patients referred with a thrombotic microangiopathy and possible diagnosis of atypical Haemolytic Uraemic Syndrome (aHUS) and provide a clinical opinion on diagnosis and treatment. 2. Authorise the initiation of treatment with Eculizumab for patients with a likely diagnosis of aHUS who may benefit from treatment and who meet the eligibility criteria.



	<ol style="list-style-type: none"> 3. Inform NHS England that Eculizumab treatment has been recommended for a patient. 4. Perform and review genetic testing for causes of aHUS and report the results to the referring consultant. 5. Provide the patient with information about aHUS and treatment with Eculizumab. 6. Initiate shared care with the referring consultant according to this agreement. 7. Review the progress of patients referred to the National aHUS Service. For patients not treated further progress review will cease upon mutual agreement between the National aHUS Service and the referring consultant 8. Offer the patient the opportunity for appointments with a consultant from the National aHUS Service (either face to face or remotely). When appropriate offer appointments to family members for genetic counselling and provide educational material to family members. Maintain written communication with the referring consultant and patient's general practitioner following all appointments. 9. Provide ongoing advice and information to the referring consultant and patient's general Practitioner as required. 10. Co-ordinate meningococcal vaccination and/or arrange measurement of antibody titres. Share results with referring team and arrange booster vaccinations when appropriate. 11. Provide support and education for healthcare professionals involved in administering Eculizumab and all other aspects of the care of patients with aHUS.
<p>Responsibilities of the Referring Consultant</p>	<ol style="list-style-type: none"> 1. Contact the National aHUS Service if a diagnosis of aHUS is suspected 2. Undertake local investigations described in the National aHUS Service protocol http://www.atypicalhus.co.uk/emergency-referrals/ to establish a cause of the thrombotic microangiopathy and indication for Eculizumab treatment. 3. Send the required samples to the National aHUS Service 4. Liaise with local microbiology laboratory to ensure urgent samples are sent for: <ol style="list-style-type: none"> a) Shiga-toxin producing <i>Escherichia coli</i> (STEC) culture to local microbiology laboratory



- b) Stool for molecular diagnostics should be sent to Bacteriology Reference Department, PHE Colindale
- c) Serodiagnosis of *E.coli* O157 and other shiga toxin producing strains should be sent to Bacteriology Reference Department, PHE Colindale
5. When Eculizumab treatment is authorised prescribe and administer Eculizumab according to the manufacturer's instructions and protocol unless a deviation from protocol is agreed with the National aHUS Service.
6. Notify the National aHUS Service of all diagnostic checklist investigations within one month of starting Eculizumab treatment.
7. Minimise the risk of meningococcal infection by vaccination and the use of prophylactic antibiotics as recommended in the National aHUS Service protocol (<http://www.atypicalhus.co.uk/ahus/eculizumab-in-the-treatment-of-ahus/risk-of-meningococcal-infection-in-patients-receiving-eculizumab/>):
 - a) Vaccinate with a tetravalent (ACWY) vaccine and Bexsero (against serotype B).
 - b) Send oropharyngeal swab to microbiology to exclude carriage of antibiotic resistant meningococcus.
 - c) Prescribe prophylactic antibiotic for the duration of Eculizumab treatment.

Make the patient/parent/carer aware of the increased risk of meningococcal infection and possible symptoms of meningococcal infection.
8. Make the patient or guardian/carer aware of the National aHUS Service and that they will be contacted directly with further information about aHUS and its treatment.
9. Arrange on-going monitoring at the recommended frequencies by the National aHUS Service, including collection of samples to be sent to the National aHUS Service for complement studies
10. Supply patient data to the National aHUS Service, as mandated by NHS England, to allow submission of regular reports of Eculizumab use and patient outcomes.
11. Report any adverse events to the National aHUS Service (consultant or specialist nurse).



	<p>12. Decisions on stopping treatment should be made in consultation with the National aHUS Service.</p> <p>13. Provide a named person (i.e. consultant, doctor or specialist nurse) for each patient who can be contacted by the National aHUS Service in case of questions relating to that patient.</p> <p>14. All patients referred to the National aHUS Service should be offered registration in RADAR and Patient View to allow automatic collection of outcome data irrespective of whether the patient is treated with Eculizumab.</p>
<p>Monitoring Required from Referring Centre</p>	<p>Monitoring of patients receiving Eculizumab treatment is required and the National aHUS Service requires this information to report to NHS England.</p> <p>Monitoring in the early stages of treatment will be based on clinical need and local protocols</p> <p>Complement blockade should be confirmed once established on maintenance dose and annually thereafter or if clinically indicated. This should include AH50 and CH50 to confirm complement blockade and will be performed by the National aHUS Service.</p> <p>In stable patients the following tests are recommended on a monthly basis:</p> <ul style="list-style-type: none"> • Renal function • Full blood count • Lactate dehydrogenase • Haptoglobin • Urinalysis • Urine Protein/Creatinine ratio <p>Further information on monitoring is available at: http://www.atypicalhus.co.uk/ahus/eculizumab-in-the-treatment-of-ahus/monitoring-of-patients-receiving-eculizumab/</p>
<p>Adverse Effects</p>	<p>Eculizumab treatment increases the risk of meningococcal infection. The protocol to minimise risk of infection is available at: (http://www.atypicalhus.co.uk/ahus/eculizumab-in-the-treatment-of-ahus/risk-of-meningococcal-infection-in-patients-receiving-eculizumab/):</p> <p>Details of other treatment related adverse effects are available at: https://www.medicines.org.uk/EMC/medicine/19966/SPC/Soliris/</p>



Referring Consultant

Signed

Date

Print name

Position

An electronic signature by typing your name in the appropriate field is acceptable.

National aHUS Service consultants

Professor David Kavanagh

Signed

Professor Neil Sheerin

Signed

Dr Sally Johnson

Signed

Dr. Edwin Wong

Signed

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