



aHUS Diagnosis Survey

Read this first

1. The Survey is being carried out by aHUS alliance Global Action, an independent registered incorporated charity.
2. The Survey is accessed only through its website, which has an SSL certificate.
3. The Survey Questionnaire is powered by Survey Monkey, a well established and respected survey provider.
4. Survey questions cover pre-symptom, early symptoms, seeking medical advice, referral to specialist stages up to diagnosis. Finally there are questions about post diagnosis treatment status and patients; overall perception of the diagnosis process.
5. The data sought is needed for a study of the experience and perception of the diagnosis process for aHUS.
6. Participant's own response data will be kept anonymously, securely and safely by the charity and not provided to any third party.
7. All data captured will be kept confidentially for two years following the release of results.
8. The results of this study may be published by aHUS alliance Global Action via newsletters, presentations, academic papers, reports and visual graphics with the multiple key aHUS stakeholders including; researchers, multi-disciplinary health care workers, policy makers and other stakeholders. It will not be possible to identify any individuals from reports or presentations.
9. All participation is voluntary and respondents are willing to complete the survey. Respondents can opt out during Survey questionnaire completion, and not answer some of the questions; but once Survey is completed no changes can be made.
10. No individual health care providers' names, nor hospital or other care organisations' names are required, only their roles and services provided are needed.

1. Do you consent to participating in this survey after having fully read the above description of the survey?

- Yes
- No



aHUS Diagnosis Survey

Section 1

Patient Experiences: Survey on the aHUS Diagnostic Process

2. Are you the patient?

- Yes
- No, I am responding on behalf of the patient

3. How old is the patient? (in years)

- Under 18
- 18-24
- 25-34
- 35-44
- 45-54
- 55-64
- 65+

4. Gender of the patient

- Male
- Female
- Other
- Prefer not to answer

5. When was aHUS diagnosed?

- Before 2011
- 2012 to 2015
- After 2015

6. How was the patient's health prior to the first symptoms?

- Very poor
- Poor
- Good
- Very Good
- Excellent

7. Was there any family history of aHUS?

- Yes
- No
- Not known

8. Was there any family history of Kidney Disease?

- Yes
- No
- Not known

9. At the time symptoms began, was the patient:

- Working
- Studying
- Other (please specify, examples: Retired or Infant)

10. Any other comments about patient life/lifestyle PRIOR to aHUS symptoms/diagnosis?



aHUS Diagnosis Survey

Section 2

First signs of noticing health issues prior to diagnosis

11. What symptoms were noticed by the patient (or their parents/caregivers)? (Tick all that apply)

- | | |
|---|---|
| <input type="checkbox"/> Headache | <input type="checkbox"/> Cold and sore throat |
| <input type="checkbox"/> Nausea | <input type="checkbox"/> Confusion or Memory issues |
| <input type="checkbox"/> Diarrhoea | <input type="checkbox"/> Pallor |
| <input type="checkbox"/> Aching Joints | <input type="checkbox"/> Facial or limb swelling |
| <input type="checkbox"/> Breathlessness | <input type="checkbox"/> None |
| <input type="checkbox"/> Other (please specify) | |

12. What was done to treat the patient's symptoms? (Tick all that apply)

- | | |
|--|---------------------------------------|
| <input type="checkbox"/> Pain or fever reducers, available at stores OTC | <input type="checkbox"/> Drank fluids |
| <input type="checkbox"/> Cold/Flu medication | <input type="checkbox"/> None |
| <input type="checkbox"/> Stayed in bed | |
| <input type="checkbox"/> Other (please specify) | |

13. How was patient health after symptoms began, but prior to seeking medical advice?

- | | |
|---------------------------------|---------------------------------|
| <input type="radio"/> Very poor | <input type="radio"/> Very good |
| <input type="radio"/> Poor | <input type="radio"/> Excellent |
| <input type="radio"/> Good | |

14. How many days passed after noticing symptoms until patient saw a doctor?

15. Any other comments?



aHUS Diagnosis Survey

Section 3

Experience of first seeking medical advice

16. Advice was first sought from which doctor?

- Primary Care Practitioner / General Practitioner (GP)
- ER / Acute Care Setting (A&E)
- Was already in hospital due to pregnancy / childbirth, transplant or other condition

17. What tests were carried out at that time? (Tick all that apply)

- Temperature taken
- Blood Pressure
- Hemoglobin for anaemia/uraemia or creatinine
- Urine Dip
- None
- Other (please specify)

18. What was the outcome of the visit?

- Common virus or cold diagnosed, returned home
- Returned home without diagnosis, told to return for care if condition continues/worsens
- Admitted to the Hospital
- Referred to a Specialist
- Other diagnosis (please specify)

19. If you were referred to a Specialist, which kind of specialist was it?

Hematologist

Paediatrician

Nephrologist

Was not referred to any Specialist at initial visit

Oncologist

Other (please specify)

20. How many times did the patient visited primary care practitioner/GP before referral to a specialist? (Please indicate total number of doctor visits.)

21. How many days was it from the first visit to Doctor to being referred to another practice?

22. What was the state of health of the patient when referred to a specialist?

Very poor

Very Good

Poor

Excellent

Good

23. Any other comments?



aHUS Diagnosis Survey

Section 4

Getting a Diagnosis of Atypical HUS

24. By the time patient reached this stage, how were the symptoms?

- Mild Critical
 Worsening Life Threatening
 Debilitating

25. Was patient admitted into the hospital?

- Yes, for regular hospital care
 Yes, into Intensive Care
 No, not admitted to hospital

26. What symptoms did the patient have? (Tick all that apply)

- Kidneys stopped working Heart-Related Issues
 Very high Blood Pressure Anemia
 Body (fluid) swelling Difficulty breathing
 Other (please specify)

27. What tests were carried out on patient? (Tick all that apply)

- Kidney Biopsy Complement Levels
 ADAMTS 13 Genetic Tests
 E. coli Imaging of heart, lungs, or other Organs/Systems
 Blood Tests for anaemia or uraemia Not known
 Blood Test for platelets (thrombocytopenia)
 Other (please specify)

28. Were any of the following given as a possible diagnosis? (Tick all that apply)

- | | |
|--|--|
| <input type="checkbox"/> Thrombotic Microangiopathy (TMA) | <input type="checkbox"/> Medical terms were used, but I didn't understand them |
| <input type="checkbox"/> Thrombotic Thrombocytopenic Purpura (TTP) | <input type="checkbox"/> Not told |
| <input type="checkbox"/> E.coli HUS | <input type="checkbox"/> Don't recall |
| <input type="checkbox"/> Other (please specify) | |

29. Were any of the following only mentioned? (Tick all that apply)

- Thrombotic Microangiopathy (TMA)
- Thrombotic Thrombocytopenic Purpura (TTP)
- E. Coli
- Don't recall
- Other (please specify)

30. If the patient was pregnant, were any of the following given as a possible diagnosis? (Tick all that apply)

- | | |
|---|--|
| <input type="checkbox"/> Eclampsia or Pre-Eclampsia | <input type="checkbox"/> Thrombotic Thrombocytopenic Purpura (TTP) |
| <input type="checkbox"/> HELLP | <input type="checkbox"/> Not told |
| <input type="checkbox"/> Anti Phospholipid Syndrome | <input type="checkbox"/> Does not apply |
| <input type="checkbox"/> Lupus | |
| <input type="checkbox"/> Other (please specify) | |

31. If the patient was pregnant or given birth, were any of the following only mentioned? (Tick all that apply)

- | | |
|---|---|
| <input type="checkbox"/> Eclampsia or Pre-Eclampsia | <input type="checkbox"/> TTP |
| <input type="checkbox"/> HELLP | <input type="checkbox"/> Not told |
| <input type="checkbox"/> Anti Phospholipid Syndrome | <input type="checkbox"/> Does not apply |
| <input type="checkbox"/> Lupus | |
| <input type="checkbox"/> Other (please specify) | |

32. Was aHUS diagnosed by first specialist referred to?

- Yes
- No

33. Which specialist gave the patient "aHUS" Diagnosis?

Hematologist

Paediatrician

Nephrologist

Diagnosis was determined by a multi-disciplinary team of doctors from more than 1 specialty area

Oncologist

Other (please specify)

34. Before changing or identifying a diagnosis, how many weeks passed before the specialist diagnosed the patient with aHUS?

35. Any other comments?



aHUS Diagnosis Survey

Section 5

After Patient's aHUS Diagnosis

36. What treatment did patient receive after being diagnosed with aHUS?

- Eculizumab
- Ravulizumab
- Plasma Infusions or Plasma Exchanges (PEX, pheresis, plasmapheresis)
- Immunosuppresants / Chemotherapy
- Other (please specify)

37. What is the current status of the patient?

- On treatment
- On treatment with transplant
- In remission without treatment
- In remission on Dialysis
- Other (please specify)

38. What is the current health status of the patient?

- Very poor
- Poor
- Good
- Very Good
- Excellent

39. Any other comments?



aHUS Diagnosis Survey

Section 6

Insights Into Attitudes about the Diagnosis & Care Plan

40. Choose which option most closely relates to your attitude regarding the overall diagnosis process

- It was not approached any differently from other healthcare issues, so I felt relaxed about getting info & options
- It was a little more complicated than I thought it would be to get an explanation but felt confident about it
- I did not know how hard and prolonged it would be to get a diagnosis of what was wrong with me , my confidence was really shaken
- I was extremely anxious that doctors did not seem to know what I had and what to do

41. What best describes the psychological impacts surrounding aHUS clinical care?

- Anxiety over treatment was not addressed by the clinical care team
- There were opportunities & resources available through our care team to deal with emotional & economic burdens
- Different treatment options were explained so that we understood the implications and challenges for patient care and lifestyle
- Our clinical team focused only on caring for the physical aspects of aHUS without addressing emotional, social, or economic impacts for me or my family

42. With hindsight what could have speeded up your aHUS diagnosis? (Pick at most 3 options)

- Greater awareness of Thrombotic Microangiopathy (TMA)
- Guidelines available on identifying a TMA
- Understanding of the different causes of TMA
- Greater ability to recognise Acute Kidney Injury
- An aHUS specific Blood Test
- Awareness of family history of aHUS/Kidney Disease
- Speedier transfers between health care providers
- Other (please specify)