deciphehr™
connecting the dots of rare disease

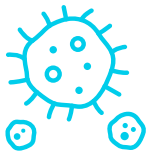
In atypical-HUS, every moment matters; leveraging your EHR may help.

- Atypical-HUS is a rare disease associated with continuous risk of complement-mediated TMA and life-threatening consequences[1,2]
- A rapid and accurate diagnosis is critical to potentially optimize patient outcomes[2,3]
- The similarity in the presentation of atypical-HUS and other TMAs combined with the absence of a single diagnostic test can make a rapid diagnosis challenging[4]

The deciphEHR™ program provides educational resources on disease characteristics and diagnostic best practices to help healthcare providers, health systems, hospitals, and specialty practices leverage their EHR systems to triage suspect patients for further clinician evaluation leading to rapid, accurate diagnoses.

atypical-HUS=atypical hemolytic uremic syndrome; EHR=electronic health record; TMA=thrombotic microangiopathy

Why is it important to rapidly diagnose atypical-HUS patients?



Atypical-HUS is a serious condition that can quickly lead to ESRD and death in adult and pediatric patients.[5*]

7 days is the **maximum** suggested time to improve the atypical-HUS diagnostic and management process.[6]

Patients with atypical-HUS may be mistreated with plasma therapy. Atypical-HUS patients have poor long-term outcomes to plasma-based therapy despite transient normalization of hematologic parameters.[3,7,8]

46% of adults and **17%** of children reached **ESRD** or **death** within **<1 month after disease onset**[5*]

*Based on a study of 214 atypical-HUS patients who received care in France, excluding all cases of secondary atypical-HUS except pregnancy. 89 of the patients were categorized as children (<16 years old) and 125 as adults (≥16 years old).

ESRD=end-stage renal disease

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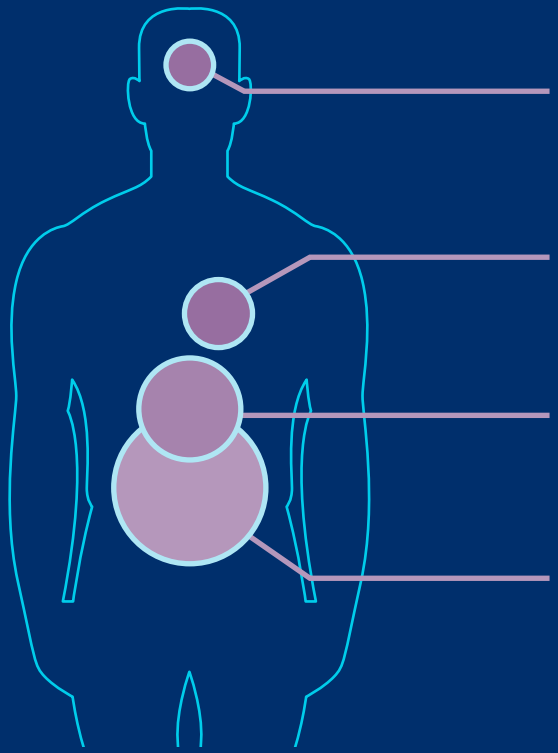
Atypical-HUS patients experience life-threatening complications[2,9*]



UP TO 25% experience neurological symptoms[1]

UP TO 33% experience cardiovascular symptoms[1]

UP TO 47% experience gastrointestinal symptoms[1]

UP TO 83% have acute renal failure at presentation[10]

Additional signs and symptoms may include*: Macrovascular complications such as peripheral arterial disease and phalangeal gangrene.[11-13] **Pulmonary pathology is also frequent in untreated atypical-HUS, but is virtually never directly involved in TTP.[14]**

*Not an exhaustive list.

TTP=thrombotic thrombocytopenia purpura

Atypical-HUS can be a challenging diagnosis[4]

Atypical-HUS can present concurrently with or be triggered by other TMAs[3]

~70% of patients with atypical-HUS present following a triggering event.[15]

Complement-triggering conditions[16]*

21.9%
Infections

18.9%
Malignancy

11.7%
Transplant (organ/bone marrow)

11.7%
Autoimmune disease (eg, SLE, APS, scleroderma)

8.5%
Malignant hypertension or hypertensive emergency

1.3%
Certain prescription medications or illicit drugs

Additional triggering events can include[3]:

- Pregnancy/postpartum/HELLP/preeclampsia
- Surgery/trauma

There is currently no confirmatory test to conclusively diagnose atypical-HUS; a clinical diagnosis requires differentiation from other TMAs and triggers.[2,3,4]

*Based on a study of 349 patients with triggering/associated events prior/up to atypical-HUS onset and enrolled in the Global aHUS Registry from 2011 to July 2021. 229 of the patients were categorized as adults (≥18 years old) and 78 as pediatric patients (<18 years old).

APS=antiphospholipid syndrome; HELLP=hemolysis, elevated liver enzymes, low platelet counts; SLE=systemic lupus erythematosus.

Missed and late diagnosis of atypical-HUS can lead to devastating consequences for patients[1,2,3]

89%
of patients are initially misdiagnosed[17]

53%
of patients remain undiagnosed beyond the suggested 7-day maximum[17]

30%
of patients remain undiagnosed for more than 31 days[17]

A quarter of patients die during the acute phase[3]

Over half of adults progress to ESRD or die within 1 year of initial presentation[5]

About 80% of patients progress to ESRD or die within 3 years of initial presentation[15]

Leveraging EHR data may help healthcare organizations rapidly triage atypical-HUS patients for further clinical evaluation[18]

The problem

- Atypical-HUS patients face **delays in diagnosis**, which can result in **life-threatening consequences**[17]
- Delays in diagnoses are driven in part due to the similarity in the presentation of TMAs combined with the absence of a single diagnostic test[4]

deciphEHR™ may be able to help

- **Your EHR system can work for you to help triage suspect atypical-HUS patients**—Alexion provides resources for you to share with your EHR team
- deciphEHR™ program identifies **clinical features** that may be used to build **suspect patient lists** in your EHR to triage patients for further clinical evaluation
- **Best practice alerts (BPAs)** and **order sets** can be utilized in your EHR system to support healthcare providers as they navigate the atypical-HUS diagnostic process

The benefits

- EHR systems can help triage patients based on existing data, prioritize resources, and provide more **coordinated care that may foster improved outcomes**[19]
- Automated **BPAs** and **order sets** assist providers in timely access to diagnostic best practices and **reduce inefficiency** by decreasing manual efforts[19-22]

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For more information on clinical features of atypical-HUS, suspect patient lists, and BPAs, consult the atypical-HUS Program Implementation Guide.

Alexion provides educational resources to help you leverage your EHR, which may decrease the diagnostic timeline for many atypical-HUS patients

- A vast majority of atypical-HUS patients are facing large delays in diagnosis and misdiagnosis[1,7]
- The similarity in the presentation of atypical-HUS and other TMAs combined with the absence of a single diagnostic test makes a rapid differential diagnosis challenging[4]
- Late or misdiagnosis of atypical-HUS is often treated inappropriately with plasma exchange, which can lead to poor long-term patient outcomes such as ESRD or death[3,7]
- Improving the diagnostic and disease management process can decrease the burden on the healthcare system[6]
- The data needed to shorten diagnostic delays may exist in your EHR
- The deciphEHR™ program has suggested EHR codes that may be used to build patient lists to flag suspect atypical-HUS patients for further clinical evaluation

Visit deciphEHRrare.com or contact your Alexion representative to find out how utilizing your EHR system can help you triage patients who would benefit from further clinical evaluation for atypical-HUS.

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