





# Hemolytic Uremic Syndrome, Post-Diarrheal Investigation Guideline

## Contents

<b>CASE DEFINITION</b> .....	<b>1</b>
<b>LABORATORY ANALYSIS</b> .....	<b>2</b>
<b>INVESTIGATOR RESPONSIBILITIES</b> .....	<b>4</b>
<b>DATA MANAGEMENT AND REPORTING TO THE KDHE</b> .....	<b>4</b>
<b>ADDITIONAL INFORMATION / REFERENCES</b> .....	<b>5</b>
<b>ATTACHMENTS</b> .....	<b>5</b>
Fact Sheet (vs. 2012) 	

*Attachments can be accessed through the Adobe Reader's navigation panel for attachments. Throughout this document attachment links are indicated by this symbol ; when the link is activated in Adobe Reader it will open the attachments navigation panel. The link may not work when using PDF readers other than Adobe.*

**Revision History:**

<b>Date</b>	<b>Replaced</b>	<b>Comments</b>
05/2018	06/2012	Notification Section and restriction section modified with requirements of new reporting regulations.
06/2012	03/2009	Formatting changes. Changes to Laboratory Analysis and Investigator Responsibilities. Addition of Notification section and removal of Standard Investigation section. (Removed references to KS-EDSS (2/2012))

# Hemolytic Uremic Syndrome, Post-Diarrheal Disease Management and Investigation Guidelines

---

## CASE DEFINITION

### Clinical Description for Public Health Surveillance:

- Hemolytic uremic syndrome (HUS) is characterized by the acute onset of microangiopathic hemolytic anemia, renal injury, and low platelet count. Thrombotic thrombocytopenic purpura (TTP) also is characterized by these features but can include central nervous system (CNS) involvement and fever and may have a more gradual onset. Most cases of HUS (but few cases of TTP) occur after an acute gastrointestinal illness (usually diarrheal).

### Laboratory Criteria for Case Classification:

The following are both present at some time during the illness:

- Anemia (acute onset) with microangiopathic changes (i.e., schistocytes, burr cells, or helmet cells) on peripheral blood smear and
- Renal injury (acute onset) evidenced by either hematuria, proteinuria, or elevated creatinine level (i.e., greater than or equal to 1.0 mg/dl in a child aged less than 13 years or greater than or equal to 1.5 mg/dl in a person aged greater than or equal to 13 years, or greater than or equal to 50% increase over baseline)

**Note:** A low platelet count can usually, but not always, be detected early in the illness, but it may then become normal or even high. If a platelet count obtained within 7 days after onset of the acute gastrointestinal illness is not less than 150,000/mm<sup>3</sup>, other diagnoses should be considered.

### Case Classification:

- **Confirmed:** An acute illness diagnosed as HUS or TTP that both meets the laboratory criteria and began within 3 weeks after onset of an episode of acute or bloody diarrhea
- **Probable:**
  - An acute illness diagnosed as HUS or TTP that meets the laboratory criteria in a patient who does not have a clear history of acute or bloody diarrhea in preceding 3 weeks or
  - An acute illness diagnosed as HUS or TTP that a) has onset within 3 weeks after onset of an acute or bloody diarrhea and b) meets the laboratory criteria except that microangiopathic changes are not confirmed

## LABORATORY ANALYSIS:

Hemolytic uremic syndrome (HUS) is a clinical diagnosis based on laboratory tests that show signs of hemolytic anemia and acute renal failure.

Because HUS has been demonstrated to be an important sequelae of *E. Coli* 0157:H7 infection, HUS can serve as a marker for *E. Coli* 0157:H7 activity and may lead to the identification of outbreaks at the state or local level.

Initial laboratory tests may include:

- Blood clotting tests (PT and PTT)
- Comprehensive metabolic panel may show increased BUN and creatinine.
- Complete blood count (CBC) may show increased white blood cell count and decreased red blood cell count.
- Platelet count is usually reduced.
- Urinalysis may reveal blood and protein in the urine
- Urine protein test can be used to show the amount of protein in the urine
- Stool culture may be positive for a type of *E. coli* bacteria or other bacteria

Identifying source of diarrhea causing organism is appropriate.

- Collection: Use an enteric kit (bottle with a Cary-Blair medium (0.16% agar))
- Specimen: Feces
- Amount: Marble size (preferred) or two rectal swabs per container.

For additional information and/or questions concerning specimens and laboratory kits call (785) 296-1620.

## EPIDEMIOLOGY

Hemolytic uremic syndrome (HUS) occurs as a complication in about 8% of diagnosed *E. coli* 0157:H7 cases, particularly children. It is also a complication in *Shigella dysenteriae* infections.

## DISEASE OVERVIEW

### A. Agent:

Most often a complication of infection with a shiga toxin-producing *E. coli* (STEC), most commonly *E. coli* 0157:H7, or with *Shigella dysenteriae*.

### B. Clinical Description:

Syndrome presenting after an acute gastrointestinal illness characterized by microangiopathic hemolytic anemia, acute renal failure and thrombocytopenia. Early clinical signs include decreased urine output, pallor and lethargy. A varying degree of renal insufficiency develops sometimes necessitating kidney dialysis or resulting in total renal failure. There is also increased risk of stroke and other complications.

### C. Reservoirs:

Cattle are the reservoir of significant public health importance; however, other animals, such as goats, sheep, and deer, are known to be carriers. Humans are a reservoir for person-to-person and *Shigella dysenteriae* transmission.

**D. Mode(s) of Transmission:**

Transmission occurs from consuming food or liquids, including water, contaminated with human or animal feces. Person-to-person transmission may occur via the fecal-oral route; including certain types of sexual contact (e.g., oral-anal contact).

**E. Incubation Period:**

In cases of *E. coli* 0157:H7, HUS occurs 2-14 days after onset of diarrhea.

**F. Period of Communicability:**

Varies with agent, for as long as the organism is excreted; typically 1 week in adults and up to 3 weeks in some children with *E. coli* 0157:H7. HUS case documented as enteric culture negative is assumed to be non-communicable.

**G. Susceptibility and Resistance:**

The infectious dose is very low and little is known about differences in susceptibility between serotypes with *E. coli*.

**H. Treatment**

Treatment of HUS is supportive including fluid and electrolyte replacement therapy and when appropriate kidney dialysis:

**NOTIFICATION TO PUBLIC HEALTH AUTHORITIES**

Suspected cases of hemolytic uremic syndrome, postdiarrheal shall be reported within 24 hours, except if the reporting period ends on a weekend or state-approved holiday, the report shall be made by 5:00 p.m. on the next business day after the 24-hour period:

1. Health care providers and hospitals: report to the local public health jurisdiction.
2. Local public health jurisdiction: report to KDHE-BEPHI (see below).
3. Laboratories: report to KDHE-BEPHI (see below).


**Kansas Department of Health and Environment (KDHE)  
Bureau of Epidemiology and Public Health Informatics (BEPHI)  
Phone: 1-877-427-7317  
Fax: 1-877-427-7318**

**Further responsibilities of state and local health departments to the CDC:**

As a nationally notifiable condition, confirmed and probable cases require a ROUTINELY NOTIFIABLE report to the Center of Disease Control and Prevention (CDC).

1. ROUTINE reporting requires KDHE-BEPHI to file an electronic report for within the next reporting cycle.
2. **Local public health jurisdiction** will report information requested as soon as possible, ensuring that the electronic form is completed within 5 days of receiving a notification of a report.

## INVESTIGATOR RESPONSIBILITIES

- 1) [Report](#) all hemolytic uremic syndrome, post-diarrheal cases to the KDHE.
  - Initiate the case investigation within 3 days of notification of a report.
  - Complete the investigation within 5 days of the notification.
- 2) Contact medical provider to collect additional information and confirm diagnosis using current [case definition](#). For all diagnosed cases:
  - Verify that the HUS is post-diarrheal.
  - Collect all data that is requested on the [HUS Reporting Form](#).
- 3) Further investigation, depends upon the confirmed or suspected etiological agent. (Refer to the associated disease investigation guideline).
  - Every attempt should be made to collect stool to identify the agent.
- 4) A case of post-diarrheal HUS with no causative agent identified is considered a suspect STEC case and should be investigated as directed in the [Shiga Toxin-Producing E. coli \(STEC\) Disease Investigation Guidelines](#).
- 5) [Record](#) data, collected during the investigation, in the KS EpiTrax system under the data's associated [\[tab\]](#) in the case morbidity report (CMR).
- 6) As appropriate, use the disease [fact sheet](#) .

## DATA MANAGEMENT AND REPORTING TO THE KDHE

- A. Accept the case assigned to the LHD and record the date the LHD investigation was started on the [\[Administrative\]](#) tab.
- B. Organize and collect data, using appropriate data collection tools including:
  - The [HUS Reporting Form](#) can be used to collect information.
  - Alternatively, investigators can collect and enter all required information directly into EpiTrax [\[Investigation\]](#), [\[Clinical\]](#), [\[Demographics\]](#), [\[Epidemiological\]](#) tabs.
- C. Report data collected during the course of the investigation via EpiTrax.
  - Verify that all data requested on the [HUS Reporting Form](#) has been recorded on an appropriate EpiTrax [\[tab\]](#)
  - Some data that cannot be reported on an EpiTrax [\[tab\]](#) may need to be recorded in [\[Notes\]](#) or scanned and attached to the record.
  - Paper report forms do not need to be sent to KDHE after the information is recorded and/or attached in EpiTrax. The forms should be handled as directed by local administrative practices.
- D. Once the investigation is completed, the LHD investigator will record the date the investigation was completed on the [\[Administrative\]](#) tab and click the "Complete" button. This will trigger an alert to the LHD Administrator so they can review the case before sending to the state.
  - The LHD Administrator will then "Approve" or "Reject" the CMR.
  - Once a case is "Approved" by the LHD Administrator, BEPHI staff will review and close the case after ensuring it is complete and that the case is assigned to the correct event, based on the reported symptoms reported. (Review the [EpiTrax User Guide, Case Routing](#) for further guidance.)


## ADDITIONAL INFORMATION / REFERENCES

- A. **Treatment / Differential Diagnosis:** Red Book: 2015 Report of the Committee on Infectious Diseases. 30th ed. Elk Grove Village, IL: American Academy of Pediatrics; 2015.
- B. **Epidemiology, Investigation and Control:** Heymann. D., ed., Control of Communicable Diseases Manual (CCDM), 20th Edition. Washington, DC, American Public Health Association, 2015.
- C. **Case Definitions:** CDC Division of Public Health Surveillance and Informatics, Available at: [wwwn.cdc.gov/nndss/](http://wwwn.cdc.gov/nndss/)
- D. **Kansas Regulations/Statutes Related to Infectious Disease:** [www.kdheks.gov/epi/regulations.htm](http://www.kdheks.gov/epi/regulations.htm)
- E. **Animals in Public Places Compendium:** [www.kdheks.gov/epi/human\\_animal\\_health.htm](http://www.kdheks.gov/epi/human_animal_health.htm)
- F. **KDHE Foodborne Illness Resources:** [www.kdheks.gov/epi/foodborne.htm](http://www.kdheks.gov/epi/foodborne.htm)
- G. **Additional Information (CDC):** [www.cdc.gov/ticks/index.html](http://www.cdc.gov/ticks/index.html)

## ATTACHMENTS

- **KDHE HUS Report Form**
- **Fact Sheet**

To view attachments in the electronic version:

1. Go to <View>; <Navigation Pane>; <Attachments> – OR – Click on the “Paper Clip”  icon at the left.
2. Double click on the document to open