

# Hemolytic Uremic Syndrome, Post-Diarrheal Investigation Guideline

## CONTENT:

## VERSION DATE:

### Investigation Protocol:

- Investigation Guideline

06/2012

### Supporting Materials found in attachments:

- Fact Sheet

05/2012

## Revision History:

Date	Replaced	Comments
06/2012	03/2009	Formatting changes. Changes to Laboratory Analysis and Investigator Responsibilities. Addition of Notification section and removal of Standard Investigation section.
02/2012	-	Removed references to KS-EDSS.

# Hemolytic Uremic Syndrome, Post-Diarrheal Investigation Guidelines

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## CASE DEFINITION (CDC 1996)

### 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 hemolytic uremic syndrome (HUS) has been demonstrated to be an important sequelae of infection with *E. coli* 0157:H7, HUS can serve as a marker for *E. coli* 0157:H7 activity in the community 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 levels of 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 certain 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 specimen collection/transport and laboratory kits call (785) 296-1620 or refer to online guidance at [http://www.kdheks.gov/labs/lab\\_ref\\_guide.htm](http://www.kdheks.gov/labs/lab_ref_guide.htm).

## 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 serve as a reservoir for person-to-person transmission and with *Shigella*.

**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**

Hemolytic uremic syndrome, postdiarrheal, shall be designated as infectious or contagious in their nature, and cases or suspect cases shall be reported within seven days:

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, shigellosis cases require a STANDARD report to the Center of Disease Control and Prevention (CDC).*

1. STANDARD reporting requires KDHE-BEPHI to file an electronic report for cases within the next reporting cycle.
  - KDHE-BEPHI will file electronic reports weekly with CDC.
2. Local public health jurisdiction will report information as requested in the Kansas electronic surveillance system, as soon as possible, ensuring that the electronic form is completed within 7 days of receiving a notification of a report.

## INVESTIGATOR RESPONSIBILITIES

- 1) [Report](#) all confirmed, probable and suspect cases to the KDHE-BEPHI.
- 2) Use the [case definition](#), to confirm the diagnosis with the medical provider.
  - Verify that the HUS is post-diarrheal.
  - Collect all data from the medical record and laboratory reports 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 a stool culture to identify the causative agent.
  - A case of post-diarrheal HUS or TTP with no causative agent identified is considered a suspect case of Shiga toxin-producing *E. coli* and should be investigated as directed in the “[Shiga Toxin-Producing E. coli \(STEC\) Disease Investigation Guidelines](#)”.
- 4) Complete and report information requested in the state electronic surveillance system.
- 5) As appropriate, use the notification letter(s) and the disease [fact sheet](#) to notify the case, contacts and other individuals or groups.

## DATA MANAGEMENT AND REPORTING TO THE KDHE

- A. Organize and collect data.
- B. Report data via the state electronic surveillance system.
  - Especially data that collected during the investigation that helps to confirm or classify a case.

**Note:** If a patient meets the case definition for both Shiga toxin-producing *E. coli* (STEC) or Shigellosis and Hemolytic Uremic Syndrome (HUS), the case should be reported for each of the conditions. A case of post-diarrheal HUS or TTP with no causative agent identified is considered a suspect case of “Shiga toxin-producing *Escherichia coli* (STEC)” and should be reported as a Shiga toxin-producing *E. coli* with the case classification marked as suspect.

## ADDITIONAL INFORMATION / REFERENCES

- A. **Treatment / Differential Diagnosis:** American Academy of Pediatrics. 2009 Red Book: Report of the Committee on Infectious Disease, 28th Edition. Illinois, Academy of Pediatrics, 2009.
- B. **Epidemiology, Investigation and Control:** Heymann. D., ed., Control of Communicable Diseases Manual, 19th Edition. Washington, DC, American Public Health Association, 2009.
- C. **Case Definitions:** CDC Division of Public Health Surveillance and Informatics, Available at: [www.cdc.gov/osels/ph\\_surveillance/nndss/casedef/case\\_definitions.htm](http://www.cdc.gov/osels/ph_surveillance/nndss/casedef/case_definitions.htm)
- D. **Quarantine and Isolation:** Kansas Community Containment Isolation/ Quarantine Toolbox Section III, Guidelines and Sample Legal Orders [www.kdheks.gov/cphp/operating\\_guides.htm](http://www.kdheks.gov/cphp/operating_guides.htm)
- E. **Kansas Regulations/Statutes Related to Infectious Disease:** [www.kdheks.gov/epi/regulations.htm](http://www.kdheks.gov/epi/regulations.htm)
- F. **KDHE Foodborne Illness and Outbreak Investigation Manual:** Available at: [www.kdheks.gov/epi/download/kansas\\_foodborne\\_illness\\_manual.pdf](http://www.kdheks.gov/epi/download/kansas_foodborne_illness_manual.pdf)
- G. **KDHE Control of Enteric Disease Outbreaks in Childcare Facilities:** [www.kdheks.gov/epi/download/Enteric\\_Disease\\_in\\_Daycare\\_Centers.pdf](http://www.kdheks.gov/epi/download/Enteric_Disease_in_Daycare_Centers.pdf)
- H. **Animals in Public Places Compendium:** [www.kdheks.gov/epi/human\\_animal\\_health.htm](http://www.kdheks.gov/epi/human_animal_health.htm)
- I. **KDHE Foodborne Illness Resources:** [www.kdheks.gov/epi/foodborne.htm](http://www.kdheks.gov/epi/foodborne.htm)
- J. **Additional Information (CDC):** [www.cdc.gov/health/default.htm](http://www.cdc.gov/health/default.htm)

## 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.