

Hemolytic Uremic Syndrome (HUS); Post-diarrheal Surveillance Protocol



Provider Responsibilities

Report all cases and any suspect cases to your local health department within 24 hours of diagnosis. Any cluster of cases should be reported to your local health department immediately.

Laboratory Responsibilities

None – diagnosis is based upon clinical findings.

The WV Office of Laboratory Services (OLS) does not provide laboratory testing for diagnosis of acute renal disease. However, OLS will test stool specimens for the presence of *Shigella*, *E.coli* O157:H7, or other Shiga toxin-producing organisms. For more information contact 304-558-3530 or see www.wvdhhr.org/labservices.

Local Health Responsibilities

1. Educate providers and the public on the connection between HUS and Shiga toxin-producing organisms, especially STEC O157.
2. Educate providers to report any case or suspected case of HUS to post-diarrheal the local health department in the patient's county of residence within 24 hours of diagnosis.
3. Obtain copy of medical records. Several pieces of clinical information will be needed to ascertain case status. For assistance in evaluating clinical information and ascertaining case status contact Division of Infectious Disease Epidemiology.
4. Since most HUS cases are secondary to STEC infection, inquire for a history of diarrheal illness. If there is a history of diarrheal illness, identify potential exposure sources (See STEC protocol for details; food, water, livestock, etc.). Inquire about high risk exposures for 2 to 10 days prior to onset of diarrhea.
5. Determine whether the case attends or works at a daycare facility and/or is a food handler. If case is in one of these high risk positions, contact Division of Infectious Disease Epidemiology to discuss the situation and any follow up that may be needed.
6. Report case information by completing all sections possible of the WVEDSS Hemolytic Uremic Syndrome Post-diarrhea Reporting Form. If laboratory testing

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has identified *E coli* O157:H7 or any other STEC, the case must also be reported in WVEDSS under that reportable condition.

State Health Responsibilities

1. Prompt and complete reporting of HUS cases to the Centers for Disease Control and Prevention (CDC) through WVEDSS.
2. Provide technical expertise and consultation regarding surveillance, investigation, control measures and prevention of HUS.
3. Notify CDC of suspected outbreaks identified in West Virginia and assist local health jurisdictions in obtaining the knowledge and resources necessary for investigation of a HUS outbreak.
4. Summarize surveillance data for cases of HUS on an annual basis.

Disease Control Objectives

Reduce the risk of additional cases by:

- Rapid and complete investigation of cases so that any unrecognized outbreak of STEC or other diarrheal pathogen can be identified and controlled.
- Early identification and appropriate exclusion of infected persons from high risk situations (day care, food handling, health care).

Disease Prevention Objectives

Since most cases of HUS are caused by *E.coli* O157:H7, reduce the risk of infection from STEC by:

- Education of the general public about hand washing as a primary means of preventing person-to-person transmission of STEC.
- Education of the general public about proper food handling, including thorough cooking of ground meat and washing of fruits and vegetables prior to consumption, and avoidance of cross-contamination.
- Education of the general public to avoid unsafe foods such as unpasteurized milk, cheese, juice, cider, and untreated water.

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Disease Surveillance Objectives

- To determine the incidence of HUS in West Virginia
- To identify demographic characteristics of persons with HUS
- To identify risk factors associated with development of HUS

Public Health Significance

Children under 5 are at the highest risk of developing HUS and occurs in 15% of children with *E. coli* O157:H7 diarrhea. Because HUS develops a week or more after onset of diarrhea, a bacterial pathogen is often not laboratory-confirmed in cases. Therefore, the proportions of cases of HUS due to specific bacterial infections are difficult to ascertain.

Clinical Description

HUS is a complication subsequent to infection with Shiga toxin-producing bacteria such as *Shigella*, or most commonly *E. coli* O157:H7. HUS is a combination of microangiopathic hemolytic anemia, thrombocytopenia, and acute renal failure. In the United States, HUS is the principal cause of acute kidney failure in children, and most cases are caused by *E. coli* O157:H7. The illness is very serious and develops during the 2 weeks after onset of diarrhea. Fifty percent of patients require dialysis and 3% to 5% do not survive. Patients with HUS can develop neurologic complications (e.g. seizures, coma, or cerebral vessel thrombosis).

Etiologic Agent

HUS is a clinical syndrome consisting of anemia from red blood cell destruction and impaired renal function. Among children, the most common cause of HUS is infection with a Shiga toxin-producing organism. The majority of cases of HUS are caused by *E. coli* O157:H7, however it can be caused by *Shigella dysenteriae*, other strains of Shiga toxin-producing *E. coli* (STEC), and very rarely by other viral and bacterial infections such as *Salmonella*, *Campylobacter*, and *Streptococcus pneumoniae*.

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Reservoir

The main reservoir for *E.coli* O157:H7 is the intestines of healthy cattle. Sheep deer and other ruminants can also serve as reservoirs for the bacteria, although to a much lesser extent. Humans are the only known reservoir for *Shigella dysenteriae* type 1.

Mode of Transmission

HUS as a clinical syndrome cannot be transmitted. However, *E.coli* O157:H7, the bacteria typically responsible for development of HUS is transmitted by a number of routes; foodborne, waterborne, direct contact with animals and their environment, and person-to-person. *Shigella*, the other bacterial cause of HUS is usually spread by the fecal oral route through person-to-person contact.

Incubation Period

HUS usually occurs 7 days following the onset of diarrhea but can occur up to 3 weeks later. In some cases, diarrhea may have resolved when HUS occurs. (For the incubation periods of the specific bacteria, refer to the protocol for Shiga Toxin-producing *E. coli* and Shigellosis.

Period of Communicability

People with HUS would only be infectious if they are shedding the bacteria which caused the clinical syndrome (e.g. *E.coli* O157:H7 (another STEC serotype) or *Shigella*) in their stool. Refer to the protocols for each specific pathogen for details on the organism's infectious period.

Outbreak Recognition

Typically, outbreaks of HUS do not occur. However, cases of HUS can serve as catalyst to recognize unreported community outbreaks of *E.coli* O157:H7 or other enteric pathogens. If multiple cases of HUS are recognized, contact Division of Infectious Disease Epidemiology immediately for help in reviewing cases of other enteric pathogens for similarities.

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Laboratory Testing

- Diagnosis is based on clinical findings
- Clinical testing to document anemia and renal injury should be done
- Stool culture when possible to identify the causative agent. Attempt stool culture on any HUS case.

Case Definition

Clinical Description of Hemolytic Uremic Syndrome (HUS)

HUS is characterized by the acute onset of microangiopathic hemolytic anemia, renal injury, and low platelet count. Thrombotic thrombocytopenic purpura (TTP) is characterized by these features but can also involve the central nervous system (CNS). Fever may be present in TTP and there may be a more gradual onset. Most cases of HUS (but a few cases of TTP) occur after an acute gastrointestinal illness (usually diarrheal).

Laboratory Criteria for Diagnosis of HUS

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

- a. Anemia (acute onset) with microangiopathic changes (i.e., schistocytes, burr cells, or helmet cells) on peripheral blood smear, and
- b. Renal injury with either hematuria, proteinuria, or elevated creatinine level (>1.0 mg/dl in a child less than 13 years of age and >1.5 mg/dl in anyone older than 13 years of age).

Case Classification of HUS

Probable:

- a. 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 the preceding three weeks, or
 - b. An acute illness diagnosed as HUS or TTP that
 - i. Has onset within three weeks after onset of acute or bloody diarrhea, and
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- ii. Meets the laboratory criteria except that microangiopathic changes are not confirmed.

Confirmed:

An acute illness diagnosed as HUS or TTP that meets the laboratory criteria and began within three weeks after onset of an episode of acute or bloody diarrhea.

Preventive Interventions

1. Wash hands thoroughly and frequently using soap, in particular before eating, and after using the bathroom/changing diapers or contact with farm animals or the farm environment.
2. Pasteurize milk, other dairy products, juices, and ciders.
3. Wash fruits and vegetables, particularly if eaten raw. Peel raw fruits when possible.
4. Avoid consumption of raw sprouts, especially by those most susceptible to severe complications of foodborne infections (young children, the elderly, pregnant women, and persons with compromised immune systems).
5. Cook beef adequately, especially ground beef, to an internal temperature of 70°C (160°F). Do not rely on cooking until all pink color is gone. Use a meat thermometer.
6. Use a separate cutting board to prepare raw meats. Use a clean plate for cooked meat. Never return cooked meat to the same plate used for raw meat.
7. Do not prepare food that will be eaten by others, attend day care, or bathe or swim with others when having diarrhea.
8. Persons with diarrhea due to HUS should not use recreational water venues for 2 weeks after symptoms resolve.
9. Strengthen control measures for exhibits that allow contact with animals or their environment in public settings, such as fairs, farms tours, petting zoos, camps, and schools, and educate populations at risk about the risks associated with attending such events.
10. Ensure adequate hygiene and frequent handwashing with soap in child care centers and in petting zoos and other animal displays.

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Treatment

HUS is a life-threatening condition usually treated in an intensive care unit. Blood transfusions and kidney dialysis are often required. More than 50% of children with HUS require dialysis. With intensive care, the death rate for HUS is 3-5%.

Surveillance Indicators

- Proportion of Cases with complete demographic information.
- Proportion of Cases with complete risk factor information.
- Proportion of cases with complete clinical severity information.
- Proportion of cases with bacterial pathogen identified.

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References

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