

# HEMOLYTIC UREMIC SYNDROME (HUS) FACT SHEET

**Agent:** Usually *E. coli* O157:H7 or other Shiga toxin producing *E. coli* (STEC).

**Brief Description:** Hemolytic uremic syndrome (HUS) is characterized by the acute onset of microangiopathic hemolytic anemia, renal injury, and low platelet count. Thrombotic thrombocytopenic purpura (TTP) is also characterized by these features but also can include central nervous system 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 Diagnosis:

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.

**Diagnostic Testing:** One of the following should be sent to the Georgia Public Health Laboratory (GPHL) in Decatur, for every HUS case:

- An isolate of the recovered *E. coli* O157;
- A stool specimen; or
- Acute and convalescent serum.

All HUS cases should be tested for *E. coli* O157 and,

if negative, other Shiga toxin producing species of *E. coli* (STEC). Collect a stool specimen as soon as possible from patients hospitalized with post-diarrheal HUS. Initially, the local clinical laboratory should test the stool specimen for *E. coli* O157. If positive for *E. coli* O157, forward the *isolate* to the Georgia Public Health Laboratory (GPHL). If negative for *E. coli* O157, forward the *stool specimen* to the GPHL. If no stool can be obtained from the case, send acute and convalescent serum to the GPHL.

## A. Typing and Shiga toxin

1. Specimen: pure culture.
2. Outfit: Culture referral.
3. Lab form: 3410.
4. Lab Test Performed: *E. coli* (enterohemorrhagic) typing, EIA for Shiga toxin, and PFGE.
5. Lab performing test: Bacteriology Laboratory, Georgia Public Health Laboratory (GPHL) in Decatur.

## B. Culture

1. Specimen: Stool
2. Outfit: Stool culture (Para-Pak TM C&S or Cary-Blair)
3. Lab Form: 3416 and Special form for submission of specimens from HUS cases
4. Lab Test Performed: Culture for *E. coli* O157:H7 or other Shiga toxin producing *E. coli* (STEC), Shiga toxin testing, and PFGE.
5. Lab Performing Test: Local clinical laboratory first, then Bacteriology Laboratory, GPHL in Decatur.

## C. Serology

1. Specimen: Acute and convalescent serum
2. Outfit: Sealed Plastic Tube
3. Lab Form: 3432 and Special form for

submission of specimens from HUS cases

4. Lab Test Performed: Testing for antibodies to *E. coli* O157:H7 and other STEC
5. Lab Performing Test: Immunology, Georgia Public Health Laboratory (CDC)

#### Case Classification:

- **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
- **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

**Comment:** Some investigators consider HUS and TTP to be part of a continuum of disease. Therefore, criteria for diagnosing TTP on the basis of central nervous system involvement and fever are not provided because cases that are diagnosed clinically as postdiarrheal TTP should also meet the criteria for HUS. These cases are reported as postdiarrheal HUS. Only HUS or TTP occurring after an acute diarrheal illness is reportable.

**Period of Communicability:** Not applicable

**Treatment:** Treatment for HUS can involve a lengthy hospital stay, including transfusions and dialysis. Details of treatment are beyond the scope of this document.

**Investigation:** Because of the high probability of *E. coli* O157 or other STEC infection, local health authorities need to identify the source and apply appropriate specific preventive measures. It is important to interview the patient quickly so that they will recall exposures accurately to prevent secondary cases.

All cases of HUS should have stool cultures performed to screen for infection with *E. coli* O157:H7 or other STEC. If stool is unavailable, acute and convalescent serum should be tested for the presence of antibodies to *E. coli* O157 and other STEC. In addition to local laboratory testing for *E. coli* O157, at least one of the following should be sent to the Georgia Public Health Laboratory for every case of HUS:

- An isolate of the recovered *E. coli* O157;
- A stool specimen packaged in an appropriate transport kit (See **Diagnostic Testing**); or
- Acute and convalescent serum.

**Reporting:** Report all cases **IMMEDIATELY** to the local health department, District Health Office, or the Epidemiology Branch at 404-657-2588. If calling after regular business hours, it is very important to report cases to the Epidemiology Branch answering service. After a verbal report has been made, please transmit the case information electronically through the State Electronic Notifiable Disease Surveillance System (SENDSS) at <http://sendss.state.ga.us>, or complete and mail a GA Notifiable Disease Report Form (#3095). Complete the special form for submission of specimens from HUS cases and send it with the specimen to the Georgia Public Health Laboratory.

## Reported Cases of Hemolytic Uremic Syndrome in Georgia, 1993-1999

Year	Number of Cases
1993	0
1994	0
1995	0
1996	0
1997	8
1998	14
1999	4

### References:

1. Centers for Disease Control. Community Outbreak of Hemolytic Uremic Syndrome Attributable to *Escherichia coli* O111:NM – South Australia, 1995. MMWR 1995; 44(29): 550-551,557-558.
2. Chin J, ed. Diarrhea Caused by Enterohemorrhagic Strains. In: Control of Communicable Diseases Manual. 17<sup>th</sup> ed. Washington, DC: American Public Health Association, 2000: 155-158.
3. Griffin PM, Tauxe RV. The epidemiology of infections caused by *Escherichia coli* O157:H7, other enterohemorrhagic *E. coli*, and the associated Hemolytic Uremic Syndrome. Epidemiologic Reviews Vol. 13, 1991: 60-98.
4. Mahon BE, Griffin PM, Mead PS, Tauxe RV. Hemolytic Uremic Syndrome surveillance to monitor trends in infection with *Escherichia coli* O157:H7 and other Shiga toxin-producing *E. coli*. Emerging Infectious Diseases, Vol. 3(3) 1997: 409-12.
5. Martin DL, MacDonald KL, White KE, Soler JT, Osterholm MT. The epidemiology and clinical aspects of the Hemolytic Uremic Syndrome in Minnesota. New England Journal of Medicine, Vol. 323(17), 1990: 1161-7.
6. Tarr PI, Neill MA, Clausen CR, Watkins SL, Christie DL, Hickman RO. *Escherichia coli* O157:H7 and the Hemolytic Uremic Syndrome: Importance of early cultures in establishing the etiology. The Journal of Infectious Disease, Vol. 162, 1990: 553-6.
7. Wong CS, Jelacic S, Habeeb RL, Watkins SL, Tarr PI. The risk of Hemolytic-Uremic Syndrome after antibiotic treatment of *Escherichia coli* O157:H7 infections. New England Journal of Medicine, Vol. 342(26), 2000:1930-6.

### Links:

CDC *Escherichia coli* O157:H7 fact sheet – [http://www.cdc.gov/ncidod/dbmd/diseaseinfo/escherichiacoli\\_g.htm](http://www.cdc.gov/ncidod/dbmd/diseaseinfo/escherichiacoli_g.htm)