Wisconsin Department of Health Services
Division of Public Health
P - 01934 (Rev 10/2017)

Communicable Disease Case Reporting and Investigation Protocol

HEMOLYTIC UREMIC SYNDROME

I. IDENTIFICATION AND DEFINITION OF CASES

A. **Clinical 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 can also 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 diarrhea). Therefore, criteria for diagnosing TTP on the basis of central nervous system involvement and fever are not provided because cases diagnosed clinically as post diarrheal TTP should also meet the criteria for HUS. These cases are reported as post diarrheal HUS.

B. **Laboratory Criteria:** Confirmatory laboratory evidence: The following are both present at some time during the illness:

- Anemia (acute onset) with microangiopathic changes (e.g., schistocytes, burr cells, or helmet cells) on peripheral blood smears, **AND**
- Renal injury (acute onset), evidenced by either:
  - Hematuria, **OR**
  - Proteinuria, **OR**
  - Elevated creatinine level (e.g., ≥1.0 mg/dl in a child <13 years of age or ≥1.5 mg/dl in an person ≥13 years of age, or ≥50% increase over baseline).

NOTE: A low platelet count can usually, but not always, be detected early in the illness; however, it may then become normal or even high as the disease progresses. If a platelet count obtained within 7 days after onset of the acute gastrointestinal illness is not <150,000/mm^3, other diagnoses should be considered.

C. **Wisconsin Surveillance Case Definition:**

- **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.
- **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 three weeks, **OR** an acute illness diagnosed as HUS or TTP, that a) has onset within three weeks after onset of an acute or bloody diarrhea and b) meets the laboratory criteria except that microangiopathic changes are not confirmed.

II. REPORTING

A. **Wisconsin Disease Surveillance Category II – Methods for Reporting:** This disease shall be reported to the patient's local health officer or to the local health officer's designee within 72 hours of recognition of a case or suspected case, per Wis. Admin. Code § DHS 145.04 (3) (b). Report electronically through the Wisconsin Electronic Disease Surveillance System (WEDSS), or mail or fax a completed Acute and Communicable Disease Case Report (F-44151) to the address on the form.

B. **Responsibility for Reporting**: According to Wis. Admin. Code § DHS 145.04(1), persons licensed under Wis. Stat. ch. 441 or 448, laboratories, health care facilities, teachers, principals, or nurses serving a school or day care center, and any person who knows or suspects that a person has a communicable disease identified in Appendix A.

C. **Clinical Criteria for Reporting:** A case of post-diarrheal HUS. Most diarrhea-associated HUS is caused by Shiga toxin-producing *Escherichia coli*, most commonly *E. coli* O157. If a patient meets the case definition for HUS and Shiga toxin-producing *E. coli* (STEC) or another reportable condition, a case should be reported for each of the reportable conditions.

D. **Laboratory Criteria for Reporting:** A case that meets the laboratory criteria for diagnosis.

III. CASE INVESTIGATION

A. **Responsibility for case investigation**: It is the responsibility of the local health department (LHD) to investigate or arrange for investigation of suspected or confirmed cases as soon as is reasonably possible. A case investigation may include information collected by phone, in person, in writing, or through review of medical records or communicable disease report forms, as necessary and appropriate.

B. **Required Documentation:**
   1. Complete the WEDSS disease incident investigation report, including appropriate, disease-specific tabs. This may be facilitated by completing a Routine Enteric Follow-Up Worksheet. See page 1 of the Worksheet for specific instructions regarding which sections should be completed during routine follow-up.
   2. Request the necessary laboratory results and documentation necessary to evaluate if the case meets the laboratory evidence for confirmation.
   3. Upon completion of investigation, set WEDSS disease incident process status to "Sent to State."

C. **Additional Investigation Responsibilities**
   1. Assess patient for high-risk settings or activities to include food handling, providing patient care or child care, or attending a child care facility.
   2. Source investigation by LHD.
   3. If the case is potentially outbreak-related, notify the Wisconsin Division of Public Health (DPH), Bureau of Communicable Diseases (BCD).

IV. PUBLIC HEALTH INTERVENTIONS AND PREVENTION MEASURES

A. In accordance with Wis. Admin. Code § DHS 145.05, local public health agencies should follow the methods of control recommended in the current editions of *Control of Communicable Diseases Manual*, edited by David L. Heymann, published by the American Public Health Association, and the American Academy of Pediatrics' *Red Book: Report of the Committee on Infectious Diseases*, unless otherwise specified by the state epidemiologist.

B. Educate the public about proper handwashing after using the toilet, changing diapers, assisting another with toileting, handling contaminated clothing or linens, before cooking, or when associating with high-risk individuals.

C. Exclude patients from high-risk settings including food handling, providing patient care or child care, or attending a child care facility or 4K program. Individuals should not return to high-risk settings following exclusion until they have been cleared by their LHD. Return to high-risk activities for people diagnosed with an STEC infection routinely requires evidence be provided to the LHD of two consecutive stool specimens negative for Shiga toxin-producing *E. coli* by culture or CIDT. Patients with post-diarrheal HUS without confirmation of STEC infections should be evaluated and tests of cure requirement may be indicated if they had an illness that was clinically compatible with STEC infection. If laboratory evidence of clearance is required, specimens should be collected a) after the individual is asymptomatic and b) at least 48 hours after discontinuance of antimicrobial therapy. Specimens should be collected at least 24 hours apart.

V. CONTACTS FOR CONSULTATION

A. Local health departments and tribal health agencies:
https://www.dhs.wisconsin.gov/lh-depts/index.htm

B. Bureau of Communicable Diseases, Communicable Diseases Epidemiology Section: 608-267-9003

C. Wisconsin State Laboratory of Hygiene: 1-800-862-1013

VI. RELATED REFERENCES

A. Heymann DL, ed. *E. coli* Diarrheal Illnesses. In: *Control of Communicable Diseases Manual*. 20th ed. Washington, DC: American Public Health Association, 2015: 158.

B. Pickering LK, ed. *Escherichia coli* Diarrhea including Hemolytic-Uremic Syndrome. In: *Red Book*: *2015 Report of the Committee on Infectious Diseases*. 30th ed. Elk Grove Village, IL: American Academy of Pediatrics, 2015: 343-347.

C. Centers for Disease Control and Prevention website: https://www.cdc.gov/ecoli/general/index.html