09-ID-37

Committee: Infectious

Title: **Public Health Reporting and National Notification for Hemolytic Uremic Syndrome (post-diarrheal)**

I. Statement of the Problem

CSTE position statement 07-EC-02 recognized the need to develop an official list of nationally notifiable conditions and a standardized reporting definition for each condition on the official list. The position statement also specified that each definition had to comply with American Health Information Community recommended standards to support "automated case reporting from electronic health records or other clinical care information systems." In July 2008, CSTE identified sixty-eight conditions warranting inclusion on the official list, each of which now requires a standardized reporting definition.

II. Background and Justification

Background[1]

Hemolytic uremic syndrome (HUS) is characterized by hemolytic anemia, kidney impairment, and low platelet count. Ninety percent of cases follow an acute diarrheal illness. Most cases of post-diarrheal HUS in North America are caused by *E. coli* O157:H7 and other Shiga toxin-producing *E. coli*, often as a result of foodborne outbreaks. About five to ten percent of persons with bloody diarrhea from *E. coli* O157:H7 develop HUS. Persons of any age can develop HUS, but the incidence is highest in children and the elderly. An estimated 1000 or more U.S. children develop HUS each year. HUS is a severe illness; virtually all patients are hospitalized, and about half require dialysis. Among children, three to five percent die during the acute illness, and of the remainder, five percent have severe high blood pressure, four percent have stroke, and four percent have severe kidney damage. The only treatment for HUS is supportive care; no specific treatment exists. Surveillance for HUS is necessary to identify and control outbreaks and to monitor the effectiveness of strategies to prevent infections with Shiga toxin-producing E. coli.

Justification

Hemolytic Uremic Syndrome (post-diarrheal) meets the following criteria for a nationally and **standard** notifiable condition, as specified in CSTE position statement 08-EC-02:

- A majority of state and territorial jurisdictions—or jurisdictions comprising a majority of the US population—have laws or regulations requiring **standard** reporting of Hemolytic Uremic Syndrome (post-diarrheal) to public health authorities
- CDC requests **standard** notification of hemolytic uremic syndrome (post-diarrheal) to federal authorities

[1] Much of the material in the background is directly quoted from the CDC's hemolytic uremic syndrome website. See the References for further information on this source.

- CDC has condition-specific policies and practices concerning the agency's response to, and use of, notifications.

III. Statement of the desired action(s) to be taken

CSTE requests that CDC adopt this standardized reporting definition for hemolytic uremic syndrome (post-diarrheal) to facilitate more timely, complete, and standardized local and national reporting of this condition.

IV. Goals of Surveillance

To provide information on the temporal, geographic, and demographic occurrence of hemolytic uremic syndrome (post-diarrheal) to facilitate its prevention and control.

V. Methods for Surveillance

Surveillance for hemolytic uremic syndrome (post-diarrheal) should use the sources of data and the extent of coverage listed in Table V below.

Table V. Recommended sources of data and extent of coverage for ascertaining cases of hemolytic uremic syndrome (post-diarrheal).

	Coverage	
Source of data for case ascertainment	Population-wide	Sentinel sites
clinician reporting	X	X
laboratory reporting	X	
reporting by other entities (e.g., hospitals, veterinarians, pharmacies)	X	
death certificates	X	
hospital discharge or outpatient records	X	
extracts from electronic medical records	X	
telephone survey		
school-based survey		
other _____		

VI. Criteria for Reporting

Reporting refers to the process of healthcare providers or institutions (e.g., clinicians, clinical laboratories, hospitals) submitting basic information to governmental public health agencies about cases of illness that meet certain reporting requirements or criteria. Cases of illness may also be ascertained by the secondary analysis of administrative health data or clinical data. The purpose of this section is to provide those criteria to determine whether a specific illness should be reported.

A. Narrative description of criteria to determine whether a case should be reported to public health authorities

Report any illness to public health authorities that meets any of the following criteria:

1. Any person diagnosed as having hemolytic uremic syndrome

2. Any person diagnosed as having thrombotic thrombocytopenic purpura

3. Any person with hemolytic anemia and renal injury as evidenced by hematuria, proteinuria or an elevated creatinine level.

4. A person whose healthcare record contains a diagnosis of hemolytic uremic syndrome.

5. A person whose death certificate lists hemolytic uremic syndrome as a cause of death or a significant condition contributing to death.

Other recommended reporting procedures

- All cases of hemolytic uremic syndrome (post-diarrheal) should be reported.
- Reporting should be on-going and routine.
- Frequency of reporting should follow the state health department's routine schedule.

B. Table of criteria to determine whether a case should be reported to public health authorities

Table VI-B. Table of criteria to determine whether a case should be reported to public health authorities. Requirements for reporting are established under State and Territorial laws and/or regulations and may differ from jurisdiction to jurisdiction. These criteria are suggested as a standard approach to identifying cases of this condition for purposes of reporting, but reporting should follow State and Territorial law/regulation if any conflicts occur between these criteria and those laws/regulations.

Criterion	**Reporting**
Clinical Evidence	
Diarrhea	
Bloody diarrhea	
Onset of illness < 3 weeks after onset of diarrhea	
Diagnosis of Hemolytic Uremic Syndrome	S
Diagnosis of thrombotic thromobocytopenic purpura	S
Death certificate lists Hemolytic Uremic Syndrome as a cause of death or a significant condition contributing to death	S

Laboratory Evidence	
Anemia	N
Microangiopathic changes on peripheral blood smear (burr cells, helmet cells, schistocytes)	N
Hematuria	O
Proteinuria	O
Increased creatinine level	O

Notes:
S = This criterion alone is Sufficient to identify a case for reporting.
N = All "N" criteria in the same column are Necessary to identify a case for reporting.
O = At least one of these "O" (Optional) criteria in each category (i.e., clinical evidence and laboratory evidence) in the same column—in conjunction with all "N" criteria in the same column—is required to identify a case for reporting. (These optional criteria are alternatives, which means that a single column will have either no O criteria or multiple O criteria; no column should have only one O.)

C. Disease Specific Data Elements:

Disease-specific data elements to be included in the initial report are listed below.

Epidemiological Risk Factors

Food history for the 10 days prior to onset of illness
- Consumption of raw, rare or undercooked ground beef
- Consumption of lettuce
- Consumption of uncooked spinach

Contact with recreational water in the 10 days prior to onset of illness
Contact with farm animals in the 10 days prior to onset of illness

VII. Case Definition

A. Narrative description of criteria to determine whether a case should be classified as confirmed or probable (presumptive):

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) 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 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/mm3, other diagnoses should be considered.

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 CNS involvement and fever are not provided because cases diagnosed clinically as postdiarrheal TTP also should meet the criteria for HUS. These cases are reported as postdiarrheal 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 both Shiga toxin-producing *E. coli* (STEC) and HUS, the case should be reported for each of the conditions.

B. Classification Tables:

Table VII-B lists the criteria that must be met for a case to be classified as confirmed or probable (presumptive).

Table VII-B. Table of criteria to determine whether a case is classified.

	Case Definition		
Criterion	**Confirmed**	**Probable**	
Clinical Evidence			
Diarrhea	N		N
Onset of illness < 3 weeks after onset of diarrhea	N	A	N
Diagnosis of Hemolytic Uremic Syndrome		O	O
Diagnosis of thrombotic thromobocytopenic purpura		O	O

Laboratory Evidence			
Anemia	N	N	N
Microangiopathic changes on peripheral blood smear (burr cells, helmet cells, schistocytes)	N	N	
Hematuria	O	O	O
Proteinuria	O	O	O
Increased creatinine level	O	O	O

Notes:
N = All "N" criteria in the same column are Necessary to classify a case.
A = This criterion must be absent (i.e., NOT present) for the case to meet the classification criteria.
O = At least one of these "O" (Optional) criteria in each category (i.e., clinical evidence and laboratory evidence) in the same column—in conjunction with all "N" criteria in the same column—is required to classify a case.

VIII. Period of Surveillance

Surveillance should be on-going.

IX. Data sharing/release and print criteria

Notification to CDC of confirmed and probable cases of Hemolytic Uremic Syndrome is recommended.

- Data will be used to determine the burden of illness due to post-diarrheal HUS, to assess the effectiveness over time of national control program, and to assess progress towards national goals for Shiga toxin-producing E. coli. Data may also be useful to compare case numbers with information from other foodborne disease surveillance systems. Electronic reports of HUS cases in the US are summarized weekly in the MMWR tables and annual case data is summarized in the yearly Summary of Notifiable Diseases.
- State-specific compiled data will continue to be published in the weekly and annual MMWR. In addition to those reports, the frequency of reports/feedback to the states and territories will be dependent on the current epidemiologic situation in the country. Frequency of cases, epidemiologic distribution, and other factors will influence communications.
- State-specific compiled data will continue to be published in the weekly reports and annual MMWR surveillance summaries. All cases are verified with the states before publication.

X. References

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