09-ID-60

Committee: Infectious

Title: Public Health Reporting and National Notification for streptococcal toxic shock syndrome (STSS)

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 definition for reporting 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]

Group A *Streptococcus* is a bacterium often found in the throat and on the skin. People may carry group A streptococci in the throat or on the skin and have no symptoms of illness. Most GAS infections are relatively mild illnesses such as "strep throat," or impetigo, of which there are several million cases each year. Occasionally these bacteria can cause severe and even life-threatening diseases.

Severe, sometimes life-threatening, GAS disease may occur when bacteria get into parts of the body where bacteria usually are not found, such as the blood, muscle, or the lungs. About 9,000-11,500 cases of invasive GAS disease occur each year in the United States, resulting in 1,000-1,800 deaths annually. These infections are termed "invasive GAS disease." Two of the most severe, but least common, forms of invasive GAS disease are necrotizing fasciitis and streptococcal toxic shock syndrome. STSS and necrotizing fasciitis each comprise an average of about 6%-7% of these invasive cases. Necrotizing fasciitis (occasionally described by the media as "the flesh-eating bacteria") is a rapidly progressive disease, which destroys muscles, fat, and skin tissue. Streptococcal toxic shock syndrome (STSS) results in a rapid drop in blood pressure and organ (e.g., kidney, liver, lungs) failure. STSS is not the same as the "toxic shock syndrome" due to the bacteria *Staphylococcus aureus,* which has been associated with tampon usage. While 10%-15% of patients with invasive GAS disease die from their infection, approximately 25% of patients with necrotizing fasciitis and more than 35% with STSS die.

[1] Much of the material in the background is directly quoted from the CDC's group A streptococcal disease Website. See the References for further information on this source.

Justification

Streptococcal toxic shock syndrome 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 streptococcal toxic shock syndrome to public health authorities
- CDC requests **standard** notification of streptococcal toxic shock syndrome to federal authorities
- 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 streptococcal toxic shock syndrome 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 streptococcal toxic shock syndrome to facilitate its prevention and control.

V. Methods for Surveillance

Surveillance for streptococcal toxic shock syndrome should use the sources of data and the extent of coverage listed in Table V.

Table V. Recommended sources of data and extent of coverage for ascertaining cases of streptococcal toxic shock syndrome.

	Coverage	
Source of data for case ascertainment	Population-wide	Sentinel sites
clinician reporting	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 person from whom group A *Streptococcus* is isolated by culture from any site *and* who is hypotensive *and* has multi-organ involvement characterized by two or more of the following:

- Renal impairment (creatinine ≥ 2 mg/dL [≥ 177 µmol/L] for adults, or greater than or equal to twice the upper limit of normal for age. In patients with preexisting renal disease, a greater than twofold elevation over the baseline level)

- Coagulopathy (platelets ≤ 100,000/mm3 or disseminated intravascular coagulation, defined by prolonged clotting times, low fibrinogen level, and the presence of fibrin degradation products)

- Liver involvement (alanine aminotransferase, aspartate aminotransferase, or total bilirubin levels greater than or equal to twice the upper limit of normal for the patient's age. In patients with preexisting liver disease, a greater than twofold increase over the baseline level)

- Acute respiratory distress syndrome (defined by acute onset of diffuse pulmonary infiltrates and hypoxemia in the absence of cardiac failure; or by evidence of diffuse capillary leak manifested by acute onset of generalized edema; or pleural or peritoneal effusions with hypoalbuminemia)

- Generalized erythematous macular rash that may desquamate

- Soft-tissue necrosis, including necrotizing fasciitis or myositis, or gangrene

Report any person whose healthcare record contains a diagnosis of streptococcal toxic shock syndrome.

Report any person whose death certificate lists streptococcal toxic shock syndrome as a cause of death or a significant condition contributing to death.

Other recommended procedures

- all cases of streptococcal toxic shock syndrome 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		
Hypotension (systolic blood pressure ≤ 90 mm Hg for adults or less than the fifth percentile by age for children aged less than 16 years)		N
Renal impairment (creatinine ≥ 2 mg/dL [≥ 177 μmol/L] for adults, or greater than or equal to twice the upper limit of normal for age. In patients with preexisting renal disease, a greater than twofold elevation over the baseline level)		O^
Coagulopathy (platelets ≤ 100,000/mm^3 or disseminated intravascular coagulation, defined by prolonged clotting times, low fibrinogen level, and the presence of fibrin degradation products)		O^
Liver involvement (alanine aminotransferase, aspartate aminotransferase, or total bilirubin levels greater than or equal to twice the upper limit of normal for the patient's age. In patients with preexisting liver disease, a greater than twofold increase over the baseline level)		O^
Acute respiratory distress syndrome (defined by acute onset of diffuse pulmonary infiltrates and hypoxemia in the absence of cardiac failure; or by evidence of diffuse capillary leak manifested by acute onset of generalized edema; or pleural or peritoneal effusions with hypoalbuminemia)		O^
Generalized erythematous macular rash that may desquamate		O^
Soft-tissue necrosis, including necrotizing fasciitis or myositis, or gangrene		O^
Healthcare record contains a diagnosis of streptococcal toxic shock syndrome	S	

Death certificate lists streptococcal toxic shock syndrome as a cause of death or a significant condition contributing to death	S	
Laboratory Evidence		
Isolation of group A *Streptococcus* by culture from a normally *non*-sterile site		O
Isolation of group A *Streptococcus* by culture from a normally sterile site (e.g., blood or cerebrospinal fluid, or, less commonly, joint, pleural, or pericardial fluid)		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.)
O^ = For Streptococcal STSS, at least two of the clinical "O" criteria are required to report a case.

C. Disease Specific Data Elements:

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

VII. Case Definition

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

Clinical description

Streptococcal toxic-shock syndrome (STSS) is a severe illness associated with invasive or noninvasive group A streptococcal (*Streptococcus pyogenes*) infection. STSS may occur with infection at any site but most often occurs in association with infection of a cutaneous lesion. Signs of toxicity and a rapidly progressive clinical course are characteristic, and the case-fatality rate may exceed 50%.

Clinical case definition

An illness with the following clinical manifestations:

- Hypotension defined by a systolic blood pressure less than or equal to 90 mm Hg for adults or less than the fifth percentile by age for children aged less than 16 years.
- Multi-organ involvement characterized by two or more of the following:
  1. Renal impairment: Creatinine greater than or equal to 2 mg/dL (greater than or equal to 177 µmol/L) for adults or greater than or equal to twice the upper limit of

normal for age. In patients with preexisting renal disease, a greater than twofold elevation over the baseline level.

2. Coagulopathy: Platelets less than or equal to 100,000/mm^3 (less than or equal to 100 x 10^6/L) or disseminated intravascular coagulation, defined by prolonged clotting times, low fibrinogen level, and the presence of fibrin degradation products.
3. Liver involvement: Alanine aminotransferase, aspartate aminotransferase, or total bilirubin levels greater than or equal to twice the upper limit of normal for the patient's age. In patients with preexisting liver disease, a greater than twofold increase over the baseline level.
4. Acute respiratory distress syndrome: defined by acute onset of diffuse pulmonary infiltrates and hypoxemia in the absence of cardiac failure or by evidence of diffuse capillary leak manifested by acute onset of generalized edema, or pleural or peritoneal effusions with hypoalbuminemia.
5. A generalized erythematous macular rash that may desquamate.
6. Soft-tissue necrosis, including necrotizing fasciitis or myositis, or gangrene.

Laboratory criteria for diagnosis

Isolation of group A *Streptococcus*.

Case classification

Probable: a case that meets the clinical case definition in the absence of another identified etiology for the illness and with isolation of group A *Streptococcus* from a nonsterile site.

Confirmed: a case that meets the clinical case definition and with isolation of group A *Streptococcus* from a normally sterile site (e.g., blood or cerebrospinal fluid or, less commonly, joint, pleural, or pericardial fluid).

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.

Criterion	Confirmed	Probable
Clinical Evidence		
Hypotension (systolic blood pressure ≤ 90 mm Hg for adults or less than the fifth percentile by age for children aged less than 16 years)	N	N
Renal impairment (creatinine ≥ 2 mg/dL [≥ 177 µmol/L] for adults, or greater than or equal to twice the upper limit of normal for age. In patients with preexisting renal disease, a greater than twofold elevation over the baseline level)	O^	O^
Coagulopathy (platelets ≤ 100,000/mm^3 or disseminated intravascular	O^	O^

coagulation, defined by prolonged clotting times, low fibrinogen level, and the presence of fibrin degradation products)		
Liver involvement (alanine aminotransferase, aspartate aminotransferase, or total bilirubin levels greater than or equal to twice the upper limit of normal for the patient's age. In patients with preexisting liver disease, a greater than twofold increase over the baseline level)	O^	O^
Acute respiratory distress syndrome (defined by acute onset of diffuse pulmonary infiltrates and hypoxemia in the absence of cardiac failure; or by evidence of diffuse capillary leak manifested by acute onset of generalized edema; or pleural or peritoneal effusions with hypoalbuminemia)	O^	O^
Generalized erythematous macular rash that may desquamate	O^	O^
Soft-tissue necrosis, including necrotizing fasciitis or myositis, or gangrene	O^	O^
Absence of another cause for the illness		N
Laboratory Evidence		
Isolation of group A *Streptococcus* by culture from a normally *non*-sterile site		N
Isolation of group A *Streptococcus* by culture from a normally sterile site (e.g., blood or cerebrospinal fluid, or, less commonly, joint, pleural, or pericardial fluid)	N	

Notes:
N = All "N" criteria in the same column are Necessary to classify a case.
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.
O^ = For Streptococcal STSS, at least two of the clinical "O" criteria are required to classify a case as confirmed or probable.

VIII. Period of Surveillance

Surveillance should be on-going.

IX. Data sharing/release and print criteria

Notification to CDC for confirmed and probable cases of STSS is recommended.

- Electronic reports of NNDSS STSS cases are summarized weekly in MMWR Tables 1 and 2. Annual case data on STSS is also summarized in the yearly MMWR Summary of Notifiable Disease.
- A second surveillance system for invasive group A streptococcal (GAS) diseases, including STSS, is maintained by the Respiratory Disease Branch (RDB). Via a case

report form, this system collects extensive information on invasive GAS cases such as underlying diseases and other risk factors, presenting clinical syndromes, and disease outcome. We hope that STSS reporting by states can be improved prior to GAS vaccine licensure. Comparing the rates of disease from national reporting and RDB's sentinel surveillance will be undertaken in the near future.

- State-specific complied data will continue to be published in the weekly and annual MMWR reports.
- State-specific compiled data will continue to be published in weekly MMWR reports and the annual MMWR Surveillance Summaries. All cases are verified with the state(s) before publication.
- The re-release of this data is provided on an as-needed basis following the NNDSS data release guidelines. Personal identifying information is not transmitted to any party.

X. References

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