

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

Background¹

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.

Source of data for case ascertainment	Coverage	
	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 that should be used by humans and machines to determine whether a specific illness should be reported.²

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:

- elevated creatinine
- coagulopathy
- elevated aminotransferase or total bilirubin
- acute respiratory distress syndrome
- generalized erythematous macular rash
- soft-tissue necrosis

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

² "Human-based" criteria (described below under "A. Narrative") can be applied by medical care providers and laboratory staff based on clinical judgment and clinical diagnosis. Machine-based criteria (described below under "B. Table") can be applied using computerized algorithms that operate in electronic health record systems, including computerized records of laboratory test orders and laboratory test results; other clinical data systems (e.g., hospital discharge data systems serving multiple hospitals); or administrative data (e.g., healthcare provider billing data, vital records, and EMS data).

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

Table VI-B. Proposed Table of criteria to determine whether a case should be reported to public health authorities. Note: The following criteria are proposed for evaluation before general implementation. For purposes of currently implementing reporting the narrative description in VI-A, should be used.

Criterion	Reporting	
<i>Clinical Presentation</i>		
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
multisystem involvement, signified by two or more of the following findings: - 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 $\leq 100,000/\text{mm}^3$ 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		N
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	
absence of another cause for the illness		
<i>Laboratory findings</i>		
isolation of group A <i>Streptococcus</i> by culture from a normally <i>non</i> -sterile site		O
isolation of group A <i>Streptococcus</i> 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 report a case.

N = All “N” criteria in the same column—in conjunction with at least one of any “O” criteria in each category (e.g., clinical presentation and laboratory findings) in the same column—are required to report a case.

O = At least one of any “O” criteria in each category (e.g., clinical presentation and laboratory findings) in the same column—in conjunction with all other “N” criteria in the same column—is 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 μ mol/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/mm³ (less than or equal to 100 x 10⁶/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. Proposed table of criteria to determine whether a case is classified. Note: The following criteria are proposed for evaluation before general implementation. For purposes of current notification, the narrative description in VII-A, should be used.

Criterion	Case Definition	
	Confirmed	Probable
<i>Clinical Presentation</i>		
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
multisystem involvement, signified by two or more of the following findings: - 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 $\leq 100,000/\text{mm}^3$ 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)	N	N

- generalized erythematous macular rash that may desquamate - soft-tissue necrosis, including necrotizing fasciitis or myositis, or gangrene		
healthcare record contains a diagnosis of streptococcal toxic shock syndrome		
death certificate lists streptococcal toxic shock syndrome as a cause of death or a significant condition contributing to death		
absence of another cause for the illness		N
<i>Laboratory findings</i>		
isolation of group A <i>Streptococcus</i> by culture from a normally <i>non</i> -sterile site		N
isolation of group A <i>Streptococcus</i> 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—in conjunction with at least one of any “O” criteria in each category (e.g., clinical presentation and laboratory findings) in the same column—are required to confirm a case.

VIII. Period of Surveillance

Surveillance should be on-going.

IX. Data sharing/release and print criteria

Notification to CDC for confirmed 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 compiled 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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XI. Coordination:

Agencies for Response:

- (1) Thomas R Frieden, MD, MPH
Director
Centers for Disease Control and Prevention
1600 Clifton Road, NE
Atlanta GA 30333
(404) 639-7000
txf2@cdc.gov

XII. Submitting Author:

- (1) David Kirschke, MD, Deputy State Epidemiologist
Tennessee Department of Health
425 5th Ave North
1st Floor, Cordell Hull Bldg
Nashville, TN 37243
Phone: (615) 532-8491
david.kirschke@state.tn.us

Co-Authors:

- (1) Associate Member
Harry F. Hull, Medical Epidemiologist
HF Hull & Associates, LLC
1140 St. Dennis Court
Saint Paul, MN 55116
(651) 695-8114
hullhf@msn.com
- (2) Associate Member
Cecil Lynch, Medical Informaticist
OntoReason
7292 Shady Woods Circle
Midvale, UT 84047
(916) 412.5504
clynch@ontoreason.com
- (3) Associate Member
R. Gibson Parrish, Medical Epidemiologist
P.O. Box 197
480 Bayley Hazen Road
Peacham, VT 05862
(802) 592-3357
gib.parrish@gmail.com