09-ID-49

Committee: Infectious

Title: National Surveillance for Smallpox

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
Smallpox is a serious, contagious, and sometimes fatal infectious disease caused by infection with variola virus. There are two clinical forms of smallpox, variola major and variola minor. Variola major is the severe and most common form of smallpox, with a more extensive rash and higher fever. There are four types of variola major smallpox: (1) ordinary (the most frequent type, accounting for 90% or more of cases); (2) modified (mild and occurring in previously vaccinated persons); (3) flat; and (4) hemorrhagic (both rare and very severe). Historically, variola major has an overall fatality rate of about 30%; however, flat and hemorrhagic smallpox usually are fatal. Variola minor is a less common and a much less severe form of smallpox, with death rates historically of 1% or less.

Generally, direct and fairly prolonged face-to-face contact is required to spread smallpox from one person to another. A person with smallpox is considered contagious with onset of fever (prodrome phase), but the person becomes most contagious with the onset of rash. At this stage the infected person is usually very sick and not able to comfortably move around in the community. The infected person is contagious until the last smallpox scab falls off. Smallpox also can be spread through direct contact with infected bodily fluids or contaminated objects such as bedding or clothing. Rarely, smallpox has been spread by virus carried in the air in enclosed settings such as buildings, buses, and trains. Humans are the only natural hosts of variola. Insects and animals do not transmit smallpox. There is no specific treatment for smallpox, and the only prevention is vaccination.

Smallpox outbreaks have occurred from time to time for thousands of years, but the disease is now eradicated after a successful worldwide vaccination program. The last case of smallpox in the United States was in 1949. The last naturally occurring case in the world was in Somalia in

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1977. After the disease was eliminated from the world, routine vaccination against smallpox among the general public was stopped because it was no longer necessary for prevention.

In the absence of smallpox disease in the world, the suggested approach to surveillance relies on a highly specific clinical case definition, which is focused on identifying the classic case presentation (ordinary type) of smallpox. Before eradication, classic (ordinary type) smallpox generally accounted for approximately 90% of smallpox cases in previously unvaccinated individuals and 70% of cases that occurred in previously vaccinated individuals who were no longer fully protected by vaccination.

Justification
Smallpox meets the definition of a nationally and **immediately** notifiable condition—as specified in CSTE position statement 08-EC-02—for the following reason(s):

- The condition has special importance for international health regulations (IHR).
- The condition is included on the list of Category A possible bioterrorism agents and toxins.
- The condition has been declared eliminated (absence of endemic disease transmission) in the United States or eradicated globally.
- A majority of state and territorial jurisdictions—or jurisdictions comprising a majority of the US population—have laws or regulations requiring **immediate** reporting of smallpox to public health authorities; the Centers for Disease Control and Prevention (CDC) requests **immediate** notification of smallpox; and the CDC has condition-specific policies and practices concerning its 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 smallpox 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 smallpox to facilitate its prevention and control.

V. Methods for Surveillance

Surveillance for smallpox should use the sources of data and the extent of coverage listed in table V.

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Table V. Recommended sources of data and extent of coverage for ascertaining cases of smallpox

	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

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. A person for whom a diagnostic test specific for smallpox has been ordered).
2. A person with generalized vesicular rash illness AND a fever ≥101ºF (38.3ºC) that
3. occurred 1–4 days before rash onset AND at least one of the following: prostration, headache, backache, chills, vomiting, *or* severe abdominal pain.
4. A person with generalized pustular rash illness AND a fever ≥101ºF (38.3ºC) that
5. occurred 1–4 days before rash onset AND at least one of the following: prostration, headache, backache, chills, vomiting, *or* severe abdominal pain.
6. A person with contact (≤ 2 meters) for (≥ 3 hours) with a laboratory confirmed case of smallpox AND a fever ≥101ºF (38.3ºC) AND at least one of the following: prostration, headache, backache, chills, vomiting, *or* severe abdominal pain.
7. A person whose death certificate lists smallpox as a cause of death or a significant condition contributing to death.

Other recommended reporting procedures

- All cases of smallpox should be reported.
- Reporting should be on-going and routine.
- Reporting should be immediate.

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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			
fever ≥101ºF (38.3ºC)	N	N	N
fever occurs 1–4 days before rash onset		N	N
prostration	O	O	O
headache	O	O	O
backache	O	O	O
chills	O	O	O
vomiting	O	O	O
severe abdominal pain	O	O	O
generalized vesicular rash		N	
generalized pustular rash			N
identifiable cause of clinical findings		A	A
death certificate lists smallpox as a cause of death or a significant condition contributing to death.	S		
Laboratory Evidence			
polymerase chain reaction (PCR) identification of variola DNA in a clinical specimen	S*		
isolation of smallpox (variola) virus from a clinical specimen	S*		
polymerase chain reaction (PCR) identification of variola DNA in an isolate from a clinical specimen	S*		
Epidemiologic Evidence			
contact (≤ 2 meters) for (≥ 3 hours) with a laboratory confirmed case of smallpox	N		

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.
A = This criterion must be absent (i.e., NOT present) for the case to meet the reporting criteria.

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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.
* A requisition for any of the "S" or "N" laboratory tests is sufficient to meet the reporting criteria

C. Disease Specific Data Elements:

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

Smallpox vaccination ever (yes/no)
Date of smallpox vaccination
Date of fever onset
Date of rash onset
Clinical type of smallpox
Known exposure to suspected/confirmed or probable case (yes/no)
Name of first hospital or isolation center where case was admitted
Outcome (complications death)
Lab confirmation
Case status (confirmed probable or suspected)

Source: Centers for Disease Control and Prevention. Draft Guide A: Smallpox Surveillance and Case Reporting; Contact Identification, Tracing, Vaccination, and Surveillance; and Epidemiologic Investigation. Atlanta: CDC; 2003 Dec 31, page 23. Available from: http://emergency.cdc.gov/agent/smallpox/response-plan/. Accessed: 2008 Aug 5.

VII. Case Definition

A. Narrative description of criteria to determine whether a case should be classified as confirmed, probable (presumptive), or suspected (possible) is provided below.

Clinical Case Definition

An illness with acute onset of fever ≥101º F (≥38.3 º C) followed by a rash characterized by firm, deep seated vesicles or pustules in the same stage of development without other apparent cause.

Clinically consistent cases are those presentations of smallpox that do not meet the classical clinical case definition: (a) hemorrhagic type, (b) flat type, and (c) variola sine eruptione. (Detailed clinical description is available on the CDC web site, see URL: http://www.bt.cdc.gov/agent/smallpox/index.asp).

Laboratory Criteria

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Polymerase chain reaction (PCR) identification of variola DNA in a clinical specimen, *or*

Isolation of smallpox (variola) virus from a clinical specimen (Level D laboratory only; confirmed by variola PCR).

Note: Indications for laboratory testing of patients with suspected smallpox should be followed as described in detail in Guide A of the CDC Smallpox Response Plan. Laboratory diagnostic testing for variola virus should be conducted in Level C or D laboratories only.

*Case Classification**

Confirmed: A case of smallpox that is laboratory confirmed, or a case that meets the clinical case definition that is epidemiologically linked to a laboratory confirmed case.

Probable: A case that meets the clinical case definition, or a case that does not meet the clinical case definition but is clinically consistent and has an epidemiological link to a confirmed case of smallpox.

Suspected: A case with a generalized, acute vesicular or pustular rash illness with fever preceding development of rash by 1-4 days.

*Exclusion Criteria: A case may be excluded as a suspect or probable smallpox case if an alternative diagnosis fully explains the illness or appropriate clinical specimens are negative for laboratory criteria for smallpox.

Note: The smallpox case definition is to be used only during post-event surveillance. The case definition described in Guide A of the Smallpox Response Plan and Guidelines (Version 3) on the CDC bioterrorism preparedness website (URL: http://www.bt.cdc.gov/agent/smallpox/response-plan/index.asp) includes different criteria for a suspected case than the smallpox case definition the Council of State and Territorial Epidemiologists approved for use in the National Notifiable Diseases Surveillance System (NNDSS). The smallpox case definition on the CDC bioterrorism web site is more sensitive and less specific than the case definition for the NNDSS, in that a "suspect" case is defined as: “a case with febrile rash illness with fever preceding the development of rash by 1-4 days.”

B. Classification Tables

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

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

	Case Definition						
Criterion	Confirmed			Probable		Suspected	
Clinical Evidence							
Fever ≥101ºF (38.3ºC)	N	N	N	N	N	N	N
Fever occurs 1–4 days before rash onset	N	N	N	N	N	N	N
Prostration	O	O	O	O	O	O	O
Headache	O	O	O	O	O	O	O
Backache	O	O	O	O	O	O	O
Chills	O	O	O	O	O	O	O
Vomiting	O	O	O	O	O	O	O
Severe abdominal pain	O	O	O	O	O	O	O
Rash with deep-seated, firm or hard, round, well-circumscribed vesicles or pustules; as they evolve, lesions may become umbilicated or confluent	N	N	N	N			
Rash with *flat*, *soft*, *focal* lesions; as they evolve, lesions may become confluent and portions of skin may slough					N1		
Lesions on any ONE part of the body (e.g., the face, arms) are all in the same stage of development (i.e., all are vesicles, or all are pustules)	N	N	N	N			
Widespread hemorrhage in skin and mucous membranes					N2		
Generalized vesicular rash						N	
Generalized pustular rash							N
Identifiable cause of clinical findings			A	A	A	A	A
Laboratory Evidence							
Polymerase chain reaction (PCR) identification of variola DNA in a clinical specimen	N			A		A	A
Isolation of smallpox (variola) virus from a clinical specimen		N		A		A	A
Polymerase chain reaction (PCR) identification of variola DNA in an isolate from a clinical specimen		N		A		A	A
Detection of orthopox-reactive IgM antibodies, 3-56 days post symptom onset					N3		
Epidemiologic Evidence							
Contact (≤ 2 meters) for (≥ 3 hours) with a laboratory confirmed case of smallpox			N		N	N	
Contact (≤ 2 meters) with a confirmed, probable, or suspected case of smallpox							

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	Case Definition						
Criterion	Confirmed			Probable		Suspected	
Contact with smallpox infected bodily fluids or contaminated objects such as bedding or clothing							
Worker in a laboratory that contains smallpox virus							

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.
1 = flat type smallpox
2 = hemorrhagic smallpox
3 = *variola sine eruptione*

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 smallpox is recommended.

- The level of investigation and reporting of smallpox cases will depend on the extent of the outbreak and the resources available to conduct these activities. In a limited outbreak, all information on the surveillance form should be obtained, whenever possible. In a larger outbreak, the state epidemiologist may decide, based on available resources, to limit data collection to selected variables. In very large outbreaks, surveillance and case reporting may be limited to aggregate reporting of cases and deaths by age group.

- This information will likely be used be state and federal authorities (representing public health and possibly law enforcement) to determine how the initial infection(s) occurred, the extent of the emergency, its progress and termination. To accomplish this, frequent interim evaluation (daily, weekly, monthly etc) of aggregate/cumulative case data is anticipated.

- In the absence of specific law enforcement concerns, state and territorial health authorities would receive updated information including the results of interim analyses as the evaluations are completed (i.e., daily, weekly, monthly, etc.)

- The schedule of release of published data will be dictated by the nature of the emergency.

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Depending on the nature of the emergency, there may be a legal requirement to provide certain types of information to law enforcement entities. WHO will be notified as per reporting requirements in the 2005 International Health Regulations.

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) Jeffrey Engel, MD
State Health Director
North Carolina Department of Health
1931 Mail Service Center
Raleigh, NC 27699-5000
(919) 707-5000
Jeffrey.engel@ncmail.net

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