09-ID-70

Committee: Infectious

Title: National Surveillance for Human Rabies

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]

Rabies virus infects the central nervous system, causing encephalitis and ultimately death. Early symptoms of rabies in humans are nonspecific, consisting of fever, headache, and general malaise. As the disease progresses, neurological symptoms appear and may include insomnia, anxiety, confusion, slight or partial paralysis, excitation, hallucinations, agitation, hypersalivation, difficulty swallowing, and hydrophobia (fear of water). Death usually occurs within days of the onset of symptoms, although treatment may increase the length of survival and very rarely even result in recovery (CDC 2004).

The number of rabies-related human deaths in the United States declined from more than 100 annually in 1900 to an average of three a year in the past five years (2003–2007). The administration of a standard regimen of postexposure prophylaxis (PEP) to persons exposed to rabies has proven extremely successful in preventing the disease. In the United States, human fatalities from rabies occur in people who fail to seek medical assistance and PEP, usually because they were unaware of the need to seek medical attention, or occasionally, not aware that an exposure occurred.

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

Justification

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

- A majority of state and territorial jurisdictions—or jurisdictions comprising a majority of the US population—have laws or regulations requiring **immediate (urgent)** reporting of human rabies to public health authorities;
- The Centers for Disease Control and Prevention (CDC) requests **immediate (urgent)** notification of human rabies to federal authorities; 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 human rabies 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 human rabies to facilitate its prevention and control.

V. Methods for Surveillance

Surveillance for human rabies 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 human rabies

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

- A person with one or more of the following clinical findings: encephalitis, myelitis, dysphagia, hydrophobia, anxiety, agitation, *or* paresthesias or pain at the wound site; AND one or more of the following laboratory findings:
    - detection by direct fluorescent antibody of rabies viral antigens in a clinical specimen (preferably the brain or the nerves surrounding hair follicles in the nape of the neck)
    - isolation (in cell culture or in a laboratory animal) of rabies virus from saliva, cerebrospinal fluid (CSF), or central nervous system tissue
    - detection of rabies viral RNA (using reverse transcriptase-polymerase chain reaction [RT-PCR]) in saliva, CSF, or tissue
    - Identification of a rabies-neutralizing antibody titer greater than or equal to 5 (complete neutralization) in the CSF

- A person with one or more of the following clinical findings: encephalitis, myelitis, dysphagia, hydrophobia, anxiety, agitation, *or* paresthesias or pain at the wound site; AND no previous vaccination for rabies; AND one or more of the above AND/OR identification of a rabies-neutralizing antibody titer greater than or equal to 5 (complete neutralization) in the person's serum

- A person whose healthcare record contains a diagnosis of rabies.

- A person whose death certificate lists rabies as a cause of death or a significant condition contributing to death.

Other recommended reporting procedures

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

[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.

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	Case Definition Confirmed
Clinical Presentation	
encephalitis	O
myelitis	O
dysphagia	O
hydrophobia	O
anxiety	O
agitation	O
paresthesias or pain at the wound site	O
ascending flaccid paralysis	O
healthcare record contains a diagnosis of rabies	S
death certificate lists rabies as a cause of death or a significant condition contributing to death	S
Laboratory findings	
detection by direct fluorescent antibody of viral antigens in a clinical specimen (preferably the brain or the nerves surrounding hair follicles in the nape of the neck)	S
isolation (in cell culture or in a laboratory animal) of rabies virus from saliva, cerebrospinal fluid (CSF), or central nervous system tissue	S
identification of a rabies-neutralizing antibody titer greater than or equal to 5 (complete neutralization) in the CSF	S
identification of a rabies-neutralizing antibody titer greater than or equal to 5 (complete neutralization) in the serum of an unvaccinated person	S
detection of rabies viral RNA (using reverse transcriptase–polymerase chain reaction [RT-PCR]) in saliva, CSF, or tissue	S
Epidemiologic linkage	
bite by a known or suspect rabid animal	

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

C. Disease Specific Data Elements:

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

See data elements collected on the CDC possible human rabies form (See references or http://www.cdc.gov/rabies/docs/ror_form.pdf) that are not listed in Tables 4 or 5.

Patient Information
-Occupation
-First Symptoms
-Date of Illness Onset
-Hospitalized
-Outpatient Visit Date
-Date Hospitalized
-Outpatient Diagnosis
-Admitting Diagnosis
-Is/was the patient in a coma
-Date of coma onset
-Date of death

Laboratory Findings
-see CDC form
-Rabies variant identified

Symptoms
-see CDC form

Epi Information
-Travel history
-Animal exposure(s)
    -date of exposure
    -species involved
    -type of exposure
- # of persons receiving PEP related to this case
    - # Healthcare workers
    - # Household contacts
    - # Other (specify)

VII. Case Definition

A. Narrative description of criteria to determine whether a case should be classified as confirmed.

Clinical description

Rabies is an acute encephalomyelitis that almost always progresses to coma or death within 10 days after the first symptom.

Laboratory criteria for diagnosis

- Detection by direct fluorescent antibody of viral antigens in a clinical specimen (preferably the brain or the nerves surrounding hair follicles in the nape of the neck), or
- Isolation (in cell culture or in a laboratory animal) of rabies virus from saliva, cerebrospinal fluid (CSF), or central nervous system tissue, or
- Identification of a rabies-neutralizing antibody titer greater than or equal to 5 (complete neutralization) in the CSF, or
- Identification of a rabies-neutralizing antibody titer greater than or equal to 5 (complete neutralization) in the serum of an unvaccinated person.

Case classification

Confirmed: a clinically compatible case that is laboratory confirmed

Comment

Laboratory confirmation by all of the above methods is strongly recommended.

B. Classification Tables

Table VII-B lists the criteria that must be met for a case to be classified as confirmed.

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
Clinical Presentation	
encephalitis	O
myelitis	O
dysphagia	O

hydrophobia	O
anxiety	O
agitation	O
paresthesias or pain at the wound site	O
ascending flaccid paralysis	O
healthcare record contains a diagnosis of rabies	O
death certificate lists rabies as a cause of death or a significant condition contributing to death	O
Laboratory findings	
detection by direct fluorescent antibody of viral antigens in a clinical specimen (preferably the brain or the nerves surrounding hair follicles in the nape of the neck)	S
isolation (in cell culture or in a laboratory animal) of rabies virus from saliva, cerebrospinal fluid (CSF), or central nervous system tissue	S
identification of a rabies-neutralizing antibody titer greater than or equal to 5 (complete neutralization) in the CSF	S
identification of a rabies-neutralizing antibody titer greater than or equal to 5 (complete neutralization) in the serum of an unvaccinated person	S
detection of rabies viral RNA (using reverse transcriptase–polymerase chain reaction [RT-PCR]) in saliva, CSF, or tissue	S
Epidemiologic linkage	
bite by a known or suspect rabid animal	

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 classify 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 classify a case.

VIII. Period of Surveillance

Surveillance should be on-going.

IX. Data sharing/release and print criteria

Notification to CDC for confirmed and probable cases is recommended.

Expectations for sharing case data

- CDC will submit aggregate national counts of reports of cases of human rabies to WHO **annually**.

Limitations on releasing case data

- Release of any identifying data to any entity other than to another state's or territory's health agency or to the CDC requires a signed data sharing agreement using a format pre-approved by the state or territorial health agency. (Refer to the *CDC-CSTE Intergovernmental Data Release Guidelines Working Group Report: CDC-ATSDR Data Release Guidelines and Procedures for Re-release of State-Provided Data* for further information.)

Restrictions on publishing case data

- CDC will only publish data on **confirmed** cases of human rabies. Provisional data will not be used until verification procedures are complete.

Rabies in humans in the United States is a rare event. Reported cases of rabies in humans has dropped since the 1940s as canine rabies has been controlled and ultimately eliminated in the United States. However, during this time period the number of wildlife associated cases of rabies reported in animals has increased. Subsequently potential human exposure to rabies and postexposure prophylaxis (PEP) remains a relatively common event in the United States (35,000 to 45,000 PEP's annually). In addition, over the past 2 decades human cases of rabies has increasingly implicated bats as the primary source of human infection. Information regarding the demographic, temporal, and geographic nature of human rabies is required to facilitate its prevention and control.

Data reported through the National Notifiable Diseases Surveillance System (NNDSS) is summarized weekly in the MMWR Tables and yearly in the MMWR Summary of Notifiable Diseases. Brief case reports are generated annually in the annual rabies surveillance summary report published in September of each year. In addition, ad hoc publication of more detailed case reports are submitted to MMWR or other peer reviewed journals. Cumulative reports of human rabies cases examining characteristics and trends are produced approximately every 10 years.

A majority of state and territorial jurisdictions—or jurisdictions comprising a majority of the US population—have laws or regulations requiring immediate reporting of human rabies to public health authorities. Response and laboratory assistance may be required/requested from state for potential contact/exposure assessment. CDC provides laboratory assistance for ante-mortem testing of rabies in humans, provides ongoing laboratory support in human cases under experimental treatment protocol for rabies, provides support to state health departments for contact tracing and exposure assessment in hospital and community setting and assists with interstate communications when necessary and potentially internationally for imported cases.

X. References

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