

Committee: Infectious

Title: National Surveillance for Tularemia

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

Background¹

Tularemia is a zoonotic disease caused by the gram-negative coccobacillus *Francisella tularensis*. Tularemia characteristically presents as an acute febrile illness. Various clinical manifestations can occur depending on the route of infection and host response, including an ulcer at the site of cutaneous or mucous membrane inoculation, pharyngitis, ocular lesions, regional lymphadenopathy, and pneumonia. A diagnosis of tularemia can be confirmed by culture of *F. tularensis* from clinical specimens or by a fourfold titer change of serum antibodies against *F. tularensis*. Presumptive diagnosis can be made by detecting *F. tularensis* antigens with fluorescent assays or by a single elevated antibody level. If untreated with appropriate antibiotics, tularemia can be fatal.

Known also as “rabbit fever” and “deer fly fever,” tularemia was first described in the United States in 1912 and has been reported from all states except Hawaii. From 1992 to 1999, the top five states in number and incidence of cases of tularemia were Arkansas, Missouri, Montana, Oklahoma, and South Dakota. Human infection can occur in several ways: being bitten by an infected tick, deerfly or other insect; handling infected animal carcasses (most commonly rodents, rabbits, and hares); eating or drinking contaminated food or water; and breathing in the bacterium. Tularemia is not known to be spread from person to person. Tularemia was removed from the list of nationally notifiable diseases in 1994, but increased concern about potential use of *F. tularensis* as a biological weapon led to its return in 2000.

¹ Much of the material in the background is directly quoted from the CDC’s Tularemia Website, Chang 2003, and CDC MMWR 2002. See the References for further information on these sources.

Justification

Tularemia 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 is included on the list of Category A possible bioterrorism agents and toxins.
- A majority of state and territorial jurisdictions—or jurisdictions comprising a majority of the US population—have laws or regulations requiring **immediate** and **standard** reporting of tularemia to public health authorities;
- The Centers for Disease Control and Prevention (CDC) requests **immediate and standard** notification of tularemia; 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 tularemia 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 tularemia to facilitate its prevention and control.

V. Methods for Surveillance

Surveillance for tularemia 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 tularemia

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. 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 for whom a diagnostic test specific for tularemia has been ordered (see table VI-B).
- A person with a clinical presentation consistent with tularemia (see tables VI-B and VII-B) AND who works—or recently worked—in a laboratory that handles tularemia specimens or *F. tularensis* cultures.
- A person with a clinical presentation consistent with tularemia (see tables VI-B and VII-B) AND a history of a tick or deerfly bite; exposure to water potentially contaminated by *F. tularensis*; or handling rodent, rabbit, or hare tissues or fluids; AND who lives in—or recently traveled to—rural areas of states with significant numbers of recent cases of tularemia.
- A person whose healthcare record contains a diagnosis of tularemia.
- A person whose death certificate lists tularemia as a cause of death or a significant condition contributing to death.

Other recommended reporting procedures

- All cases of tularemia 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	Reporting		
<i>Clinical Presentation</i>			
fever (> 38° C)		N	N
cutaneous ulcer		N1	N1
regional lymphadenopathy		N1,N2	N1,N2
conjunctivitis		N3	N3
preauricular lymphadenopathy		N3	N3
stomatitis		O4	O4
pharyngitis		O4	O4
tonsillitis		O4	O4
cervical lymphadenopathy		N4	N4
intestinal pain		N5	N5
vomiting		N5	N5
diarrhea		N5	N5
pleuropneumonitis		O6	O6
hilar lymphadenopathy		O6	O6
healthcare record contains a diagnosis of tularemia	S		
death certificate lists tularemia as a cause of death or a significant condition contributing to death	S		
<i>Laboratory findings</i>			
elevated serum antibody titer(s) to <i>F. tularensis</i> antigen (without documented fourfold or greater change)	S*		
detection of <i>F. tularensis</i> in a clinical specimen by fluorescent assay	S*		
isolation of <i>F. tularensis</i> from a clinical specimen	S*		
fourfold or greater change in serum antibody titer to <i>F. tularensis</i> antigen	S*		
detection of <i>F. tularensis</i> in blood by nucleic acid amplification (PCR)	S		

<i>Epidemiological risk factors</i>			
history of tick bite			O
history of deerfly bite			O
exposure to water potentially contaminated by <i>F. tularensis</i>			O
handling rodent, rabbit, or hare tissues or fluids			O
hunting or trapping rabbits or hares			O
working in a laboratory that handles tularemia specimens or <i>F. tularensis</i> cultures		N	
living in—or recently traveling to—rural areas of states with significant numbers of recent cases of tularemia (e.g., Arkansas, Missouri, Montana, Oklahoma, or South Dakota)			N

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 the “O” criteria in each category (e.g., clinical presentation and laboratory findings) in the same column—are required to report a case. A number following an “N” indicates that this criterion is only required for a specific clinical presentation (see below).

O = At least one of these “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 number following an “O” indicates that this criterion is only required for a specific clinical presentation (see below).

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

1 = Ulceroglandular tularemia: cutaneous ulcer with regional lymphadenopathy (21–87% of cases in the U.S.)

2 = Glandular tularemia: regional lymphadenopathy with no ulcer (3–20% of cases in the U.S.)

3 = Oculoglandular tularemia: conjunctivitis with preauricular lymphadenopathy (0–5% of cases)

4 = Oropharyngeal tularemia: stomatitis or pharyngitis or tonsillitis and cervical lymphadenopathy (0–12% of cases)

5 = Intestinal tularemia: intestinal pain, vomiting, and diarrhea

6 = Pneumonic tularemia: primary pleuropulmonary disease (7–20% of cases)

7 = Typhoidal tularemia: febrile illness without early localizing signs and symptoms (5–30% of cases)

C. Disease Specific Data Elements:

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

Epidemiological information

Evidence or history of a tick or deerfly bite

Exposure to tissues of a mammalian host of *Francisella tularensis*

Exposure to potentially contaminated water

Work in a laboratory that handles tularemia specimens or *F. tularensis* cultures

Living in—or recently traveling to—rural areas of states with significant numbers of recent cases of tularemia

VII. Case Definition for Case Classification

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

Clinical description

An illness characterized by several distinct forms, including the following:

- Ulceroglandular: cutaneous ulcer with regional lymphadenopathy
- Glandular: regional lymphadenopathy with no ulcer
- Oculoglandular: conjunctivitis with preauricular lymphadenopathy
- Oropharyngeal: stomatitis or pharyngitis or tonsillitis and cervical lymphadenopathy
- Intestinal: intestinal pain, vomiting, and diarrhea
- Pneumonic: primary pleuropulmonary disease
- Typhoidal: febrile illness without early localizing signs and symptoms

Clinical diagnosis is supported by evidence or history of a tick or deerfly bite, exposure to tissues of a mammalian host of *Francisella tularensis*, or exposure to potentially contaminated water.

Laboratory criteria for diagnosis

Presumptive

- Elevated serum antibody titer(s) to *F. tularensis* antigen (without documented fourfold or greater change) in a patient with no history of tularemia vaccination, *or*
- Detection of *F. tularensis* in a clinical specimen by fluorescent assay

Confirmatory

- Isolation of *F. tularensis* in a clinical specimen, *or*
- Fourfold or greater change in serum antibody titer to *F. tularensis* antigen

Case classification

Probable: a clinically compatible case with laboratory results indicative of presumptive infection

Confirmed: a clinically compatible case with confirmatory laboratory results

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>			
fever (> 38° C)	N	N	N
cutaneous ulcer	N1	N1	N1
regional lymphadenopathy	N1,N2	N1,N2	N1,N2
conjunctivitis	N3	N3	N3
preauricular lymphadenopathy	N3	N3	N3
stomatitis	O4	O4	O4
pharyngitis	O4	O4	O4
tonsillitis	O4	O4	O4
cervical lymphadenopathy	N4	N4	N4
intestinal pain	N5	N5	N5
vomiting	N5	N5	N5
diarrhea	N5	N5	N5
pleuropneumonitis	N6	N6	N6
hilar lymphadenopathy			
healthcare record contains a diagnosis of tularemia			
death certificate lists tularemia as a cause of death or a significant condition contributing to death			

<i>Laboratory findings</i>			
elevated serum antibody titer(s) to <i>F. tularensis</i> antigen (without documented fourfold or greater change)		N	
detection of <i>F. tularensis</i> in a clinical specimen by fluorescent assay			N
isolation of <i>F. tularensis</i> from a clinical specimen	O		
fourfold or greater change in serum antibody titer to <i>F. tularensis</i> antigen	O		
detection of <i>F. tularensis</i> in blood by nucleic acid amplification (PCR)			
<i>Epidemiological risk factors</i>			
history of tularemia vaccination		A	
history of tick bite			
history of deerfly bite			
exposure to water potentially contaminated by <i>F. tularensis</i>			
handling rodent, rabbit, or hare tissues or fluids			
hunting or trapping rabbits or hares			
working in a laboratory that handles tularemia specimens or <i>F. tularensis</i> cultures			
living in—or recently traveling to—rural areas of states with significant numbers of recent cases of tularemia (e.g., Arkansas, Missouri, Montana, Oklahoma, or South Dakota)			

Notes:

N = All “N” criteria in the same column—in conjunction with at least one of the “O” criteria in each category (e.g., clinical presentation and laboratory findings) in the same column—are required to classify a case. A number following an “N” indicates that this criterion is only required for a specific clinical presentation (see below).

O = At least one of these “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. A number following an “O” indicates that this criterion is only required for a specific clinical presentation (see below).

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

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

1 = Ulceroglandular tularemia: cutaneous ulcer with regional lymphadenopathy (21–87% of cases in the U.S.)

2 = Glandular tularemia: regional lymphadenopathy with no ulcer (3–20% of cases in the U.S.)

3 = Oculoglandular tularemia: conjunctivitis with preauricular lymphadenopathy (0–5% of cases)

- 4 = Oropharyngeal tularemia: stomatitis or pharyngitis or tonsillitis and cervical lymphadenopathy (0–12% of cases)
- 5 = Intestinal tularemia: intestinal pain, vomiting, and diarrhea
- 6 = Pneumonic tularemia: primary pleuropulmonary disease (7–20% of cases)
- 7 = Typhoidal tularemia: febrile illness without early localizing signs and symptoms (5–30% of cases)

VIII. Period of Surveillance

Surveillance should be on-going.

IX. Data sharing/release and print criteria

Notification to CDC for confirmed, probable, and suspected cases of tularemia as well as all cases prior to classification is recommended.

- Immediate (extremely urgent) notification to CDC should occur for all suspected, probable and confirmed cases if intentional release of tularemia is suspected as the cause of infection.
- Standard notification should be used for all other cases.
- Provisional data on tularemia cases reported through NETSS/NNDSS are summarized weekly in the MMWR, and finalized data are published annually in the Summary of Notifiable Diseases. Longer articles describing and interpreting national trends are published in the MMWR on an ad-hoc basis (approximately once every other year).
- State-specific compiled data will continue to be published in the weekly and annual MMWR.
- Provisional state-specific compiled data will continue to be published in the weekly reports. Finalized data will be published in the annual MMWR Surveillance Summaries, following verification by each state. The frequency of release of additional publication of this data will be dependent on the current epidemiologic situation in the country. These publications might include annual epidemiologic summaries in the MMWR or manuscripts in peer-reviewed journals.

X. References

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