

RICKETTSIAL DISEASES (RICKETTSIOSES)

Includes Typhus Fever and Spotted Fever Rickettsioses

Background

This case definition was developed by the Armed Forces Health Surveillance Branch (AFHSB) for the purpose of epidemiological surveillance of a condition important to military-associated populations.

Clinical Description

Rickettsial infections are caused by bacteria from the family Rickettsiaceae and the genera *Rickettsia* and *Orientia*. Not included are the genera *Anaplasma*, *Ehrlichia*, *Neorickettsia*, and *Neoehrlichia*.¹ Most infections are transmitted by arthropod vectors including ticks, fleas, mites and lice. Clinical presentation varies with the causative agent and patient; however, common symptoms that typically develop within 1-2 weeks of infection include fever, headache, malaise, rash, nausea, or vomiting. Many infections are also accompanied by a maculopapular, vesicular, or petechial rash and an eschar at the site of the tick or mite bite. Diagnosis is based on clinical recognition, epidemiological context and serologic testing. Treatment includes immediate empiric treatment with tetracycline.²

Case Definition and Incidence Rules

For surveillance purposes:

A *confirmed* case of rickettsial disease is defined as:

- One record of a reportable medical event *with* laboratory or epidemiological confirmation.

A *possible* case of rickettsial disease is defined as:

- *One hospitalization* with a case defining diagnosis of rickettsial disease (see ICD9 and ICD10 code lists below) in *any* diagnostic position.

A *suspected* case of rickettsial disease is defined as:

- One record of a reportable medical event *without* laboratory or epidemiological confirmation; or
- *One outpatient medical encounter* with a case defining diagnosis of rickettsial disease (see ICD9 and ICD10 code lists below) in the *first* or *second* diagnostic position.

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¹ Clark L, Williams VW. Historical Review: Rickettsial Diseases and Their Impact on U.S. Military Forces. *Medical Surveillance Monthly Report (MSMR)*. 2019 August; 26(8):28–33.

² Nicholson, WL, Paddock, CD. Rickettsial (Including Spotted Fever & Typhus Fever Rickettsioses, Scrub Typhus, Anaplasmosis, and Ehrlichiosis). *CDC Health Information for International Travel 2020: The Yellow Book*. Available at: <https://wwwnc.cdc.gov/travel/yellowbook/2020/travel-related-infectious-diseases/rickettsial-including-spotted-fever-and-typhus-fever-rickettsioses-scrub-typhus-anaplasmosis-and-ehr>. Accessed August 2019.



Case Definition and Incidence Rules *(continued)*

Incidence rules:

For individuals who meet the case definition:

- The incidence date is considered the date of onset documented in a reportable medical event report, or the first hospitalization or outpatient medical encounter that includes a defining diagnosis of rickettsial disease.
- An individual is considered an incident case only *once per lifetime* for each type of rickettsial disease.
- Confirmed cases are prioritized over possible cases and suspected cases, respectively.

Exclusions:

- None

Codes

The following ICD9 and ICD10 codes are included in the case definition:

Condition	ICD-10-CM Codes	ICD-9-CM Codes
Typhus fever rickettsiosis	<i>A75 (typhus fever)</i>	
	A75.0 (epidemic louse-borne typhus fever due to <i>Rickettsia prowazekii</i>)	080 (louse-borne (epidemic) typhus)
	--	<i>081 (other typhus)</i>
	- A75.1 (recrudescent typhus [Brill's disease])	- 081.1 (Brill's disease)
	- A75.2 (typhus fever due to <i>Rickettsia typhi</i>)	- 081.0 (Murine (endemic) typhus)
	- A75.3 (typhus fever due to <i>Rickettsia tsutsugamushi</i>)	- 081.2 (Scrub typhus)
	- A75.9 (typhus fever, unspecified)	- 081.9 (typhus, unspecified)
Spotted fever rickettsiosis	<i>A77 (spotted fever [tick-borne rickettsioses])</i>	<i>082 (tick-borne rickettsioses)</i>
	A77.0 (spotted fever due to <i>Rickettsia rickettsii</i>)	082.0 (spotted fevers)
	A77.1 (spotted fever due to <i>Rickettsia conorii</i>)	082.1 (boutonnesuse fever)

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	A77.2 (spotted fever due to <i>Rickettsia siberica</i>)	082.2 (North Asian tick fever)
	A77.3 (spotted fever due to <i>Rickettsia australis</i>)	082.3 (Queensland tick typhus)
	A77.8 (other spotted fevers)	082.8 (other specified tick-borne rickettsioses)
	A77.9 (spotted fever, unspecified)	082.9 (tick-borne rickettsiosis, unspecified)
Other rickettsioses	A79.1 (Rickettsialpox due to <i>Rickettsia akari</i>)	083.2 (rickettsialpox)
	- A79.89 (other specified rickettsioses)	083.8 (other specified rickettsioses)
	A79.9 (rickettsiosis, unspecified)	083.9 (rickettsiosis, unspecified)

Development and Revisions

- This case definition was developed in August of 2019 by the *Medical Surveillance Monthly Report (MSMR)* staff for surveillance of rickettsial diseases. The definition was developed based on reviews of the ICD9 and ICD10 codes, the scientific literature, and previous AFHSB analyses.

Case Definition and Incidence Rule Rationale

- For the purposes of counting new incident cases, AFHSB uses a once per lifetime incidence rule unless a specific timeframe is more appropriate and is specified (e.g., individuals may be counted as an incident case once every 365 days). Historically, a "once per surveillance period" incidence rule was used due to limited data in the Defense Medical Surveillance System (DMSS), but that is no longer necessary.

Code Set Determination and Rationale

- In addition to the ICD9 and ICD10 codes used in the case definition documented above, the ICD-10-CM Manual includes the following codes in the category Rickettsioses (ICD10 A75-A79) (see table below).

The intent of this case definition is to capture those illnesses which have historically been associated with military field operations (including war) or other activities which carried an increased risk of exposure to the vectors of the agents originally classified as rickettsiae. Modern microbiology has permitted the clarification of the classification of a range of agents once thought to be rickettsiae. This case definition is focused on those bacteria now identified as members of the family Rickettsiaceae, (i.e., the genera *Rickettsia* and *Orientia*). Not included in this case definition are members of another family that is similar to Rickettsiaceae, (i.e., the family Anaplasmataceae), which includes the genera *Anaplasma*, *Ehrlichia*, and *Neoehrlichia*. Those genera are candidates for a separate case definition for anaplasmosis and ehrlichiosis. Trench fever is not included in this case definition because that louse-borne infection is caused by the *Bartonella quintana*, and would be better grouped with the agents of sandfly-transmitted Carrion's disease (*B. bacilliformis*) and cat scratch disease (*B. henselae*). Q-fever is not included because the causative agent *Coxiella burnetii* is no longer classified as a rickettsia and it is not transmitted by insect vectors. Molecular studies have reclassified *C. burnetii* as a Proteobacteria in the same group as Legionella.³

³ Centers for Disease Control and Prevention. *Q Fever*. Available at: <https://www.cdc.gov/qfever/>. Accessed August 2019.



Condition	ICD-10-CM Codes	ICD-9-CM Codes
Ehrlichiosis	A77.4 (ehrlichiosis)	082.4 (ehrlichiosis)
	- A77.40 (ehrlichiosis, unspecified)	- 082.40 (ehrlichiosis, unspecified)
	- A77.41 (ehrlichiosis chafeensis [<i>E. chafeensis</i>])	- 082.41 (ehrlichiosis chafeensis [<i>E. chafeensis</i>])
	- A77.49 (other ehrlichiosis)	- 082.49 (other ehrlichiosis)
	- A79.81 (rickettsiosis due to ehrlichia sennetsu [<i>E. sennetsu</i>])	-
Other rickettioses	A79.0 (trench fever)	083.1 (trench fever)
Q fever	A78 (Q fever)	083.0 (Q fever)

Reports

AFHSB reports on rickettsial diseases in the following reports:

- Periodic *MSMR* articles

Review

- Sep 2019 Case definition reviewed and adopted by the AFHSB Surveillance Methods and Standards (SMS) working group.
- Aug 2019 Case definition developed and adopted by the AFHSB *MSMR* staff.

Comments

None