

*** Proposed new case definition; to be confirmed by AFHSC following analyses of data**

SYPHILIS

Includes Early Syphilis (Primary, Secondary, Early Latent) and Late Syphilis (Tertiary, Late Latent); Does not include Congenital Syphilis

Background

This case definition was developed in 2013 by the Armed Forces Health Surveillance Center (AFHSC) for the purpose of epidemiological surveillance of a sexually transmitted infection important to military associated populations.

Clinical Description

Syphilis is a systemic, sexually transmitted, disease caused by the bacterium *Treponema pallidum*. The disease is transmitted from person to person by direct contact with syphilis sores that occur primarily on the external genitals, vagina, anus, and rectum. Sores can also occur on the lips and mouth.¹ The disease is divided into overlapping stages based on the appearance of symptoms. The early (primary and secondary) stage is characterized by a painless sore(s) at the site of inoculation that usually lasts 3 to 6 weeks. If the disease is not treated at this stage, it progresses to the secondary stage characterized by skin rashes, often on the palms of the hands and soles of the feet, mucous membrane lesions, fever, lymphadenopathy, weight loss, and fatigue. If not treated, the symptoms usually resolve in 3 to 6 weeks and the disease progresses to the early latent and late (tertiary) stages of infection. Approximately 15% of individuals not treated for syphilis develop late stage disease. Symptoms in this stage can appear 10-30 years after infection and are the result of damage to the central nervous system, the aorta, and, less often, other internal organs.² Syphilis is effectively treated with early detection and stage-specific treatment with antibiotics.

Case Definition and Incidence Rules – Early Syphilis (Primary, Secondary, Early Latent)

For surveillance purposes, a case of *early* syphilis is defined as:

- *One record of a hospitalization* with any of the defining diagnoses of early syphilis (see ICD9 and ICD10 code lists below) in the *primary, secondary, or tertiary* diagnostic position; or
- *One outpatient medical encounter* with any of the defining diagnoses of early syphilis (see ICD9 and ICD10 code lists below) in the *primary or secondary* diagnostic position; or
- *One record of a reportable medical event* of syphilis.

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¹ <http://www.cdc.gov/std/Syphilis/STDFact-Syphilis.htm>; accessed Sept 16, 2013

² <http://www.cdc.gov/std/treatment/2010/syphilis-infections.htm>; accessed October 2013.



Case Definition and Incidence Rules *(continued)*

Incidence rules:

For individuals who meet the case definition:

- The incidence date is considered the date of the first reportable medical event, hospitalization or outpatient medical encounter with a defining diagnosis of early syphilis.
- An individual is considered a new incident case if *at least 365 days* have passed since the last medical encounter with a case defining diagnosis of early syphilis (see explanation of “gap” rule below).

Exclusions:

- Individuals with a diagnosis of syphilis prior to the surveillance period.

Case Definition and Incidence Rules – Late Syphilis (Tertiary, Late Latent)

For surveillance purposes, a case of *late* syphilis is defined as:

- *One record of a hospitalization* with any of the defining diagnoses of late syphilis (see ICD9 and ICD10 code lists below) in the *primary, secondary, or tertiary* diagnostic position; or
- *One outpatient medical encounter* with any of the defining diagnoses of late syphilis (see ICD9 and ICD10 code lists below) in the *primary or secondary* diagnostic position; or
- *One record of a reportable medical event* of syphilis.

Incidence rules:

For individuals who meet the case definition:

- The incidence date is considered the date of the first reportable medical event, hospitalization or outpatient medical encounter with a defining diagnosis of late syphilis.
- An individual is considered a new incident case of late syphilis *once per lifetime*.

Exclusions:

- Individuals with a diagnosis of syphilis prior to the surveillance period.



Codes

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

Condition	ICD-10-CM Codes	ICD-9-CM Codes
Early Syphilis	A51 Early syphilis	091 Early syphilis symptomatic
	- A51.0 (primary genital syphilis)	091.0 (genital syphilis, primary)
	- A51.1 (primary anal syphilis)	091.1 (primary anal syphilis)
	- A51.2 (primary syphilis of other sites)	091.2 (other primary syphilis)
	- A51.3 (secondary syphilis of skin and mucous membranes)	--
	- A51.31 (condyloma latum)	091.3 (secondary syphilis of skin or mucous membranes)
	- A51.32 (syphilitic alopecia)	098.82 (syphilitic alopecia)
	- A51.39 (other secondary syphilis of skin)	091.3 (above)
	A51.4 (other secondary syphilis)	--
	- A51.41 (secondary syphilitic meningitis)	091.81 (acute syphilitic meningitis; secondary)
	- A51.42 (secondary syphilitic female pelvic disease)	091.89 (other forms of secondary syphilis; other)
	- A51.43 (secondary syphilitic oculoopathy)	091.5 (uveitis due to secondary syphilis) - 091.50 (syphilitic uveitis, unspecified) - 091.51 (syphilitic chorioretinitis; secondary) - 091.52 (syphilitic iridocyclitis; secondary)
	- A51.44 (secondary syphilitic nephritis)	091.89 (above)
	- A51.45 (secondary syphilitic hepatitis)	091.62 (secondary syphilitic hepatitis)
	- A51.46 (secondary syphilitic osteopathy)	091.61 (secondary syphilitic periostitis)
	- A51.49 (other secondary syphilitic conditions)	091.4 (adenopathy due to secondary syphilis) 091.6 (secondary syphilis of viscera and bone) 091.69 (secondary syphilis, other viscera) 091.7 (secondary syphilis, relapse) 091.8 (other forms of secondary syphilis) 091.89 (above) 091.9 (unspecified secondary syphilis)

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A51.5 (early syphilis, latent)	092 Early syphilis, latent 092.0 (early syphilis, latent, serologic relapse after treatment) 092.9 (early syphilis, latent, unspecified)
A51.9 (early syphilis, unspecified)	092.9 (above)

Condition	ICD-10-CM Codes	ICD-9-CM Codes
Late Syphilis	A52 Late syphilis	
	--	093 Cardiovascular syphilis
	A52.0 Cardiovascular and cerebrovascular syphilis	--
	- A52.00 (cardiovascular syphilis, unspecified)	093.9 (cardiovascular syphilis, unspecified)
	- A52.01 (syphilitic aneurysm of aorta)	093.0 (aneurysm of aorta, specified as syphilitic)
	- A52.02 (syphilitic aortitis)	093.1 (syphilitic aortitis)
	- A52.03 (syphilitic endocarditis)	093.2 (syphilitic endocarditis) - 093.20 (syphilitic endocarditis; valve unspecified) - 092.21 (syphilitic endocarditis of mitral valve) - 093.22 (syphilitic endocarditis of aortic valve) - 093.23 (syphilitic endocarditis of tricuspid valve) - 093.24 (syphilitic endocarditis of pulmonary valve)
	- A52.04 (syphilitic cerebral arteritis)	- 093.89 (other specified cardiovascular syphilis)
	- A52.05 (other cerebrovascular syphilis)	- 093.89 (above)
	A52.06 (other syphilitic heart involvement)	093.8 (other specified cardiovascular syphilis) - 093.81 (syphilitic pericarditis) - 093.82 (syphilitic myocarditis)
	A52.09 (other cardiovascular syphilis)	- 093.89 (above)
	--	094 Neurosyphilis
	A52.1 (symptomatic neurosyphilis)	--
	- A52.10 (symptomatic neurosyphilis, unspecified)	094.9 (neurosyphilis, unspecified) <i>(continued on next page)</i>



- A52.11 (tabes dorsalis)	094.0 (tabes dorsalis)
- A52.12 (other cerebrospinal syphilis)	094.9 (above)
- A52.13 (late syphilitic meningitis)	094.2 (syphilitic meningitis)
- A52.14 (late syphilitic encephalitis)	094.81 (syphilitic encephalitis)
- A52.15 (late syphilitic neuropathy)	- 094.84 (syphilitic optic atrophy) - 094.85 (syphilitic retrobulbar neuritis) - 094.86 (syphilitic acoustic neuritis)
- A52.16 (Charcot's arthropathy, tabetic)	094.0 (above) with 713.5 (arthropathy associated with neurological disorders)
- A52.17 (general paresis)	094.1 (general paresis)
--	094.8 (other specified neurosyphilis_
- A52.19 (other symptomatic neurosyphilis)	- 094.82 (syphilitic Parkinsonism) - 094.83 (syphilitic disseminated retinochoroiditis) - 094.87 (syphilitic ruptured cerebral aneurysm) - 094.89 (other specified neurosyphilis)
A52.2 (asymptomatic neurosyphilis)	094.3 (asymptomatic neurosyphilis)
A52.3 (neurosyphilis, unspecified)	094.9 (above)
A52.7 (other symptomatic late syphilis)	095 (other forms of late syphilis with symptoms)
- A52.71 (late syphilitic oculoopathy)	095.0 (syphilitic episcleritis)
- A52.72 (syphilis of lung and bronchus)	095.1 (syphilis of lung)
- A52.73 (symptomatic late syphilis of other respiratory organs)	095.8 (other specified forms of late symptomatic syphilis)
- A52.74 (syphilis of liver and other viscera)	095.2 (syphilitic peritonitis) 095.3 (syphilis of liver)
- A52.75 (syphilis of kidney and ureter)	095.4 (syphilis of kidney)
- A52.76 (other genitourinary symptomatic late syphilis)	095.8 (above)
- A52.77 (syphilis of bone and joint)	095.5 (syphilis of bone)
- A52.78 (syphilis of other musculoskeletal tissue)	095.6 (syphilis of muscle) 095.7 (syphilis of synovium, tendon, bursa)
- A52.79 (other symptomatic late syphilis)	095.8 (above) 095.9 (late symptomatic syphilis, unspecified)

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	A52.8 (late syphilis, latent)	096 (late syphilis, latent)
	A52.9 (late syphilis, unspecified)	097.0 (late syphilis, unspecified)
	A53 (other and unspecified syphilis)	097 (other and unspecified syphilis)
	- A53.0 (latent syphilis, unspecified as early or late)	097.1 (latent syphilis, unspecified)
	- A53.9 (syphilis, unspecified)	097.9 (syphilis, unspecified)

Development and Revisions

- In July of 2014 the case definition was updated to include ICD10 codes. The SMS working group proposed updated case definition to include new incidence rules for late syphilis.

- In 2013 AFHSC updated the case definitions for sexually transmitted infections (STIs) to be more specific for each of the individual STIs. For syphilis the case defining criteria were updated to include hospitalizations with case defining ICD9 codes in the primary, secondary, and tertiary positions only, and outpatient medical encounters with ICD9 codes in the primary and secondary diagnostic positions only. The incidence rule for an individual to be considered a new case was updated from a 30 day absolute rule to a 365-day gap rule. Reports using this case definition were featured in a March 2013 Webinar on “STDs in Military Populations.”³

This case definition was developed based on reviews of the ICD9 codes, the scientific literature, and previous AFHSC analyses. The definition was developed by the AFHSC Medical Surveillance Monthly Report (MSMR) staff for a February 2013 article on syphilis and other STIs.⁴

- In September 2009, a case definition for sexually transmitted infections (STIs) was developed by AFHSC and a Department of Defense (DoD) HIV/STI Prevention working group. The definition was used for a descriptive epidemiology report on the most frequent sexually transmitted infections among active component members of the U.S. Armed Forces.⁵ The case defining criteria in this definition included hospitalizations, used ICD9 codes in *any* diagnostic position, and were the same for all STIs in the report.

Case Definition and Incidence Rule Rationale

- *One record of a hospitalization* with any of the defining diagnoses of syphilis (see ICD9 and ICD10 code lists below) in the *primary, secondary, or tertiary* diagnostic position.
- *For early syphilis* this case definition uses a 365-day “gap in care” incidence rule for medical encounters to define a new incident case of early syphilis infection. Use of this methodology presumes that medical encounters for syphilis infection that occur within 365 days of a previous such encounter constitute follow-up care of the previously diagnosed case. The goal of the “gap in care” rule is to lessen the frequency with which encounters for follow-up care are treated as new incident cases of infection.

³ Focus on the Treatment of STDs in Military Populations – 2010 STD Treatment Guidelines Webinar. See <http://www.cdc.gov/std/training/webinars.htm>.

⁴ Armed Forces Health Surveillance Center. Sexually Transmitted Infections, Active Component, U.S. Armed Forces, 2000-2012. *Medical Surveillance Monthly Report (MSMR)*. 2013 February; 20(2): 5-10.

⁵ Armed Forces Health Surveillance Center. Sexually Transmitted Infections, U.S. Armed Forces, 2004-2009. *Medical Surveillance Monthly Report (MSMR)*; 2010 August; Vol.17(8): 2-10.



The “gap in care” rule differs slightly from an absolute 365-day incidence rule in which an individual may be considered an incident case once every 365 days. Analyses comparing the two incidence rules show an approximate 2% difference between the methodologies, with the gap in care rule being slightly more specific. Use of the “gap in care rule” for this case definition is consistent with the methodology used by other investigators doing syphilis surveillance.

- *For early syphilis* a period of 365 days between encounters is used to allow for adequate resolution of infection after treatment and to avoid confusing medical encounters for follow-up care with medical encounters for a new infection.
- *For late syphilis* this case definition uses an incidence rule that permits an individual to be counted as a case of late syphilis only once per lifetime.

Code Set Determination and Rationale

- This case definition is intended to capture cases of syphilis in an adult population. Therefore, ICD9 codes 090.x (congenital syphilis) are not include in the code set.

Reports

AFHSC reports on syphilis in the following reports:

- Monthly: AFHSC Reportable Events Monthly Report. Available on the AFHSC website at: <http://www.afhsc.mil>; see “Reports and Publications”.
- Weekly: DoD Communicable Disease Weekly Report; Summary of Communicable Reportable Events by Service; Available on the AFHSC website at: <http://www.afhsc.mil>; see “Reports and Publications”.

Review

Jul 2014	Case definition reviewed and updated by the AFHSC Surveillance Methods and Standards (SMS) working group.
Feb 2013	Case definition reviewed and adopted by the AFHSC MSMR staff.

Comments

Late syphilis: During SMS review, service partners noted there are basically no cases of late syphilis in their surveillance populations so they do not include codes for late syphilis in their case definition at all. The SMS workgroup felt that for AFHSC purposes (i.e., possible surveillance of beneficiaries and VA population) it would still be worthwhile to separate the case definition into early and late infection and to document as such. A lifetime incidence rule vs a 2 year absolute or gap incidence rule was proposed.

