Syphilis, Congenital (rev. 01/2015)

Clinical description

A condition caused by infection in utero with *Treponema pallidum*. A wide spectrum of severity exists, from inapparent infection to severe cases that are clinically apparent at birth. An infant or child (aged less than 2 years) may have signs such as hepatosplenomegaly, rash, condyloma lata, snuffles, jaundice (nonviral hepatitis), pseudoparalysis, anemia, or edema (nephrotic syndrome and/or malnutrition). An older child may have stigmata (e.g., interstitial keratitis, nerve deafness, anterior bowing of shins, frontal bossing, mulberry molars, Hutchinson teeth, saddle nose, rhagades, or Clutton joints).

Laboratory criteria for diagnosis

Demonstration of *Treponema pallidum* by:

- Darkfield microscopy of lesions, body fluids, or neonatal nasal discharge, or
- Polymerase chain reaction (PCR) or other equivalent direct molecular methods of lesions, neonatal nasal discharge, placenta, umbilical cord, or autopsy material, or
- Immunohistochemistry (IHC), or special stains (e.g., silver staining) of specimens from lesions, placenta, umbilical cord, or autopsy material.

Case classification

*Probable*: a condition affecting an infant whose mother had untreated or inadequately treated* syphilis at delivery, regardless of signs in the infant, or an infant or child who has a reactive non-treponemal test for syphilis (Venereal Disease Research Laboratory [VDRL], rapid plasma reagin [RPR], or equivalent serologic methods) AND any one of the following:

- Any evidence of congenital syphilis on physical examination (see Clinical description)
- Any evidence of congenital syphilis on radiographs of long bones
- A reactive cerebrospinal fluid (CSF) venereal disease research laboratory test (VDRL) test
- In a nontraumatic lumbar puncture, an elevated CSF leukocyte (white blood cell, WBC) count or protein (without other cause)

Suggested parameters for abnormal CSF WBC and protein values:

1. During the first 30 days of life, a CSF WBC count of >15 WBC/mm3 or a CSF protein >120 mg/dL.
2. After the first 30 days of life, a CSF WBC count of >5 WBC/mm3 or a CSF protein >40 mg/dL, regardless of CSF serology
The treating clinician should be consulted to interpret the CSF values for the specific patient.

*Confirmed: a case that is laboratory confirmed*

**Syphilitic Stillbirth**

**Clinical case definition**

A fetal death that occurs after a 20-week gestation or in which the fetus weighs greater than 500 g and the mother had untreated or inadequately treated* syphilis at delivery.

*Adequate treatment is defined as completion of a penicillin-based regimen, in accordance with CDC treatment guidelines, appropriate for stage of infection, initiated 30 or more days before delivery.*

**Comment**

Congenital and acquired syphilis may be difficult to distinguish when a child is seropositive after infancy. Signs of congenital syphilis may not be obvious, and stigmata may not yet have developed. Abnormal values for CSF VDRL, WBC count, and protein may be found in either congenital or acquired syphilis. Findings on radiographs of long bones may help because radiographic changes in the metaphysis and epiphysis are considered classic signs of congenitally acquired syphilis. While maternal antibodies can complicate interpretation of serologic tests in an infant, reactive tests past 18 months of age are considered to reflect the status of the child. The decision may ultimately be based on maternal history and clinical judgment. In a young child, the possibility of sexual abuse should be considered as a cause of acquired rather than congenital syphilis, depending on the clinical picture. For reporting purposes, congenital syphilis includes cases of congenitally acquired syphilis among infants and children as well as syphilitic stillbirths.