

Congenital Syphilis

Revision Dates

Case Definition	July 2012
Reporting Requirements	July 2012
Remainder of the Guideline (i.e., Etiology to References sections inclusive)	July 2012

Case Definition

Confirmed Case

Early Congenital Syphilis (within 2 years of birth)

Laboratory confirmation of infection:

- Identification of *Treponema pallidum* by dark-field microscopy, fluorescent antibody or equivalent examination of material from nasal discharges, skin lesions, placenta, umbilical cord or autopsy material of a neonate (up to and including 28 days old)
OR
- Reactive serology (non-treponemal and treponemal) from venous blood (not cord blood) in an infant/child with clinical, laboratory or radiographic evidence of congenital syphilis^[1], whose mother is without documented evidence of adequate treatment
OR
- Detection of *T. pallidum* nucleic acid (e.g., PCR) in an appropriate clinical specimen.

Syphilitic Stillbirth

- A fetal death that occurs after 20 weeks gestation where the mother had untreated or inadequately treated syphilis at delivery;
AND
- Laboratory confirmation of infection (ie: detection of *T. pallidum* DNA in an appropriate clinical specimen, fluorescent antibody or equivalent examination of material from placenta, umbilical cord or autopsy material).

Probable Case

Early Congenital Syphilis (within 2 years of birth)

- Reactive serology (non-treponemal and treponemal) from venous blood (not cord blood) in an infant/child without clinical, nor laboratory, nor radiographic evidence of congenital syphilis who's mother had untreated or inadequately treated syphilis at delivery.

Syphilitic Stillbirth

- A fetal death that occurs after 20 weeks gestation where the mother had untreated or inadequately treated infectious syphilis at delivery with no other cause of stillbirth established.

^[1] Includes any evidence of congenital syphilis on physical examination (e.g. hepatosplenomegaly), evidence of congenital syphilis on radiographs of long bones, a reactive CSF VDRL, an elevated CSF cell count or protein without other cause.

Reporting Requirements

1. Physicians/Health Practitioners and others

- Physicians, nurses, nurse practitioners, midwives, persons in charge of an institution, or operators of a supportive living accommodation as listed in Sections 22(1) or 22(2) of the *Public Health Act* shall notify the Medical Officer of Health (MOH) (or designate) of all confirmed, probable cases of congenital syphilis by the Fastest Means Possible (FMP) i.e., direct voice communication.
- For out-of-zone, out-of-province and out-of-country reports, the following information should be forwarded to the CMOH (or designate) by phone, fax or electronic transfer within 48 hours (two days) including:
 - name,
 - date of birth,
 - current health care number,
 - current address of residence and phone number,
 - attending physician (locally and out-of-province),
 - positive laboratory report (faxed) and
 - date of exposure.

2. Laboratories

- Section 23(b) of the *Public Health Act* (1) requires that all laboratories, including the Provincial Laboratory for Public Health (ProvLab), shall report all positive laboratory results by mail, fax or electronic transfer within 48 hours (two days) to the:
 - CMOH (or designate), and
 - Attending/ordering physician or health practitioner.

3. Alberta Health Services

- The Medical Officer of Health (MOH) (or designate) is responsible for ensuring investigation, treatment and follow-up of all reported cases.
- The MOH (or designate) of the zone where the case currently resides shall ensure that all confirmed and probable cases are reported to the CMOH (or designate), using the preliminary Notifiable Disease Report (NDR) form and Congenital Report Form, within seven days (one week) of notification and the final NDR (amendments) within two weeks of notification.

Etiology

Syphilis is caused by the spirochete *Treponema pallidum* ssp. *pallidum*. It is an extremely fragile organism, surviving for only a short period of time outside of the host. The organisms are slender, tightly coiled, unicellular and helical cells. The organism moves with a drifting, rotary, corkscrew motion and usually has a characteristic flexuose or wave-like movement about its center. This distinctive feature is used to distinguish *T. pallidum* from other treponemes. Unlike most bacteria, *T. pallidum*'s genome lacks apparent compatible elements with other bacteria, suggesting it is extremely conserved and stable. This may explain why it has remained exquisitely sensitive to penicillin for over 70 years. (2;3)

Clinical Presentation

Infections of the fetus can occur in utero at any stage of maternal infection. It is most likely to occur in early syphilis stages (from 3 – 90 days post infection). Fetal risk of infection decreases after the early stages. Infection of the fetus before the fourth month of gestation is rare; therefore early abortion is an unlikely result of syphilis. Depending on the severity of the infection, late abortion, stillbirth, neonatal death, neonatal disease or latent infection may be seen.(4-6)

In the neonate as with adults, the clinical pattern is variable. Most infected infants are asymptomatic at birth with approximately 2/3 of these infants developing symptoms by 3 – 8 weeks. Symptoms are often subtle and non-specific. The earliest sign is often rhinitis (snuffles), which is quickly followed by a diffuse, maculopapular, desquamative rash with severe sloughing of the epithelium, especially on the palms and soles, and around the mouth and anus. Almost all infected neonates exhibit symptoms by three months of age. A clinical presentation that is distinctly characteristic of congenital syphilis is necrotizing funisitis. This is an inflammatory process involving the matrix of the umbilical cord and is characterized by inflammation around and within the cord's blood vessels. The umbilical cord will appear swollen and discoloured red, white and blue, resembling a barber's pole.(7;8)

Neonatal splenomegaly, anemia, thrombocytopenia and jaundice are a result of a heavily infected liver. This infection may lead to inflammatory changes in almost any organ of the body. Neonatal death is usually a result of liver failure, severe pneumonia, hypopituitarism or pulmonary hemorrhage. Kidney complications may also develop at about four months of age.(9-11)

- Early congenital syphilis manifestations include (but are not limited to): (10)

Manifestation	Approximate Time of Onset
Necrotizing funisitis	At birth
Hematological abnormalities	At birth or delayed onset
Characteristic mucopurulent or blood-stained nasal discharge (snuffles)	first 3-8 weeks
Characteristic vesiculobullous eruptions	Birth to 8 weeks
Failure-to-thrive	Birth to 8 weeks
Generalized lymphadenopathy	Birth to 8 weeks
Hepatosplenomegaly	Birth to 8 weeks
Irritability	Birth to 8 weeks
Macular, copper-colored rash on the palms and soles	Birth to 8 weeks
Papular lesions around the nose, mouth and diaper area	Birth to 8 weeks
Petechial lesions	Birth to 8 weeks
Osteochondritis (chondroepiphysitis), especially of the long bones, ribs	May begin at birth becoming permanent eventually
Saddle nose (no bridge to nose)	Late onset (can be < 2 years)
Mulberry Molars	13 – 19 months

Diagnosis

- Diagnosis of congenital syphilis is complicated by the transplacental transfer of maternal nontreponemal and treponemal IgG antibodies to the fetus. This complicates the interpretation of reactive serologic tests for syphilis in infants.(12)
- Serology [including both treponemal (EIA) and non-treponemal testing (RPR)] from venous samples from mother and baby.(2;8)
 - Maternal history, including stage of syphilis, history of treatment, and syphilis serology results need to be considered in interpreting reactive antibodies in the neonate.
- In newborns, fluid may be taken from nasal discharge, skin lesions, placenta, umbilical cord or autopsy material. Note: Cord blood is not suitable.(7)
- Placenta, neonatal nasal discharge or skin lesions may be examined by dark-field microscopy or DFA/IFA for *T. pallidum*.(6;13)
- CSF examination should be performed in high-risk cases.(14;15)
- Long bone x-rays should be performed in high-risk cases.(14;15)

Epidemiology

Reservoir

Humans are the only known reservoir.

Transmission

Congenital syphilis infection generally occurs in utero, however infection can occur via contact with an infectious lesion during delivery. The risk of transmission in an untreated pregnant woman with primary or secondary syphilis is 70 – 100%.(4) In early latent syphilis the transmission risk is 40%, and in late latent is low but has been reported as high as 10%. Approximately 40% of pregnancies in women with infectious syphilis end in fetal demise.(8;16)

The majority of infants with congenital syphilis are infected in utero after the fourth month of gestation, but infection can occur as early as nine weeks gestation.(14)

Incubation

Incubation period is not clearly defined, as exposures happen in utero. Identification of a timeline from exposure to symptom development is difficult to determine due to the pregnancy.(17)

Period of Communicability

The infection is communicable from mother to fetus during primary, secondary and early latent stages. Sores and lesions, especially with drainage are considered infectious in the neonate. Congenitally infected newborns are generally non-infectious following at least 24 hours of adequate antibiotic therapy.(14)

Host Susceptibility

Susceptibility is universal.(7)

Occurrence

General

Congenital syphilis occurs worldwide. Literature suggests syphilis rates are rising internationally (Europe, South Africa, sub-Saharan Africa, and Canada).(13;18-20)
Consequently, the rate of congenital syphilis is on the rise as well.

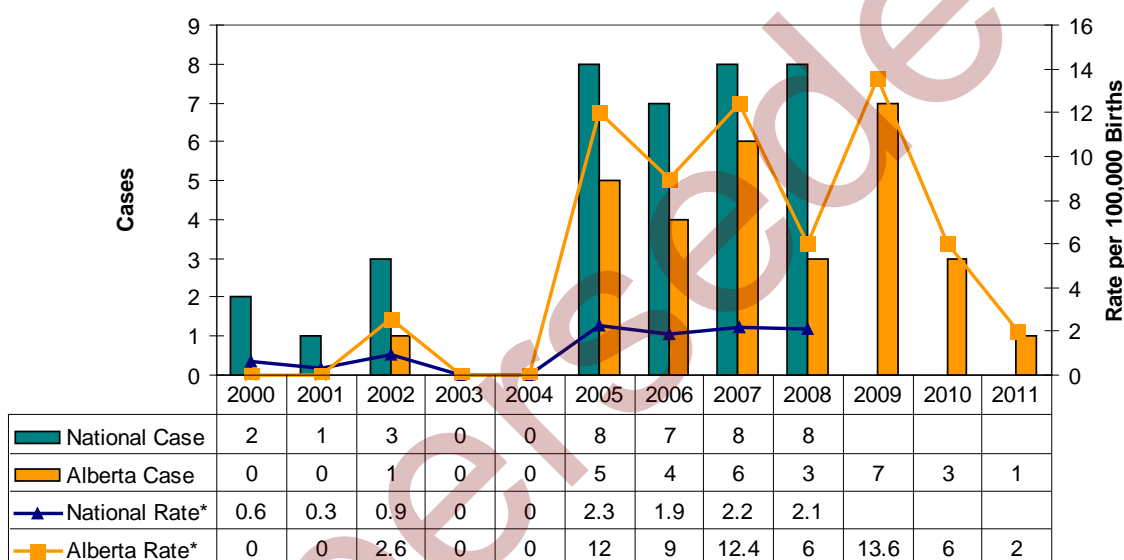
Canada

The national rates of congenital syphilis increased sharply in 2005. They have remained steady from 2005 to 2008. Data for 2009 and 2010 is not available. (See below table).(21)

Alberta

Beginning in 2005 and continuing until 2010, Alberta experienced a significant resurgence in infectious syphilis spreading to all regions of the province. In March 2007, a provincial syphilis outbreak was officially declared. The following chart contains National and Alberta specific congenital syphilis cases and rates from 2000 to 2011. (22, 23)

Congenital Syphilis Cases and Rates, 2000-2011



*2009-2011 National Case numbers and rate information not available at time of publication.

*2011 Alberta data: Source Communicable Disease Reporting System (CDRS), April 2012.

Key Investigation

Single case/cluster

- Review maternal health history. Confirm prenatal syphilis testing was completed. If unable to verify prenatal care or testing, perform testing as outlined in diagnosis section.
- If maternal history reveals positive syphilis test, ensure appropriate treatment was given. Consultation with or referral to a STI specialist, STI clinic or Pediatric Infectious Disease specialist is recommended.(13)
- Infection of the fetus may result in stillbirth or prematurity. Infants may or may not be symptomatic at birth. Signs may present within the first two years of life or later, and may include involvement of the central nervous system, bones and joints, teeth, eyes and skin (see clinical presentation section).(12;14)

Control

Management of a Case

- Treatment of choice is intravenous crystalline penicillin G for 10 days. (50,000 units/kg given every 12 hours for infants < 1 week old, every 8 hours for infants 1 – 4 weeks old, and every 6 hours for infants > 4 weeks old).(14;16)

- Refer to: Congenital syphilis: No longer of historical interest by Canadian Paediatric Society.
<http://www.cps.ca/english/statements/ID/CongenitalSyphilis.htm>

Treatment of a Case

Management of Infants born to women with reactive treponemal test (TTs) during pregnancy.

Scenario	Baseline and monthly assessment for signs or symptoms of congenital syphilis for the first three months	Syphilis serological tests (RPR and TT) with clinical assessment each time	Long-bone radiographs, complete blood cell count and differential, and sampling of CSF for cell count and differential, glucose, protein and VDRL, with a low threshold for doing ophthalmologic and audiologic assessments	Treatment for congenital syphilis
Mother has a well documented history of adequate treatment of any stage of syphilis before pregnancy, with no rise in her RPR titre during the pregnancy and no known risk factors for re-infection	No	No	No	No
Mother was treated for primary, secondary or early latent syphilis during pregnancy more than four weeks before delivery, with adequate fall in her RPR titres and no evidence of relapse or re-infection.	Yes	0,3,6 and 18 months	No	No
Mother was treated for late latent syphilis anytime during or following pregnancy	No	0,3,6 and 18 months	No	No
Mother had untreated primary or secondary syphilis during pregnancy, treponemes are detected on direct examination of specimens from infant, infant's RPR titre is fourfold or greater (higher than the mother's at birth), or there is a fourfold rise in the infant titre, OR child has any findings compatible with congenital syphilis at any age, OR infant has a reactive RPR (and TT) at 12 months of age or a reactive TT (confirmed with a second type of TT) at 18 months of age	Yes	0,3,6 and 18 months	Yes	Yes
Mother was treated for primary, secondary, or early latent syphilis within four weeks before delivery, or was treated with an antibiotic other than penicillin, OR mother was treated for primary, secondary or early latent syphilis before or during the pregnancy and her RPR titre did not show the expected decline or inadequate time has passed to assess the decline	Yes	If treated for congenital syphilis, do at 0,3, 6 and 18 months of age; If not treated, also do at 1, 2, and 12 months of age	Yes	Usually
Mother was treated for primary, secondary or early latent syphilis before pregnancy, but there are doubts about the adequacy and her follow-up RPR was not obtained OR mother was treated for any type of syphilis during pregnancy but long-term infant follow-up cannot be assured	Yes	If treated for congenital syphilis, do at 0,3, 6 and 18 months of age; If not treated, also do at 1, 2, and 12 months of age	Depends on risk, but mandatory if mother had primary, secondary, or early latent syphilis and follow-up is not likely to occur, or if clinical or serological findings are abnormal.	Depends on risk and on results of assessments
Infant has a reactive RPR (and TT) at six months of age	NA	Depends on timing of last serology	Yes	Usually

Reference: Congenital syphilis: No longer of historical interest. Used with Permission (14)
<http://www.cps.ca/english/statements/ID/CongenitalSyphilis.htm>

Follow-up

- **Consultation with or referral to a STI specialist, STI clinic or Pediatric Infectious Disease specialist is recommended.**
- For pregnant women with reactive syphilis serology and infants born to mothers with reactive serology, follow up will depend on maternal and neonatal history; advice should be sought from CMOH (or designate) or STI expert.

Screening

- All pregnant women should be screened for syphilis during pregnancy. Screening should be performed: (14)
 - in the first trimester and at time of delivery for **all** pregnant women, and
 - more frequently for women at high risk of acquiring syphilis. This includes women:
 - who have had contact with a known case of syphilis,
 - who are sex trade workers,
 - who are street involved/homeless,
 - who are injection drug users,
 - of Aboriginal ethnicity,
 - with multiple sexual partners,
 - with a history of syphilis,
 - with HIV and other STI,
 - originating from or having sex with an individual from a country with a high prevalence of syphilis,
 - living in areas experiencing outbreaks of heterosexual syphilis,
 - with sexual partners of any of individuals with the preceding characteristics and
 - victims of sexual assault.

Preventive Measures

- Ensure appropriate treatment of syphilis for all cases.
- Make prenatal services culturally appropriate, readily accessible and acceptable, regardless of economic status.
- Educate the parent about symptoms, transmission and prevention of syphilis infection.
- Ensure testing is offered to all pregnant women.

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