Alberta Public Health Disease Management Guidelines

Congenital Syphilis

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Health and Wellness Promotion Branch

Public Health and Compliance Branch

Alberta Health

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Case Definition

Confirmed Case

Early Congenital Syphilis (Within Two Years of Birth)

Laboratory confirmation of infection:

- Identification of *Treponema pallidum* by dark-field microscopy, fluorescent antibody or equivalent examination[(A)] 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[(B)], whose mother is without documented evidence of adequate treatment for her syphilis infection;

OR

- Molecular detection of *T. pallidum* nucleic acid (e.g., NAAT) in an appropriate clinical specimen.[(C)]

Syphilitic Stillbirth

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

AND

- Laboratory confirmation of infection (i.e., 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 Two 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 whose mother had untreated or inadequately treated syphilis at delivery.

Syphilitic Stillbirth

- A fetal death that occurs after 20 weeks gestation or in which the fetus weighs greater than 500 g where the mother had untreated or inadequately treated infectious syphilis at delivery with no other cause of stillbirth established.

[(A)] In Alberta, diagnosis by this method is exceedingly rare and only available in specialized clinics.
[(B)] 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 counts or protein without other causes.
[(C)] Guidance and direction on lab specimen collection can be found at Alberta Precision Laboratories.

Reporting Requirements

Physicians, Health Practitioners and Others

Note: This includes First Nations and Inuit Health Branch

- Physicians, health practitioners and others shall notify the STI Medical Director[(D)] via Sexually Transmitted Infection Centralized Services (STICS) of all confirmed and probable cases within 48 hours (two business days).

Laboratories

- All laboratories shall report all positive laboratory results by mail, fax or electronic transfer within 48 hours (two business days) to the STI Medical director via STICS and the Chief Medical Officer of Health (CMOH) (or designate).

Alberta Health Services: STICS

Contact Information: **Toll free: 1-855-945-6700 option 4**
Fax: 780-670-3624

The STI Medical Director/STICS are responsible for ensuring investigation and follow-up of all reported confirmed and probable cases and contacts.

- The STI Medical Director/STICS shall forward the initial *Perinatally Acquired Notifiable Disease Enhanced Report Form* of all confirmed and probable cases to the CMOH (or designate) within two weeks of notification and the final Notification of STI form within four weeks or at confirmation of diagnosis staging.
- For out-of-province and out-of-country case reports, the following information (when available) should be forwarded to the CMOH (or designate) by phone, fax or electronic transfer as soon as possible:
  - name,
  - date of birth,
  - out-of-province health care number,
  - out-of-province address and phone number,
  - positive laboratory report, and
  - other relevant clinical / epidemiological information.
- For out-of-province and out-of-country contacts the following information (when available) should be forwarded to the CMOH (or designate) as soon as possible:
  - name,
  - date of birth,
  - date of exposure, and

[(D)] The STI Medical Director is the Provincial Medical Director of Alberta Health Services' Sexually Transmitted Infection Centralized Services (STICS) and is also a Medical Officer of Health.

- out-of-province / country contact information.

Additional Reporting Requirements

In all cases, where a person under 18 is suspected or confirmed to have an STI, an assessment should be carried out by the clinician to determine if additional reporting is required.

To Alberta Child and Family Services

- The clinician should determine whether there are reasonable and probable grounds to believe that they are in contact with "a child in need of intervention" [as per Section 1 of the *Child, Youth and Family Enhancement Act (CYFEA)*] and shall report to a director pursuant to Section 4 of the *CYFEA*.[1]
- Reporting is done by contacting the local Child and Family Services office or calling the **CHILD ABUSE HOTLINE: 1-800-387-5437 (KIDS)**. For local office contact information see: www.humanservices.alberta.ca/services-near-you/15010.html.

To Law Enforcement Agency

- Consent is a key factor in determining whether any form of sexual activity is a criminal offence. Children under 12 do not have the legal capacity to consent to any form of sexual activity. The law recognizes that the age of consent for sexual activity is 16. However, the law identifies the exception for minors between 12 and 16 years as having the ability to consent, in "close in age" or "peer group" situations.[2]
- Reporting is done by contacting your local City Police Detachment or RCMP Detachment at: www.rcmp-grc.gc.ca/detach/en/find/AB.

For additional information:

- Alberta Child, Youth and Family Enhancement Act at: https://open.alberta.ca/publications/c12
- Age of Consent to Sexual Activity at: www.justice.gc.ca/eng/rp-pr/other-autre/clp/faq.html
- Criminal Code of Canada at www.laws-lois.justice.gc.ca/eng/acts/C-46/ [3]

Epidemiology

Etiology

Syphilis is caused by the spirochete *Treponema pallidum* ssp. pallidum, an extremely fragile organism that can survive for only a short period of time outside of the host. 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.[4]

Clinical Presentation

Infections of the fetus can occur in utero at any stage of maternal infection. It is most likely to occur in early infectious syphilis stages. Pregnant women with untreated syphilis have the highest risk of transmission to the fetus. Depending on the severity of the infection, miscarriage, stillbirth, neonatal death, neonatal disease or latent infection may be seen.[5–8]

At delivery, the umbilical cord can appear swollen and discoloured red, white and blue, resembling a barber's pole due to necrotizing funisitis, an inflammatory process involving the matrix of the umbilical cord that results in inflammation around and within the cord's blood vessels. Further, the placenta is generally larger, thick and pale.

In the neonate, as with adults, the clinical pattern is variable. Most infected infants are asymptomatic at birth with approximately two thirds 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.[7,9–12]

An infected infant, without adequate timely treatment during pregnancy and post-natally, can go on to develop a spectrum of manifestations.

Early congenital syphilis manifestations include (but are not limited to):[13]

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
Mulberry Molars	13–19 months

- Late congenital manifestations are rare in developed countries. However, they do persist in untreated survivors and are the corollaries of earlier lesions. These can include: [10,14]

- saddle-nose deformity,
- eighth nerve deafness,
- saber shins,
- rhagades,
- hutchinson teeth, and
- keratitis.

Diagnosis

The transplacental transfer of maternal nontreponemal and treponemal IgG antibodies to the fetus complicates diagnosis of congenital syphilis. This obscures the interpretation of reactive serologic tests for syphilis in infants. Serology [including both treponemal (EIA) and non-treponemal testing (RPR)] should be taken from both mother and baby. 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, clinical specimens may be taken from nasal discharge, skin lesions, placenta, umbilical cord or autopsy material. Note: Cord blood is not suitable. CSF examination and long bone x-rays should be performed in high-risk cases.[4,7,9,15,16]

Treatment

- Collaborative management should occur between STICS and a Pediatric Infectious Disease specialist.[15,17]
- Refer to Appendix 1: Management of Infants Born to Pregnant Women with Syphilis Reactive Results During Pregnancy

Reservoir

Humans are the only known reservoir.[4,12]

Transmission

Congenital syphilis infection primarily 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%. In early latent syphilis, the transmission risk is 40%, and in late latent is low but has been reported as high as 10%. Approximately
30–40% of pregnancies in women with infectious syphilis end in fetal demise. 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.[5,9–13]

Incubation Period

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.[18]

Period of Communicability

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.[9,10]

Host Susceptibility

Susceptibility is universal.[7]

Incidence

In Alberta the congenital syphilis case counts and incidence rate reflect the rise of infectious syphilis diagnoses.[19] This parallels the rising rates experienced nationally and internationally as well.[20] Provincial outbreaks have been declared for Alberta in 2007 and again in 2019.

See the Interactive Health Data Application and the most recent Alberta STI Report for more information.

Public Health Management

Key Investigation

For all deliveries:

- Review maternal health history,
- Review prenatal syphilis testing status, and
- Repeat testing at delivery per *Alberta Prenatal Screening Program for Select Communicable Diseases*.

If maternal history reveals positive syphilis test, ensure appropriate treatment was given. Consult with STICS for maternal treatment history and guidance.[21]

Management of Contacts

- Interview mother, identify and ensure appropriate treatment and follow-up of disease for self and sexual partner(s).
- For any stillbirth occurring after 20 weeks or if fetus is greater than 500 g, maternal testing and investigation is recommended.

Preventative Measures

- Ensure appropriate treatment of syphilis for cases and contacts.
- Make prenatal services culturally appropriate, readily accessible and acceptable.
- Educate the parent about symptoms, transmission and prevention of syphilis infection and other STIs.
- Ensure screening for all pregnant women.
- Ensure testing is available for their partners.

Screening

- All pregnant women should be screened for syphilis during pregnancy as per the *Alberta Prenatal Screening Program for Select Communicable Diseases*.

Appendix 1: Management of Infants Born to Pregnant Women with Syphilis Reactive Results 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

Appendix 2: Revision History

Revision Date	Document Section	Description of Revision
September 2021	Complete Document	• Updated references where available
	Case Definition	• Updated testing to reflect current lab practice in Alberta
	Reporting Requirements	• Adjustment of reporting structure to reflect AHS STICs role
	Epidemiology	• Link to IHDA added
	Treatment of Case	• Removal of specific treatment recommendations and indication of appropriate consultation needed
	Screening	• Link to Alberta Prenatal Screening Program for Select Communicable Diseases
	General	• Diagnosis and Treatment sections moved to Epidemiology • Updated weblinks

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