SYPHILIS (venereal)[1]

Outline:

I. Introduction
II. Clinical
  a. General
  b. Primary
  c. Secondary
  d. Tertiary
  e. During pregnancy
  f. Congenital
  g. Syphilis + HIV
III. Similar Diseases
IV. Treatment

I. Introduction

Cause: Spirochete.
Synonyms: Lues, Treponematosis.
Regional Notes: E, F, I, M, O, R, S, U
Definition: Syphilis is an infection with the spirochete Treponema pallidum.

Includes: Venereal syphilis only, in this entry.

Not ill to very ill; Class 1-3; Contagious; Worldwide and very common in the tropics.

Note: Venereal syphilis is sexually transmitted; endemic syphilis is transmitted non-sexually. There are three kinds of endemic syphilis: PINTA (Americas only; see M Index), TREPONARID (arid areas only; see this Index and *Regional Indices*), and YAWS (humid areas only; see this *Index* and *Regional Indices*). Venereal syphilis, PINTA, TREPONARID, and YAWS are all closely related diseases; each gives partial immunity to the others. SYPHILIS (i.e., venereal syphilis) is described here; see the separate listings for the three kinds of endemic syphilis.

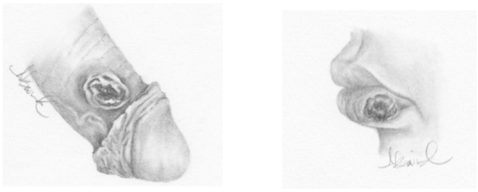
Age: Adults; sexually abused children; babies of infected mothers. **Who:** Those who have had a sexual relationship outside of faithful marriage; partners with an unfaithful spouse; children born to infected mothers. **Onset in adults:** *Incubation:* 9-90 days, from infecting contact until the primary ulcer. *Incubation* 2 weeks to 6 months until symptoms of secondary syphilis. Usually there are no symptoms at all for the next two years; *Incubation* 1-20 years until symptomatic tertiary syphilis. **Onset in newborns:** *Incubation* to primary and secondary, up to 2 years. *Incubation* to tertiary, a few months to 20 years. **Onset with HIV INFECTION:** the symptoms of primary and secondary syphilis may occur at the same time, and progression of the disease to tertiary may be much faster. The latent period between secondary and tertiary may be abolished; the patient may have symptoms of secondary and tertiary at the same time.

[1] See the PowerPoint lecture on the DVD entitled Syphilis.

II. Clinical

- **Primary Syphilis:**

This starts with a (usually) single, round or oval ulcer on the genital area, 0.3-3 cm in diameter; it is relatively painless (but possibly tender) and it is swollen on the rim and underneath. The presence of swelling beneath is most specific. With some sexual practices the mouth, buttocks, anus, or fingers might have the primary chancre which is usually painful but not swollen underneath. Usually there is only one ulcer unless the patient also has HIV INFECTION. Usually there is no visible pus; the ulcer looks clean, and it does not bleed readily. It persists for at least 2 weeks before starting to heal. Painless enlargement of the groin (or the neck) lymph nodes starts a week later, usually on both sides for genital ulcers, possibly one side elsewhere. The enlarged nodes are firm, and they are not stuck to the flesh around them. The skin over top has a normal appearance. They persist for weeks or months after the primary ulcer heals.



A syphilis chancre is usually roughly round or oval, with swelling underneath it. On the genitals, it is usually non-tender to touch. On other sites with sexual contact appearance and tenderness vary.

- **Secondary Syphilis:**

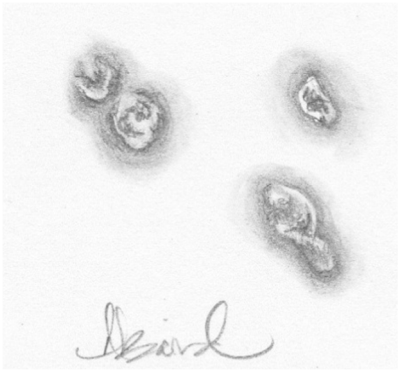
The healing primary or the scar thereof may still be present. This develops about 6 weeks after primary syphilis, but it may develop at the same time in patients who are HIV infected. Secondary syphilis may occur without the ulcer of primary syphilis, or primary syphilis without the symptoms of secondary. In the latter case, primary syphilis advances directly to tertiary. The secondary stage happens, but the patient is not aware of it.

These are the most common scenarios:

- On fair or light brown skin, a symmetrical, red-or brown- spotted rash (which may be subtle) develops mainly on the trunk and face;

it may also include the limbs, palms, and soles[2]. Most often the spots can be seen but not felt; they may be hard to see. There may be few or many. It is usually not itchy in Whites but it may itch and appear dark-color or white in those with dark skin. The rash, if it occurs, arises over a week or two, not hours or days. The patient is not ill to mildly ill. These are some appearances

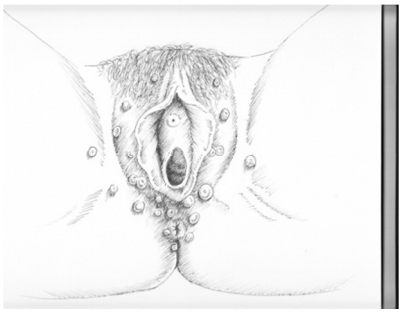
- A measles-type rash, few spots or none on the face.
- Circles, irregular shapes, or targets, sometimes with rims, usually in groups rather than single; common in non-Whites.
- Like acne, but more bumps than scars. With acne there are more scars than bumps.



These are magnified spots of a secondary syphilis rash. They are irregular shapes with rims. The size and appearance may vary.

- The patient may have fever, headache, enlarged and tender lymph nodes, weight loss, loss of appetite, and fatigue. Usually the headache is prominent. The enlarged nodes are those near the primary ulcer plus the nodes by the elbow(s) and the back of the neck.
- The patient may have slightly raised, round, relatively painless, white patches on moist, pink surfaces of the mouth and genitals, silver-gray in color, with red halos. These may ulcerate. Usually they are toward the front of the mouth and on one side only[3]. He may have splits in the skin by the side of his mouth, long grooves or an ulcer on his tongue. He may have bleeding gums so the condition resembles SCURVY.
- He may have flat-topped, pale, moist "warts" near the site of the primary ulcer[4], sometimes elsewhere, usually on warm, moist body parts. Genital warts are common.The warts are flat, without stems. These are common in females.

This is the appearance of a single syphilis wart



This gives an idea of the appearance and distribution of syphilis warts in a female.

- Patches of hair on his head or eslewhere may fall out. The scalp has a moth-eaten appearance. The affected areas have fine hairs in them; the patient is not totally bald in these areas. There is no visible scarring. There is no predilection for one part of the scalp or another.
- Secondary syphilis may cause spontaneous ABORTION. Rarely there is HEPATITIS, IRITIS, MENINGITIS.

➢ **Tertiary Syphilis:**

This may be totally without symptoms. When there are symptoms, they occur 5-30 years later in about 2/3 of those who have had primary or secondary syphilis. In tertiary syphilis, 2/3 of the patients have no history of primary or secondary disease. This may affect many different parts of the body, as follows, listed from most common to least common:

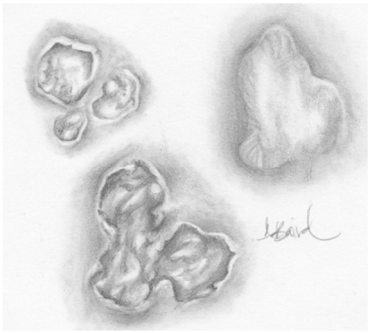
- **Skin**: A patient may have lumps under his skin. The lumps that form may break open to

[2] Sole spots in the absence of palm spots are normal in persons of African genetic heritage; they do not indicate syphilis or any other disease. Other diseases may also cause a rash that affects the palms and soles, but in these cases the patient is always very ill. With secondary syphilis, the patient is not very ill.

[3] Thus they can be distinguished from similar patches due to DIPHTHERIA which usually occur on both sides and tend to be toward the back of the mouth.

[4] Not all genital warts are due to syphilis. Some are viral. Warts due to syphilis are usually moist and they have a broad rather than narrow base. The majority of the warts are single or in small groups, not 10 and more together like viral warts.

become ulcers resembling TROPICAL ULCERS which heal very slowly if at all.



This starts out with swelling below. Then a group of spontaneous ulcers form as in the upper left. The ulcers coalesce to make a large scalloped ulcer, like the lower left. Then the ulcer heals, leaving wrinkled, thin scar tissue in an area with scalloped edges, the upper right. The whole process is totally painless.

- **Limbs:** Slow-growing, painful lumps may form in the bones. These are initially tender but not so tender later on. The shin bone is most commonly affected, leading to saber shins5.
- **The large joints** may be swollen symmetrically (usually) and move abnormally. Sometimes a joint may be grossly swollen and deformed but relatively painless. There may be skin changes with this. There may be lightning pains from trunk to limb.
- **Heart:** There may be an irregular pulse, especially with lying down, and ANGINA (chest pain) or HEART FAILURE. His pulse pressure may be high. He may be dizzy because of this. He will not gladly tolerate exercise.
- **Head/Brain:** There is a stuttering or slow onset, with episodes of improvement even without treatment. It may cause STROKE, SEIZURES; paralysis (usually stiff); leaking stool or urine; sharp, shooting pains in limbs; insanity; impotence; double vision; blindness; unequal pupils; or uncoordination. His gait will be abnormal. Uncoordination is always worse in the dark; the patient cannot tell where his limbs are without looking at them. He will probably have trembling, poor handwriting, slurred speech, and pupils which change size with focusing but not with light shone in them. He finally becomes bedridden and dies with BRAIN DAMAGE.
- **Mental symptoms:** If the patient becomes insane, this usually begins at age 35-50. It starts with headache and insomnia, trouble concentrating, and easy fatigue. Then his personality changes, becoming either stupid or grandiose. He may have day-night reversal. Usually he is unaware of his poor functioning.
- **Eyes:** He may not be able to fully open his eyes. He may have trouble with eye movements so he sees double or has crossed eyes. He may have unequal pupils, KERATITIS or IRITIS, see flashes of light, and have distorted vision. This is particularly common in the presence of HIV INFECTION.
- **Mouth/Nose:** A painless bump may form on the tongue in the midline. This may break down to become an ulcer There may be longitudinal cracks in the tongue or holes in the roof of the mouth. It may destroy the center divider of the nose so the bridge of the nose is sunken..
- **Pelvic area:** Painless lumps may form in the testicles. The bladder might not empty all the way so the patient retains urine. This is likely to become infected, leading to KIDNEY FAILURE. He is likely to have impotence and a leaky bladder.
- **Abdomen:** There are commonly recurrent, severe abdominal pains or episodes of vomiting or both, extremely exhausting. They come on suddenly, last days, and then suddenly stop, leaving the patient exhausted.

Syphilis During Pregnancy:

Syphilis during pregnancy causes miscarriages during the last 6 months as well as stillbirths. Syphilis acquired early in pregnancy causes either miscarriage or else symptoms of tertiary syphilis at birth. Syphilis acquired late in pregnancy, causes symptoms of primary or secondary syphilis at or shortly after birth. If there is latent syphilis (without symptoms, between secondary and tertiary syphilis), the woman has a 70% chance of having an infected baby.

Congenital Syphilis[6]:

Most symptoms occur after 4 months of age and many after years. The early and late-onset symptoms are listed separately here, but in real life there is no definite dichotomy.

- **In infants, birth to 2 years:**
- The placenta is abnormally large.
- The baby may fail to gain weight; he may not nurse vigorously.
- His liver and spleen may be large

[5] With saber shins, the middle 1/3 of the shin is thicker in the front-back direction than the upper and lower thirds. Hence the shin bows forward so its shape resembles a saber.

[6] Be sure to see the illustrations of this condition on the DVD that goes with this volume.

- He may have swollen fingers, like sausages, usually sparing the end joints. Other causes of sausage fingers involve the end joints.
- He may have a continually runny nose, possibly with blood, as if he had a cold that would not go away. The lips tend to develop splits.
- The soft spot on the head will stay open for a long time, and there is delayed eruption of the baby teeth (similar to HIV infection and RICKETS).
- He develops shiny, red, possibly peeling skin on his palms and soles, white patches in his mouth, hoarseness, splits in his skin, or skin bumps that look like warts. The skin changes tend to be on the mouth, palms, soles, and genitals. They may be on the whole body. They may be red spots (as shown below), blisters, or both red spots and blisters together.
- His limbs become tender or swollen or both. They may be fragile, fracturing with slight movement.
- He may appear like a tiny, wrinkled, old man.

Summary of diagnosis of congenital syphilis in a newborn, in the absence of laboratory support:

The patient should have either two major criteria or one major and one minor criterion.

Major criteria:

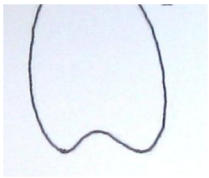
A. An enlarged liver, with or without an enlarged spleen.
B. Limb problems, usually symmetrical,: swelling or the infant refuses to move them or they are tender to touch or an x-ray of the limbs is abnormal.

Minor criteria

A. A generalized rash on the skin or abnormal whitish patches within the mouth. If the rash involves the palms and soles, then this becomes a major criterion.
B. A runny nose, like a cold that will not go away.
C. Anemia: either paleness inside the lower eyelid or the tongue, or a drop of blood on a piece of paper is paler than normal.
D. Bilirubin in the urine by dipstick. An alternative is that if you shake the patient's urine in a clear glass bottle, you can see yellowish foam on top.

➢ **Symptoms developing later in life (2-25 years old)—some or all of the following:**

- Clouding of the cornea with redness around, pain, and tearing.
- There may be a delay in eruption of the first teeth and susceptibility to decay. Upper front permanent teeth have notched cutting edges[7] and are smaller than normal. This may be on one or both sides.



The abnormal shape of a tooth due to congenital syphilis consists in its having a single, large notch. Multiple tiny notches are normal. The sides of the syphilic tooth are tapered in, from the cutting edge to the gum line.

- Sunken bridge of the nose with small cheek bones.
- Deafness; sudden or gradual onset, usually ages 5-25, commonly with ringing in the ears or dizziness or both beforehand.
- Changes in bones: one or more of the following.
  - Shins that are thickened in the middle third; the front is sharp and bowed forward. OR
  - Collar bones that are thickened in their entirety or else only the joint near the breast bone. This may be one or both sides. OR
  - Symmetrically swollen large joints, particularly the knees.
- Any symptom of tertiary syphilis such as paralysis, uncoordination, and insanity.

If there are symptoms at birth, the prognosis is poor.

Syphilis and HIV:

- There may be multiple primary ulcers.
- The rash of secondary syphilis may be asymmetrical.
- There is usually no latent period; syphilis jumps directly from secondary to tertiary.

[7] Multiple notches are normal. Syphilis causes a single notch. Also, the sides of the teeth taper inward so they are shaped like pegs. Normal teeth have perpendicular sides or have a wider cutting edge. .

- Relapses are common and genital warts are common with relapses.

III. Similar Conditions:

Very many, too many to list all. See the relevant protocols in the differential diagnosis section.

Primary syphilis is similar to other SEXUALLY TRANSMITTED DISEASE's. Its distinguishing characteristics are that there is usually only one ulcer, there is swelling beneath the ulcer, and it is both round or oval and painless. The lymph nodes of syphilis are distinct (not stuck together), and they do not break down and drain.

Secondary syphilis See Protocol B-2. Syphilis can be confused with INFLUENZA or MEASLES or any other illness with a fever, but the rash of syphilis develops slowly, over weeks. Syphilic warts do not resemble ordinary WARTS since they have moist, flat, light-colored surfaces. They differ from the wart-like skin bumps of DONOVANOSIS in that they are skin color or pale whereas those caused by DONOVANOSIS are red. The warts of syphilis clear within a week with PENICILLIN but those of DONOVANOSIS take more than two weeks with any antibiotic.

Tertiary syphilis affecting the heart may be similar to HEART FAILURE due to bad valves. BERI-BERI causes leg weakness also, but the trouble walking is no worse in the dark and the pains of BERI-BERI are more burning. The lightning pains are similar to SHINGLES. The bone problems can be similar to the other diseases listed in Protocol B-6. Some MALNUTRITION can be nearly identical but it is reversed (slowly) with MULTIVITAMINS. Tertiary syphilis is one of many causes of BRAIN DAMAGE. See Protocol B-10 B: Confusion and/or Lethargy.

Congenital syphilis: RICKETS, SCURVY, HIV INFECTION, THALLASEMIA.

Higher-Level Care. Speedy referral for syphilis in a newborn and tertiary syphilis is very desirable. See Volume I, Appendix 13.

Laboratory: Fairly simple blood tests are available. These tests (RPR and VDRL) are not sensitive in primary and tertiary syphilis, but they are sensitive in secondary syphilis. The primary ulcer must be present for at least a week before the test turns positive. There are occasional false-positives, usually in the presence of old age or some other diseases, most notably LEPROSY and some kinds of ARTHRITIS. More reliable tests are more expensive and less available. The blood tests for syphilis should become negative a year after treatment for primary syphilis, two years after treatment for secondary syphilis, and five years after treatment for tertiary syphilis. They may remain positive in the presence of HIV.

Since venereal syphilis and the various forms of endemic syphilis (PINTA, YAWS, TREPONARID) are all closely related, all laboratory tests will be positive regardless of which form (venereal or endemic) of the disease the patient has. This should be kept in mind when missionary parents are confronted with a question of childhood sexual abuse. **A positive blood test for syphilis can readily be acquired non-sexually in the tropics**.

Practitioner: An abstinence-based, sexually transmitted disease clinic is ideal for primary and secondary. A pediatrician should treat a newborn. An infectious disease specialist should manage tertiary syphilis.

IV. Treatment

Prevention: Find and treat sexual contacts. Wash your hands well. Wear gloves while doing physical exams on high-risk patients.

Patient Care: Unless you are very sure of the diagnosis, treat according to Protocol B-1 rather than for syphilis alone. PENICILLIN is still the drug of choice for this. Do not use AZITHROMYCIN.

Caution: With the first dose of PENICILLIN the patient will probably have a DRUG ERUPTION. Do not stop the medication for that.

An hour before the first dose, give one dose of DIPHENHYDRAMINE, if you have it.. Then give PENICILLIN. Syphilis requires continual treatment without missing doses. Many of the effects of tertiary SYPHILIS will not change in spite of treatment, but treatment may prevent their worsening. DOXYCYCLINE, ERYTHROMYCIN,[8] AZITHROMYCIN,[9] CEPHALOSPORIN or CHLORAMPHENICOL in usual doses are also effective. Because of problems with keeping people on medicine, WHO recommends benzathine PENICILLIN; one injection replaces ten days of drug by mouth. However, benzathine PENICILLIN does not adequately treat syphilis that has affected the brain. Infants born with the disease should have weekly benzathine PENICILLIN injections for 4 weeks.

[8] Don't use the ERYTHROMYCIN estolate; any other type of erythromycin is fine.

[9] Reportedly this works most of the time but there is some resistance to it.