

# ***Congenital syphilis***

- Congenital syphilis can be passed on from an infected mother to her unborn child during intrauterine life through placenta by *spirochaeta pallida* (when the spirochete in the mother's body simply crosses over placental barriers to enter the infant's body).

- **Intrauterine infection develops at the end of the fifth month of gestation when placenta starts functioning.**
- **The invasion of spirochetes into the fetus over placenta, umbilical vein, lymphatic split occurs due to placenta injury by treponema toxins.**

- Specific septicemia develops and treponemae penetrate into different organs of the fetus (liver, spleen, adrenal glands) and associated with abscess formation. Intrauterine death and miscarriage occur as a result of metabolic disorder and malnutrition of the fetus.
- Especially severe changes are revealed in the lungs in the form of so-called "**white pneumonia**".

- Affected pulmonary tissue is airless, of white-greyish colour because of infiltration of alveolar septa and desquamation of alveolar epithelium.
- **"White pneumonia"** usually results either in intrauterine death or infant's death after several inspirations (sometimes infant lives for some days).

- **As stated above immature and still birth of macerative fetus on the sixth-seventh month of gestation is typical for untreated pregnant women.**
- **Another outcome is the birth of a sick infant, or the birth of a healthy one, or miscarriage.**

- **In untreated pregnant women with secondary syphilis (especially during first three years of illness) infection occurs in 100% of cases; less commonly in patients in the late stage of the disease (30%); and almost never in patients with primary syphilis.**

# Classification

- Depending on clinical symptoms and terms of manifestation, the following stages of congenital syphilis are singled out:
- Early congenital syphilis. Any congenital manifestations of syphilis in infants under 2 years of age belong to this stage.

- **-Early congenital latent syphilis.**  
Absence of clinical manifestations is typical; serological reactions of blood and cerebrospinal fluid in infants under 2 are positive.



- - Late congenital Syphilis. Any congenital syphilitic conditions which develop in two or more years after birth.
- - Late congenital Syphilis latent. Without any clinical symptoms; this stage is characterized by positive serological reaction and normal fluid composition.

## Early congenital syphilis

- In the majority of cases congenital syphilis is manifested clinically from the first three months after birth.
- Moreover, the earlier syphilitic lesions develop, the worse prognosis is and the higher the risk of lethality is.
- Infants with a severe form of syphilis look like "little old people".

- **The symptoms are as follows: vivid hypotrophy; skin of dirty yellow color with practically absolute lack of subcutaneous fatty tissue resulting from decubitus [bed sores] formation.**
- **Arms and legs resemble sticks covered with skin.**

- Such infants cannot put on weight properly, they cry constantly.
- Sometimes at night they give a high-pitch cry (**Sisto's symptom**).
- Left untreated and without proper care such infants usually die.

- Injury of the skin and mucosa are specific and never occur in acquired syphilis.
- 1) Pemphigus syphiliticus. Early changes of the skin develop at birth or appear on the first days of life and occur in 5-20% of infants with congenital syphilis.

- Eruptions in the form of pea-sized or cherry-sized blisters are filled with serous or serous purulent exudate.
- As a rule, they are located at palms and soles, less commonly at flexor surfaces of extremities and trunk.

- **2) Diffuse papular Hochsinger's infiltration** is a pathognomonic symptom of the syphilis in infancy.

It usually develops on the eighth - tenth weeks after birth in the form of diffuse infiltration and erythema on palms, sole, face, scalp, thigh. The skin becomes smooth, glittering, bright red, as if varnished.

- Later on the skin becomes macerative, and skin folds typically appear ("**hands of a washerwoman**").
- In other areas of the body this process usually results in desquamation.



- But the most typical picture develops in diffuse infiltration of the skin of the face. Lips are thickened and edematous.
- Deep cracks and ulcers develop due to constant impairment from cry. Whitish radial scars around the mouth (Robertson-Fournier's symptom) remain after healing and persists through out the life.

- **3) Syphilitic rhinitis (Hochzinger's nasal cold) may be the only sign of congenital syphilis.**

**It develops during the first days of life, less commonly during the first month after birth; and it is associated with impairment of the mucous membrane of the nose, mainly of its front part.**

- **Constriction of the nasal orifice resulting from hyperplasia makes it difficult to breathe through the nose, which, in turn, impedes feeding.**

**Profuse fetid discharges with a large amount of spirochetes from the nose are typically observed.**

- If the patient does not undertake specific treatment, deep ulceration of the mucosa can spread over cartilages and bones with subsequent necrosis of the nasal membrane and sinking of the dorsum of the nose ("saddle-like nose").

## *Skeletal system affection*

It occurs in 85% of patients.

Osteochondritis develops but it is not an inflammation, it is only impairment of calcium deposit and retardation of osteoblast development.

- There are few trabeculae in the area of cartilage calcification in this stage, that is why in the case of any trauma epiphysis separates from diaphysis with the development of intraepiphyseal fracture which manifests clinically as false paralysis-like condition (4)Parrot's pseudoparalysis).

## Parrot's pseudoparalysis

- An infant gives a cry trying to move; and there are no active movements in the affected extremity. Upper extremities hang like strings, lower ones are drawn to the abdomen which resembles a contracture.

## Specific lesion of the inner organs

begins during intrauterine life.

- The liver is enlarged; its changes are associated with anemia and flaccidity.
- Both liver injury (75%) and splenomegaly (the spleen is enlarged 10 times) are typically observed.



- Convulsions and epileptic attack develop in some infants due to sharp increase of intracranial pressure.
- Hydrocephaly results in modification of the form of the head with the formation of "buttock-like skull".

# Late congenital Syphilis

## *Lesion of the inner organs*

occurs at the age of 2 till 17, but sometimes even in some decades. in case of late congenital syphilis develops less commonly than in early congenital syphilis.

- Hepatomegaly , splenomegaly , dysendocrinia (nanism, infantilism, obesity and etc.) are noted.

Either clinical manifestations may be a continuation of the processes of congenital syphilis of infancy stated above, or these symptoms develop in the state of "complete health".

- In diagnosis of late congenital syphilis one can single out true, probable signs and stigmata (atrophy).

True signs are pathognomonic and are represented by the triad of Hutchinson's symptoms:

# parenchymatous keratitis

1. develops suddenly at the age of 5-15 and may be the only symptom of late syphilis.

The patients complain of lacrimation, blepharospasm, photophobia, and further sometimes - blindness.

# **labyrinthine deafness**

2. is usually bilateral, it develops suddenly, persists consistently; it is caused by the inflammation of the labyrinth and degeneration of acoustic nerve.

Deafness is resistant to treatment.

# **Hutchinson's teeth**

3. are symptoms of dystrophy of upper front incisors.

They have barrel-shaped form or the form of screwdriver widening to the gingival edge, pointing to the cutting edge and the presence of semilunar incisure.

## Probable signs

- The presence of probable signs permits us to suspect congenital syphilis but to confirm the diagnosis additional evidence are essential, e.g. associated clinical presentations or the results of the examination of the family.



# Probable signs are as follows:

- 1- sword-like leg - consequences of osteochondritis in infancy.
  - Bones of the shin are curved outside and forwards.

- 2- radial scars - Robertson-Fournier's symptom (consequence of Hochzinger's diffuse infiltration)
- 3- "saddle-like nose" (as a result of Hochzinger's syphilitic rinitis)

4- "bump-like skull" (consequence of hydrocephaly)

5- syphilitic Hutchinson's teeth (screwdriver, "tightened" at gingival edge)

## *Stigma*

- is the manifestation of dystrophy which does not occur only in congenital syphilis.

Its presence is an alarming symptom of syphilitic infection.

- 1- Avsitidisky's symptom (thickening of thoracic edge of the right collar bone)
- 2- Olympic forehead (enlargement of frontal and parietal tubers of the skull)
- 3- high "Gothic" palatine

4- axiphoidia (absence of xiphoid process)

5- Gasher's diastema (a gap between two upper incisors )

- 6- infantile little finger (shortened, underdeveloped)
- 7- hypertrichosis of the forehead (low disposed interface of hair growth at the forehead)
- 8- assymetry of the auricles

- **Detection of several dystrophies, their association with true signs or several probable ones, with positive serologic reaction in the infant and parents is the base for making a diagnosis of late congenital syphilis.**