



Resident's Page

Laws and signs of congenital syphilis

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Sir William Osler aptly quoted, "He, who knows syphilis, knows medicine." Perhaps, no other infection has been studied so keenly as syphilis till date. Nevertheless, the florid multisystem manifestations in congenital syphilis have fascinated humankind so much that certain laid down laws and numerous eponyms have emerged.

LAWS OF CONGENITAL SYPHILIS

1. COLLES' OR BAUMIS LAW (1837) after Colles – A newborn child affected with congenital syphilis, even though it may have symptoms in the mouth, never causes ulceration of the breast which it sucks, if it is the mother who suckles it, though continuing capable of infecting a strange nurse.^[1]
2. PROFETA'S LAW (1865) after Profeta G – At birth, an infected infant may appear healthy and may not develop signs till it is some weeks old, but in other cases, signs are present at birth.^[2]
3. DIDAY-KASSOWITZ LAW (1876) after Paul Diday and Kassowitz – If a woman with untreated syphilis has a series of pregnancies, the likelihood of infection of the fetus in later pregnancies becomes less. Thus, in successive pregnancies, she may have miscarriage at the 5th month, next occasion-stillbirth at the 8th month, next, birth of an infant with congenital syphilis who dies within a few weeks followed by birth of 2 or 3 infants with congenital syphilis who survive, finally, 1 or more healthy children may be born.^[1]
4. FOURNIER-FINGER CRITERIA (FOR THIRD-GENERATION SYPHILIS) after Fournier (1891) and Finger (1900):
 - (a) Acquired syphilis must be demonstrated in grandmother and preferably also in grandfather.
 - (b) Prenatal syphilis must be demonstrated in the mother. Acquired syphilis must be excluded in her case and father must be proved healthy.
 - (c) There must be incontrovertible evidence of prenatal syphilis in the third generation.
 - (d) Manifestations must appear soon after birth in the second and third generations.^[3]

SIGNS IN CONGENITAL SYPHILIS

- BARBER-POLE APPEARANCE OF UMBILICAL CORD – Interspersion of blue and pink areas along with chalky white coloration on umbilical cord in spiral configuration due to necrotizing funisitis.^[4]
- BATTLEDORE PLACENTA – Umbilical cord is inserted at or near the placental margin.
- BUCKET HANDLE SIGN – Fractures through degenerated metaphysis.

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- BULLDOG FACIES – Saddle nose, short maxilla, and prominent mandible which are stigmata of early stages.
- BULLDOG JAW – Normal mandible that appears relatively longer and bigger due to maxillary hypoplasia.^[1]
- CAFÉ-AU-LAIT TINT – Brownish-yellow discoloration of skin in early congenital syphilis due to the combination of anemia, jaundice, and hyperpigmentation.^[1]
- CELERY-STALK SIGN – Plain film appearance of metaphyses characterized by longitudinally aligned sclerotic bands. Also seen in congenital rubella, toxoplasma and cytomegalovirus infections.
- CLUTTON'S JOINTS – Chronic, painless, and insidious joint effusion of knees in late congenital syphilis.^[5]
- CONDYLOMA LATA – Seen in early congenital syphilis as moist and eroded papules in intertriginous region, especially napkin area which may later become hypertrophic, identical to those of secondary stage in acquired syphilis.^[1]
- DUBOIS' SIGN – Short incurved little finger that occurs as stigmata.^[6]
- FURUNCLE OF BARLOW – Indolent purplish cutaneous swelling which contains scanty secretion of ill-formed pus, seen toward the end of the 1st year of life.
- GHOST VESSELS – Empty blood vessels extending from sclerotic to deeper layers of cornea occurring as stigmata of interstitial keratitis.
- GROUND-GLASS CORNEA – Hazy cornea due to cellular exudation into deeper layers in late stages.
- HENNEBERT'S SIGN – Nystagmus caused by pressure applied to a sealed external auditory canal. Also positive in Meniere's disease.
- HIGOUMENAKI'S SIGN – Unilateral enlargement of sternoclavicular articulation in late congenital syphilis.
- HUTCHISON'S TEETH (Screwdriver teeth) – Abnormal permanent upper central incisors that are peg shaped and notched, usually with obvious thinning and discoloration of enamel in the area of notching; they are widely spaced and shorter than lateral incisors; width of the biting surface is less than that of the gingival margin.^[7] It is one of the commonly observed stigmata.
- HUTCHISON'S TRIAD – Hutchison's teeth+interstitial keratitis+sensorineural deafness.
- INFANTILE ALOPECIA OF EYEBROWS – Suggestive of early congenital syphilis.
- KRISOVSKI'S SIGN – Cicatricial lines radiating from the mouth.
- LEUKOMELANODERMA – Hypo- and hyperpigmented macules over palms and soles.
- MULBERRY MOLARS (Moon/Fournier molars) – The biting surface of the first molars is dome shaped and has multiple underdeveloped and poorly enameled cusps that occur as a part of stigmata.^[1]
- NATIFORM SKULL (Hot cross bun skull) – Frontoparietal bossing along with prominent suture lines of skull, stigmata due to healed gummatous osteoperiostitis.
- OLD MAN LOOK – In early congenital syphilis, loss of weight produces wrinkled skin, likened to that of elderly people.
- OLYMPIAN BROW (Beetled brow) - Bony prominence of forehead – one of the classical stigmata of congenital syphilis.
- ONION PEEL PERIOSTEUM – Successive layers of bone are laid down on the surface of the cortex in regular fashion giving this radiographic appearance in early stages.
- OPERA GLASS NOSE – Due to nasal chondritis, lower nose appears to be pushed into the intact upper nose.
- PARROT'S NODES – Localized osteoperiostitis of skull vault leads to the formation of round bony swellings in frontoparietal region followed by permanent thickening of bones in late congenital syphilis.
- PSEUDOPARALYSIS OF PARROT – Extremely tender bones leading to decreased movements in early stages.^[1,8]
- RHAGADES (Parrot's radial scars) – Radiating fissures at angles of mouth produced by the movement of lips that become secondarily infected, heal as linear scars, in early congenital syphilis
- SABRE SHIN (Sabre-blade tibia/Fournier's sign) – In late congenital syphilis, sharp anterior convexity of tibia occurs due to thickening of middle third of shaft.
- SADDLE NOSE (Fleur de lis nose) – Stigmata due to flattening of bony bridge of the nose due to improper development. Can be seen in a variety of conditions such as nasal trauma, Hansen's disease, relapsing chondritis, and granulomatosis with polyangiitis.
- SALMON PATCH – Due to circumcorneal vascularization, a dull pink patch occurs at the periphery of cornea in late stages.
- SALT AND PEPPER FUNDUS – Chorioretinitis giving rise to tiny light specks interspersed among dark specks, can be seen in all stages.
- SARCOPHAGUS SIGN – Periosteal new bone formation resulting in layers of marrow trapped between layers of subperiosteal bone encasing entire shaft resembling a sarcophagus.^[9]
- SILEX'S SIGN – Radial furrows around mouth.
- SNUFFLES – Rhinitis, earliest manifestation of congenital syphilis due to mucous patch like lesions in mucoperiosteum of nasal cavity.^[1]
- SPLIT PAPULES – Mucous patches at angle of mouth or corner of nose in early congenital syphilis.
- STOKES FACIES – in alert, sleepy, tired, fagged, clouded, dreaming, or obscured appearance of upper face; an

appearance of veiledness as if a smudge had lightly swept across the brows and eyes and the nasal bridge of a crayon portrait.

- SYPHILITIC DACTYLITIS – Painless, fusiform swelling of fingers due to osteoperiostitis of proximal phalanges in early congenital syphilis
- SYPHILITIC ONYCHIA – Loosening and shedding of nail
- SYPHILITIC PEMPHIGUS – Bullous lesions, earliest skin manifestation.
- SYPHILITIC WIG – Abundant coarse hair which stops short abruptly at hair margin in early congenital syphilis
- TOWER SKULL – High cranium
- VIRCHOW'S SIGN – Tongue with smooth base in congenital syphilis.^[10]
- VON GIE JOINT – Bone erosions within involved joints.
- WHITE PNEUMONIA (Pneumonia alba) – Firm and pale lungs due to inflammation and fibrosis in alveolar septa.^[11]
- WIMBERGER SIGN (Cat bite sign) – Localized bilateral metaphyseal destruction of medial proximal tibias in early congenital syphilis. Also seen in scurvy.
- ZIG-ZAG SIGN (Saw-tooth metaphysis/Wegner sign) – Serrated appearance at the metaphysis of long bones.^[11]

CONCLUSION

Awareness of the basic laws and signs of congenital syphilis is imperative for any dermatovenereologist to identify early lesions and takes timely interventions to prevent future complications and stigmata of the disease.

Declaration of patient consent

Not required as there are no patients in this article.

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Conflicts of interest

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