

Endemic syphilis in Europe

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Abstract Nonvenereal syphilis (endemic syphilis) has existed in Europe since the 16th century. Main characteristics of the disease are its presence for a longer time in a specific territory and its transmission regardless of age and sex, mainly extragenitally in unsanitary living conditions. Nonvenereal syphilis was described under different names in almost all regions of Europe. The primary genital chancre was absent, and lesions were most frequently found in the mouth and affected mostly children. The disease spread in rural areas with poor economic and hygienic conditions. The disease was eradicated in Europe in the 20th century, but it is still present in some rural regions of the Arabian Peninsula, Southwest Asia, and North Africa. © 2014 Elsevier Inc. All rights reserved.

Introduction

Human treponematoses (some prefer singular, treponematosis) are chronic infectious diseases caused by noncultivable, strictly parasitic bacteria of the genus *Treponema* (formerly *Spirochetae*), characterized by a very small genome (about 1.1 Mb) and accordingly limited metabolic capabilities, thus deriving important macromolecules from the host.¹

Some authors believed there was only one disease, treponematosis,^{2,3} with different clinical patterns, linked to various climatic, socioeconomic, and hygienic conditions, while others thought that there were four diseases (treponematoses) caused by different pathogenic *Treponemas*, more or less widely distributed in the world.

Cecil J. Hackett (1905-1995) has proposed dividing treponematoses in two distinct groups⁴:

1. venereal (or sporadic) treponematosis, ie, venereal syphilis caused by the subspecies *Treponema pallidum*,

transmitted mostly by sexual contact and very rarely by accidental transmission or from mother to fetus

2. nonvenereal treponematoses (endemic) or extravenereal treponematoses for which the living conditions are essential, affecting especially children, and acquired by social contacts in undeveloped rural regions. These nonvenereal treponematoses encompass endemic (nonvenereal) syphilis caused by the *Treponema pallidum* subspecies *endemica*; yaws (framboesia) mostly occurring in the tropics and caused by the subspecies *Treponema pallidum* subspecies *pertenue*; and finally pinta (carate) present in Central and South America, caused by *Treponema carateum*, a disease confined only to the skin.⁵

The etiologic agent(s) of these treponematoses are closely related and impossible to distinguish by dark field microscopy, electron microscopy, serology, and even histopathologic examination.^{6,7} The treatment of treponematoses is also characterized by similar response to drugs. This makes the diagnosis of a specific disease based not on the etiologic agent, but on the clinical manifestations, epidemiology, and geographic factors. Recently, owing to new investigations,

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the issue of their diagnosis can be improved by subtle genetic differences.^{8,9}

Characteristics of endemic syphilis in Europe

There are several possibilities to define endemic diseases, and some believe the term is not completely appropriate. According to the opinion of Franjo Kogoj (1894-1983) (Figure 1), Josip Fleger (1896-1966), Kristian Grön, and others, 10-12 endemic treponematoses have been characterized by the presence for a longer or shorter time in a specific territory and transmitted regardless of age and sex, mainly extragenitally in unsanitary living conditions. Sometimes, with changing local conditions, endemic syphilis can turn into venereal syphilis and vice versa.13 Interestingly, like venereal syphilis, endemic syphilis also had numerous local names in various countries and European regions. Frequently, the names were connected with the places where it originated; ie, morbus Brunogallicus, Scherlievo or Skrljevo disease, morbus Brenensis, Jutland disease, and so on. To differentiate these diseases from venereal syphilis, they were also termed syphiloids or trepanoid. Today, few physicians know the true meaning of these names.

In the past, as for other treponematoses, the diagnosis of nonvenereal syphilis was made only on clinical grounds. Prior to the 20th century, the etiologic agents were not discovered nor were there serologic reactions to demonstrate the presence of treponemal antibodies. Transmission of nonvenereal treponemas from one individual to the other can be via direct skin to skin or mucosa to mucosa contact, or indirect through eating or drinking utensils usually in undeveloped rural territories, crowded conditions, and poor hygienic conditions.¹⁴

The diagnosis, in view of lacking the discrimination by means of serology, of nonvenereal syphilis is characterized



Fig. 1 Academician Franjo Kogoj.

by the absence of a genital chancre. Sometimes, chancres can be found on the oropharyngeal mucosa or on the nipple in women nursing an infant, accompanied by regional lymphadenitis. The second stage follows with mucous patches, localized papules on the skin, condylomata in intertriginous areas, nocturnal pain in the long bones of the extremities, and in tertiary stage development of gummata on the skin, bones, and nasopharynx. Cardiovascular and central nervous involvement is possible but rare.¹⁵ Infection in early childhood is common, while congenital transmission is rare. The largest number of patients with endemic syphilis was diagnosed in the secondary or latent stage.¹⁶ Significance for the diagnosis depends on the presence in some European countries. Numerous data on the history of endemic syphilis can be found in E. Lanceareaux's book¹⁷ in the chapter on endemic epidemic of syphilis and in K. Grön's monograph in J. Jadasshon's Handbook,¹⁰ even if he did not differentiate completely endemic from accidental syphilis and believed that in the first half of the 20th century endemic syphilis was only present in Russia, whereas it is known that it was also present on the Balkans (Bosnia, Croatia, Serbia).

Today, endemic or nonvenereal syphilis is present in some rural regions of the Arabian Peninsula (Bejel), Southwest Asia, and North Africa.^{14,15} Endemic syphilis disappeared in Europe in the second half of the 20th century after elimination of the last foci in the Balkans due to penicillin treatment and the improvement of socioeconomic and hygienic conditions.

Endemic syphilis in the 16th century

Syphilis, after its emergence in Europe in the 1490s, soon spread as a venereal disease throughout Europe and even worldwide. It became clear that it could also propagate in other ways. As early as 1524, Erasmus from Rotterdam (1466?-1536) described transmission of syphilis in some Denmark baths.¹⁰

In 1575, in Brno (formerly Brünn), a strange contagious disease appeared affecting about 200 people in the urban area and more in the rural parts, over a period of several months. The disease was unknown and characterized with pain in the bones, headache, and exanthema. Various theories had been advanced to explain the new disease. It seems that it appeared after the visit of some traveler and the spread was connected with the use of a communal bath and bloodletting. Sometimes, ulcers appeared at the sites where the cupping glasses had been applied by barbers, using no disinfection.¹⁰ Thomas Jordanus (1539-1585) called the disease morbus Brunogallicus and the earliest notice about the malady one can find in his "Lues novae in Moravia anno 1577 ex ortae descriptio" (Frankfurt, 1580). A similar endemic disease appeared in 1591 in Bern (Switzerland) and was also linked with the use of bathing and cupping.¹⁷

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