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Hutchinson's incisors and Moon's molars: accurate diagnosis to restore form and function

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Abstract: Congenital syphilis is endemic in our country, with thousands of new cases per year and increasing prevalence, especially in the Southeast region. Among its manifestations are those involving the teeth: Hutchinson's incisors and Moon's molars. From

the discussion about the mechanisms that lead to its occurrence, the criteria for its diagnosis and forms of treatment, it was sought to revisit the subject, contributing so that Dentistry professionals may recognize it in a precise way and contextualize it in the patient's

systemic condition in order to treat the teeth involved, both aesthetically and functionally. For this, the subject must be known, reviewing it as presented: a new public health problem. **Keywords:** Hutchinson's incisors. Moon's molars. Mulberry molars. Congenital syphilis.

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WHY THIS ARTICLE?

1 - Some diseases, such as smallpox and measles, have been eradicated at a certain point; others have become historical and rare; some, however, return with the same characteristics or new forms and surprise the biomedical world. When this happens, new professionals should be able to interpret their signs and symptoms, in addition to complementary tests.

2 - Syphilis is a unique example. It has, again, become endemic in many countries.^{3,6} In Brazil, between 2014 and 2015, there was an increase of 32%, with 228 thousand new cases, the great majority in the Southeast.⁵

In our country, this happens due to an unusual reason: the syphilis medicine to which the bacterium does not develop resistance, penicillin, is very efficient, but extremely cheap, and its manufacture started not to be so interesting from an economic point of view. Governments then have difficulty in acquiring it on the market, and patients go untreated.

3 - To dramatize even more the carriers' situation, many are women and become pregnant at risk of passing syphilis to their children, who will be born with the congenital version of the disease. Among pregnant women, syphilis has increased by 20%. There are thousands of children with congenital syphilis and many do not even know it.

In 2015, there were 6.5 live births with congenital syphilis per thousand births in Brazil,⁵ which represents 13 times what the WHO considers acceptable. Many patients with congenital syphilis' mothers were not treated due to the lack of social assistance, available medicine and lack of adequate diagnosis by health professionals. For example, in Bauru, in the state of São Paulo, a city with 350 thousand inhabitants in 2014, according to the Municipal Health Department 413 pregnant women were diagnosed with

syphilis; in 2015 were 535 and in 2016 until October there were 407.⁴

4 - One of the most classic and pathognomonic manifestations of congenital syphilis affects the incisor teeth and the first molars, which for a long time have become so "historical" that the great majority of professionals have never diagnosed them in their treatment (Fig 1-5).

In this article, we sought to revisit knowledge about congenital syphilis and its dental manifestations, inducing reflections on its treatment.

ABOUT SYPHILIS: A SYNTHESIS OF THE CLINICAL AND EVOLUTIONARY PICTURE

Syphilis is not an STD only when transmitted from the mother to the fetus through the placenta. It had great prevalence until World War II, when the use of penicillin caused its incidence to decrease to no longer worrying indexes. As of 1955, the indexes have risen to the present day in a frightening and endemic way. Syphilis is an exclusively human disease.



Figure 1: Clinical aspects of Hutchinson's incisor: maxillary central incisors with barrel shape. The central bevel decreases as the tooth wears out during use over the years.



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Figure 2: Morphological aspects of Hutchinson's incisor specimens: **A)** Maxillary lateral incisor; **B)** maxillary central incisor; and **C)** mandibular incisor. The central chamfering stands out.

Its causative agent, *Treponema pallidum*, grows and survives well on the genitalia and mouth. Ninety to ninety-five percent of the diagnosed cases are by sexual transmission, and the mouth is the most common extragenital region of occurrence.

Rarely (and generally doubtfully), there is indirect transmission by contaminated objects, since *Treponema pallidum* has no penetrating power in the skin. Its invasion occurs in intact mucous membranes or areas of abraded skin, as may occur during the sexual act. Transmission through blood transfusions is extremely rare.

Treponema pallidum was discovered by Schaudinn and Hoffmann in 1905.⁷ It is a thin, delicate spirochete with 6 to 14 spirals, measuring 6 to 15 micrometers long by 0.2 micrometers in diameter (Fig 6), with no possibility of being cultivated in the laboratory.

There is a classic classification of the evolutionary course of the disease in adults that divides it into primary, secondary and tertiary, intermediated by periods of clinical latency. The primary and secondary phases are the most infecting.



Figure 3: Morphological aspects of Hutchinson's incisors specimens and Moon's molars from the collection of the Hunterian Museum of the Royal College of Surgeons of England (London) presented by Nissanka-Jayasuriya, Odell and Phillips⁶ in 2016.



Figure 5: Morphological aspects of Moon's molar with the constricted occlusal face of the first maxillary molar.

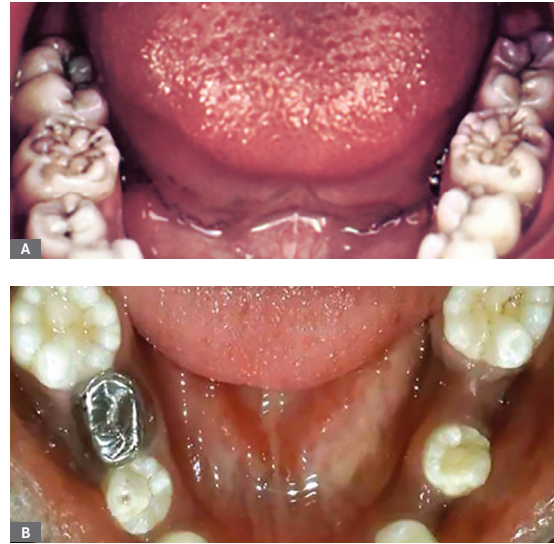


Figure 4: Morphological aspects of Moon's molars, or mulberry, presented by Robinson et al. 9, 1990, and by Chowdhary et al.² in 2014.

Primary phase - When penetration occurs, just below the epithelial lining, treponema propagates easily, gaining the perivascular spaces. In a few hours, it reaches the regional ganglia into the bloodstream. Part of the microorganisms is in the place of penetration and proliferates, generating a characteristic lesion of the disease: the primary cancer. The incubation period varies from 15 to 30 days, with an average of 3 weeks.

At the site of inoculation, primary cancer, usually isolated and painless, may or may not occur. Initially, it is a macula, then a papule and, with its growth and infiltration, it can evolve into erosion. When the ulcer is formed, it has beveled and narrow edges, it is bronze in color, well-delimited and very firm, receiving the name of hard cancer. In the mouth, it occurs mainly on the lips and tongue. Eventually, it can occur in the eyes of dentists and assistants. The cancer lasts for 2 to 6 weeks, progressing to fibrosis and spontaneous scarring.

At all stages and locations of the disease – when a biopsy is performed, as syphilis was not suspected – the microscopic mark of the syphilitic inflammatory reaction is obliterating endarteritis. The small arteries and arterioles in the inflammatory reaction exhibit swelling and proliferation of endothelial cells, producing concentric lesions in onion peel form, with marked narrowing of light. Around these vessels, there is a prominent perivascular infiltrate of plasmacytes in the form of a cuff. Such alterations lead the pathologist to suggest, microscopically, the diagnosis of syphilis. In cases where there is clinical suspicion of the disease, the most practical and effective means of diagnosis are serological tests, VDRL type and not biopsy.

Secondary stage - The primary manifestation may or may not occur. With or without this primary manifestation, there is a period of 2 to 4 years of clinical latency, broken by generalized mucocutaneous manifestation: secondary syphilis.

Rashes are red or pink macules or papules, 5 to 10mm in diameter. In the body, they tend to be symmetrical and distributed mainly in the trunk and extremities, especially in the soles and palms, as well as in the face. Lesions may become scaly or pustular, but are not pruritic. They can affect the hair follicles and promote hair loss, characterizing alopecia areata.

In the mucosa, these lesions appear slightly grayish, high and small, with no necessary lymphadenitis, usually present when the lesions are cutaneous. In 10 to 15% of the patients, mucous plaques are formed on the lips, tongue, palate, pharynx, vulva, vagina, penis and foreskin.

In about 25% of cases, the patient and the professional's diagnosis is confused with "allergic" manifestations. The secondary lesions disappear and recur in two years until total dis-

appearance, when they lie dormant indefinitely.

Tertiary phase - When untreated in secondary manifestations, after many years (10-20 years), the tertiary phase occurs in 1/3 of the cases. Tertiary lesions are necrotic, extensive, deep, and deforming. They are large inflamed chronic hyperergic areas with necrosis, local fibrosis and severe, sometimes dramatic, sequelae. These areas may be mucocutaneous, skeletal-muscular, parenchymal, highly destructive, including vessel walls such as the aorta and central nervous system.

Diagnosis and treatment - The diagnosis and treatment of syphilis must be finished and applied by the doctor specialized in infectious diseases, since the disease is necessarily systemic and requires notification for public health control.

The diagnosis is made through serological tests, such as VDRL, which is positive 5 to 6 weeks after infection, just before the cancer heals. In the secondary phase, the levels are strongly positive, from 1:32 to 1:256, unlike the primary phase, when they are 1: 2. In the tertiary phase, positivity levels are reduced to 1: 4 to 1: 8. Other tests are usually requested, such as FTA- Abs, or Fluorescent Treponemal Antibody Absorption.

Treatment is done with antibiotics and *Treponema pallidum* does not present resistance. In cases of early latent syphilis, primary and secondary syphilis, as well as after the contact with syphilitic patients, IM penicillin G benzathine is administered. In allergic patients, oral doxycycline for 14 days. In cases of late latent syphilis and unknown progression, as well as in the tertiary phase without neurosyphilis, the doses are almost triplicate.

Another classification - Syphilis is also classified as early or late, not only to facilitate



Figure 6: *Treponema pallidum* among ameloblasts in germ, presented in the classic work of Pindborg⁷ in 1970.

diagnosis, but also to apply standardized therapeutic protocols. Early syphilis last up to 2 years and comprise what is called in the previous classification primary, secondary and early latent. Late syphilis would correspond to the late latent phase and tertiary phase.

CONGENITAL SYPHILIS: THE TRANSMISSION PATH IS PLACENTAL

Transmission of *Treponema pallidum* through the placenta occurs in the fourth or fifth month, when the placental barrier is thinner, more precisely between the 16th and 18th weeks of intrauterine life.

If the patient with congenital syphilis survives, during his development he may present a series of signs and symptoms known as the Hutchinson's triad, described in 1858-1863, by Sir Jonathan Hutchinson (1828-1913),¹¹ and consisting of:

1) Diffuse interstitial keratitis with corneal opacification in the eyes, leading to blindness and manifesting after 5 to 25 years of age.

2) Labyrinth disease, which can lead to deafness, due to changes in the 8th cranial nerve.

3) Hutchinson's incisors and Moon's molars (Fig 1 to 5).

Other signs of congenital syphilis are: maxillary hypoplasia, ogival palate, nasal sealing, mandibular prognathism, saddle nose and pronounced frontal gibbosity.

CONGENITAL SYPHILIS IS BACK: HUTCHINSON'S INCISORS AND MOON'S MOLARS AND THEIR CLINICAL CHARACTERISTICS

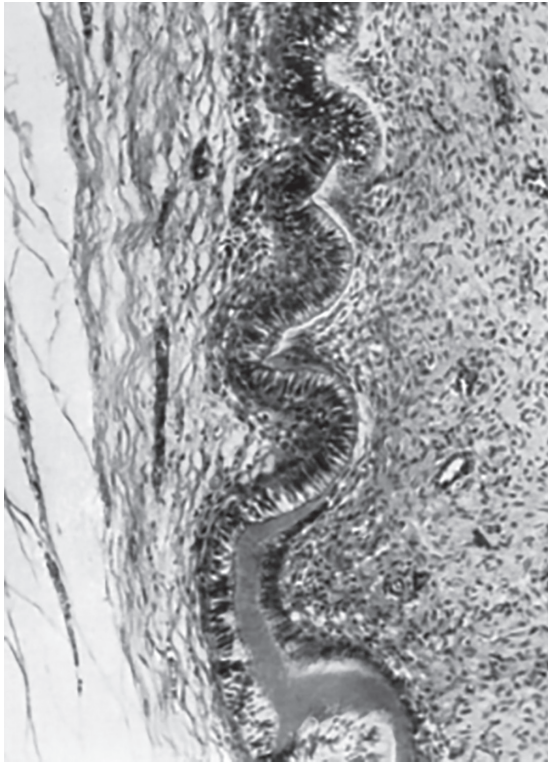
The prevalence of dental changes of congenital syphilis varies widely. In a sample of 36 cases of congenital syphilis in the USA, 67% of the patients had dental alterations. In Finland, of 254 patients with congenital syphilis, 45% had changes in the incisors and 22% in the molars.⁸

After the initial description, it was noted that these typical alterations of the maxillary central incisors also occurred in other incisors, canines, as well as in the first molars.

The typical Hutchinson's incisor is smaller than the normal central incisor.³ The crown has its sides converging from cervical to incisal, with its proximal angles well rounded, which gives them the shape of a barrel (Fig 1, 2, 3). The enamel of Hutchinson's incisors may be very slightly grayish.

In many cases, the incisal margin may reveal a central chamfer, but it may also not be present, which causes the altered form not to draw the clinician's attention so much to this diagnosis. The chamfer may also disappear as the tooth wears off at the incisal margin (Fig 1).

A diastema between the maxillary central incisors is present in patients with congenital syphilis, with 1/3 of the patients having anterior open bite.



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Figure 7: Irregularity and focal proliferations of the ameloblasts and intermediate strata in the region of the future amelodentin junction, which would lead to the tuberosities in the enamel production in the teeth affected by *Treponema pallidum*, in the original work of Bradlaw¹ in 1953.

The typical first molar of congenital syphilis is also known as mulberry molar, first described in 1876 by British surgeon Henry Moon (1845-1892). In general, it has its crown smaller than the first and second normal molars.⁸

The main characteristic of Moon's molars is the reduction of the crown's diameter on its occlusal surface, which is constricted (Fig. 5) and/or with numerous irregular protrusions or cusps, resembling the surface of a mulberry (Fig 3,4).^{2,9} In many cases, these changes are very discrete. Although Hutchinson's incisors and Moon's molars are symmetrical in almost all occurrences, there are asymmetric cases.

The involvement of deciduous teeth in congenital syphilis is extremely rare because, in general, transplacental infection occurs between 16th to the 18th weeks of pregnancy, when the germs of deciduous teeth have passed the morphodifferentiation phase.

HOW DOES THE FORMATION OF THESE DENTAL CHANGES OCCUR IN CONGENITAL SYPHILIS? MECHANISMS AND OBSERVATIONS

Enamel mineralization is normal in the enamels of the involved teeth, but there are irregularities in the amelodentin junction,¹ as seen in one of the illustrations of the original work (Fig 7). The dentin, in turn, is hypomineralized, with many areas of interglobular dentin and cell inclusions.

In fetus and newborns with congenital syphilis, an infiltrate of plasmocytic cells and vascular changes in the pericoronyal follicle and the papilla/dental pulp were noted. In the enamel organ, disturbances in the amelogenesis are observed, because *Treponema pallidum*, when invading the dental germ, is lodged between the ameloblasts (Fig 6).¹

It is assumed that the involvement of the dental germ by *Treponema pallidum* leads to a proliferation of the intermediate layer and the ameloblasts, forming an irregular and wavy amelodentin junction, determining small and nodular enamel protuberances in the place of the cusps and tubers, especially in the molars.

The detailed analysis of two specimens of Hutchinson's incisors by Hillson et al,³ in 1998, allowed us to observe that changes in the shape of these teeth by *Treponema pallidum* occur after the patient's birth, when the process of amelogenesis begins in permanent teeth.

TREATMENT OF AFFECTED TEETH IN CONGENITAL SYPHILIS

Hutchinson's Incisors - Because it is a marker sign of congenital, pathognomonic syphilis, it should be reanatomized with aesthetic material even if discrete. In agreement with this approach, if there are diastemas between the central incisors and/or anterior open bite, the orthodontic measures for their correction and/or interception must be proposed and executed. The patient's smile should not allow others to recognize their congenital syphilis condition.

From a structural point of view, Hutchinson's teeth should not be considered completely normal. The occurrence of these defects occurs due to the interference of *Treponema pallidum* in enamel production, which represents enamel hypoplasia. The amelodentinal junction becomes irregular, wavy and the underlying dentin with a greater amount of interglobular dentin. The present enamel has a normal structure. Although there is no such reconstructive approach in the pertinent literature, it seems logical to us that a return to normal anatomy and physiology should be provided for these Hutchinson teeth.

Moon's Molars - The mulberry occlusal surface, with numerous protuberances and cusps, can generate occlusal interferences to be corrected with adequate adjustment. A greater amount and irregularity of pits and fissures may require the use of sealants and/or even the restoration of the occlusal face if cavities are detected.

FINAL CONSIDERATIONS

Literature is very restricted when searching for information on the occurrence of Hutchinson's incisors and Moon's molars, especially about its aesthetic and functional restorative approach.

In spite of the large number of cases of congenital syphilis in the country, the diagnosis of the disease and/or the treatment of the aesthetic and functional sequelae of the affected teeth are rare in written works or presentations at congresses and other scientific events. Finally, some questions to induce reflections on the subject:

1) Cases of Hutchinson's incisors and Moon's molars do not occur or they are not diagnosed/recognized by professionals?

2) From an aesthetic and functional point of view, is the professional fit or trained to correct the anatomy and physiology of these teeth?

3) Regarding the systemic situations involved and the manifestations of congenital syphilis in other body regions: will the dentist be able to recognize other signs and symptoms, as well as interact with the medical team to participate in the treatment and follow-up of these patients?

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