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Endemic Nonvenereal Treponematosis (Bejel) in Saudi Arabia

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A total of 2,515 individuals attending a large military hospital in Saudi Arabia who had appropriate radiologic evidence of treponematosis were studied clinically and serologically. The indications are that nonvenereal treponematosis (bejel) exists in considerable numbers among the nomadic communities living in rural areas. In contrast, venereal syphilis is less common in this population and is found almost exclusively in urban populations. Some of the high-risk regions for bejel have been identified. Many individuals from nomadic communities complained of persistent pain in the lower limbs, which was often associated with radiologic evidence of osteoperiostitis of the long bones. It also appears that within the last 30 years bejel has become clinically attenuated, with the majority of seropositive individuals having latent disease. A hypothesis is put forward that persistent lesions are sustained by superinfection and that improvements in hygiene have resulted in a decrease in the incidence of reexposure. Measures to control the infection are outlined.

Endemic syphilis, called bejel in the Middle East, is a nonvenereal treponematosis that has its onset in early childhood and is transmitted from child to child by close skin-to-skin contact, by kissing, and possibly by fomites such as communal drinking vessels.

Major features of bejel and venereal syphilis are compared in table 1. The classic form of bejel is easily recognized as a clinical entity; the initial infectious mucous patches in the mouth usually are followed by the appearance of a generalized nonirritating rash such as that seen in syphilis but much more marked and persistent. In some patients a late stage develops during which gummata of the skin, bone, and cartilage progress, sometimes resulting in the formation of destructive lesions, especially of the nose and palate. Painful osteoperiostitis of the tibia and fibula is common. This florid form was first described by Hudson in Syria [1], and the description was later amplified by Hudson [2] and Csonka [3] in Iraq. Bejel has been reported from Yugoslavia, where it has now been completely eradicated [4]. The disease is still present in parts of Africa and Southeast Asia [5]. It is confined to nomadic and seminomadic communities living in the remoter rural areas, where the standard of hygiene is low and access to static health services is limited. The florid form of this disease appears to have been significantly modified in a generation, having been replaced by a milder form in which the number, severity, and duration of both early and late lesions are reduced. The reason for this change is not clear. Since the sera of individuals with bejel are positive for lues to a degree that is similar to that for individuals with venereal syphilis at the same stage, a simple serologic survey of rural communities should provide the epidemiologic data essential for mapping the location of endemic foci, information that can serve as a basis for the introduction of control measures.

In a recent report the World Health Organization (WHO) linked the patchiness of the distribution of areas with persistent nonvenereal treponematosis not only to inadequate medical services but also to the increase in the number of atypical and latent cases, which are not identified by clinicians unfamiliar with these forms of the disease [5]. While bejel was contained in parts of the Middle East after the introduction of treatment campaigns in the 1950s, persistent foci of the infection were reported in 1954 by a team from WHO [6] and again in 1979 by Sebai and Baker [7] and have been confirmed by us [8]. Such information is important, as the social consequences of mistaking nonvenereal bejel for venereal syphilis can be catastrophic. These considerations encouraged us to extend our investigations. We compared evidence (anamnestic, clinical, serologic, and radiologic data) on treponematoses for those attending the hospital who were nomads and those who were born and bred in towns.

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Table 1. Major features of bejel compared with those of venereal syphilis.

Feature	Venereal syphilis	Bejel
Causative organism	*Treponema pallidum*	*T. pallidum*
Geographical distribution	Worldwide	Middle East, Africa, Southeast Asia
Climate	Any	Hot and humid or dry
Transmission		
Venereal	Yes	No
Congenital	Yes	No*
Nonvenereal contact	Very rare	Yes
Contaminated fomites	Very rare	Yes
Age group infected	Mostly young adults	Young children
Sex	Males predominate	Females predominate
Infectious lesions	Early genital sores, open skin and mucous membrane lesions	Mucous patches in the mouth, early open skin lesions; status during early latency undetermined
Late complications		
Skin	Becoming very rare	Becoming rare
Skeletal	Uncommon	Common
Cardiovascular	Yes	No*
Central nervous system	Yes*	No*
Stages of disease	If present, clearcut and separated by time	If present, less clearcut, persists with overlapping
Predominant course of untreated disease	Latency often leads to cure	Latency often leads to cure
Eradication by mass treatment	Not applicable	Possible—achieved in Yugoslavia
Response to penicillin		
Early stages	Excellent	Excellent
Late stages	Variable	Variable

* Generally assumed, but certain proof is not available.

Subjects and Methods

Subjects. In a first retrospective study, the medical records of 105 seropositive patients seen at Abdul Aziz Hospital (Tabuk, Saudi Arabia) between 1976 and 1980 who had a diagnosis of venereal syphilis were assessed, and the findings for nomads and urban dwellers were compared. In a second prospective investigation, 527 women attending the antenatal clinic and 263 of their husbands were examined, and clinical findings and rates of seropositivity among nomads and urban dwellers were compared. In a third prospective study, 1,620 nomadic patients who came from all parts of the country to attend the primary care unit of the hospital were screened. Patients found to have venereal syphilis received 2.4 million units of benzathine penicillin im once a week for three weeks. Patients thought to have bejel were given a single im injection of 1.2 million units of benzathine penicillin.

Serologic tests. The Venereal Disease Research Laboratory (VDRL) and the *Treponema pallidum* hemagglutination (TPHA) tests were used routinely for antibody determinations. The fluorescent treponemal antibody-absorption (FTA-ABS) test was used when results of other tests were inconclusive.

Results

When the medical records of 105 seropositive patients were reviewed in the prospective study, it appeared that all 99 nomadic patients had endemic nonveneral syphilis and that all six city dwellers had venereal syphilis. The rates of seropositivity (by the FTA-ABS test) for the 790 Saudi nationals attending the antenatal clinic in Tabuk were 0.3% (two of 609) for the city dwellers and 19.3% (35 of 181) for the nomads. No cases of "congenital" bejel were observed in infants born of untreated, seropositive nomadic mothers. The rate of seropositivity was greater among the female than the male nomads attending the primary care unit in Tabuk; 20.5% (133

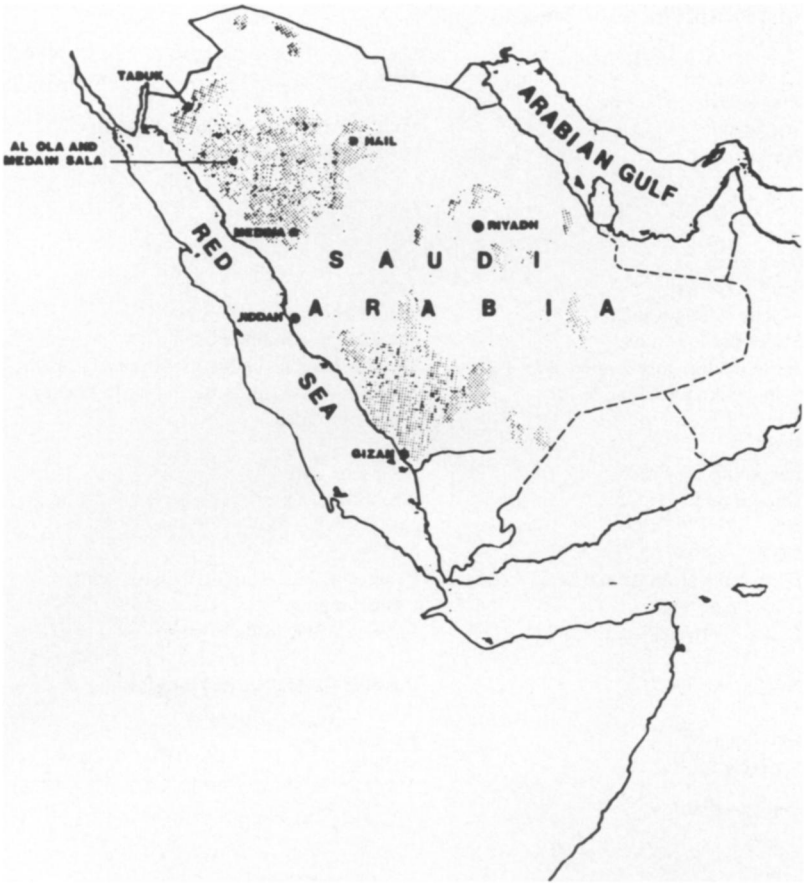
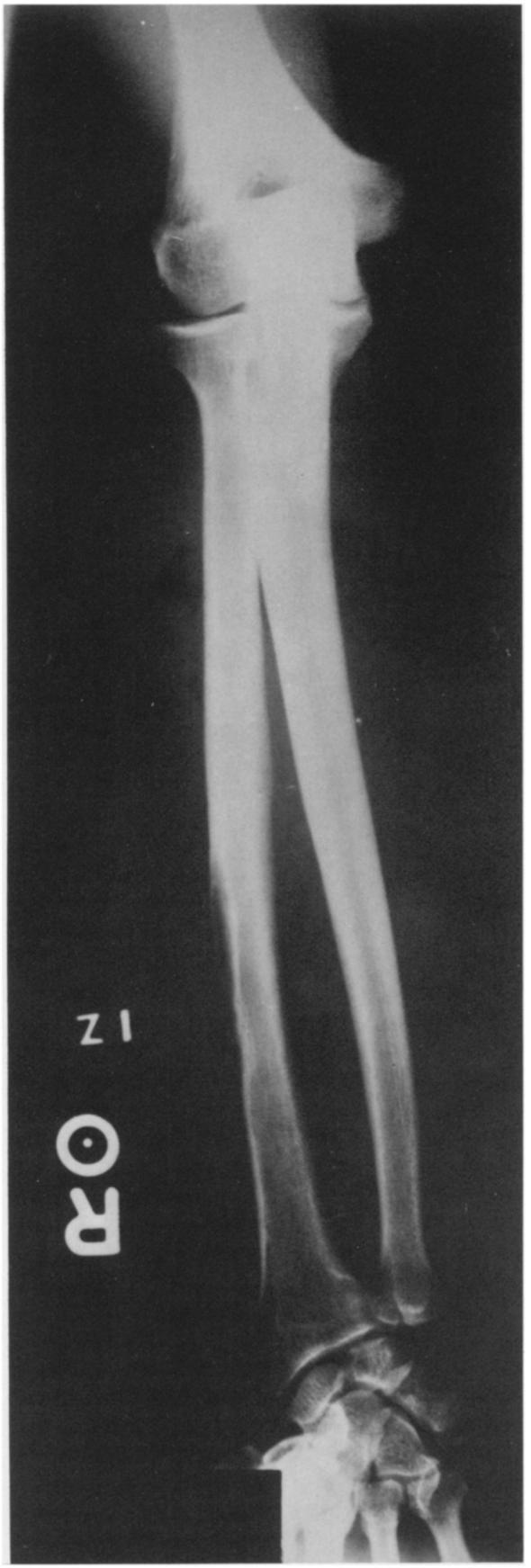


Figure 1. Map of Saudi Arabia; stippling indicates the areas within which 276 nomads seropositive for treponema antibody live.

of 648) of the females and 14.7% (143 of 972) males were seropositive. The majority of the patients in this group were between 15 and 35 years of age. The domiciles of seropositive patients in this group are shown in figure 1; there is clustering in two areas. Radiographs of the long bones of the lower limbs and occasionally of the forearms were taken selectively for the first 200 of these patients, but thereafter radiology of the tibia and fibula was part of the routine examination; 15% of these patients showed evidence of osteoperiostitis. Some of the characteristic radiologic features (figure 2) are similar to those in a patient with active bejel in Iraq who was seen by one of us 30 years ago (figure 3). The majority of patients with radiologic changes complained of nocturnal pain in their lower limbs, and pain was sometimes severe enough to interfere with work and even to curtail all activities. Other late lesions were observed comparatively rarely; these included gummata of the skin and destruction of the palate. One patient, a Bedouin woman with long-standing hoarseness was thought to have tuberculous laryngitis but did not respond to antitubercular therapy. Results of her serum VDRL and FTA-ABS tests were positive, and the biopsy specimen showed granulomatous lesions compatible with treponemal infection. Symptoms cleared promptly with penicillin therapy. Another seropositive patient presented with extreme destruction of the face and nasooropharynx and is at present undergoing a series of plastic repair operations (figure 4).

Discussion

The evidence for the 2,515 individuals in this study, 418 (16.6%) of whom were seropositive, suggests that the incidence of positive serology is significantly higher among nomads than among persons born and living in towns. The absence of young children attending the hospital may have been in part responsible for the absence in this series of patients with early lesions of mucous membranes in the mouth



and early generalized rash. In our experience in neighboring Iraq during the height of the bejel epidemic a generation ago, skin lesions often continued into adolescence; therefore, there appears to have been a genuine decrease in the incidence of clinically obvious early lesions. In a small survey undertaken by one of us in Saudi Arabia in 1978, which included the screening of a number of nomadic children, the absence of frank clinical lesions in children who were seropositive was noted [8]. In contrast, recent reports from the two largest cities in Saudi Arabia, Riyadh and Jeddah, show that classic primary and secondary syphilis and latent syphilis for which there was a history of early venereal syphilis appeared to be the only cause of treponemal infection [9, 10].

The incidence of seropositivity among nomads in the present series is of interest; a larger percentage of females than males were seropositive, a preponderance that is typical of endemic nonvenereal treponematosis. For venereal syphilis males are much more frequently affected. It is therefore becoming clear that one is dealing with two different populations at risk of treponemal infection: the nomadic or seminomadic Bedouins, who appear to have a high incidence of endemic treponematosis; and the people born and bred in towns, who do not get endemic treponematosis but may develop venereal syphilis because the opportunity of exposure is much greater than among the nomads. The present findings suggest that in the nomadic communities bejel accounts for the majority of positive serum reactions and that in the country as a whole bejel probably outweighs venereal syphilis.

Bejel seems to have undergone a change, and more recently fewer clinically observable lesions are observed; this phenomenon has also been reported from areas of the world with other types of endemic nonvenereal treponematoses, but the reason for this change is not known [5].

The only common late manifestation noted by us was painful chronic osteoperiostitis of the tibiae and fibulae. Interestingly, many such patients considered the pain to be due to bejel, which they remember having had in early childhood.

Penicillin was given to patients with skeletal lesions, but it is too early to assess its efficacy; past experience in Iraq indicated that, whereas penicillin halted progression of these lesions, its influence on

Figure 2. Osteoperiostitis of the tibia and fibula of a seropositive patient in Saudi Arabia, 1981.

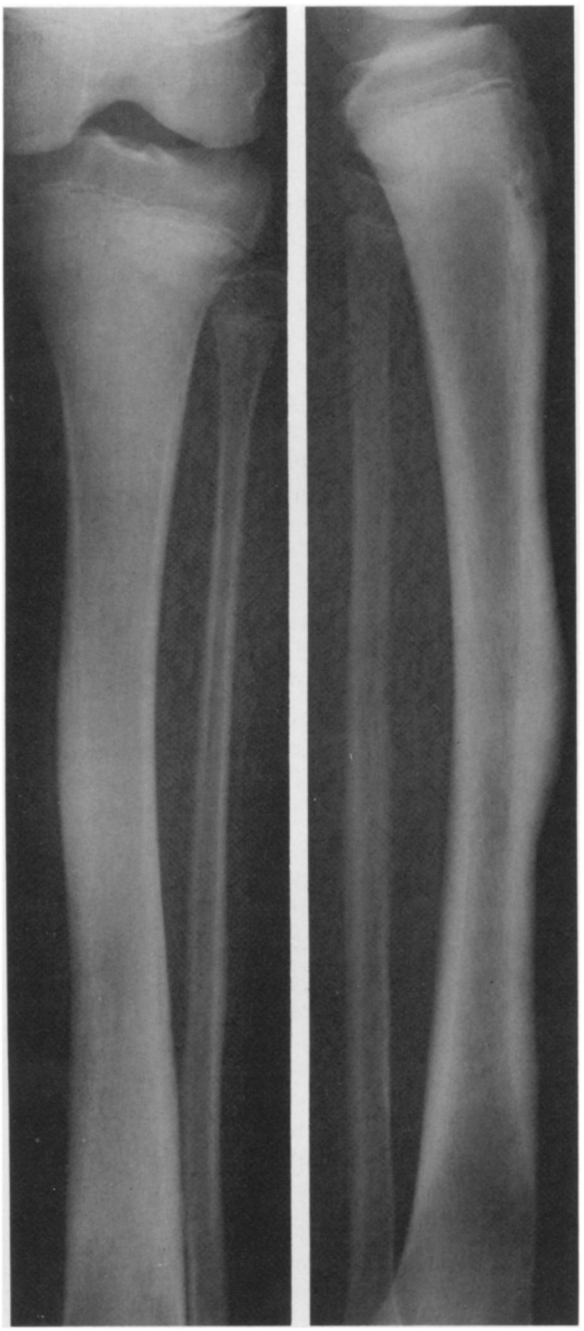


Figure 3. Osteoperiostitis with some osteolytic changes involving mainly the tibia in an Iraqi patient with bejel, 1951.

pain was variable. Destructive lesions of the palate and nasal septum are particularly distressing features of untreated bejel; they are commonly associated with difficulties in speech and eating and are disfiguring. Such an extreme case as that illustrated in figure 4 appears to be very rare, but a similar case in an adolescent boy with bejel in Iraq was reported by Jones [11].

Figure 4. "Mutilating bejel," with almost complete destruction of the nose, nasal septum, and palate, in a patient in Saudi Arabia.

We feel that the time has come to start field studies in Saudi Arabia, since we have identified several high-risk communities. The inhabitants of these areas, including all children, should be systematically screened clinically and serologically to establish the prevalence of bejel and to determine the chain of infection under present circumstances. The most suitable serologic test is the quantitative VDRL reaction, which correlates well with activity. In our hands, however, this test results in false-negative results in 20% of cases and should therefore be paired with a specific test such as the TPHA. If our assumption from the available evidence is correct—that $\leqslant 5\%$ of individuals living in endemic areas will have early active disease—such patients and their immediate

contacts should be treated. The treatment recommended by WHO consists of a single im injection of 1.2 million units of benzathine penicillin for adults and 0.6 million units for children younger than 10 years of age. In the event of there being >5% of such patients in a community, they, their household contacts, and all children younger than 10 should receive treatment. Annual resurveys are essential until proof of eradication has been obtained. A factor that was not significant in the past is people's increasing mobility, which can lead to the spread of the infection across frontiers; thus cooperation of all countries adjacent to an area with endemic treponematosis is necessary to limit such extension. While field studies are proceeding, nomads attending the major hospitals of the country should be screened for endemic treponematosis by the simple VDRL and TPHA tests and the results should be entered on a master map. One can expect to have soon a fairly complete picture, which by constant updating can show the main foci of endemic treponematosis and be used for planning screening and treatment campaigns. It is likely that after such campaigns the infection will have been contained or even eradicated in Saudi Arabia. Some observers question the wisdom of eliminating endemic nonvenereal treponematosis, since this might remove some immunologic protection against venereal syphilis, i.e., by functioning as a sort of natural vaccination. We believe that one should eradicate endemic nonvenereal treponematosis, which can cause avoidable chronic ill health, and treat venereal syphilis as it arises.

The intriguing question, as yet unanswered, is why bejel—and indeed all other nonvenereal treponematoses—have become attenuated within a generation even in communities that until recently have rarely been exposed to antibiotics. It is possible that the early persistent lesions were sustained by frequent superinfection and that with improvement in hygiene such repeated contacts have become uncommon, a change that has resulted in a decline in the frequency, severity, and duration of early lesions. As regards late lesions, there is good evidence that for patients who have had untreated endemic treponematosis, reexposure to fresh infection years later precipitates late lesions [4]. Consequently, as the chances of reexposure diminish so does the development of late lesions. It might be argued that, if this is true, raising the standard of hygiene would by itself eliminate nonvenereal treponematoses. This may be the case in the long run, but the intelligent use of penicillin now, when we know it is still fully effective, can be expected to speed this process. Furthermore, the opportunity given by treatment campaigns can be used to improve hygiene for the benefit of all.

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