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A PROPOSED STUDY OF CONJUGAL LEPROSY WITH REFERENCE TO CONTAGION AND HEREDITARY SUSCEPTIBILITY

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Leprosy is unique amongst the infectious diseases in its tendency to regional, racial, and familial occurrence. The reason for these long observed and striking restrictions in the distribution of the disease has been the subject of continuous controversy. A little over 100 years ago Sir James Y. Simpson's emphatic conclusion that "Few facts in the history of tubercular leprosy seem to be more universally admitted by all writers on the disease, both ancient and modern than the transmission of the predisposition to it from parents to offspring," would seem to have settled the controversy.(1) But from Danielssen and Boeck in 1848 to Hansen, the prevailing view was that the disease itself was hereditary. From Hansen's discovery in 1873 until recent years, contagion, but not without the support of the doctrine of prolonged and intimate exposure, has been the preferred explanation for the epidemiologic features which are unique to leprosy. Ordinarily, in controversial questions of this sort, it is not long before some new advance in knowledge either in the same subject or in collateral fields, forces a decision. The reason why the two opposed schools of thought in leprosy have gone on for so long would appear to lie to a large extent in the fact that there are validities as well as discrepancies in each point of view, and that the measurable evidence in support of either theory is one and the same, namely; history of other cases in the family.

Under usual circumstances the previous occurrence of leprosy in relatives of a patient is capable of interpretation in support of either conception. As a matter of fact, the two conceptions were actually derived from opposed interpretations from essen-

tially the same observations. On the one hand, the high degree of restriction of the disease to families in which there have been other cases, a restriction far exceeding that exhibited by any other known contagion, points strongly to familial susceptibility. On the other hand, the frequency with which other cases have been found in members of the immediate families of patients certainly introduces the possibility that patients have been subjected to more "prolonged and intimate exposure."

One approach to distinguishing between the two conceptions as determinants in the familial occurrence of leprosy would be a demonstration that "spread" within the family actually follows known laws of contagion or that the disease occurs within the family according to distinct laws of heredity. Because of the many vagaries which would be encountered, such as the lack of preciseness of incubation period, the impossibility of determining the actual degree of exposure, and the inability to collect sufficient data to establish Mendelian functions, it is not likely that any attempts at detailed study in this connection would succeed. But there is already one basic observation which would appear to be out of keeping with the operation of any known law of contagion, but more in keeping with the Mendelian operation of hereditary susceptibility—the occurrence of the disease and within families in males and females in a ratio of about two to one.

It would be only under exceptional circumstances that a history of leprosy in the relatives of patients could be more clearly interpreted as being indicative of the operation of one or the other of the two mechanisms under discussion. If a patient comes from a family in which leprosy has occurred but if he has not been exposed to his leprous relatives and develops the disease following exposure to other sources of infection, it would suggest familial susceptibility. If a patient who has no leprous relatives develops the disease following prolonged and intimate exposure it would suggest contagion.

As distinguished from familial leprosy in general where the individuals are at the same time blood relatives (heredity) and living in the same household (prolonged and intimate exposure), conjugal leprosy presents a situation which in a number of ways might provide an opportunity for differentiation between familial susceptibility and prolonged and intimate exposure as major determinants of the disease.

Conjugal mates of patients are not (as a rule) blood relatives. They live under conditions not of prolonged and intimate

exposure in the usual sense, from birth throughout their childhood (so much emphasized as a requirement for infection), but under conditions of exposure which are intimate to be sure but are limited both in point of age and of time.

Finally, the rarity of conjugal leprosy (as compared with familial leprosy in general) affords an opportunity for comparative studies of regional, racial, and familial factors in conjugal mates of patients who develop the disease and in those under similar marital circumstances of exposure who escape it.

The infrequency with which the disease is transmitted from patients to their spouses has usually been interpreted by those of the contagionist school of thought not so much as a failure of contagion but rather as an indication of adult insusceptibility to infection. As a matter of fact, the occurrence of a large portion of cases before adult life which has suggested greater susceptibility of young could equally well be interpreted merely as an indication of opportunity for exposure earlier in life (as in children in families with cases).

Under the ascendency of the contagionist point of view even the remote possibility of prolonged and intimate exposure (and there is almost always a history of exposure) has over-shadowed other factors such as familial susceptibility as the determining factor in the limited occurrence of disease amongst those who have been exposed. For example, the well known experimental inoculation of Arning has been considered doubtful because the inoculated patient "may have contracted his subsequently developing disease through (previous) contact with two infected relatives," although it does not seem to be a matter of record that the patient had actually been exposed to these relatives. Thus, a history of leprosy in relatives of a patient has come to be considered synonymous with prolonged and intimate exposure.

This point of view introduces a difficulty into the study of conjugal leprosy with reference to familial susceptibility or prolonged and intimate exposure. For example, a family history of leprosy in a conjugal mate who develops the disease would, in the mind of many, lead to the inference that the disease was the result of childhood exposure to relatives with leprosy rather than to conjugal exposure (in adult life, and because of familial susceptibility as indicated by a history of leprosy in relatives).

It is believed that a study of conjugal leprosy which takes into consideration both the interval after conjugal exposure and the age at which the disease occurs would make it possible to distinguish between prolonged and intimate exposure and fa-

milial susceptibility as the probable determinant in conjugal infection.

In a previous study it has been shown that leprosy in veterans of American wars followed distinct patterns as to time and age of development of the disease as well as to place of origin of patients, dependent on whether or not military service was seen in areas where there is a risk of exposure to infection(2). Veterans of the Spanish American War whose military service was in areas which are foci of the disease, for the most part came from areas within the United States which are not foci of the disease and developed their disease on the average of 23 years following their military service in these

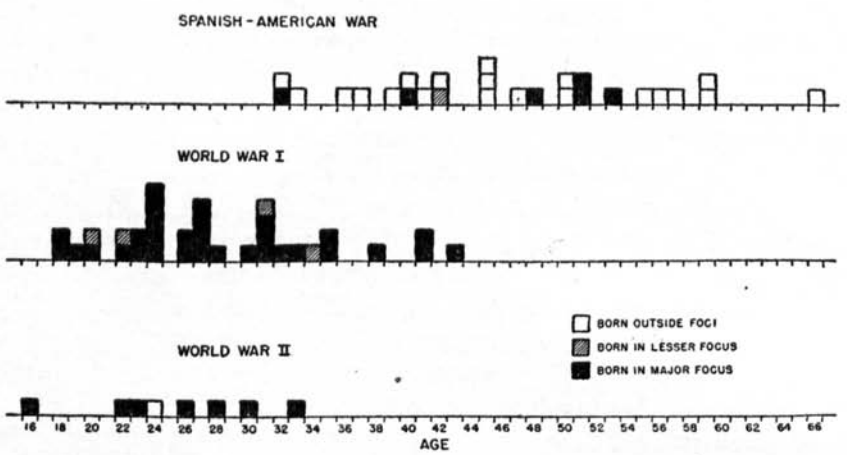

Chart 1. Leprosy in veterans of American wars in years following military service. (2) Spanish American War, average 23.7 years; World War I, average 6.5 years; World War II, average 2.4 years.

foci,* and at an average age of 46 years. In sharp contrast veterans of World War I (not in foci of leprosy) and World War II (so far) practically all came from areas in this country which are foci of the disease. On the average the disease has developed (or at least has been diagnosed) during their military service or within a very few years. The average age of these patients is 26 years. (Charts 1 and 2.) In respect to World War I, there is no reason for supposing that military service constituted a risk of exposure. It is believed that the occurrence

*This does not mean exposure was always confined to the brief period of the Spanish American War. Many of the veterans who developed the disease had remained in foci of the disease for years following the Spanish American War.

of a large proportion of these cases during their military service represents the recognition of disease which had already existed in them. This likewise applies to veterans of World War II who have been recognized as leprous up to the present time. It is anticipated that within a period of years additions to leprosy in World War II will come from veterans of campaigns which were in foci of the disease (the Pacific) and that they will follow the pattern of leprosy in Spanish American War veterans in respect to both time and age of development of disease as well as in respect to origin.

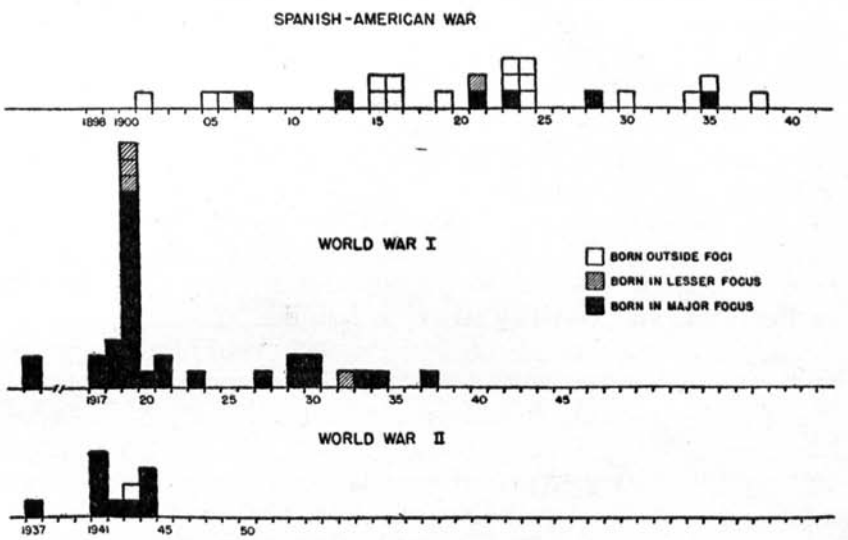

Chart 2. Leprosy in veterans of American wars according to age at which first symptoms occurred. (2) Spanish American War, average age 46.3 years; World War I, average age 27.9 years; World War II, 25.3 years.

It is clear from the fact that Spanish American War veterans coming largely from nonleprous regions where previous exposure can in all probability be excluded, received their initial exposure at the time of their military service. On the other hand, World War I and World War II veterans have been recognized as having the disease during or shortly after their military service but coming from foci of the disease had been exposed to infection in these foci long before entrance into military service. This is borne out by the fact that Spanish American War veterans developed the disease on an average of 24 years after their military service; while World War I and World War II veterans have developed the disease on the average of within

a few years after the beginning of their military service. That exposure to infection in Spanish American War veterans took place on an average of 20 years later in the lives of Spanish American War veterans than in the World War veterans is again indicated by the fact that the average age at which leprosy developed in Spanish American War veterans was 46 years; while in World War veterans it was 26 years of age.

Following the same pattern in the study of marital leprosy as was used in the study of military leprosy, it is anticipated that conjugal cases may likewise be divided into two groups which will enable a more precise evaluation of the influence of exposure and familial susceptibility. Thus, it is expected that there would be some cases developing within a short period of exposure to their conjugal mates and at younger ages which might rightly be attributable to previous exposure in their own families. On the other hand, if a second group of cases is found who develop their disease at a longer interval from exposure to their leprous spouses and at correspondingly older ages (like Spanish American War veterans) it would appear that the disease was contracted from their leprous spouses and not from exposure in earlier years to cases in their own families. In other words, a family history of leprosy in such patients could be interpreted more precisely as an indication of familial susceptibility. A final step in such a study would be a comparative study of family histories of cases in this category with that of the conjugal mates of leprous patients who, under similar conditions of exposure, do not develop the disease. The absence of the history of leprosy in the families of such individuals as well as their failure to contract the disease from similar exposure to their leprous spouses would be in keeping with the idea of familial susceptibility as the determinant.

It is anticipated that instances may be encountered, particularly in leprous areas, where spouses who actually come from leprous families do not contract the disease from their mates. Such individuals proving to be "insusceptible" both to exposure in their families and to conjugal exposure might in reality represent that portion of leprous families who, in accordance with genetic expectancy, are not themselves susceptible. In this connection exceptional instances of conjugal leprosy in individuals who have *not* been exposed to their leprous relatives would be instructive. On the other hand, particularly where spouses do not come from leprosy areas, instances may be encountered where the disease *is* contracted from leprous mates even though there is no family history. The absence of leprosy in such fami-

lies (although susceptible) might in reality be due to the fact that none had been previously exposed to infection.

Again exceptional instances may be encountered where conjugal leprosy occurs in an individual from a family living in localities where exposure to infection is unlikely but where other members have developed the disease following definite exposure in foci of the disease—as in certain of the cases in Spanish American War veterans.

It is believed that analysis of data in even a relatively small number of instances of conjugal leprosy, such as indicated on the accompanying questionnaire, and with special reference to the exceptional instances just mentioned, will shed light on the relative importance of prolonged and intimate exposure and heredity susceptibility as determinants in the occurrence of the disease.

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2. AYCOCK, W. L. and GORDON, J. E. Leprosy in veterans of American wars. Am. J. Med. Sci. **214** (1947) 329-339.

CONJUGAL LEPROSY

Date.....

	Husband			Wife		
Name (maiden name of wife)						
1.) Sex 2.) Race 3.) Descent	1.	2.	3.	1.	2.	3.
Address						
Year of Birth						
Place of Birth						
Year of Marriage						
Year Marital Living Terminated						
Has Leprosy Developed						
Year of Onset of Symptoms						
Year of Diagnosis						
Type of Disease						
History of Exposure—Other than to Husband or Wife						
Living in Locality where Leprosy Occurs						
Living in Household with a Case						
Other Contact						
Duration of Contact						
History of Leprosy in Relatives (Exact Relationship)						

Give any other details which may be pertinent, particularly with reference to History of Exposure and Leprosy in Relatives, on reverse side of this sheet.

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