THE DISAPPEARANCE OF LEPROSY IN A SEMI-ISOLATED POPULATION (NIIHAIU ISLAND, HAWAII)\textsuperscript{1}

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The "epidemic curve" of leprosy evolves over a period of many decades. Consequently, studies of the course of this disease in a particular population must deal with past events, and are therefore dependent on the accuracy and availability of old records. Hawaii is almost alone in having had a relatively high prevalence of leprosy during the past 7 or 8 decades with a continuing case-finding program, fairly good medical records, and good demographic data for the same period of time. The racial and socioeconomic composition of the population has radically changed during the last 100 years, thus complicating any attempt at analysis of the course of leprosy in Hawaii. Census data for Hawaii are shown in Fig. 1 (\textsuperscript{3}4).

The greatest changes in the racial composition of the Hawaiian population have taken place since the second quarter of the 19th century, during which period leprosy was introduced, probably from China (\textsuperscript{1}). The socioeconomic changes during the same period of time can be summarized by stating that, a century ago, 83 per cent of the people of Hawaii lived in rural areas, mostly as subsistence farmers, fishermen, or cowboys. The plantation system was just developing, and the influx of oriental plantation laborers had just begun. Today only 24 per cent of the people of Hawaii live in rural areas, mostly connected with the sugar or pineapple plantations (\textsuperscript{6}4). During the massive immigration (Fig. 1), many cases of leprosy were discovered in which the disease was undoubtedly acquired in the immigrants' homelands. These cases will be excluded from consideration here, and only those cases in native-born people of Hawaiian or part-Hawaiian ancestry will be considered. These have made up over 75 per cent of all cases of leprosy in Hawaii.

The island of Nihihiu offers a unique situation for epidemiologic studies in Hawaii. It is 72 square miles in area, and it lies some 20 miles to the leeward of the larger island of Kauai across a very rough channel. Nihihiu has no airfield or good harbor. It has been a cattle and sheep ranch entirely in the ownership of one family for the past 100 years. During this period it has had a population of 180 to 250 people, almost all of whom are of pure Hawaiian ancestry.

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These people have lived in the same pastoral setting in the same one or two small villages since about 1865, protected from most of the changes taking place in the rest of Hawaii. A few people have left Niihau for Kauai or Honolulu, mostly during the summer months, and this is largely limited to friends and relatives among the Hawaiian population in the adjacent Waiman district of Kauai (in which district Niihau is included for administrative purposes); that population numbered about 2,500 in 1860, but has now dwindled to about 250 (5).

The leprosy case-finding program in Hawaii began in 1865, was somewhat spasmodic until about 1890, but has been fairly consistently carried out since then. In the past all clinically active cases were isolated, but in recent years only bacteriologically positive cases. Since 1865 there has almost always been a government physician in residence in the Waiman district of Kauai, whose responsibilities have included the finding and reporting of cases of leprosy. Until well into the 1950’s a physician made trips across to Niihau fairly regularly to examine its population, but visits have been sporadic since then, and the people now go over to Waiman for medical care. Judging from the data presented below, it seems likely that before the turn of the century many of the people of Niihau, especially women and children, may have remained out of sight during these medical inspections, as happened in many places in Hawaii during earlier years (5).

A careful search of the records of the Hawaii Department of Health from its inception in 1850 through June 1961, plus correspondence and an interview with the owners of Niihau island, have disclosed a total of 34 cases of leprosy in people born there, including 3 cases.
diagnosed in Honolulu among people who had emigrated from Niihau.
No case has been reported from Niihau since 1933, or from among
Niihau-born people since 1939. During the same period of time, 87
cases of leprosy were reported among Hawaiians and part-Hawaiians
in the adjacent Waimea district, but none since 1940, and no cases
among immigrants in that district since 1954 (the last group of immi-
grants having arrived in 1946 from the Philippines).

COMPARATIVE RATES

As can be seen from Fig. 2, leprosy has disappeared from among
the approximately 250 people living in Niihau and from the approxi-
mately equal Hawaiian population living in the adjacent Waimea dis-
trict (\(1\)). The last case from either of these populations was reported
over 20 years ago. If the most recent state-wide rates for Hawaiians
and part-Hawaiians (0.59 cases per 1,000 population per year) were
applied, there would be at least 6 cases expected from each of these
two populations during this period of 20 years. It is exceedingly un-
likely that any such number of cases would have gone unreported un-
der modern conditions.

In the Waimea district there also live about 7,000 non-Hawaiian
people, largely of Japanese and Filipino descent. These non-Hawaiian

![Graph](image-url)
people formerly produced as many as 10 cases each decade (virtually all adult male immigrants), but in the last 25 years only one case has appeared, a 69-year-old man from Japan, diagnosed in 1954. It therefore appears that leprosy has stopped being transmitted in the entire population of this rural district of Kauai and the adjacent small island of Niihau.

Before 1880 the place of origin of patients was not consistently recorded; so the 3 cases shown in Fig. 3 during the 1870's undoubtedly represent only a part of those occurring in Niihau. The gap in reported cases during the 1880's probably represents a period when cases diagnosed on Niihau were segregated in a separate small village on that island, at the request of the people and owners of the island because of the reports of bad conditions then existing at the leprosarium on Molokai. The 10 cases reported from 1890 to 1899 all came out of Niihau on the same day in July 1893, and they probably represent the termination of this unwritten agreement with the Board of Health or its local agents (*). These cases were undoubtedly the survivors of a larger number accumulated during the previous 10 or 15 years.

Age.—After the turn of the century the cases came out of Niihau one or two at a time, and there was a sharp drop in their median age
at the time of diagnosis (Fig. 4). This pattern probably reflects a change in attitude toward case-finding and the isolation of cases, and lends confidence in the completeness of the data after 1899.

Examination of the records of the 21 cases reported from Niihau after 1899, when case-finding presumably became quite complete and relatively prompt, reveals that cases were removed from the island shortly after diagnosis. The age distribution of these 21 cases is presented in Fig. 5.

This curve, with its very sharp peak in the 10-19 age group, shows that almost all cases were being diagnosed and removed from the population before they were likely to have become parents. This curve also suggests strongly that most of these cases were exposed to leprosy as very young children. For comparison the age curves for two other groups—69 cases who were born in the Kalaupapa leper settlement and removed from contact by age 3 or 4 (1), and all 114 Hawaiian and part-Hawaiian cases reported in Hawaii during the decade 1950-1959—are presented in Fig. 6.

It is to be seen that of the cases exposed in their infancy in the leper settlement, virtually all were diagnosed during the first 3 decades of their lives, while of the total of Hawaiian and part-Hawaiian cases of the past decade, 38 per cent were diagnosed after age 29 (almost all in the early stages of the disease), and therefore probably had been
exposed at some age after infancy, when they had ventured outside of their immediate home environment. The secondary hump that appears in the curve of state-wide cases after age 40 almost certainly represents exposure during adulthood.

Sex ratio.—It is commonly stated (5, 7, 11) that in Hawaii there is a predominance in the number of male leprosy patients over females in the ratio of roughly 2:1. This holds true for the 13 Ni’ihau cases reported before 1900 (10 male and 3 female), but among the 21 cases reported since then, 10 were male and 11 were female, approximately a 1:1 ratio, and significantly different from the numbers expected in a 2:1 ratio (by chi-square test at the 1% level). Likewise, among the 144 Hawaiian and part-Hawaiian cases in the entire state in the past decade, only 53 were male, and 61 were female—also not far from a 1:1 ratio, but with females actually predominating (by 15%).

This apparent change in the sex ratio may represent a real change in the pattern of the disease in Hawaii, or it may merely mean that in former years, when the tendency was to hide leprosy cases as long as possible, it was easier to keep a woman at home and out of public notice than it was to hide a man. In the rather complete case-finding that has prevailed on Ni’ihau since 1899 and in the state as a whole during recent years (now that people have learned about the sulfone drugs) the unequal sex ratio has disappeared, lending support to the conclusion that it was an artifact.
Heredity and exposure.—Genealogic records of the people of Niihau are fairly well preserved back to 1860. There has been a considerable amount of intermarriage, as would be expected in this small, semi-isolated population; so it is probable that most of those alive today are not more distantly related than 4th or 5th cousins. In Fig. 7 are shown the year of birth and the year of diagnosis of each of the 21 cases of leprosy reported from Niihau since 1899.

These data add nothing definitive regarding the importance of genetic susceptibility in the acquisition of leprosy, but they do allow the very general observations that even in this small, relatively inbred population over one-third of the cases appear to have no direct, close genetic relationship with other cases, and that the disease has disappeared although the inbreeding has continued. The removal of most of the cases before the age of reproduction could scarcely be a powerful enough selective force to have accounted for the disappearance of the disease on a purely genetic basis in so short a time (a little over one generation). A careful study of these data shows that a few undetected cases of leprosy (perhaps only one) living in this small community from about 1885 to 1908 (see vertical lines in Fig. 7) could have directly exposed 15 of the 21 cases while they were still children, di-
rectly exposed 2 more cases as adults, and have been indirectly re-
sponsible for the remaining 4 cases by infecting the case marked
"uncle" in Fig. 7 in 1908, while he subsequently (1916-1919) infected
his two young nephews and the first two sporadic cases (see vertical
dashed line in Fig. 7).

Therefore, to explain the epidemiology of leprosy on Niulan during
its final 35 or 40 years, one does not have to invoke any obscure genetic
mechanism, any transmission of the disease through large number of
subclinical carriers, or any "exhaustion of susceptibles" in a continu-
onously-exposed population. One simply has to make the entirely reason-
able postulate that one or two cases were living in this small commu-
nity for a few years at the turn of the century. The age distribution
and chronologic evolution of the resulting cases can be explained on the
basis of the only two well-established facts about the epidemiology of
leprosy: (1) that the latent period after exposure to the appearance of
symptoms is long—usually between 5 and 15 years; and (2) that when
exposed equally, children are more susceptible to the disease than are
adults.
These data fit the hypothesis that, at about the turn of the century, the people or the owners of Niihau began to cooperate fully in the effort to isolate cases of leprosy as soon as they appeared. At that time quite a few people in the community had already acquired the infection, perhaps from a very few sources. Seventeen of these people, mostly children, progressed to the clinically active disease during the next two or three decades (last case in 1939), and were removed from the community as soon as they were discovered. One of these cases was an 11-year-old boy who was removed from Niihau in 1919, but not before exposing his two infant nephews and two other infants who subsequently developed the disease, the last one in 1939 at the age of 23. And that was the end of leprosy in this community, whose members have been protected from further infection by their relatively isolated situation and by the concomitant disappearance of leprosy in the Waimea district of Kauai, with which they have the greatest contact.

SUMMARY AND CONCLUSIONS

A review of leprosy records of Hawaii reveals that among Hawaiians and part-Hawaiians, who provide the bulk of the cases, the incidence per 1,000 population per year has fallen to 0.59 during the decade 1950-1959, but for the 250 or so Hawaiians on semi-isolated Niihau island and a like number in the adjacent Waimea district on the island of Kauai the disease has disappeared, with no case found in over 20 years. The disease has likewise virtually disappeared in the approximately 7,000 non-Hawaiian people (largely Japanese and Filipino) in the Waimea district, with only one case reported in the past 25 years.

The median age at the time of diagnosis of cases of leprosy from Niihau dropped sharply during the first decade of this century (to under 20 years of age), which probably marked the beginning of cooperation with the leprosy case-finding program. An analysis of the 21 cases from Niihau diagnosed from 1900 to 1939 (when the last case was found) shows that all but 4 were diagnosed before age 30 and were therefore presumably exposed during childhood. On the other hand, a similar analysis of all 114 Hawaiian and part-Hawaiian cases from the entire state during the decade 1950-1959 shows that 38 per cent were diagnosed after age 29, almost all in the early stages of the disease, and therefore they were presumably exposed as adults.

The 13 cases reported from Niihau before 1900 had a sex ratio of 10 males to 3 females, but among the 21 most recent cases (1900-1939) the sex ratio was 10 males to 11 females, approximately a 1:1 ratio, as is also true in the 114 recent state-wide cases mentioned. It is suggested that the predominance of male cases formerly reported in Hawaii was an artifact due to the greater ease of keeping women hidden from the authorities, and that this artifact disappears in the presence of co-
operation, as has been the case on Niilau since 1900, and in the state as a whole since the knowledge of the sulfone drugs has become general during the past decade.

A review of genealogic patterns of the 21 most recent Niilau cases shows that in this relatively inbred population 8 of the cases were sporadic, with no apparent close relationship with any other case. The other 13 cases were in four apparently unrelated families. Inbreeding has continued, but the disease has disappeared.

A close study of the history of these 21 cases shows that a case (or cases) of leprosy living in this small community from about 1895 to 1908 could have directly exposed 15 of them while they were children, 2 others as adults, and could have been indirectly responsible for the remaining 4 cases through one of the 15 children mentioned.

It is concluded that the disappearance of leprosy in this semi-isolated population can be adequately explained by the cooperation of the people in the isolation of cases as they appeared over a period of about 55 years. This same set of circumstances has now pertained for the entire state of Hawaii for about 15 years (large scale immigration from the Orient having ceased, and the sulfone drugs having been widely publicized to promote public cooperation). By analogy, this holds well for the disappearance of leprosy from the Hawaiian islands within the next 20 to 25 years.

RESUMEN Y CONCLUSIONES

Un repaso de las actas de la lepra en Hawaii revela que, entre los humanos y semi-humanos, que forman la mayoría de los casos, la incidencia por 1,000 habitantes al año ha bajado a 0,20 durante el decenio de 1950-1959, pero que para los 250 humanos más o menos en la isla de Niilau y un número semejante en el adyacente distrito de Waimea en la isla de Kauai, la lepra ha desaparecido, sin que se haya observado un caso en más de 20 años. Así también la enfermedad ha virtualmente desaparecido entre unos 7,000 sujetos no humanos (en gran parte japoneses y filipinos) del distrito de Waimea, no habiéndose denunciado más que un solo caso en los últimos 25 años.

La edad media para la fecha del diagnóstico de casos de lepra en Niilau descendió anualmente durante el primer decenio de este siglo (a menos de 20 años), lo cual probablemente señala la existencia de la cooperación con el plan de desinfección de casos de lepra. Un análisis de los 21 casos de Niilau diagnosticados de 1900 a 1939 (cuando se describió el último caso) muestra que todos menos 4 fueron diagnosticados antes de la edad de 20 años, y por lo tanto habían estado previamente expuestos durante la infancia. Por otro lado, un análisis semejante de otros 111 humanos y semi-humanos de todo el Estado durante el decenio de 1950-1959 muestra que 38 por ciento fueron diagnosticados después de la edad de 20 años, casi todos en los periodos inmediatos de la enfermedad, y por lo tanto habían estado expuestos siendo ya presumiblemente adultos.

Los 15 casos desconocidos de Niilau antes de 1900 mostraron una proporción sexual de 10 varones y 3 mujeres, pero entre los 21 casos más recientes (1900-1939) la proporción fue de 10 varones y 11 mujeres, aproximadamente una proporción de 1:1, lo cual reza también con los 111 casos recientes mencionados para todo el Estado. Se puede que el predominio de casos masculino descrito en Hawaii fue un artefato debido lo más fácil que es mantener a las mujeres escondidas de las autoridades, y que este artefato desaparece cuando existe cooperación, como ha sucedido en Niilau desde 1900, y en el
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Un estudio minucioso de la historia de estos 21 casos revela que un caso (o casos) de lepra viviendo en esta pequeña colectividad aproximadamente de 1880 a 1908 pudo haber expuesto directamente a 15 de ellos cuando eran niños, a 2 más cuando adultos, y haber sido indirectamente causante de los 4 casos restantes por conducto de uno de los 15 niños mencionados.

Debemos que cabe explicitar adecuadamente la desaparición de la lepra en esta población semi-ludá en la cooperación de la gente en el aislamiento de los casos al ir éstos apareciendo durante un periodo de unos 35 años. Esta misma serie de circunstancias ha intervenido ya para todo el Estado de Hainzi durante unos 13 años (habiendo escasa la inmigración en gran escala del Oriente y habiéndose divulgado todo lo relativo a las sulfonas para fomentar la colaboración del público). Por analogía, éste es un buen augurio de la desaparición de la lepra de las islas de Hainzi en términos de los próximos 20 a 25 años.

RESUME

D'après la compilation des archives de la lepré à Hainzi, il apparaît que parmi les hawaïiens de souche pure ou mis, qui forment la majorité des cas, Persévérence par 1,000 personnes par an est tombée à 0,29 durant la décennie 1956-1959. Par contre, la maladie a disparu chez les quelques 250 hawaïiens résidant sur l'île à demi isolée de Hainzi, et aussi chez ceux, d'un effectif semblable, qui habitent le district limitrophe de Waiman sur l'île de Kauai; aucun cas n'a été trouvé en plus de 20 ans. Parallèlement, la maladie a pratiquement disparu parmi les 7,000 non-hawaïiens environ (pour la plupart japonais et filipinos) qui peuplent le district de Waiman : un seul cas a été rapporté durant les 25 dernières années.

Chez les cas rapportés à Hainzi, l'âge moyen au moment du diagnostic de lepre est tombé au cours de la décennie de ce siècle (pour descendre jusqu'en dehors de 20 ans), ce qui marque probablement le début de la collaboration avec le programme de dépistage. Une analyse des 21 cas de Hainzi dépistés entre 1900 et 1939 (le dernier cas en date se situant cette année) indique que tous, sauf 4, ont été diagnostiqués avant qu'ils n'avaient pas 30 ans, suggérant par là qu'ils avaient vraisemblablement été exposés durant l'enfance. Par ailleurs, une analyse analogue, pour la décennie 1950-1959, des 114 hawaïiens de souche pure ou mis, pour l'ensemble de l'État, montre que 38% ont été diagnostiqués sous 20 ans, presque tous aux étades précoces de l'affection, ce qui permet de supposer clairement qu'ils avaient probablement été exposés à la maladie alors qu'ils étaient adultes.

Les 13 cas relevés à Hainzi avant 1900 comprennent, d'après le sexe, une proportion de 10 hommes pour 3 femmes, mais parmi les 21 cas plus récents (1900-1939), le rapport par sexe a été de 10 hommes pour 11 femmes, approximativement au rapport 1:1, semblable à celui qui a été ainsi trouvé pour les 114 cas rapporté récemment dans l'ensemble de l'État. On peut croire que la prédominance de l'élément masculin parmi les ces rapportés précédemment à Hainzi est un artefact du au fait qu'il était plus facile de soustraire les femmes aux autorités, et que cet artefact disparaît lorsqu'il y a collaboration, ainsi que c'est le cas à Hainzi depuis 1908, et dans l'ensemble de l'État depuis que, durant la décennie, la connaissance des médications sulfamées s'est étendue.

Une revue des arbres généalogiques des 21 derniers cas de Hainzi montre que, dans
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cette population à la consanguinité relativement importante, 8 des cas étaient spéciaux, sans apparente d'une relation étroite avec aucun autre cas. Les 33 autres cas sont survenus dans 4 familles qui semblent non apparentées. Les mariages consanguins ont continué, mais la maladie a disparu.

Une étude minutieuse de l'histoire de ces 21 cas nous a appris qu'un (ou plusieurs) malades atteints de lèpre, ayant vécu dans cette petite communauté depuis 1885 environ jusqu'à 1909, pouvaient avoir directement exposé 15 membres de la communauté alors que ceux-ci étaient enfants, 3 comme adultes, et qu'ils pourraient avoir été indirectement responsables des 4 cas restant par l'entremise d'un des 15 enfants mentionnés.

On en conclut que la disparition de la lèpre dans cette population semi-islâle peut être justement expliquée par la collaboration mise en place par ses ressortissants à isoler les cas au fur et à mesure qu'ils apparaissaient, durant plus de 35 ans à peu près. La même conjoncture existe à présent dans l'ensemble de l'État de Hawaï depuis environ 33 ans (l'immigration orientale sur une grande échelle ayant cessé, et les maladies surnuméraires ayant fait l'objet d'une vaste publicité visant à promouvoir la coopération du public),

On peut donc, par analogie, accepter l'augure de la disparition de la lèpre aux îles Hawaï d'ici les prochaines 20 à 25 années.

REFERENCES


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