Burden of Leprosy

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ABSTRACT

Hansen disease, also known as leprosy, is a curable infectious disease that continues to cause great disability and stigma globally, including in the United States. Nurse practitioners may believe that Hansen disease is a rare and ancient disease localized to underdeveloped countries, but it still exists in the US and should be included in the differential diagnosis. Hansen disease, although curable, can lead to deformity and disability if not diagnosed early. This article aims to raise awareness of Hansen disease by providing an introductory understanding of the disease process, diagnosis, and treatment.

Keywords: differential diagnosis, global health, Hansen disease, leprosy, neglected tropical disease

Hansen disease (HD), also known as leprosy, is a curable infectious disease of biblical notoriety that continues to cause great disability and stigma globally, including in the United States.[1] The World Health Organization (WHO) has designated HD as a neglected tropical disease (NTD). NTDs are a group of communicable diseases that predominately occur in tropical and subtropical regions among populations who live in close proximity to disease vectors and those struggling with poverty, poor hygiene, and community sanitation.[2] The designation of NTD provides a platform to bring awareness to providers and policy makers of diseases that cause significant global morbidity and mortality and allows for inclusion in international health agendas. The purpose of this article is to bring to light this neglected disease that continues to evade diagnosis, treatment, and eradication globally. The aim is to make nurse practitioners (NPs) more aware of HD, shed light on the great stigma that it brings, and stress the importance of accurate diagnosis and early treatment of this highly curable disease.

HD is a chronic mycobacterial infection caused by *Mycobacterium leprae*. Since the introduction of multidrug therapy in the early 1980s, there has been a decline in the incidence of HD.[2-4] The global burden has dropped from 5.2 million infected in 1985 to 211,973 new cases in 2015.[3,5] Although rates of infection have dramatically decreased, the disease remains endemic in many parts of the world, including the US. The largest number of cases in the US has been reported in Florida, California, Massachusetts, New York, Louisiana, Texas, and Hawaii.[6,7] There are approximately 6,500 people currently diagnosed with HD in the US, with half of those requiring active medication treatment.[8] The majority of new cases reported annually throughout the US are among populations born outside of the US; however, a low incidence of HD among indigenous residents persists.[7] There were 178 new cases reported in 2015 with 57% of patients born outside of the US.[7] Of the new cases reported in 2015, 96 cases were reported from Texas (21), Louisiana (16), Arkansas (2), Mississippi (2), Georgia (4), and Florida (49), and of those, 63 were indigenously acquired infections, occurring in people born in the US without a history of residing elsewhere.[7] In fact, the prevalence of HD in the aforementioned states has remained steady, except for Florida, where rates are increasing.[7]

EPIDEMIOLOGY

In 2015, there were 178 new cases of HD in the US, occurring primarily in Arkansas, California, Florida, Hawaii, Louisiana, Texas, and New York.[6] In the US, few new cases of leprosy occur each year, and misdiagnosis is common.[1,9] The subsequent delay in treatment can result in tissue damage and severe

disability, which can have lifelong ramifications for patients.[1,3,9] HD is curable with multidrug therapy. Permanent deformity and disability may be avoided with timely diagnosis and treatment.[1,3] When NPs and other health care providers lack familiarity with the presentation of HD, a delay in diagnosis occurs.

BACKGROUND

Although the occurrence of HD is tracked and epidemiologic data are analyzed by governmental organizations, there is little ongoing research in the US. Studies completed in the US evaluating the time between symptom onset and diagnosis of HD are infrequent and have focused solely on a single geographic location, clinic, or state. To date, there are no studies completed at a national level that evaluated the length of time between symptom onset and diagnosis. Leon et al[10] reported 30 Atlanta-area HD patients, 36% of whom had nerve damage at diagnosis, experienced a median length of time between symptom onset and diagnosis of 12 months. Marco et al[11] completed a study in Mississippi that included only 4 HD patients and revealed a time span of 5 to 16 months from the onset of a rash to diagnosis. Guerra[12] completed the next most recent study as a master's thesis and showed the average length of time from onset of symptoms to correct diagnosis for males and females in Texas was 4.04 and 4.3 years, respectively. Outside the US, a 2001 study conducted by Lockwood and Reid[9] showed that the diagnosis of HD is also delayed in the United Kingdom, with the mean of the onset of symptoms to correct diagnosis being 3.1 years.[13]

TRANSMISSION

Although HD has caused great deformity and disability since ancient times, the disease is still not well understood. The causative bacterium, *M leprae,* has an incubation period of approximately 5 years, with the presentation of symptoms taking as long as 20 years to appear.[3] *M leprae* is unable to be cultured, further hindering advancement in research.[14-16]

Within the scientific community, many questions remain regarding the transmission of HD. The long-held belief is that HD is spread from person to person through contaminated respiratory droplets,[2,16,17] in utero transplacentally and through breast milk, and through skin contact;[16] however, some recent studies point toward genetic variants that limit susceptibility to the bacterium to people carrying only certain genes.[16,18] The genetic predisposition to *M leprae* contributes to the differences seen in disease progression and symptom manifestation among infected patients.[1,19] The vast majority of individuals exposed to *M leprae* do not become infected.[19] Several genes have been identified as key to host resistance and may explain why the majority of the general population has an innate immune response against the disease[16,19]; additional research is needed into the genetic links to susceptibility.[16]

In the Southeastern US, an additional factor related to HD transmission must be taken into account—the presence of the 9-banded armadillo (Figure 1) as a native species.[20] The armadillo has proven to be an ideal reservoir because of its low body temperature in which *M leprae* can thrive.[15] Recent findings indicate that humans and 9-banded armadillos share the same strain of *M leprae*, indicating that HD is a zoonotic infection and is also a factor in the regional disease persistence.[12,20] Armadillo-to-armadillo transmission of *M leprae* occurs,[20] and the distribution limits of this armadillo have been expanding.[21] During the past few decades, states as far north as Missouri, Illinois, and Indiana, in addition to southern states of Kentucky, Tennessee, South Carolina, Arkansas, Georgia, Florida, Alabama, Mississippi, and Texas, are now home to the 9-banded armadillo.[21] Although the incidence of HD is low in the US, people living within the distribution limits of the 9-banded armadillo should be cautious. Although the transmission of *M leprae* can occur through fomites like soil, the vast majority of people infected by armadillos experienced extensive contact with the animals including direct contact with an armadillo while preparing the animal for cooking and consuming the animals.[15,20]

DIAGNOSIS

HD is known as a great imitator and may present with a variety of manifestations.[22] Classically, HD manifests with hypopigmented, hypoesthetic lesions and peripheral nerve enlargement, with or without neuropathy.[16] The initial presentation may also be as a nonhealing erythematous patch, poorly defined