Kuru

Kuru is a rare, degenerative, and fatal brain disorder that occurs primarily among the Fore natives in the highlands of New Guinea. The disease is thought to result from the practice of ritualistic cannibalism, in which relatives consume the tissues (including brain) of a deceased family member. Brain tissue from individuals with kuru is highly infectious, and can transmit the disease through either oral or peripheral routes of infection (e.g., cuts, sores). Cannibalism was prevalent before the 1960s, but government discouragement of the practice led to a continuing decline in the disease, which has now essentially disappeared. Kuru belongs to a class of infectious diseases called **transmissible spongiform encephalopathies** (TSEs), also known as prion diseases. Other TSEs include Creutzfeldt-Jakob disease in humans, fatal familial insomnia in humans, bovine spongiform encephalopathy in cattle (also known as mad cow disease), scrapie in sheep and goats, and chronic wasting disease in deer and elk. “Prion” is the name given to an abnormal form of protein that appears to be able to cause more and more of its normal form to become malformed, eventually leading to deposits in cells of the central nervous system, and eventually killing these cells. Prion diseases are most easily transmitted by direct transfer of infected tissue to the CNS, although (like kuru) they can also be transmitted orally.

TREATMENT

There are no treatments that can control or cure the disease, nor is there currently a diagnostic test for kuru or other TSEs. The only way to confirm a diagnosis of a TSE is through a brain biopsy or autopsy. In a brain biopsy, a neurosurgeon removes a small piece of tissue from the patient's brain for examination. Usually, this is done only to rule out other, potentially treatable, diseases, like encephalitis or meningitis that might mimic the symptoms of a TSE disease.

WHAT WILL HAPPEN

Like other TSEs, kuru has a long incubation period and it may take years or even decades before an infected person shows symptoms. Because kuru mainly affects the cerebellum, which is located at the base of the brain and is responsible for coordination, the usual first symptoms are those of incoordination, such as unsteady gait, tremors, and slurred speech. Unlike most of the other TSEs, dementia is either minimal or absent; instead, there is usually a marked change in mood, often taking the form of indifference, or even euphoria. Eventually, patients become unable to stand or eat, and they die in a helpless comatose state from 6 to 12 months after the onset of illness.

RESEARCH BEING DONE

The NINDS funds research grants to institutions throughout the country in an effort to learn how to better understand the genetic, molecular, and cellular mechanisms that underlie kuru and other TSE diseases.

Research is continually being done to learn more about this problem.

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