Creutzfeldt-Jakob Disease

National Institute of Neurological Disorders and Stroke (NINDS)

Source

Creutzfeldt-Jakob disease (CJD) is a rare, degenerative, fatal brain disorder. Onset of symptoms typically occurs at about age 60. There are three major categories of CJD: sporadic (the most common form, in which people do not have any known risk factors for the disease); hereditary (in which the person has a family member with the disease and tests positive for a genetic mutation associated with CJD), and acquired (in which the disease is transmitted by exposure to brain and nervous system tissue, usually through certain medical procedures). A form called variant CJD can be acquired by eating meat from cattle affected by a disease similar to CJD, called bovine spongiform encephalopathy (commonly called “mad cow” disease). CJD cannot be transmitted through the air or through touching or most other forms of casual contact. Initial symptoms of CJD include problems with muscle coordination, personality changes including progressive and impaired thinking and judgment, vision problems that may lead to blindness, and involuntary muscle jerks called myoclonus. People eventually lose the ability to move and speak, and enter a coma. Tests that help in the diagnosis of CJD include electroencephalography (which records the brain's electrical pattern), detection of certain proteins in the fluid that surrounds the brain and spinal cord, and magnetic resonance imaging. The only way to confirm a diagnosis of CJD is by brain biopsy or autopsy. A brain biopsy is discouraged unless it is needed to rule out a treatable disorder. CJD belongs to a family of diseases known as prion diseases--derived from “protein” and “infectious.”