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Subacute Sclerosing Panencephalitis

National Institute of Neurological Disorders and Stroke (NINDS)

Source

National Institute of Neurological Disorders and Stroke (NINDS). <u>Subacute Sclerosing</u> <u>Panencephalitis Information Page.</u>

Subacute sclerosing panencephalitis (SSPE) is a progressive neurological disorder of children and young adults that affects the central nervous system (CNS). It is a slow, but persistent, viral infection caused by defective measles virus. SSPE has been reported from all parts of the world, but it is considered a rare disease in developed countries, with fewer than 10 cases per year reported in the United States. The incidence of SSPE declined by at least 90 percent in countries that have practiced widespread immunization with measles vaccine. The incidence of SSPE is still high in developing countries such as India and Eastern Europe. There is a higher incidence among males than females (male/female: 3/1). Most youngsters with SSPE have a history of measles infection at an early age, usually younger than 2 years, followed by a latent period of 6 to 8 years before neurological symptoms begin. Despite the long interval between the measles infection and the onset of SSPE, researchers think that the infection of the brain occurs soon after the primary bout with measles and progresses slowly. Why it persists and progresses still isn't clear. The initial symptoms of SSPE are subtle and include mild mental deterioration (such as memory loss) and changes in behavior (such as irritability) followed by disturbances in motor function, including uncontrollable involuntary jerking movements of the head, trunk or limbs called myoclonic jerks. Seizures may also occur. Some people may become blind. In advanced stages of the disease, individuals may lose the ability to walk, as their muscles stiffen or spasm. There is progressive deterioration to a comatose state, and then to a persistent vegetative state. Death is usually the result of fever, heart failure, or the brain's inability to continue controlling the autonomic nervous system.