



NINDS Progressive Multifocal Leukoencephalopathy Information Page

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What is Progressive Multifocal Leukoencephalopathy?

Progressive multifocal leukoencephalopathy (PML) is caused by the reactivation of a common virus in the central nervous system of immune-compromised individuals. Polyomavirus JC (often called JC virus) is carried by a majority of people and is harmless except among those with lowered immune defenses. The disease occurs, rarely, in organ transplant patients; people undergoing chronic corticosteroid or immunosuppressive therapy; and individuals with cancer, such as Hodgkin's disease, lymphoma, and sarcoidosis. PML is most common among individuals with acquired immune deficiency syndrome (AIDS). Studies estimate that prior to effective antiretroviral therapy, as many as 5 percent of people with AIDS eventually developed PML. For them, the disease was most often rapidly fatal.

With current HIV therapy, which effectively restores immune system function, as many as half of all HIV-PML patients survive, although they sometimes have an inflammatory reaction in the regions affected by PML. The symptoms of PML are the result of an infection that causes the loss of white matter (which is made up of myelin, a substance that surrounds and protects nerve fibers) in multiple areas of the brain. Without the protection of myelin, nerve signals can't travel successfully from the brain to the rest of the body. Typical symptoms associated with PML are diverse, since they are related to the location and amount of damage in the brain, and evolve over the course of several days to several weeks. The most prominent symptoms are clumsiness; progressive weakness; and visual, speech, and sometimes, personality changes. The progression of deficits leads to life-threatening disability and death over weeks to months. A positive diagnosis of PML can be made on brain biopsy, or by combining observation of a progressive course of the disease, consistent white matter lesions visible on a magnetic resonance image (MRI) scan, and the detection of the JC virus in spinal fluid.

Is there any treatment?

Currently, the best available therapy is reversal of the immune-deficient state. This can sometimes be accomplished by alteration of chemotherapy or immunosuppression (even if it means losing non-vital transplanted organs). In the case of HIV-associated PML, immediately beginning anti-retroviral therapy will benefit most individuals.

What is the prognosis?

The mortality rates for those with HIV-PML have fallen dramatically from approximately 90 percent to around 50 percent according to most reports. For non-AIDS individuals with PML, the prognosis remains grim; the disease usually lasts for months and 80 percent die within the first 6 months, although spontaneous improvement has been reported. Those who survive PML can be left with severe neurological disabilities.

What research is being done?

The National Institute of Neurological Disorders and Stroke (NINDS) and other institutes of the National Institutes of Health (NIH) conduct research related to PML in laboratories at the NIH, and support additional research through grants to major medical institutions across the country. Much of this research focuses on finding better ways to prevent, treat, and ultimately cure disorders such as PML.

NIH Patient Recruitment for Progressive Multifocal Leukoencephalopathy Clinical Trials

- ▶ [At NIH Clinical Center](#)
- ▶ [Throughout the U.S. and Worldwide](#)

Organizations

[National Organization for Rare Disorders \(NORD\)](#)

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Publicaciones en Español

- ▶ [Leucoencefalopatía multifocal progresiva](#)

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