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Two case reports of rare stiff person syndrome

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Two female patients achieved clinical remission from the rare, debilitating neurological disease called stiff person syndrome (SPS, which can be marked by a "tin soldier" gait) after an autologous (from your own body) stem cell transplant that eventually allowed them to return to work and regain their previous functioning.

SPS is a disease characterized by stiffness of the skeletal muscles, painful muscle spasms and, in severe cases, the disease can prevent movement and walking. Autologous hematopoietic stem cell transplantation (auto-HSCT) has been used to successfully treat patients with autoimmune diseases such as multiple sclerosis and scleroderma, which are resistant to more conventional treatment. A regimen of high-dose chemotherapy and antilymphocyte antibodies rid the body of diseased immune cells (immunoablation) before the immune system is regenerated with auto-HSCs.

The Ottawa Hospital Blood and Marrow Transplant Program performed immunoablation and auto-HSCT on the two women with severe SPS based on a regimen used for multiple sclerosis.

One of the women was diagnosed in 2005 at age 48 after having progressive leg stiffness, spasms, falls and walking with a "tin soldier" gait. The auto-HSC was performed in 2009. One month after the transplant, her SPS symptoms were resolved and she was fully mobile six months after the transplant and returned to work and playing sports. She remains asymptomatic nearly five years after transplantation.

The second woman was an otherwise healthy woman who had had periodic leg muscle stiffness that lasted several hours and she was eventually diagnosed with SPS in 2008 at age 30 years. She had stopped working, driving and moved back in with her parents before undergoing auto-HSCT in 2011. Her post-transplant course was complicated by four periods of severe muscle spasms within 18 months of transplantation. The woman has been able to return to work and her previous activities. She has not had SPS symptoms in more than a year.

"To our knowledge, this is the first report documenting that immunoablation followed by auto-HSCT can produce long-lasting and complete remission of SPS." Sheilagh Sanders, M.D., of the University of Ottawa, Canada, and colleagues said in their *JAMA Neurology* article.

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