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Stiff man syndrome and related conditions.

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Source

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Abstract

The stiff man syndrome (SMS) and its variants, focal SMS, stiff limb (or leg) syndrome (SLS), jerking SMS, and progressive encephalomyelitis with rigidity and myoclonus (PERM), appear to occur more frequently than hitherto thought. A characteristic ensemble of symptoms and signs allows a tentative clinical diagnosis. Supportive ancillary findings include (1) the demonstration of continuous muscle activity in trunk and proximal limb muscles despite attempted relaxation, (2) enhanced exteroceptive reflexes, and (3) antibodies to glutamic acid decarboxylase (GAD) in both serum and spinal fluid. Antibodies to GAD are not diagnostic or specific for SMS and the role of these autoantibodies in the pathogenesis of SMS/SLS/PERM is the subject of debate and difficult to reconcile on the basis of our present knowledge. Nevertheless, evidence is emerging to suggest that SMS/SLS/PERM are manifestations of an immune-mediated chronic encephalomyelitis and immunomodulation is an effective therapeutic approach.

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