

eTable 1 Diagnostic criteria for stiff-person syndrome adopted from the work of Dalakas MC¹

Major criteria (all criteria must be present)
(1) Insidious onset of muscular rigidity in the limbs and axial muscles, most prominent in the abdominal and thoracolumbar paraspinals, with difficulty in turning or bending. †
(2) Superimposed painful spasms precipitated by unexpected noises, tactile stimuli, or emotional upset.
(3) Continuous motor unit activity in agonist and antagonist muscles, as confirmed clinically.
(4) Normal motor and sensory examinations except for difficulty in active movement. §
Minor criteria
(5) Positive for anti-GAD65 antibodies (or other autoantibodies ¶) in serum as assessed by immunocytochemistry, Western blot, enzyme-linked immunosorbent assay (ELISA), enzyme immunoassay (EIA), or radioimmunoassay (RIA).
(6) Electromyographic evidence of continuous motor unit activity in agonist and antagonist muscles.
(7) Clinical response to benzodiazepines or sleep.
Exclusion criteria
(8) Absence of other neurologic diseases that can explain stiffness and rigidity (e.g., dystonia, Isaacs' syndrome).
Clinical classification
Classic SPS, stiff-limb syndrome (SLS), progressive encephalomyelitis with rigidity and myoclonus (PERM)
Diagnostic categories
Definite: Major criteria + all items of Minor criteria + Exclusion criteria
Probable: Major criteria + 2 items of Minor criteria + Exclusion criteria
Possible: Major criteria + 1 item of Minor criteria + Exclusion criteria

† SPS may start focally from one lower limb.

§ PERM shows brainstem dysfunction or dysautonomia, aside from SPS features.

¶ Autoantibodies against amphiphysin, glycine receptor α_1 subunit, DPPX, gephyrin, and GABA_AR.