Stiff person syndrome (Moersch–Woltmann syndrome)

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Condition for which IVIg has an established therapeutic role.

Specific Conditions	• Stiff person syndrome
Indication for IVIg Use	• Treatment of significant functional impairment in patients who have a verified diagnosis of stiff person syndrome.
Level of Evidence	Evidence of probable benefit – more research needed (Category 2a)
Description and Diagnostic Criteria	Patients with stiff person syndrome present with symptoms related to muscular rigidity and superimposed episodic spasms. The rigidity insidiously spreads, involving axial muscles, primarily abdominal and thoracolumbar, as well as proximal limb muscles. Typically, co-contraction of truncal agonist and antagonist muscles leads to a board-like appearance with hyperlordosis. Less frequently, respiratory muscle involvement leads to breathing difficulty and facial muscle involvement to a mask-like face. Investigations that may be useful for diagnosis include auto-antibodies to GAD-65 or GAD-67, electromyography recordings from stiff muscles that may show continuous discharges of motor unit, and cerebrospinal fluid oligoclonal bands.
Justification for Evidence Category	The Biotext (2004) review included one randomised, double blind, placebo-controlled trial with a crossover design of 16 patients with stiff person syndrome and anti-GAD-65 antibodies. A significant treatment effect with intravenous immunoglobulin (IVIg) was seen, resulting in patients' decreased stiffness and heightened sensitivity scores. According to expert consensus, considering the disabling progressive course of stiff person syndrome, IVIg should be offered as the first-line treatment. Although periodic infusions would be required in the majority, further studies are needed to determine optimal dosage and duration (Kornberg 2004, Asia–Pacific Advisory Board).
Diagnosis Requirements	A diagnosis must be made by a Neurologist or a General Medicine Physician. Where the diagnosis was not made by a Neurologist the diagnosis must be verified by a Neurologist.
Qualifying Criteria for IVIg Therapy	Treatment of significant functional impairment in patients who have a verified diagnosis of stiff person syndrome. • Significant functional impairment as measured by stiffness, activities of daily living (ADL) or other functional/disability scale.

Review Criteria for Assessing the Effectiveness of IVIg Use	Review by a Neurologist is required within six months and, for stable patients on maintenance treatment, at least least annually. Clinical effectiveness can be demonstrated by objective findings of improvement in symptoms of stiffness. Documentation of clinical effectiveness is necessary for continuation of IVIg therapy. **On review of the initial authorisation period** Clinical effectiveness of Ig therapy may be demonstrated by: • Improvement in disability and stiffness compared to the qualifying assessment, as measured by activities of daily living (ADL) or other functional/disability/stiffness scales. **On review of a continuing authorisation period** Clinical effectiveness may be demonstrated by: • Improvement in, or stabilisation of, disability and stiffness compared to the previous review assessment, as measured by activities of daily living (ADL) or other functional/disability/stiffness scales.
Dose	• **Induction Dose** - 2 g/kg in 2 to 5 divided doses. • **Maintenance Dose** - 1-2 g/kg 4-6 weekly. Aim for the minimum dose to maintain optimal functional status. **The aim should be to use the lowest dose possible that achieves the appropriate clinical outcome for each patient.** **Dosing above 1 g/kg per day is contraindicated for some IVIg products.** **Refer to the current product information sheet for further information.**

Bibliography

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