

Motor and/or Sensory Neuropathies

Overview

Ganglioside Autoantibodies

Autoantibodies to ganglioside monosialic acid (GM₁) are present in 50% of patients with multifocal motor neuropathies (MMN) and chronic inflammatory demyelinating polyneuropathy (CIDP),^{1,2} with 100% specificity based on a cutoff to rule out low titer autoantibodies seen in amyotrophic lateral sclerosis (ALS).³ **Because clinical utility depends on the assay system used, Specialty employs the most analytically sensitive EIA protocol.**⁴ GM₁ autoantibodies can cause destruction of motor neurons, myelin, muscle spindles, motor end plates and axons.⁵ Asialo-GM₁ autoantibodies are found predominantly in lower motor neuron syndromes with early weakness in proximal muscles.⁶ GD_{1b} autoantibodies are found in some sensory dominant and sensorimotor polyneuropathies, in the absence of GM₁ reactivity.⁷ GD_{1a} and GQ_{1b} autoantibodies are reported in patients with Guillain-Barré syndrome (GBS), Miller-Fisher syndrome, dementia, inner ear disease, ALS, Bickerstaff encephalitis and sensory polyneuropathy.⁸⁻¹²

MAG Autoantibodies

Monoclonal reactivities against myelin-associated glycoprotein (MAG) are detected in 50% of patients with IgM paraproteinemia and sensory polyneuropathy.^{13,14} Non-para-protein (polyclonal) IgM MAG autoantibodies are detected in some patients with multiple sclerosis, GBS, chronic polyneuropathy, inner ear disease and myasthenia gravis. Low concentrations of IgM MAG autoantibodies are also detected in some patients with SLE or rheumatoid arthritis and in normal individuals.^{15,16} The detection of ganglioside or MAG autoantibodies in patients with peripheral neuropathy helps identify a subset of patients likely to respond to treatment by plasmapheresis or immunosuppressive drugs.¹⁷⁻¹⁹ MAG autoantibodies should be confirmed by immunoblot** because ganglioside autoantibodies can bind to MAG antigen.²⁰

SGPG Autoantibodies

Autoantibodies to SGPG (sulfoglucuronyl paragloboside) can be detected in sensory and sensorimotor neuropathies, CIDP with or without gammopathy, GBS and to a lesser extent motor neuron disease.²¹⁻²³ Differences in autoantibody fine specificities exist; hence, not all MAG-reactive sera are SGPG-positive and vice versa.^{24,25} To enhance sensitivity, *Specialty* assays SGPG and MAG autoantibodies separately.

Neuronal Nuclear (Hu) Autoantibodies

Neuronal nuclear autoantibodies are present in some patients with paraneoplastic sensory neuropathy and are usually associated with an underlying small-cell lung cancer.^{26,27}

Monoclonal Gammopathies

Immunofixation Electrophoresis (IFE) is the method of choice for detection of paraproteins in the CSF of patients with peripheral neuropathies.^{1,28} A new method, Capillary Electrophoresis and Immunosubtraction (CE/I), is being used for serum samples, giving the same accuracy as IFE with better throughput. Very high MAG or GM₁ autoantibody concentrations are commonly due to the monoclonal immune response to MAG or GM₁. In patients with multifocal motor neuropathies, decreases in GM₁ autoantibodies following treatment with immunosuppressive agents correlate with marked clinical improvement.²⁹ MAG autoantibodies reduce MAG content in myelin and are used to differentiate monoclonal gammopathy of undetermined significance (MGUS) from CIDP.^{30,31}

Carbohydrate-deficient Transferrin UltraQuant[®]

CDT is present in high concentrations in the serum of alcohol misusers.³² *Specialty's* research has shown that nearly 20% of serially received specimens for peripheral neuropathy testing are positive for CDT in the absence of autoantibodies, suggesting an alcohol-related etiology.³³ Alcohol misuse can cause both non-inflammatory myopathies and idiopathic neuropathies.³³

Ordering Information & Specimen Requirements

Test Code	Test Name	Specimen Requirements
4020	Motor & Sensory Neuropathy EvaluatR[™] GM ₁ ? Asialo-GM ₁ ? MAG IgM ? SGPG IgM ? GD _{1a} ? GD _{1b} ? GQ _{1b} ? Hu Autoabs ? IFE Paraproteins ? CDT UltraQuant [®]	6 mL Serum; Ambient, Refrigerated or Frozen. Collect by venipuncture, clot and separate within 4 hrs.

Related Tests

- 4030 Sensory Neuropathy EvaluatR™
CDT UltraQuant® ? IFE Paraproteins ? MAG IgM ? Neuronal Nuclear (Hu) ? SGPG IgM Autoabs
- 4026 Motor Neuropathy EvaluatR™
CE/I Paraproteins ? GM₁ ? MAG IgM ? GQ_{1b} ? Asialo-GM₁ ? GD_{1a} ? GD_{1b} ? SGPG Autoabs

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