PRIMEVIEW

NEUROMYELITIS OPTICA

nature reviews disease primers

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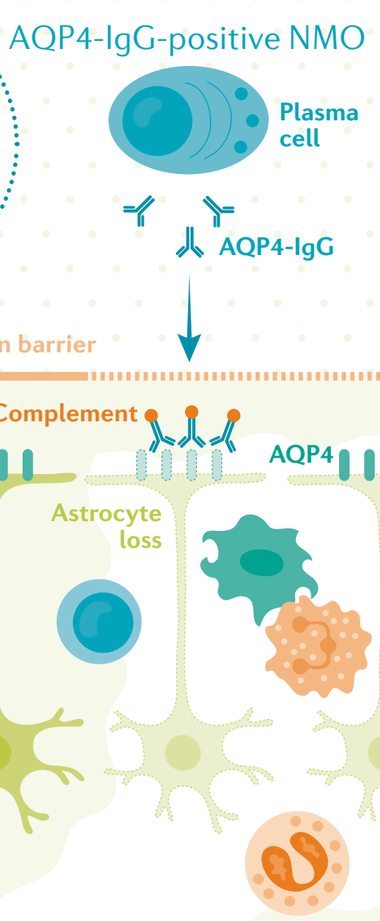
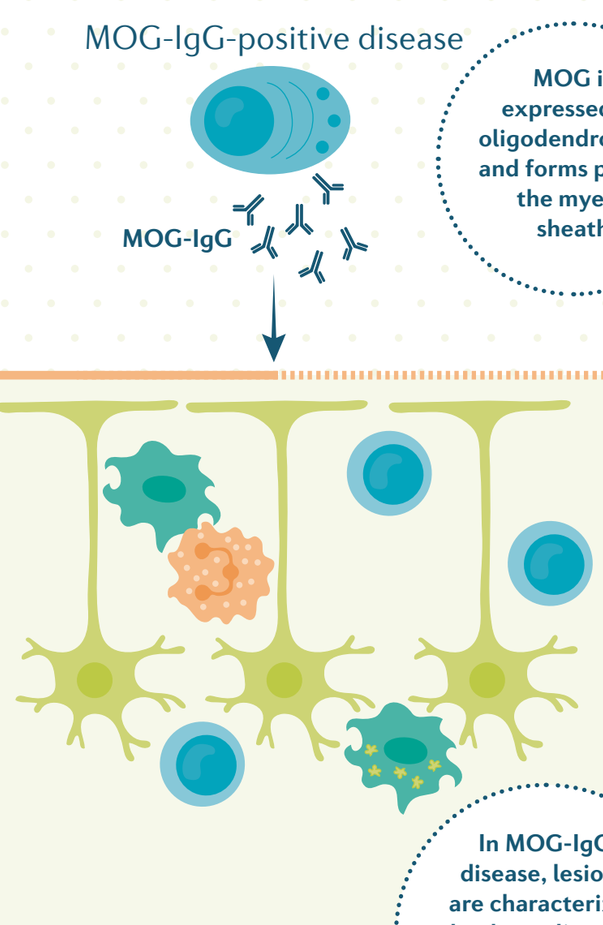
Neuromyelitis optica (NMO) is an autoimmune condition of the central nervous system. More than 80% of patients have antibodies to aquaporin 4 (AQP4-IgG); 10–40% of the remaining patients have antibodies to myelin oligodendrocyte glycoprotein (MOG-IgG), whereas others are seronegative for both AQP4-IgG and MOG-IgG.

MECHANISMS

AQP4 is a water channel protein that is concentrated in the astrocytic endfeet





IgG and complement deposits, and infiltration of various immune cells, are found in both AQP4-IgG NMO and MOG-IgG-positive disease

DIAGNOSIS

NMO is characterized by attacks of optic neuritis (inflammation of the optic nerve) and transverse myelitis (inflammation of the spinal cord). In addition, symptomatic brainstem and brain involvement can occur, both in patients with MOG-IgG and in those with AQP4-IgG. Accurate diagnosis is important to differentiate NMO from other neurological disorders with similar presentations, such as multiple sclerosis. Diagnosis includes MRI of the brain, spinal cord and optic nerve to identify lesions, and serology to detect autoantibodies. Cerebrospinal fluid cytology, optical coherence tomography, fundoscopy and electrophysiology can also be used to assist diagnosis.

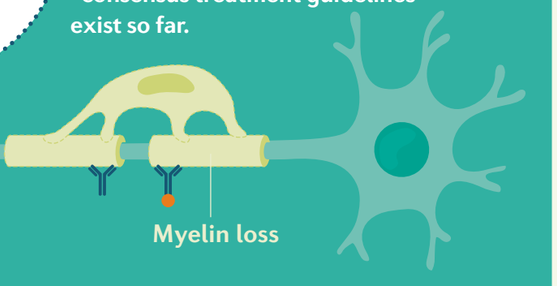
EPIDEMIOLOGY

The median age at onset is ~40 years but is lower in non-white individuals and those with MOG-IgG disease; AQP4-IgG-positive NMO shows a strong female preponderance (1:9). Risk factors for NMO include female sex, variants in genes encoding HLA and, possibly, low vitamin D levels. Infectious diseases precede the first attack in some patients, although no specific infections have been strongly linked to NMO onset or recurrence.

! The interval between attacks of both AQP4-IgG-positive NMO and MOG-IgG-positive disease varies from few months to several years. Some patients with MOG-IgG disease have a monophasic disease course.

MANAGEMENT

Acute attacks can be treated with intravenous methylprednisolone (IVMP) as first-line therapy and with plasma exchange or immunoadsorption as an early escalatory treatment in patients who do not sufficiently respond to IVMP. Immunosuppressants can be used to reduce the risk of relapse. Several phase III trials have been completed, which have resulted in FDA approval of three new long-term therapies. However, no consensus treatment guidelines exist so far.



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Written by Louise Adams; designed by Laura Marshall