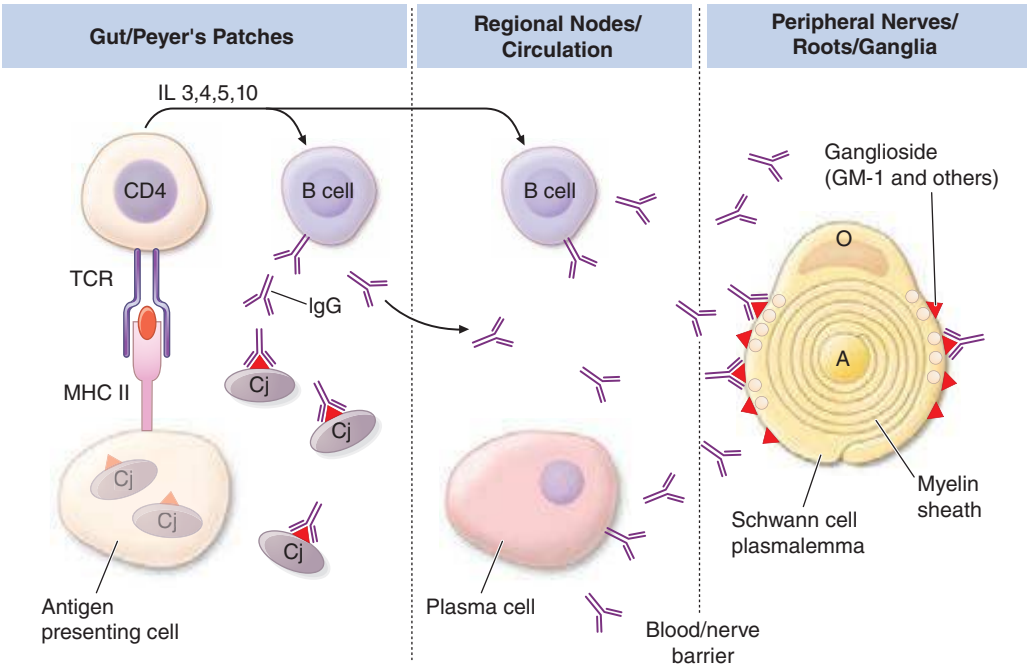

FIGURE 460-1 Postulated immunopathogenesis of Guillain-Barré syndrome (GBS) associated with *Campylobacter jejuni* infection. B cells recognize glycoconjugates on *C. jejuni* (Cj) (*triangles*) that cross-react with ganglioside present on Schwann cell surface and subjacent peripheral nerve myelin. Some B cells, activated via a T cell–independent mechanism, secrete primarily IgM (not shown). Other B cells (*upper left side*) are activated via a partially T cell–dependent route and secrete primarily IgG; T cell help is provided by CD4 cells activated locally by fragments of Cj proteins that are presented on the surface of antigen-presenting cells (APCs). A critical event in the development of GBS is the escape of activated B cells from Peyer's patches into regional lymph nodes. Activated T cells probably also function to assist in opening of the blood-nerve barrier, facilitating penetration of pathogenic autoantibodies. The earliest changes in myelin (*right*) consist of edema between myelin lamellae and vesicular disruption (*shown as circular blebs*) of the outermost myelin layers. These effects are associated with activation of the C5b-C9 membrane attack complex and probably mediated by calcium entry; it is possible that the macrophage cytokine tumor necrosis factor (TNF) also participates in myelin damage. A, axon; B, B cell; MHC II, class II major histocompatibility complex molecule; O, oligodendrocyte; TCR, T cell receptor.

TABLE 460-2 PRINCIPAL ANTIGLYCOLIPID ANTIBODIES IMPLICATED IN IMMUNE NEUROPATHIES

Clinical Presentation	Antibody Target	Usual Isotype
Acute Immune Neuropathies (Guillain-Barré Syndrome)		
Acute inflammatory demyelinating polyneuropathy (AIDP)	No clear patterns	IgG (polyclonal)
	GM1 most common	
Acute motor axonal neuropathy (AMAN)	GD1a, GM1, GM1b, GalNAc–GD1a (<50% for any)	IgG (polyclonal)
Miller Fisher syndrome (MFS)	GQ1b (>90%)	IgG (polyclonal)
Acute pharyngeal cervicobrachial neuropathy (APCBN)	GT1a (? most)	IgG (polyclonal)
Chronic Immune Neuropathies		
Chronic inflammatory demyelinating polyneuropathy (CIDP) (75%)	P0, myelin P2 protein, PMP22, neurofascin	No clear pattern
CIDP-M (MGUS associated) (25%)	Neural binding sites	IgG, IgA (monoclonal)
Chronic sensory > motor neuropathy	SPGP, SGLPG (on MAG) (50%)	IgM (monoclonal)
	Uncertain (50%)	IgM (monoclonal)
Multifocal motor neuropathy (MMN)	GM1, GalNAc–GD1a, others (25–50%)	IgM (polyclonal, monoclonal)
Chronic sensory ataxic neuropathy	GD1b, GQ1b, and other b-series gangliosides	IgM (monoclonal)

Abbreviations: CIDP-M, CIDP with a monoclonal gammopathy; MAG, myelin-associated glycoprotein; MGUS, monoclonal gammopathy of undetermined significance.

Source: Modified from HJ Willison, N Yuki: Brain 125:2591, 2002.

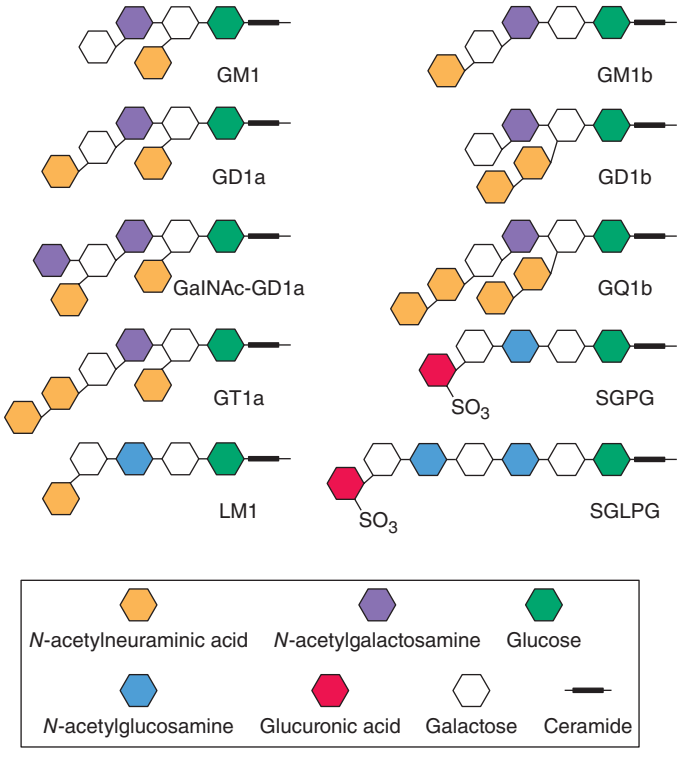

FIGURE 460-2 Glycolipids implicated as antigens in immune-mediated neuropathies. *(Modified from HJ Willison, N Yuki: Brain 125: 2591, 2002.)*