

after a viral encephalitis (herpes simplex or Japanese encephalitis); (2) some syndromes predominate in young individuals and children; (3) despite the severity of the symptoms, patients usually respond to treatment of the tumor, if found, and immunotherapy (e.g., glucocorticoids, IVIg, plasma exchange, rituximab, or cyclophosphamide); and (4) for many of these disorders, the antibody pathogenicity has been demonstrated in models using cultures of neurons or passive transfer of patients' antibodies to animals (**Fig. 94-3**).

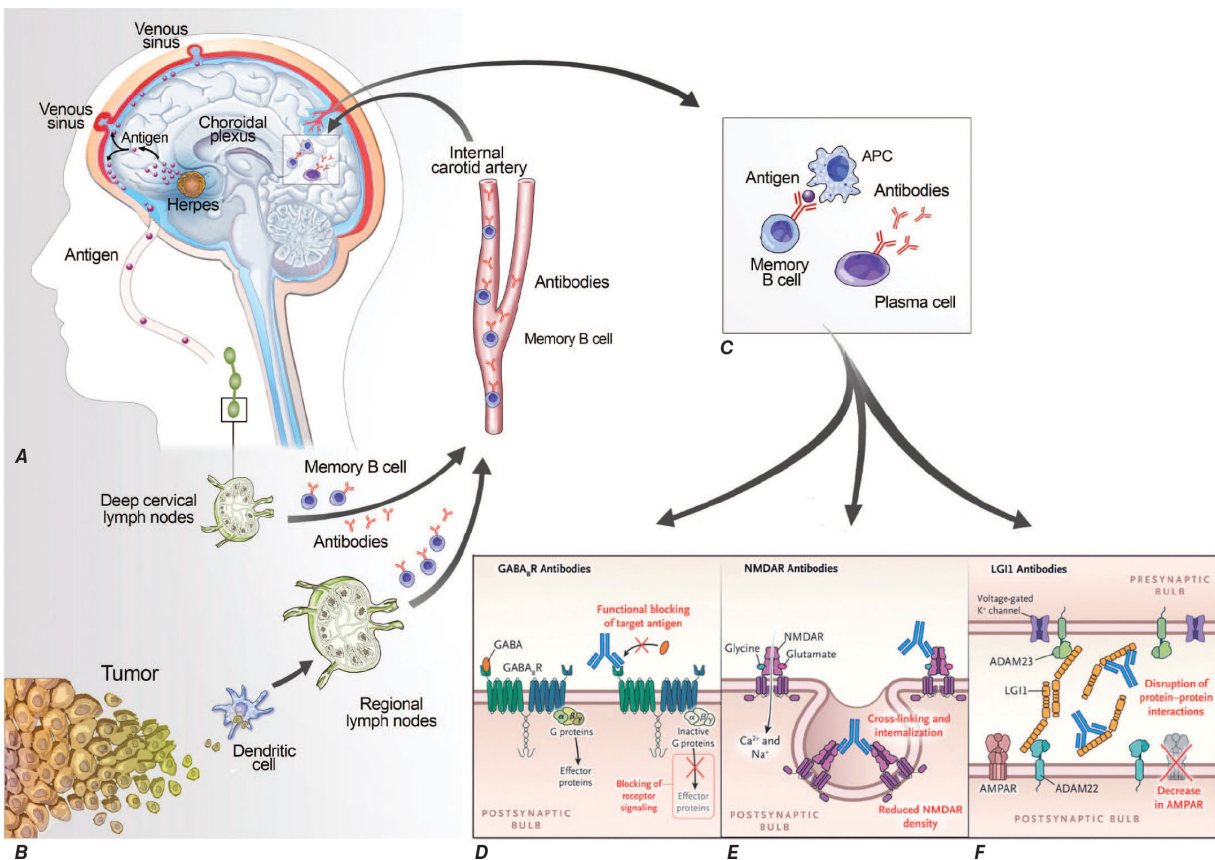


FIGURE 94-3 Proposed mechanisms of disease and functional interactions of autoantibodies with neuronal proteins. The graph shows a multistep process that results in antibody-mediated neuronal dysfunction; some of the steps have been demonstrated in reported studies, whereas others are based on proposed hypotheses. Two well-known triggers of autoimmune encephalitides are represented: herpes simplex encephalitis (**A**) and systemic tumors (**B**); the genetic susceptibility of some autoimmune encephalitides and unknown immunologic triggers are not depicted. It is postulated that antigens released by viral-induced neuronal destruction or apoptotic tumor cells are loaded into antigen-presenting