

Autoantibody-mediated brain disorders are a relatively novel disease category, providing clinicians with a series of immunotherapy-responsive diseases, many of which are associated with clinically-characteristic findings. Here, I will describe the clinical and laboratory features of these 'not to miss' diseases, with particular attention to some of the most recent observations.

The commonest autoantibody-mediated brain diseases are associated with autoantibodies to leucine-rich glioma inactivated 1 (LGI1) and the NMDA receptor. Patients with LGI1-antibodies have a variety of frequent focal seizure semiologies, of which Faciobrachial dystonic seizures (FBDS) are the most distinctive. In addition, around 75% of patients also have prominent amnesia as part of a limbic encephalitis. The seizures show a rapid response to immunotherapies, but are relatively refractory to anti-epileptic drugs (AEDs). Furthermore, seizure cessation is associated with the prevention of otherwise incipient cognitive impairment, and hence improved long term recoveries (Fig. 1). Patients with LGI1-antibodies have a very strong association with HLA-

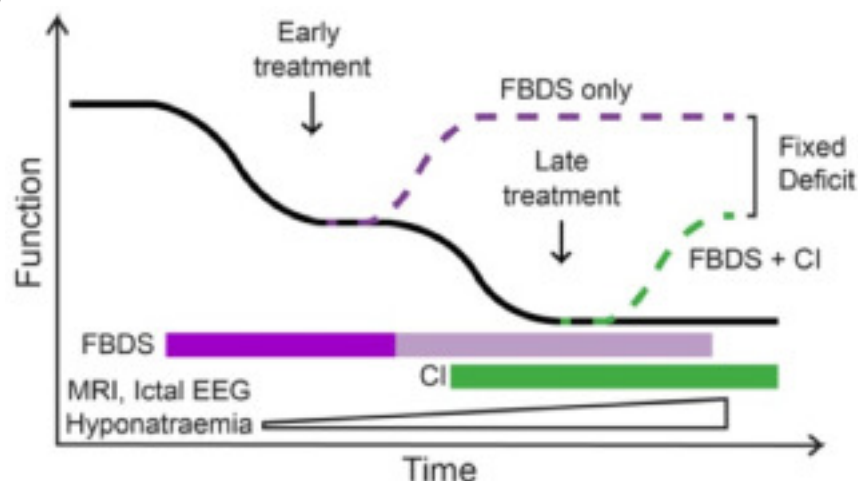


Figure 1. From Thompson et al 2018, Brain

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NMDA-receptor antibodies are typically found in young patients with psychiatric features and a movement disorder. Recent clinical characterisations have revealed that the movement disorder is complex and typically a combination of dystonia, stereotypes and chorea. Also, the psychiatric features are similarly complex and span a number of typical DSM-IV based diagnostic categorisations. These features make the disorder highly-distinctive and clinically-recognisable.

The other autoantibody mediate brain disorders will be discussed more briefly.