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TitleOutcome of early immunotherapy in 103 patients with faciobrachial dystonic seizures

BodyFaciobrachial dystonic seizures (FBDS) and limbic encephalitis closely associate with LGI1-antibodies. FBDS have a clinically-recognisable semiology and often predate the onset of the cognitive impairment (CI). We describe 103 consecutive patients with FBDS and LGI1-antibodies and characterise the clinical, therapeutic and serological differences between those with and without cognitive impairment (CI), and to determine whether cessation of FBDS can prevent CI. The 22/103 patients without CI typically had normal brain imaging, EEGs, sodium levels ($p<0·0001$) and almost exclusive IgG4 LGI1-antibodies ($p=0.009$), versus the frequent IgG1-antibodies in patients with CI ($p=0.03$). Overall, cessation of FBDS with antiepileptic-drugs alone occurred in only 9/89 (10%) patients. In contrast, 30 days after immunotherapy initiation, 51% had cessation of FBDS ($p<0·0001$); an effect which was more rapid in those without CI ($p=0.038$). Every week of delayed immunotherapy conferred a 5% relative reduction in the probability of FBDS cessation, and shorter time to immunotherapy ($p=0·031$) and absence of CI ($p=0·0014$) predicted reduced disability at 24-month follow-up. Furthermore, of 80 patients with FBDS as their initial feature, 56% developed CI after 90 days of active FBDS. In contrast, only one patient developed CI after cessation of FBDS ($p<0·0001$). FBDS show a striking time-dependent response to immunotherapy. Prompt immunotherapy appears to prevent development of CI, maybe via inhibiting IgG1-mediated complement deposition.

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