

# Teaching Video NeuroImages: Acquired focal neuromyotonia in LGI-1 autoimmunity

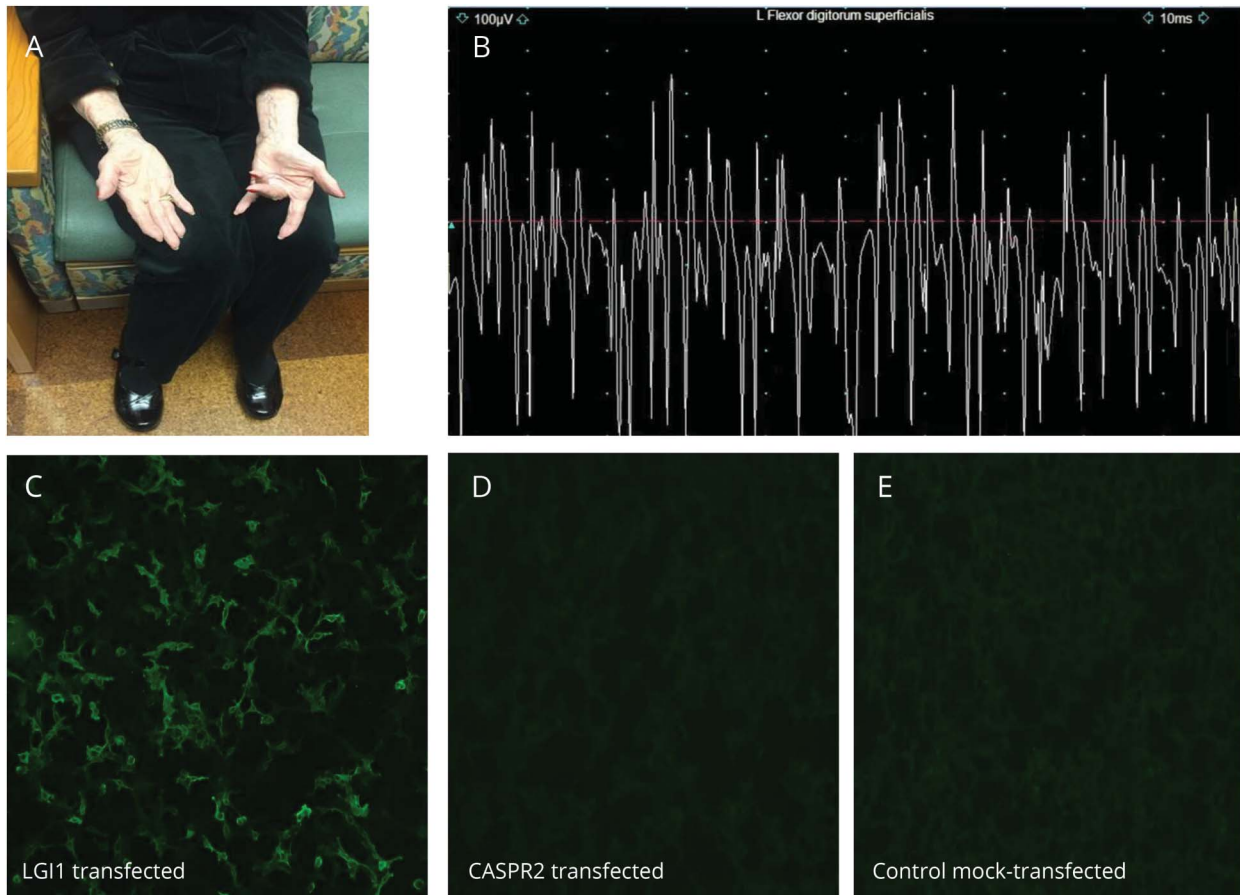
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**Figure** Physical examination, EMG, and serologic testing



(A) Persistent left III and IV digit flexion. (B) EMG shows abnormal high intraburst frequency discharges. Serum was tested for immunoglobulin G (IgG) reactive with LGI1 or CASPR2 proteins by a cell-based immunofluorescence assay using as substrate fixed HEK 293 cells that were nontransfected or transfected with plasmid-encoding human LGI1 or CASPR2 proteins. We detected bound IgG by use of fluorescein isothiocyanate conjugated goat IgG specific for human IgG. Serum is reactive with LGI1 transfected cells (C), but not CASPR2 transfected cells (D) or control mock-transfected cells (E).

An 81-year-old woman was diagnosed with corticobasal degeneration (cognitive decline and left hand posturing). Hyponatremia and bilateral faciobrachial dystonic seizures (FBDS) ensued. Neurologic examination (figure, A; video) revealed persistent involuntary continuous left 3rd and 4th finger flexor activation and delayed relaxation. EMG revealed high-frequency spontaneous discharges, including neuromyotonia and fast myokymia of the left flexor digitorum superficialis and pronator teres (figure, B). LGI1 immunoglobulin G (IgG)<sup>1</sup> was detected, but not CASPR2-IgG (figure, C–E). Oncologic evaluation revealed

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breast ductal carcinoma in situ. Encephalopathy and FBDS resolved after IV immunoglobulin. Encephalopathy coexisting with peripheral nerve hyperexcitability can mimic neurodegeneration, and should prompt exclusion of LGI1/CASPR2 autoimmunity.<sup>2</sup>

### Author contributions

Dr. Lopez contributed to the conception and design of the study; collection, analysis, and interpretation of the data; drafting and critical revision of the article; and generation/collection of the figures. Dr. Matsumoto contributed to the conception and design of the study; collection, analysis, and interpretation of the data; and generation of the figures. Dr. Sorenson contributed to the conception and design of the study; collection, analysis, and interpretation of the data; and generation of the figures. Dr. Klein contributed to the conception and design of the study; collection, analysis, and interpretation of the data; and generation of the figures. Dr. McKeon contributed to the conception and design of the study; collection, analysis, and interpretation of the data; drafting and critical revision of the article; and generation/collection of the figures. All authors gave final approval of the article.

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