Efgartigimod as Add-On Treatment in Refractory Guillain–Barré Syndrome: *A Case Report*

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Background
Despite the efficacy of standard treatments, that is, plasma exchange (PLEX) on alternate days for five exchanges or 2 g/kg intravenous immunoglobulin (IVIg), about 20% of Guillain–Barré syndrome (GBS) cases evolve to flaccid tetraparesis and need mechanical ventilation [1]. In such cases, available data do not support additional benefits from a second course of IVIg or PLEX, or from the combination of PLEX and IVIg. Efgartigimod (EFG) is a monoclonal antibody that binds the FcRn receptor and increases the degradation of IgG antibodies [2].

Case report
A 60-year-old healthy woman, who received flu vaccination 7 days before, developed in 6 h a severe flaccid tetraparesis with generalized areflexia, stocking-glove hypoesthesia, progressive dysphonia, dysphagia, and respiratory failure. Within 12 h, the patient was bedridden and required mechanical ventilation in the Intensive Care Unit (**Fig. 1**).

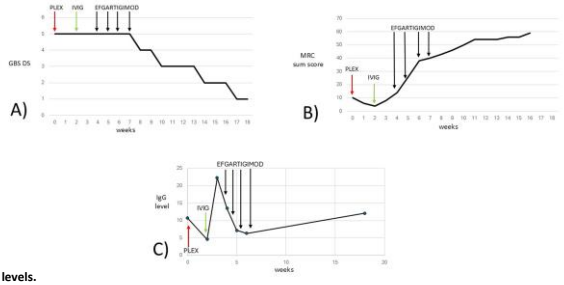

Figure 1. Evolution of disease course, muscle strength, and serum IgG levels.
The *x*-axis shows the time after admission (=week 0). The effect of different treatments on patient disability, represented by GBS disability score. The effect of different treatments on patient muscle strength, represented by the MRC sum score. The nadir of weakness was reached after 2 weeks. Serum IgG level was normal at admission, reduced after PLEX, increased after IVIg, and then progressively reduced after EFG infusions. IgG level was normal again after 10 weeks.

The patient's medical history was unremarkable. Nerve conduction studies (NCS) (**Fig. 2**) showed an increase in distal latencies, a reduction in conduction velocities, and reduced persistence of F waves. The sural nerve sensory action potential (SAP) was normal. Cerebrospinal fluid analysis, performed 6 h after admission, showed normal findings, whereas whole spinal cord MRI showed gadolinium enhancement of the cauda equina. The patient was admitted to the Intensive Care Unit because she was tetraplegic, requiring intubation, mechanical ventilation, and nasogastric tube for nutrition. The next morning, PLEX 3 L/session was started on alternate days, for five times (**Fig. 1**). Tracheostomy was performed 8 days after admission, and gastrostomy was positioned 14 days after admission. At the end of PLEX treatment, the patient was still severely tetraparetic and mechanically ventilated (**Fig. 1**). Blood tests and serological tests (IgM and IgG for CMV, EBV, Mycoplasma Pneumoniae, and HEV) were all negative. Serum collected at admission was tested for anti-ganglioside antibodies and antibodies directed against nodal/paranodal structures, with negative results. At day 14, NCS showed inexcitable nerves in the lower limbs and severe reduction of CMAP of median and ulnar nerves (**Fig. 2**). The SAP of median and ulnar nerves was bilaterally absent. Needle EMG showed spontaneous activity in the right tibialis anterior muscle, with no voluntary activity in proximal muscles.
The next day (15 days after admission) we started IVIg 0.4 g/kg/day for 5 consecutive days. Nine days after the last IVIg infusion (1 month from admission), the patient was still severely tetraparetic, bedridden, and mechanically ventilated, with minimal improvement in the MRC sum score (**Fig. 1**). We thus started (at day 30 after disease onset) **EFG 10 mg/kg intravenously weekly for 4 cycles**, observing already within a few hours an improvement in upper limb strength. The patient was transferred to the rehabilitation clinic. After the second and the third infusion, strength in the upper limbs reached MRC score grade 4, but the patient was still bedridden.
One week after the last EFG infusion (2 months after admission), the tracheotomy could be removed, and 2 weeks later the patient walked with aid (**Fig.1**). At 4 month follow-up visit, apart from absent deep tendon reflexes and mild stocking hypoesthesia, the neurological examination was normal, with normal strength in the four limbs (**Fig. 1**). The same day, NCS showed marked improvement of CMAP amplitude of all nerves (**Fig. 2**), whereas needle EMG did not detect any spontaneous activity.

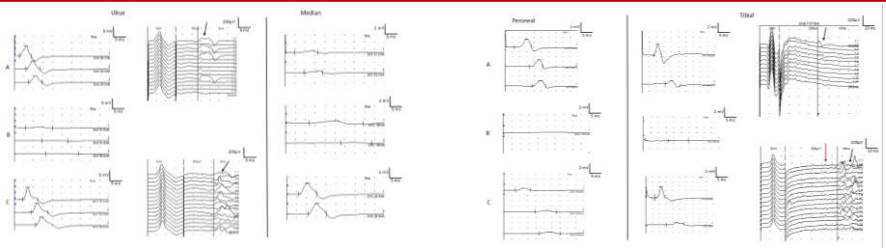

Figure 2. Nerve conduction studies of the upper limbs (left) and lower limbs (right). The evolution of the curves of the same CMAP over time can be appreciated comparing different time points: at admission (A), 14 days later (B) and at 4-month follow-up (C). At admission, distal latencies were increased and CMAP reduced. F waves (black arrows) had reduced persistence. After 2 weeks, nerves became almost inexcitable, and F-waves could not be elicitated. Four months after admission, the CMAP amplitude were diffusely increased, and F-waves become again visible, with normal persistence. At this stage, A-waves were also recorded in the tibialis stimulation (red arrows).

Interpretation
In our patient, suffering from a fulminant course of AIDP, EFG led to clinical improvement and was safe, but it is possible that we could have obtained the same clinical effect by repeating additional courses of PLEX (progressively reducing IgG levels) or waiting longer for the effect of the IVIg course. Recently, in China, other severely affected GBS patients have been treated with EFG as first-line treatment [3], or as sequential treatment after PLEX and IVIg [4–6], with a described favorable outcome.

References
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