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Tapering of Corticosteroids in Patients With Generalized Myasthenia Gravis Treated With Efgartigimod: A Case Series

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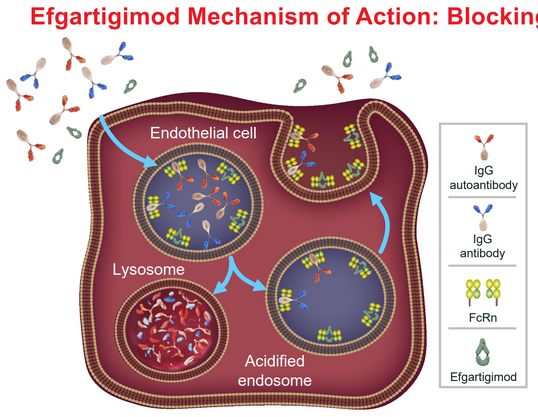
BACKGROUND

Efgartigimod Mechanism of Action: Blocking FcRn



- FcRn recycles IgG antibodies and albumin. This recycling and salvage from lysosomal degradation results in IgG antibodies having the longest half-life and being the most abundant of all immunoglobulins[1-3]
- Blocking FcRn to selectively reduce IgG levels is therefore a rational therapeutic approach in patients with IgG-mediated autoimmune diseases[1,2]
- Efgartigimod is an IgG1 antibody Fc fragment that has been engineered for increased affinity to FcRn, and is uniquely composed of the only part of the IgG antibody that normally binds FcRn[1]
- Efgartigimod selectively reduces IgG by blocking FcRn-mediated IgG recycling without impacting antibody production, albumin levels, or other parts of the immune system[1,4,5]
- Efgartigimod prevents IgG recycling by blocking IgG antibodies from binding to FcRn, with unbound IgG antibodies being degraded[1,4]

Corticosteroids and gMG

- Corticosteroids are a mainstay of treatment for many autoimmune diseases, including gMG[6]
- Corticosteroids are associated with multiple adverse events that have a major impact on patient quality of life[7,8]
- There is currently limited available information on how novel therapies impact corticosteroid use in patients with gMG

SUMMARY

- 6 patients presented to the clinic with symptoms of fatigable muscle weakness, with the time from onset of symptoms to evaluation ranging from 2 months to 12 years
- Patients received 3 to 12 treatment cycles of efgartigimod concurrently with corticosteroids and pyridostigmine
- Improvements were observed in disease-specific scales that evaluated ability to perform activities of daily living and objective measures of strength
- Patient-reported outcomes included improvement in bulbar symptoms, appendicular symptoms, and ocular symptoms
- Treatment with an FcRn-blocking therapy in these cases improved patient muscle weakness symptoms and allowed for tapering of the dose and/or dosing frequency of corticosteroids

PATIENT CASES

Patient Case Overview

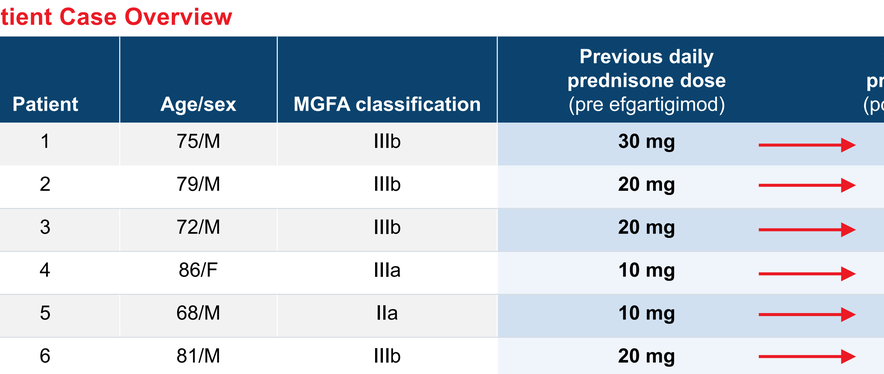
Patient	Age/sex	MGFA classification	Previous daily prednisone dose (pre efgartigimod)	Current daily prednisone dose (post efgartigimod)	Baseline MG-ADL (pre efgartigimod)	Most recent MG-ADL (post efgartigimod)	Baseline MGC (pre efgartigimod)	Most recent MGC (post efgartigimod)	# of efgartigimod cycles
1	75/M	IIIb	**30 mg**	**10 mg**	**6**	**3**	**8**	**5**	12
2	79/M	IIIb	**20 mg**	**5 mg**	**6**	**0**	**9**	**0**	4
3	72/M	IIIb	**20 mg**	**None**	**10**	**0**	**18**	**2**	5
4	86/F	IIIa	**10 mg**	**2 mg**	**7**	**1**	**11**	**2**	7
5	68/M	IIa	**10 mg**	**5 mg**	**4**	**1**	**8**	**0**	5
6	81/M	IIIb	**20 mg**	**5 mg**	**6**	**2**	**10**	**2**	3
					Range: 0-24 (lower is better)		**Range:** 0-50 (lower is better)		

Patient Case 1
Male Aged 75 Years With AChR-Ab+ gMG

History
- Hyperlipidemia
- Hypothyroidism
- GERD
- Hypertension
- Pulmonary embolism
- Osteopenia

Symptoms
- Weakness rising from chair
- Diplopia and eyelid droop
- Shortness of breath
- Dysphagia

Prior treatments
- Prednisone 30 mg QD
- Pyridostigmine 30 mg TID
- Mycophenolate 1000 mg BID
- Eculizumab 1200 mg Q2W



Continuation of treatment

Efgartigimod Treatment
- Continuation of 4 weekly infusions followed by 4 weeks between cycles
- Received **12 cycles** to date

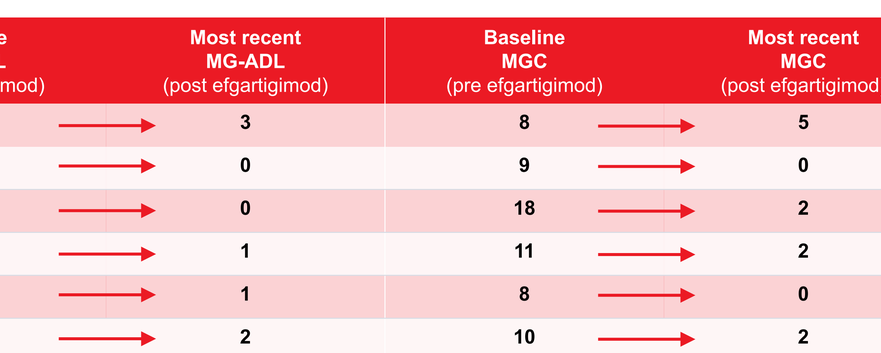
Current Treatment
- Prednisone **10 mg QD**
- Pyridostigmine 30 mg TID (Patient rarely takes due to AEs)
- Mycophenolate 1000 mg BID

Patient Case 2
Male Aged 79 Years With AChR-Ab+ gMG

History
- Hypertension
- Hyperlipidemia
- Psoriasis
- Aortic stenosis s/p TAVR
- Length-dependent polyneuropathy

Symptoms
- Dysphagia
- Dysarthria
- Lower facial muscle weakness

Prior treatments
- Prednisone 20 mg QD
- Pyridostigmine 60 mg TID



Continuation of treatment

Efgartigimod Treatment
- Continuation of 4 weekly infusions followed by 4 weeks between cycles

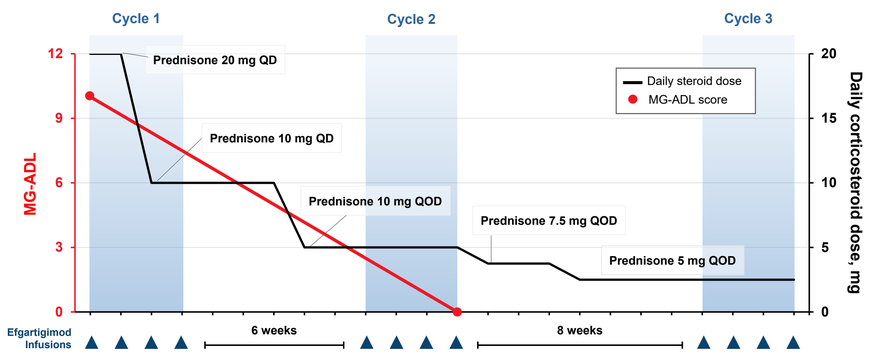
Current Treatment
- Prednisone **5 mg QD**

Patient Case 3
Male Aged 72 Years With AChR-Ab+ gMG

History
- Hypertension
- Osteoarthritis

Symptoms
- Bulbar symptoms
- Proximal weakness
- Right-sided ptosis

Prior treatments
- Prednisone 20 mg QD
- Pyridostigmine 60 mg TID
- Mycophenolate mofetil 1000 mg BID



Continuation of treatment

Efgartigimod Treatment
- Continuation of 4 weekly infusions followed by 8 weeks between cycles
- Received **5 cycles** to date

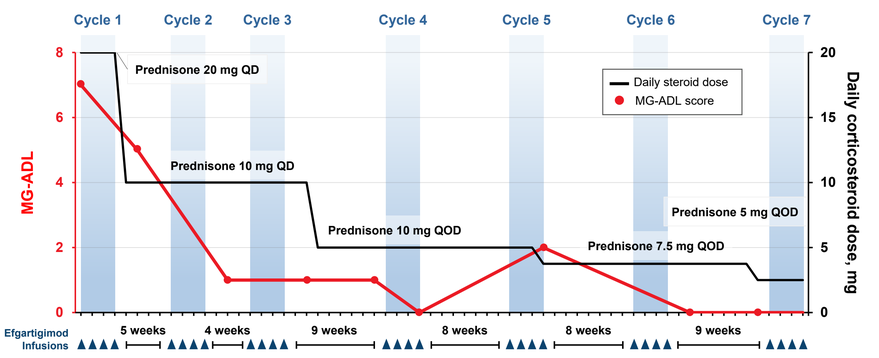
Current Treatment
- ***Prednisone discontinued***
- Pyridostigmine 60 mg **PRN**
- Mycophenolate mofetil 1000 mg BID

Patient Case 4
Female Aged 86 Years With AChR-Ab+ gMG

History
- Multifactorial anemia
- Tubular/tubulovillous polyps
- Peripheral neuropathy
- Hypothyroidism
- Osteopenia

Symptoms
- Persistent left-side eyelid droop
- Intermittent diplopia
- Significant fatigue
- Trouble rising from chair

Prior treatments
- Prednisone 20 mg QD
- Azathioprine[a]
- Mycophenolate mofetil[a]



Continuation of treatment

Efgartigimod Treatment
- Continuation of 4 weekly infusions followed by 8 weeks between cycles

Current Treatment
- Prednisone **2 mg QOD**
- **Pyridostigmine 60 mg PRN**

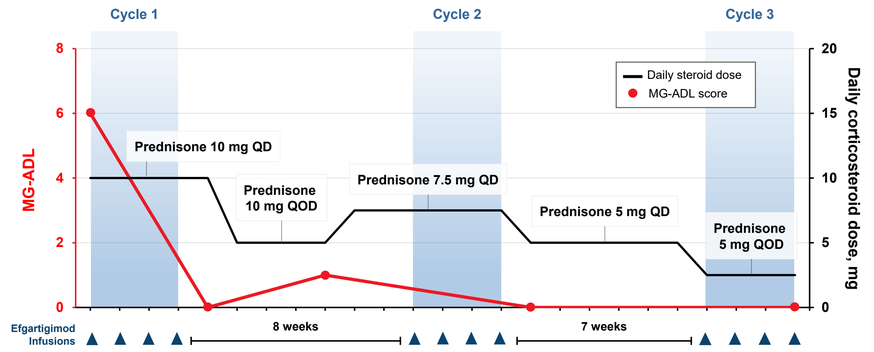
[a]Historical treatments that were discontinued prior to being evaluated by Dr Macwan.

Patient Case 5
Male Aged 68 Years With AChR-Ab+ gMG

History
- Allergic rhinitis
- Solitary lung nodule
- Gout
- Hypothyroidism
- Hypertension
- Hyperlipidemia
- Normocytic anemia
- Stage 3b CKD

Symptoms
- Trouble rising from chair
- Trouble chewing
- Difficulty raising arms above shoulders
- Shortness of breath

Prior treatments
- Prednisone 10 mg QD
- Pyridostigmine 60 mg TID
- Methotrexate 7.5 mg QW



Continuation of treatment

Efgartigimod Treatment
- Continuation of 4 weekly infusions followed by 8 weeks between cycles
- Received **5 cycles** to date

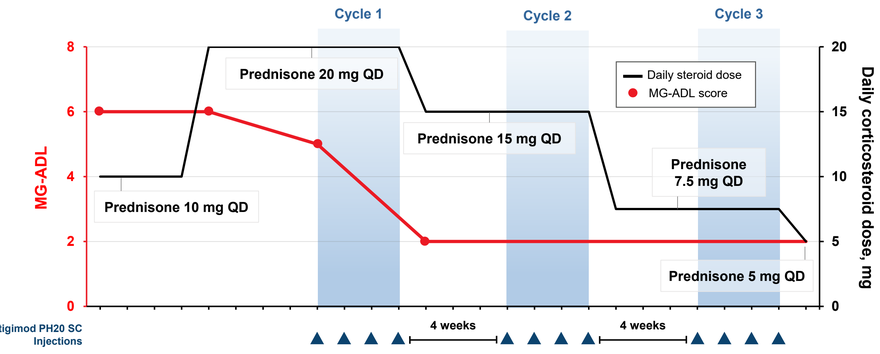
Current Treatment
- Prednisone **5 mg QOD**
- Pyridostigmine 60 mg **PRN**
- Methotrexate 7.5 mg QW

Patient Case 6
Male Aged 81 Years With AChR-Ab+ gMG

History
- DVT
- Dyslipidemia

Symptoms
- Trouble chewing
- Trouble swallowing
- Trouble rising from chair

Prior treatments
- Prednisone 10 mg QD
- Pyridostigmine 60 mg TID
- IVIg 2 g/kg divided over 4 days



Continuation of treatment

Efgartigimod PH20 SC Treatment
- Continuation of 4 weekly injections followed by 4 weeks between cycles
- Received **3 cycles** to date

Current Treatment
- Prednisone **5 mg QD**
- Pyridostigmine 60 mg **PRN**

ABBREVIATIONS
Ab, antibody; AChR, acetylcholine receptor; AE, adverse event; BID, twice daily; CKD; chronic kidney disease; DVT, deep vein thrombosis; Fc, crystallizable fragment; FcRn, neonatal Fc receptor; GERD, gastroesophageal reflux disease; gMG, generalized myasthenia gravis; Ig, immunoglobulin; IVIg, intravenous immunoglobulin; MG-ADL, Myasthenia Gravis Activities of Daily Living; MGC, Myasthenia Gravis Composite; MGFA, Myasthenia Gravis Foundation of America; PRN, as needed; Q2W, once every 2 weeks; QD, once daily; QOD, once every other day; QW, once weekly; SC, subcutaneous; s/p, status post; TAVR, transcatheter aortic valve replacement; TID, 3 times daily.

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ACKNOWLEDGMENTS AND DISCLOSURES: Cases are based on the clinical experience of Dr. Samir Macwan; details may have been modified to ensure patient confidentiality.

SM: AbbVie, Alexion, argenx, Catalyst, Grifols, Kabafusion, Octapharma, and UCB. **JT:** No disclosures to report. Medical writing and editorial support for this presentation was provided by Precision AQ and funded by argenx.

Presented at The Neuromuscular Study Group (NMSG) Annual Scientific Meeting; September 20-22, 2024; Tarrytown, New York