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Fast-acting treatment of myasthenic crisis with efgartigimod from the perspective of the neonatal intensive care unit

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Abstract

Background Myasthenic crisis (MC) refers to rapid deterioration of myasthenia gravis (MG), affecting lung and bulbar muscles and causing breathing difficulties. Currently, efgartigimod has shown good therapeutic effects in patients with generalized myasthenia gravis (GMG). This retrospective real-world study explored the effectiveness of efgartigimod in patients with MC.

Method Reviewing the clinical data of five patients (including four patients with refractory MC) with MC who received efgartigimod at the First Affiliated Hospital of Sun Yat-sen University, all of these patients were admitted from September 2023 to December 2023.

Results Each patient received 20 mg/kg of efgartigimod on the first and fifth day. After discharge, all patients showed a clinically meaningful decrease in Myasthenia Gravis Activities of Daily Living (MG-ADL) scale (a decrease of ≥ 2 points) and an improvement in their lung function. Additionally, all patients had a decrease in IgG levels (58.59 ± 18.48% after one cycle of efgartigimod). We also explored the ICU stay and mechanical ventilation (MV) duration for these five patients, and found no significant improvement compared to a large sample data. In terms of safety, four patients experienced adverse events (AEs), all of which were mild. At the last follow-up, four patients achieved the minimal symptom expression (MSE) status (an MG-ADL score of 0 or 1) after 6.25 ± 3.30 weeks. Only one patient experienced a worsening of symptoms in the second week after discharge, but she also achieved the MSE status after receiving a second cycle of efgartigimod treatment.

Conclusions Given the conclusion that intravenous efgartigimod is a non-invasive fast-acting treatment with fewer AEs, this may provide NICU workers with another option for managing patients with MC.

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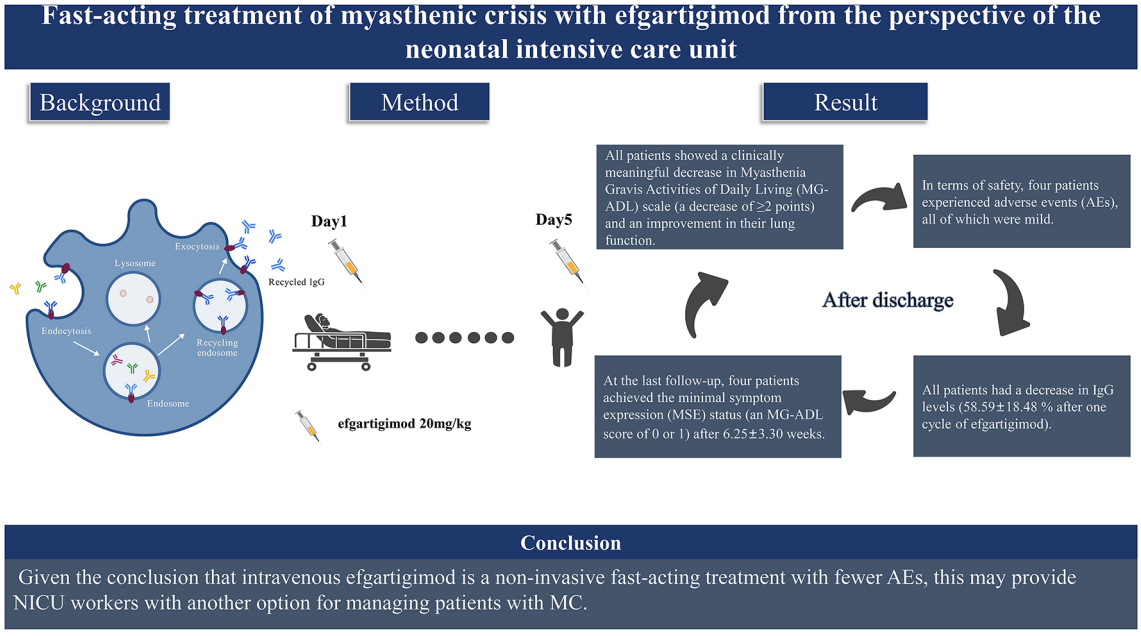
Graphical abstract



Keywords Myasthenic crisis, Myasthenia gravis, Efgartigimod, Fast-acting treatment, The neonatal intensive care unit

Introdution

Myasthenic crisis(MC) is defined as quickly worsening of myasthenic weakness requiring intubation or noninvasive ventilation due to either respiratory weakness or significant bulbar muscular weakness, is the leading cause of death in individuals with myasthenia gravis(MG) [1, 2]. It imposes a substantial burden on individuals and society, severely decreasing the quality of life of MC patients. MG occurs in 10–20% of patients, the majority of whom develop MC within 2 years of MG diagnosis [3, 4].

Fast-acting treatment is defined as aggressive application of non-oral fast-acting immunotherapies including plasmapheresis (PE), intravenous immunoglobulin (IVIG) and/or intravenous high-dose methylprednisolone (IVMP) as first-line treatment, which is recommended by various guidelines for MG [5]. The neonatal intensive care unit (NICU) is the most appropriate place for MC treatment, which is experienced in fast-acting treatment.

MG is an autoimmune disease that targets the neuromuscular junction, potentially affecting skeletal muscles throughout the body, which results in limb weakness, dysarthria, head drop, and dysphagia. This disorder is caused by the production of pathogenic autoantibodies such as acetylcholine receptor antibodies (AChR-Ab), muscle-specific tyrosine kinase antibodies (MUSK-Ab), and low-density lipoprotein receptor-related protein 4 antibodies (LRP4-Ab) [6, 7]. All these antibodies are of the IgG type, with a long half-life of approximately 20–23 days due to reuptake by the neonatal Fc receptor (FcRn). The FcRn represents a type I MHC molecule that recycles immunoglobulins in a pH-dependent manner, inhibiting IgG degradation by lysosomes and prolonging its half-life. Effective plasma exchange and immunoadsorption can efficiently clear autoantibodies in patients with MG; thus, blocking FcRn is a rational therapeutic approach for targeting the key pathogenic driver of generalized myasthenia gravis(GMG) [8, 9]. Efgartigimod is a human IgG1 Fc fragment that competitively binds to pathogenic antibodies, decreasing IgG recycling and inducing their degradation. Thus, efgartigimod could decreases the serum levels of pathogenic antibodies [10, 11].

Currently, 10 mg/kg/quaque week(qw) efgartigimod in patients with GMG has been approved in several countries. A multicenter real-world study conducted in China demonstrated that among nine patients with MC, the average Myasthenia Gravis Activities of Daily Living (MG-ADL) score decreased by four points within the first week following treatment, with continued improvement over time [12]. Positive outcomes were also observed in patients with MUSK-Ab positive MC [13]. As MC is an emergency condition, a fast-acting treatment is required. The value of applying high doses of efgartigimod remains unknown, although an ongoing trial (NCT05701189) of Guillain Barré Syndrome (GBS) applied intravenous 20 mg/kg administration of efgartigimod on days one and five. Currently, the maximum dose of efgartigimod used

in Phase III clinical trials is 50 mg/kg, which has been deemed safe. To address this knowledge gap, this retrospective real-world study of intravenous high-dose efgartigimod was performed in patients with MC, evaluating detailed clinical information.

Methods

Patients

From September to December 2023 in the Department of Neurology, the First Affiliated Hospital of Sun Yat-sen University, five patients diagnosed with MC and administered efgartigimod as the fast-acting treatment were included in this analysis. All the five MC cases examined met the international definition of manifest MC: worsening of myasthenic weakness requiring intubation or noninvasive ventilation to avoid intubation, except when these measures are applied in routine postoperative management [14]. These cases were categorized as Myasthenia Gravis Foundation of America (MGFA) class V [15]. After hospital admission, the patients were transferred to the NICU due to severe condition.

Treatment

In this cohort, prior to efgartigimod administration or while in a pre-crisis state, the patients may have been administered other fast-acting treatments such as IVMP, PE, or IVIG. However, these treatments were unsuccessful in improving the patients' conditions. First, the five patients were all administered efgartigimod intravenously at a dose of 20 mg/kg on the first and fifth days. Next, based on weekly response to efgartigimod during and after treatment, whether a second cycle of efgartigimod treatment should be administered was determined for each patient. According to the patient's condition, a treating neurologist determined whether to switch to a fast-acting drug or to continue with the current treatment plan if the previous treatment was ineffective.

Evaluation of clinical effectiveness

The measure utilized in this analysis was the MG-ADL scale. Clinically meaningful improvement in MG-ADL scores was defined as ≥ 2 point change in MG-ADL after one cycle of administration or dischargement compared with scores when the first administration was given. Minimal symptom expression (MSE) is defined as an MG-ADL score of 0 or 1 [16–18]. The reduction in patients' IgG levels and lung function improvement were also included to analyse the effiency of the efgartigimod. Lung function improvement was defined as successful weaning from the ventilator and removal of the tracheal tube. The concentration of total IgG in plasma was measured by an immunoturbidimetric assay. Clinical evaluations were performed at treatment initiation and weekly until the last follow-up. During the efgartigimod treatment period, no changes were made in the types and doses of other immunosuppressive agents.

Safety evaluation

All adverse events(AEs) during and after efgartigimod treatment were recorded. White blood cell count, serum albumin et al. were measured at the initial administration of efgartigimod and at the last follow-up.

Result

Clinicodemographic characteristics of MC patients

The age at admission was 43.80 ± 16.48 years old with a female-to-male ratio of 4:1. The disease duration was 4.08 ± 5.63 years from the onset to the current crisis. The proportion for thymoma and thymectomy was 60% (3/5) and 40% (2/5), respectively. In this study, a female patient (Case 5) was diagnosed with MuSK antibody-related myasthenia gravis during hospitalization, with initial symptoms indicating myasthenic crisis (Table 1).

Treatment descriptions

All patients completed two infusions of efgartigimod (Fig. 1). Each patient was administered 20 mg/kg twice, on the first and fifth days. Case 1 received two cycles of efgartigimod treatment. Prior to treatment, most patients were administered first-line treatments such as IVIG and IVMP, alongside corticosteroids and immunosuppressants. The immunotherapies before the current MC included corticosteroid (80%,4/5), tacrolimus (40%,2/5), mycophenolate mofetil (MMF) (20%,1/5), and IVIg and/or TPE maintenance (80%, 4/5) (Table 2).

Clinical effectiveness of efgartigimod for MC treatment

MG-ADL scores

The MG-ADL scores of all patients indicated clinically meaningful improvement (MG-ADL score decease by ≥ 2 points at discharge versus scores at admission). MG-ADL score reduced from 21.00 ± 3.00 at the baseline to 11.6 ± 3.2 at discharge. However, in Case 5, after the first dose of efgartigimod, lung function improved slowly, and MG-ADL score showed no decrease.

At the last follow-up, 4 patients ultimately reached the MSE state after 6.25 ± 3.30 weeks. The follow-up period was 10.60 ± 3.13 weeks. MG-ADL scores in 4 patients continued to show meaningful improvement until the last follow-up, while 1 patient had increased MG-ADL scores (Fig. 2).

Lung function

All five cases underwent tracheal intubation and received MV. One patient underwent tracheostomy. All patients had tracheal tubes removed before discharge and were successfully weaned from ventilation.

Table 1 Clinical characteristics for the patients

	Age at diagnosis	Sex	MG duration	Abs	Pathology of thymus	Thymectomy	Underlying disease	Skeletal muscle weakness of MG	Triggers for MC	The times of MC onset	Treatment before MC
Case 1	49	Female	5 months	Ach-R-Ab、Titin-Ab、RyR-Ab	Thymoma			Bulbar, respiratory and limb muscles	No obvious precipitant	1	CS 8 mg, Pyd 60 mg t.i.d, IVMP, IVIG
Case 2	42	Female	14 years	Ach-R-Ab、Titin-Ab	Thymoma		HTN	Ocular, bulbar, respiratory and limb muscles	Upper respiratory infection	1	CS 8 mg, Pyd 60 mg t.i.d, MMF 250 mg b.i.d, IVMP, IVIG
Case 3	32	Male	2 years	Ach-R-Ab、Titin-Ab	Thymoma	Yes		Ocular, respiratory and limb muscles	Aspiration pneumonia	1	CS 20 mg, Pyd 60 mg t.i.d, tac 2 mg, IVMP, IVIG
Case 4	27	Female	3 years	Ach-R-Ab、Titin-Ab	Thymic hyperplasia	Yes		Ocular, bulbar, neck, respiratory and limb muscles	No obvious precipitant	2	CS 20 mg, Pyd 60 mg q6h, tac 3 mg, IVMP, IVIG
Case 5	69	Female	1 year	Musk-Ab	Normal		PE、PMV、Tr	Bulbar, respiratory and limb muscles	Aspiration pneumonia	1	No

MG = myasthenia gravis; Ach-R-Ab = acetylcholine receptor antibodies; Titin-Ab = Titin antibodies; Musk-Ab = muscle-specific tyrosine kinase antibodies; RyR-Ab = Ryanodine receptor antibodies; HTN = hypertension; PE = pulmonary embolism; PMV = prolapse of mitral valve; TR = tricuspid regurgitation; CS = prednisone/methylprednisolone; Pyd = pyridostigmine; MMF = mycophenolate mofetil; Tac = tacrolimus

IgG levels

After one cycle of efgartigimod treatment, immunoglobulin levels decreased. Specifically, IgG levels in all patients decreased by 58.59 ± 18.48%. IgG levels reduced to 5.39 ± 1.33 g/L from 17.28 ± 13.11 g/L after one cycle of efgartigimod (Fig. 3).

ICU stay and MV duration

ICU stay and MV duration for all five patients were recorded. The length of stay in the ICU for the five patients was 25.00 ± 13.15 days, with a median of 16 days. The MV duration was 24.40 ± 15.76 days, with a median of 18 days.

Safety profile of efgartigimod for MC treatment

Four patients experienced AEs, with no recorded infusion-related reaction. During hospitalization, 3 patients had leukocytosis, 2 had elevated aspartate aminotransferase (AST) levels, 1 had elevated creatinine (Cr), 1 had elevated uric acid (BUN) and 1 experienced infection (Fig. 4).

After discharge, 1 patient (Case 3) experienced upper respiratory tract infections twice without fever, accompanied by mild headache (Table 3).

Discussion

This study reports 5 patients with MC who were administered efgartigimod. All five patients responded well to efgartigimod treatment, with MG-ADL scores decreased meaningfully compared with admission values, successful weaning from MV and removal of tracheal intubation. One patient (Case5) showed no significant improvement in lung function in the first three days of efgartigimod monotherapy, suggesting that the treatment effect of efgartigimod was not evident. Blood gas analysis revealed that the partial pressure of CO2 remained high (40–47 mmHg). When the machine-controlled respiratory rate was decreased, the ventilator repeatedly indicated suffocation, and the machine-controlled respiratory rate needed to be kept at 8 breaths/minute. After controlling the infection, steroids and IVIG were added to intensify immunotherapy and help alleviate the condition. All patients had decreased IgG levels.

In terms of long-term effectiveness, 4 patients had a sustained decrease in MG-ADL scores, which was clinically meaningful. The four patients maintained their immunotherapy medications throughout their hospitalization, with dosage adjustments made progressively under the guidance of treating neurologist. At the last follow-up, 4 patients ultimately reached the MSE state. Case 1 developed dysarthria and dysphagia approximately 2 weeks after discharge, and subsequently returned to our hospital. This was suspected to be due to noncompliance with the instructions from the neurologist in using

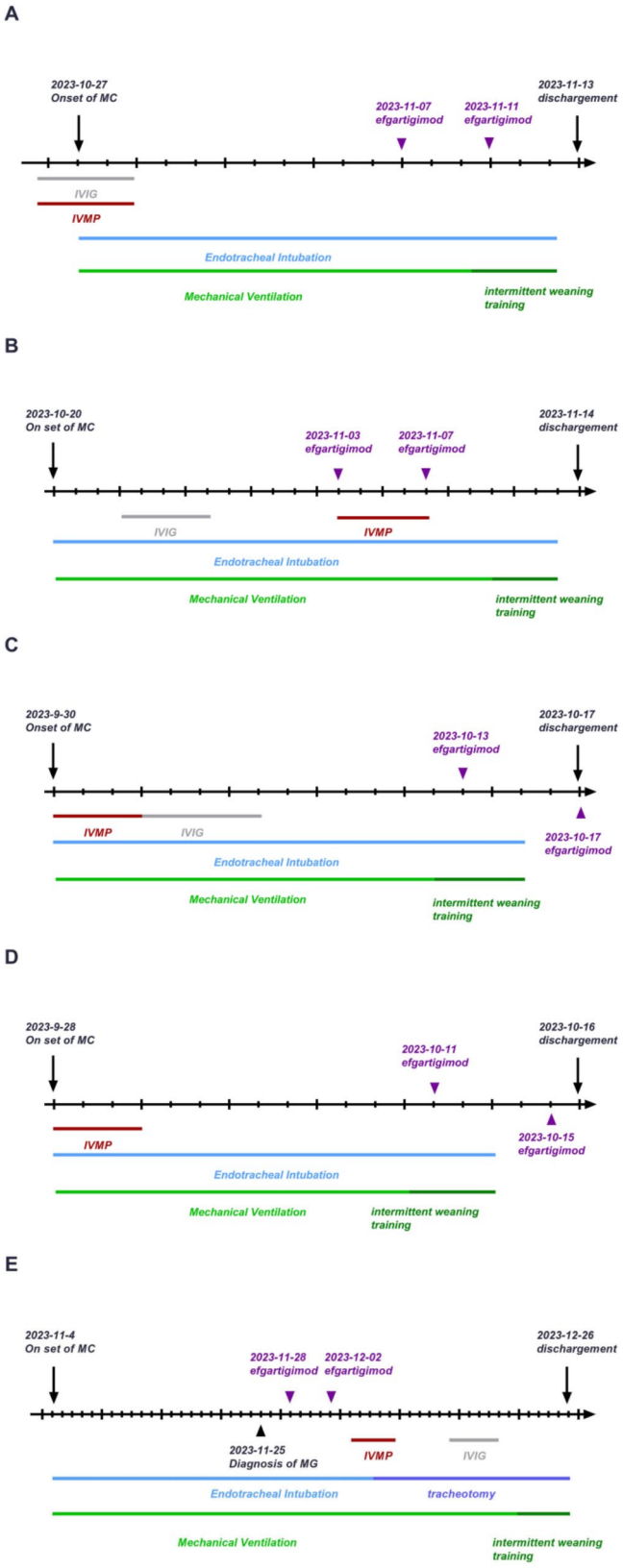


Fig. 1 Treatment descriptions for case 1 (**A**), case 2 (**B**), case 3 (**C**), case 4 (**D**) and case5 (**E**). The figure illustrates a timeline depicting the entire process for five MC patients, encompassing the stages of diagnosis, treatment, and subsequent recovery. The purple markers indicate the timing and frequency of efgartigimod infusions. Additionally, changes in treatment are depicted along the trajectory. MG = myasthenia gravis; IVMP = intravenous high-dose methylprednisolone; IVIG = intravenous immunoglobulin

Table 2 Treatment descriptions of the patients

	IVMP	IVIG	PE	Efgartigimod	Concomitant medications	Ventilator weaning	Tracheotomy	Decannulation	Duration of ICU stay	Duration of MV
Case 1	Yes	Yes	No	Yes	CS 40 mg, Pyd 60 mg q6h, Tac 3 mg	Yes		Yes	17	16
Case 2	Yes	Yes	No	Yes	CS 40 mg, Pyd 60 mg t.i.d, MMF 500 mg	Yes		Yes	24	23
Case 3	Yes	Yes	No	Yes	CS 40 mg, Pyd 60 mg q8h, Tac 2 mg	Yes		Yes	18	16
Case 4	Yes	No	No	Yes	CS 40 mg, Pyd 60 mg q6h, Tac 3 mg	Yes		Yes	18	15
Case 5	Yes	Yes	No	Yes	CS 40 mg, Pyd 60 mg q8h, Tac 3 mg	Yes	Yes	Yes	48	52

IVMP = intravenous high-dose methylprednisolone; IVIG = intravenous immunoglobulin; PE = plasma exchange; ICU = intensive care unit; MV = mechanical ventilation

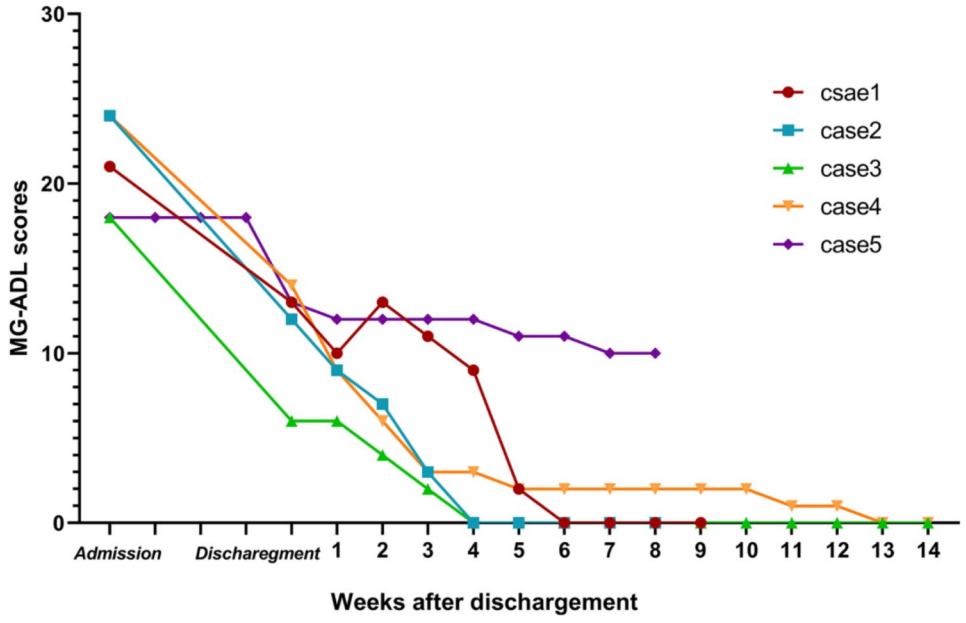


Fig. 2 Changes in Myasthenia Gravis Activities of Daily Living (MG-ADL) scores. The changes in MG-ADL scores for five patients were monitored during hospitalization, throughout the treatment process, and after discharge on a weekly basis

the gastric tube in the first week after discharge. After evaluating her symptoms, a second cycle of efgartigimod was administered at 10 mg/kg once a week, for a total of 4 doses. After infusion of the second cycle, the patient's MG-ADL score decreased to 0. After the patient reached a stable condition, thymectomy was performed. In Case 5, the patient underwent three fast-acting treatment, specifically IVMP, IVIG and efgartigimod. As a result, the patient was able to successfully wean off the ventilator and was discharged following the last follow-up. Regrettably, this patient was subsequently lost to follow-up, and we were unable to acquire any further information regarding his/her condition.

In terms of safety, 3 patients experienced leukocytosis. However, it is noteworthy that all five patients were concurrently administered oral or intravenous steroids, so it is challenging to rule out steroid-induced leukocytosis. Some patients had elevated AST, uric acid, or creatinine levels after efgartigimod treatment, but these levels mostly returned to normal before discharge. The patients used multiple medications simultaneously during hospitalization, making it difficult to determine whether the symptoms were caused by the toxic effects of efgartigimod or interactions with other drugs.

In this study, the ICU stay and MV duration of patients were also compared with those reported by large-sample studies. A retrospective analysis of 250 patients with MC revealed a median MV duration of 12 days (with a range of 1-219 days), a median ICU stay of 16 days, and a median hospital stay of 26 days [19]. In this cohort, MV duration was relatively longer. However, in four out of five cases, other immunotherapeutic methods did not

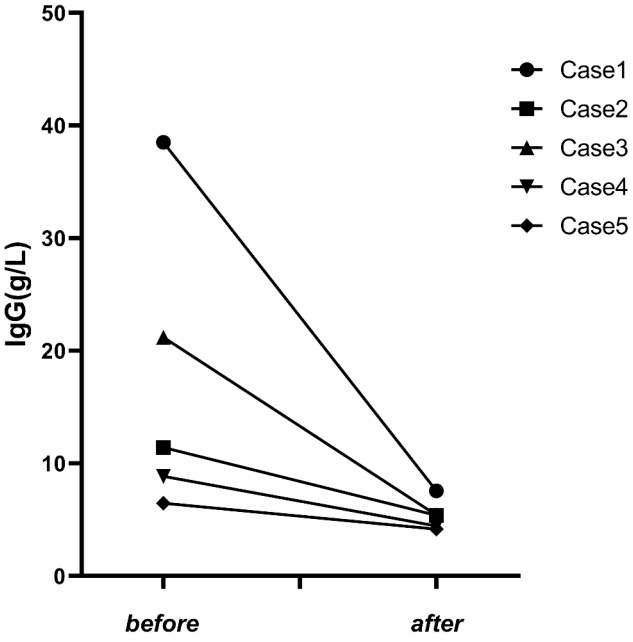


Fig. 3 Changes in immunoglobulin levels. The changes in IgG levels for these five patients were measured before and after treatment with efgartigimod

yield good results, indicating a refractory myasthenia crisis (RMC). Moreover, considering that two of the five patients were elderly, two had pneumonia, and two had comorbidities, all of which are risk factors for prolonged MV duration, indicating severe conditions for these five patients, rendering weaning from ventilation difficult. After efgartigimod administration, most patients were successfully weaned from ventilation within one week. Case 5 experienced extended MV duration and ICU stay, possibly due to the delayed diagnosis of MC, which was detected on the 21st day after onset. This led to a delayed start of first-line treatment. Additionally, the patient was elderly and had more severe baseline MG, more comorbidities, severe pneumonia, and MuSK antibodies, all of which contribute to prolonged intubation and ICU stay [19, 20].

Currently, IVMP, IVIG and PE are the most commonly applied first-line treatment approaches for MC patients. While IVMP is effective, it carries the risk of exacerbating MG symptoms. Additionally, corticosteroids can impact both innate and acquired immunity, with their immunosuppressive effects increasing the risk of infection in a dose-dependent manner. Nevertheless, the significant side effects of high-dose steroids, including hyperglycemia, gastrointestinal reactions, skin changes, and obesity, should be considered. Although IVIG exerts positive treatment effects, it has poor efficacy in patients with Musk antibody positivity [21, 22] and VNTR2/3 gene expression [23].PE does not solely clear IgG antibodies and requires central venous access, but potentially induces infection and thrombosis. Meanwhile, it requires trained and specialized nursing staff and single-use

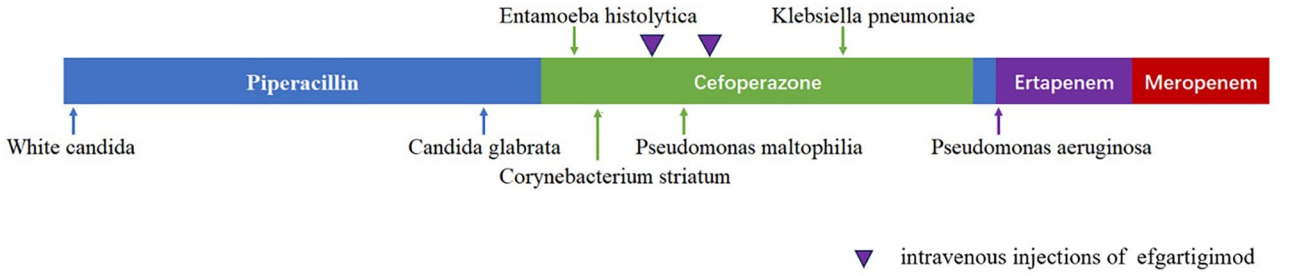


Fig. 4 Infection history and primary medication history of main antibiotic for Case 5. Case 5 was admitted with aspiration pneumonia and elevated infection markers. Subsequent sputum cultures detected infections with Candida albicans, Acinetobacter lwoffii, Pseudomonas maltophilia, Klebsiella pneumoniae subsp, Burkholderia cepacia, and Pseudomonas aeruginosa. Except for Pseudomonas aeruginosa, bacterial sputum cultures for other bacteria were negative after antibiotic treatment

Table 3 Summary of adverse events

	Any adverse event	Leukocytosis	hypalbuminemia	Any infection	Elevated AST	Elevated BUN	Elevated Cr	Upper respiratory tract infection	Headache
Case 1	Yes	Yes			Yes				
Case 2	Yes	Yes	Yes	Yes	Yes	Yes			
Case 3	Yes							Yes	Yes
Case 4	No								
Case5	Yes	Yes		Yes			Yes		

BUN = Blood urea nitrogen, Scr = Serum creatinine, AST = Aspartate aminotransferase

equipment. It also has higher incidence rates of moderate to severe AEs, e.g., catheter-related bloodstream infections, venous thrombosis, allergic reactions, and hypersensitivity reactions, which may also cause patient aversion. This suggests the need for a faster-acting and safer treatment option.

Currently, efgartigimod is approved for patients diagnosed with GMG. Phase I, II, and III trials have demonstrated good efficacy and safety in antibody-positive patients with GMG [11, 24, 25]. Reduced IgG1-4 levels were reported by clinical trials, without significant decreases in serum albumin, IgA, and IgM levels. In the ADAPT trial, the standard dosing regimen for efgartigimod was 10 mg/kg once weekly for a total of 4 doses. However, this regimen is not suitable for patients with MC in the NICU. In a phase I trial of efgartigimod, Ulrichts et al. concluded that IgG levels gradually decreased with dosage. However, only a slight reduction in IgG1 levels was observed 28 days after delivery among the 10, 25, and 50 mg/kg groups, and the levels of IgG subclasses 2, 3, and 4 were similarly reduced.In terms of safety, patients administered high-dose treatment appeared to show a higher frequency of AEs. However, most reported AEs were considered mild, and no serious AEs related to efgartigimod infusion were detected [11].

This new dosing regimen compresses the original 4-week treatment to 5 days, and the single dose was doubled while maintaining the total dose unchanged. This dosing approach is expected to decrease the treatment duration and facilitate the recovery of various functions in patients with MG, which is important in the NICU. Further, as a non-invasive treatment option, efgartigimod is safer than traditional fast-acting treatments for MC, with lower incidence and milder severity of AEs. Most patients can afford such treatment. Efgartigimod administration does not require specialized equipment, and the procedure is simple, making it feasible in primary care hospitals, which provides convenience for early intervention in MC patients.

Currently, no studies have reported the use of efgartigimod in MC patients with thymoma. In this study, three MC cases with thymoma achieved long-term remission after efgartigimod treatment. Previously, Katyal et al. reported four MG patients with thymoma administered a cycle of efgartigimod treatment. Three cases showed clinically meaningful improvement in MG-ADL score, with a decrease of more than 2 points after efgartigimod administration [18]. Kawama et al. reported two MG patients with thymoma who showed disease exacerbation after several cycles of efgartigimod treatment. Given that these are case reports with inconsistent conclusions, the associations of thymoma, thymectomy and efgartigimod need further investigation [26].

In this study, Case 5 tested positive for Musk antibodies, which are classified as IgG4 antibodies. In phase I clinical trials, use of efgartigimod accelerated the clearance of IgG4 antibodies, although IgG4 antibodies had less pronounced reduction compared with IgG1-3 [11]. In a phase III trial, three MuSK antibody-positive patients were reported, as MG-ADL responders (≥ 2-point MG-ADL improvement sustained for ≥ 4 weeks), indicating that efgartigimod still demonstrates good efficacy in MuSK antibody-positive MG patients [25].

In this study, simultaneous use of efgartigimod and IVMP (Case 1) may lead to improved treatment outcomes. While IVMP may temporarily worsen myasthenic symptoms and potentially impact the process of weaning, it can still be considered for patients requiring MV. However, efgartigimod may quickly improve MG symptoms by clearing antibodies, partially mitigating the exacerbation of MG induced by IVMP and further synergizing with the immunosuppressive effects of steroids to assist in respiratory function recovery.

Concerns about the use of efgartigimod for the treatment of MC still exist. For instance, patients administered efgartigimod then continue with IVIG, which may accelerate the removal of antibodies and immunoglobulin, thereby reducing the effectiveness of IVIG. Therefore, it is unknown whether these two methods can be used in sequence, as well as the appropriate interval between their administrations. Further clinical trials are required to address this question. The FCGRT gene encodes Fc receptors. Compared with individuals harboring the VNTR3/3 genotype, those with the VNTR2/3 genotype have an 18% lower expression of FcRn [27]. The mechanism of action of efgartigimod involves competing with Fc receptors and interfering with IgG recycling, thereby reducing circulating IgG levels. Therefore, patients with the VNTR2/3 genotype are likely to show a poorer response to efgartigimod. However, further investigation is required to confirm this notion. Unfortunately, genetic testing for this gene was not conducted for the five patients in this study.

The strength of this study lies in the novel application of a 20 mg/kg dosage in MC cases, administered twice on the first and fifth days, with good efficacy and safety. This dosing regimen, while maintaining the same dosage, shortened the treatment time, providing a novel fast-acting treatment option for patients, which is especially suitable for NICU patients. The limitations of this study should be mentioned. First, the sample size was limited, with a short follow-up period. In addition, the bias of baseline severity of patient condition and the degree of infection may affect the actual efficacy of efgartigimod in treating major depression. The current study utilized MG-ADL scores to evaluate the patients' conditions, primarily focusing on changes over a week. This may

not be the most suitable measurement approach for MC patients. Additionally, in the NICU, MG cases are often stabilized with analgesics and sedatives to prevent agitation and discomfort, which could potentially impact the assessment of Quantitative Myasthenia Gravis (QMG) scores. Currently, there is no specialized scoring system suitable for patients with MC, and future research should address this gap.

Conclusion

Efgartigimod is an innovative treatment for MG, with promising therapeutic effects in patients with GMG. In this article, four refractory MC cases (those with no improvement with IVMP, PE or IVIG treatment) were administered efgartigimod as the fast-acting treatment, achieving significant disease remission and reaching the MSE state. Case 5 was treated with efgartigimod in combination with IVIG and IVMP, and achieved disease remission upon discharge. These findings suggest that high-dose efgartigimod treatment shows a positive therapeutic effect on refractory myasthenia crisis (RMC), with acceptable safety. Larger prospective cohort studies are warranted to confirm the actual effectiveness of efgartigimod in patients with MC.

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Author contributions
F.S. and R.L. performed the manuscript writing and the statistical analysis. L.F., H.Z. and X.S. acquired the data. C.S., J.F., and Z.X. contributed to data analysis and interpretation. H.W. and H.F. contributed to the experimental design and manuscript revision, and handled funding and supervision. H.W. and H.F. were the co-corresponding authors, and had full access to all the data in the study and takes responsibility for the integrity of the data and the accuracy of the data analysis. All authors contributed to manuscript revision, read, and approved the submitted version.

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Data availability
No datasets were generated or analysed during the current study.

Declarations

Ethics approval and consent to participate
All procedures with human participants' involvement were following the ethical standards of the institutional and/or national research committee and with the 1964 Helsinki Declaration and its later amendments or comparable ethical standards. This is an observational study, and the local ethics committee for clinical research has confirmed that no ethical approval is required. Before administration of the efgartigimod, we clearly informed the efficacy and AEs of it and fully explained the purpose and content of this study. Moreover, as this is a retrospective study with no additional interventions, the requirement for written informed consent was waived. Every patient who filled out the questionnaire indicated agreement to participate.

Consent for publication
All patients indicated agreement for publication. No personal information of the participants has been disclosed in this manuscript.

Competing interests
The authors declare no competing interests.

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