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Brief Research Report

Intravenous immunoglobulin is an effective steroid-sparing drug in inflammatory myositis

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ABSTRACT

INTRODUCTION. Glucocorticoids are the cornerstone of treatment of idiopathic inflammatory myositis (IIM) but are associated with significant side effects, especially during long-term use. Intravenous immunoglobulin (IVIG) has emerged as a promising steroid-sparing alternative, particularly for refractory cases.

METHODS. This retrospective cohort study included patients aged ≥ 18 years treated with IIM who received IVIG treatment between August 2018 and August 2023. Outcomes included changes in myositis-related symptoms, functional exercise tests, creatinine kinase (CK) levels and prednisolone dosage at three and six months.

RESULTS. A total of 22 patients were included, with the most common IIM subtypes being polymyositis (36%), immune-mediated necrotizing myopathy (23%) and dermatomyositis (18%). Dysphagia improved in 87.5% of patients, and cardiac symptoms resolved in all affected individuals by six months. Significant improvements were observed in functional performance and CK levels at both time points. Prednisolone dosage decreased from 44.7 mg/day (standard deviation (SD): ± 29.7) at baseline to 9.9 mg/day (SD: ± 12.8) at six months, representing a 77% reduction.

CONCLUSIONS. IVIG is an effective treatment and steroid-sparing therapy in IIM, yielding substantial reductions in glucocorticoid use, symptom improvement and functional recovery. These findings suggest that IVIG may be considered earlier in the disease course, especially in patients with contraindications to glucocorticoids.

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TRIAL REGISTRATION. This study was not registered as it was a quality improvement project and did not meet criteria for trial registration.

Idiopathic inflammatory myositis refers to a diverse group of immune-mediated diseases that affect the muscles and often involve other organs [1]. Glucocorticoids remain first-line therapy [2], despite side effects and suboptimal therapy success rates [3].

Long-term glucocorticoid therapy can lead to a range of adverse effects, including osteoporosis, increased risk of infection, hypertension and psychological effects. Additionally, glucocorticoids may induce a progressive chronic myopathy, which can exacerbate muscle weakness in patients with idiopathic inflammatory myositis [4]. These risks highlight the need for steroid-sparing alternatives. Intravenous immunoglobulin (IVIG) has emerged as a promising alternative, particularly in refractory cases [5, 6].

Herein, we report an IVIG down-scaling strategy that leads to restricted glucocorticoid use, and which was effective in 20/21 patients with various idiopathic inflammatory myositis subtypes.

Methods

We conducted a retrospective cohort study using data from the electronic medical files and the hospital registry between August 2018 and August 2023. Patients were followed for six months or until death or emigration. This study was approved as a quality project, ensuring its compliance with Danish legislation.

We included all patients aged 18 years or older who received IVIG treatment and were registered with an international classification of diseases, 10th version (ICD-10) code for idiopathic inflammatory myositis (M330, M331, M332, M339, M360, M609) at a single tertiary centre. Based on review of the electronic medical files, the patients were sub-grouped into one of the following idiopathic inflammatory myositis subtypes: polymyositis (PM), immune-mediated necrotizing myopathy (IMNM), dermatomyositis (DM), immune checkpoint inhibitor-associated myositis (ICI myositis), anti-synthetase syndrome (ASS), overlap myositis or amyopathic dermatomyositis.

Outcomes were assessed at three and six months after IVIG initiation and included the following: I) changes in myositis-related symptoms, including dysphagia and cardiac involvement, II) performance on functional exercise tests, measured by the Manual Muscle Test-8, Six-minute Walk Test (6MWT), and grip strength, III) plasma creatine kinase (CK) concentrations and IV) prednisolone dosage.

Medical records were used to collect data on demographics, disease manifestations, serology, comorbidities and adverse effects. Adverse events such as deep vein thrombosis and pulmonary embolism were noted. The index date was defined as the date of the first treatment with IVIG during the study period.

Categorical data are presented as counts and percentages, and continuous data as mean ± standard deviation (SD) or median with interquartile range, depending on distribution. Outcomes were calculated as the average percentage change from baseline to three and six months. Statistical analyses were conducted using Excel (v16.60), and GraphPad Prism was used for graph preparation.

Trial registration: This study was not registered as it was a quality improvement project and did not meet criteria for trial registration.

Results

A total of 22 patients with various idiopathic inflammatory myositis subtypes were included (Supplementary table 1): PM (36.4%), IMNM (22.7%) and DM (18.2%). ICI myositis (13.6%) and ASS/overlap myositis (9.0%). Fourteen of the patients (63.6%) were male, and the median disease duration was 1.2 years (range: 0-7.8 years). Twenty-one patients had disease manifestations from internal organs, including dysphagia (n = 16), myocarditis (n = 4) and interstitial lung disease (n = 1). At baseline, 14 patients (63%) received immunosuppressive treatment, most commonly methotrexate (n = 6), azathioprine (n = 5) and rituximab (n = 5) (Supplementary Table 1). The most frequent indications for IVIG treatment were insufficient effect of prior immunosuppressive medication (n = 18) and/or dysphagia (n = 5). The most frequently used prior immunosuppressive treatments were azathioprine (n = 11), methotrexate (n = 9) and rituximab (n = 9). Treatment with IVIG was given intravenously at a dose of 0.4 g per kilogram of body weight/day for five days at baseline. Further treatment series of IVIG were given intravenously over three or five consecutive days in one or more series as required. Eleven patients received only IVIG during the first three months of follow-up.

One patient died due to metastatic cancer during follow-up, and 21 patients completed the six-month study period. Among the 16 patients with dysphagia, improvement was observed in nine patients (56.3%) after three months and in ten patients (62.5%) after six months. Only one out of 16 patients (6.3%) experienced dysphagia relapse. Cardiac improvement was seen in three out of four patients (75.0%) after three months and in all four

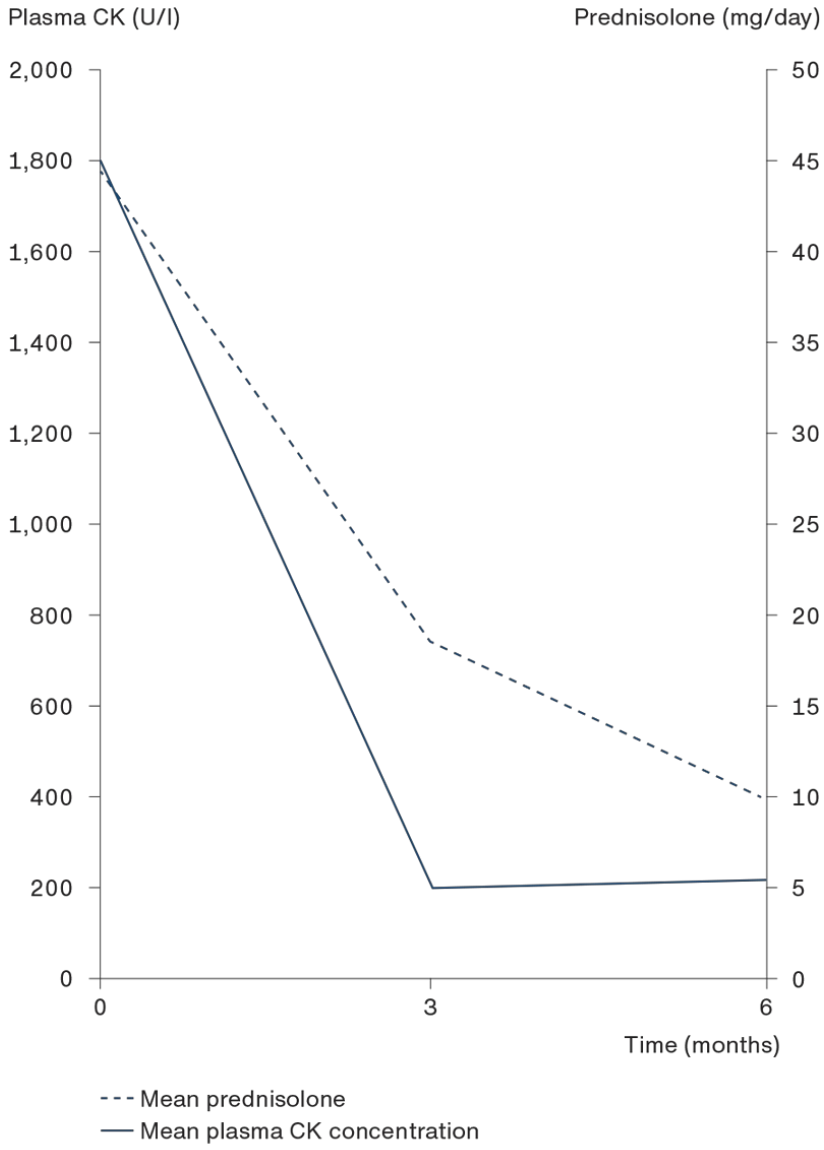
affected patients after six months. Functional improvements were observed in all tests at three and six months (**Table 1**). Among the eight patients who could not complete the 6MWT at baseline, one patient could perform the test at three months and two additional patients at six months. The average change in plasma CK concentrations was –1,628.5 U/l from baseline to month three (**Figure 1**), and 23.1 U/l from month three to month six. The mean daily prednisolone dose was significantly reduced, from 44.7 mg/day (SD: ± 29.7 mg/day) to 9.9 mg/day (SD: ± 12.8 mg/day) after six months. The largest reduction in prednisolone use occurred within the first three months (26.1 mg/day), with a further reduction by month six (34.8 mg/day).

TABLE 1 Functional test results, plasma creatine kinase concentrations, prednisolone dose and intravenous immunoglobulin dose at baseline and during follow-up. Data on functional tests (Manual Muscle Test-8, Six-minute Walk Test, and grip strength), plasma creatine kinase concentrations, daily prednisolone dose and cumulative intravenous immunoglobulin dose at baseline (intravenous immunoglobulin initiation) and at three and six months after treatment.

		Change from baseline to	
	Baseline	month 3	month 6
MMT8[a], mean ± SD (95% CI), score	25.63 ± 4.1	4.4 (1.6-7.3)	4.3 (0.7-7.8)
6MWT[b], mean ± SD (95% CI), m	341.8 ± 146.6	76.8 (–86.8-240.4)	8.3 (–188-204.8)
Grip strength[c], mean ± SD (95% CI), kg	58.2 ± 25.7	15.3 (–8.3-38.5)	13.5 (–16.8-43.8)
Plasma creatine kinase concentration, mean (95% CI), U/l	1,827.7	–1,628.5 (–2,554-–703)	–1,605.4 (–2,530-– 681)
Prednisolone dose, mean (95% CI), mg/day	44.7	–26.1 (–37-–15)	–34.8 (–48-–22)
Cumulative IVIG dose (% of total dose), g/kg	0	11.1 (77.3)	14.3 (100)

6MW = Six-minute Walk Test; IVIG = intravenous immunoglobulin; MMT8 = Manual Muscle Test-8.
a) Based on valid test results for 16 patients.
b) Based on valid test results for 7 patients.
c) Based on valid test results for 15 patients.

FIGURE 1 Mean levels of plasma creatine kinase (CK) concentration and prednisolone dose at the initiation of intravenous immunoglobulin (IVIG) therapy and during follow-up. The figure shows the mean serum CK levels and mean daily prednisolone dose at baseline (IVIG initiation) and at three and six months after treatment.



At the end of the six-month follow-up period, 17 patients were receiving immunosuppressive therapy. The most commonly used agents were methotrexate (n = 9), mycophenolate mofetil (n = 6) and rituximab (n = 5). An adverse event was recorded in one patient (4.5%) who was diagnosed with kidney cancer and developed a pulmonary embolism within three months of follow-up.

Discussion

This study shows that IVIG is an effective treatment and steroid-sparing agent in patients with various idiopathic inflammatory myositis subtypes, leading to a 77% reduction in glucocorticoid use after six months. Plasma CK

concentrations also significantly decreased. However, a slight increase from month three to six was observed, likely due to improved muscle function and mass during recovery. Additionally, we noted that the patients in this study successfully tapered to lower IVIG doses relatively quickly.

Although no standardised glucocorticoid tapering protocols have been studied for IVIG treatment in idiopathic inflammatory myositis, our findings are consistent with previous case series showing substantial glucocorticoid reductions (50-86%) in DM and PM, depending on phenotype and IVIG regimen [3-6]. Ohad et al. reported a mean 66.0% reduction in glucocorticoid use (mean initial dose: 53 mg/day; mean end dose: 18 mg/day) after a median of 31 months of follow-up in a case series comprising 23 patients with DM and PM who were successfully treated with IVIG [7]. Cherin et al. showed a reduction exceeding 50% in initial prednisone dose in 35 patients with refractory PM treated with IVIG (mean start dose: 32.7 mg/day; end dose: 21.9 mg/day) [8].

The main limitations of this study were the small sample size and missing outcome data due to the retrospective design.

Conclusions

IVIG appears to be an effective treatment in idiopathic inflammatory myositis, and its use could be considered earlier in the disease course, particularly in patients with contraindications to glucocorticoids. Further studies are needed to evaluate glucocorticoid tapering protocols in combination with IVIG treatment.

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