

LETTERS TO THE EDITOR

Pulsed intravenous methylprednisolone combined with oral steroids as the initial treatment of inflammatory myopathies

Pulsed intravenous methylprednisolone (IVMP) was given to 11 patients with polymyositis and dermatomyositis (group A) whom we studied prospectively. The outcome was compared with that of 14 patients who had been treated with oral steroids alone (group B). Group B comprised all cases of polymyositis and dermatomyositis with sufficient information in our past medical records studied retrospectively. Definite diagnoses of polymyositis and dermatomyositis were made in all cases.¹ Cases of myositis associated with cancer and inclusion body myositis were excluded.

For group A, methylprednisolone sodium succinate (500 mg diluted in 200 ml of 5% dextrose in water) was infused for two hours. A course of treatment consisted of three consecutive daily infusions, and was repeated three to nine times at intervals of a week, except in one patient who had only one course of IVMP. Oral steroids were given as in group B except on the days of IVMP.

For group B, treatment with oral prednisolone (1 mg/kg body weight per day) was started, maintained for two months, and then tapered down gradually over two years. When recovery was inadequate, the cases were kept either on the initial dose for up to four months, or the dose was increased up to 100 mg daily, or immunosuppressive agents were added. After two years, steroids were tapered off in some patients, and a small dose was maintained in others.

Muscle power was assessed both by neurologists and physiotherapists. The physiotherapists were not informed of how the steroids were given. The handicap was rated according to the disability grade.² The patients were followed up for one to five years.

Remission was defined as recovery of the strength of the weakest muscle or muscles by one grade or more on the British Medical Research Council (MRC) scale, in association with normalisation of the serum creatine kinase activity (falling to an activity lower than 200 IU/l). Partial remission was defined as recovery in either muscle strength or creatine kinase alone. Recovery of strength was judged in the same way as in the remission group. When neither muscle strength nor creatine kinase satisfied the standard of recovery, the case was included in the no improvement group. When recurrence occurred after remission, the patients were classified as remission and recurrence. Here recurrence was defined as worsening of muscle strength by one grade or more.

Groups A and B were not exactly comparable because of the difference in method of sampling, but to estimate characteristics of IVMP treatment, the two groups were compared statistically with Fisher's exact probability or Wilcoxon rank sum tests. The *p* values <0.05 were regarded as statistically

Comparison of cases of groups A and B and their outcome

Groups	A (IVMP + oral steroids)	B (oral steroids)
Number of cases	11	14
Age (y)*	54 (19)	43 (12)
Sex (female/male)	10/1	11/3
Diagnoses (PM/DM)	8/3	10/4
Duration (months)*	12.6 (15.8)	9.8 (6.6)
Oral prednisolone (mg)*	54.6 (9.3)	61.1 (13.9)
Disability grade**:		
Before steroids	6 (1)	4 (1)
On steroids	1 (1)	2 (1)
sCK*:		
Before steroids	3241 (2991)	4732 (3368)
On steroids	73 (62)	457 (614)
Weeks needed to normalise sCK*	6.6 (3.6) (n = 10)	11.7 (5.1) (n = 7)
Outcome:		
Remission	10	6
Remission/recurrence	1	4
Partial remission	0	2
No improvement	0	2

* Mean (standard deviation); **median (semi-interquartile range).

significant. Null hypotheses were tested two sided.

The table summarises the results. For group A, on average, 4.3 courses of IVMP was given to a case. Ten cases had persistent remission (remission group). Another, who had only one course of IVMP, had a recurrence after two years of remission (remission and recurrence group), requiring three more courses of IVMP to induce remission again. None needed immunosuppressive agents. One case required treatment for diabetes mellitus and candida stomatitis, but no change in the steroid treatment was needed.

For group B, six cases out of 14 had persistent remission (remission group). Four had a recurrence after a period of remission (remission and recurrence group). Two were classified as having partial remission, as they did not have recovery of muscle strength as defined, despite normal creatine kinase. Two belonged to the no-improvement group. Three had immunosuppressive agents six to 24 months after the initial treatment.

There was no significant difference between the groups for age, duration of the illness, or the maximum dose of oral prednisolone. The disability grade before treatment was higher in group A than in group B (*p* = 0.002).

Six months after initiation of steroids, half of the cases of group B and all the cases of group A were in remission. The difference was significant (*p* = 0.014). Persistent remission was seen in 10 out of 11 cases of group A and six out of 14 of group B (*p* = 0.034). There was no difference in the outcome between the cases of polymyositis and dermatomyositis.

The time needed for serum creatine kinase activity to return to normal was examined in cases in which the creatine kinase activity exceeded 500 IU/l before treatment and returned to normal later. The time was shorter in group A (*n* = 10) than in group B (*n* = 7; *p* = 0.014).

Studies of the effect of IVMP on adult dermatomyositis and polymyositis have been few.³⁻⁵ The effectiveness of IVMP at the initial stage of treatment of polymyositis and dermatomyositis has been mentioned.^{6,7}

In this study IVMP led to remission more often and serum creatine kinase returned to normal more rapidly than oral steroids alone. Among our cases, whereas none of those with a duration of illness longer than 24 months had remission with oral steroids alone, two cases with a duration of 30 and

54 months had a remission with IVMP.

Recurrence in a case of group A hinted that only one course of IVMP was not sufficient to induce sustained remission. Although immunosuppressive agents were not used in group A, they might enable us to decrease the number of courses of IVMP.

IVMP can help to decrease the complications of long term high dose oral steroids by shortening the time needed to induce remission and by increasing the rate of remission. Although this trial was open and small, the results suggest a beneficial effect of IVMP and warrant further studies.

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- Bohan A, Peter JB. Polymyositis and dermatomyositis (first of two parts). *N Engl J Med* 1975;292:344-7.
- Walton JN. *Disorders of voluntary muscle*. 4th ed. London: Churchill Livingstone, 1981: 452.
- Hrncir Z, Pidman V, Bradna P. Comment on the article by Laxer *et al* (letter). *Arth Rheum* 1989;32:505.
- Yanagisawa T, Sueishi M, Nawata Y, *et al*. Methylprednisolone pulse therapy in dermatomyositis. *Dermatologica* 1983;167: 47-51.
- Fessel WJ. Megadose corticosteroid therapy in systemic lupus erythematosus. *J Rheumatol* 1980;7:486-93.
- Walton J. The idiopathic inflammatory myopathies and their treatment. *J Neurol Neurosurg Psychiatry* 1991;54:285-7.
- Engel AG, Emslie-Smith AM. Inflammatory myopathies. *Curr Opin Neurol Neurosurg* 1989;2:695-9.

Familial inflammatory demyelinating polyneuropathy: a Guillain-Barré syndrome variant without autoimmune predilection

The putative pathogenesis of acute inflammatory demyelinating polyneuropathy (AIDP) is immune mediated.¹ Unlike other autoimmune diseases, such as systemic lupus erythematosus, rheumatoid arthritis, juvenile diabetes mellitus, and multiple