Treatment of chronic inflammatory demyelinating polyradiculoneuropathy (CIDP): steroids versus IVIg

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Chronic inflammatory demyelinating polyradiculoneuropathy (CIDP) is a chronic and often disabling neuropathy (1) that often respond to immune therapies including corticosteroids, plasma exchange and high-dose intravenous immunoglobulin (IVIg) (2) as also summarized in recent Cochrane reviews (3-5). It is however difficult to decide what therapy should be first used in CIDP. This decision should consider the short-term and long–term efficacy, the cost and side effects of each these therapies. A few randomized trials have shown a comparable short-term efficacy of IVIg and oral corticosteroids (6) and of IVIg and plasma exchange (7) in CIDP. Plasma exchange is considered however less suitable for long–term treatment than IVIg and is often reserved to patients with an insufficient response to IVIg or corticosteroids (8). A randomized trial comparing the six-month efficacy of monthly therapy with IVIg or intravenous methylprednisolone (IVMP) showed that IVIg was more frequently effective and tolerated (87.5%) than corticosteroids (47.6%) during the first six month of treatment (9). When effective however, corticosteroids were less frequently associated with deterioration than IVIg in the six months following therapy discontinuation. This was confirmed in the follow-up extension of the study (10) showing that the median time to deterioration was significantly longer after discontinuing IVMP (14 months) than IVIg (4.5 months). A similar proportion of patients however eventually deteriorated in the 42 months of median follow- up after discontinuing IVIg (87%) or IVMP (79%) confirming that none these therapies eventually cured CIDP. A similar discrepancy in the prolonged efficacy of steroids and IVIg can be derived from previous studies individually assessing the frequency of deterioration after IVIg or corticosteroids discontinuation. In a study comparing six-month treatment with IVIg and placebo, discontinuation of IVIg was followed by deterioration in 45% of the patients after 24 weeks (5.6 months) (11). Similar data were obtained in a study comparing the efficacy of interferon-beta and placebo in preventing disease progression after IVIg suspension. Clinical deterioration was observed within 16 weeks (3.7 months) in 48% of the patients suspending IVIg in the placebo group (12). On the other hand the extension of the PREDICT study that compared the efficacy of six month therapy with pulsed oral dexamethasone and daily oral prednisolone in CIDP showed that the median time to relapse after therapy discontinuation was 11 months for oral prednisolone and 17.5 months for pulsed oral dexamethasone, similar to the 14 months of our study (13). A similar more prolonged efficacy of steroids than of IVIg can be also assumed from a five year follow-up study of 70 patients with CIDP showing that the possibility to stop treatment tended to be more frequent in patients who responded to steroids than to IVIg (14). The long-term efficacy of continuous treatment with steroids was also shown in an uncontrolled retrospective study on 20 patients with CIDP, 15 of whom were continuously treated for 5 years with monthly high-dose of intravenous methylprednisolone irrespective of the possible phase of reactivity or remission of the disease (15). The improvement in these patients was maintained up to 5 years and, in those further treated, up to 10 years. Considering the safety of therapy, no significant differences in the proportion of patients experiencing adverse events was observed after six month therapy with IVIg or IVMP, even if it is not possible to exclude that this might occur after a more prolonged use of these therapies (9, 15). In addition it was recently confirmed that the annual cost per patient was considerably higher for patients with CIDP treated with IVIg (49,000£) than with other therapies (9400£) (16). In conclusion, even if IVIg are more frequently effective and possibly safer than steroids as initial treatment in CIDP, the latter have a more prolonged efficacy and consistently lower cost than IVIg.

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