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EFNS TASK FORCE

European Federation of Neurological Societies/Peripheral Nerve Society guideline on management of chronic inflammatory demyelinating polyradiculoneuropathy: report of a joint task force of the European Federation of Neurological Societies and the Peripheral Nerve Society

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Numerous sets of diagnostic criteria have sought to define chronic inflammatory demyelinating polyradiculoneuropathy (CIDP) and randomized trials and systematic reviews of treatment have been published. The objective is to prepare consensus guidelines on the definition, investigation and treatment of CIDP. Disease experts and a patient representative considered references retrieved from MEDLINE and Cochrane Systematic Reviews in May 2004 and prepared statements which were agreed in an iterative fashion. The Task Force agreed on good practice points to define clinical and electrophysiological diagnostic criteria for CIDP with or without concomitant diseases and investigations to be considered. The principal treatment recommendations were: (1) intravenous immunoglobulin (IVIg) or corticosteroids should be considered in sensory and motor CIDP (level B recommendation); (2) IVIg should be considered as the initial treatment in pure motor CIDP (Good Practice Point); (3) if IVIg and corticosteroids are ineffective plasma exchange (PE) should be considered (level A recommendation); (4) If the response is inadequate or the maintenance doses of the initial treatment are high, combination treatments or adding an immunosuppressant or immunomodulatory drug should be considered (Good Practice Point); (5) Symptomatic treatment and multidisciplinary management should be considered (Good Practice Point).

Objectives

To construct guidelines for the definition, diagnosis and treatment of chronic inflammatory demyelinating polyradiculoneuropathy (CIDP) based on the available evidence and, where adequate evidence was not available, consensus.

Background

The first proposal for diagnostic clinical criteria for CIDP was published by Dyck *et al*. [1,2] and included progressive course at 6 months, usually slowed nerve conduction velocities (and occurrence of conduction block), spinal fluid albumino-cytological dissociation, and nerve biopsy demonstrating segmental de- and remyelination, subperineurial or endoneurial oedema, and perivascular inflammation. Exclusion criteria were associated diseases, monoclonal gammopathy, and evidence of hereditary neuropathy. This descriptive proposal was the basis for a formalized set of criteria [3]. Mandatory inclusion and exclusion criteria reduced the required disease progression time to 2 months. Major laboratory criteria consisted of nerve biopsy abnormalities, motor conduction slowing to $<70\%$ in two nerves, and spinal fluid protein >450 mg/l. Fulfilment of all criteria was necessary for a definite diagnosis. Fulfilment of only two and one laboratory criteria led to the diagnostic categories of probable and possible, respectively. Research criteria were proposed by an American Academy of Neurology (AAN) in 1991 [4]. Fulfilment of clinical, physiological, pathological, and

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spinal fluid criteria led to three diagnostic categories (definite, probable and possible). Fulfilment of pathological criteria was necessary for a definite diagnosis. Physiological criteria for primary demyelination were very detailed, but restrictive when applied clinically as three of four nerve conduction parameters were required to be abnormal, even for the diagnosis of possible CIDP. However, the criteria for partial motor conduction block and abnormal temporal dispersion were probably not restrictive enough, as suggested by the American Association of Neuromuscular and Electrodiagnostic Medicine (AAEM) consensus criteria for the diagnosis of partial conduction block [5]. Patients who meet AAN research criteria certainly have CIDP, but many patients diagnosed as CIDP do not meet these criteria. In research studies of therapy of CIDP, several different sets of diagnostic criteria for CIDP have been created. These have been reviewed in a longer version of this paper which is available on the European Federation of Neurological Societies (EFNS) website (http://www.efns.org). For the present needs of the EFNS and Peripheral Nerve Society we offer the present diagnostic criteria to balance more evenly specificity (which needs to be higher in research than clinical practice) and sensitivity (which might miss treatable disease if set too high).

Since the first treatment trial of prednisone of Dyck *et al*. [2] a small body of evidence from randomized trials has accumulated to allow some evidence-based statements about treatments. These trials have been the subject of Cochrane reviews on which we have based some of our recommendations.

Search strategy

We searched MEDLINE from 1980 onwards on July 24, 2004 for articles (on 'chronic inflammatory demyelinating polyradiculoneuropathy' AND 'diagnosis' OR 'treatment' OR 'guideline') but found that the personal databases of Task Force members were more useful. We also searched the Cochrane Library in September 2004.

Methods for reaching consensus

Pairs of task force members prepared draft statements about definition, diagnosis and treatment which were considered at a meeting at the EFNS congress in September 2004. Evidence was classified as class I–IV and recommendations as level A–C according to the scheme agreed for EFNS guidelines [6]. When only class IV evidence was available but consensus could be reached the Task Force offered advice as good practice points [6]. The statements were revised and collated into a single document which was then revised iteratively until consensus was reached.

Results

Diagnostic criteria for CIDP

New criteria are currently being developed for defining CIDP from first principles by a group led by C.L. Koski but in the meantime the Task Force was obliged to develop their own criteria based on consensus. Criteria for CIDP are closely linked to criteria for detection of peripheral nerve demyelination. At least 12 sets of electrodiagnostic criteria for primary demyelination have been published, not only to identify CIDP (for review, see [7]). Nerve biopsy, usually the sural sensory nerve, is considered useful for confirming the diagnosis, but is a mandatory criterion for a definite diagnosis of CIDP only in the American Academy of Neurology criteria [4]. The available evidence indicates that sural nerve biopsy can provide supportive evidence for the diagnosis of CIDP, but positive findings are not specific and negative findings do not exclude the diagnosis. Increased spinal fluid protein occurs in at least 90% of patients. Therefore, increased protein levels can be used as a supportive but not mandatory criterion for the diagnosis. Integration of magnetic resonance imaging (MRI) abnormalities of nerve roots, plexuses, and peripheral nerves in diagnostic criteria for CIDP may enhance both sensitivity and specificity and may therefore be useful as a supportive criterion for the diagnosis. As most patients with CIDP respond to steroids, plasma exchange, or intravenous immunoglobulin (IVIg), a positive response to treatment may support the diagnosis and has been suggested as another diagnostic criterion [8]. There is only class IV evidence concerning all these matters. Nevertheless the Task Force agreed good practice points to define clinical and electrophysiological diagnostic criteria for CIDP with or without concomitant diseases (Tables 1–6).

Investigation of CIDP

Based on consensus expert opinion, CIDP should be considered in any patient with a progressive symmetrical or asymmetrical polyradiculoneuropathy in whom the clinical course is relapsing and remitting or progresses for more than 2 months, especially if there are positive sensory symptoms, proximal weakness, areflexia without wasting, or preferential loss of vibration or joint position sense. Electrodiagnostic tests are mandatory and the major features suggesting a diagnosis of CIDP are listed in Table 2. Minor electrodiagnostic features are greater abnormality of median than sural nerve sensory action potential, reduced sensory nerve conduction velocities and F-wave chronodispersion. If electrodiagnostic criteria for definite CIDP are not met initially, repeat

Table 1 Clinical diagnostic criteria

I. Inclusion criteria
A. Typical CIDP
Chronically progressive, stepwise, or recurrent symmetric proximal and distal weakness and sensory dysfunction of all extremities, developing over at least 2 months; cranial nerves may be affected, and
Absent or reduced tendon reflexes in all extremities
B. Atypical CIDP
One of the following, but otherwise as in A (tendon reflexes may be normal in unaffected limbs)
Predominantly distal weakness (distal acquired demyelinating sensory, DADS)
Pure motor or sensory presentations, including chronic sensory immune polyradiculoneuropathy affecting the central process of the primary sensory neuron [27]
Asymmetric presentations (multifocal acquired demyelinating sensory and motor, MADSAM, Lewis–Sumner syndrome)
Focal presentations (e.g. involvement of the brachial plexus or of one or more peripheral nerves in one upper limb)
Central nervous system involvement (may occur with otherwise typical or other forms of atypical CIDP)
II. Exclusion criteria
Diphtheria, drug or toxin exposure likely to have caused the neuropathy
Hereditary demyelinating neuropathy, known or likely because of family history, foot deformity, mutilation of hands or feet, retinitis pigmentosa, ichthyosis, liability to pressure palsy
Presence of sphincter disturbance
Multifocal motor neuropathy
Antibodies to myelin-associated glycoprotein

Table 2 Electrodiagnostic criteria

I. Definite: at least one of the following
A. At least 50% prolongation of motor distal latency above the upper limit of normal values in two nerves, or
B. At least 30% reduction of motor conduction velocity below the lower limit of normal values in two nerves, or
C. At least 20% prolongation of F-wave latency above the upper limit of normal values in two nerves (> 50% if amplitude of distal negative peak compound muscle action potential (CMAP) < 80% of lower limit of normal values), or
D. Absence of F-waves in two nerves if these nerves have amplitudes of distal negative peak CMAPs at least 20% of lower limit of normal values + at least one other demyelinating parameter [a] in at least one other nerve, or
E. Partial motor conduction block: at least 50% amplitude reduction of the proximal negative peak CMAP relative to distal, if distal negative peak CMAP at least 20% of lower limit of normal values, in two nerves, or in one nerve + at least one other demyelinating parameter[a] in at least one other nerve, or
F. Abnormal temporal dispersion (> 30% duration increase between the proximal and distal negative peak CMAP) in at least two nerves, or
G. Distal CMAP duration (interval between onset of the first negative peak and return to baseline of the last negative peak) of at least 9 ms in at least one nerve + at least one other demyelinating parameter [a] in at least one other nerve
II. Probable
At least 30% amplitude reduction of the proximal negative peak CMAP relative to distal, excluding the posterior tibial nerve, if distal negative peak CMAP at least 20% of lower limit of normal values, in two nerves, or in one nerve + at least one other demyelinating parameter [a] in at least one other nerve
III. Possible
As in 'I' but in only one nerve

To apply these criteria the median, ulnar (stimulated below the elbow), peroneal (stimulated below the fibular head) and tibial nerves on one side are tested. Temperatures should be maintained to at least 33°C at the palm and 30°C at the external malleolus. (Good Practice Points). Further technical details are given in the accompanying web document (http://www.efns.org) and see van den Bergh and Piéret [7] .

[a]Any nerve meeting any of the criteria A–G.

electrodiagnostic testing in more nerves or at a later date, cerebrospinal fluid (CSF) examination, MRI of the spinal roots, brachial or lumbar plexus and nerve biopsy should be considered (Table 6). The nerve for biopsy should be clinically and electrophysiologically affected and is usually the sural, but occasionally the superficial peroneal, superficial radial, or gracilis motor nerve. Sometimes the choice of nerve may be assisted by MRI. The minimal examination should include paraffin sections, immunohistochemistry and semithin resin sections. Electron microscopy and teased fibre preparations are highly desirable. There are no specific appearances. Supportive features are endoneurial oedema, macrophage-associated demyelination, demyelinated and to a lesser extent remyelinated nerve fibres, onion bulb formation, endoneurial mononuclear cell infiltration, and variation between fascicles. During the diagnostic workup investigations to discover possible concomitant diseases should be considered (Good Practice Points, Table 6).

Treatment of CIDP

Corticosteroids

In one unblinded randomized controlled trial (RCT) with 28 participants prednisone was superior to no treatment [2,9] (class II evidence). Six weeks of oral prednisolone starting at 60 mg daily produced benefit which was not significantly different from that produced by a single course of IVIg 2.0 g/kg [10,11] (class II evidence). However there are many observational studies reporting a beneficial effect from corticosteroids except in pure motor CIDP in which they have sometimes appeared to have a harmful effect [12]. Consequently a trial of corticosteroids should be considered in all patients with significant disability (level B recommendation). There is no evidence and no consensus about whether to use daily or alternate day prednisolone or prednisone or intermittent high dose monthly intravenous or oral regimens [13].

Plasma exchange

Two small double-blind RCTs with altogether 47 participants showed that PE provides significant short-

Table 3 Supportive criteria

A. Elevated cerebrospinal fluid protein with leucocyte count $< 10/mm^3$ (level A recommendation)
B. Magnetic Resonance Imaging showing gadolinium enhancement and/or hypertrophy of the cauda equina, lumbosacral or cervical nerve roots, or the brachial or lumbosacral plexuses (level C recommendation)
C. Nerve biopsy showing unequivocal evidence of demyelination and/or remyelination in ≥5 fibres by electron microscopy or in > 6 of 50 teased fibres
D. Clinical improvement following immunomodulatory treatment (level A recommendation)

Table 4 CIDP in association with concomitant diseases

One of the following is present
(a) Conditions in which, in some cases, the pathogenesis and pathology are thought to be the same as in CIDP
Diabetes mellitus
HIV infection
Chronic active hepatitis
IgG or IgA monoclonal gammopathy of undetermined significance
IgM monoclonal gammopathy without antibodies to myelin-associated glycoprotein
Systemic lupus erythematosus or other connective tissue disease
Sarcoidosis
Thyroid disease
(b) Conditions in which the pathogenesis and pathology may be different from CIDP
Borrelia burgdorferi infection (Lyme disease)
IgM monoclonal gammopathy of undetermined significance with antibodies to myelin-associated glycoprotein[a]
POEMS syndrome
Osteosclerotic myeloma
Others (vasculitis, haematological and non-haematological malignancies, including Waldenström's macroglobulinaemia and Castleman's disease)

[a]Patients with antibodies to myelin-associated glycoprotein are considered to have a disease with a different mechanism and are excluded. See Table 1

Table 5 Diagnostic categories

Definite CIDP
Clinical criteria I A or B and II with Electrodiagnostic criteria I; or Probable CIDP + at least one Supportive criterion; or Possible CIDP + at least two Supportive criteria
Probable CIDP
Clinical criteria I A or B and II with Electrodiagnostic criteria II; or Possible CIDP + at least one Supportive criterion
Possible CIDP
Clinical criteria I A or B and II with Electrodiagnostic criteria III
CIDP (definite, probable, possible) associated with concomitant diseases

Table 6 Investigations to be considered

To identify CIDP
Nerve conduction studies
CSF cells and protein
MRI spinal roots, brachial plexus and lumbosacral plexus
Nerve biopsy
To detect concomitant diseases
Serum and urine paraprotein detection by immunofixation (repeating this should be considered in patients who are or become unresponsive to treatment)
Oral glucose tolerance test
Complete blood count
Renal function
Liver function
HIV antibody
Hepatitis B and C serology
Borrelia burgdorferi serology
C reactive protein
Antinuclear factor
Extractable nuclear antigen antibodies
Thyroid function
Angiotensin-converting enzyme
Chest radiograph
Akeletal survey (repeating this should be considered in patients who are or become unresponsive to treatment)
To detect hereditary neuropathy
Examination of parents and siblings
PMP22 gene duplication or deletion (especially if slowing of conduction is uniform and no evidence of partial motor conduction block or abnormal temporal dispersion)
Gene mutations known to cause Charcot-Marie-Tooth (CMT)1 or hereditary neuropathy with liability to pressure palsies

term benefit in about two-thirds of patients but rapid deterioration may occur afterwards [14–16] (class I evidence). Plasma exchange might be considered as an initial treatment (level A recommendation). However because adverse events related to difficulty with venous access, use of citrate and haemodynamic changes are not uncommon, either corticosteroids or IVIg should be considered first (Good Practice Point).

Intravenous immunoglobulin

Meta-analysis of four double blind RCTs with altogether 113 participants showed that IVIg 2.0 g/kg produces significant improvement in disability lasting 2–6 weeks [11,17–20] (class I evidence). Because the benefit from IVIg is short lived, treatment, which is expensive, needs to be repeated at intervals which need to be judged on an individual basis. Crossover trials have shown no significant short-term difference between IVIg and plasma exchange [21] or between IVIg and prednisolone [10] but the samples were too small to establish equivalence (both class II evidence).

Immunosuppressive agents

No RCTs have been reported for any immunosuppressive agent except for azathioprine which showed no benefit when added to prednisone in 14 patients [22,23]. Immunosuppressive agents (Table 7) are often used together with corticosteroids to reduce the need for IVIg or PE or to treat patients who have not responded

Table 7 Immunosuppressant and immunomodulatory drugs which have been reported to be beneficial in CIDP (class IV evidence, see 23 for review)

Anti-CD20 (rituximab)
Azathioprine
Cyclophosphamide
Ciclosporin
Etanercept
Interferon alpha
Interferon beta-1a
Mycophenolate mofetil

to any of these treatments but there is only class IV evidence on which to base this practice [23]. More research is needed before any recommendation can be made. In the meantime immunosuppressant treatment may be considered when the response to corticosteroids, IVIg or PE is inadequate (Good Practice Point).

Interferons

One crossover trial of interferon (IFN) beta-1a for 12 weeks did not detect significant benefit [24] but the trial only included 10 patients. In a more recent non-randomized open study of intramuscular beta IFN-1a 30 μg weekly seven of 20 patients treated showed clinical improvement, 10 remained stable and three worsened [25]. An open study of IFN-α showed benefit in nine of 14 treatment-resistant patients [26] and there have been other favourable smaller reports. In the absence of evidence IFN treatment may be considered when the response to corticosteroids, IVIg or PE is inadequate (Good Practice Point).

Initial management (Good Practice Points)

Patients with very mild symptoms which do not or only slightly interfere with activities of daily living may be monitored without treatment. Urgent treatment with corticosteroids or IVIg should be considered for patients with moderate or severe disability, e.g. when hospitalization is required or ambulation is severely impaired. Common initial doses of corticosteroids are prednisolone or prednisone 1 mg/kg or 60 mg daily but there is a wide variation in practice [13]. The usual first dose of IVIg is 2.0 g/kg given as 0.4 g/kg on 5 consecutive days. Contraindications to corticosteroids will influence the choice towards IVIg and vice versa. For pure motor CIDP IVIg treatment should be first choice and if corticosteroids are used, patients should be monitored closely for deterioration.

Long-term management (Good Practice Points)

No evidence-based guidelines can be given as none of the trials systematically assessed long-term management. Each patient requires assessment on an individual basis. For patients starting on corticosteroids, a course of up to 12 weeks on their starting dose should be considered before deciding whether there is a no treatment response. If there is a response, tapering the dose to a low maintenance level over 1 or 2 years and eventual withdrawal should be considered. For patients starting on IVIg, observation to discover the occurrence and duration of any response to the first course should be considered before embarking on further treatment. Between 15% and 30% of patients do not need further treatment. If patients respond to IVIg and then worsen, further and ultimately repeated doses should be considered. Repeated doses may be given over 1 or 2 days. The amount per course needs to be titrated according to the individual response. Repeat courses may be needed every 2–6 weeks. If a patient becomes stable on a regime of intermittent IVIg, the dose per course should be reduced before the frequency of administration is lowered. If frequent high dose IVIg is needed, the addition of corticosteroids or an immunosuppressive agent should be considered. Approximately 15% of patients fail to respond to any of these treatments. Some probably do not appear to respond because of severe secondary axonal degeneration which takes years to improve.

General treatment

There is a dearth of evidence concerning general aspects of treatment for symptoms of CIDP such as pain and fatigue. There is also a lack of research into the value of exercise and physiotherapy and the advice which should be offered concerning immunizations. International and national support groups offer information and support and physicians may consider putting patients in touch with these organizations at http://www.guillain-barre.com/or http://www.gbs.org.uk (Good Practice Point).

Recommendations

Good Practice Points for defining diagnostic criteria for CIDP:

1 Clinical: typical and atypical CIDP (Table 1);
2 Electrodiagnostic: definite, probable and possible CIDP (Table 2);
3 Supportive: including CSF, MRI, nerve biopsy and treatment response (Table 3);
4 CIDP in association with concomitant diseases (Table 4);
5 Categories: definite, probable, and possible CIDP with or without concomitant diseases (Table 5).

Good Practice Points for diagnostic tests:

1 Electrodiagnostic tests are recommended in all patients (Good Practice Point);
2 CSF, MRI and nerve biopsy should be considered in selected patients (Good Practice Point);

3 Concomitant diseases should be considered in all patients but the choice of tests will depend on the clinical circumstances (Table 6).

Recommendations for treatment

For induction of treatment:

1 IVIg or corticosteroids should be considered in sensory and motor CIDP in the presence of troublesome symptoms (level B recommendation). The presence of relative contraindications to either treatment should influence the choice (Good Practice Point).
2 The advantages and disadvantages should be explained to the patient who should be involved in the decision making (Good Practice Point).
3 In pure motor CIDP IVIg should be considered as the initial treatment (Good Practice Point).
4 If IVIg and corticosteroids are ineffective PE should be considered (level A recommendation).

For maintenance treatment:

1 If the first-line treatment is effective continuation should considered until the maximum benefit has been achieved and then the dose reduced to find the lowest effective maintenance dose (Good Practice Point).
2 If the response is inadequate or the maintenance doses of the initial treatment are high, combination treatments or adding an immunosuppressant or immunomodulatory drug may be considered (Table 7) (Good Practice Point).
3 Advice about foot care, exercise, diet, driving and lifestyle management should be considered. Neuropathic pain should be treated with drugs according to EFNS guideline on treatment of neuropathic pain (N. Attal, in prep.). Depending on the needs of the patient, orthoses, physiotherapy, occupational therapy, psychological support and referral to a rehabilitation specialist should be considered (Good Practice Points).
4 Information about patient support groups should be offered to those who would like it (Good Practice Point).

Anticipated date for updating this guideline

Not later than October 2008.

Conflicts of interest

The following authors have reported conflicts of interest as follows: R. Hughes personal none, departmental research grants or honoraria from Bayer, Biogen-Idec, Schering-LFB and Kedrion; D. Cornblath personal honoraria from Aventis Behring and Baxter, A Hahn personal honoraria from Baxter, Bayer, Biogen-Idec; C. Koski personal honoraria from American Red Cross, Baxter, Bayer, ZLB-Behring; J.M. Léger personal none, departmental research grants or honoraria from Biogen-Idec, Baxter, Laboratoire Français du Biofractionnement (LFB), Octapharma; E. Nobile-Orazio personal from Kedrion, Grifols, Baxter, LFB (and he has been commissioned by Kedrion and Baxter to give expert opinions to the Italian Ministry of Health on the use of IVIg in dysimmune neuropathies); J. Pollard departmental research grants from Biogen-Idec, Schering; P. van Doorn personal none, departmental research grants or honoraria from Baxter and Bayer. The other authors have nothing to declare.

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