

Guidance on the management of CFS/M.E.

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■ Introduction

Chronic Fatigue Syndrome (CFS/M.E.) is a common disorder, or spectrum of disorders, that causes substantial ill health and disability in people of all ages. It has characteristic features but is highly variable, including severity and duration, and lacks specific disease markers. Patients can be helped substantially by quite straightforward approaches.

This guide has been produced by Action for M.E to assist GPs in the assessment and management of patients with CFS/M.E. It is based on the Chief Medical Officer's Working Group Report on CFS/M.E.¹ The Report:

- Recognises CFS/M.E. and its impact.
- Sets out an approach to clinical diagnosis.
- Outlines some useful general principles to guide management, based on evidence and experience; no approach is universally beneficial, and none is a cure.
- Emphasises that each individual needs a flexible management plan, using specific strategies and therapies tailored to their circumstances.
- Describes treatment strategies including cognitive behavioural therapy, graded exercise and pacing. Intrusive symptoms should be controlled.
- Indicates that most people can expect some improvement with time and appropriate treatment, justifying a positive but realistic attitude towards rehabilitation and recovery.
- Reinforces that children have particular needs, and especially require a multidisciplinary approach.



■ Epidemiology

- The population prevalence of CFS/M.E. is 0.2% – 0.4%.
- It can occur at any age, most commonly early 20s to mid-40s, and, in children, between 13 and 15.
- It is about twice as common in women, and affects all social classes and ethnic groups.

■ Prognosis

Prognosis is extremely variable. Many patients have a fluctuating course with some setbacks. Most will improve to some degree.

- Many can improve relatively quickly.
- Some have more prolonged illness; in an important minority, the duration is very long.
- Prognosis is better if a diagnosis is made and appropriate treatment and support are provided; substantial improvement in both function and symptoms may be seen.

■ Aetiology and pathogenesis

The aetiology and pathogenesis of CFS/M.E. are not known. There is commonly an infection or other trigger, and individuals may have predisposing factors that make them more vulnerable to the occurrence or persistence of a fatigue state.

Various immune, endocrine, musculoskeletal, and neurological abnormalities have been described which could be primary processes or secondary consequences.

■ Symptoms

The characteristic features of CFS/M.E. are *overwhelming fatigue* and *malaise*, affecting both physical and cognitive functioning, often with a wide range of other symptoms.

Typically, increased physical or mental activity prompts worsening of symptoms with a delayed impact, felt later the same day, the next day or even later. This is followed by a recovery period, which may take days or weeks. In some, a *cumulative* impact is seen after a period of sustained activity.

Common symptoms include:

Fatigue

- physical and cognitive fatigue/exhaustion typically delayed; often profound, quite unlike everyday tiredness

Cognitive impairment

- poor concentration
- poor short-term memory
- difficulties with word finding and complex or multiple tasks

Malaise

- pervasive, often flu-like malaise, typically exacerbated by exertion

Pain

- muscle pain (sometimes with twitching or cramps)
- joint pain
- neuropathic pain (+/- paraesthesiae)
- fibromyalgia
- headache (often migrainous)

Sleep disturbance

- insomnia
- early morning waking
- unrefreshing sleep
- disturbed sleep/wake cycle
- hypersomnia

Digestive disturbances (IBS)

- nausea
- loss of appetite
- indigestion
- excessive wind
- bloating
- abdominal cramps
- alternating diarrhoea and constipation
- food intolerance

Other symptoms and signs

- temperature disturbance
- vertigo
- postural hypotension
- light or noise sensitivity
- alcohol intolerance
- sensitivity to medication and other substances
- recurrent sore throat
- lymphadenopathy (tender and/or enlarged)

■ Evaluation and diagnosis

Making a firm, or even a provisional diagnosis of CFS/M.E. is an essential first step in active management. It should be a positive diagnosis, based on pattern recognition of the characteristic symptoms and the way they behave, together with the exclusion of alternative diagnoses. It is not simply a diagnosis of exclusion. Formal diagnostic criteria for clinical use have not been developed yet, and existing research criteria are too restrictive for clinical practice.

Full clinical history

As the crucial tool in diagnosis is the clinical history, sufficient time should be allowed for patients to give a narrative account of their illness experience. Current status should be assessed in relation to previous healthy functioning. Mental state assessment and psychosocial assessment are needed, because mood disorder may result from CFS/M.E. or coexist and interact with it, as well as being a differential diagnosis.

Physical examination

Findings on physical examination are frequently normal in CFS/M.E., but it may be helpful in excluding other conditions.

Basic screening tests

Simple screening tests should be undertaken to exclude other conditions; additional tests may be needed to evaluate specific symptoms or features.

- full blood count
- C-reactive protein (CRP)
- blood biochemistry including creatinine, urea and electrolytes, calcium, liver enzymes and glucose
- thyroid function tests
- urinalysis

Differential diagnosis

This may include:

- adrenal insufficiency
- anaemia
- chronic infection
- coeliac disease
- immunodeficiency
- malignancy
- mood disorders (anxiety, depression)
- multiple sclerosis
- myasthenia gravis
- primary sleep disorder
- rheumatic diseases
- somatisation disorder
- thyroid disease

If symptoms suggest that any of these are likely, appropriate investigations should be done.

Onset

The onset of CFS/M.E. can be sudden or gradual. In cases of sudden onset, the condition commonly follows an acute infective episode.

Timescale

A working or interim diagnosis is better than none and allows active management to begin. For most adults, six weeks from the onset of abnormal fatigue is a time to be considering CFS/M.E. as a differential diagnosis; advice and management should reflect the possibility. By six months, if symptoms persist, a provisional diagnosis can usually be confirmed. These timescales should be shorter in children (see page 6).

■ Treatment and rehabilitation

In addition to making a clear diagnosis and ongoing monitoring and support, clinicians should use their generic skills in chronic disease management.

Three specific strategies have been identified as potentially beneficial in modifying this illness:

- **graded activity/exercise**
- **cognitive behavioural therapy**
- **pacing**

Continuing care will entail advice on balancing rest and activity, and maximising potential by setting realistic goals. Basic lifestyle management, including **pacing**, should be informed by the principles and practice of graded rehabilitation, flexibly applied to achieve sustainable progress. Some patients may benefit from a more structured approach to rehabilitation using **cognitive behavioural therapy** and/or **graded activity/exercise, with a therapist who has appropriate training and experience**.

Intrusive symptoms or those which are likely to maintain illness, such as sleep disturbance, pain and mood changes should be controlled. Assistance with benefits, employment and education can also make a substantial difference.

Activity Management

Patients will need guidance on this, although pacing can largely be a self-management technique. The aim of the underlying approach to **pacing** is to establish sustainable activity levels that avoid the 'boom and bust' pattern so often seen when patients attempt too high a level of functioning. Too much activity or too much rest can each be unhelpful. The actual level depends on the severity of the limitation. A consistent baseline of activity (mental as well as physical) should be established that avoids delayed setbacks. Baselines may sometimes need to start at a very low level. A diary may help to establish patterns. From this baseline, gradual incremental increases can be attempted and modified in the light of experience. It is important to adjust any programme if there is an adverse reaction rather than increase the activity regardless. Appropriate goals can be set and then built up as tolerance increases. Balance is the watchword throughout: balance between activity and rest/relaxation; balance between physical and mental tasks; balance between work and leisure; balance between needs and wants.

Making these steps can be hard to achieve, either at a practical level or in terms of personal adjustment of attitude and behaviour. This is where guidance from others, especially suitably trained professionals, can make a difference. An ambulant patient, who is able to manage all basic activities of daily living, could benefit from a formal programme of **graded aerobic exercise under supervision**; however, patients with a lower level of functioning can use their normal daily tasks as the basis for a structured approach to graded increases in activity. Different patients will experience different difficulties in modifying activity, attitude or behaviour which is where **cognitive behavioural therapy** may prove beneficial; the type and amount of assistance, advice and guidance offered should reflect their own needs.

Symptom control

Symptom control is important for patients with CFS/M.E., not only to improve quality of life, but also to facilitate the patients' ability to engage in specific therapy programmes. **Patients with CFS/M.E. are often relatively intolerant of medication**, so it is wise to start with lower doses and to use agents that are less likely to have adverse effects. Some agents, including tricyclics, may encourage weight gain, so patients who are physically inactive should be warned to take care over calorie intake when on them.

Pain

This can be burdensome and intrusive. Simple analgesics may suffice, on a regular or as required basis, to gain a level of control. Agents that 'gate out' pain, such as low dose tricyclics, amitriptyline or nortriptyline, or the anticonvulsants gabapentin, sodium valproate or carbamazepine can be useful, especially for neuropathic pain. If muscle pain is accompanied by twitching or cramps, muscle relaxants (eg baclofen) may be helpful. Other approaches for chronic pain management may be valuable, including the use of psychological techniques.

Sleep

Establishing a sleep routine should be the first approach, looking at factors that enhance sleep (e.g. setting a routine, avoiding caffeine). If medication needs to be used, low doses of tricyclic and related agents are often effective in restoring sleep quality and rhythm, and are preferable to hypnotic agents. Amitriptyline, nortriptyline, doxepin and trazodone are useful. Short-term or intermittent use of mild, short-acting hypnotics may help re-establish sleep routines.

Mood disorders

Patients with CFS/M.E. commonly suffer from depressive and anxiety (mood) disorders. These can exacerbate CFS/M.E. directly (e.g. increasing fatigue and cognitive difficulties) or indirectly (e.g. through insomnia). Mood disorders significantly increase risk of suicide and are associated with worse prognosis. Appropriate treatment of depression and anxiety improves outcomes and quality of life. Treatments include medication and/or psychotherapy. It must be made clear that the agents are being used to treat mood and that they are not treatments for CFS/M.E.

The usual agents may be considered, but it is important to take account of the particular sensitivity of CFS/M.E. patients to medication, especially psychotropic medication, in both choice of agent and in dosage used. Account must also be taken of sleep pattern in the choice of agents or combinations of agents.

CFS/M.E. patients with depression are often well suited by citalopram or sertraline, which can also reduce anxiety and panic attacks; they are often poorly tolerant of more activating agents, such as fluoxetine, paroxetine and venlafaxine, especially if they have sleep disturbance and/or limited functional capacity.

Tricyclic and related agents, which in low dosage are used for sleep and pain, may also be useful as antidepressants, but intolerance is more of a problem where higher doses are needed. Agents with less anti-muscarinic activity, such as doxepin or trazodone, may be better tolerated.

Limited and short-term use of anxiolytics may sometimes be helpful as part of a wider management plan.

Patients may need psychotherapy or anxiety management, alone or in conjunction with such agents. Expert guidance should be sought where major mood disturbance is present, and urgently where there is suicidal ideation or risk.

Headaches

These sometimes have a migrainous character and agents used for migraine can be helpful. Where they are less frequent or severe, triptans can help. More frequent headaches may merit a trial of migraine prophylaxis, such as low-dose tricyclics, pizotifen, or sodium valproate (beta-blockers are often poorly tolerated in CFS/M.E.). Some patients find that dietary change can reduce such headaches, and trials of dairy or wheat exclusion may be worth considering in patients with recalcitrant headaches.

Dizziness

This is commonly of two types: postural hypotension or rotational vertigo. Both may be assisted by advice and care with changes in position. Cardiovascular exercises may help restore orthostatic responses. In some patients, vestibular sedatives such as cinnarazine or betahistine can be helpful, when symptoms are intrusive or for travel and other likely triggers.

Abdominal symptoms

Patients with CFS/M.E. often have features of irritable bowel syndrome. Usual approaches to this syndrome should be tried, such as reducing fibre in diet and use of antispasmodics such as mebeverine or alverine. In some patients, exclusion diets have proved helpful, notably those excluding wheat and/or dairy products; these may be worth a trial. Specialist referral may be appropriate.

■ Approach to management

A mutually agreed management plan is crucial. Such management plans are ideally patient-centred and utilise appropriately trained professionals in multidisciplinary teams.

GPs should usually be able to manage most patients with CFS/M.E., as with many other chronic conditions. Shared care with specialists, for diagnosis or development of a management plan, may help where problems are complex, severe or prolonged.

The severely affected, e.g. bed-bound or house-bound, will not only have the greatest need, but will also be those who are least able to access care and support. Appropriate domiciliary provision should be provided. Special difficulties arise from being physically unable to access the many services that now require patients to be ambulant, or to travel to the point of service assessment or delivery. Immobility and isolation can easily lead to what some people describe as 'invisibility'.

Successful management of any long-term illness includes the patient as a partner in their own care, resulting in greater self-reliance, better adherence, higher satisfaction, and greater continuity of care. A partnership approach acknowledges that patients must cope continuously with their illness. Education on the illness and on self-management can empower patients to take an active role in their care and regain control of their lives. Information and support are needed from early on. Action for M.E. offers information and support to patients with CFS/M.E. and their carers.

■ Children and young people

Children and young people can be profoundly affected by CFS/M.E. The principles of care of children and young people are common to many chronic conditions.

Their rights to be heard, to have their views taken into account, to access quality medical treatment, and to be protected from abuse both by individuals and by systems, need particular attention.

Nature and impact

Most of the issues covered for adults also apply to children, but some need different emphasis.

CFS/M.E. can threaten physical, emotional and intellectual development, and can disrupt education, social and family life. Social isolation and school absence cause particular difficulties. Most studies suggest that young people have a better prognosis than adults.

Characteristic clinical features which are similar include impaired physical and cognitive functioning plus malaise and a wide range of other symptoms (in particular, abdominal pain, nausea, and appetite changes, leading to weight gain or loss). 'Fatigue' as such may not be a prominent complaint, but physical and/or cognitive activity are usually restricted, and interfere with schooling. Increasing activity prematurely often delays recovery, although younger patients do not always obviously have the typical delayed exacerbation of symptoms seen in adults.

Evaluation and diagnosis

An especially prompt and clear diagnosis is needed, while other illnesses and complications must remain in mind. **When a child or young person has symptoms that have affected school attendance for 15 days, active steps should be undertaken to identify the cause, from a list that includes CFS/M.E.** Evaluation should be as for adults, but with specific attention paid to sleep, mental health, physical and social activities, family interactions and education.

Other conditions that present with school absence are important differential diagnoses that need to be considered early and ruled out or treated. Possibilities include physical and mental illness, particularly depression, as well as school phobia, eating disorders, and, rarely, child abuse.

Approach to management

Clinicians face additional difficulties in supporting and managing younger patients and their families. **Listening to the young person and their family, hearing, and understanding what they say is vital.**

Ideal management also includes:

- Patient-centred, community-based, multidisciplinary, co-ordinated care.
- A clinician co-ordinating care and involvement of other agencies such as the Local Education Authority and school to meet the child's educational needs.
- A detailed, flexible treatment plan, mutually agreed and regularly reviewed in partnership with the child and family.
- A common approach to the condition among professionals as far as possible.

An ideal care pathway would involve:

- Early recognition in primary care with specialist confirmation, if appropriate.
- Evolution of management and follow-up adapted to individual and family circumstances, local expertise and specialist interest.
- Follow up in primary care, or by mutually agreed community or hospital specialist, for those children not in school and receiving home tuition.
- Community paediatric and/or specialist services available widely, and to all with prolonged school absence.
- Hospital admission reserved for difficult diagnostic assessment and for dealing with severe complications, serious intercurrent illness, or other specific problems.

Primary care services may require additional or specialist support. For severely affected young people, GPs can access the domiciliary visiting service by consultants. Community-based members of professions allied to medicine are available to provide additional multidisciplinary support to patients (e.g. social worker, joint work or referral to Child and Adolescent Mental Health Services [CAMHS], occupational therapy or physiotherapy services). Community nurses, paediatric nurse specialists and consultants, school nurses and home-care teams may be particularly valuable to support and co-ordinate care and management.

Information and support

All clinicians can:

- Listen to the patient, recognise and believe their individual experience.
- Agree a name for the condition.
- Offer some form of psychological support to the family, including acknowledgement of uncertainty and the impact of this uncertainty.
- Provide information on and discuss the condition, self-management, helpful therapies, and other sources of support and services.
- Offer support and advice on improving well-being and symptomatic treatment.

Valuable support includes keeping a diary, advice in managing limited energy and balancing types of activity. Desired outcomes and a timetable for achievement of defined goals can be mutually agreed. All concerned need to be aware that setbacks, or an inability to reach certain goals, are not uncommon. Ultimately, an individualised non-coercive programme for return to education and social functioning can be agreed.

Education

The Department for Education and Skills states that 'a child or young person who is unable to attend school because of medical needs should have their educational needs identified and receive educational support quickly and effectively.

For children and young people's needs to be identified early, cross-agency working and liaison between health services, social services, and learning services is essential. This cannot be achieved without the support of the National Health Service. There should be close liaison between hospital consultants, GPs, schools and Local Education Authorities (LEAs) so that ill pupils can be provided with educational support quickly and effectively and ongoing monitoring can be facilitated. Ideally, an early diagnosis should be made by a consultant paediatrician. The co-ordinating clinician might then be responsible for referral to the Education Welfare Service to ensure that education is minimally disrupted. School nurses, and for older children, their Connexions personal adviser, can play a pivotal role in linking agencies and supporting a child or young person.

LEAs must have arrangements to ensure that a pupil with medical needs who is away from school for any period has access to education. If a pupil is expected to be away from school for more than 15 working days, education, in whatever form, should begin immediately the pupil is absent from school. It is the total time of predicted absence from school that is important, not merely the hospital stay. Regular analysis of medical absences, by the school or Education Welfare Officer, can be used to develop regular monitoring of pupils with medical needs, including those referred to the home and hospital teaching service'.²

Child protection

Neither the fact of a child or young person having unexplained symptoms, nor the exercising of selective choice about treatment or education for such a child, constitutes evidence of abuse. Nonetheless, children with CFS/M.E. may suffer harm, and this is part of the differential diagnosis. **It is important to listen to the child, as well as to family members and parents/carers, to respect their experiences, and to give due weight to their views, especially the child's.** The young person should be given the opportunity to speak with the clinician, with or without their parents/carers.

In cases of CFS/M.E., evidence clearly suggestive of harm should be obtained before convening child protection procedures or initiating care proceedings in a family court. Social Services should be made aware that medical opinion in this area is divided, and consideration should be given to obtaining a further opinion from an expert medical practitioner with specialist knowledge of CFS/M.E. The Department of Health guidelines, *Working together to Safeguard Children*³, sets out the inter-agency arrangement to protect and safeguard children's welfare. This should be followed when there are concerns that a child may be, or is likely to, suffer significant harm.

Symptom control

Various treatments can be used to relieve disabling symptoms. Specific therapies can be chosen on the basis of advice (e.g. guidelines or reviews, and see adults section) and adapted to the individual. Drug therapies are best started at lower doses, with agents least likely to have adverse effects.^{4,5} Many products will not be licensed for such indications or age groups, so if doubt exists, specialist advice is needed. Ideally, all children should have access to dedicated pain and symptom control services, but these are patchy.

Follow-up

A balance between primary and specialist/hospital care is needed that is acceptable to patient and family, with the most appropriate clinician(s), locally where possible. During follow-up, the child needs to be reviewed for:

- Progress made towards specific patient-directed goals in various fields.
- Complications (physical, psychological, or social) that need management.
- Newly arising and alternative diagnoses that require treatment.

Transition to adult services

Any transition may require increased input from services – e.g. from childhood to adolescence to adulthood, starting or stopping school – and represents an opportunity, if not a requirement, to review the management plan with patient and parents/carers.

Impact on family/carers

Lack of blame needs to be specifically stated, and this key message can open doors to achieving necessary support for child and family. Other measures to minimise the impact of illness on carers and on family life include:

- Offering parents/carers opportunities to speak when the child is not present, though the child's consent and agreement for this must be obtained.
- Communication over and agreement on the illness and management plan.
- Prioritising areas at particular risk of long-term damage (e.g. social contact).
- Anticipation, recognition, and management of secondary difficulties affecting the child or other family members, including siblings.
- Identifying child and family strengths that can be developed despite the illness.



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Action for M.E. is a national charity working to improve the lives of people with M.E. We offer information and support to patients, carers, health and social care professionals. Please contact Action for M.E. at the above address for further information

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References and additional resources

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