Annexes to the

Report of the Working Group on CFS/ME

Report to the Chief Medical Officer
of an Independent Working Group

Annex 6: Management of CFS/ME – Report Summary

Annex 7: Management of CFS/ME – Children and Young Persons Summary

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Contents

Annex **6**

Management of CFS/ME – Report Summary

1. Preface

In this annex is presented a shortened version of the findings of the report, with particular reference to the work of clinicians who provide care for people with CFS/ME. Additional material is provided on case management.

Although the material is presented here in a form which is rather longer than might be useful in day-to-day clinical practice, this clinical material can be seen as a resource which can be summarised in various ways, to provide the basis for local guidance on the management of children, young people and adults with CFS/ME.

2. Introduction

CFS/ME is a genuine condition that imposes a substantial burden on patients, carers, and families.

Chronic fatigue syndrome (CFS)/myalgic encephalomyelitis (ME) is now generally accepted as being a discrete disorder or spectrum of disorders. This condition may also be known as postviral or postinfectious fatigue syndrome. Some believe that ME is separate from CFS, but little evidence currently exists to support this division. Concerns remain over the names given to the condition, but while awaiting review of the terminology, this clinical guide refers to the disorder as CFS/ME in line with the remit given to the Working Group.

This guidance provides information for clinicians to improve the care and management that people with CFS/ME, both adults and children, receive from the National Health Service (see also **Annex 7**). The advice is supported by the balance of published evidence (see **Annexes 1**, **2**, and **5**) where possible, and also takes account of the reported experiences of people with CFS/ME and their carers and broad clinical opinion.

3. Definition

CFS/ME is a quite common, very heterogeneous condition of adults and children that lacks specific disease markers but is clinically recognisable. The broader impact of the disease can be extensive even in its milder forms and may be under-recognised. The characterising features are overwhelming fatigue, related effects on both physical and cognitive functioning, and malaise, accompanied by a wide range

of other symptoms. The fatigue is evidently very different in character from everyday tiredness and is accompanied by a profound lack of energy; it is commonly described as like no other in type and severity.

Perhaps the prime indicator of the condition is that physical or mental exertion beyond the individual's tolerance typically prompts worsening of symptoms with a characteristic delayed impact, which may be felt later the same day, the next day, or even later. This is followed by a recovery period, which again may last for days or even weeks. In some instances, the patient can sustain a level of activity for some time, but a cumulative impact is seen, with a setback after several weeks or more. Common symptoms include:

Persistent/excessive tiredness or fatigue – Although physical 'fatigue' (or other words used by patients to convey their experience) is an essential symptom, its severity varies, and other symptoms may be equally or more conspicuous. Cognitive fatigue is almost invariably experienced. These forms of fatigue need to be explored by the clinician in the same way, as, for example, the varying characteristics and experience of pain.

Cognitive impairment – In addition to general cognitive fatigue, other more specific difficulties in cognitive functioning occur, including: reduced attention span, reported impairment of short-term memory, word-finding difficulties, inability to plan or organise thoughts, spatial disorientation, and loss of powers of concentration.

Postexertional malaise – The malaise that follows exertion may comprise 'flu-like symptoms or other constitutional features.

Pain – This is typically persistent and often difficult to alleviate with standard analgesics. The pain may be muscular pain, joint pain, neuropathic pain (with or without parasthesiae), head pain and/or headache (often migrainous in type).

Sleep disturbance – This may take several forms such as early morning wakening, insomnia, hypersomnia, unrefreshing sleep, and disturbed sleep/wake cycle.

Other symptoms related to neuroendocrine function – Patients may experience symptoms apparently related to the neurological and/or endocrine systems, including: temperature disturbance; dizziness, including vertigo, rotational dizziness, postural hypotension, and dizziness on standing; and increased sensitivity to sensory stimuli, such as light and sound. Those severely affected may have more substantial neurological impairments, including unwanted muscle activity and feeding difficulties.

Recurrent sore throat – with or without lymphadenopathy.

Digestive disturbances – These include: nausea, loss of appetite, indigestion, excessive wind, bloating, abdominal cramps, and alternating diarrhoea and constipation. The features are similar to those of irritable bowel syndrome and may be exacerbated by certain foods (e.g. wheat, dairy products).

Intolerances – Intolerance and sensitivity/altered tolerance are common in CFS/ME. Alcohol intolerance is very common; many patients also experience

intolerance of some foods, some medications (especially psychotropic medication), and other substances (sometimes described as "multiple chemical sensitivity").

In addition to symptoms that occur in the majority, individual symptom complexes seem to vary according to the individual's medical history and activity pattern. In some individuals, recurrence of symptoms from the triggering event is part of the symptom profile (e.g. recurrent sore throats and lymphadenopathy after glandular fever, or vertigo after labyrinthitis). In others, old symptoms or susceptibilities relapse or recur with development of CFS/ME (e.g. pain from old injuries, headaches or migraines in predisposed individuals, mood disturbance in patients with previous anxiety or depression).

Symptoms can reflect the predominant activity, whether they are prompted by the activity or highlighted as a result of effects on the activity (e.g. muscle pain in the physically active, concentration impairment in those who rely extensively on cognitive performance). Symptoms such as postural hypotension and dizziness can in part reflect the secondary effects of inactivity and/or isolation resulting from enforced inactivity. Over and above these patterns, some patients seem to have a dominant locus of symptoms (e.g. 'flu-like malaise, neuromuscular symptoms, cognitive impairment, or gastrointestinal disturbance). In some women, premenstrual or menstrual exacerbation of symptoms is reported as is use of hormone replacement in those patients with low levels of serum oestradiol. Ultimately, an individual's symptom profile will be modified by the impact of illness on the affected person and those around them.

4. Aetiology and pathogenesis

The aetiology of CFS/ME is not fully understood, but some predisposing, triggering, and maintaining factors have been identified, with the consensus being that the condition is caused by multiple factors that interact in an individual. CFS/ME is slightly more common in women than in men. Triggers may include certain infections, such as glandular fever, viral meningitis, and viral hepatitis. Maintaining factors include sleep difficulties, which may involve excessive sleep or insomnia, and mood disorders, which commonly coexist with CFS/ME. Although inactivity is a consequence of CFS/ME, it seems that it can sometimes also maintain the disorder through physical deconditioning, balance and temperature control problems, loss of confidence, and sleep difficulties. Certain strongly held beliefs about the illness may also act to delay recovery in a minority of patients.

Down-regulated hypothalamic-pituitary-adrenal axis activity, abnormal autonomic nervous system function, immune dysfunction, and subtle signs of encephalopathy are among the reported abnormalities in CFS/ME, but their roles in pathogenesis are not yet established.

5. Approach to management

A management plan, including treatment strategies, that is mutually agreed between clinician, patient, and carers when appropriate, is a key component of the approach. Such management plans are ideally patient-centred and utilise appropriately trained professionals in multidisciplinary teams. Establishing a good

doctor-patient relationship is a priority, as in any condition. Clinical wisdom would suggest that this is enhanced by good two-way communication and establishing a mutual understanding of what is wrong and why.

GPs should usually be able to manage patients with CFS/ME, as with many other chronic conditions. Only when individual clinicians feel that their patient requires access to particular clinical or therapeutic skills that the clinician does not possess, is referral likely to be helpful. Availability of expertise and application of generic clinical skills and understanding are more important than specialty of the referral.

Severe manifestations of the illness, including being bed-bound or house-bound, are as much of a problem in this condition as they are in many other chronic conditions. People who are severely affected often become 'invisible' to those responsible for delivery of services, be they healthcare, education, or other services. Clinicians, service providers, and others responsible for delivery of care should ensure that appropriate provision is made for those who are unable to attend centres where services are provided.

A key component to the successful management of any long-term illness is involvement of the patient as a partner in care. Taking patients' views into consideration leads to higher satisfaction, better compliance with treatment, and greater continuity of care. A partnership approach acknowledges that the patient must continue to cope with their illness throughout, rather than simply during intermittent clinical interventions. Education on the illness and on self management can empower patients to become experts and take an active role in their care. Information and support are likely to be needed from early on in the illness. Several registered charities/voluntary organisations offer information, training, education, and support to patients with CFS/ME and their carers.

6. Evaluation and diagnosis

Making a diagnosis of CFS/ME can be seen as the first step in actively managing the condition. Evaluation and the process of diagnosis are very important steps in the acceptance and acknowledgement of CFS/ME by both clinicians and people with the illness. A positive diagnosis of CFS/ME is important, rather than a diagnosis by exclusion. Because there is no diagnostic test to confirm the presence of the condition, initial evaluation should allow enough time to gather sufficient information to make a diagnosis of CFS/ME.

Currently there are no universally agreed diagnostic criteria for CFS/ME. Those often used are research criteria, and are often considered too narrow for clinical practice. They also tend to require an arbitrary duration of 6 months before a diagnosis of CFS/ME is made. Hence, a diagnosis of CFS/ME, as with several other chronic diseases of uncertain aetiology, should rely on the presence of a set of characteristic symptoms together with the exclusion of alternative diagnoses.

Clinical assessment

The intended purpose of initial clinical assessment is: to increase the probability of a correct diagnosis of CFS/ME; to rule out other conditions; to confirm the diagnosis; to identify any clinical subgrouping relevant to the patient; and to

identify and characterise clinically significant consequences. Symptoms or signs not typical of CFS/ME should be considered separately. Separate consideration should also be given to new or subsequent symptoms, which were not part of the initial diagnostic pattern. The following are usually are involved:

Full clinical history – At present, the crucial diagnostic procedure for CFS/ME is the clinical history, including detailed drug history that encompasses complementary and recreational agents. Sufficient time should be allowed for patients to give a narrative account of their illness experience. Full assessment is needed of previous healthy functioning and the individual's current status, including the broader impact of the illness such as occupational or educational functioning.

Mental health evaluation – The patient may have concurrent mental health difficulties, particularly mood disorders, either as a result of CFS/ME, as an overlapping problem, or as a separate diagnosis. Assessment for mental health problems including mental state assessment and psychosocial assessment at an early stage is important, so that any such problems can either be dealt with or excluded, but the clinician must recognise that the patient may feel sensitive about this kind of assessment.

Sleep evaluation – Sleep problems are common in patients with CFS/ME, and can exacerbate other symptoms. Primary sleep disorders must be excluded in people with unexplained fatigue, especially if drowsiness or day-time somnolence are prominent. Common conditions include sleep apnoea, restless leg syndrome, periodic leg movement disorder, and delayed sleep phase syndrome. Secondary sleep problems may be due to mood disorders or pain.

Physical examination – The physical examination is essential, and it should be repeated as indicated by symptoms and clinical course. Findings are frequently normal in CFS/ME, but the examination may be helpful in excluding other conditions.

Basic screening tests – There is no validated diagnostic test for CFS/ME but several basic screening tests should be undertaken to exclude a wide range of conditions.

Basic screening tests

The tests include:

- full blood count
- C-reactive protein (CRP) concentration
- blood biochemistry tests including concentrations of creatinine, urea, electrolytes, calcium, phosphate, glucose, liver enzymes, and markers of thyroid function
- simple urine analysis

Other tests will be determined by the history or examination – e.g. rheumatoid factor and ANA with prominent joint or skin features and antibodies to gliadin and endomysium when coeliac disease is part of the differential diagnosis.

Specialised tests – These may be required within the differential diagnostic process, so as to exclude particular conditions that are suggested by specific types or mixtures of symptoms, or abnormal findings on physical examination or investigation. For example, in patients with suggestive symptoms, it is wise to carry out screening tests for a range of rheumatic diseases (including tests for rheumatoid factor and ANA [antinuclear antibodies]), because these disorders are in the differential list. By contrast, tests that are currently used in research, for example for specific immune markers or neuroimaging, are not necessary as part of routine clinical evaluation.

7. Differential diagnosis

This assessment will usually exclude conditions that are similar to CFS/ME, as well as identifying other coexisting illnesses. On occasions, other tests may be indicated.

Alternative diagnoses include:

- Addison's disease
- anaemia (haematological conditions and other causes)
- chronic infections (e.g. Lyme disease)
- chronic somatisation disorder
- coeliac disease
- immunodeficiency
- malignancy
- mood disorders, including both anxiety and depressive disorders
- multiple sclerosis
- myasthenia gravis
- primary sleep disorder (e.g. sleep apnoea, restless leg. syndrome)
- rheumatic diseases (e.g. Sjögren's disease)
- thyroid disease

8. Timescale

The diagnosis of CFS/ME should be based on the characteristic pattern of symptoms, once alternative diagnoses have been excluded. Given the complex nature of the assessment, several consultations may be necessary before the diagnosis is finally made. However, it is important that this in itself does not delay the process of diagnosis. A working or interim diagnosis is better than none and allows active management to begin. Delay may often add to the impact of the disease and may have important consequences not only for healthcare but for aspects of the patient's life such as employment and education. Criteria developed for research purposes suggest that symptoms should last for 6 months before a diagnosis of CFS/ME can be made. But this threshold is arbitrary, and the clinical diagnosis can and should be made well before this time, especially in children and

young adults. By 6 months, if symptoms persist, a provisional diagnosis usually should be confirmed. But the plan for managing the illness and its consequences should already have been put in place.

For most presentations in adults, a 6-week point from onset of abnormal fatigue is a more useful marker for considering CFS/ME as one of the differential diagnoses. Subsequent clinical presentation should then guide management decisions, rather than any external or arbitrary timescales about disease stages which vary widely between individuals.

9. Children and young adults

The timescale for the diagnostic process for children and young adults should be shorter because of the potential impact on development, education, and social functioning (see **Annex 7**). The factors that should alert clinicians to consider the possibility of CFS/ME also differ from those in adults. Missing school is an important sign. In particular, once a child has missed around 4 weeks of school, action should be taken to speed up the diagnostic process. An approximate 4-week duration of persistent fatigue is an appropriate time for management actively to begin for CFS/ME, if that proves to be the diagnosis.

Reported pain is common in children with CFS/ME – e.g. tummy ache, headache – and children are also more likely to experience loss of appetite or weight gain, abdominal pain, and nausea. Recognition of the impact on the family and provision of support is very important, for there may be consequences both for caring responsibilities and family dynamics, as well as the patient's well-being.

10. Treatment and care

Although treatments aimed at the underlying cause(s) are not possible (because the aetiology of the illness is not known), much can be done. Clinical wisdom suggests that management of limited energy and supervision of the balance between activity and rest (physical, mental, emotional, etc) are an essential part of ongoing care. Many maintaining factors and symptoms of the condition can be alleviated. Co-morbid conditions and symptom relief can be managed as they would be in other clinical situations.

The evidence for treatments that have an impact on CFS/ME overall is sparse. The Working Group reviewed and synthesised available evidence from research findings and patient reports with clinical opinion and likely resource implications. We identified three specific strategies as potentially beneficial in modifying the illness experience: graded exercise, cognitive behavioural therapy and pacing. Although opinions differ widely on the potential benefits and disadvantages of each of these approaches, all can be considered as potential management options, when applied with best practice and adapted to the circumstances of each individual patient.

The common aim behind all these treatments is sustainable improvement in functioning and adaptation to illness, via gradual steps that are mutually agreed and regularly reviewed by patient and clinician. As with any treatment, an explanation of the benefits and possible harms of the approach should be provided

before decisions are made to offer and accept treatment. It is important to recognise that people vary in response to these treatments and that it is appropriate to review therapy if symptoms appear to worsen as a result.

Insufficient evidence exists currently to recommend any other treatments for CFS/ME. Moreover, there have been insufficient studies of treatment to be able to make definite recommendations for treatment of children and young people or the most severely affected patients of any age. Open studies suggest that similar approaches can be beneficial, so long as the approach is adapted to the patient's level of disability.

11. Applying the research evidence in clinical practice

The clinician, in assessing the patient, needs to identify how best to utilise the available therapeutic strategies for their specific needs and circumstances. These should then be discussed and agreed with the patient. The advice must be informed by the research evidence, insofar as it relates to their circumstances; this should be shaped and adapted to the individual, their level of functioning, the areas of activity and adaptation where their needs are greatest, and the available resources.

For example, 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, ambulant patients with a lower level of functioning could use their normal daily tasks as the basis for a structured approach to graded increases in activity, where possible, using similar principles. This could be guided by a therapist or their doctor, depending how readily the patient can achieve the plan and how much regular guidance they need. More severely affected patients may need to focus on finding a way of sustaining some essential and valuable activities of daily living, until such time as they can carefully explore possible increases. Pacing, as applied by many patients and clinicians, is similar in approach.

Similar principles apply to the use of cognitive behavioural therapy, which is a tool for constructively modifying attitude and behaviour in such a way as to maximise the patient's ability to improve and to manage their condition more effectively. Some patients will have aspects that they find difficult, where a mutually agreed course of CBT delivered by a trained therapist focusing on these areas could help. Others may be able to adapt more readily without such closely supervised treatment, once they have been diagnosed and been given an overall management plan that is informed by the research evidence and adjusted to their own circumstances; regular follow-up can identify progress, problems and any additional needs.

In devising a management plan for the individual, the clinician and patient should assess and agree which components of therapy are appropriate, how they should be applied, and how supervised. During follow-up, the clinician should continually reassess the needs and strategies used, as these may change and evolve over time. This may be because of different levels of disability, setbacks, changing symptomatology, changes in co-morbid conditions, or problems relating to duration of illness. One important example is in patients who achieve some improvement; they may become more aware of their limitations and paradoxically may have more difficulty in adjustment than they did when they were more unwell.

The following sections outline some further principles in energy and activity management, as well as identifying some of the more important aspects of symptom control and treatment of some co-morbid conditions, which are based on generic evidence adapted to clinical experience in CFS/ME.

12. Energy/activity management

Clinicians can assist patients to manage limited energy but it is important that all concerned adopt a similar approach. Rest is reportedly beneficial in the early stages of the illness, but too much rest can be harmful. Similarly, overactivity can prompt setbacks ('boom and bust' cycles) so patients should aim to improve within limits that avoid too little or too much activity. Some adverse reactions to activity may be usual and expected (e.g. resulting from long-unused muscles) whereas others may indicate 'bust' or that a programme is not proving beneficial. Balance is the watch-word: balance between activity and rest/relaxation (physical, mental, and emotional); balance between physical and mental tasks; balance between work and play; and balance between needs and wants.

Two elements seem key to energy management, used both in a specific therapy programme, such as cognitive behavioural therapy, and as part of a generic multi-dimensional approach that also includes information provision, psychological support, and symptom control.

Setting daily activity levels – An individualised, positive, and consistent approach is important to establish a daily level of activity. Useful tools can be deployed, such as diaries, allowing the individual to relate activity levels to improved or worse symptoms. Once the condition is stabilised, and an assessment of baseline functioning is made, the amount of activity is then gradually increased and the amount of rest decreased as the patient becomes more confident. Goals are initially negotiated using baseline diaries; they typically involve a variety of specific tasks but include a mixture of social, work, and leisure activities. Short walks or tasks carried out in even chunks throughout the day are ideal and are interspersed with rests. The goals are gradually built up as tolerance to symptoms increases and until longer-term targets are reached. This usually takes several months. Fatigue levels may not decrease very much initially, so the emphasis should be on consistency and sustainability. Ideally, management of fluctuations in health, such as setbacks and plateaux, should be agreed in advance, or at least jointly recognised before a mutually agreed adjustment is made to the routine.

Establishment of a sleep routine – A sleep routine should be established as quickly as possible, although the pace of changes to the sleep routine will depend on the severity of the problem. Early on, patients can be asked to keep a diary of bedtime, sleep time, wake up time, the time they get up, and 'catnaps'. The total number of hours spent asleep is calculated. Going to bed and getting up at a pre-planned time, while simultaneously attempting to reduce or eliminate daytime sleeping as far as possible, may help to improve quality of sleep. However, daytime 'catnaps' are not proven to disrupt night-time sleep, and some individuals will need to have a 'siesta' or similar period of daytime rest.

13. Symptom control

Symptom control is important for patients with CFS/ME, not only to improve quality of life, but also to facilitate the patient's ability to engage in specific therapy programmes. Clinicians should access their usual 'toolkit' of symptomatic measures where appropriate, adapted to the patient's need – e.g. advice on and support for adaptations in behaviour or diet, or medication(s) to treat or to prevent symptom(s). The options, with their rationale, advantages, and possible side-effects, should be explained. It is often worth suggesting that such interventions be used as a therapeutic trial for a defined period (unless severe intolerance occurs), during which the impact of the approach to the particular patient can be evaluated. Ultimately, discussions can be held on whether or not to continue.

Specific therapies can be chosen based on advice from relevant guidelines or reviews. In some cases, that advice may need to be adapted to the clinical setting of CFS/ME and refined on the basis of the individual patient's previous experience. Since patients with CFS/ME are often relatively intolerant of medication, it is usually wise to start with lower doses and to make use of agents that are less likely to have adverse effects where possible. If the initial approach is not effective or tolerable, variations or alternatives can be attempted by agreement after suitable discussion. If intolerance to medication is a major difficulty, other strategies are worth exploring with the assistance of specialist therapists, as appropriate. Patients report that some complementary therapists may be helpful in this regard.

Difficulties with pain, sleep, and mood should be specifically elicited and treated, as each seem to compound the others. Specific measures for control of key symptoms include:

Pain – Pain 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, or the anticonvulsants carbemazepine, sodium valproate, and gabapentin, can be especially useful for pain with neuropathic quality (and the related paraesthesiae). Other approaches used in chronic pain management may be valuable, including the use of psychological techniques; referral to a specialist pain team may be valuable.

Muscle pain may be accompanied by twitching, fibrillation, muscle jumps, or cramps. Muscle relaxants (such as baclofen) may be helpful in reducing unwanted unco-ordinated spontaneous muscle activity, which can reduce pain and sometimes weakness.

Sleep – Once primary and secondary sleep disorders have been excluded, education on establishing a sleep routine should be the first approach before considering medication. If medication needs to be used, low doses of tricyclic agents, if tolerated, are often effective in restoring sleep quality and rhythm, and are preferable to hypnotic agents, because of the risk of dependency with the latter.

Mood disorders – Both children and adults with CFS/ME commonly suffer from depressive and anxiety (mood) disorders. Mood disorders can exacerbate CFS/ME directly (for example causing worse fatigue and cognitive difficulties) and indirectly (for example, through insomnia). Mood disorders significantly increase risk of

suicide and also lead to a worse prognosis; active management of these conditions can improve outcomes.

The presence of a comorbid mood disorder is a management "red flag", which should lead to appropriate treatment, or referral on to the appropriate professional (clinical psychologist or psychiatrist) in patients of all ages. Similarly, the presence of suicidal thoughts requires an urgent referral to a psychiatrist, on the same day, if indicated. Appropriate treatment of depressive and anxiety disorders improves outcomes and quality of life. Possible treatments include cognitive behavioural therapy and either antidepressant or anxiolytic medication. It is important to note that antidepressants have been found ineffective for CFS/ME per se. Little research is available on these issues in children.

The usual agents may be considered for treatment of mood disturbance, but it is important to take account of the particular sensitivity of CFS/ME 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/ME 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 agents in low dosage can be used as an adjunct for sleep disturbance and other symptoms. Tricyclic and related agents may also be useful as antidepressants, but care is needed both in type and dosing, to avoid intolerance 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.

Headache – Headaches sometimes have a migrainous character and agents used for migraine can be helpful. Where migraines are less frequent or severe, triptan drugs can reduce intrusiveness of such symptoms. 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/ME). 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 – Dizziness 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 discomfort – Patients with CFS/ME 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. In some patients, exclusion diets have proved helpful, notable those excluding wheat and/or dairy products; these may be worth a trial. Specialist referral may be appropriate.

14. Setbacks

Some patients suffer from set-backs or more substantial relapses, when they find their symptoms recur or increase in intensity, with consequent increased disability. Management should start with a review of possible triggers for the set-back, such as a stressful life event, an infection, or a particular period of over-exertion, which can usually be identified. It can also be helpful to identify those factors that predispose the patient to the set-back (e.g. inability to say "no"). This enables both the clinician (or therapist) and patient to understand the reasons for the set-back, thus reducing its impact, which may otherwise be enhanced by the uncertainty. The patient and clinician (or therapist) can then adapt the management plan to take into account the triggers, predisposing factors, and the current disability. Thus the set-back can improve the management plan with increased knowledge of those factors that predispose to and trigger a patient's illness.

15. Information and support

Information and support is vital to empower patients to act as partners in their care. Several registered charities and voluntary organisations provide valuable information, support, education, and other resources for patients and carers.

What clinicians can do:

- Listen to the patient, recognise and believe his or her individual experience.
- Acknowledge uncertainty and the impact that this has on the patient, family, and carers.
- Provide support and encouragement – e.g. during setbacks.
- Provide information on and discuss: the nature of the condition, approaches to self management, helpful therapies, and how to access other agencies and services.
- Agree a name for the condition.
- Give advice on symptomatic treatment.

16. Prognosis

As with many chronic conditions, the emphasis should be on improvement and adjustment rather than 'cure'. For patients with CFS/ME:

- Prognosis is extremely variable; Although many patients have a fluctuating course with some setbacks, most will improve to some degree. However, health and functioning rarely return completely to the individual's previous healthy levels.
- Of all the people in the community who fulfil criteria for CFS/ME, many experience the majority of their improvement relatively quickly.
- In those who do not recover relatively quickly, the illness has a tendency to become more prolonged and in a minority, the duration is very long.

- Prognosis is better with treatment, and in the long term, many patients who receive some form of appropriate treatment will substantially improve in both function and symptoms.

17. Ongoing care

Ongoing assessment is a key part in the management of this condition, and is particularly true when there appears to be little or no improvement despite treatment. In this clinical scenario, it is important to review the diagnosis and co-morbid conditions, then review any maintaining factors, including symptoms such as pain or sleep disturbance. Finally, evidence suggests that looking for barriers to recovery may be worthwhile.

Recovering from a chronic illness can bring on problems of its own, particularly in an illness that affects patients' confidence in their functioning. A patient may doubt whether he or she can cope with the stresses and responsibilities of normal life again. Clinical evidence would support a gradual and negotiated return to as normal a life as possible, if and when health improves sufficiently. This can include "stepping stones", such as part-time voluntary work, therapeutic earnings, and returning to paid work on a part-time basis.

Return to work or school may be difficult and may require the clinician to support the patient if employment or schooling difficulties arise during this phase.

Annex 7

Management of CFS/ME – children and young persons summary

1. Introduction

Children and young people (of school age) do get and can be profoundly affected by CFS/ME. This summary provides information for clinicians to improve the care that such patients receive from the National Health Service, supported by the balance of published evidence where available, with evidence from patients and clinical opinion.

The principles of care of children and young people are common to many chronic conditions. Children's rights are protected by UN Convention.[1] The 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.

2. Nature and impact

The disorder or spectrum of disorders known as CFS/ME is becoming more commonly diagnosed among school-age children, and even in children as young as 5 years. Available evidence suggests increased onset at secondary school age (around 14–15 years) and a prevalence of 0·07%, with one study attributing 51% of all long-term school absence in 333,000 young people to CFS/ME. Most studies suggest a better prognosis than for adults, and many children recover even after long illnesses.

Characteristic clinical features 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' may not be prominent, but usually physical and/or cognitive activity is limited, and school absence occurs. Increasing activity often delays recovery, although younger patients do not always have the typical delayed exacerbation of symptoms seen in older patients. Symptoms wax and wane over time and in severity, and are influenced by individual circumstances, previous functioning, and responses to illness.

As with other chronic illness, CFS/ME potentially threatens physical, emotional, and intellectual development, and can disrupt education, social, and family life. Social isolation and school absence cause particular difficulties, as do the wider professional and public uncertainties surrounding the illness.

3. Evaluation and diagnosis

An especially prompt and authoritative diagnosis is needed, while the possibility of other illnesses and complications must remain in mind. CFS/ME lacks disease markers, and specific diagnostic criteria for CFS/ME are poorly defined in children. So, the diagnostic process is the familiar one of assembling positive clues, while ruling out other conditions. Second opinions are needed if doubt exists.

When a child or young person has symptoms affecting school attendance for at least four weeks, active steps should be undertaken to identify the cause from a list that includes CFS/ME. Evaluation should include clinical history, physical examination, and basic screening tests (general haematological and biochemical blood tests, urinalysis, and screening for rheumatic diseases and coeliac disease as indicated; see **Annex 6**). Specific evaluation of sleep, mental health, activities, and education is also needed.

Other conditions that present with school absence are important differential diagnoses that need to be considered early and dismissed or treated, since they can also complicate CFS/ME. Possibilities include physical and mental illness, particularly depression, as well as school phobia, eating disorders, and, rarely, child abuse.

4. 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, including educational and family 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 of CFS/ME in primary care with specialist confirmation if appropriate.
- Evolution of the treatment plan, with management and follow-up agreed according to individual and family circumstances, and local expertise and specialist interest.
- Most children missing school cared for and managed in their homes, with follow-up in primary care or by a mutually agreed community or hospital specialist.
- 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 via GPs to provide additional support to patients (e.g. social worker during assesment, joint work or referral to Child and Adolescent Mental Health Services [CAMHS] in the context of a multidisciplinary approach, occupational therapy or physiotherapy services for general care and specific needs such as equipment, etc). Community nurses, paediatric nurse specialists and consultants, and school nurses and home-care teams may be particularly valuable to support and co-ordinate care and management.

5. Information and support

All clinicians can:

- Listen to the patient, recognise and believe his or her 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 assistance to keep a diary, manage limited energy and other resources, and balance the approach to activity. Desired outcomes and a possible timetable 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.

6. Education

Education is critical in the young person's management, as is play for the younger child. Those likely to have special needs, such as home tuition, need to be identified and referred early to the Education Welfare Service, preferably with involvement of a consultant paediatrician. Most young people can aim for attending their school for short periods, resting or working as needed (e.g. in a separate area), and maintaining some social contact, before gradual reintegration. Some may need home tuition and/or distance learning by means of information and communications technology, which can also improve social contact. Those too severely affected to study at all need encouragement to set a level of sustainable activity, with regular review.

7. 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/ME 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/ME, 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 a specialist knowledge of CFS/ME. Advice in *Working Together to Safeguard Children*[2] (Department of Health, Department for Education and Employment, and Home Office) should be followed when there are concerns that a child is at risk of significant harm.

8. 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 adapted to the individual. Drug therapies are best started at lower doses, with agents least likely to have adverse effects.[3,4] 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.

9. 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; and
- 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.

10. 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 child's 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.

References

1 Convention of the Rights of the Child: Second Report to the UN Committee on the Rights of the Child by the United Kingdom. Executive Summary. London: Social Care Group, Department of Health, 1999.

2 Working Together to Safeguard Children: a guide to inter-agency working to safeguard and promote the welfare of children. Department of Health, Home Office, Department for Education and Employment *London: The Stationery Office, 1999*.

3 Royal College of Paediatrics and Child Health. Medicines for children. London: RCPCH, ISBN 1-900954-39-9.

4 BMA and Royal Pharmaceutical Society of Great Britain. British National Formulary. London: BMJ Books, ISBN 0-85369-465-6