

GUIDELINES

Diagnosis and management of chronic fatigue syndrome or myalgic encephalomyelitis (or encephalopathy): summary of NICE guidance

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Why read this summary?

Data from other countries indicate that chronic fatigue syndrome (also known as myalgic encephalomyelitis or myalgic encephalopathy) (CFS/ME) is relatively common (affecting at least 0.2-0.4% of the population), although good epidemiological data for the United Kingdom are lacking. Many different potential aetiologies for CFS/ME have been investigated, including neurological, endocrine, immunological, genetic, psychiatric, and infectious, but the aetiology cannot yet be fully explained. CFS/ME can cause prolonged illness and disability and substantially affect patients and their families. Although most patients have mild or moderate symptoms, some have severe CFS/ME and are housebound or even unable to move from their bed. Uncertainties about diagnosis and management may exacerbate the impact of symptoms, and patients often encounter delays in diagnosis and difficulty accessing information, support, and potentially helpful therapies.¹ This article summarises the most recent guidance from the National Institute for Health and Clinical Excellence (NICE) on diagnosing and managing this condition.²

Recommendations

NICE recommendations are based on systematic reviews of best available evidence. When minimal evidence is available (as with CFS/ME), a range of consensus techniques is used to develop recommendations. In this summary, recommendations derived primarily from consensus techniques are indicated with an asterisk (*).

General principles

- Acknowledge the reality and impact of the condition and symptoms.*
- Provide information on possible causes of the condition, its course, and management strategies (including benefits and risks of therapies, return to work or education), tailored to individual circumstances.*
- Offer information about self help and support groups for patients, families, and carers.*
- Be aware that all patients have the right to refuse or

withdraw from any component of their care plan without this affecting other aspects of care or future choices about care.*

- Aim to establish a supportive, collaborative relationship with patients and their carers and families (especially those of children, young people, and people with severe CFS/ME).*

Presentation

- Consider the possibility of CFS/ME if a person has fatigue with all of the following features:
 - New or specific onset (that is, it is not lifelong)
 - Persistent and/or recurrent
 - Unexplained by other conditions
 - Resulted in a substantial reduction in activity level
 - Characterised by post-exertional malaise and/or fatigue (typically delayed, for example by at least 24 hours, with slow recovery over several days)
- together with one or more of the following symptoms:
 - Difficulty with sleeping, such as insomnia, hypersomnia, unrefreshing sleep, a disturbed sleep-wake cycle
 - Muscle and/or joint pain that is multisite and without evidence of inflammation
 - Headaches
 - Cognitive dysfunction (such as difficulty thinking; inability to concentrate; impairment of short term memory; and difficulties with word finding, planning or organising thoughts, and processing information)
 - General malaise or influenza-like symptoms
 - Sore throat
 - Painful lymph nodes without pathological enlargement
 - Dizziness and/or nausea
 - Palpitations in the absence of identified cardiac pathology
 - Worsening of symptoms upon physical or mental exertion.
- “Red flag” symptoms that might indicate another serious illness include significant weight loss; clinically significant lymphadenopathy;

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localising or focal neurological signs; features of inflammatory arthritis, connective tissue disease, or cardiorespiratory disease; and sleep apnoea.*

- Do not delay advice on symptom management, which should be provided as in usual clinical practice even before establishing a diagnosis of CFS/ME.*

Diagnosis

- CFS/ME is diagnosed clinically as insufficient evidence exists for routinely using diagnostic tests, such as the head-up tilt test, auditory brainstem responses, and electrodermal conductivity.³⁻⁵
- Usual testing to exclude other diagnoses should include:
 - Urine analysis for protein, blood, and glucose
 - Full blood count and erythrocyte sedimentation rate or plasma viscosity
 - C reactive protein
 - Serum urea, creatinine, electrolytes, and calcium
 - Random blood glucose
 - Creatine kinase
 - Liver function
 - Thyroid function
 - Screening blood tests for gluten sensitivity
 - Serum ferritin (children and young people only).*
- Do not do serological testing for viral or bacterial infections unless the history indicates such an infection.
- CFS/ME should be diagnosed in an adult after symptoms have persisted for four months and after exclusion of other likely causes of the symptoms. In a child, the condition should be diagnosed (or the diagnosis confirmed) by a paediatrician after symptoms have persisted for three months and after exclusion of other likely causes.*

General management and referral

- After diagnosis, manage symptoms as in usual clinical practice, which may include drugs and dietary changes. Other interventions that may improve function and quality of life include sleep management (for example, identifying common changes in sleep patterns seen in CFS/ME), appropriate use of rest periods, relaxation, pacing,* and further dietary changes,* as needed.
- During a setback (or relapse) with increased symptoms, advise patients to maintain physical activity if possible. If not possible, aim for a gradual return to previous exercise and functional routines.*
- Pharmacological treatments (such as antidepressants, steroids, thyroxine in euthyroid patients) are not recommended as evidence for their overall benefit is equivocal.*
- Advise patients to maintain a well balanced diet. Dietary supplements (including vitamins and minerals) and complementary therapies are not recommended as there is insufficient evidence of benefit. However, patients may wish to try these

therapies for symptom control as part of a self management strategy.*

- Advise on fitness for work and education and recommend flexible adjustments or adaptations to work or studies for return to these when the patient is ready and fit enough.* With the patient's consent, liaise with employers, education providers, and support services such as occupational health services, disability services through Jobcentre Plus (www.jobcentreplus.gov.uk/), schools, home education services, and local education authorities, and/or disability advisers in universities and colleges.*
- Consider referral to a specialist on the basis of the person's needs and symptoms: offer referral within six months of presentation to those with mild symptoms, within three to four months to those with moderate symptoms, and immediately to those with severe symptoms.*

Specialist care

- Collaborate with the patient on an individualised programme, aiming to sustain or gradually extend the patient's physical, emotional, and cognitive capacity, and to manage the physical and emotional impact of symptoms on the individual and his or her carers.
- Offer cognitive behaviour therapy and/or graded exercise therapy to people with mild or moderate CFS/ME and provide these therapies to those who choose them, as these interventions show clearest evidence of benefit. These interventions should only be delivered by appropriately trained professionals with experience in CFS/ME, and with appropriate clinical supervision.
- Cognitive behaviour therapy should follow the usual principles and include asking the patient to self monitor activity, rest, thoughts, feelings, and behaviours; discussing with the patient his or her adjustment to the diagnosis; and encouraging acceptance of current functional limitations.
- Graded exercise therapy should include the establishment of a baseline followed by planned increases in duration of low intensity physical activity, followed by gradual increases in intensity leading to aerobic exercise (which increases the pulse rate). It should be based on current level of activities and daily routines and on the patient's own goals. Both patient and healthcare professional should recognise that it may take weeks to years to achieve these goals.
- Diagnosis, investigation, management, and monitoring for people with severe CFS/ME should be supervised or supported by a specialist in the condition. This may include providing domiciliary services or using methods such as telephone or email as appropriate.

Overcoming barriers

- A lack of services (and resources)¹ and misperceptions about the nature of CFS/ME¹ may

hinder implementation of these recommendations. Recognising the condition and the impact it can have on the lives of patients and their families is therefore a necessary preliminary to improving care.

- People with severe CFS/ME who are housebound or need prolonged bed rest require specialised care, but evidence is lacking about which treatments are most effective to reduce their symptoms.
- Little research exists on CFS/ME generally, and specifically on its causes and diagnosis.
- The guideline development group recognised the need for wider research on aetiology and pathogenesis, as a basis for developing new treatments.
- For all people with CFS/ME the guidelines recommend an integrated, multidisciplinary approach, incorporating health and social care, with support for education and work when and if the patient is ready. This comprehensive approach can be difficult to achieve unless one named professional has responsibility for coordinating the patient's care.

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- 2 National Institute for Health and Clinical Excellence. *Chronic fatigue syndrome/myalgic encephalomyelitis (or encephalopathy): diagnosis and management of CFS/ME in adults and children*. London: NICE, 2007. <http://guidance.nice.org.uk/CG053>
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A PATIENT'S JOURNEY

Ehlers-Danlos syndrome

Frances Gawthrop,¹ Rae Mould,² Amanda Sperritt,³ Fiona Neale⁴

These three case histories illustrate the many problems facing patients with Ehlers-Danlos syndrome in its various forms

Patient 1: Jacqueline Mould

At birth my daughter, Jacqueline, had very visible veins on her head and body. She bruised badly when she fell. The doctors said it was nothing—just thin skin. Then, when she was 12 years old she developed an unusual localised rash on her knees. A dermatologist said she had perforating elastoma. She was an “interesting case,” but he did not know the cause. At age 23 Jacqueline developed pain in her legs. The doctor said it was just varicose veins—there was nothing to be done except to avoid standing and to wear support stockings. The pain got worse and Jacqueline went for a hospital consultation. At the clinic blood was taken and she bled for 13 minutes. She was sent to a joint consultation with a haematologist and a dermatologist, who said she should see a specialist interested in the genetics of Ehlers-Danlos syndrome. Still we had no idea of what was going on.

We waited two years to see the geneticist. He commented on her facial features and tested her joints, which were not hypermobile. He told us that Jacqueline had the vascular type of the syndrome. No one in the family was like her—her condition was caused by a mutation. She was told that she should not have children, must not take part in contact sport, and must avoid physical

stress. We were invited to ask questions, but it was all too much of a shock. We went home in silence. Jacqueline was particularly distressed about not being able to have children.

When I read the literature of the Ehlers-Danlos Syndrome Support Group I realised the full horror of probable sudden death. I felt numb. Jacqueline was a fit young woman in full time employment who was enjoying her life. I left the literature for her to read if she wished. She decided not to tell her friends about her illness. She had episodes of depression, but together we decided to carry on as normal.

Two years later, at work, Jacqueline's legs suddenly went numb. She was taken to the local hospital. The doctors didn't know about Ehlers-Danlos syndrome. They found nothing wrong and said she could go home. At this point Jacqueline collapsed. After resuscitation it was realised that she was bleeding internally. A renal artery had ruptured and despite surgery Jacqueline died on the operating table.

Today I work with the Ehlers-Danlos Syndrome Support Group to improve awareness of this rare condition and to help support those who have to face its problems.

Patient 2: Amanda Sperritt

I am 36 years old and have hypermobile Ehlers-Danlos syndrome. As a child I was “double jointed”—able to do party tricks that made people say “yuk.” When I was 15

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