A Report of the

CFS/ME Working Group

Report to the Chief Medical Officer
of an Independent Working Group

January 2002

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Foreword

In 1998 the Working Group on CFS/ME set out to consider how the NHS might best provide care for people of all ages who have this complex illness.

We have approached the work in two stages. First, to listen to, and to try to understand, the patient and carer perspective on the very wide range of issues surrounding the management of the illness. Unsurprisingly, when so little is known about the cause of CFS/ME, there are concerns from patients and their representatives, and from a broad range of clinicians, over the way the illness is managed. These concerns on management apply to the NHS, to other government Departments and to the private sector. In particular, patients and health professionals involved in the care of CFS/ME find much disbelief about the nature of the illness and of its impact. Perhaps as a result, in many areas of the country there appears to be a lack of appropriate health care facilities.

In the second stage of our work, in which contributors from the Working Group were assisted by a Reference Group and many other commentators, we sought to bring together knowledge on CFS/ME to support initiatives to improve care for patients. This has been an intricate process, drawing on research evidence, the experience of patients and diverse clinical opinion. We were assisted in this endeavour by a systematic review of the evidence which was commissioned by the Department of Health. Although it was not always possible to resolve some of the differences of view, for perspectives and opinions on some issues ranged widely across the contributors in the Working Group, we have been able to bring together a Report which meets our brief of advising on ways of improving care for children, young people and adults with CFS/ME.

This is an illness that most clinicians will encounter, although to varying extents. We found that it can, and should, be approached and managed clinically like any other chronic illness. That is, it can be managed by drawing on evidence and knowledge of what works best for a particular group of patients, and by using the usual generic clinical skills for assessment, therapy and care, adapted to the particulars of the condition and of the individual patient.

The production of this report has been a complex and challenging task and considerable effort has been required to bring this Report together. Many people have given their time to this enterprise and I wish to record my thanks to all of them. I see this Report as one unique step in a process of improving care for people with CFS/ME. There is much to do. But I hope this Report may act as a means of assisting health care professionals and public and private services to begin that process.

ALLEN HUTCHINSON
Chairman of the Working Group

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Chapter 1

Introduction

Chronic fatigue syndrome (CFS/ME) is a genuine illness and imposes a substantial burden on the health of the UK population. Improvement of health and social care for people affected by the condition is an urgent challenge. This report proposes ways in which clinicians and the NHS might respond to that challenge. Widespread uncertainty surrounds this condition, so the Working Group does not claim to present a definitive answer. Rather, we have aimed to bridge gaps in understanding of CFS/ME and between concerned parties – by highlighting areas where general agreement exists or by delineating controversy where possible – and to detail the many positive suggestions and models we have encountered. Where research evidence exists we have been guided by it.

During the course of preparing the report, the Working Group has continued to be concerned at the widespread controversy surrounding the existence and nature of CFS/ME. Patients, their carers, and healthcare professionals encounter different levels and varying manifestations of disbelief and prejudice against people affected by the condition. The disbelief and controversy over CFS/ME that exists within the professions has done nothing to dispel public disbelief in the existence of such a seemingly varied and inconstant illness.

Our aim has been to hear, understand, and address the disbelief and controversy where possible. During this process, the Working Group agreed that existing controversy cannot and should not be used as an excuse for inaction or unsuitable practice. Instead, we adopted a view that is considered appropriate for many other conditions: that every patient's experience is unique and their illness must be considered and treated flexibly in its own right.

1.1 Background to the report

On the 16 July 1998, at a scientific briefing to the press at the Royal College of Physicians, the then Chief Medical Officer Sir Kenneth Calman said:

> "I recognise chronic fatigue syndrome is a real entity. It is distressing, debilitating, and affects a very large number of people. It poses a significant challenge to the medical profession."

At the briefing, he announced establishment of a Working Group on CFS/ME.

On the 4 November 1998, Sir Kenneth's successor Professor Liam Donaldson announced the membership of the CFS/ME Working Group. Professor Donaldson stated in his press release:

> "This initiative will provide us with a real opportunity to review the practical care and support for patients, carers, and healthcare professionals alike."

After an initial exploratory period in which the principal aspects of the brief were examined, evidence and opinions sought from many quarters, and a systematic search of the international evidence on CFS/ME commissioned, the brief of the Working Group's report was confirmed:

> "To review management and practice in the field of CFS/ME with the aim of providing best practice guidance for professionals, patients, and carers to improve the quality of care and treatment for people with CFS/ME, in particular to:
>
> - develop good clinical practice guidance on the healthcare management of CFS/ME for NHS professionals, using best available evidence;
> - make recommendations for further research into the care and treatment of people with CFS/ME;
> - identify areas which might require further work and make recommendations to CMO."

1.2 Policy context

The first Task-Force Report provided a starting point for subsequent deliberations and assessments. A report of a Joint Working Group between the Royal College of Physicians, the Royal College of General Practitioners, and the Royal College of Psychiatrists, *Chronic Fatigue Syndrome* (published in 1996), set out to assess the field and provided a starting point to inform medical opinion. In addition there have been two further useful reports by the National Task Force on Chronic Fatigue Syndrome, Post Viral Fatigue Syndrome, and Myalgic Encephalomyelitis, an initiative of the registered charity Westcare covering NHS services, and children and young people.

However, none of these reports was accepted by all and, indeed, some aspects were perceived by some patients, their carers, and voluntary organisations to be potentially harmful. The aim of this Working Group has been quite different from previous work. The focus of our report is to provide advice on clinical management that reflects the importance of individualised, holistic care, and tailoring approaches based on the best possible evidence to reflect particular needs of the patient and their carers.

The NHS Plan published in July 2000 sets out new arrangements for public and patients' involvement. Improving the experience of each patient is at the centre of this programme, and the plan sets out new mechanisms to increase the influence of the public and patients over ways in which the NHS is run. One of the most

fundamental aspects of *The NHS Plan* is that the system will be redesigned to meet the needs of patients in the 21st century.

New mechanisms are being put in place to support patients, including a Patient Advice and Liaison Service and an increased emphasis on the role of patients as experts. Key changes anticipated include professional education on and support of self management, improved information to and communication with patients, and greater choice of healthcare options to support the concept of individualised care. *A First Class Service*, published in September 1998, presents arrangements for setting quality standards in the NHS. The National Institute for Clinical Excellence promotes clinical efficacy and cost-effectiveness through its clinical guidelines and Appraisal of Technology Programme.

1.3 Working Group process

The Working Group was established by the CMO with three constituent groups, to reflect the impact of the illness on all age groups and to draw on the wide experience of patients, carers, and healthcare professionals (**Appendix I**). These three groups – the Key Group, the Children's and Young People's Group, and the Reference Group – had differing but complementary roles.

The Key Group was responsible for surveying the evidence, developing the main report, and agreeing the final recommendations to the CMO. It was supported by the Children's and Young People's Group, who focused on issues pertinent to this age group. In undertaking this task, the group worked closely and in parallel with the Key Group. The Reference Group had an ad-hoc advisory/consultative role, and members had a wide range of expertise and opinion. A wider clinical network including practitioners outside of these groups was also consulted. A mix of generalists and various specialists with different backgrounds and experiences were consulted to ensure that the report would be informative and useful from diverse perspectives. We are grateful to all colleagues who provided the challenge to sustain the impetus for this work.

A small subgroup of the Working Group was established to produce a paper on CFS/ME and the benefits system. This working paper was then submitted to the CMO in April 2000. Professor Donaldson formally copied the paper to the Chief Medical Advisor of the Department of Social Security to inform that Department's Working Group, which was established to review the benefits system for people with chronic illness.

1.3.1 Methodology and sources of evidence

Initially, the work of the Key and Children's Groups was mainly exploratory, seeking to define the extent of the illnesses' impact and approaches to care and treatment from both the patients' and the health-service's perspective. Various types of evidence were used to inform the decisions of the Working Group, and to support exploration of the issues and the derivation of the guidance and recommendations in this report. Overall, sufficient research evidence was lacking, and in many areas the quality of research was not optimal. Therefore, we have used good-quality research evidence where possible to support commentary on the nature and extent of the range of illness presented in CFS/ME and on the effectiveness of possible therapies.

In the case of advice on therapeutic interventions, much of the evidence base was drawn from a systematic review of management strategies, commissioned by the Policy Research Programme Division of the Department of Health, and undertaken by the NHS Centre for Reviews and Dissemination at the University of York (see **Annex 5**). This systematic review examined the evidence for the effectiveness of all available interventions for CFS/ME among adults and children. However, the Group did not adopt the conclusions of this review in isolation. Instead, we aimed to synthesise three lines of evidence in a "trident" approach: research findings, the qualitative experience of patients, and broader clinical opinion. The summary for clinicians (**Annex 6**) and the clinical summary for children and young people (**Annex 7**) were developed as brief management tools to improve management and service provision for patients with CFS/ME in the NHS.

1.3.2 Collecting patients' experience

Harnessing the views of patients, parents, families, and carers to underpin the guidance was afforded high priority by the Working Group (**Annex 3**). Two 'Sounding Board' events were designed to capture some patient voices and to ensure that major stakeholders could identify pertinent issues. These events, together with analysis of supporting material and surveys sponsored by Action for ME, the ME Association, the 25% ME Group, the Association of Young People with ME, and the TYMES Trust, have informed the report throughout (see **Annex 3**). Above all, we have sought to avoid bias in drawing together a picture of the impact of CFS/ME. The Group aimed to capture views from individuals with special interests or expertise and from a wider constituency, then to structure this material to reflect the range of views without giving undue weight to those restricted to certain individuals or groups.

The Working Group was particularly conscious of the need to reflect the views of individuals severely affected by the condition, who tend to be overlooked and under-represented in research, service development, and policy. We are particularly grateful to: the 25% ME Group, which represents patients who remain severely affected; the 12 severely affected participants at the Sounding Board events; two members of the Key Group; and others for sharing their experiences.

1.3.3 Development of the report

This report represents the Working Group's efforts to review, synthesise, and distil the wide range of available evidence and opinion, including qualitative patient evidence and clinical opinion. Throughout, we have aimed where possible to base our commentary and recommendations on the best quality evidence, and from a range that includes randomised, controlled trials and clinical anecdote. In the absence of research evidence to inform many issues, the bulk of the report is derived from a synthesis of patients' and clinical experience. Where some data exist, albeit incomplete and not fully agreed, we used the trident approach, together with the likely resource implications to inform our conclusions.

An original draft was developed from extracts written by members of the Key Group, according to their expertise. The report has subsequently undergone a repeated process of review by the Working Group and has been informed by the Reference Group and wider clinical network. The drafting process has been directed by the Editorial Team and undertaken by independent medical editorial staff.

1.3.4 Limitations of the process

In many cases, the Working Group was limited by the lack of available good-quality research. In particular, little is known about individuals who make a full recovery from this condition, and we acknowledge that more should be done to capture the experience of such patients.

Although the Sounding Board events were wide-ranging, we acknowledge that these views represent only some patients' voices, and we did not adequately capture the views of black and other minority groups affected by CFS/ME. The Working Group acknowledged that initial work commissioned by Action for ME from the SAMEC Trust has highlighted the difficulties faced by disadvantaged groups. This is an area where more research is needed to ensure that these patients are able to access appropriate health and social care.

1.4 Clinical context

To review management and practice of any clinical condition, certain questions must first be answered: What is the condition under review? How many individuals are affected? What is the impact and outlook of the condition? Thus, an early step in our process was to review available evidence on definitions and terminology, epidemiology, and prognosis (see **Annexes 1–3**).

1.4.1 Definitions and terminology

The CMO assembled the Working Group to report on CFS/ME. Many correspondents with the Group noted that the term CFS/ME covered subgroups of patients who might have different aetiology, symptom complexes, or response to various treatments. The members of the Key Group, the Children's Group, and the Reference Group also reflected this perspective to varying degrees.

Many terms have been used to encompass this condition or clinical entities within this disease spectrum. Currently, CFS and ME are classified as distinct illnesses in the World Health Organisation's *International Classification of Diseases*. In recent years, CFS has been the preferred medical term for this disorder, or group of disorders, although the large majority of patients' support organisations use the term ME. The term ME has been applied to the syndrome – or, according to some interpretations, a subset of it – and is widely used in the community. The Working Group is conscious that some patients, especially those who are severely affected, consider the use of the name CFS to be unrepresentative of their illness experience.

The Working Group decided that the most important requirement concerning terminology is the need for patients and clinicians to agree a satisfactory term as a means of communication. A consensus on definitions and terminology is urgently needed. Although a resolution is beyond the scope of this report, discussions are underway in the USA with international input, which will, we hope, propose a long-term solution acceptable to all parties. While awaiting such a solution, the Working Group suggests that the composite term CFS/ME is used as an umbrella term and considered as one condition or a spectrum of disease for the purposes of this report. This approach is consistent with our original terms of reference and ensures as far as possible an inclusive approach to our review.

1.4.2 Epidemiology

CFS/ME affects many people and their families in the UK and elsewhere in the world. Information about actual numbers of people with the condition is very hard to come by because of the difficulty in defining the illness precisely. Use of restrictive case definitions may mean that such estimates are likely to under-represent the true scale of the disease, particularly the number of people with severe CFS/ME. Some useful studies have been done (see **Annex 1**), but despite some large initial sample sizes, the numbers of cases identified were relatively small and the estimates statistically imprecise, so not too much reliance can be placed on the findings.

The lack of epidemiological data means that many estimates of incidence and prevalence are based on extrapolation of results from other populations. There is, though, no evidence to suggest that similar rates of incidence and prevalence occur in different populations or ethnic groups. Thus, such extrapolations could be unreliable. The likelihood is that natural variation does exist between populations and geographical locations, and that results obtained in one study cannot be extrapolated to another with any degree of accuracy. However, even this cannot be said with certainty.

Overall, evidence suggests:

- A population prevalence of at least 0.2% – 0.4%.
- The commonest age of onset is early twenties to mid-forties.
- In children, the commonest age of onset is 13–15, but cases can occur as young as five years old.
- CFS/ME is about twice as common in women as in men.
- It affects all social classes to a similar extent.
- It affects all ethnic groups.

Little more is known about the incidence and prevalence of CFS/ME. As a result, a key piece of information is missing – one that is needed in order to undertake health-needs assessment as a prelude to provision of an adequate network of services. This gap needs to be filled if the NHS and other agencies are to meet the needs of these patients in a comprehensive and equitable way.

It is clear that local community-based studies alone are not sufficient to answer the outstanding questions about incidence and prevalence. In the UK, the organisation of primary care services creates a unique opportunity to conduct prevalence studies on the national scale necessary to generate requisite data. Such studies could be usefully augmented by community-based studies to detect and quantify variations in prevalence between communities, and to validate predictions from national studies (see **Chapter 6**).

1.4.3 Prognosis

Although current research evidence on prognosis indicates that only a small minority of patients recover to previous levels of health and functioning, this finding must be tempered by the likelihood of selection bias in studies towards inclusion of those with poorer prognosis. The likelihood is that most patients will show some degree of improvement over time, especially with treatment. A substantial number of patients will pursue a fluctuating course with periods of relative remission and relapse, while a significant minority become severely and perhaps permanently disabled. Gradually progressive deterioration is unusual in CFS/ME and should always prompt a further detailed clinical review to ensure that there is no other explanation that has been missed.

However, progressive deterioration can occur in some patients with CFS/ME; the existence of such patients, many of whom are among the more severely affected, must be recognised. Many need special attention in the delivery of care and the provision of support.

As with epidemiological studies of the incidence and prevalence of CFS/ME, knowledge of the prognosis of the illness is limited both by a lack of agreement on definitions (with the consequence that different researchers may not be always studying the same condition) and by a shortage of good studies (see **Annex 2**).

However, researchers appear agreed on three points:

- 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; most of those who feel recovered stabilise at a lower level of functioning than that before their illness.
- Of all the people in the community who fulfil criteria for CFS/ME, many experience the majority of their improvement relatively quickly – thus, the distribution of duration of illness is uneven, with greater numbers having shorter than average duration of disease.
- In those that do not recover relatively quickly, the illness has a tendency to become more prolonged and in a minority, the duration is very long.

Overall, there is wide variation in the duration of illness, with some people recovering in less than two years, while others remain ill after several decades. Those who have been affected for several years seem less likely to recover; full recovery after symptoms persist more than five years is rare. Irrespective of the statistics, each individual requires the necessary assistance to maximise their chances of an early recovery and minimise the impact of the illness.

Chapter 2

Evidence from Patients

During the Working Group process, the strongest message received was that patients' voices are not being listened to and understood. This chapter aims to provide a brief summary of the views of people with CFS/ME and their carers. The views represented here are those of some patients that the Working Group felt captured the key issues arising from the body of patient evidence. These views have been gathered formally and informally then synthesised by the Working Group: formally, from major voluntary organisations and two Sounding Board conferences organised by the Working Group; and informally, through correspondence and conversation with patients and carers, and the experience of clinicians and other healthcare professionals. The Sounding Board events, in particular, also generated a host of positive suggestions for improvement of healthcare provision, which are detailed here, with solutions offered in **Chapter 4**. Issues of definition and terminology are covered in **Chapters 1** and **3** and **Annexes 1** and **4**. Details of the Sounding Board process and further information on the impact of CFS/ME are available in **Annex 3**.

This chapter is an attempt to reproduce the views as expressed by patients and their carers.

Key messages

- Patients' voices are not being listened to and understood.
- People affected by CFS/ME indicated improvements needed in three main areas:
  - Recognition, diagnosis, acceptance, and acknowledgement;
  - Healthcare service provision;
  - Care of groups with special circumstances.
- Patients reported the need for more healthcare professionals who know about and understand CFS/ME. Public awareness campaigns, professional education, and information for patients and carers are accorded high priority.
- Experiences of primary care are polarised. Positive experiences are characterised by: "willingness" of clinicians to treat the patient as an equal; supportive attitudes; belief in the patient's experiences; and early recognition and diagnosis.
- Experiences of further care are predominantly negative. Needs identified include access to specialists and respite-care services.
- Those severely affected by CFS/ME (up to 25% of patients) feel "severely overlooked" by services. They experience isolation, lack of understanding, and particular barriers to accessing all forms of care.

Key messages

- Children and young people are profoundly affected by public and professional uncertainties over the illness. Young people also suffer from impact on their families and from lack of support and expertise within the education system.
- Individuals with CFS/ME from disadvantaged class/ethnic groups face special difficulties, yet they are under-represented in research.
- Carers, particularly of young people, need more recognition, support, and respite.

2.1 Principles

2.1.1 Importance of the patient voice

> "My GP and others feel helpless because they don't know what to do. I was told: 'You'll probably hear about anything before I do'."

Acceptance is growing that patients, particularly those with chronic illnesses, have a wealth of experience about management of their condition. This is acknowledged by the Government in the White Paper *Saving Lives: Our Healthier Nation* published in July 1999, which announced the intention to help people with chronic disease maintain their health and improve their quality of life. The report *The Expert Patient – A new approach to chronic disease management for the 21st Century* (September 2001), sets out how the NHS will empower those living with chronic long term conditions to become key decision makers in their own care. (See also *The NHS Plan*, July 2000, and **Chapter 1**, **section 1.2**)

A central activity of the Working Group has been to listen to the voices of patients, carers, and clinicians when they have described the impact of the illness on patients and thus to try to characterise these impacts (see **Annex 3**).

2.1.2 General themes

> "Where to go for help? What is wrong? When will 'it' go?"

This quote from a Sounding Board participant encapsulates the bewildering number of issues that can arise throughout the course of CFS/ME. Patients, their carers, and representatives generally share concerns in three areas:

- Recognition, diagnosis, acceptance, and acknowledgement;
- Significant variation and shortcomings in healthcare service provision;
- Issues particular to groups with special circumstances.

Specific concerns shared by many were identified:

- Importance of early diagnosis;
- Need for early information and advice;
- Problems arising from disbelief and lack of understanding;
- Difficulties in accessing specialist opinions;

- Lack of professional education and training;
- Need for improved public awareness.

These concerns were backed up by many positive suggestions for improvements in every area, which are detailed in specific subsections.

2.1.3 Recognition, diagnosis, acknowledgement, and acceptance

Many issues surrounding the management of people with CFS/ME have their origins in knowledge (or lack of) and attitudes. Key issues are:

- Poor recognition of CFS/ME by professionals
- Difficulties that arise over diagnosis
- Lack of professional and public acceptance and acknowledgement

Respondents highlighted initial experience as a time when the responses of health-care services are particularly important. The majority of patients first present to their GPs with no preconceived ideas on diagnosis. They are experiencing a complex, difficult, and uncertain illness, with no "hard or fast rules" to follow that would alleviate symptoms or expedite recovery – "an unplanned journey with no clear map or directions". Problematic responses include a lack of belief in the illness and other difficulties with diagnosis, notably a "slowness" to recognise that symptoms might be CFS/ME.

The Working Group heard that national support groups and other voluntary organisations have an important role to increase understanding in the wider community. However, much needs to be done within the NHS. Many of those consulted are keen to see an appropriate media campaign to improve awareness.

2.2 Service provision

2.2.1 Patient information and public needs

Many members of the medical profession do not themselves know where to go to for advice on how to manage CFS/ME, and they are therefore unable to provide patients with information they require. They often suggest that information can be obtained from charities or voluntary organisations. Patients try to inform themselves because they perceive their GPs to be ill-informed; however, they then can be caricatured as "obsessive". Carers voice concern about the lack of supportive and helpful information available throughout the NHS. They also perceive a lack of communication between healthcare professionals, the impact of "prejudice", and an overall lack of understanding of the impact on the family.

Patient information and public needs – patients' suggestions

- Computer-based database of information on good practice for primary care staff
- Formal record of arrangements for care between GP and other healthcare professionals, such as patient-held records
- Information on CFS/ME available from NHS Direct
- Other online information sources for patients
- Credible and correct explanations to patient, backed up with information/ education/leaflets.

2.2.2 Professional education

> "GPs could do with more training in how to support people with incurable/untreatable illnesses and in dealing with their own feelings of powerlessness."

Participants felt that the widespread lack of understanding of the condition is not specific to clinicians but includes other healthcare and social care professionals. This lack of knowledge was identified by the majority of those consulted, together with a lack of communication and advice, especially in the early stages, on how to cope in general with long-term illness for families and sufferers.

Those consulted consider a crucial area to be addressed is raising of awareness within the NHS. In particular, healthcare professionals should be made aware of the significant physical, psychological, and social suffering and disability caused by the illness. The need to increase the number of clinicians who have a special interest in CFS/ME and who are up-to-date on research is perceived as essential to improving NHS services.

Professional education – patients' suggestions

- Clinicians with experience in different fields of management and up-to-date with new developments.
- CFS/ME included as clinical condition(s) in medical curricula.
- Accurate, representative, up-to-date information in textbooks.
- CFS/ME as a topic in medical practitioners' study sessions.
- NHS 'roadshows' to facilitate education, information exchange, and service development.

2.2.3 Primary care

> "My GP was brilliant. He said he didn't know how to cure me, but we would work together to make me better."

Patients have a range of experiences of general practice: some have supportive and helpful GPs, some report mixed experiences, and others have unhelpful or bad experiences. Where experiences of GPs are positive, the key factors are "willingness" to treat the patient as an equal, and recognition and belief of the patient's experiences. The most important aspects of good clinical services for patients in primary care are seen as a supportive attitude and early diagnosis. Some were

concerned that their illness is not taken seriously, that some GPs show indifference, or that they do not always understand the illness and its possible severity. Some participants suggested that GPs and the wider primary healthcare team need improved knowledge and skills to support people with chronic or incurable illness in general and with CFS/ME specifically.

Patients identified concerns about referral pathways and what they perceived as inappropriate referrals, such as those seemingly directed by the clinician's particular view of or belief about the illness not shared by the patient. A particular concern is if a referral to mental health services is made because the illness is being characterised as "all in the mind". There is evidence that some patients "fight" for referrals, and in general GPs are confused over where to refer patients, so diagnosis can be a "hit and miss" affair. This has led patients to pursue their own specialist referrals for diagnosis; though in that situation the expressed experiences suggest that ongoing advice and support are lacking.

Primary care – patients' suggestions

- More listening on both sides; recognition and belief of patients' experiences.
- Joint GP/specialist/patient dialogue.
- Information for GPs on diagnosis of CFS/ME and support of patients with chronic or incurable illness.
- Prompt information provided soon after onset of illness.
- Proactive management of symptoms by clinicians.
- Increased information on rehabilitative approaches, such as pacing and graded activity.
- Telephone and/or email follow-up consultations to supplement home visits.
- Management strategies targeted at the newly diagnosed and long-term severely affected.
- Acknowledgement of carers' needs.

2.2.4 Further care

Sadly, the overall experience of specialist and hospital services among participants was predominantly negative. In contrast, an example of a positive experience highlights the experience that could be gained:

> "This was positive: a diagnosis of CFS was made and I was advised how to manage my energy; my symptoms were not dismissed as psychological and I was seen within 3 months of referral."

Beyond primary care level, the issue that causes most concern is the lack of specialists and services. For some patients there is "suspicion" and lack of trust in respect to specialist advice. The value of consultants with specialist knowledge and expertise is acknowledged, but, for CFS/ME, they are perceived as few and geographically difficult to access. Some patients find themselves in geographical "black holes" that lack specialist provision.

Further care – patients' suggestions

- Access to consultants with interest/expertise in CFS/ME.
- Fast-track referral system to multidisciplinary centres.
- Provision for severely affected in inpatient and outpatient settings, with outreach services to avoid travelling problems.
- Development of specialist nursing roles and active involvement of other professionals such as occupational therapists and physiotherapists.
- Provision for respite care.

2.2.5 Complementary approaches

In general, from individual comments and surveys by charities, patients find that alternative practitioners are more understanding and have a gentler approach to the illness than clinicians; they treat the person as an individual, and encourage self healing.

> "Brilliant ... the practitioners listen, they're open minded."

Practitioners most commonly consulted were chiropractors, nutritionists, allergy specialists, homeopathy practitioners, reflexologists, and herbalists. Patients reported that some of these approaches were helpful to alleviate some symptoms.

> "If we cannot be cured at least we can be comfortable."

Some individuals raised concerns at costs and access. There is also concern that therapists need to be trained and regulated, and that no therapist (whether NHS, private, or voluntary) should promise a "wonder cure".

Complementary approaches – patients' suggestions

- Availability on the NHS, through primary care teams.
- Development of an evidence-based approach.
- Access to reliable information on which approaches are potentially helpful – e.g. acupuncture for pain relief.
- Regulation of practitioners.

2.3 Groups with special circumstances

Repeatedly, the Working Group has heard from those affected by CFS/ME of the importance of treating each patient as an individual. However, we are also cognisant of the fact that certain groups of individuals share particular circumstances and needs that should be especially highlighted. These groups are the severely affected, children and young people, patients of disadvantaged class or minority group, and carers of those affected. This section highlights additional issues and positive suggestions for improvement.

2.3.1 People who are severely affected

> "Severely ill are severely overlooked – just ignored and invisible."

Estimates suggest that up to 25% of people with CFS/ME are so seriously affected that they are unable to perform most basic personal tasks and are confined to bed or spend the majority of the day in bed. Such patients feel particularly alone and isolated. The severity, complexity, and longevity of the illness are poorly understood.

> "Severely affected are particularly vulnerable from lack of medical attention, understanding, and home attention."

2.3.1.1 Primary care

Views were expressed that when GPs were confronted with a patient whose illness is complex and does not improve with treatment then they can feel helpless, and may effectively "withdraw to the sidelines" leaving the patient to feel totally alone.

> "Dumped in the community – totally invisible."

The lack of agreement over advice and management compounds the difficulties of the severely affected. In general, this group is excluded from research, so they may not fulfil criteria used to test evidence-based approaches. For example, many comment on the inappropriateness of extreme exercise regimens that have been studied in less adversely affected patients. Some report that they want to believe doctors and feel "frightened to say no", or that they do not have the energy to disagree. Fears were also expressed over: being "branded" as a "difficult patient", losing benefits, letting people down, not trying, losing the love of the family, and being labelled as mentally ill.

Severely affected – patients' suggestions (primary care)

- Supportive GPs and home visits.
- Assessment for home support services and liaison with Local Education Authority.
- Outreach services and trained nursing support.
- Appropriate referrals to specialists.
- Appropriate advice and management strategies.
- Respite care.
- Support and advice to carers.

2.3.1.2 Further care

> "Very difficult to seek any specialist attention – cannot get to hospital."

Severely affected individuals find several aspects of specialist care inadequate: in general, specialist advice is more difficult to obtain; outpatient and inpatient care are woefully inadequate for their needs; and such services do not take account of the needs of people whose symptoms may include painful sensitivity to light, noise,

and chemicals, who have special dietary requirements, and who need to rest more than other patients.

Clinicians who are interested and experienced in CFS/ME are few in number and patchy in location. Long travelling distances and lack of choice particularly influence the severely affected. Medical interventions and research generally "miss" those severely affected because of their mobility problems and a perception that they are "too unstable" for inclusion in trials. Therefore, the patients' suggestion is that these issues are kept in mind when any treatment programmes for them are under consideration or development.

> An analogy was drawn that Alzheimer's disease is not known as "Chronic Forgetfulness Syndrome!"

This group reflected strong loathing of the name CFS because fatigue is often not perceived to be their main problem; ME is a preferred term by many.

Severely affected – patients' suggestions (further care)

- Appropriate arrangements for outpatient consultation.
- Follow-up consultations by telephone.
- Appropriate referrals between specialists.
- Universally accepted grading system for the severely affected.
- Development of outreach services including nursing and occupational therapy.
- Home monitoring and visits from specialists.

2.3.2 Children and young people

The uncertainties surrounding the illness are particularly difficult for children and young people.

> "People saying what's not wrong with you and never what is."

2.3.2.1 Awareness and understanding

Ignorance and lack of understanding of the condition are perceived to exist among healthcare professionals as in society, with a widespread perception that "children don't get CFS/ME". Patients want supportive and sympathetic clinicians who believe in their illness and can provide information, advice, and access to other services.

> "I don't see my friends at all and it's the ones that I don't know as well that stood by me but I'm still isolated."

Younger people do not always understand their friend's illness, and some may share the widespread disbelief surrounding CFS/ME. These problems are compounded by absence from school, which inevitably restricts contact, leading to feelings of isolation.

Children and young people – patients' suggestions (awareness and understanding)

- Action to address misconceptions.
- Central involvement of the Royal College of Paediatrics and Child Health in bringing about a change in attitudes.
- High-profile awareness campaign – TV, newspapers, and doctors' surgeries.
- Fact sheet for children and young people.
- Training and dissemination of good practice.
- Funded research programme into the causes, intended to lead to better approaches in management.

2.3.2.2 Service provision

The Sounding Board event identified several general aspects of care important to children and young people with CFS/ME and their families. Overall, the message is that the underpinning philosophy of service development should focus on the young person's perspective. In doing so, it could provide a more efficient, effective, and supportive care pathway.

Delays in obtaining a diagnosis result in delays in accessing appropriate services. Once a diagnosis is obtained, there is a lack of informed professional advice, monitoring, and support in the everyday management of the young person, and limited access to counselling services specifically for people with CFS/ME. Services may be limited for patients who are too ill to stay at home, and hospitals are generally unable to offer appropriate care for the severely affected younger person. Participants at the Sounding Board events noted the lack of services focused on young people in which healthcare professionals have a broad understanding of the illness.

Strong polarised views which professionals may hold can deny young people access to appropriate supportive services, or result in inappropriate referrals. Professional controversy can contribute to creating "a difficult time at home", with the child or young person conscious of the burden placed on other family members. Disruptions to family functioning have led some professionals to assume that the cause of the illness is "overprotective" parents, with further adverse impact on the child and family.

The lack of public understanding is reflected in the lack of interest from schools in providing appropriate contact and support. School nurses' and health-visiting services' understanding of CFS/ME is considered to be limited.

Children and young people – patients' suggestions (service provision)

- Development of services around the young person's perspective.
- Listening to the young person, with or without their parents according to their choice.
- Awareness by professionals that "what they say" has a significant impact on the young person.
- Avoidance of the assumption that parents are "overprotective".
- Avoidance of the message to young people that it is "all in their mind".
- Appropriate NHS care provision to support family and self management concept.
- Partnerships between clinical services and voluntary organisations.
- Access to benefits facilitated by sympathetic assessment by professionals with experience of CFS/ME in children and young people.
- Improved liaison between health professionals, teachers, and Local Education Authorities.
- More school/community nurses who know about CFS/ME.

In addition to these suggestions of general principles for service provision, those consulted advocated key steps in the management pathway.

Children and young people – patients' suggestions (management)

- Early diagnosis.
- GP referral for specialist advice from paediatrician with an interest or CFS/ME specialist.
- A balance of service provision between primary and further care to be determined for the individual.
- Information, advice, and ongoing support via regular visits to or from the GP or specialist nurse, for both the child/young person and the family.
- Full explanation in accessible language of the nature of the illness and management choices, agreement on an individualised management plan, and support for self management, including provision for fluctuations.
- Mitigation of symptoms, such as pain, nausea, dizziness.
- Offer of psychiatry/psychology if needed as part of a wider choice of help available.
- Regular, supportive follow-up with a multidisciplinary team, including GP, paediatric nurses, school nurses, physiotherapists, occupational therapists, psychologists, social workers, usually co-ordinated by a community paediatrician.
- An individually tailored education programme, for return to school, college, work as recovery proceeds.
- Respite care, especially for those severely affected.

2.3.3 Class/ethnic differences

Cultural and class issues may pose special difficulties for patients from different social and ethnic minority groups. Issues can arise from individuals' differing expectations of their role within the family and social structures, which may be especially difficult for women. Further, cultural or religious groups have very

different health beliefs and attributions, and such differences can create additional social tensions and misunderstanding. Stigma and prejudice surrounding the disease can be especially problematic, not only for the individual with CFS/ME, but also within the social group.

Certain ethnic groups are under-represented in the gathering of statistics on CFS/ME; a social class bias in research is also suggested by some. Under-representation is an issue of equality or social justice, but also:

- Undermines the accuracy of incidence and prevalence statistics;
- Questions the basis of present organisation of services and the pattern of future development;
- Has serious and often negative consequences for the individual's experience of the illness.

2.3.4 Carers

Having a loved one affected by CFS/ME has a profound effect on every part of life. Many carers reported that their world felt it had been "turned upside down" and they had feelings of "despair". The difficulties experienced by patients around the widespread disbelief and misunderstanding compound problems for carers. One important aspect for carers – considered by some to be the most difficult to come to terms with – is the uncertainty about restoration of the previous level of functioning, and restructuring of both the patient's and carer's lives. Increased access to respite care would have a substantial positive impact on carers. Otherwise, for carers, particularly for those looking after severely affected patients, there is no rest.

Some carers were clearly distressed about being ignored by GPs, and some reported unpleasant behaviour. Some reported that treatment regimes were occasionally advocated by clinicians against "expressed" wishes leading to loss of trust. In contrast, some reported supportive GPs who admitted their limited knowledge and "feeling out of depth" with the illness, and treated the family with respect. Carers respect honesty and prefer not to be given an over-optimistic prognosis.

2.3.4.1 Carers of children and young people

Parents expressed feelings of isolation resulting from society's misconceptions of the illness and the change in family circumstances owing to the impact of the illness. The lack of understanding of the fluctuating nature of the condition, the range of symptoms, and variation between "sufferers" creates barriers beyond access to health care. Parents and young people drew attention to the difficulties in obtaining help and services, particularly practical help (e.g. wheelchairs, mobility badges). The impact on families from loss of earnings is marked when a parent has to assume a full-time caring role.

As with any other chronic illness, children, young people, and their families identified help that was required to cope with the psychological and social impact of illness. Personal advocates can be helpful to assist with negotiation between the Local Education Authority, the school, and the Benefits Agency. A key suggestion was for co-ordination of help from other agencies at government level; this includes the Department for Education and Skills and the Department for Work and Pensions.

Chapter **3**

Nature and impact of CFS/ME

The term CFS/ME encompasses a relatively common and characteristic clinical disorder or spectrum of disorders that can lead to substantial ill health and disability in people of all ages. The aetiology (cause) and pathogenesis (disease mechanisms) are unclear, although research is increasingly providing important clues, notably on factors that predispose, precipitate, or perpetuate the condition. Although the disorder is clinically recognisable, CFS/ME assumes many different clinical forms and is highly variable in severity and duration, but lacks specific disease markers.

These factors are likely to have contributed to the poor recognition of the nature of the disease, its clinical and personal impact, and its wider societal effects. The many names it has been given, and the various case definitions proposed (see **Appendix II** and **Annex 1**), have added to the difficulty of its characterisation, impeding both clinical care and substantive research. This chapter reviews what can be said about the nature and impact of the condition in adults. Much is applicable to children and younger people, although important differences and special considerations are given in **Chapter 5**. Further material on general concepts and philosophy of disease is available in **Annex 4**.

Key messages

- CFS/ME is a relatively common condition of adults and children that is clinically heterogeneous and lacks specific disease markers but is clinically recognisable.
- The broader impact of the disease even in its milder forms can be extensive; people who are severely affected and/or with longstanding disease are profoundly compromised, and improvement of their care is an urgent challenge.
- The aetiology (cause) of CFS/ME is unclear, although several predisposing factors, disease triggers, and maintaining factors have been identified.
- The pathogenesis (disease process) underlying CFS/ME is also unclear. Research has demonstrated immune, endocrine, musculoskeletal, and neurological abnormalities, which could be either part of the primary disease process or secondary consequences.
- One highly heterogeneous disease might exist that encompasses CFS/ME or several related pathophysiological entities may exist; these distinct hypotheses should be studied.
- Current evidence does not allow complete distinction between CFS and ME, or useful delineation of subgroups. Every patient's experience is unique, and the illness should be managed individually and flexibly.

3.1 Introduction

CFS/ME has been increasingly recognised in recent decades, though it may have existed for centuries. It is not clear whether the disease is more common now than previously, or whether this impression is due to increased awareness and ascertainment of cases.

There have been several attempts to harmonise a case definition to assist surveillance and research, although none has been completely satisfactory. Nevertheless, case definitions have led to more reliable epidemiological data and to more consistent approaches for studies on clinical recognition and management. However, uncertainties about the nature and cause of the illness, and disputes about these from different perspectives, have delayed recognition of the disorder by healthcare professionals, by other individuals and agencies with whom patients and carers interact, and also by the public.

Despite the many remaining uncertainties about its aetiology and pathogenesis, we examine the nature and impact of the condition with the following aims:

- To support a more consistent approach to prompt clinical recognition and appropriate management of CFS/ME in primary care and specialist/hospital practice;
- To encourage investment in broad research covering clinical, epidemiological, health-services, and basic scientific aspects of CFS/ME and its management; and
- To assist families, carers, employers, educational institutions, and benefits and other agencies to develop more appropriate responses to people affected by CFS/ME.

3.2 Definitions and terminology

Many terms have been used to encompass this condition or clinical entities within this disease spectrum. Some of the more common definitions, including those used in research, are to be found in **Annex 4** and **Appendix II**.

The term “syndrome” is widely used in medicine to encompass a variable pattern of disease, whether or not the syndrome has one unique causal mechanism. “Chronic” indicates long duration. For some, the term “fatigue” is problematic and considered demeaning because it is common parlance for the physiological experience of tiredness, whereas patients’ experience in CFS/ME is profoundly different. Also fatigue, though invariably present, may not be the major symptom. “Myalgic” is similarly inappropriate for those patients with little muscle pain. “Encephalomyelitis”, meaning inflammation of the brain and spinal cord, is incorrect because the term implies a pathophysiological process for which no evidence exists.

“Encephalopathy” has been suggested because this term lacks the implication of inflammatory change, while suggesting a significant focus of disordered function in the central nervous system. The term “immune dysfunction” is unsatisfactory, because the relevance of observed abnormalities and an immune cause for the disease are not established.

The Working Group decided that the most important requirement in terminology is for patients and doctors to agree a satisfactory term as a means of communication. We recognise that no current terminology is satisfactory, so in line with our original terms of reference, we have used the composite CFS/ME for the purposes of this report, acknowledging that CFS is widely used among clinicians and ME among patients and the community. A group in the USA, with international input, is currently discussing terminology for this condition, and this work will, we hope, lead to an internationally acceptable terminology for patients and professionals.

3.3 Aetiology, pathogenesis, and disease associations

The aetiology (cause) and pathogenesis (disease process) of CFS/ME are not clearly elucidated, and uncertainty continues to surround these issues. Although CFS/ME has certain characteristic features, the condition is heterogeneous either in causative factors or in its clinical nature. The heterogeneity could represent the range of a single condition (as with other diseases, such as diabetes), or could mean that several distinct diseases are being bracketed together because of the similarity of their clinical appearance (as with severe combined immunodeficiency). These possibilities complicate the consideration of aetiology and pathogenesis, as they do other aspects of the condition.

Research has demonstrated immune, endocrine, musculoskeletal, and neurological abnormalities. To what extent these abnormalities are part of the primary disease process or secondary consequences remains the subject of debate. The possibility that one highly heterogeneous disease might exist that encompasses CFS/ME or that several similar pathophysiological entities occur should be kept in mind so that these opposing hypotheses can be tested in research studies (see also **Annex 4**).

Several overarching possibilities, which are not mutually exclusive, have been proposed to explain the occurrence of CFS/ME, including:

- CFS/ME is an umbrella term for several different illnesses.
- One (or more) ‘core’ disorder(s) exist.
- Several different causative factors trigger a common disease process.
- The aetiology and/or pathophysiology are multifactorial.
- Certain factors are necessary but not sufficient to cause CFS/ME.
- Certain factors can influence individual manifestations or duration.
- Some features are downstream (secondary) consequences of the primary disease process.

There is good-quality evidence that some factors trigger CFS/ME, while others maintain it, although evidence of predisposing factors is limited.

3.3.1 Predisposing factors

Gender – Incidence in females exceeds that in males of any age.

Familial – The familial incidence of CFS/ME is slightly higher than expected, which suggests that familial factors may play a part in susceptibility. Twin studies suggest a hereditary component but family environmental factors also may have an influence.

Personality – There is evidence both for and against the possibility that certain personality traits might predispose people to develop CFS/ME. Positive findings from retrospective studies could be explained by the effect of chronic disability on personality.

Other disorders – Some patients have a past or current history of other disorders, particularly fibromyalgia and irritable bowel syndrome. The association of other disorders with CFS/ME could reflect different manifestations of a similar process or different expressions of a common predisposition. Alternatively, other disorders could in some way predispose a person to CFS/ME.

Previous mood disorder – Most, but not all, studies have found a history of mood disorders in individuals with CFS/ME. This finding might simply reflect the fact that previous mood disorders predict future mood disorders, which often coexist with chronic illnesses, including CFS/ME. Alternatively, this finding could reflect a common predisposition to both mood disorders and CFS/ME.

3.3.2 Triggers

Infections – Good-quality evidence indicates that certain infections are more common triggers for CFS/ME than others. Glandular fever, viral meningitis, and viral hepatitis are followed by CFS/ME in about 10% of cases of the primary infection. CFS/ME can follow infections with herpes viruses, entero viruses, hepatitis viruses, and some other viruses, and also non-viral infections such as Q fever. CFS/ME has been reported after salmonellosis, toxoplasmosis, and brucellosis. Influenza and 'flu-like infections can trigger CFS/ME, but common upper respiratory tract infections do not seem to. Available evidence suggests that abnormal persistence of infectious agents does not occur in CFS/ME, although certain chronic infections can cause similar symptoms.

Immunisations – A few case reports have suggested that CFS/ME has occurred after immunisations, though intercurrent events, including infection, might have played a part in the disease process. It is biologically plausible that some processes seen after infections could also occur after immunisations, but this has yet to be confirmed by a good quality cohort study in the case of CFS/ME. Current advice to avoid immunisations during infections is designed to avoid such triggering.

Life events – The evidence that life events can trigger CFS/ME is weak. Severe life events are much more likely to provoke a mood disorder, which can be misdiagnosed as CFS/ME. However, clinical and patient experience suggests that increased "stress" may be common around the onset of symptoms or a triggering event, such as infection. It is unclear whether this is as a triggering, a predisposing or a maintaining factor. Stress is also recognised as a trigger for setbacks.

Physical injuries – These may be more likely to trigger the seemingly related condition of fibromyalgia than CFS/ME, though instances of CFS/ME after physical or operative trauma have been described.

Environmental toxins – Reports have suggested an association between exposure to environmental toxins, such as organophosphorus compounds, and development of disease in isolated cases. The balance of evidence indicates that this is not a common or widespread trigger.

3.3.3 Maintaining factors

Sleep disturbance – The majority of patients with CFS/ME experience sleep difficulties, which are generally independent of mood disorders, but can contribute to cognitive dysfunction. Poor sleep quality will also exacerbate fatigue and other symptoms.

Mood disorders – Disorders of mood, especially depressive and anxiety disorders, occur in a large minority of CFS/ME sufferers. They are important to identify or exclude because they can either mimic or coexist with CFS/ME. Mood disorders can exacerbate, modify, or contribute symptoms, and can affect adaptation and recovery. However, mood disorders can also be misdiagnosed in patients with CFS/ME because of the overlap of key symptoms.

Inactivity – A decrease in activity is an obvious consequence of CFS/ME. If prolonged, inactivity may then become a problem in its own right, with consequent loss of physical fitness, problems with balance and temperature control, autonomic dysfunction, loss of confidence, and sleep disturbance. Research evidence suggests that patients with CFS/ME seem no more physically unfit than sedentary people and may be as fit as non-sedentary people, although one study suggested that pervasive inactivity occurs in a quarter of patients. The importance of deconditioning in the disease process is contested. Although clinical wisdom and some research suggests that the degree of physical deconditioning is likely to be linked with severity of disease, this finding is not universal, and other as yet undetermined factors must also predict the extent of impairment. Pervasive inactivity can predict non-response to certain treatments.

Overactivity – Activity beyond the level that an individual can usually tolerate will prompt a delayed worsening of symptoms. Observation suggests that patients who show cycles of overactivity followed by setbacks (‘boom and bust’) may have a more protracted course.

Intercurrent stressors – Clinical experience suggests that emotional and physical stressors, “stress” including intercurrent infections, vaccinations, and surgical operations can cause setbacks in some CFS/ME patients.

Iatrogenic illness – As with all conditions, clinical management strategies can sometimes contribute to maintenance of the disease. These could include inappropriate advice (to exercise too much, or to rest too much), misdiagnosis (e.g. diagnosing a psychiatric disorder when one is not present, or missing such a disorder or other diseases because of misattribution of symptoms to CFS/ME), and inappropriate prescribing. Failure to acknowledge the patient’s illness or to provide supportive care can cause additional distress and alienation, and might encourage the patient to seek unconventional/untested remedies, some of which may cause harm.

Illness beliefs – The way in which abnormal illness behaviour and illness attributions (especially about cause) may be perpetuating ill health and disability in some

CFS/ME patients remains a contentious issue. It is thought that certain strongly held beliefs about the cause of the illness can impede progress. These include the view that the illness is entirely physical or is caused by a persistent virus. These beliefs could be partially correct – e.g. a virus could have provoked a persistent or prolonged change in physical functioning. However, they could also act as obstacles to recovery or to necessary treatment. It seems important that patients and professionals keep open minds since knowledge continues to grow. Positive attitudes and cooperation based on mutual respect seem likely to produce best outcomes.

3.3.4 Possible disease mechanisms

The research literature contains several hypotheses and proposals to explain how CFS/ME may be caused or maintained. The quality of the evidence is variable, however, and many suggested mechanisms are as yet based on associations rather than cause or linkages. This overview outlines the scope of the ideas:

Biomedical model – In this overarching conceptual framework, CFS/ME is seen as a condition like many other medical conditions where illness results from a specific pathological defect in physiological functioning, mediated at organ, tissue, cellular and/or molecular level, by as yet undefined mechanisms. It is not incompatible with the following, but implies that a primary disease entity exists and that the biopsychosocial aspects are consequential.

Biopsychosocial model – The biopsychosocial model of pathophysiology, applicable to all disease, suggests that once an illness has started its expression is affected by beliefs, coping styles, and behaviours, while consequential physiological and psychological effects act in some ways to maintain and/or modify the disease process.

Immune – Immunological abnormalities are common in patients with CFS/ME. The findings are mostly non-specific, and their relationship to the illness has not been established. The pattern suggests some immune dysregulation, with activation or suppression of different components, as indicated by changes in cytokine concentrations and cell surface markers. In atopic patients, case reports suggest that allergic manifestations can be exacerbated or triggered.

Hypothalamic-pituitary-adrenal axis – Several studies have found subtle neuroendocrine abnormalities, particularly hypoactivity of the hypothalamic-pituitary-adrenal axis. It is also possible that disturbances in hypothalamic function could contribute to some CFS/ME symptoms such as fatigue, sleep problems, and disturbed thermoregulation. The possibility remains that these changes are directly or indirectly involved in pathogenesis.

Central nervous system – Many of the symptoms of CFS/ME suggest dysfunction of the central nervous system. These could include cognitive disturbance, central fatigue (e.g. when movement requires increased mental effort), and disrupted neural regulatory mechanisms (e.g. those involved in sleep and temperature regulation). These changes could be primary or secondary to some widespread process.

One suggested primary change in the central nervous system of patients with CFS/ME is abnormal brain blood flow, particularly involving the brain stem.

However, many of these findings are inconsistent. Furthermore, regional brain blood flow can be altered by factors unrelated to any disease process; it is also subject to autoregulation, which means that local changes in blood flow could reflect altered activity in that brain region rather than the cause of altered activity. Measurement techniques are still being developed and selection of controls with other brain disorders is crucial to interpretation of the subtle changes seen.

Magnetic resonance imaging studies have found subtle white matter abnormalities in some individuals, more common in those without coexistent psychiatric conditions, leading to the hypothesis that some CFS/ME patients have a subtle encephalopathy. The numerous studies on cognitive functioning have not always found consistent results and can be criticised for not reflecting the severity of subjective complaints. However, it seems likely that cognitive dysfunction in CFS/ME cannot be explained solely by the presence of a coexistent psychiatric disorder. Vestibular dysfunction is proposed to explain the widely reported symptom of "dysequilibrium".

Peripheral lesions – The roles of dysfunction in the peripheral nervous system and muscles are uncertain, though some indirect evidence and specific symptoms in individuals have implicated them.

Autonomic nervous system – Autonomic dysfunction seems to play a part but its role is not established. There is inconsistent evidence as to whether autonomic abnormalities, in particular neurally mediated hypotension, are part of a primary disease process or due mainly to inactivity associated with CFS/ME. For example, one study has found evidence of lower cardiac stroke volumes – a finding that may indicate covert cardiac dysfunction or reduced blood volume. It should also be noted that low fluid and salt intake with or without increased loss may be important in causing orthostatic problems in some patients.

3.4 Spectrum of illness

3.4.1 Subgroups

The issue of subgroups or discrete entities within CFS/ME was the subject of much debate by the Working Group. We are conscious that some sectors strongly hold the view that the term ME defines a subgroup within CFS, or even a distinct condition. The Working Group accepts that some patients' presentation and symptoms align more closely to the original clinical description of ME[1] than to the current definition of CFS by the US Centers for Disease Control and Prevention.[2] However, there is currently no clear scientific evidence to allow formal differentiation of ME from CFS on the basis of pathophysiology or response to treatment. Therefore, for the purposes of this report, we regard CFS/ME as a single, albeit diverse, clinical entity.

We hold the view that every patient's experience is unique and his or her illness must be treated flexibly in its own right, from a range of options that are generically applicable to the disorder but individually adapted. This approach is similar to that for many other conditions.

3.4.2 Symptom profiles

Patients with CFS/ME experience an individual array of symptoms from the overall range seen in the illness. Some, such as physical and/or cognitive fatigue are seen in almost all patients, though their extent can vary. Others are very common, such as pain, disturbed sleep, and gastrointestinal disturbance.

In addition to symptoms that occur in the majority, individual symptom complexes may 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 patients, symptoms remain relatively constant in type, whereas others experience an evolution through different 'layers' of symptoms, in some cases with a recurrence of early symptoms during recovery. In some women, premenstrual or menstrual exacerbation of symptoms is reported by patients.

The profusion of symptoms in several body systems can be confusing and alarming for patients, who can find this hard to encompass in a single overarching explanation. Consequent fear over the possible significance of discrete symptoms can be very intrusive and distressing. This uncertainty is difficult for patients and those around them. As in other chronic conditions, a new symptom can raise new questions about the reliability of the diagnosis, or concern that a quite different pathology could be missed if the symptom is immediately attributed to CFS/ME.

An individual's symptom profile is modified by the impact of illness on the person affected and those around them. Patients' experience can be one of frustration about the inability to function at previously normal levels, about continual setbacks, and about the lack of understanding or disbelief from people around them. Anxiety or depression, anger, and withdrawal from social interaction are relatively common consequences in response to the impact of any chronic illness on personal and social functioning. These understandable reactions add to distress, and in some cases become part of, or even dominate, the clinical picture in CFS/ME. In vulnerable patients or at difficult times, suicidal ideation can occur, and suicide becomes a serious risk.

Despite these difficulties, most patients establish a tolerable level of functioning, especially with appropriate support of family, friends, and professionals. The extent of constructive adaptation to their very changed circumstances and expectations is often remarkable.

3.4.3 Severity

Severity of any illness can be understood in several different dimensions. So for each individual, account must be taken of the way in which his or her symptoms, reduced levels of physical and cognitive activity, and altered social functioning have an unwelcome impact on normal life, goals, and expectations. Above all, each patient must be assessed on his or her own expression of the illness and actual functional level.

The Working Group is concerned that it is necessary to make these points for CFS/ME, when such considerations are self-evident and part of usual clinical practice for other disorders that are better recognised. Appreciation and understanding are essential if the needs of different patients are to be met effectively and appropriately, in medical and social care, education, and employment, as well as in attitudes of family, friends, and society.

The term 'severely affected' has been widely applied to patients whose physical disability is most severe, leading to serious restrictions in mobility and functioning. In many, these restrictions are accompanied by other markers of severity, such as cognitive impairment or prolonged course. This degree of physical restriction, especially if prolonged, has profound effects on personal and social functioning, which in turn substantially affects the patient's ability to access health and social services, and has an impact on the patient's carers.

A recent description has suggested four categories of severity in CFS/ME:[3] although care must be taken not to diminish inadvertently the experience of any patient by descriptors of severity.

> "**Mild** – Are mobile and can care for themselves and can do light domestic tasks with difficulty. The majority will still be working. However, in order to remain in work, they will have stopped all leisure and social pursuits, often taking days off. Most will use the weekend to rest in order to cope with the week."
>
> "**Moderate** – Have reduced mobility and are restricted in all activities of daily living, often having peaks and troughs of ability, dependent on the degree of symptoms. They have usually stopped work and require rest periods, often sleeping in the afternoon for one or two hours. Sleep quality at night is generally poor and disturbed."
>
> "**Severe** – Will be able to carry out minimal daily tasks only, face washing, cleaning teeth, have severe cognitive difficulties and be wheelchair dependent for mobility. These people are often unable to leave the house except on rare occasions with severe prolonged after-effect from effort."
>
> "**Very severe** – Will be unable to mobilise or carry out any daily tasks for themselves and are in bed for the majority of the time. These people are often unable to tolerate any noise, and are generally extremely sensitive to light."

Indeed, there may be severe impact on people's lives even of less overtly severe CFS/ME, as the descriptions offered by Cox and Findley[3] for mild and moderate CFS/ME suggest. Such patients may suffer most impact through the discrepancy between what they were able to achieve previously and what they can now do. Even

less prolonged illness, whatever the severity, can have very substantial personal and social impact, mainly intrusions on the individual, relationships, work, and finances. Self-confidence and self-esteem are severely eroded in many cases.

Attempts by individuals to maintain activity close to a previous level of functioning can be unrealistic and unsustainable. This realisation can cause additional distress, compounded by the responses of those around the patient to the confusing signals they receive, and the unpredictability of the patient's levels of functioning.

3.4.3.1 People with severe illness

The descriptions above give an indication of the functional impact of severe disease and an indication of consequent needs. Current provision of services falls well below what is needed for the vast majority of severely and very severely affected patients.

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'. The lack of simple technical solutions and the great difficulty that some professionals and others have in facing the uncomfortable reality of the illness, especially in a severe form, can compound the problem.

The duration of illness and disability due to CFS/ME can itself become part of the severity of the disease's impact, for any duration of illness can be intrusive and cause substantial problems. Severe illness that continues over many years with no sense of improvement has a profound cumulative personal and social impact. A minority of those with CFS/ME remain permanently severely disabled and dependent on others. Yet, even if we lack easy solutions, professionals can still support, care, and provide for many patients' needs by reaching such patients in their homes, maintaining contact, and continually exploring potential options. Those who are most severely affected need acknowledgement, encouragement, and support to remain optimistic.

3.5 Socioeconomic impact

3.5.1 Work, finance, and education

Evidence from patients and clinicians suggests that there can be a substantial impact on work, finance, and education. Many people's circumstances are linked to continuing income from full-time working, and if an individual is unable to work, the consequences can be considerable. If available, sick pay is often halved within six months and then may cease within a relatively short time. People with CFS/ME frequently experience problems with accessing state benefits. This is partly because of the variable nature of CFS/ME and uncertain prognosis, but sufferers may also have difficulty obtaining a diagnosis, and thus in obtaining benefits. Improved knowledge and understanding of the condition among clinicians will help to eliminate this.

Such factors, of both a financial and social nature, can often dominate patients' early approaches to their illness, sometimes encouraging premature return to unsustainable levels of work, which exacerbates their own and their employers' loss of confi-

dence. Return to work, even after prolonged absence, can be hard to negotiate at levels realistic for these patients, and the potential for a 'benefits trap' is only partially ameliorated by current rules on therapeutic work and therapeutic earnings.

Another problematic gap can occur for patients who are too ill to work for long periods, but are unable to access remuneration from insurance policies or ill-health retirement. This often occurs because the loss of functioning that prevents work is different from that deemed necessary to claim from health insurance or allow ill-health retirement. The most common obstacles are duration or the need to demonstrate permanence of the condition.

The period of absence from work that can lead to complete loss of earned income is typically 12 months, but ill-health retirement is usually only considered after illness of some 3 years' duration. The requirement of many pension or personal health insurance schemes to demonstrate permanence (usually through medical reports) requires a level of prognostic foresight that may not be realistic. Also, such a requirement could potentially encourage a negative and fatalistic view of long-term rehabilitation so that people can access sufficient financial support to cope through prolonged illness.

Patients can encounter arbitrary and poorly informed decision-making on other issues such as home help and mobility badge schemes, as well as sheer resource limitation. Failure to access appropriate support from social services can be compounded if doctors fail to provide clear guidance about diagnosis and need.

Similar issues arise over education, not only school but also higher education. Access to educational institutions represents a serious barrier for children and young people with CFS/ME. Once there, the individual's inability to sustain his or her expected normal rate and level of achievement can cause further difficulties. Peer pressure and disruption from usual peer-group activities have a particularly adverse impact. Premature pressure to return to education may be particularly damaging.

3.5.2 Social impact

The pervasive social impact of this illness and its consequences can be glimpsed from comments in the Sounding Board events, and in many individual and collective representations to the Working Group (see **Chapter 2** and **Annex 3**). Patients, carers, and the professionals working with them continually face the practical constraints that the illness imposes and the limited support available through statutory services, in both volume and type.

Much more complex issues may also face patients as a result of their illness. As in much other chronic illness, their role can change radically. They cease to function as the bread-winner, the parent, the spouse, or the one who sorts things out, and the loss of their working role and other social functioning can all cause serious adjustment difficulties. Loss of confidence, self-esteem, and self-efficacy can be demoralising and demeaning, and an inability to cope, because of its unfamiliarity, can mean that patients lack defined pathways for seeking help.

Chronic illness can alienate patients from usual social contacts, who may find its persistence, its unpredictability, and its invisibility both confusing and threatening.

This is particularly troublesome for children. Disruption of usual support mechanisms can greatly exacerbate the direct effect of the illness, increasing upset and anger. Guilt about the illness and the patient's reduced functioning is very common; not only adding to distress but also encouraging patients to attempt tasks and levels of functioning that they cannot sustain.

A proportion of patients feel alienated from clinical professionals by early responses to their symptoms, illness experience, and disability. Actual or perceived dismissiveness, incomprehension, or even disbelief are encountered, and have profoundly negative impacts. Such attitudes can also lead the patient to seek help from alternative and complementary therapists, without feeling that they can obtain advice about such therapies from orthodox clinicians. Thus, patients may come to rely excessively on unproven, unregulated approaches, rather than the regulated, evidence-based services that can and should be available through the NHS. Many complementary therapists are supportive, and they often provide time and personal attention, while some report achieving positive benefits with various approaches.

However, there is concern that some therapists can instil confusing or misleading health beliefs, recommend unnecessary, unvalidated tests or potentially hazardous therapies, or encourage the patient to spend considerable sums of their limited resources.

Key references

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Chapter **4**

Management of CFS/ME

Appropriate management and service provision for patients with CFS/ME and their carers are urgent priorities. Of the areas addressed by the Working Group process, the nature of appropriate management is the area perhaps most beset by confusion and controversy. However, many constructive steps can be taken. This chapter details the general principles and some specific advice for management of the condition(s) CFS/ME, as defined by the Working Group (see also **Chapter 3** and **Annex 4**). The research evidence base is detailed further in **Annex 5**. The aim of this chapter and the accompanying summary for clinicians (**Annex 6**) is to lay down the current perceptions of best practice, based on research evidence where available, clinical experience, and wider consultations with those affected and their advocates. We appreciate that not all areas are addressed with ideal scope and certainty, and this chapter also aims to highlight those areas for which consensus has not been reached and where further research would be most informative. Additional considerations and special circumstances for children and young people are covered in **Chapter 5** and **Annex 7**.

Key messages

- Initial professional responses to CFS/ME can have major impact on the patient and carers. Clinicians should listen to, understand, and help those affected to cope with the uncertainty surrounding the illness.
- Early recognition with an authoritative, positive diagnosis is key to improving outcomes. Symptoms are diverse, but increased activity frequently worsens fatigue, malaise, and other symptoms with a characteristically delayed impact.
- All patients need appropriate clinical evaluation and follow-up, ideally by a multidisciplinary team. Care is ideally delivered according to an agreed flexible management plan, tailored from a generically applicable range of options.
- Therapeutic strategies that can enable improvement include graded exercise/activity programmes, cognitive behaviour therapy and pacing; intrusive symptoms and co-morbid conditions may also require specific management.
- The overall aim of management must be to optimise all aspects of care that could contribute to any natural recovery process. Management strategies need regular review to guide their application and adaptation to the individual.
- Education and support, plus measures to tackle the broader impact of the disease, need to be initiated as early as practicable. Much support is provided by the voluntary sector. Patients can be empowered to act as partners in care.

Key messages

- Review of the evidence highlights the lack of good-quality research to support effectiveness of various therapies. Patient responses suggest that no approach is universally beneficial and that all can cause harm if applied incorrectly.
- The goal of rehabilitation or re-enablement will often be adjustment to the illness; improvement is possible with treatment in the majority of people.

4.1 Principles

> "A Physician who does not admit to the reality of the disease can not be supposed to cure it." William Cullen (1710-90)

CFS/ME is a genuine condition that imposes a substantial burden on patients, carers, and families. The lack of certainty surrounding CFS/ME, as for other chronic illnesses with no certain cause or disease process, also poses very real problems for healthcare professionals. Although the Working Group acknowledges this uncertainty, our conclusion is that clinicians need to apply current knowledge despite the remaining uncertainty; inaction due to ignorance or denial of the condition is not excusable.

The ability of professionals to guide the patient effectively through the illness may be undermined by uncertainties over aetiology and pathogenesis, prevalence, natural history and prognosis, the nature and characterisation of CFS/ME, diagnostic tools, clinical management and therapies, and where and when to refer. These uncertainties, together with a lack of knowledge and perceived lack of skill, can disempower professionals and reduce their ability to access generic clinical skills. However, positive things can be done, both by adopting principles of good practice for management of chronic illnesses, and by taking account of special considerations in the individual's illness.

A key difficulty faced by the Working Group in considering management is the divergence of views on general models of disease. The nature of such divergence can be illustrated by considering polar views: one view holds that a disease is caused by an external disorder that "strikes people down" and for which one can seek a cure or learn to live with the disease burden; an opposing view (the biopsychosocial model) holds that illness arises out of an interplay between a set of external and internal circumstances, which may include physical, psychological, and social factors that precipitate and/or modify the condition. Another classic divergence is the designation of disease as purely physical or psychological, although others adopt a more holistic view – i.e. that physical, psychological, and other features are inter-related.

Divergence of views on the model of disease can influence clinical management of CFS/ME, since the model of disease held can inform management sought and offered. In the example considered above on disease causation, the former view implies that management should be symptomatic only, while seeking a cure for the specific cause; moreover, any behavioural, psychological, or social aspects of the individual's disease may be viewed as maladaptive responses and necessarily treatable. The second view implies that management strategies should target any factors

that seem modifiable in the individual and address triggers and modifiers as part of the disease process rather than as symptomatic therapy. The Working Group has attempted to synthesise aspects of these styles of management, since ideally all approaches are applicable irrespective of one's view of the disease.

During the Working Group process, we agreed that we could:

- Identify approaches to management for which there is evidence of clinical effectiveness;
- Identify approaches that are considered 'common sense' clinically or are reportedly beneficial to patients, for which there is limited scientific evidence; and
- Develop as annexes to this report, resource tools to guide diagnosis and clinical management (**Annexes 6** and **7**).

The Working Group has not aimed to achieve consensus in all areas but rather to delineate explicitly agreement and difference of views where they exist.

4.1.1 Recognition, acknowledgement, and acceptance

Much of the distress surrounding CFS/ME is caused by difficulties in recognition, acknowledgement, and acceptance of the condition and its impact, by both professionals and the public. This distress in turn can affect patients' and their carers' adjustment to the illness. If such issues affect the individual doctor-patient relationship, they can cause difficulties in obtaining a diagnosis and may further affect adjustment to illness. In the NHS, as in society, increased awareness, knowledge, and supportive attitudes are needed.

The Working Group agreed that a positive therapeutic relationship, built from the time the patient first approaches clinical services and based on a recognition of the impact of the illness, will lead to a more successful outcome. Healthcare professionals should adopt an understanding attitude and should not get into disputes with patients about what to call the illness, or about the belief that "it doesn't exist". A name or 'label' for the illness should be agreed to facilitate communication.

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 information on and discuss: the nature of the condition, approaches to self management, helpful therapies, and how to access other agencies for support and services.
- Agree a name for the condition.
- Give advice on symptomatic treatment.

4.1.2 Approach to management

> "The doctor's job should be to 'heal sometimes, relieve often, comfort always'."

No management approach to CFS/ME has been found universally beneficial, and none can be considered a "cure". However, general principles can be outlined to guide management. Most people with CFS/ME can expect some degree of improvement with time and treatment, so a positive attitude towards recovery needs always to be encouraged. Each individual is best managed according to a unique flexible management plan, in which specific strategies and therapies are tailored to his or her particular circumstances. All clinical interventions carry a potential risk of harm, especially if applied incorrectly; for CFS/ME in particular, imposed, rigid programmes can be actively harmful.

The aim for management will in most cases be rehabilitation or re-enablement, according to the patient's needs and circumstances. Re-enablement should encompass cognitive, emotional, and social aspects as well as physical aspects. Management strategies supervised by a therapist, including activity management, cognitive behavioural therapy (CBT) and so on, can be beneficial, provided that they are agreed and viewed as a partnership. Any rehabilitation or increase in activity should start from an agreed, and possibly very low, baseline and should be gradual. It seems important that all practitioners working with an individual are consistent in approach, and share professional perspectives, while utilising their distinct skills and experience.

Most patients can expect some improvement, especially with treatment. Although a return to previous levels of functioning in the short to medium term is often unrealistic, patients can be encouraged to set targets that involve steadily increasing both physical and mental activities once their condition has started to stabilise. Fluctuations in the condition are natural, potentially resulting in the need to recognise natural plateaux, setbacks or more substantial relapses. Such fluctuations should not be seen as reasons to abandon the management plan, but rather to reassess or perhaps slow down. The fact that not all patients will benefit means that any therapy needs to be carefully supervised.

A multi-disclipinary assessment is key to the provision of a supportive package of health care and social care provision. Although care packages need to be individually tailored, where appropriate they should include visits from primary care teams and assessment of the need for equipment and practical assistance.

4.2 Diagnosis and evaluation

CFS/ME should be treated in the same way as any other chronic illness of unknown aetiology. The aim is to develop a supportive relationship, and provide information and education to assist the patient, families, and carers towards self management with support. Management plans and therapeutic approaches require continual assessment, supervision, and re-evaluation. The frequency of the evaluation will be based on the severity of the illness, and on the plan agreed jointly between clinician and patient, and, in the case of children, the child and parents.

An appropriate evaluation is sometimes difficult to achieve. When faced with complex decisions on management, primary care physicians have sometimes found it difficult to identify an expert from whom to seek additional advice. Furthermore, people with severe and long-standing symptoms, who may be house-bound or bed-bound, may find accessing primary care difficult and help from more specialised care services almost impossible to access. A point made consistently by patients is that the exertion involved and impact of attending hospitals (and to a lesser extent primary care services) have a negative effect on their health and their ability to communicate effectively with practitioners. These obstacles must be overcome in practical ways if we are to ensure that the most affected do not continue to be the least supported.

Evaluation requires an acceptance on behalf of clinicians, patients, and their family/carers that management is dynamic, even though sometimes change occurs slowly. Within health service management and commissioning structures, there must be an acceptance of the need to provide the additional sources of advice and support that primary care teams need. Patients' needs are key to determine appropriate referral pathways, irrespective of the speciality. Openness on the referral and the reasoning behind it is vital.

Adults, young people, and children can obviously develop new illnesses while they are suffering from CFS/ME. Evaluation of new symptoms needs always to be from first principles, to ensure appropriate recognition and therapy, if necessary through referral to specialist care.

4.2.1 Diagnostic process

A key principle for effective management is to establish a working diagnosis early, to give patients and carers a name to put to their experience, and to allow a management plan to be discussed and put in place. The importance of an early diagnosis is a key issue highlighted by patients, because of the potential harm done by late diagnosis. There is also some evidence from other areas of chronic illness management that early diagnostic labelling can minimise the psychosocial impact of illness. An early, authoritative, positive diagnosis is crucial to minimise the impact of the uncertainty surrounding the illness and early responses to it, such as attempting to "work through fatigue". Furthermore, some patient evidence (**Annex 3**) indicates that the lack of a name for the condition, sometimes until quite an advanced stage, prevents people from coming to terms with their illness, and may also limit the ability to implement an effective management plan.

A positive diagnosis of CFS/ME is needed, rather than one of exclusion. Without a validated test for the illness, diagnosis is based on recognition of the typical symptom pattern (see **Annex 6**) together with exclusion of alternative conditions. Thus, a positive diagnosis can usually be made from clinical history, examination, and a few appropriate laboratory investigations, as in other chronic illnesses of uncertain nature.

In the management of any potentially serious illness that lacks a specific diagnostic test, in which the clinical picture is unfolding, and in which time plays a crucial part in the process, a diagnostic triage approach can prove useful. This process recognises that some conditions require more urgent management than others, and these conditions should be explicitly considered and excluded as a priority. For example,

CFS/ME should be considered at an early stage as part of the differential diagnosis when individuals of any age present with symptoms of excessive tiredness or fatigue. The diagnostic process then becomes the familiar one of assembling positive clues from the history and examination, while ruling out other likely differential diagnoses, usually by laboratory investigations (see **Annex 6**).

During this process, a working diagnosis of CFS/ME may emerge as the symptoms start to form a recognisable pattern. Depending on the constellation of symptoms presented and the time-span of the symptom presentation, the clinician will decide on appropriate investigations to be undertaken. When other diagnoses have been excluded and CFS/ME remains as one of the possible diagnoses, a limited set of investigations is usually appropriate. However, this should not dissuade clinicians from pursuing lines of clinical inquiry that will alter management or reduce uncertainty for patients and clinicians. In addition, clinicians may wish to conduct investigations that may improve our understanding of aetiology and pathogenesis, and better treatment; such clinical research, with appropriate consent, is important, but it must be explicitly distinguished from normal clinical care.

4.2.1.1 Diagnostic criteria

Current diagnostic criteria are useful only for research purposes, and no clinically recognised set of diagnostic criteria exists. Moreover, the Royal Colleges Report in 1996 noted that meeting the definitions of the criteria for CFS/ME does not constitute a diagnosis. Whereas the researcher needs to be able to define a disease or syndrome narrowly, the clinician requires a more inclusive approach, but one that is not so broad as to include other similar conditions that require different management. Conversely, it is important to be able to include patients with coexisting diagnoses (separate or interacting) in a way that a research definition would exclude. Weighted criteria-based diagnostic scores have not been validated, and cannot be used as a diagnostic tool until their value has been studied formally. A list of diagnostic criteria appear at **Appendix II**.

A diagnosis of CFS/ME, as with several other chronic illnesses of uncertain aetiology relies on the presence of a set of characteristic symptoms together with the exclusion of alternative diagnoses. The key symptoms of CFS/ME in adults, as listed below, broadly fit with existing research criteria, and form a recognisable pattern of characteristic symptoms of CFS/ME, although the list is by no means exhaustive. One of the most common and characteristic complaints of adults, particularly in the early stages of the illness, is of intolerance to both physical and mental exertion with delayed impact. So perhaps the key pointer to a diagnosis of CFS/ME is the way in which the symptoms behave after increased activity.

Persistent fatigue should be differentiated from acute fatigue, which may follow illnesses such as influenza. It should also be differentiated from other kinds of fatigue (for instance, tiredness due to overexertion, weakness due to neuromuscular disease, and loss of motivation and pleasure due to major depression). These other fatigue states do not present with the characteristic delayed fatigue seen in CFS/ME. Another distinguishing feature of the illness, in comparison with other ‘fatigue states’, is its prolonged relapsing and remitting course over months or years.

4.2.1.2 Characteristic features

The characterising features of CFS/ME are overwhelming fatigue, related effects on both physical and cognitive functioning, and malaise, typically exacerbated after physical or mental exertion, accompanied by a wide range of other symptoms. The fatigue is commonly described as like no other in type and severity, and is evidently very different from everyday tiredness.

Perhaps the prime indicator of the condition is the way in which symptoms behave after activity is increased beyond what the patient can tolerate. Such activity, whether physical or mental, has a characteristically 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. The amount of activity that provokes increased symptoms is related to the severity of the disorder, and in some individuals is very modest. Delayed fatigue, postexertional malaise, or increase in other symptoms after activity can be helpful to make a diagnosis. However, the consequent variability in functional disability can make adaptation to the illness very difficult for the patient, and can be confusing to those around the patient or who assess them.

Characteristic or 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 also almost invariably seen as part of the picture of CFS/ME. These forms of fatigue, both physical and mental, need to be explored in the same way, as, for example, the varying characteristics and experience of pain.

Cognitive impairment – In addition to general cognitive fatigue, other difficulties include 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 – Malaise after exertion may comprise 'flu-like symptoms or other constitutional features.

Pain – This is typically persistent and often difficult to alleviate with standard analgesia. Pain may be muscular, rheumatic, neuropathic (with or without parasthesiae), head pain or headache (often migraine-like).

Sleep disturbance – This takes many forms, such as early wakening, insomnia, hypersomnia, and disrupted sleep/wake cycle.

Other symptoms – 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. In a minority of severely affected patients, serious neurological symptoms include: double vision, blackouts, atypical convulsions, loss of speech, and loss of swallowing necessitating nasogastric feeding.

Recurrent sore throat – with or without lymphadenopathy.

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

Intolerances – Intolerance or sensitivity/altered tolerance are common. Alcohol intolerance is very common, and many patients are intolerant of some foods, some medications (especially psychotropic medication), or other substances (sometimes called "multiple chemical sensitivity").

4.2.1.3 Onset and course

The onset of CFS/ME can be sudden or gradual. In cases of sudden onset, the condition commonly follows an acute infective episode. These infections are typically viral, but CFS/ME has also been reported after other infections. Patients with gradual onset disease often have an episodic course, or a stepwise decline. An insidious and gradually progressive course is uncommon. Antecedents or triggers are harder to identify in patients with less acute onset of disease, but other factors have been associated with onset of CFS/ME in individual cases, including: some immunisations (possibly during intercurrent illness), surgery, chemotherapy, and exposure to some chemical agents (e.g. organophosphorus compounds). The importance of such factors in the population burden of CFS/ME is as yet unclear.

Many patients report that they attempt at first to keep going with usual activities, or to return to work or education before being fully recovered, and then are repeatedly or progressively unable to sustain previous levels of activity. Others have had additional physical or psychological stressors around the time of the onset. Factors relating to the development of CFS/ME have been considered as including predisposing, triggering, and maintaining factors (see **Chapter 3**), which is conceptually helpful, but is fraught with problems of interpretation and attribution at the level of the individual.

4.2.1.4 Predictors of chronicity

Little is known about the reasons for variations in prognosis, though research has indicated several factors associated with prolonged disease. Severity of illness is a major factor, with a tendency for illness to persist longer in more severely affected patients. However, the research evidence, while sometimes appearing convincing, is not conclusive, because most studies share several faults. Firstly, most are uncorroborated by other studies. Secondly, although they may have speculated about causation, mostly what has been demonstrated is an association. For example, the various psychological factors claimed to be causal may be a consequence of severe, prolonged CFS/ME, and for the most part the study designs adopted would not enable the question of causality to be resolved. Thirdly, definitions of severity have varied widely, and in some cases are quite vague. Few attempts have been made to quantify severity in ways that are reproducible, using validated instruments. The role of various demographic factors and blood markers (e.g. immunological) is not yet clear from the literature and needs further investigation.

Individuals with acute onset illness appear to have a better prognosis than those with gradual onset, and epidemic or 'clustered' disease has a better prognosis than sporadic disease. In children, as among adults, the prognosis appears worse for more severe cases. Overall, the duration of disease appears shorter in younger people than among adults, and a high proportion of children appear to recover. Recovery may rarely be complete (**Chapter 1**, **section 1.4.3**), and some children will relapse in adult life.

Other factors that appear to be associated with poor prognosis include:

- the coexistence of psychiatric and other chronic illnesses with CFS/ME;
- a long duration of symptoms of CFS/ME;
- older age.

4.2.1.5 Timescale

Research criteria for CFS/ME stipulate that a diagnosis can be made only after the presence for six months of a cluster of symptoms. However, in clinical practice, six months should be viewed as an endpoint for the diagnostic process, as patients will need help to manage the illness much before then. An approximate timing of the diagnostic pathway for adults might be:

Six weeks – At about six weeks after the onset of symptoms that include excessive tiredness, fatigue, or malaise, a differential diagnosis is considered that includes acute fatigue syndrome. Appropriate history, clinical examination, and investigations are done to exclude other illnesses. As with any potential chronic illness, discussion with the patient includes management of symptoms and the impact of illness on daily living.

Three months – At three months, in patients whose symptoms persist and for whom other causes of the symptoms have been excluded through careful history taking, examination, and investigation, a provisional diagnosis of CFS/ME might be made. What justifies and determines the future course of action is the clinical presentation, rather than the application of time-based definitions derived for epidemiological research purposes.

In common with many other chronic illnesses, the diagnosis may be pieced together over a period of time through a series of consultations. The resulting discussions with the patient should be seen as the beginning of a positive therapeutic relationship in which sharing of information is part of the process of helping the patient to manage the impact of symptoms.

Six months – By six months, if symptoms persist, the provisional diagnosis should have been confirmed. But the plan for managing the early stages of the illness and its consequences will already have been put in place.

4.2.2 Clinical evaluation

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

that are not typical of CFS/ME and newly arising symptoms can be considered separately. In general, the following steps are involved (see **Annex 6**):

Full clinical history – At present, this is crucial diagnostic procedure for CFS/ME, and sufficient time should be allowed for patients to give a narrative account of their illness experience, its broader impact, and the extent of their current disability against the background of the patients previous levels of functioning.

Sleep evaluation – Exclusion of primary sleep disorders in people with unexplained fatigue is important, especially if drowsiness or day-time somnolence are prominent.

Mental health evaluation – Assessment for mental health problems including mental state assessment and psychosocial assessment at an early stage is important, so that any such problems can be either dealt with or excluded. The clinician should recognise that the patient may feel sensitive about this kind of assessment.

Physical examination – The physical examination is essential and may be helpful in excluding other conditions.

Basic screening tests – These include full blood count, C-reactive protein (CRP) concentration, blood biochemistry, thyroid function tests and urinalysis.

Specialised tests – These may be required to exclude differential diagnoses that are suggested by particular symptom patterns, or abnormal findings on physical examination or investigation (e.g. blood markers of rheumatic diseases or antibodies to gliadin or endomysium to identify coeliac disease). Tests used in research, such as specialist neuroimaging, do not currently seem necessary as part of routine care.

4.2.2.1 Specialist referrals

In many cases, much, if not all, of the initial clinical evaluation and diagnostic process can be satisfactorily undertaken by the primary care team. A general practitioner should be able to make a firm diagnosis of CFS/ME in most instances among adult patients. However, there is a proportion of cases in which referral to a specialist experienced in CFS/ME may be useful in confirming a diagnosis, or where complex issues or symptom patterns give rise to uncertainty. In some instances, a patient may request a second opinion.

Specialist referral would usually be to a local consultant with an interest and expertise in CFS/ME. Patients may also need to see other specialists if consideration of differential diagnoses requires assessment. Sufficient tertiary specialists in CFS/ME are needed to support primary and secondary care for the most difficult clinical problems, and to act as a resource for teaching, training, and research. Currently, there are too few identified and resourced specialists at secondary and tertiary level in much of the country, and the few that exist are overburdened; they are also inappropriate for care of the most severely affected, who cannot travel, or even less severely affected patients, for whom travel over long distances is liable to cause setbacks.

Children should usually be at least known to community paediatric services, and many should be referred to a paediatrician to confirm both the diagnosis and because of the impact of the illness on their education and their social relationships (see **Chapter 5** and **Annex 7**).

Education and support, plus measures to tackle the broader impact of the disease, should be initiated as early as practicable in management. This will often include assistance on how to contact other services, including in the voluntary sector.

4.3 Information and support

For patients, carers, and clinicians, CFS/ME presents many layers and types of uncertainty. Yet human beings cope poorly with uncertainty, which can undermine personal and professional responses to the illness and its impact. Acknowledgement of this impact and strategies to help patients and their carers cope with uncertainty are an essential part of management. Essential components include: helping patients to come to terms with the illness, coping with uncertainty, and developing a partnership with the patient to ensure that activities are carried out at a steady and regular pace. A sympathetic approach and understanding of the range of uncertainties is crucial.

4.3.1 Information

Patients with CFS/ME and their families/carers may have diverse information needs, although all need clear clinical communication on options, progress, and prognosis, as a minimum. Information on the nature of the condition and self management seems to facilitate adjustment to the illness, and a better outcome. Such education is also particularly important for anticipating and managing fluctuations or more substantial remissions and relapses. Good-quality communication will also be needed on behalf of the patient with employers, schools, universities, benefits agencies, private health insurers, pensions and health-insurance schemes, social services, and so on, as well as between other professionals involved in care.

Several charities and voluntary organisations offer information, training, education, and support as well as investment in research on CFS/ME. Patients may need advice on how to access support from these and other agencies.

Patients' questions that clinicians can answer

- Initially, whether they are ill and what their illness is.
- Is it their fault? Are they getting old, going mad, developing Alzheimer's disease, etc?
- Will it get worse? If they improve, will they relapse?
- Which treatments are worth trying?
- How will others react?
- To whom can they safely talk about how they feel?
- What will the impact of their illness be on them, their work, or their friends and family relationships?
- How will they cope financially? Can they work?

4.3.2 Self management

Research in chronic disease management consistently shows that a key component to the successful management of a long-term illness is involvement of the patient as a partner in care, whatever their age. An approach of taking patients' views into consideration leads to higher satisfaction, better compliance with treatment, and greater continuity of care. A partnership approach to management acknowledges that the patient must continue to cope with their illness throughout, rather than just during intermittent clinical interventions.

Important approaches to enabling people with long-term conditions to act as partners in the management of their care are: education on obtaining information, with the aim of the patient being an expert in their condition; and education on self management.

Such education can be provided by clinicians, appropriate therapists, charities and voluntary organisations, and through other resources – e.g. self management programmes, which are delivered by trained and accredited lay people who have had the long-term condition.

4.3.3 Equipment and practical assistance

In many chronic illnesses, daily functioning, including mobility, cooking, cleaning, dressing, personal care, and social support, can be improved dramatically by sympathetic provision of appropriate practical assistance. Even simple equipment may be valuable for increasing independence (e.g. rails, bathing aids, kitchen adaptations), particularly for those severely affected. Wheelchairs and other mobility aids can be particularly useful for some people affected by CFS/ME, to increase the physical range and mobility of disabled patients, enabling them to do or participate in more things, and see people who would otherwise be inaccessible. Experience suggests that provision of a wheelchair or other mobility aid does not stop patients working towards mobility without the equipment in the long term; indeed such aids probably assist remobilisation, with suitable supervision e.g. from a physiotherapist.

An assessment by social services is needed when there may be a need for social care provision. This might include: home care, general support, short breaks for carers, or direct payments for patients to purchase their own services where this is appropriate. Careful discussion with the patient, carers, and the rest of the team supporting them will be needed to determine the nature of the services required. Occupational therapists may also need to assess for the longer term provision of equipment for daily living where the person is not able to undertake such tasks without assistance. More complex equipment or adaptations to the home – e.g. stairlifts – would only be provided where the person has a substantial disability that is likely to continue for at least six months, which may be difficult to determine with a fluctuating condition. The prognosis given by the medical practitioner will be crucial in such cases and will need to be realistic without disheartening the patient.

4.3.4 Socioeconomic support

Chapter 3 sets out the impact of the illness on finance, work, and education. Amelioration of this impact is an important aspect of clinical management, and it should be considered as early as practicable. **Chapter 5** contains further material on educational support for children and young people.

Support from clinicians is needed for the provision of medical reports and assistance with negotiations with the Benefits Agency, employers, educational institutions, and insurance companies. Medical advisers for the Benefits Agency need to be aware of the incapacity experienced by CFS/ME patients. This has been highlighted in detail in our earlier paper (see chapter 1.3) which has been passed on to the Chief Medical Advisor to the DWP.

4.3.5 Support to family/carers

For carers and family of those affected, life is turned "upside down". They will need support and advice on ways in which to cope with their loved ones' illness. Voluntary organisations and local carers groups can be helpful in this respect. Clinicians may need to advise on various matters.

Carers' questions that clinicians can answer

- Will they need to leave their jobs?
- How long will the illness last, and how disabled will their loved one be during this time?
- What can the carer do, and what should they not do?
- Will their doctors believe them?
- Where can they get advice about benefits?

4.4 Ongoing care

Once a diagnosis of CFS/ME has been made, ongoing care is subject to the same principles and approach as previously detailed. In particular, information and support is needed as patients and carers continue to cope with an evolving illness. Treatment should always be a collaboration between the patient and the clinician, and not something imposed. Good communication and a good therapeutic relationship can make an appreciable difference to the response of the patient who feels the need to be understood and listened to empathetically. For example, there are many techniques for self help which clinicians can make easier with guidance and support[1]. Each patient needs repeated assessment of his or her illness to guide individual adaptation of management strategies from a generically applicable range of options.

A key question the Working Group sought to answer was whether any specific approaches are effective in management of CFS/ME. In consultation with the Key Group, the Department of Health for England commissioned an assessment of the available evidence on the effectiveness of treatments.[2] The report highlighted the paucity of good-quality evidence. The Key and Children's Groups found the report to be a good review of available evidence from randomised trials, but some members were concerned that the review was limited to quantitative studies rather than also including qualitative studies. Submissions to the Working Group identified other approaches as helpful, though these lacked research evidence. Moreover, some patients and practitioners reported that evidence-based strategies could be detrimental when applied inappropriately or inflexibly.

4.4.1 People who are severely affected

Not enough is known about severe forms of the condition CFS/ME that are reported to affect up to 25% of patients. Severe disease has profound effects on health, social functioning, and all other aspects of life for such patients and their carers. These patients suffer from additional problems of invisibility, barriers to accessing all forms of care, variable responses to treatments, and under-representation in research. The Working Group is very aware, from evidence gathered through the Sounding Board events and surveys undertaken by voluntary organisations, that provision of health care for these severely affected patients is often seriously inadequate. However, we found insufficient evidence available to guide specific management of those people who are severely affected.

Severely affected patients report different responses to management strategies applied with some success to other individuals with CFS/ME. However, the Working Group found insufficient evidence to clarify whether this difference in responses was quantitative – due to the severity of the individual's illness and the lack of adaptation of programmes – or qualitative – representing some definable difference in the way management is applied or how an individual responds. In general, a rehabilitative approach has not been researched for the patients most severely affected. Any such approach must therefore be adopted with caution in this group. It seems best on present evidence to recognise the need to adapt therapies to the functional level of the patient, and to adjust them further in response to feedback from the patient during therapy.

Care of people who are severely affected is an urgent challenge that must be addressed in appropriate and imaginative ways, drawing from service models applied to other severe chronic disabilities. Healthcare and social service professionals are responsible for finding ways of supporting and guiding patients and their carers for the duration of illness, ensuring access to available support, keeping in contact, constantly re-evaluating the options, maintaining morale, enabling respite, and minimising consequences of prolonged disease. Patients with severe or prolonged illness will usually need support in their homes and communities. Thus, local domiciliary services linked to existing primary and community-care structures are key, although such services can be informed and supported at a distance by more specialist services.

We suggest that the prevalence and impact of severe disease, the pathways to chronicity and to becoming severely affected, and strategies that would benefit such individuals urgently need further study. Moreover, this needs to be kept in mind by clinicians when devising a management plan with someone who is severely affected. However, such constraints are not an excuse for failing to guide and support individuals with severe disease and/or disability. Further special considerations for those severely affected are noted in the relevant sections of this chapter. Care of severely affected patients should be developed with full acknowledgement of these special considerations and other particular circumstances noted in **chapters 2** and **3**.

4.4.1.1 Response to treatment

Some patients with CFS/ME might not respond, or might even respond adversely, to certain treatments found effective in other patients. For example, most intervention studies have examined people who are sufficiently mobile to attend services

for repeated treatments/assessments. When such treatments are applied to more severely affected patients, some have noted adverse effects. This discrepancy might be explained by a failure to adapt the therapy sufficiently to the severely affected patient's very different level of functioning or because the treatment is simply inappropriate.

In view of concerns raised over patients' experience, the question of whether differential treatment response represents some distinct difference in disease merits carefully planned research. Since some patients with particular features (e.g. severity of physical disability) will have intrinsic limitations in their ability to participate, care must be taken in study design to overcome the hazard of selection bias. Future studies will need to control for possible differences in treatment response due to age, gender, ethnicity, severity, duration, triggering event, coexistent conditions, and symptom profile.

4.4.2 Therapeutic Strategies

The Working Group agreed that there is no cure for CFS/ME but identified three specific strategies as potentially beneficial in modifying the illness: graded exercise, cognitive behavioural therapy, and pacing. Members of the Working Group expressed widely differing opinions on the potential benefits and disadvantages of these approaches. However, we agreed that all could be considered as management options, in line with general principles outlined here, and adapted to the circumstances of each individual patient. Clinical wisdom suggests that management of limited energy and supervision of any increases in physical or mental activity are an essential part of ongoing care for individuals with CFS/ME. The Group also found it important for clinicians to use the pharmacological and non-pharmacological means available to relieve disabling symptoms. Patient experience suggests that some complementary therapies can also play a role in this respect.

Often, the most essential aspect of continuing care will be for clinicians to provide advice on appropriate ways of relieving symptoms, balancing rest and activity, and maximising potential. In primary care, this might involve advice on pacing, prescription medication, or basic lifestyle management counselling. A proportion of patients benefit from more structured specialist approaches, such as graded exercise or cognitive behavioural therapy.

The success of any specific approach is dependent upon many factors, not least the way in which the approach is applied. In cases where an approach does not succeed or is found harmful, it is important to distinguish those where the approach is not appropriate for the individual from those where the approach is inappropriately or poorly applied. In addition, the Working Group note the following general principles that govern good practice:

- The decision to recommend a particular approach is best guided by the individual's illness and circumstances.
- The content and development of any such approach should be mutually agreed by both clinician and patient and informed by up-to-date specialist knowledge.
- Ideally, a decision to refer from primary care would be mutually agreed and guided by the degree of uncertainty surrounding the patient's illness or its management.

- Specialist therapies (e.g. graded exercise and cognitive behavioural therapy) are likely to be most effective when supervised and regularly monitored by therapists who have appropriate training and experience.
- All interventions need to be administered with thought and care and in accordance with revised Department of Health recommendations on informed consent.[3]
- It is not appropriate that participation in a particular treatment regimen is made an absolute condition for continuation of sickness/disability payments.

The Working Group deemed it helpful to highlight specific therapies that had reasonable evidence of effectiveness. Given the prevailing uncertainty surrounding the condition and its therapies and the limitations of the existing range of research highlighted in the York review, we recognised that the evidence we sought needed to encompass both the precision of findings from rigorous randomised controlled trials and the breadth of clinical experience and patient reports. To do this, we used a trident approach to review and synthesise three lines of evidence: research findings, patient reports, and clinical opinion. We also considered resource implications, although cost-effectiveness is the least studied of all aspects of CFS/ME management.

4.4.2.1 Graded exercise

As a general principle, the Working Group agreed that both activity and rest can be harmful when overdone and yet be beneficial when carried out with the appropriate degree of balance. Graded exercise is a form of structured and supervised activity management that aims for gradual but progressive increases in aerobic activities such as walking or swimming. It is based on a principle – contested by some – that a principal factor maintaining the illness is inactivity, subsequent physical deconditioning, and its physiological consequences, which graded and supervised increases in exercise can help to reverse. In addition, it may act as a rehabilitative behavioural therapy by gradually exposing the patient to an activity (exercise) that has been avoided. Gradual, supervised exposure within the individual's limits is thought also to help improve confidence in physical ability.

One key controversy that exists over graded exercise rests on whether the nature of the treatment is appropriate for the nature of the disease, at least in some individuals. Existing concerns from voluntary organisations and some clinicians include the view that patients have a primary disease process that is not responsive to or could progress with graded exercise, and that some individuals are already functioning at or very near maximum levels of activity.

Research findings – The York review found promising results for graded exercise: all three randomised controlled trials so far found varying degrees of improvement in fatigue and disability with differing graded exercise regimens compared with no treatment and two control treatments. These trials all scored highly in the validity assessment, although, as with most clinical trials, the findings encompass only the range of patients able to meet the entry criteria. The York review found that people who were unable to attend outpatient clinics were excluded from these and other trials of treatment effectiveness.

In the trials, very few participants reported feeling worse with graded exercise, although the drop-out rate was just under a third in one of the trials – thought to be related to demands of the programme.

No randomised, controlled trials of graded exercise have been conducted in patients unable to attend outpatient clinics or in children. Several open studies suggest that graded exercise can be helpful to improve disability in more severely affected patients, so long as the treatment is carefully planned, regularly reviewed, and mutually agreed with the patient: however these studies lack the rigour needed to make/allow definitive comment on the role of this approach in severely affected patients as pointed out in the York review.

Patient reports – Voluntary organisations, as well as the Sounding Board events, note that graded exercise therapy can be effective in some individuals, but substantial concerns exist regarding the potential for harm, particularly when such therapy is applied inflexibly or without mutual agreement with the patient. The non random survey of people who were severely affected found that out of 1214 who had tried graded exercise, 417 believed it was "helpful", 187 reported "no change", and 610 believed it had made their condition "worse"(see also **Annex 3**, **section 3**). Similar adverse comments were also reported in patient group survey results from less severely affected patients, and no other treatment – pharmacological or non-pharmacological – received such negative feedback in patient surveys.

Clinical opinion – As with pacing, there is disagreement among clinicians about the value of graded exercise. Some clinicians consider graded exercise an effective therapy because of the evidence base, whereas others believe that CFS/ME involves a primary disease process that is not responsive to this type of approach, and that many of their patients are already functioning at or near to maximum levels of activity. However, the Working Group did agree that whenever graded exercise is being undertaken, activity levels should be initially based on current physical capacity. The programme should be mutually agreed between patient and therapist, it should be regularly adapted according to the clinical response, and patients should be carefully monitored to ensure that exertion does not exceed target levels.

Resource implications – Best practice in this area indicates that the initial stages of any graded exercise programme should only be carried out by therapists (i.e. occupational therapists, physiotherapists, exercise physiologists, sports therapists, etc) who have the necessary expertise to manage CFS/ME patients. At present, very few therapists are available with such expertise.

Synthesis – The majority of the Working Group agreed that appropriately supervised, graded exercise therapy, applied by appropriately trained individuals, can benefit many, though not all, ambulant outpatients with CFS/ME.

A successful outcome probably depends on the therapy being initially based on current physical capacity, mutually agreed between the therapist and patient, and adapted according to the clinical response. Appropriate education regarding the rationale and cautions of this therapy needs to be given to potential candidates for graded exercise. Patients who drop out of therapy need to be followed up swiftly to review the reasons and reassess their management plan.

The place of this therapy for more severely affected patients is currently uncertain, but a suitably modified (initially low intensity) exercise or activity programme may reverse the adverse consequences of pervasive inactivity, if this is perceived to be a key factor in the individual's illness.

4.4.2.2 Cognitive behavioural therapy

Although there is no cure for CFS/ME, the condition has been found to improve in most patients both with and without treatment; it is good practice to encourage patients to become experts in self-management and to choose between treatment options.

Cognitive behavioural therapy is known to be helpful to some patients with physical and psychiatric illnesses to improve quality of life and day-to-day functioning. Re-enablement based on the cognitive behavioural model aims to empower patients to identify, understand, and modify their belief systems and behaviours, to maximise their own functioning and well-being with support and guidance from the therapist. It involves personal actions – i.e. "what we do" and "what we think" – that can affect physiological processes; for example, smoking, excessive alcohol intake, and stress can all contribute to illness. Application of a cognitive behavioural model to CFS/ME has been found successful in most patients in the trials.

It is important to note that a specific or shared belief system is not essential to apply the principles of cognitive behavioural therapy to CFS/ME. However, the wider uncertainty surrounding the nature of CFS/ME does impact on perceptions and delivery of the therapy in individuals. Difficulties can also arise when therapist and patient share differing beliefs about the individual's illness, and the nature of CFS/ME. Patients may have an understandable apprehension about increasing activity, so it is important that changes are mutually agreed and the patient is supported through the process.

The specific model and components of cognitive behavioural therapy vary between services, disciplines, and between therapists. The core components of a cognitive behavioural approach to CFS/ME would include energy/activity management, establishment of a sleep routine, goal setting, and psychological support. The general principles of this holistic approach can be administered by a range of therapists and nurses with experience of CFS/ME, provided they have had appropriate training in the techniques.

Research findings – The York review found that cognitive behavioural therapy showed positive results in adults able to attend outpatient clinics. Three of the four randomised, controlled trials evaluating this therapy found a positive overall effect of the intervention, with the majority of those who had the therapy demonstrating varying degrees of improvement in both function and fatigue. These studies scored highly on validity assessment in the systematic review.

Few patients reported feeling worse after treatment in the trials, but few reported complete recovery. The best results seem to be obtained by therapists with knowledge of CFS/ME. In one trial, global improvements were maintained after five years; however, there was no difference (between intervention and control groups) in fatigue, physical functioning, and other measures. Few adverse events were

reported from the trials. Drop-out rates varied; one trial had drop-out rates in all three study groups (including the control group), with a 20% rate in the group of patients assigned cognitive behavioural therapy, who started therapy after randomisation. Other trials had low drop-out rates of around 10% in both study groups.

There have been no published randomised controlled trials of cognitive behavioural therapy for children or for the severely affected, although open studies suggest it may be helpful for the latter if applied appropriately.

Patient reports – These suggest wide variation in both the practice of and the individual response to cognitive behavioural therapy. Although there is general acceptance that the therapy can help some patients, some comments point to difficulties with inflexibility in the therapists' views or in the treatment plans. Some patients are reluctant to receive what they perceive to be a "psychological treatment" for a "physical" disorder. A persistent concern is that cognitive behavioural therapy is viewed by some clinicians as the sole proven treatment strategy. A further observation was that services are often unavailable locally or available only after a long wait.

In one patient-group survey, only 7% of respondents found the therapy "helpful", compared with 26% who believed it made them "worse". The remaining 67% reported "no change" (see also **Annex 3**, **section 3**). Harm was suggested by the report to occur if activity scheduling was too rigid, if the therapist displayed scepticism of the patient's views or experiences, or if they implied that their illness was "all in the mind". Patient reports indicate that patients find a holistic, practical, occupational therapy based service to be an acceptable approach. While mental health workers and therapists from other disciplines may also offer acceptable services, it has been suggested that therapists accustomed to working solely with psychiatric populations may need additional training and experience to engage and work appropriately with patients suffering from CFS/ME.

Clinical opinion – There was disagreement among clinicians as to the precise value and place of cognitive behavioural therapy, which partially reflected the varying models of the therapy and disease. However, there was agreement that when applied appropriately, with mutually agreed approaches and goals, it can undoubtedly benefit some patients. Some clinicians, using trial evidence and clinical experience, feel that it is beneficial to the majority of patients, whereas others feel it only benefits a minority. We also noted that misunderstanding, misplaced concern, and poor practice in this area could potentially undermine the beneficial application of this therapy or its principles in patients with CFS/ME.

Resource implications – Outside specialist referral centres, it is currently difficult to find therapists with the necessary experience.

Synthesis – The majority of the Working Group accepts that appropriately administered cognitive behavioural therapy can improve functioning in many ambulatory patients with CFS/ME who attend adult outpatient clinics. Preferably, the therapist should have experience in CFS/ME or have some training in this field. Patients who might benefit can expect to receive a logical explanation of why cognitive behavioural therapy might help them, based on their specific history and general principles. Where such services are not available or the patient decides against a trial of therapy, patients might be managed by usual clinicians, using the principles of and informed by the practice of cognitive behavioural therapy with other approaches.

Cognitive behavioural therapy for people with CFS/ME is currently unavailable or very difficult to obtain in much of the UK. Local expertise would be increased by the training of more cognitive behavioural therapists and by increased education on and experience of CFS/ME for existing therapists. The value of a trained therapist who has a good understanding of CFS/ME is that they are competent to raise concerns about the way the patient handles their illness, so the patient is empowered to manage their illness and its consequences more effectively. Good practice involves a flexible approach which encourages useful change, undertaken with mutual respect between therapist, the patient, and their family or carers.

Whether cognitive behavioural therapy is useful for children or adolescents is not known, although some clinicians believe that selected patients may benefit from a trial of the therapy by appropriately trained therapists. The place of the therapy for patients more or less severely affected than those who participated in research is currently uncertain.

Further research is needed to identify which CFS/ME patients derive most benefit from the therapy as well as trials that compare cognitive behavioural therapy to other rehabilitative approaches (pacing, graded exercise, etc), to delineate essential aspects of successful rehabilitation. Findings of such research would assist decisions on referral – e.g. on whether priority needs to be given to those patients who are experiencing obvious difficulties in coping with either the emotional, social, or activity management of their illness.

4.4.2.3 Pacing

'Pacing' is an energy management strategy in which patients are encouraged to achieve an appropriate balance between rest and activity. This usually involves living within physical and mental limitations imposed by the illness, and avoiding activities to a degree that exacerbates symptoms or interspersing activity with periods of rest. The aim is to prevent patients entering a vicious circle of overactivity and setbacks, while assisting them to set realistic goals for increasing activity when appropriate. Although the research evidence base for this therapy is very limited, many voluntary organisations and a proportion of clinicians consider that pacing has an important place to play in the management of CFS/ME.

Pacing is based on the 'envelope' or 'glass ceiling' theories of CFS/ME, which suggest that energy is finite and limited, and that the best way for a patient to manage their illness is to live within this envelope – i.e. not constantly breaking through the ceiling (some therapists advise never going beyond 70% of a patient's perceived energy limit). The underlying hypothesis is that if patients use their energy wisely, their limited energy will increase gradually. The therapy involves daily monitoring of energy and activity levels, reviewing the effects, and making appropriate adjustments. An individual approach is planned after a full assessment of previous healthy functioning and an agreement of the baseline activity possible for the individual. Little evidence exists to indicate harmful effects, although clinical wisdom suggests that the strategy is not universally appropriate and some clinicians consider that pacing may perpetuate illness. The underlying principle might be viewed as being somewhat contradictory to the underlying principles of more active rehabilitative strategies.

Pacing accommodates various stages (acute, stabilisation/transition, and recovery) and degrees of severity (mild, moderate, severe) reported by patients with CFS/ME. During an acute phase, appropriate rest then convalescence is advised. The first goal of subsequent stabilisation or transition phase(s) is to establish a baseline of sustainable activity, from which gradual stepwise increases in activity are encouraged. The individual is assisted to find an appropriate balance between various physical, mental, and emotional activities, and to review and adapt their activity schedule if setbacks occur. For patients who enter a recovery phase, the principles of pacing can be applied to a return to work or education. For those who remain severely unwell and make no substantial progress, pacing therapy may also involve passive physiotherapy and other measures that can help prevent complications of prolonged immobility. The principles and practice of pacing are detailed in the 1994 Task Force report.[4]

Research findings – Research on pacing is sparse. One controlled trial found that six sessions of pacing therapy were no more helpful than "guided support" in helping fatigue, depression, and symptom scores. This non-randomised study had several limitations and is unlikely to be representative.

Patient reports – Considerable support exists for pacing among patients and voluntary organisations, particularly for those who are more severely affected. A survey of more than 2,000 members of a voluntary organisation who were or had been severely unwell showed that 89% of group members found pacing "helpful" (**Annex 3**, **section 3**). Similar findings are reported from surveys of less severely affected patients. Voluntary organisations believe that sufficient periods of rest are particularly beneficial in the early stages of the illness.

Clinical opinion – Disagreement exists among clinicians who treat patients with CFS/ME over the value of pacing. Many clinicians, including some in the Working Group, routinely recommend the approach while others are less convinced of its benefits, their experience suggesting that pacing may prolong a patient's illness. The Working Group noted that disagreement also exists over what is included in the term "pacing".

Resource implications – Advice to patients about pacing principles involves few additional resources. The basic principles are readily available in lay language from the voluntary sector.

Synthesis – Despite the lack of research on pacing, the Working Group recognises that this form of energy management is popular with patients, voluntary organisations, and some clinicians. The Working Group notes some general principles that may assist clinicians to help some patients manage their energy (see **Annex 6**, **section 5**). The principles of, and tools used in, pacing, as well as those of the more active strategies, can be incorporated into a care plan for CFS/ME patients in both primary and secondary care.

Because of the shortage of good research evidence of the effectiveness of pacing, there is an urgent need for randomised controlled trials of pacing therapy, particularly in early illness (for example, in comparison with rehabilitation therapies such as cognitive behavioural therapy and/or graded exercise, and other forms of support such as counselling).

4.4.3 The use of counselling

Counselling describes both a skill used by clinicians in their daily work and a structured form of therapy. The principle is to create a supportive environment by the way in which the practitioner relates and responds to the patient, to provide them with the opportunity to explore, clarify, and make progress on personal issues with the goal of increasing resourcefulness towards improving wellbeing. Clinical wisdom suggests that some form of counselling on coping with a longterm illness is an important part of the ongoing approach to management of CFS/ME. Clinicians and patients have sometimes been disappointed by the lack of guidance and the non-directiveness from some counselling styles.

Further research is warranted in the form of a larger, randomised, controlled trial to examine the possible benefits of counselling compared with other rehabilitative approaches in patients who have CFS/ME.

4.4.4 Symptom control

Patients with CFS/ME characteristically have many symptoms. Although some of these may be tolerable with explanation and reassurance, some symptoms are intrusive and unpleasant. Moreover, some may act to cause a descending cycle, exacerbate the impact of the illness, and impede recovery and/or adaptation. Sleep, mood disturbance, and pain are notable in this regard, because they are common and have substantial impact, yet are often treatable. In some women, pre-menstrual or menstrual exacerbation of symptoms is reported as is use of hormone replacement in those patients with low levels of serum oestradiol. As part of the diagnostic process, each symptom should be carefully assessed and, where appropriate, further investigation should be undertaken to characterise the process and rule out other illness.

Clinicians can access their usual “toolkit” of symptomatic measures where appropriate, adapted to patient need. Substantial efforts should be made to specifically elicit and manage difficulties with pain, sleep, and mood, not least because they are common and without treatment may compound each other. Management of more intrusive symptoms could involve advice on and support for adaptations in behaviour or diet, for example, or the use of medication(s) to treat or to prevent the symptom(s). In many cases, the clinician will consider approaches that are used when the same symptoms occur in other disease settings. The options, with their rationale, advantages, and possible side-effects, should be explained. This enables the patient and/or carers to decide whether or not they would like to try one or more such approaches and, if so, when. They will generally be quite clear about whether symptoms are sufficiently intrusive and/or frequent to justify symptom-control strategies. 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 will need to be adapted to CFS/ME and refined based on 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 choices are available. If the initial approach does not succeed or is not tolerated,

variations or alternatives can be attempted after discussion and agreement. If intolerance to medication is a major difficulty for the individual, other strategies are worth exploring with the assistance of specialist therapists, as appropriate.

4.4.5 Complementary approaches

Complementary approaches are popular with patients. The Sounding Board events and surveys undertaken by the voluntary organisations indicate that patients report benefit from several different therapies, partly owing to the perceived approach of the practitioners (see **Chapter 2**). However, the limited research evidence is acknowledged by voluntary organisations, and patients participating in the Sounding Board events expressed concerns about some complementary practitioners who make unrealistic claims of success, as do other orthodox clinicians. Charges for complementary approaches can also be prohibitive for patients who experience financial difficulties.

Clinicians may feel they lack the knowledge to advise patients on complementary therapies. Such therapies do impact – positively and negatively – on health and clinical care, so clinicians should be aware of their use by the individual. The patient can be advised to adopt a similar approach to complementary therapies to that they would adopt for symptom control.

4.4.6 Follow-up, transitions, and recovery

As for other patients with chronic disease or ongoing disability, regular follow-up to guide and monitor the patient is important, especially in primary care. Apart from the therapeutic benefit of maintaining contact and dialogue, it provides the opportunity to refine and adjust the therapeutic approach, assess new or changing symptoms or alterations in functional capacity, and advise on the wider repercussions and on self management. Unfortunately, some patients show little or no response to existing treatment options, and may show no improvement over long periods; while both patient and clinician should continue to review possible ways of improving their situation, the patient must not be made to feel that they are to blame for the lack of response, nor forced into therapies that are inappropriate, unwanted, or ineffective for that individual.

As with many chronic illnesses, the fluctuating nature of CFS/ME means that remissions, setbacks or more substantial relapses may occur. Education about the expected nature of the illness can mitigate some of the impact of setbacks. However, improvements can also create uncertainty and other difficulties, as can other transitions in illness, including moving services. Any transition may require increased input from services – e.g. from childhood to adolescence to adulthood, starting or stopping work. A transition represents an opportunity to review the management plan with patient and carers. The nature of the transition may make re-evaluation a necessity – e.g. during an apparent relapse or a move from paediatric to adult services.

A gradual and mutually negotiated return to work or education can improve outcome. Thus, part of the therapeutic package to be considered for both adults and young people should be a mutually agreed and gradual return to previous activities including work and education. This should be, supported by appropriate negotiations with educational institutions, businesses, insurance companies, and

the Department of Social Security and the Benefits Agency, all of whose medical advisers need to be aware of and sympathetic to the incapacity experienced by CFS/ME patients.

Negotiations with insurance companies and the Department of Social Security about proportional and rehabilitation benefits and therapeutic work can improve outcomes, and health professionals have an important role to play by providing support and advice in these negotiations. The same level of understanding needs to be shown by medical advisors to insurance companies and the Benefits Agency about the condition, its natural course, prognosis, and range of available approaches to recovery.

4.5 Service models

Provision of services specifically designed for patients with CFS/ME is either limited or non-existent. While patients have access to the normal range of primary, secondary, and tertiary care services, few are specifically tailored and staffed with appropriate expertise to meet the specific needs of this patient group. Specialist services for children and young people, including inpatient facilities, are limited to a few nationwide. The Working Group confirmed deficits reported by patients and support organisations by inquiries to Regional Offices. A substantial number of patients are referred from primary care for a consultant opinion to one or more of several system-based specialists (general physicians, immunologists, neurologists, haematologists, and psychiatrists). Referrals also occur to tertiary centres for assessment and management.

Where the limited number of services have developed, this has been due to either the enthusiasm of clinicians who have been instrumental in “championing” the development of NHS services or the involvement of the charitable/voluntary sector.

The lack of locally based services is a problem to both patients who need a service and to commissioners of health services who wish to reduce the cost of out-of-area treatments.

4.5.1 Service need

There is insufficient good-quality evidence available to guide precise estimates of service need (see **Annexes 1** and **2**). However, on the basis of a reasonable estimate of adult population prevalence of 0.4%, a general practice with a population of 10,000 patients is likely to have 30 – 40 patients with CFS/ME, about half of whom may need input from services. The proportion of the latter patients who are severely affected by the disease is thought to be up to 25%. This group, who may be house-bound or bed-bound, have a considerable level of need yet they face considerable barriers to accessing services.

The burden of illness and service needs arising for children and young people are less clear. Much of the burden of care may rest on existing child health services, although there is a need for greater education on CFS/ME and services specifically designed around the needs of the young people (**Chapter 5**, **section 5.2.2**).

4.5.2 Developing local services

The incremental development of a locally based service, including provision of domiciliary care for severely affected patients, would significantly improve care for all patients with CFS/ME. Ideally, services would be patient-centred, and adopt a biopsychosocial model or a holistic view of care (**see 3.3.4**). The general components of such a service are:

- Medical care;
- Support for adjustment and coping (including CBT);
- Facilities for energy/activity management (including GET);
- Nursing and personal care.

4.5.2.1 Primary care

The development of primary care services is important because where possible CFS/ME should be managed in the community. The development of a local service with locally available expertise is key to providing support to GPs in their task of providing ongoing advice and care to patients and carers. The development of a skilled multidisciplinary team to support rehabilitation programmes and adjustment to the disease and disability is considered to be a cost-effective development that will reduce referrals to secondary care consultants. This team should have the capacity to develop a local network of services to support in particular the severely affected, house-bound and bed-bound patients who are currently unable to access services.

4.5.2.2 Further care

The provision and optimisation of specialist expertise and facilities should be considered as part of the commissioning process for primary care teams. The majority of the provision will be for secondary care: outpatient consultations, diagnostics, and clinical investigation, and to develop the medical components of management, including symptom control and specific interventions, patient education, GP advice, specialist clinical referral, and referral to multidisciplinary teams or other services (e.g. nutritional advice and support). Identifying a physician or other specialist to champion the development of local specialist services seems one key to success; an alternative model is the appointment of a suitably qualified and experienced GP. Involvement with the voluntary sector is likely to be beneficial to the development of services.

Dedicated inpatient services for CFS/ME are lacking. Development of inpatient services will be required in the immediate future to provide advice, evaluation, and inpatient care for those with complex needs.

Hospital-based services can play an important role in providing education to healthcare practitioners and can act as a knowledge resource to both primary and secondary care organisations.

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Chapter 5

Children and young people

"It is important to believe the child and listen to their views."

Children and young people (defined as being of school age) do get and are profoundly affected by CFS/ME, contrary to some professional and public perceptions. However, there are important differences as well as similarities between children and adults both in the nature and impact of the disease and in its management. This chapter reviews special considerations for the disease and its management in children and young people, but it should not be read in isolation. Rather, the rest of the report should serve as the context in which to place particular aspects of care in young patients.

The principles of care of children and young people with chronic illness are well established. Management of such conditions in England is guided by principles laid down by the Department of Health for England. Children's rights are safeguarded by UN convention and need to be respected at all times by professionals and parents/carers. 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.

Key messages

- CFS/ME represents a substantial problem in the young – "children do get it", though many recover, even after prolonged illness.
- Important differences exist between children and adults in the nature and impact of the disease and its management.
- The condition potentially threatens physical, emotional, and intellectual development of children and young people, and can disrupt education and social and family life at a particularly vulnerable time of life.
- Clinicians face additional difficulties in supporting and managing the younger patients and their families and parents/carers.
- An especially prompt and authoritative diagnosis is needed in the young, while the possibility of other illnesses and complications must also remain in mind.
- Ideal management is patient-centred, community-based, multidisciplinary, and co-ordinated, with regular follow-up. Community paediatric services need to be available for most children and all with prolonged school absence.
- The clinician who co-ordinates care needs to consider educational needs and impact on the family and parents/carers as early as practicable.
- Care is best delivered according to a specific, flexible, patient-focused treatment plan, designed and reviewed regularly with patient and family.
- Future services need to be developed around the needs of the child or young person and their family.

5.1 Nature and impact of CFS/ME in children and young people

Ill health imposes great demands on children and young people, so it is remarkable how many meet the challenges positively. Isolation from peer groups and loss of schooling cause a young person concern even if for short periods and for a well-known disorder. When their condition is less well known, or when the young person meets disbelief or perceives professional hostility or criticism of his or her parents/carers, the impact of the illness is compounded. No more is known about CFS/ME in children and young people than in adults, but this should not prompt professional indifference – quite the opposite. The Working Group notes that the Royal Colleges have a continuing role to play in this respect. We would welcome 'bridge-building' initiatives between children and young people, their parents/carers, voluntary organisations, and paediatricians and child psychiatrists through their respective Royal Colleges.

Young people with CFS/ME, especially of long duration, are vulnerable to negative effects on growth, including physical, emotional, and intellectual development, which may perpetuate impairment. The potential influence of any chronic disorder on education is of particular concern, as is the broader effect of the illness on parents/carers and family life. The relative lack of professional certainty over CFS/ME especially compounds difficulties for children with learning difficulties or those from minority groups, who also can be affected by CFS/ME. In general, diagnostic criteria for CFS/ME are poorly defined in children, and the disease may present differently in those with coexisting disadvantage or disability.

5.1.1 Epidemiology

During the past decade, CFS/ME has become more commonly diagnosed among school-age children and even in children as young as 5, although evidence suggests increased onset at secondary school age and around 14-15 years. Information on the disease burden in young people is scanty. Two studies have suggested a prevalence rate of 0.07% for children and young people. Dowsett and Colby[1] found that 51% of all long-term sickness absence in a studied school population of 333,000 was due to CFS/ME. Regardless of whether these figures are more widely applicable, CFS/ME clearly represents a substantial and widespread problem in the young.[2] Few children in ethnic minority groups have been diagnosed with CFS/ME, although more children seem to be accessing services.

5.1.2 Clinical profile

In general, CFS/ME that develops in a child or young person is recognisably the same clinical entity that develops in adults (detailed in **Chapter 4**), with some differences. Children usually have a symptom pattern similar to adults, but they are particularly prone to abdominal pain, nausea, and variations in appetite, leading to weight gain or loss. Younger patients do not always experience the delayed onset of symptoms after increased physical or cognitive activity, but they generally do have a prolonged recovery period after activity. However, even in those with a mild form of the illness, physical activity is usually limited and loss of schooling occurs.

As with adults, symptoms wax and wane over time and in severity, and are affected by the individual's circumstances, previous healthy functioning, and responses to

the illness. Aetiological and pathological considerations, including psychological factors, seem similar to those in adults, but children seem to be more vulnerable to the misconception that the disease is “all in the mind” or worse, that it does not exist. In addition, children do not always listen to caution from adults, teenagers may deny they have the illness, and very young children are often not able to understand the fluctuating nature of the condition. The expected duration of the illness in younger people is unknown because, as in adults, it is unique to the individual. However, most studies suggest that prognosis is better for children and young people than for adults, and many children recover even after long illnesses.

5.1.2.1 Severely affected children and young people

Some children and young people with CFS/ME are so severely affected by the disease that they become bed-bound, with a similar degree of cognitive and physical impairment to that experienced by patients with severe neurological conditions. Sensitivity to light, sound, and touch are characteristic symptoms in this group, together with visual deficits, hypotonia, and myoclonic jerks. Other aspects of the condition, such as nutrition, may also be severely affected. Care of such children and young people presents particular challenges. Diagnosis maybe difficult and require particular care. In general, such patients would benefit from mobilisation of full community supportive services. Additional support and services may be required (e.g. tube feeding). Where they exist, community children’s nursing teams are ideal to co-ordinate care with these patients and their parents/carers. Such support may need to continue for months or years.

5.1.3 Social impact

Within a family, any individual affected with CFS/ME has a profound impact on family life, particularly when that individual is a young person. The family unit experiences increased stress, stopping of normal activities, and in some instances the inability of other family members to accept the illness. The need for ongoing care means that parent(s) may have to give up work. In turn, the young person’s progress towards increasing independence is impeded, children become more reliant on their parents/carers, and separation from them can cause distress. Many young people lose contact with friends and peers, which means that often the individual’s only peer-group contact is with siblings. Other children in the family can feel left out as attention is focused on the ill child. CFS/ME can cause social isolation and an end to ‘normal’ family life. Clinicians and other professionals caring for the child can play a valuable part in minimising the impact of the disease on the family. Referral to the Disability Social Work Team should also be considered, to assess the need for support to the child and to help the family access disability related services. [see **5.3**].

5.2 Management

CFS/ME needs to be managed and services developed in keeping with general principles applicable to any chronic disease in the young: around the needs of and in partnership with the young person and their family.[4] The uncertainties that surround CFS/ME are particularly difficult for children and young people, in whom an incorrect diagnosis presents an additional risk. Clinicians face additional difficulties in supporting and managing younger patients and their families,

including issues over off-licence or off-label prescribing and obtaining consent to research or treatment in minors. Fortunately, children's health services are well placed to give optimum care for CFS/ME. Many child patients and their families speak warmly of the support they have received.

5.2.1 Diagnosis

> "People saying what's not wrong with you and never what is."

A diagnosis of CFS/ME in the young must be especially prompt, accurate, and authoritative, and second opinions are needed if doubt exists. As with other medical conditions that lack a diagnostic test, the diagnostic process for CFS/ME is the familiar one of assembling positive clues from the history and examination, while simultaneously ruling out other conditions, usually by laboratory and imaging investigations. During this process and throughout the illness, it is as important to consider symptoms in the context of the entire length and breadth of the illness as it is to focus on a single symptom for fear of overlooking an alternative condition (e.g. headache and brain tumour).

Other conditions that present with school absence are important differential diagnoses in the young, since they can mimic or complicate CFS/ME. Such disorders need to be considered early and excluded or treated. The possibilities including physical illness such as hypothyroidism, musculoskeletal, neurological or cardiac disorders and mental and social conditions, particularly depression, but also eating disorders, refusal syndromes and rarely child abuse. Primary and secondary/psychiatric co-morbidity will need recognition and management. Some children with chronic unexplained symptoms will not fit a unifying diagnosis but will still have medical and other needs to be met. (see also **Chapter 4** and **Annexes 6** and **7**).

Almost all healthy children and young people are in full-time education. Given the potential impact of illness on education, the timescale for establishing a working diagnosis and management plan needs to be minimal, whatever the cause. When a child or young person has suffered excessive tiredness and/or other symptoms leading to fragmentary school attendance or absence for at least 15 working days, active steps should be undertaken to identify the cause from a list of diagnoses that includes CFS/ME. Excessive tiredness or 'fatigue' may not be a presenting problem in children with CFS/ME and might only emerge from careful history taking.

5.2.2 Approach to management

An ideal approach to management is patient-centred (see also **Chapter 4**), and involves early recognition of CFS/ME by primary care services, and confirmation by a specialist if appropriate. A treatment plan can then evolve depending on the degree of incapacity, with follow-up by the clinician(s) at an appropriate level for the incapacity, and according to local expertise and specialist interest. The plan needs to be developed with the patient and family, and revised according to feedback. Valuable clinical support can be offered to assist the family in keeping a diary, managing the child's limited energy, and developing a balanced approach to activity.

Desired outcomes and an approximate timetable for their achievement need to be agreed with all concerned. Setbacks or an inability to reach certain goals are not uncommon and all concerned need to be aware of this possibility, to avoid inadvertent criticism or censure. Ultimately, as recovery proceeds, an individually tailored reintegration programme for return to education and social functioning can be developed that is mutually agreed and non-coercive. Throughout, the child or young person needs to be listened to, understood, and allowed as much control as possible over their care. The principles of obtaining consent should be followed as set out by the Department of Health (see http://www.doh.gov/consent) and due weight given to the child's assent or otherwise. The Gillick principles may need to be considered.

Few chronic medical conditions in childhood are managed by a single discipline and many require specialist follow-up. The hallmark of successful chronic disease management in children is integrated multidisciplinary support, provided locally and usually co-ordinated by a paediatrician in partnership with family/carers and children. Involvement of a social worker early in the assessment process may be helpful to support families and professionals. Most children who are missing school can be cared for and managed in their homes, with follow-up in primary care or by a specialist such as a community paediatrician. Given the variability in professional expertise and education on CFS/ME, the speciality of the co-ordinating clinician is not as important as the need for someone to adopt that role. If needed, hospital follow-up can be with a general paediatrician or other specialist mutually agreed by the child, their family, and GP. In general, a child who has prolonged school absence needs to be under the care of a specialist.

Other specialists, including child psychiatry when appropriate and professions allied to medicine are important in management. Affected children and young people will benefit from psychological support in some form. Joint work or referral to the Child and Adolescent Mental Health Services (CAMHS) may be helpful. Early referral to occupational therapy or physiotherapy services can be made if a specific need (e.g. for equipment) is identified early on. More expertise is required in school nursing and health visiting services for children and young people with CFS/ME.

In general, local services are preferred, although non-local services might be accessed for respite care and in specialist/hospital care for second opinions and for complicated cases. Hospital admission is mainly reserved for difficult diagnostic assessment and for dealing with severe complications, serious intercurrent illness, or other specific problems. Whether convalescent inpatient services are needed for children and young people with CFS/ME is debatable. An acute ward is not the optimum environment for an adolescent with a long-term medical condition, but few other options now exist.

5.2.3 Primary care

The affected child's GP is pivotal to orchestrating an effective response to the illness, and to alerting community paediatric services. The GP may also be the most appropriate clinician to co-ordinate care in some patients, although this would usually be done with input from a paediatrician. When a paediatrician is the key clinical co-ordinator, the GP may be aware of or best placed to treat other

medical conditions, particularly those causing school absence, and to consider the impact on family and parents/carers.

GPs may require additional or specialist support because they may be unaware of all potential options for assistance. 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. 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, including nutritional needs and medication. Although many healthcare professionals have not had specific education on CFS/ME, all should follow general principles for care of children with chronic illness. Primary care workers need at least to be aware of ongoing liaison with the child's Local Education Authority, particularly over provision of services such as transport and home tuition [see **section 5.2.6**.].

Many children and young people with CFS/ME have unpleasant symptoms, just as adults do. Pharmacological and other treatment can be needed to relieve such symptoms. Clinicians should apply the same general principles to prescribing for this group as for adults, while recognising that many products will not be licensed for such indications or for patients of particular age. If doubt exists, advice needs to be sought from a specialist. *Medicines for Children* (from the Royal College of Paediatrics and Child Health) and the *British National Formulary* are useful sources of reference.

5.2.4 Further care

The ideal ongoing management plan (including pharmacological, psychological, educational, and rehabilitative aspects) would depend on the individual's condition and circumstances, and be revised according to feedback. Various therapists and teachers may be involved, either throughout the illness or for specific periods, including: home tutors, physiotherapists, occupational therapists, nutritional experts, specialist nurses, play therapists, mental health professionals, and doctors in other medical specialities (e.g. surgery). Therapists and educationalists need to adopt a common approach to the condition as far as possible, in partnership with the child and family. Children's pain and symptom control services are developing but provision is patchy. Ideally, all children should have access to such services.

Selective joint work or specialist referral to Child and Adolescent Mental Health teams (CAMHS) can be important in the assessment and management of children and young people with CFS/ME. Firstly, a psychiatric opinion can be key to diagnosis, to identify a depressed child presenting with symptoms similar to those in CFS/ME or depression as a secondary consequence of CFS/ME. Secondly, children and young people with chronic illness – whether that be malignant, renal, respiratory disease, or CFS/ME – can develop emotional or behavioural complications that would benefit from specialist management as a physical complication would. Thirdly, CAMHS may be well placed to liaise with educational and other services.

Specialist referral to CAMHS needs particular sensitivity to the beliefs and fears of the young person and their family. Younger people may be more vulnerable to the general stigma surrounding mental illness, and the specific potential for misattributing CFS/ME as "all in the mind". As with any other service, referral needs

explanatory discussion and agreement with the patient and their family, to place the offer of psychological support in the appropriate context of a broad model of the illness and the benefits of psychological support in all forms of chronic illness.

Advice from physiotherapy or occupational therapy services may be sought for general energy/activity management, and particularly for sympathetic consideration of equipment needs. Provision of a wheelchair to a child can be perceived as a dramatic step to take, but wheelchairs can increase independence and allow families to regain a sense of normality in their social life. The notion of "once in a wheelchair, never out" is prejudicial: each case must be assessed according to clinical and functional need.

All services need to be audited and opportunities taken to create service networks to maintain standards, promote research, and share good practice. So-called 'postcode provision of services', when access to services is limited by the patient's residence, is unacceptable.

5.2.5 Follow-up

Most children with chronic medical conditions need specialist follow-up, with the frequency and scope dictated by the condition and its progress. A balance between primary and specialist/hospital care needs to be found that is acceptable to patient and family, with the most appropriate clinician co-ordinating follow-up, whether in primary or further care. In keeping with the principle of locally accessible services, primary care should bear prime responsibility for follow-up where possible. However, more complex cases will need follow-up by clinicians more experienced in their management. For school-age children for whom education is so vital, the most appropriate service to follow up certain coexisting conditions, such as neurological, learning, and seizure disorders, may be community paediatrics.

During follow-up, particular attention needs to be paid to progress made towards specific patient-directed goals in various rehabilitative fields. The clinician who follows up the patient must be aware of potential complications, whether physical, psychological, or social, to detect them and offer appropriate treatment where necessary. Children with chronic medical conditions can develop new medical conditions, which could be physical, psychological, or social, and may require treatment in their own right. Sometimes, new symptoms that may mimic those of the primary condition (e.g. children with chronic abdominal complaints can develop appendicitis). The original diagnosis needs to be kept under review.

5.2.6 Education

Nearly all children who are severely affected and many who are moderately affected will require the provision of home tuition and/or distance learning. A critical element of the child's management is assessment and provision of educational needs. An educational plan is not an optional extra but an integral part of therapy, just as play is for the younger child. A young person who is likely to have special needs, including home tuition, should be identified early in the diagnostic process, preferably by a GP or paediatrician. The co-ordinating clinician is then responsible for early referral to the Education Welfare Service to ensure that education is minimally disrupted. Adequate provision of continuing education needs close

liaison between GP, community paediatric services, education services, the young person, and their family.

Some young people will be too severely affected by their illness to participate in any form of education, even at home. A resumption of education, in whatever form, should be managed in keeping with the general principles of activity management as outlined in **Chapter 4** and **Annex 6**. Specifically, a young person with CFS/ME should never be forced to study but instead should be encouraged to set a pace that is likely to be sustainable, then have their progress regularly reviewed.

With support and reassurance, both schools and families can reach a position where the child is attending their school for short periods, is working in a separate area quietly if need be, can rest or work as their ability to concentrate fluctuates through the day, and can maintain some contact with their peers. Gradually they can be reintegrated into the mainstream education system. The advantage of this approach is that it minimises the isolation of the child once he or she is able to get out of the house. It does require sensitive negotiation with the school and a tolerance on all sides.

Some more severely disabled children may need home tuition and/or distance learning on a longer-term basis. In addition to the time of a tutor or therapist, this may require information and communications technology, which can also help improve social contact.

5.2.7 Transition to adulthood

Children with CFS/ME grow up: the onset of adolescence and adulthood needs to be anticipated and the potentially retarding effects of a chronic medical condition such as CFS/ME on emotional, physical, sexual and social development should be minimised. This is a unique period of a person's life. Services for young people with CFS/ME should be tailored to their progress to adulthood: in particular arrangements for transition from paediatric secondary care to adult medical services need to be put in place well before it happens.

5.2.8 Child protection

On occasions, families of child sufferers with more severe CFS/ME have been the subject of child protection concerns. The Working Group notes that 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 patient by the parents/carers and/or young person 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. *Working*

together to safeguard children, issued jointly by the Department of Health, the Department for Education and Skills, and the Home Office, sets out the interagency 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.

5.3 Managing the impact on family/carers

All professional involvement with family and carers requires a clear awareness of the implications for parents/carers of uncertainty over their child's illness. Clinicians who are sympathetic and responsive to the increased parental anxiety engendered by diagnostic uncertainty and other specific fears are likely to minimise additional impact on the child. A background of disbelief and misattribution can pave the way for parents/carers experiencing a sense that their parenting is under scrutiny and liable to criticism, and for feelings of doubt, blame, or guilt. Clinicians need to take steps to empower parents/carers through communication over and agreement on the nature of their child's illness and the management plan that follows from this.

As with other chronic childhood illness, professional work needs to be founded on a clear awareness of the crucial role of parent(s)/carers in undertaking the main care of the child, and aim to equip them adequately to support the child's progress. Several factors can affect parents' capacity to support and care for their child, all of which could influence progress and all of which may change during the course of the illness.

These include:

- The nature of their relationship with the child (illness may accentuate pre-existing vulnerabilities);
- Energy levels (adequacy of sleep, respite, practical support, health);
- Emotional resilience (anxiety, depression, other life events);
- Other commitments (family and personal priorities, professional commitments, etc);
- Knowledge, skills, and understanding of the condition and the care the child requires; and
- Adequate support (spouse, extended family, friends, professionals).

Practitioners can support parents/carers by recognising the need to offer them opportunities to speak when the child is not present, though the child's consent and agreement should be obtained before this goes ahead. Parents/carers are likely to be concerned with protecting the child from a sense of being a burden, from the problematic implications of confused perceptions of the illness, and from the consequences of their own anxieties. It may be difficult for parents/carers to ask for help if it is not offered, since it is the child who is the patient. Risks for the main carer mirror those for the child (social isolation, work difficulties, loss of leisure, coping with the misperceptions of others, perceived blame, loss of confidence, depression, anxiety, etc).

Clinicians and other practitioners may be well-placed to equip parents/carers to improve the child's and their own situation. Potential steps include:

- Identifying child and family strengths that can be developed despite illness;
- Prioritising areas in which long-term damage is a particular risk (e.g. social integration);
- Recognition of secondary depression, anxiety, development of secondary gain, difficulties of reintegration, social isolation, overdependency, etc;
- Awareness of the implications of the child's increasing maturation, if illness is prolonged;
- Anticipation and recognition of difficulties for siblings (e.g. jealousy, perceived favouritism, difficulties of 'making allowances' over months or years, etc).

Awareness of these issues needs to be matched by awareness of the importance to the child's care of parents' confidence in perceiving their own child's needs. It is important that parental confidence is actively protected, since it may be made vulnerable by both lay and professional responses to the illness. Careful listening and respect for parents/carers' opinions are important factors. Although a family environment will inevitably impact on the course of any chronic childhood illness, the lack of blame needs to be specifically stated, and this is the key message that can open doors to achieving necessary support for the child and family.

Key references

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Chapter 6

Recommendations of the Working Group

CFS/ME is a relatively common clinical condition, which can cause profound, often prolonged, illness and disability, and can have a very substantial impact on the individual and the family. It affects all age groups, including children. The Working Group has encountered extensive evidence on the extent of distress and disability that this condition causes to patients, carers, and families. It has examined the evidence on the effectiveness of interventions used in the management of this condition.

The Working Group is concerned about several issues. Patients and carers often encounter a lack of understanding from healthcare professionals. This lack seems to be associated with inadequate awareness and understanding of the illness among many health professionals and in the wider public. Many patients complain of the difficulty of obtaining a diagnosis in a timely manner. There is evidence of underprovision of treatment and care, with patchy and inconsistent service delivery and planning across the country. Finally, there is a paucity of good research evidence and very little research investment for a serious clinical problem that in likelihood has a pervasive impact on the individual and the community. Insufficient attention has been paid to differential outcomes and treatment responses in children and young adults, the severely affected, cultural, ethnic and social class groupings.

The Working Group has identified measures that should be taken with some urgency to address the current situation.

6.1 Recognition and definition of the illness

- The NHS and healthcare professionals should recognise CFS/ME as a chronic illness that, despite uncertain aetiology, can affect people of all ages to varying degrees, and in many cases substantially.
- In view of current dissatisfaction among some groups over the nomenclature applied to this illness, we recommend that the terminology should be reviewed, in concert with other international work on this topic.

6.2 Treatment and care

- Patients of all ages with CFS/ME must receive care and treatment commensurate with their health needs and the disability resulting from the illness.
- Healthcare professionals should have sufficient awareness, understanding, and knowledge of the illness to enable them to recognise, assess, manage, and support the patient with CFS/ME. Healthcare workers who feel they need extra skills should seek and receive help from those experienced in this area.
- General Practitioners should usually be able to manage most cases in the community setting, but must be able to refer patients for specialist opinion and advice where appropriate (e.g. because of complexity in diagnosis and treatment).
- CFS/ME of any severity in a child or young person – defined as of school age – is best co-ordinated by an appropriate specialist – usually a paediatrician or sometimes a child psychiatrist – in concert with the GP and a paediatric or CAMHS multidisciplinary team.
- Sufficient tertiary level specialists in CFS/ME should be available to advise and support colleagues in primary and secondary care.
- Management should be undertaken as a partnership with the patient, should be adapted to their needs and circumstances, and should be applied flexibly in the light of their clinical course.
- The support of the patient with CFS/ME and the management of the illness should usually extend to the patient's carers and family.
- Clinicians must give appropriate and clear advice, based on best national guidance, on the nature and impact of the illness to those involved in providing or assessing the patient's employment, education (primary, secondary, tertiary, and adult), social care, housing, benefits, insurance, and pensions.

6.3 Health service planning

- Service networks should be established to support patients in the primary care and community setting, to access when necessary the skills, experience, and resources of secondary and tertiary centres, incorporating the principles of stepped care. Services should be configured so that individual professionals and aspects of the service can meet individual needs, particularly in the transition from childhood to adult life.
- Health service commissioning through primary care organisations, supported by health authorities or wider consortia, must ensure that local provision for these patients is explicitly planned and properly resourced, and that health professionals are aware of the structure and locale of provision. Health commissioners should be requested to take immediate steps to identify the current level of service provision for CFS/ME patients within their locality.
- Each Strategic Health Authority should make provision for secondary and tertiary care for people with CFS/ME, based on an estimated annual prevalence rate of approximately 4,000 cases per million population in the absence of more refined data.

- People who are so severely affected that their disability renders them house-bound or bed-bound have particular constraints in regard to their access to care. These specific needs must be met through appropriate domiciliary services.
- The NHS should make use of the wide range of support and resources available through partnership arrangements with voluntary agencies, enabling suitable self-management by the patient.

6.4 Education and awareness

- The education and training of doctors, nurses, and other healthcare professionals should include CFS/ME, as an example of the wider impact of chronic illness on the patient, on carers and family, and on many aspects of society.
- Healthcare professionals, especially in primary care and medical specialities, should receive postgraduate education and training so that they can contribute appropriately and effectively to the management of patients with CFS/ME of all ages.
- GPs and medical specialists should consider CFS/ME as a differential diagnosis in appropriate patients, and should at least be able to offer initial basic guidance after diagnosing this condition (**Annexes 6** and **7**).
- Awareness and understanding of the illness needs to be increased among the general public, and through schools, the media, employers, agencies, and government departments.

6.5 Research

A programme of research on all aspects of CFS/ME is required.

Government investment in research on CFS/ME should encompass health-services research, epidemiology, behavioural and social science, clinical research and trials, and basic science.

In particular, research is urgently needed to:

- Elucidate the aetiology and pathogenesis of CFS/ME;
- Clarify its epidemiology and natural history;
- Characterise its spectrum and/or subgroups (including age-related subgroups);
- Assess a wide range of potential therapeutic interventions including symptom-control measures;
- Define appropriate outcome measures for clinical and research purposes; and
- Investigate the effectiveness and cost-effectiveness of different models of care.

The research programme should include a mix of commissioned or directed research alongside sufficient resource allocation for investigator-generated studies on the condition.

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Appendix **I**

Membership of the Working Group

Professor Allen Hutchinson	Chair
Professor Anthony J Pinching	Deputy Chair (from May 2000)
Dr Timothy Chambers	Chair of Children's Sub-Group (from July 2000)
Judith Waterman	Former Chair of Children's Sub-Group
Former Member	(to May 2000)
Naomi Wayne	Co-Chair/Former Chief Executive, Action for
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We are grateful to the members of the Working Group and Children's Group for their active participation, debate, and contribution to the formulation of this report.

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Appendix **II**

Existing diagnostic criteria (adults)

US Centers for Disease Control and Prevention, 1988 (CFS) (Holmes 1988)	6 months duration of fatigue Functional activity – 50% decrease in activity Six or eight symptoms required; physical signs sometimes required Neuropsychiatric symptoms – may be present New onset required *Exclusions:* Extensive list of known physical causes, psychosis, bipolar disorder, substance abuse
US Centers for Disease control and Prevention, 1994 (CFS) (Fukuda 1994)	6 months duration of fatigue Substantial functional impairment Four symptoms required Cognitive or neuropsychiatric symptoms may be present New onset required *Exclusions:* Clinically important medical conditions, melancholic depression, substance abuse, bipolar disorder, psychosis, eating disorders
Australia, 1990 (CFS) (Lloyd)	6 months duration of fatigue Substantial functional impairment – disruption of daily activities Postexertional fatigue No symptoms specified Cognitive or neuropsychiatric symptoms required New onset not required *Exclusions:* Known physical causes, psychosis, bipolar disorder, substance abuse, eating disorders
UK, 1991 "Oxford Criteria" (CFS) (Sharpe)	6 months duration of fatigue Disabling functional impairment – affects physical and mental functioning No symptoms specified Cognitive or neuropsychiatric symptoms – may be present Definite onset required *Exclusions:* Known physical causes, psychosis, bipolar disorder, eating disorder, organic brain disease, substance abuse Other psychiatric disorders (depressive illness, anxiety disorders) are not reasons for exclusion

London, 1990 (ME) (Derived from Dowsett & Ramsay)	Complaint of general or local muscular fatigue following minimal exertion with prolonged recovery time Neurological disturbance, especially of cognitive, autonomic, and sensory functions Variable involvement of cardiac and other systems, a prolonged relapsing course Syndrome commonly initiated by respiratory and/or gastrointestinal infection but an insidious or more dramatic onset after neurological, cardiac, or endocrine disability
World Health Organization, 1994	The WHO's International Classification of Diseases (ICD) provides a system of categories for international systematic recording. These are not diagnostic criteria, and are not used by clinicians as such. The current version, ICD-10, includes categories for 'Neurasthenia'/ 'Fatigue Syndrome' and for 'Post-Viral Fatigue Syndrome'/'Benign Myalgic Encephalomyelitis'.

Appendix **III**

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