MYALGIC ENCEPHALOMYELITIS (ME)

THE IMPACT ON SUFFERERS: IS HEALTH POLICY IN SCOTLAND ON THE RIGHT PATH?

'ME is a systemic* disease with many systemic features, but it is characterised primarily by CNS (central nervous system) dysfunction, of which fatigue is only one of many components.' (Hyde, 1992, p18)

**relating to, or affecting the whole body*

'……CFIDS* (ME) is not about being tired. Researchers have demonstrated numerous abnormalities of the immune, muscular, cardiovascular, and central nervous systems in people with CFIDS. It is truly a multi-system disease with a strong component of immune dysfunction.' (Congressional statement by DeFreitas, 1991, quoted in Shannon, 2000)

** used in the US – Chronic Fatigue and Immune Dysfunction Syndrome*

- ME is a serious, debilitating and chronic disease with no known cure affecting people of all ages, social classes and ethnic groups. It occurs in both sporadic and epidemic forms and can have a sudden or gradual onset (Carruthers *et al.*, 2003, p9). Families may have more than one member affected by the illness.

- The World Health Organisation classifies myalgic encephalomyelitis (ME) as a neurological disease, i.e. a disease of the central nervous system, in the International Classification of Diseases (ICD) 10, section G93.3. The UK Dept of Health formally accepts the ICD classification.

- **'ME is not T.A.T.T. – 'tired all the time''** (The National ME Centre)
  As with many chronic illnesses, fatigue may be present in many ME patients. However, the fatigue is post-exertional, often delayed, disproportionate to effort and quite unlike the 'fatigue' experienced by healthy people.

- **'ME is not depression, nor does depression cause ME'** (The National ME Centre)

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In consultation with the Cross Party Group on ME

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Key Points

- ME is defined by the World Health Organisation as a neurological condition. It is a serious and chronic disease with a spectrum of severity and no known cure.

- ME sufferers experience considerable deterioration in their quality of life which is more severe than in many other chronic illnesses.

- Sound epidemiological data does not exist – key information in planning services is therefore lacking.

- The introduction of the name 'chronic fatigue syndrome' (CFS) in place of ME fails to acknowledge a critical distinction between unexplained fatigue and ME. The impact on healthcare is significant as the implications for health care and management are quite different for these two conditions

- Health services for ME patients are inadequate and often inappropriate. Biomedical research which supports physiological and biochemical abnormalities is ignored in clinical practice. Many sufferers, irrespective of the severity of their condition, receive no medical care.

- Claims are made for the appropriateness and effectiveness of rehabilitative approaches but biomedical research and patient surveys challenge these claims and clinical opinion is divided.

- Clear clinical guidelines are essential and already exist in Canada. These have not yet been adopted in Scotland.

- The Scottish Executive's recent invitation to health boards to submit proposals for healthcare services for ME has revealed unsatisfactory proposals from most boards. They do not inspire confidence amongst ME patients that their illness is being addressed appropriately.

- Increased funding for research and services, proper dialogue and real consultation with researchers and patients are necessary for the inclusion in health care of ME patients.

Is ME a new disease?

- ME is **not** a new disease but has been documented in medical literature since at least the 1930s, with the term ME first used in the 1950s. It does, however, appear to be increasingly prevalent.
- A review of early outbreaks found that **'clinical symptoms were consistent in over sixty recorded epidemics of ME spread all over the world'** (Scottish Executive, 2003, p7).

What does ME mean?

ME: Myalgic Encephalomyelitis

- Myalgic: **Myalgia** means pain in a muscle or group of muscles.
- Encephalomyelitis: **'Encephalo-'** refers to the brain; **'-myel-'** to the spinal cord and **'-itis'** denotes inflammation.
  - Although the accuracy of '-itis' is questioned by some, research evidence demonstrating brain abnormalities supports the presence, or consequences, of inflammation (Dowsett, 2004).

What causes ME?

- We do not yet understand why people develop ME, but a number of triggering factors have been identified.
  - Viral infection: enteroviruses have been shown to be present in many outbreaks of ME (Dowsett, 2004).
  - Vaccinations e.g. hepatitis B
  - Toxins and pesticides

- In a minority there is no clear precipitating factor (Shepherd & Chaudhuri, 2001, p5)

- Claims as to why patients fail to recover, although acknowledging the above triggers, have become, and remain, influential: in particular,
    - physical deconditioning as a result of low levels of activity
    - a continuing belief that one is ill.
  - However, these claims are challenged by strong evidence from the clinical assessment of patients and biomedical research. (see Appendix 2)

How many people have ME?

There has been no systematic attempt to establish the incidence and prevalence of ME in Scotland, i.e. an epidemiological study.

- The Scottish Health Executive's Short Life Working Group Report (2003) suggests figures based on an estimated prevalence ranging from 2 to 4 per 1000 of the adult population – up to 20,500 sufferers (Scottish Executive, 2003, pp9-10).
  - This range is taken from the Chief Medical Officer's Report (Dept. of Health, 2002, p6), and is described by the report as a *minimum* prevalence range.
  - A total of 20,000 sufferers was described as a "conservative" estimate by Professor Jung, Scottish Chief Scientist, speaking at the Edinburgh Science Festival (9.4.04).

- For children and young people, the estimated prevalence is 7 per 10,000 – 600 children in Scotland.
  - Children as young as 5 years have been diagnosed (Scottish Executive, 2003, pp9-10).
  - **"ME is the biggest cause of Long Term Absence from School"** (Tymes Trust, 2003, p5).

- Three local studies – carried out in the north of Scotland, Fife, and the Western Isles (all unpublished) – yielded widely varying results, ranging from 3 to 27 per 1000 of population.

No one knows the accurate number of adults and children who are ill with ME or its spread within the population. The CMO's report acknowledges the lack of sound epidemiological data:

> **"……a key piece of information is missing – one that is needed in order to undertake a health-needs assessment as a prelude to provision of an adequate network of services."**
> **(**Dept. of Health, 2002, p6**)**

The Cross Party Group on ME's petition, lodged in September 2001, calls for an epidemiological study - an essential tool for the planning of services.

What are the distinguishing features of ME?

The clinical profile of ME is unique and does not mimic any other illness.

> **"ME was known to run a chronic course and patients had disabilities due to persistent symptoms of pain, fatigue and loss of endurance to normal physical activities with conspicuous deterioration of symptoms after exercise (post-exertional malaise)"** (Scottish Executive, 2003, p7).

- Post-exertional malaise is a key defining feature.
  - Patients experience a considerable exacerbation of symptoms which may precipitate a significant relapse, even after minor amounts of physical exertion.
  - Mental activity can also exacerbate symptoms.

- Cardinal symptoms are:
  - **"Muscle fatiguability whereby, after even a minor degree of physical effort, three, four or five days, or longer, elapse before full muscle power is restored is unique.**" (Ramsey, 1988, p 30)
  - Muscle pain (myalgia) that may include tenderness.
  - Cognitive / neurological dysfunction affecting e.g. memory; concentration, balance, vision, hearing, sleep rhythm, temperature, appetite, hormone production.

- A special feature of this disease is that the condition waxes and wanes.
  - Symptoms tend to vary and fluctuate in severity from hour to hour and day to day.
  - The illness often follows a pattern of flare-ups interspersed with periods of relative remission.
  - Some patients experience no remission at all.
  - The illness persists over the long-term.

- The pattern of symptoms and their severity varies from person to person. However the cardinal symptoms are common to all.

With acknowledgment to: CFS Research Foundation; Ramsey, 1988, 1991; Shepherd & Chaudhuri, 2001.

How does ME affect sufferers' lives?

- People with ME experience a marked deterioration in their quality of life.
  - **"The quality of life (QOL) of ME/CFS* patients shows marked diminution which is more severe than in many other chronic illnesses"** (Carruthers *et al*, 2003, p29, referring to the findings of six published studies) *CFS is discussed below

- There is a spectrum of severity.
  - Approximately **25% may be termed severely affected** i.e. severely restricted in mobility and ability to carry out essential daily tasks and attend to personal care.
  - At its most extreme, sufferers may be totally bedbound, in constant pain, unable to tolerate light or noise and even requiring to be tube-fed.
  - Fatalities, although rare, do occur (Carruthers *et al*, 2003, p34).

- Symptoms tend to fluctuate during the day and from day to day, making planning of routine activities very difficult. They inhibit employment for all but the mildest cases.
  - **"[Other] major sources of work disability in ME/CFS are the lack of endurance, the unpredictable symptom dynamics and the presence of delayed reactive fatigue and pain and cognitive dysfunction"** (Carruthers *et al,* 2003, p34).

- Regarding **prognosis**, affected individuals rarely experience a return to previous levels of health and functioning (Dept. of Health, 2002, p7).
  - In a nine year study of 177 patients only 12% reported recovery. **"Other studies** [five research papers are referenced] **suggest that less than 10% of patients return to premorbid levels of functioning"** (Carruthers *et al*, 2003, p29).

ME and 'chronic fatigue syndrome' (CFS) – is there a difference in terms of diagnosis?

- ME can be diagnosed clinically on the basis of its distinguishing features.
  - These features are reflected in published guidelines by Carruthers *et al*, 2003. [Discussed below]
  - Most doctors are unaware of the unique and distinctive presentation of this illness.

- The term ME is rarely used now in medical circles. Instead, people presenting with the clinical features of ME are generally given a diagnostic label of Chronic Fatigue Syndrome (CFS).

"In 1988, the term 'Chronic Fatigue Syndrome' was introduced as a **diagnostic term to define all chronic fatiguing disorders that are otherwise unexplained by known medical conditions"** (Scottish Executive, 2003, p7).

- This has changed the medical perception of ME
  - **ME is now commonly perceived in terms of chronic fatigue which has no medical basis.** Chronic fatigue is a symptom common to many illnesses.

- CFS is a broad term which is interpreted in different ways, not all relevant to ME, and this has caused confusion and confounding in research and clinical practice to the detriment of ME sufferers.
  - As one patient, giving evidence to the English Chief Medical Officer's Working Group, observed:

    "Alzheimer's Disease is not known as 'Chronic Forgetfulness Syndrome'!" (Dept. of Health, 2002, p15)

- The varied interpretations of CFS pose a problem for epidemiological studies.
  - These need to have a clear and agreed definition of the illness being studied to be effective.

- The use of the expression CFS fails to acknowledge a critical distinction:
  - Unexplained fatigue is listed as a mental and behavioural disorder in the World Health Organisation International Classification of Diseases (ICD) 10, section F48.0.
  - ME, as noted above, is classified as neurological (section G93.3)
  - Some researchers and clinicians, however, use a strict definition of CFS which equates with ME (see Carruthers et al, 2003).

- Many researchers and some doctors now acknowledge that the 'diagnosis' CFS almost certainly includes several different patient groups, i.e. different illnesses.

- For the individual patient, the illness is very real and their suffering is exacerbated by the diagnostic mess.

What healthcare is provided?

- Following ME/CFS diagnosis, no further explanations are generally sought for the range and types of symptoms.
  - Current NHS practice recommends routine, but limited, investigations which are intended only to exclude a range of conditions with related symptoms.

- Many medical practitioners have no knowledge of the origin, development and resultant effects of this illness.

- At best, a degree of palliative care is provided, but medical practitioners could be much better informed about the options available.

- For many patients, the reality is that they lose contact with their GP and are left to soldier on alone, often for many years.

- Many severely affected sufferers receive no medical care and the full extent of the severity of the illness is neither observed nor understood.
  - According to patient surveys, it would appear that around half of those most severely affected by this illness have no contact with the health service as a result of GP reluctance to carry out home visits. (Action for ME, 2001; 25% ME Group, 2004)

BIOMEDICAL RESEARCH

Research groups across the world have been uncovering physiological and biochemical abnormalities in groups of ME/CFS patients.

Some recent examples of **biochemical, vascular, brain and muscle research** are set out in appendix 2.

- Not all sufferers will exhibit all of these abnormalities and many researchers and doctors now acknowledge the need to identify **subgroups among patients** in order to make progress with medical understanding and appropriate management of the condition.
- **Biological research results are not applied in clinical practice**.
  - In consequence, patients are generally labelled as having 'nothing physically wrong'. This can be extremely detrimental, particularly to those who are unable to work and depend on sickness and disability benefits to provide a basic income.

BEHAVIOURAL INTERVENTIONS

Opinion is deeply divided regarding the appropriateness and effectiveness of behavioural interventions aimed at rehabilitation.

- The Chief Medical Officer's Report on ME stated that "**no management approach to CFS/ME has been found universally beneficial, and none can be considered a cure"** (Dept of Health, 2002, p34).
- Nevertheless, two behavioural interventions are frequently cited as beneficial, and potentially curative, treatments for ME
  - **Cognitive Behaviour Therapy** (CBT) is a psychological intervention which aims to alter the ways patients view or cope with their illness to facilitate improved functioning.
  - **Graded Exercise Therapy** (GET) involves structured and supervised activity management which aims to increase previously avoided activities.

The sole source of evidence which would support behavioural interventions is research studies aiming to address the needs of patients with unexplained chronic fatigue, <u>not</u> ME.

- The evidence for GET in respect of ME is disputed. (Appendix 3)
  - In one survey of severely affected patients, 8 out of 10 reported that their illness had been made worse by graded exercise.
  - **Some of these patients were not severely affected before graded exercise therapy**.
  - **"no other treatment (sic) – pharmacological or non-pharmacological – received such negative feedback in patient surveys"** (Dept. of Health, 2002, p47)

- Biomedical research evidence supports the inappropriateness, and at worst harmfulness, of graded exercise to patients with ME.
- It is unacceptable that CBT may be applied in ways which encourage ME sufferers to believe that their illness does not have a biomedical basis.
- **Duty of care** is called into question if behavioural interventions are the only approaches offered to ME sufferers.
    - **"To ignore the demonstrated biological pathology of this illness, to disregard the patient's autonomy and experience and tell them to ignore their symptoms, all too often leads to blaming patients for their illness and withholding medical support and treatment"** (Carruthers *et al*., 2003, p47).

The petition of the Cross Party Group on ME identifies the need for a strategic needs review assessment.

CLEAR CLINICAL GUIDELINES

The adoption of the Canadian clinical guidelines, and recognition of their implications, would constitute a major step forward in health policy for ME in Scotland.

- This recently published paper, setting out clinical criteria, and developed following input from world leaders in research and clinical management, is authoritative.
    - It is **"based on the consensus panel's collective extensive clinical experience diagnosing and/or treating more than twenty thousand (20,000) ME/CFS patients…"** (Carruthers *et al*., 2003, p 9-10).
    - The Canadian clinical guidelines use the term 'ME/CFS' in defining the illness. Their definition is generally accepted as appropriate to ME by biomedical researchers and patient groups.
    - They incorporate an effective diagnostic protocol.

        "We present a systematic clinical working case definition that encourages a diagnosis based on characteristic patterns of symptom clusters, which reflect specific areas of pathogenesis." (*ibid*., page 7-8)
    - They set out valuable advice for the investigation of symptoms and their management.

- **The Canadian clinical guidelines are not yet endorsed by the Scottish Parliament's Health Executive.**

The Scottish Executive and ME – the way forward.

This paper has described ME and the impact on sufferers of the debates within research and medicine about its nature. The ways in which health policy responds to these debates also have an impact. From the perspective of ME patients, problems exist in three areas:

- funding of services
- organisation of services
- official perception of the illness.

FUNDING OF SERVICES FOR ME

Following the Short Life Action Group's report (2003), the Health Executive invited health boards to submit proposals for the development of services for 'CFS/ME'. This has taken considerable time.

- Finance is a problem.
  - Patients who were represented in health board discussions on service development noted that health boards frequently commented on the lack of funds to implement services.
  - The Health Executive has said that no extra funding will be available.
  - The Department of Health in England has released £8.5 million for the development of services for ME.

Parity in the funding of services is essential if ME patients in Scotland are to have an equal level of service as those in England.

ORGANISATION OF SERVICES

The proposals for the organisation of services submitted to the Health Executive were presented to the Cross Party Group on ME by Dr Mac Armstrong, Chief Medical Officer, on 24 November 2004.

- This exercise has produced unsatisfactory results. Alex Fergusson, convenor of the CPG on ME, commented to the Health Committee on 1 February 2005, that:
  - The proposals represented a piecemeal approach to ME services with a 'post-code lottery' effect: health boards proposed different models of provision; four boards proposed no service at all.

- Only two health boards have engaged in meaningful patient consultation. In the majority consultation has been minimal, unsatisfactory or non-existent, contrary to Health Executive guidelines.

- Health service provision for ME has been neglected for a considerable time. As a result, there is little expertise within health boards on which to build appropriate and effective services.

The Cross Party Group on ME's petition calls for the establishment of a centre of excellence.

- **The advantage of a centre of excellence would be to reverse ME's neglect by:**
  - **Co-ordinating epidemiology, research and strategic needs**
  - **Disseminating research information and best clinical practice to clinical teams and GPs at a local level**
  - **Facilitating a feedback of information about ME patients to the centre**

The Scottish Executive has not adequately assessed the needs of the majority of ME sufferers.

The aims of the CPG on ME's petition have not yet been met.

OFFICIAL PERCEPTION OF THE ILLNESS

"The philosophy behind management/treatment programmes is of the utmost importance" (Carruthers et al, 2003, p37).

The way in which the illness is understood determines what care is provided.

Dr Mac Armstrong, Chief Medical Officer, has acknowledged that ME patients have been excluded from appropriate care within the health service.

- However, the view that inclusion is achieved by absorbing ME patients within services for chronic fatigue is unfounded.
  - ME and unexplained chronic fatigue are not the same illness.
  - Rehabilitative approaches which are beneficial for chronic fatigue patients are already known to be inappropriate and ineffective in treating ME patients.

Inclusion of ME patients in appropriate and effective health care requires Scottish Executive support for:

- **Biomedical research**
- **Increased funding for services**
- **New perspectives in the organisation of services**
- **Consultation with medical researchers and patients about the illness**

The Scottish Executive has stated its aim of inclusion for all. The problem of how to include ME sufferers will only be resolved by proper dialogue - real consultation based on real and relevant information.

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Appendix 1

The Petition of the Cross Party Group on ME

The aims of the petition are to:

- carry out a Strategic Needs Review Assessment on ME and CFS in Scotland
- establish the size of the ME and CFS population
- establish the proportion severely affected and establish the Benefits entitlement & uptake of these
- establish a centre of excellence for the treatment of and research into ME and CFS
- ensure that GPs are informed about the advances in diagnosis and treatment
- ensure the GPs are informed about the new centre and liaise with it.

Appendix 2

Biomedical Research

BIOCHEMICAL

- Oxidative stress – (Richards et al., 2000; review by Pall, 2001; Kennedy et al., 2003; Vecchiet et al., 2003)
- Dysregulation of anti-viral pathways – i.e. abnormal activity of the anti-viral immune responses (Suhadolnik et al., 1994; De Meirleir et al., 2000; Tiev et al., 2003)

VASCULAR

- Endothelial dysregulation – i.e. abnormal responses of small blood vessels selectively to acetylcholine (Spence et al., 2000; Khan et al., 2003 and 2004)
- Altered brain perfusion – i.e. areas of reduced blood flow in the brain (Ichise et al., 1992; Costa et al., 1995; Tirelli et al., 1998)
- Orthostatic hypotension – i.e. physiological changes to blood pressure/cardiovascular mechanisms on standing (Streeten et al., 2001; Naschitz et al., 2002; Stewart et al., 2003)

BRAIN

- Metabolic abnormalities – e.g. alterations of brain choline (important in brain function) (Tomoda et al., 2000; Puri et al., 2002; Chaudhuri et al., 2003)

MUSCLE

- Altered metabolism – e.g. changes in muscle composition or use of fuel (Fulle et al., 2000; Vecchiet et al., 2003; Fulle et al., 2003)
- Abnormal response to exercise (Lane et al., 1998; Paul et al., 1999; McCully et al., 2004)
- Enteroviral sequences in muscle – i.e. evidence of a persisting virus in some CFS patients (Lane et al., 2003; Douche-Aourik et al., 2003)

(Spence, 2003)

Appendix 3

Evidence on Behavioural Interventions

Surveys conducted by patient charities have consistently indicated that rehabilitative approaches are harmful to a considerable proportion of patients, and clinical opinion is deeply divided on the subject. The table below summarises the evidence on behavioural interventions (Dept. of Health, 2002, p 46-50)

SOURCE of EVIDENCE	**ASSESSMENT OF EVIDENCE**	
	GRADED EXERCISE	COGNITIVE BEHAVIOUR THERAPY
RESEARCH FINDINGS	*"promising results"*	*"positive results"*
PATIENT REPORTS	*predominantly harmful or ineffective*	*wide variation; predominantly ineffective, substantial minority harmed*
CLINICAL OPINION	*"disagreement"*	*beneficial to some "when applied appropriately"*

- The assessment of 'promising'/ 'positive' results is based on the reported findings of 7 published studies (3 on graded exercise; 4 on cognitive behaviour therapy). One of the 7 studies found no benefit to patients. This study used tighter criteria for the selection of research subjects.
  - The limitations of these trials have been discussed in the British Medical Journal e.g. Abbot, N. & Newton, D. (2002): *Question marks over the evidential basis of claims for psychosocial therapies*: the main points are summarised in MERGE (2002): *Unhelpful Counsel?*.

- Research into behavioural interventions has received considerably greater funding than biomedical research.
  - According to figures provided by the Chief Scientist Office in Scotland, approximately £500,000 has been awarded to psychosocial research compared with only £9,000 allocated to biomedical research.