9 SEVERE ME/CFS

There are serious deficiencies in the way that severely affected ME/CFS patients (i.e. those who spend a considerable period of time house-, wheelchair- or bed-bound) are managed in both primary and secondary care. Although no accurate figures are available, it is estimated that around 25% of all people with ME/CFS will fall into this category at some stage in their illness.

Feedback to The ME Association indicates that a great deal more could be done at a primary care level in relation to the provision of regular home assessments by GPs, involvement of community nurses, occupational therapists and physiotherapists, investigation of new or prominent symptoms, effective symptom control and pain management, support from social services and availability of local respite care. When it comes to secondary care, there are very few hospitals with in-patient facilities to which severely affected patients can be referred for both clinical assessment and management (McDermott *et al* 2014). Even where facilities exist, more consideration needs to be given to practical problems faced by the severely affected when attending out-patient appointments. The situation is even worse in relation to the provision of suitable facilities for in-patient management and for domiciliary assessment of severely affected patients who are unable to travel.

9.1 Assessment and diagnosis of people with severe ME/CFS

Where someone presents with severe symptoms and the diagnosis has not been established, and probably delayed for some time since the onset of symptoms, a far more thorough clinical assessment is required to make sure that other diagnostic explanations have been excluded.

The pre-diagnosis clinical assessment of people with more severe symptoms should also include a nutritional assessment, preferably carried out by a dietician, and making use of a validated nutritional screening tool such as the Malnutrition Universal Screening Tool (MUST) or the Mini Nutritional Assessment (MNA). These tools are designed to record and monitor a person's body mass index (BMI) along with any recent weight loss. Clinical signs of malnutrition can include loss of hair, changes in facial features, a red swollen tongue, loss of skin elasticity when pinched, brittle nails and muscle wasting.

Consideration should also be given to the presence of complications such as vitamin D deficiency where exposure to natural sunlight has been significantly or completely reduced.

Baxter *et al* (2021) have reviewed life-threatening malnutrition in severe ME/CFS and noted that there can often be a significant delay in implementing proper care allowing the patient to become severely malnourished. They noted that healthcare professionals may fail to recognize that these problems are a direct consequence of very severe ME, preferring to postulate psychological theories rather than addressing the primary clinical need. They present five case reports in which delay in instigating tube feeding led to severe malnutrition of a life-threatening degree.

9.2 Management of severe ME/CFS

Cognitive dysfunction, pain and sensitivity to light and/or noise can be very disabling for people with severe ME/CFS. Serious neurological symptoms can also be present. These can include double vision, blackouts, atypical convulsions, loss of speech, and loss of swallowing necessitating nasogastric/tube feeding.

Severely affected patients often report different responses to management strategies that are commonly applied to people with mild or moderate CFS/ME. Health professionals must therefore recognise the need to adapt both drug and non-drug therapies to the functional level of the patient, and to adjust them further in response to patient feedback. This is particularly important when it comes to any form of management that involves physical or cognitive activity. Management of people with severe ME/CFS should also include assessment of vitamin D status, regular monitoring of weight and nutritional status, along with guidance on and to minimise the risk of potential problems (eg pressure sores, osteoporosis) that are associated with prolonged immobility.

The 2002 Chief Medical Officer's Report stated that the care of people who are severely affected is an urgent challenge that needs to be addressed by drawing on service models applied to other severe chronic disabilities. Healthcare and social service professionals are, in theory, responsible for supporting and guiding patients and their carers for the duration of any long-term illness. Health professionals should therefore be ensuring access to all available support, keeping in contact, constantly re-evaluating the options, maintaining morale, enabling respite, and minimising the consequences of prolonged disease. Those with severe or prolonged illness will normally need support in their homes and communities. Local domiciliary services linked to existing primary and community-care structures are key components here. Sadly, although this urgent call for action was made in 2002, there has been no significant improvement in the situation.

9.3 Research into severe ME/CFS

It should also be noted that very few of the research studies into either pathoaetiology or management of ME/CFS have ever involved severely affected patients or children – a fact that should be borne in mind when considering the general use of controversial management approaches such as CBT. As noted in section 6.4.3, a number of more severe neurological symptoms can occur in the severely affected group. Problems with swallowing may require nutritional support through either tube feeding or through a percutaneous endoscopic gastrostomy. The possibility of co-existent vitamin D deficiency should also be considered – see sections 6.6 and 7.8.4.

Research funded by The ME Association into factors that may be involved in the development of severe ME/CFS (Pheby and Saffron 2009) has shown that the type of early illness management appears to be the most important determinant of severity. Having a mother with ME/CFS was also important – possibly related to mitochondrial inheritance. Pre-morbid personality was not a risk factor, and neurotic traits were more frequent among the less severely ill. Conscientiousness overall was not related to severity.

One recent study (Pendergrast *et al* 2016), from a research group at Newcastle University, reported that people with ME/CFS who are housebound are significantly more impaired in relation to physical functioning, bodily pain, vitality, social functioning, fatigue, post-exertional malaise, sleep, pain and neurocognitive, autonomic, neuroendocrine (hormonal) and immune-system functioning than people with ME/CFS who are not housebound.

The ME Biobank at the Royal Free Hospital, which we fund, is collecting blood samples from people with severe ME/CFS in their homes – so that researchers who require blood samples and clinical details from people with severe ME/CFS will now be able to do so.

Strassheim *et al* (2017) carried out a scoping review to assess what is known about severe and very severe ME/CFS. The review highlights the limited research focusing on the severely affected ME/CFS population. The heterogeneity of the condition contributes to the lack of consensus concerning definitive diagnostic criteria and functional measures to assess disability. They concluded that focused research to understand the disease characteristics of the most severely ill will help to advance our understanding of possible phenotypes associated with distinct severity categories.

9.4 Key points from the 2021 NICE guideline on the management of people with severe and very severe ME/CFS (1.17.1 – 1.17.4)

Be aware that people with severe or very severe ME/CFS may experience the following symptoms that significantly affect their lives, including their mobility, emotional wellbeing and ability to interact with others and care for themselves:

- severe and constant pain, which can have muscular, arthralgic or neuropathic features
- hypersensitivity to light, sound, touch, movement, temperature extremes and smells
- extreme weakness, with severely reduced movement
- reduced ability or inability to speak or swallow
- cognitive difficulties that limit the person's ability to communicate and take in written or verbal communication
- sleep disturbance such as unrefreshing sleep, hypersomnia and altered sleep pattern
- gastro-intestinal difficulties such as nausea, incontinence, constipation and bloating
- neurological symptoms such as double vision and other visual disorders, dizziness
- orthostatic intolerance and autonomic dysfunction, such as postural orthostatic tachycardia syndrome (POTS) and postural hypotension.

Recognise that symptoms of severe or very severe ME/CFS may mean that people:

- need a low-stimulus environment, for example a dark quiet room with interaction at a level of their choice (this may be little or no social interaction)
- are housebound or bedbound and may need support with all activities of daily living, including aids and adaptations to assist mobility and independence in activities of daily living (for example, a wheelchair)
- need careful physical contact when supported with activities of daily living, taking into account possible sensitivity to touch
- cannot communicate without support and may need to choose someone to be their advocate and communicate for them
- are unable to eat and digest food easily and may need support with hydration and nutrition
- have problems accessing information, for example because of difficulty with screens, sound and light sensitivity, headaches affecting their ability to read, or brain fog affecting their concentration

Personal care and support for people with severe or very severe ME/CFS should be carried out by health and social care practitioners who are:

- known to the person and their family or carers wherever possible
- aware of the person's needs.

Risk assess each interaction with a person with severe or very severe ME/CFS in advance to ensure its benefits will outweigh the risks (for example, worsening their symptoms) to the person. For people with very severe ME/CFS, think about discussing this with the person's family or carers on their behalf (if appropriate), while keeping the focus of the engagement on the person with ME/CFS.

9.5 Hospital care (1.8.4 and 1.17.7)

Discuss with people who need inpatient care whether any aspects of where their care will be provided could cause problems for them, including:

- where a bed is situated on a ward – if possible, aim to provide a single room
- the accessibility of toilets and washrooms
- environmental factors such as lighting, sound, heating and smells

When planning hospital care for people with severe or very severe ME/CFS:

- discuss with the person (and their family or carers, as appropriate) what to expect when they come into hospital
- aim to minimise discomfort and post-exertional malaise during transfer to hospital, for example by planning the route in advance, avoiding noisy areas and admitting them straight to the ward on arrival

- discuss the person's care and support plan with them, including information on comorbidities, intolerances and sensitivities, to plan any reasonable adjustments that are needed
- aim to provide a single room if possible
- keep stimuli to a minimum, for example by:
    - seeing them one-to-one
    - using calm movements and gestures
    - not duplicating assessments
    - being cautious about the pressure of touch – keeping lights dimmed
    - reducing sound
    - keeping a stable temperature
    - minimising smells

9.6 Further information

The ME Association has a recently updated information leaflet on severe ME/CFS. The 25% Group is a charity that provides information and support for people with severe ME/CFS. 25% Group website:

https://25megroup.org/blog

As part of the Dialogues for ME/CFS series, two excellent videos covering the diagnosis and management of severe and very severe ME/CFS have now been released:

https://www.dialogues-mecfs.co.uk/films/severeme

Montoya *et al* (2021) have published a very useful paper on caring for people with severe or very severe ME/CFS.

Review of information on severe ME/CFS that was prepared for the NICE guideline committee:

https://tinyurl.com/32aky9sj