An open response from parents and carers of people with Severe Myalgic Encephalomyelitis to 'Management of Nutritional Failure in People with Severe ME/CFS: Review of the Case for Supplementing NICE Guideline NG206'

Background and Purpose

We are a group of parents and carers of people with Severe Myalgic Encephalomyelitis (ME). The ages of our loved ones range from mid-teens to 70s. Many of those we care for have experienced hospital care in the UK or are at risk of an emergency admission due to challenges maintaining adequate nutrition and hydration at home. Group members share strikingly similar experiences. We have read *Management of Nutritional Failure in People with Severe ME/CFS: Review of the Case for Supplementing NICE Guideline NG206* (Edwards, 2024), henceforth referred to as 'the paper', with interest and welcome the pragmatic approach in seeking to improve care for those suffering from life-threatening complications due to ME. We are offering this response as a constructive contribution.

The paper highlights the '*serious problems with nutrition*' for people with Severe Myalgic Encephalomyelitis (referred to in the paper as ME/CFS) due to difficulties with eating and drinking. It also points to the lack of provision of adequate services, misunderstandings / conflict over diagnosis and approaches to managing severe symptoms.

The paper is presented within a particular context in England where there have been several recent high-profile cases of young women with Severe ME who have been denied appropriate management of nutrition and fluids, resulting in further harms. In some cases, this has resulted in preventable deaths.

Whilst we welcome the paper's recommendations, our group suggests several additional factors not included in the preprint, which we see as critical to the analysis of the issues highlighted. This includes the need to acknowledge the growing body of scientific and clinical evidence explaining the underlying pathology in patients with Severe ME. We believe this to be critical to countering the misinformation and bias that often influences clinical and managerial decision-making about patients with this condition.

Acknowledging the biomedical evidence

Whilst the paper seeks to find a middle ground, it does not reference the biomedical evidence relating to ME. Although we appreciate the need for pragmatism in improving care for patients, we feel there are risks inherent in adopting an approach which seeks to find a middle ground between settled science and the misinformed beliefs of some health care workers and managers. Indeed, without acknowledgement of the biomedical evidence and a recognition of appropriate clinical expertise (e.g. Bateman et al. 2021; Montoya et al. 2021; Snell and Stevens, 2018, Leslie et al. 2023), it is unlikely that there will be a middle ground that patients and carers can safely accept.

Despite the prevailing notion of *'limited evidence', 'speculative diagnoses', 'unknown mechanisms'*, etc., decades of research (albeit poorly funded) have contributed to the understanding of ME, with a significantly expanding literature since Covid-19. Pertinent examples from this year alone include: Appelman et al. (2024); Armstrong et al. (2024); Chen et al (2024); Eastman (2024); Keller et al (2024); Peluso et al (2024); Pretorius et al. (2024); Ryabkova, et al (2024); Walitt et al. (2024). The common pathophysiology between post-COVID syndrome and ME is now well-described, with Arron et al. (2024) providing a cohesive model which calls for a holistic approach to diagnosing, researching, and treating '*one of the most misunderstood conditions in modern medicine'* (Arron et al. 2024).

As recognised by the National Institute for Health and Care Excellence (NICE), (National Institute for Health Care and Excellence, 2021), Myalgic Encephalomyelitis is indisputably a biomedical condition for which Graded Exercise Therapy (GET) and variations thereof are harmful. Keller et al (2024) spotlight the '*robust evidence of disrupted immune function, neural activity, and metabolic function*' and '*clinical evidence of impaired energy production and recovery'*.

For severe patients (an estimated 25% of the total ME patient population), simply rolling over in bed can exceed their lowered anaerobic threshold. These patients are often extremely sensitive to light, noise, movement, touch, and smells, and are orthostatically intolerant, often on account of reduced blood volume (Newton et al., 2016; van Campen et al., 2018). Many are bedridden constantly, tube-fed (Baxter et al., 2021; Rowe et al., 2017) and require total care i.e., 0% on the Bell Disability Scale (Bell, 1995).

The impact on the functional status of the patient with Severe ME is immense and results in a quality-of-life score that has been recognised to be lower than all of the major diseases to which it has been compared (Hvidberg et al 2015; Nacul et al 2011; Vyas et al. 2022). At its most severe, ME can be fatal as evidenced in the cases of, for example, Merryn Crofts, Sophia Mirza, Maeve Boothby O'Neill, Emily Collingridge, Brynmoor John and Kara Jane Spencer.

As identified in the paper, the key diagnostic symptom of ME is PESE / PEM (Post Exertional Symptom Exacerbation / Post-Exertional Malaise), an often-delayed exacerbation of all symptoms when the patient exceeds the lowered anaerobic threshold (Weir and Speight, 2021; Snell et al., 2013; Keller et al., 2014) that is characteristic of the disease. What is often overlooked is that this includes physical (including eating and drinking), emotional, and cognitive activity (Twisk, 2015; Holtzman et al., 2019; Hartle et al., 2021; Chu et al., 2018).

As carers, we strongly believe that the onus should be on acknowledging what is now known and knowable. This alone would strongly refute the notion of a need for a middle ground and would instead focus attention on the need to generate solutions from both evidence-based practice and practice-based evidence for managing symptoms, not only in relation to nutritional failure, but also (but not limited to): orthostatic intolerance, dysautonomia, hypovolemia, autoimmunity, sleep disorders, fluid balance, gastroparesis, mast cell activation, and pain.

Implementation of NICE Guideline [NG206] and Focus on Clinicians' Judgment

The report '*Patchy, Misunderstood, and Overlooked'* (Action for ME, 2023) identified that only 28 per cent of NHS trusts and integrated care boards had implemented the NICE Guideline [NG206]. This figure resonates with the reported lived experience of patients and their carers. In this context, the suggestion in the paper that patients can trust '*the judgment of health care professionals with responsibility for resolving these difficult problems'* is currently unpalatable, especially to those who are at the greatest risk of harm.

While clinicians often rely on their clinical judgment, there is a high risk of cognitive bias in the current context for ME patients as this judgement is highly likely to be informed by misunderstandings about the condition and limited familiarity with NICE's evidence-based guidance. The case histories of patients who have been harmed due to clinicians following their judgment and not engaging with evidence-based expertise on managing ME do not reassure patients and their carers (McPhee et al.2021; Tillman, 2018; Invest in ME Research, 2006). They also represent a serious institutional concern from the perspective of patient safety. This is reinforced by Dr Anthony Hemsley, Medical Director of the Royal Devon & Exeter Hospital, revealing in written evidence to the coroner for the inquest into the death of Maeve Boothby O-Neill that there were no commissioned specialist services for ME locally, regionally or nationally. Dr Hemsley has stated that this needs to be rectified and '*action is required at the highest level'* (Humphries, 2023)

Due to the widespread failure to implement the NICE Guideline [NG206], people with ME are still regularly subjected to harmful practices and inappropriate investigations and diagnoses / formulations, which would be prevented through adherence to the NICE Guideline. A lack of training, limited awareness and bias among medical professionals create a reciprocal network of compounding factors that effectively disenfranchise the most vulnerable ME patients. Cases where things have gone badly wrong for patients with Severe ME accessing healthcare point to various recurrent themes. Cognitive bias, unconscious incompetence and a disregard for the NICE Guideline feature prominently.

Despite a plethora of established biomedical evidence, harmful notions based on flawed science about ME being psychologically or socially derived (e.g. deconditioning, fear avoidance, false sickness beliefs, catastrophising, hypervigilance, etc.) remain prevalent.

Cognitive bias on the part of clinicians and health care workers too often adversely impacts decision-making and ultimately compromises patient safety and well-being. In turn, this also creates insurmountable barriers to appropriate support for patients with severe ME. Director of ME Action, Jaime Seltzer, is clear: '*Hospital is seldom the best place for these patients'* (Seltzer, 2024). Fundamentally, she argues that the most dramatic improvement for Severe ME patients would be access to hospital at home services including home saline infusions ('*which can often really help with a lot of the different symptoms that are triggered in a severe episode of PEM, including MCAS and orthostatic intolerance'*). In most circumstances, domiciliary care would enable patients to remain within their own environment, thus minimising PEM / PESE.

The paper focuses on nutritional management. However, there is also overwhelming evidence from the lived experience of patients with Severe ME that maintaining appropriate hydration /

fluid balance constitutes an equally important challenge. Patients with ME have not only been denied appropriate nutritional management but have also been denied appropriate management of fluids. The Health and Social Care Act 2008 (Regulated Activities) outlines requirements to ensure that the nutritional and hydration needs of patients are identified and met (Care Quality Commission, undated). The NICE Guideline CG32 (National Collaborating Centre for Acute Care, 2006) addresses oral nutrition support, enteral tube feeding and parenteral nutrition for malnourished or 'at-risk' individuals in hospitals, care homes or their own homes.

Due to misinformation and bias about this condition, patients with severe ME are particularly vulnerable to erroneous and therefore unhelpful psychiatric / psychological formulations relating to refusal, withdrawal and avoidance (Baxter et al, 2021). This has often led to a distorted view of the situation and created trauma, delayed / denied basic care and resulted in patient harms and even fatalities, as in the case of Sophia Mirza (Invest in ME Research, 2006). Rowe et al (2017) highlight that a '*lack of familiarity with the clinical diagnostic features*' and *'skepticism about its existence'* have led to children and young people with Myalgic Encephalomyelitis being misdiagnosed. Typical misdiagnoses listed by the authors include: Fabricated or Induced Illness (FII), school refusal, Pervasive Refusal Syndrome (PRS), and somatoform disorder. Where children and young people are severely affected, patients and their families are often treated with suspicion by professionals who are not familiar with Severe Myalgic Encephalomyelitis and subjected to harmful narratives and ill-informed safeguarding referrals. In some cases, children have been removed from the care of their parents (Weir and Speight, 2021).

We believe our group has a unique vantage point and first hand lived experience from which to view this phenomenon. It is our collective view that there is a dangerous lack of understanding and awareness of the underlying pathology, especially in relation to Severe ME in NHS hospitals, which leaves patients vulnerable to harm and serious misunderstandings. People with ME and their carers have experienced decades of marginalisation, discrimination, neglect, and often even a lack of compassion and understanding from some of the people they turn to for help. Once again, we very much welcome this contribution that spotlights managing nutritional failure in patients with ME and would ask that the barriers to this and fluid management are scrutinised more closely. We agree with Dr Hemsley that action is indeed required at the highest level to address the need for informed, patient-led and safe care for the loved ones we care for, many of whom currently live with a rational fear of hospital admissions.

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