Pioneering National Nutrition Standards

How one dietitian is blazing a trail for the provision of nutritional care for sickle cell patients

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Nearly 4 years following the publication of the first article on nutrition in sickle cell disease (SCD) in *Dietetics Today*,[1] the first ever national nutrition standards in sickle cell disease (SCD) were published as part of the 2nd Edition (2018) of the Sickle Cell Society's 'Standards for the Clinical Care of Adults with Sickle Cell Disease in the UK'.[2] This is a significant victory and recognition of the importance of nutrition in the management of SCD and, more importantly, promoting the role that the dietitian has to play as the recognised nutrition and diet expert in this emerging speciality. The publication of nutrition standards in SCD is widely welcomed by the international sickle cell community.

What is sickle cell disease?

SCD is the most common and fastest growing[3] autosomal recessive genetic blood disorder in the UK and around the globe.[4] In SCD, haemoglobin (Hb), the oxygen carrying molecule present in red blood cells, is defective and when deoxygenated becomes rod like, deformed and sickle shaped. It is characterised by a range of pathophysiological consequences, including the following: defects in the structure and function of haemoglobin, the integrity of red blood cell membranes, the density of erythrocytes, endothelial activation, microvascular tone, inflammatory mediators, and coagulation.[5]

The main clinical features of SCD can be grouped into four broad categories: anaemia and its sequelae; vaso-occlusive crises and bone marrow fat embolisation syndrome; infection (from functional asplenia); and organ dysfunction.[5] A host of complications result from these clinical features and include stroke, retinopathy, nephropathy, liver disease or pulmonary arterial hypertension. SCD is considered as a multisystem condition, which involves the main organs and systems of the body, including the skeletal, genito-urinary, gastrointestinal, spleen, hepatobiliary, cardiopulmonary and central nervous systems.[6, 7]

The hallmark clinical feature of SCD is the acute vaso-occlusive event or painful crisis,[8] which is the main reason for A+E visits and hospitalisations.[9, 10] Sickle crisis is also a measure of disease severity and predictor of early death in adults.[11] There are a wide range of factors which precipitate a sickle cell crisis, including: hypoxia, acidosis, dehydration, infection, extreme fatigue, trauma, temperature changes (sudden), stress/anxiety and increase physical/physiological demand, such as pregnancy and physical exercise.[7, 12] In most cases, sickle crisis requires immediate hospitalising and the main treatment modalities include anti-inflammatory drugs, non-steroidal analgesics, hydroxyurea, opioid analgesia, rehydration and in severe cases transfusion,[13] which may cause other long-term side effects.

Why the need for the national nutrition standards?

The findings from a cross-sectional survey conducted amongst dietitians in 2015, exploring their involvement, knowledge and attitudes towards SCD, identified that the primary reason for the lack of dietetic involvement in SCD was related to the absence of nutritional guidelines and standards of care, and the poor knowledge and understanding of the nutritional implications of SCD.[14] This gives significance to the need for the nutrition standards in SCD, as a means of providing dietitians with information and evidence to support their dietetic management and involvement in SCD.

The nutrition standards[2] also address a key oversight, the gross lack of translation of existing scientific literature into the role of nutrition in SCD. The resulting paucity of nutritional evidence base in SCD, is a fundamental cause for the lack of nutritional service provision in SCD. Moreover, the lack of nutritional evidence base, despite the plethora of existing research in SCD, has resulted in poor recognition of nutrition as a viable treatment option for SCD, even though nutrition is identified to be a part of the standard clinical management of SCD.[15]

What are the national nutrition standards for sickle cell disease?

SCD Nutrition Standard 1: Dietitians should be included in the multi-disciplinary team caring for patients with SCD.[2]

Significantly, the nutrition standards are a public acknowledgement from within the sickle cell community that nutrition has a role to play in the management of SCD and, therefore, the health and wellbeing of the sickle cell patient population. This is a major milestone in changing the nutritional management in SCD. The standards therefore aim to reform the nutritional landscape in SCD and recognises the role that the dietitian must play in effectively managing the nutritional needs of the SCD patient population in the UK and, therefore, also globally. The dietitian should be an active participant of the MDT caring for sickle cell patients. As a result, dietitians can be a 'visible force' and an 'audible voice', playing an active role in developing the nutritional management of vulnerable SCD patients.

SCD Nutrition Standard 2: Patients with SCD should be screened for malnutrition/risk of malnutrition by healthcare professionals with appropriate skills and training (National Institute for Health and Care Excellence, 2012a).[2]

Screening patients with and at risk of disease-related malnutrition (DRM) is a recognised and essential part of the management of DRM. The standards recognise the importance of DRM screening being available to SCD patients. More importantly, sickle cell patients identified to be at risk of DRM should be referred to a dietitian, who is the only qualified professional able to assess, diagnose and treat diet and nutrition problems at an individual and wider public health level.[16] Dietitians have the responsibility to comprehensively assess the nutritional needs of patients, make accurate nutritional diagnosis and provide patients with a nutritional care plan. However, the standards also highlight the multifactorial nature of SCD and the need for dietitians to consider the wider determinants of health and the psychosocial factors that may affect the health and wellbeing of sickle cell patients.[17]

SCD Nutrition Standard 3: Patients with SCD who have been identified as high risk for malnutrition should be offered a nutritional assessment by an appropriately (SCD) trained dietitian and should receive a nutritional management care plan.[2]

More importantly, the standards recognise the nutritional needs of sickle cell patients to be more than just healthy eating. Healthy eating has been portrayed as the main nutritional need of sickle cell patients over the years, a direct consequence of the poor translation of the findings of previous research which has identified undernutrition as a real consequence of the pathophysiology of SCD. Undernutrition is synonymously used with malnutrition[18] and the standards acknowledges and recognises the real risk of DRM,[19] which potentially threatens the health and wellbeing of sickle cell patients both within the hospital and the community. BAPEN highlights the disease and cost burdens associated with under recognised and under treated DRM.[20]

Background evidence considered for the development of the nutrition standards

The standards briefly draw attention to some of the key specific nutritional aspects contributing to the DRM observed in SCD, which are a consequence of the pathophysiology of the condition.[2] SCD has been theorised to produce a form of 'protein energy malnutrition'.[15] This is not solely due to poor intake but to an increased energy demand because of a high protein turnover and high energy expenditure,[15] caused by chronic haemolytic anaemia. Chronic haemolytic anaemia is one of the main clinical features of SCD, due to the shortened lifespan of sickle red blood cells. (16-20 days lifespan compared to 120 days for normal red blood cells).[3] The impact of chronic haemolytic anaemia results in decreased red cell count, chronic anaemia and fatigue and increased myocardial demand.[15]

In keeping with the impact of haemolytic anaemia on energy demand, direct measurements of high protein and energy metabolism, despite adequate dietary intakes, has led to the theory of a relative shortage of macro- and micronutrients in SCD, which is required for normal growth and development.[21] Hypermetabolism in SCD has gained traction as a theory. Hypermetabolism is characterised by a state of increased energy demand due to a higher rate of catabolism (nutrient breakdown) and anabolism (nutrient build up) and in SCD there is a shift towards catabolism – leading towards an increased nutrient demand.[22, 23] Furthermore, individuals with SCD have a higher resting energy expenditure (REE) attributed to an increase in myocardial energy demand and the production of proinflammatory cytokines.[24] Therefore, when compared to age-and sex-matched healthy controls, individuals with SCD have a higher REE.[24-26]

Low plasma levels of zinc observed in individuals with sickle cell is associated with delayed growth and sexual maturation.[27] It is also associated with low levels of serum testosterone in males[28] and decreased pubertal development in general.[27] Providing micronutrient supplements to individuals with SCD has led to improvement in growth and maturation by way of improved testosterone levels.[15] Supplementing other micronutrients, like vitamin A,[29] vitamin B and magnesium,[30] has resulted in improved growth, decreased hospital emergency room visits, decreased frequency of pain crisis, and reduced frequency of infection. Micronutrient supplementation has also led to improvement in muscle function, cognition and coordination, decreased inflammation, and improved antioxidant and anaemia status.[15]

Supplementation studies with omega-3 fatty acids also need consideration. One of the first studies using omega-3 fatty acids showed reductions in inflammation, oxidative stress, red cell density and pain episodes.[31] A more recent study[32] reported that docosahexaenoic and eicosapentaenoic acid supplementation prevents vaso-occlusive and haemolytic crises in patients with the severe form of sickle cell. However, the authors recommend that conducting a larger, multicentre, study is needed to further prove that omega-3 fatty acids are a safe, effective and affordable treatment for SCD.

Implications of the standards

As seen above, the nutritional needs of sickle cell patients are complex and there is still a long way to go to optimise the specific nutritional management of the DRM observed in SCD. However, the SCD nutrition standards take us a step closer to developing tailored guidelines, based on the research which has already been done over the last 60 years. Therefore, the standards highlight the urgency to comprehensively translate the findings from the existing scientific literature into a meaningful evidence-base that can be used to improve the nutritional management of SCD patients in the UK and worldwide. My Doctoral research project seeks to add to the nutritional evidence base in SCD. My research is aimed at integrating nutrition into sickle cell healthcare provision using a health education intervention. The research project uses a Learning Alliance Methodology,[33] to form a nutrition alliance between sickle cell service users and service providers/stakeholders, with the aim to co-develop a whole systems management strategy to improve the knowledge, awareness and understanding of the nutrition needs of sickle cell patients. The research is positioned as a health promotion project and uses a socio-ecological approach[34] to comprehensively understand the factors influencing the nutrition needs of SCD patients.

The nutrition standards have therefore become a vehicle to improve the knowledge and awareness of the nutritional implications of SCD, not only for healthcare professionals, but healthcare providers, stakeholders and the sickle cell patients themselves. The international sickle cell community stand to benefit from it too. The nutrition standards form part of the 'National Clinical Standards of Care for Adult sickle Cell Patients in the UK', and are available from the Sickle Cell Society's website:[2] www.sicklecellsociety.org/wp-content/uploads/2018/05/Standards-for-the-Clinical-Care-of-Adults-with-Sickle-Cell-in-the-UK-2018.pdf.

Next steps

In addition to the nutrition standards, a few key recommendations are included to support the implementation of the standards to a wider audience.[2] The aim is to further promote and raise awareness of the specific nutritional implications of SCD and the importance of including nutrition in all contact with SCD patients. It also emphasises the important role research plays in building the evidence base required to support nutrition assessment, diagnoses and treatment tailored to the needs of the sickle cell patient population. Below are the recommendations.

Overall recommendations[2]

1. *The British Dietetic Association (BDA) should consider the development of a sickle cell specific nutritional risk assessment tool to allow consistent dietary assessment.*
2. *Sickle cell centres could consider offering an annual review with a dietitian to advise on optimal nutrition.*
3. *Health and social care practitioners should be aware of how diet can enhance healthy living and general well-being among patients with SCD.*
4. *Further research into the role of nutritional support for patients with SCD is needed.*

Moving forward

There is a lot more that should be done, but without a scientific evidence base change is impossible. Dietitians, however, stand to make a significant contribution to developing nutrition service provision in SCD in the UK and globally. It will require ongoing attempts to bring SCD under the radar of health planners and authorities to increase the knowledge and awareness of the DRM needs associated with SCD, as the disease and cost burden of SCD continues to rise.

Therefore, the commissioning and inclusion of the first ever national nutrition standards for SCD is a personal victory on behalf of the dietetic profession. The nutrition standards not only acknowledge the nutritional implications of SCD but acknowledges the important role that the dietitian plays as the go to diet and nutrition expert. As such, we must capitalise on this opportunity to lobby for more funding opportunities to research and develop the nutrition service provision in SCD. Together we can be the difference, that makes the difference.

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