

Nutritional Implications of Sickle Cell Disease



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Nutrition in sickle cell disease (SCD) remains greatly underused and under-recognised as a management strategy for the condition; a lack of understanding of the nutritional implications of the condition was identified in a recent national survey¹ as one of the main reasons why dietitians do not have a greater involvement in SCD.

SCD, a genetically inherited red blood cell disorder, affects the nutritional status of, particularly, Haemoglobin SS (HbSS) patients, the severe form of the condition. Slowed growth, delayed sexual maturation and poor immunological function are some of the physiological manifestations.²

Furthermore, SCD is characterised by high non-elective admission rates and extensive length of stay (LOS). These factors provide additional incentives in support of raising the level of nutritional input/involvement from dietitians, along with the evidence that shows improved nutritional input has been reported to speed up recovery and decrease LOS. It is well-known that non-elective admission rates observed in SCD places a high financial burden on NHS resources to the tune of £16.2 million in 2010/11, with 11,109 admissions for painful crisis as the main reason for admission.³

Undernutrition a critical feature of SCD

Many complications associated with this disease have a nutritional underpinning and undernutrition, identified as a critical feature of SCD, is considered as a serious complication of the disease that should be managed as part of the required clinical care.² Malnutrition can be acute or chronic and is characterised by a deficiency or imbalance of energy, protein, and other nutrients causing measurable and adverse effects on body composition, function, and clinical outcomes.^{4,5} Under nutrition refers to a state of nutrient deficiency which is connected with adverse consequences on physical functions or clinical outcomes and results from an imbalance among nutrient intake and nutrient needs. A number of factors affect its severity and its impact on clinical outcomes, which include the following:⁶

- Difference between energy intake and expenditure
- Nutritional status and energy reserves at the onset of undernutrition

- Extend of adaptive processes to undernutrition
- Potential incidence of stress response (e.g. inflammation, surgery) during a period of undernutrition.

Interestingly, in a review of the role of nutrition in SCD,² a number of proposed mechanisms for undernutrition have been identified, including:

- Protein hypermetabolism
- Decrease dietary intake possibly due to interleukin-6-related appetite suppression
- High cardiac demand/expenditure
- Increased red cell turnover.

In considering the myriad of clinical consequences of malnutrition, which includes: impaired immune response, reduced muscle strength, impaired wound healing, impaired psycho-social function, impaired recovery from illness and surgery, and poor clinical outcomes,⁷ a clear parallel can be drawn between the clinical and nutritional consequences of SCD based on the pathophysiology of the condition, which will be discussed later in the article.

A number of contributing factors leading to under nutrition are identified; the two main factors being disease and social and psychosocial factors.⁶ Failing to consider the multifactorial nature of SCD would, therefore, be ludicrous; the nutritional risk of this patient population is hugely affected by the wider determinants of health and the psychosocial impact it has on the individual patient. In a recent article, published in the July/August edition of CN Magazine,⁸ four main components to managing nutrition in SCD were identified. In addition to the social and psychosocial considerations, there are specific nutritional (infection risk, dehydration, frailty and gastrointestinal intolerances) and medical considerations (hydroxyurea, transfusion treatment, iron overload) which impact the nutritional risk of SCD patients.⁸ This makes a compelling case in favour of the urgency to develop services to effectively manage the nutritional implications of SCD.

As a consequence of nutrition in SCD being greatly underused and under-recognised, a large proportion of sickle cell patients in the UK are left vulnerable. To date, there are no specialist dietetic services available to this patient population which directly impact patient outcomes, in particular, younger adults transitioning from paediatric services.

Pathophysiology explained

The hallmark clinical features of SCD are increased red cell turnover (haemolysis) and vaso-occlusion. Sickle red blood cells have a shortened lifespan of 16-20 days as opposed to 120 days for normal red blood cells, resulting in a chronic anaemia.⁹ Chronic anaemia is manifested in the following presentations: tiredness, irritability, dizziness and light-headedness, fast heart rate, difficulty breathing, pale skin colour, jaundice, slow growth, and delayed puberty.¹⁰

Vaso-occlusion, caused by sickle shaped red blood cells blocking small blood vessels, results in tissue infarction and unpredictable painful crisis. Endothelial damage is caused

by repeated episodes of vaso-occlusion, complicated by vasoconstriction due to nitric oxide (endogenous vasodilator) deficiency. This results in damage to the large blood vessels, leading to a number of complications affecting a wide range of organs and systems of the body,⁹ including the skeletal, genitourinary, gastrointestinal, spleen, hepatobiliary, cardiopulmonary and central nervous system.¹¹

The clinical manifestations of SCD, as a result of the ongoing tissue infarction and endothelial damage in blood vessels, are quite variable and the complications associated with these affected organs and systems make the disease very complex to manage medically and, therefore, nutritionally. It goes without saying that the clinical manifestation of SCD, due to its complex pathophysiology, places SCD patients at high risk of malnutrition with its associated nutritional problems and financial burden on NHS resources.

The dietitians role

As dietitians we are governed by an ethical code and compelled to work within our individual clinical expertise, level of competence and capability. However, SCD presents a host of complex nutritional problems and complications owing to the multifaceted nature of its pathophysiology and multi-organ and system involvement (see **Table One**). **Table One** further illustrates the wide ranging clinical manifestations that a SCD patient can present with, including any of the following: increased risk of infection (particularly streptococcal infection), neutropenia (resulting from hydroxyurea treatment), inflammatory symptoms, stroke, to longer term problems including renal failure, liver failure requiring liver transplantation, cardiac failure due to cardiac iron overload and chronic lung disease. All these potential problems coexist on the background of chronic anaemia and vaso-occlusion precipitating the onset of unpredictable acute or acute on chronic painful crisis, requiring prompt hospitalisation.¹³

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Table One: Main Clinical Manifestation of affected Organs and Systems¹²

System/Organ	Clinical Manifestation/Complication
Skeletal	Osteonecrosis, Osteomyelitis, leg ulcers, avascular necrosis
Genitourinary	Chronic renal insufficiency, priapism, chronic renal failure
Gastrointestinal	Cholelithiasis, viral hepatitis from transfusion, liver failure
Spleen	Splenic enlargement and fibrosis, acute aplastic anaemia, parvovirus B19, functional asplenia, leucocytosis
Hepatobiliary	Indirect hyperbilirubinemia
Cardiopulmonary	Pulmonary hypertension, cardiomegaly, cardiac failure
Central nervous	Stroke, silent infarcts

Yet, in reality, many SCD patients are managed by dietitians who often lack the knowledge and understanding of the complex nature of the condition; a recent survey found that 84% of responding dietitians agreed that they have limited knowledge about the nutritional needs of SCD patients.¹ Currently, SCD remains an untapped specialism in the dietetic profession. Dietitians, though, are nutrition experts! By definition, dietitians are qualified health professionals who assess, diagnose, and treat diet and nutrition problems at individual and wider public health level.¹⁴ Importantly, they help service users achieve their health-related aspirations using leadership skills and evidence-based practice.¹⁴

A 'specialist' dietetic approach!

Notwithstanding, the effective management of SCD is NOT straightforward; it is complex and multifactorial and therefore requires a specialist dietetic approach. The breadth and extent of the clinical, medical and nutritional factors which constitutes this long-term condition is so extensive that it deserves not only a specialist approach, but more recognition, if we are to do justice to the patients. I wonder if it's time to reconsider the merit of SCD patients being managed by a dietitian with a specialist knowledge and understanding of the condition.

Looking at the Model and Process for Nutrition and Dietetic Practice,¹⁴ we as dietitians, have a responsibility to assess our patients comprehensively in order to elicit

the facts to make an accurate nutritional diagnosis. Once a nutritional diagnosis is made, in addition to having a thorough knowledge and understanding of the complexities of the specific condition, we need access to evidence-based guidelines, requirements and resources, which are sadly non-existent with regards to SCD. The only conclusion to make is that we are managing a complex specialist condition with a general dietetic approach!

High resting energy expenditure, poor immunity, high protein turnover, low body mass index and poor exercise tolerance are all factors which increase the nutritional requirements of SCD patients.² Moreover, current national standards of care for the clinical management of adult SCD patients does not include nutrition at all, but that is about to change! I have just been invited to be a contributor, as part of the overall review of the 2008 national SCD standards,⁷ to lead on the development of the nutrition/dietetic standards of care. This will include improvements to the availability of SCD specific nutritional resources available for both healthcare professionals and patients.

Change is on the horizon

Pioneering work is underway, aimed at developing nutrition services to improve the nutritional management of SCD patients in the UK. Much needed SCD specific nutritional guidelines, standards of care and resources are being planned and developed. The acceptance of two nutrition in SCD abstracts, for poster presentation at the 9th Annual Guys and St Thomas's Sickle Cell and Thalassaemia

Advanced Conference in London,¹⁵ gives recognition to the developmental initiatives underway.

One of the key scoping projects undertaken was a national cross sectional survey, aimed at exploring the involvement, knowledge and attitudes of dietitians towards SCD in the UK.¹ Despite the low response rate, the main findings of the survey provided pertinent themes in support of recommendations to be made for the commissioning of specialist dietetic services for SCD. The survey brought to light a number of invaluable insights explaining the lack of involvement of dietitians in SCD. The two main reasons being: 1) A lack of understanding amongst dietitians of the nutritional implications of SCD; and 2) The lack of SCD specific nutritional guidelines and resources. Poor referral rates requesting dietetic input for SCD patients have also been highlighted in the survey.

It is early days yet, but without the full backing of the dietetic profession and support from the British Dietetic Association, as well as support from the wider SCD community and associated government departments (e.g. Specialist Commissioning), patients living with SCD in the UK will not be able to capitalise on the full 'worth' and 'capability' of the 'specialist expertise' dietitians have to offer to enhance the nutritional outcomes of this patient population.

Please feel free to contact me on email: claudine.matthews@homerton.nhs.uk should you wish to offer support, or if you have any questions or comments.

Supporting those with sickle cell disease

Sickle Cell Society - <http://sicklecellsociety.org>

The Sickle Society is a registered charity which was set up by a group of patients, parents and health professionals who were concerned about the lack of understanding and treatment for sufferers of sickle cell disorders. Claudine has recently written a guest post for the Society, visit:

<http://sicklecellsociety.org/nutrition-in-sickle-cell-underused-and-under-recognised>

The Homerton Hope - www.homerton.nhs.uk/about-us/homerton-hope-charity

Homerton Hope is Homerton Hospital's Charitable Fund, one of their goals on their 'wish list' is to generate funding for the Sickle Cell unit to offer drop-in alternative therapy sessions like reflexology, acupuncture and massage for patients, along with day trips for the elderly patients.

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