



Understanding the contribution of sickle cell and thalassaemia specialist nurses: a summary report

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Professor Elizabeth Anionwu & Dr Alison Leary



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Introduction

This report highlights the vital role that specialist sickle cell and thalassaemia nurses play – both in the community and in acute care – in the management of these complex, multisystem conditions. It draws on evidence from a specially-commissioned project that explores current provision and considers its value.

For the first time, information is available to commissioners, managers and nurse leaders to take forward the debate. We are very grateful to Roald Dahl's Marvellous Children's Charity for making this possible by funding the project.

This is a summary of a longer report, which includes detailed references. See back page for more information.

What are sickle cell and thalassaemia?

Sickle cell and thalassaemia are genetic blood disorders, passed on by parents to children. Over 650,000 people in England are healthy carriers of haemoglobin variants, of which sickle cell and thalassaemia are the most common. When two healthy carriers have a baby together, their child has a 1 in 4 chance of inheriting a full disorder.

Sickle cell

Sickle cell is the most common serious genetic disorder in England – more common than cystic fibrosis or haemophilia. It affects an estimated 12,500 people and causes acute episodes of severe pain (sickling crisis), tissue damage and serious long term complications such as stroke.

Thalassaemia

About 800 people in England are living with thalassaemia. It affects the body's ability to make red blood cells, and patients must undergo blood transfusions every few weeks to reduce the risk of organ damage, restricted growth, liver disease and heart failure. They must also comply with a challenging regime to filter excess iron from their blood.

As with all long term conditions, sickle cell and thalassaemia can be psychologically and socially very challenging to patients and their families.

Who gets sickle cell and thalassaemia?

Sickle cell and thalassaemia mainly tend to affect certain ethnic minority communities. This is because being a healthy carrier of unusual haemoglobin may help protect against malaria in childhood, so in places where malaria has been widespread, the genes have become more common – including countries around the Mediterranean (like Cyprus, Turkey, Italy and Greece), Africa, the Caribbean, the Middle East, parts of India, Pakistan, south and south-east Asia. People whose ancestors are from these areas are more at risk of inheriting one of these conditions.





Understanding the contribution of sickle cell and thalassaemia specialist nurses

Over the last three decades, great strides have been made in the provision of expert, multidisciplinary care for people with sickle cell and thalassaemia (SC&T) – from the world's first linked antenatal and newborn screening programme, to dedicated health professionals supporting service users of all ages and at every stage of the care pathway.

At the heart of this provision are SC&T specialist nurses, who are ideally placed to meet and manage the complex needs of people living with these challenging disorders.

In England, there are two main groups of specialist nurses: one community based and one acutely based. They perform a range of activities including genetic counselling, disease monitoring and symptom control, psycho-social interventions, family support, information and referral to appropriate agencies – caring for paediatric and adult patients in all settings. A rapid analysis of nurses included in the 2012 'STAC* list' suggests that there are around 117 SC&T specialist nurses in England including 76 counsellors, 39 acute and 2 combined counsellor-acute posts. With 41 SC&T nurses covering the acute sector in England, this equates to a caseload per nurse of 325. About 55% of specialist nurses are based in Greater London and 45% outside London.

*STAC: Sickle Cell & Thalassaemia Association of Counsellors

An evidence-led approach

In March 2011, the All Party Parliamentary Group for Sickle Cell & Thalassaemia (APPG) held a meeting to discuss current economic constraints and the implications for specialist nursing posts. It was agreed that a detailed exploration of their value was required in the context of commissioning sustainable, high quality care.

A project was commissioned to identify the contribution of sickle cell & thalassaemia specialist nurses in acute and community settings. The project was designed to build on existing work assessing the contribution of specialist nursing; to incorporate existing standards and frameworks in sickle cell and thalassaemia care and treatment (NHS Sickle Cell & Thalassaemia Screening Programme 2012); and to take full account of patients' views.

A number of techniques such as complex activity analysis, in depth interviews and a review of the available literature were used to gather data which was subsequently mined.



"My son goes every 4 weeks for a transfusion. We know the specialist nurse and she knows my son, and if the nurse is there it all goes smoothly."

"I have a specialist nurse who I usually see every 4 weeks ... I get a very good response from my nurse and she is available by phone."



Findings

Specialist nursing practice

In total, the project captured over 8,966 nursing events undertaken by 22.6 whole time equivalent SC&T specialist nurses – equating to over 1,639 hours – just under half of which took place in acute settings and the remainder in community settings. The ratio of clinical to non-clinical/clerical work was 80:20 in acute settings and 74:26 in community settings.

Nurses were engaged in a wide range of clinically complex, interrelated activities – including symptom control; managing disease crises; addressing psychosocial issues; directing ward rounds; nursing assessment and complex care planning; supporting and educating other professionals; leading new and innovative services such as exchange transfusion; administrative tasks and making referrals (figures 1 – 4).

The expertise and flexibility of SC&T specialist nurses is particularly significant. They manage caseloads throughout the patient journey and at all stages of life – antenatal, paediatric, teenage, adult and older people. Moreover, sickle cell conditions are particularly unpredictable, with unexpected sickling crises often occurring at night. Although types of nursing intervention may be similar to those for other long term conditions, patterns of activity are very different.

Sickle cell and thalassaemia impact on patients’ lives and the lives of their families in many different ways. In addition to physical symptoms, they face increased risk of anxiety, biographical disruption and distress. Specialist nurses place a strong emphasis on helping patients to become experts in their own condition and on

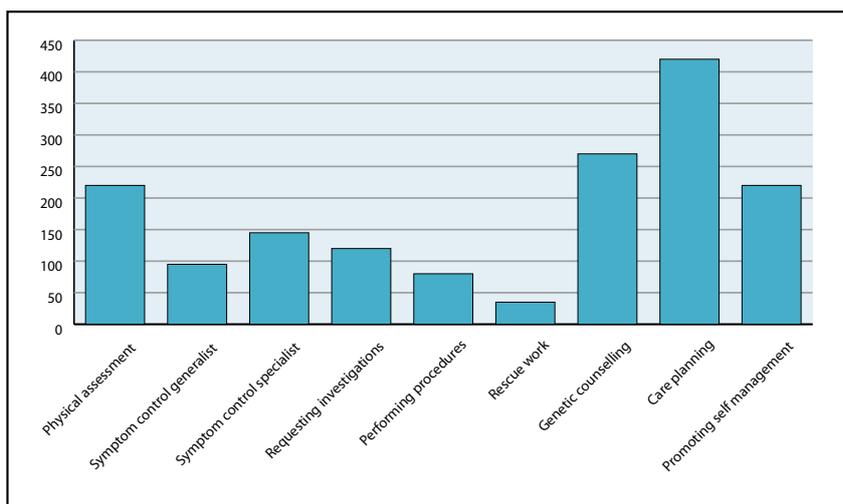


Figure 1: the distribution of activity in physical interventions (community) n=1598 nursing events (33% of total for community group)

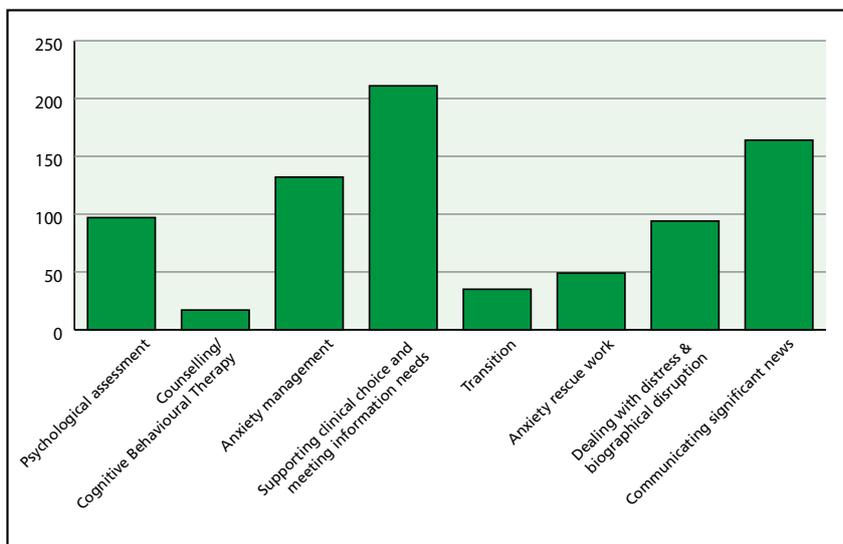


Figure 2: the distribution of activity in psychological interventions (community) n=799 events (13% of total for community group) (Note: ‘communicating significant news’ was mostly around delivering the diagnosis of a disorder)



teaching effective techniques for self management. Helping parents and children to build up their confidence early on encourages a positive approach from the start, which in turn can reduce the number of hospital admissions and visits to A&E. This has been reflected in local studies on admission avoidance.

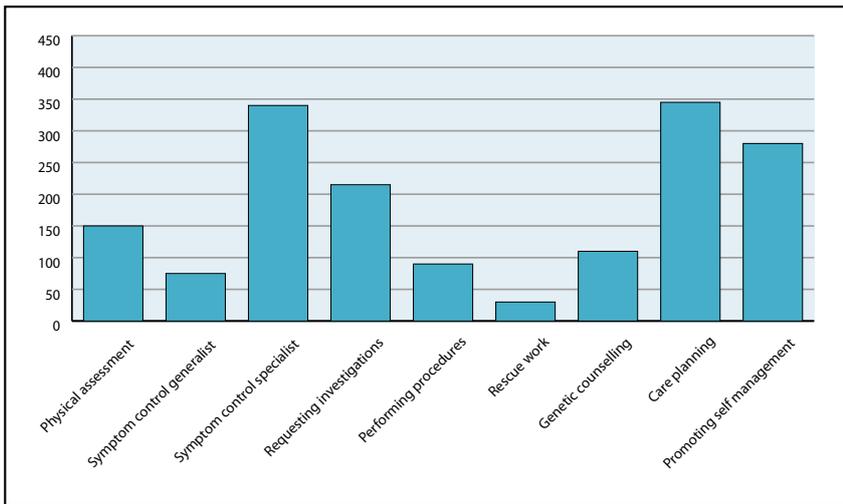


Figure 3: the distribution of activity in physical interventions (acute) n=1624 events (42% of total for acute group)

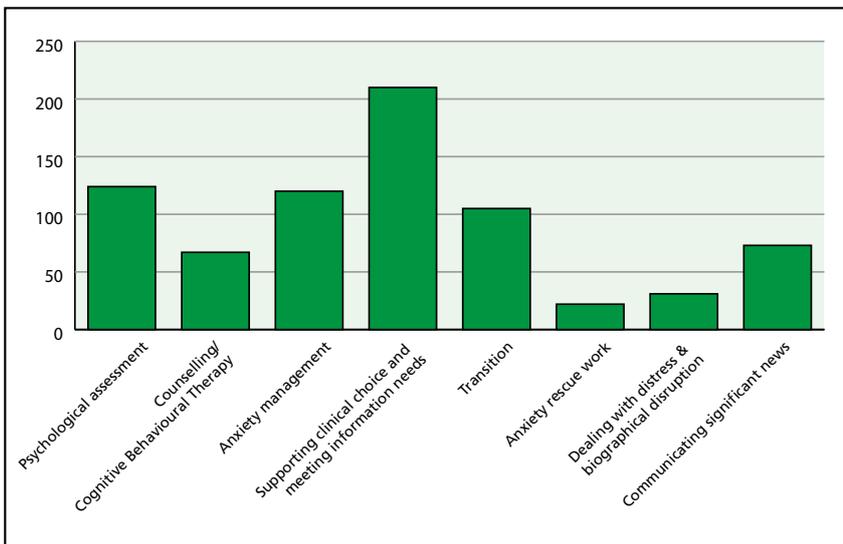


Figure 4: the distribution of activity in psychological interventions (acute) n=752 events (19% of total for acute group)

SC&T specialist nurses work in a cultural context, managing issues such as the taboo nature of the conditions, denial as a coping mechanism and brokering with those who have influence in the community – for example, negotiating with church leaders in the case of a parent relying solely on prayer to help their child. Their transcultural approach to care enables them to work effectively with groups affected by these conditions, and they play a key role in helping other health care professionals to deliver safe and appropriate care. The social dimension of the work is small but has high impact – and when such issues arise, the specialist nurse has the expertise to refer onto to other agencies at the appropriate time. This study revealed the complex network of health and social care professionals that the nurses utilise to manage care.

The unremitting nature of these conditions and the amount of nursing vigilance applied in the management of a multisystem disease is reflected in these data and also in the interviews. Nurses described their vigilance work, understanding of risk, and using supported self-management of patients to a point where they intervened if required.

The cultural dimension of sickle cell and thalassaemia permeates every aspect of the work of the specialist nurse



The acute group tended to provide specialist and complex symptom control more often and in the context of inpatient, outpatient and emergency care. The community nurses provided the majority of specialist symptom control within the context of outreach, nurse-led clinics and telephone work.

Procedures performed ranged from low intensity venepuncture to exchange transfusion and central line insertion. These procedures require a high degree of technical skill, nursing vigilance (observations, possible reactions, infections), brokering (for example with radiologists, radiographers, bed managers, scientists) and in terms of role substitution offer a viable cost-effective alternative to medical staff.

Much of the nursing work was in the form of nurse-led clinics or telephone assessment/interventions (around 40% of the work overall). SC&T nurses spent 7-9% of their activity delivering clinic appointments to new-to-service patients. More information can be found in the full report.

Organisational barriers to practice

During interview, nurses identified a number of organisational barriers to practice, such as a perceived lack of support for prescribing and a lack of administrative support – particularly relating to data collection. This is reflected in the amount of clerical work the nurses recorded. Extrapolated to one year, this represents a considerable amount of non clinical time which could be released to manage clinical care (around 5-7 hours per WTE per week).

Many nurses found that their roles were being subjected to a review process by external (non-nurse) management consultants. This had a negative impact on their own perception of the role, with many expressing feelings of lack of understanding from managers.



“Without the committed help and advice from my specialist nurse to me and my family, I don’t think I would have pulled through as well as I have”

“... now (I have a child) there is more than just me, so I am much more self managing and I listen to my body”



Education

Specialist education plays a vital role in the provision of comprehensive, effective and high quality SC&T services – supporting both genetic counselling and clinical nursing activities.

Training for counselling at-risk couples and giving newborn screening results to parents has been developed by the NHS SC&T Screening Programme and training requirements are included in the Programme standards. Two new modules replacing the old 'PEGASUS' course have been developed by King's College London, which will run from summer 2012. A competency framework for SC&T counselling is also in development, including detailed signposting to web-based learning materials (NHS Sickle Cell & Thalassaemia Screening Programme 2012).

Competences have also been developed for SC&T clinical nursing care, accredited by the RCN in 2010 (RCN 2011). This framework has been enthusiastically received but needs further work to make it more user-friendly in the workplace. A new (or modified existing) course might focus the syllabus on these competences and offer one or more modules at post graduate level, so that nurses specialising in SC&T have the opportunity to take accredited postgraduate courses in both clinical and counselling care.

However, in the interview studies for this project, nurses highlighted increasing difficulties in accessing specialist post-registration and postgraduate education. This appeared to be due to workload and financial constraints of employers. They also articulated an unmet need for specialist SC&T education and advanced practice skills, such as physical assessment courses and prescribing.

It is a key concern that rising university fees and reduced NHS training budgets will put SC&T specialist education at risk with consequent threat to the quality of nursing care.

Specialist and advanced practice education is likely to represent a good return on investment for this group.

Specialist SC&T nursing and the contribution to Quality, Innovation, Productivity and Prevention (QIPP)

In other specialisms, specialist nursing case management has led to quicker discharge of patients from hospital and prevention of patients coming into A&E (RCN 2010, Quinn 2011, Baxter & Leary 2011, Oliver & Leary 2012, Mahaffey et al 2012).

In SC&T there have been some local studies examining the cost-effectiveness of the role. Over 1000 hospital 'bed days' were saved at a major London hospital in one year through a nurse-led case management system. Similar studies have been conducted in two other hospitals. A community matron model has been developed by one PCT, providing patients with safe and personalised care at home that might otherwise be carried out in hospital. It is estimated that this model has saved over £70,000 in secondary care avoidance costs.

Sickle cell and thalassaemia are 24-hour conditions, requiring expert out of hours support. In one area of London, a fifth of emergency beds are taken up with haemoglobinopathy patients, and in the case of sickle cell, 90% of acute admissions are due to painful crises. In rheumatology and cancer, some Trusts are running 24-hour expert nurse telephone services which contribute to preventing unscheduled care including A&E admissions. A similar approach for SC&T would benefit patients and makes sound economic sense.

Several centres gave examples of innovative case management designed to reduce hospital admissions – for example, the use of community matrons enabling expert nurses and consultants to use their time more appropriately.





Advantages and disadvantages of different models for community provision were considered. For example, are patients best treated in stand-alone SC&T centres, or by a central team specialising in long term conditions? Issues to balance included efficiency, economies of scale and pooling of resources, specialist expertise, cultural understanding and community awareness.

SC&T specialist nurses are among a group of specialist nurses who perform exceptionally well against the NHS QIPP agenda (Quality, Innovation, Productivity & Prevention – see figure 5). They help to prevent unscheduled admissions – and many take the lead on service improvement, redesign and reform, as they understand patient need. A high proportion of work is carried out in cost-effective nurse-led (rather than consultant-led) clinics. Specialist nursing interventions enable patients to self manage – which has already been shown to be cost-effective in other long term conditions.

In the absence of a specialist nurse, patients rely on hospitals and A&E (particularly for sickle crises). However, generic services may lack the expertise and time to deal with the complexity of these multisystem disorders, which may lead to suboptimal care and extended hospital stays. It is therefore cost-effective to employ specialist nurses where there is clinical need.

“One night in A&E, it took 25 attempts to get the (transfusion) line in.”

“The unit is mainly for cancer ... my specialist nurse is not always available, and I feel quite isolated. There is no patient drop in”



Figure 5: SC&T specialist nurses and the NHS QIPP challenge



Recommendations

This project has demonstrated the immense contribution of SC&T specialist nurses in providing affordable, expert and accessible care to patients who are from diverse ethnic backgrounds. To maximise the benefit of the role to patient care, the following recommendations are made.

- **Commission SC&T specialist nurses where they are needed.** Lack of expert care is costly in the long term, causes distress and results in poor clinical outcomes & experiences. Patient/specialist SC&T nurse ratios should be on a parity with other long term conditions.
- **Undertake a full national census of current SC&T specialist nurse posts** to gain a better understanding of the workforce and inform workforce planning.
- **Safeguard the jobs of existing specialist SC&T nurses.** Expert nursing care keeps people out of hospital (reducing costs) and living higher quality, healthier, productive lives (improving outcomes in line with the NHS Outcomes Framework).
- **Release time to care, maximising the use of specialist nurse time.** Clerical work can be delegated – for example, investing in clerical support/data collection staff would release about 5-7 hours per whole time equivalent per week back into clinical work.
- **Focus specialist nursing resources on self management and reducing complications.** Help patients and their families to understand their conditions and look after themselves, preventing unscheduled admission where possible.
- **Listen to patients and families.** Regular work to capture their views and experiences ensures that nursing care meets patient needs. Support expert patients & carers to inform service delivery.
- **Recognise and encourage a greater contribution to the NHS QIPP agenda by engaging with specialist nurses.** Trusts can increase productivity and use the insight that nurses provide to perform robust service reviews.
- **Establish clear job descriptions** to eliminate the multiplicity of job titles and define the role of the SC&T specialist nurse.
- **Ensure access to appropriate accredited education and resources for specialist nurses.** Offer support through bursaries, commissioned places and study time.
- **Organise nursing services based on clinical need** and to enable seamless, integrated specialist nursing care for the patient across acute and community settings.
- **Offer equitable access to expert SC&T nursing care across all geographical areas.** Given the variation in prevalence throughout England, there will be different service models and organisational structures according to local needs. However, the two principles of effective governance and the provision of equitable and seamless services based on clinical need are always paramount.
- **Address the lack of health care research in sickle cell and thalassaemia.** Recommendations will be more robust and will benefit from further research.



This report has brought together for the first time data about the complexity of specialist nursing activity for patients with sickle cell and thalassaemia. The evaluation undertaken will be of immense value to commissioners and health care providers, and provides a solid basis for future work – in particular, further exploration of cost benefits, and lobbying for sickle cell and thalassaemia to be recognised as ‘ambulatory care sensitive conditions’ (ALCs), requiring active management to keep patients well and out of hospital.



“We’ve come a long way since 1979 when I was appointed as the first ever Sickle Cell & Thalassaemia specialist nurse in England. We have a national screening programme for sickle cell and thalassaemia; we’ve built up clinical services for people living with these debilitating conditions; and we’ve established national standards for training and practice. Specialist nurses have played a pivotal role in this success story – but they still have a lot to do. Every patient deserves the highest quality of care, and specialist nurses are best placed to deliver it.”

Professor Elizabeth Anionwu, CBE, FRCN
Emeritus professor of nursing,
University of West London

SC&T specialist nurses provide expert, life-saving and culturally sensitive care and if compared with other long term conditions are likely to represent excellent value for money. Their informed, coordinated and empathetic approach meets very specific needs at different times through the life course, from pre-conception, through childhood and adolescence, and on to adult life and old age. They are highly valued by patients and families, giving them the tools they need to take control.



References

Baxter J, Leary A (2011) Productivity gains by specialist nurses. *Nursing Times* Aug 2-15; 107 (30-31):15-7

Mahaffey K et al (2012) The role of the diabetes inpatient specialist nurse in preventing hospital admission from A&E. *Journal of Diabetes Nursing* 16(2) 57-61

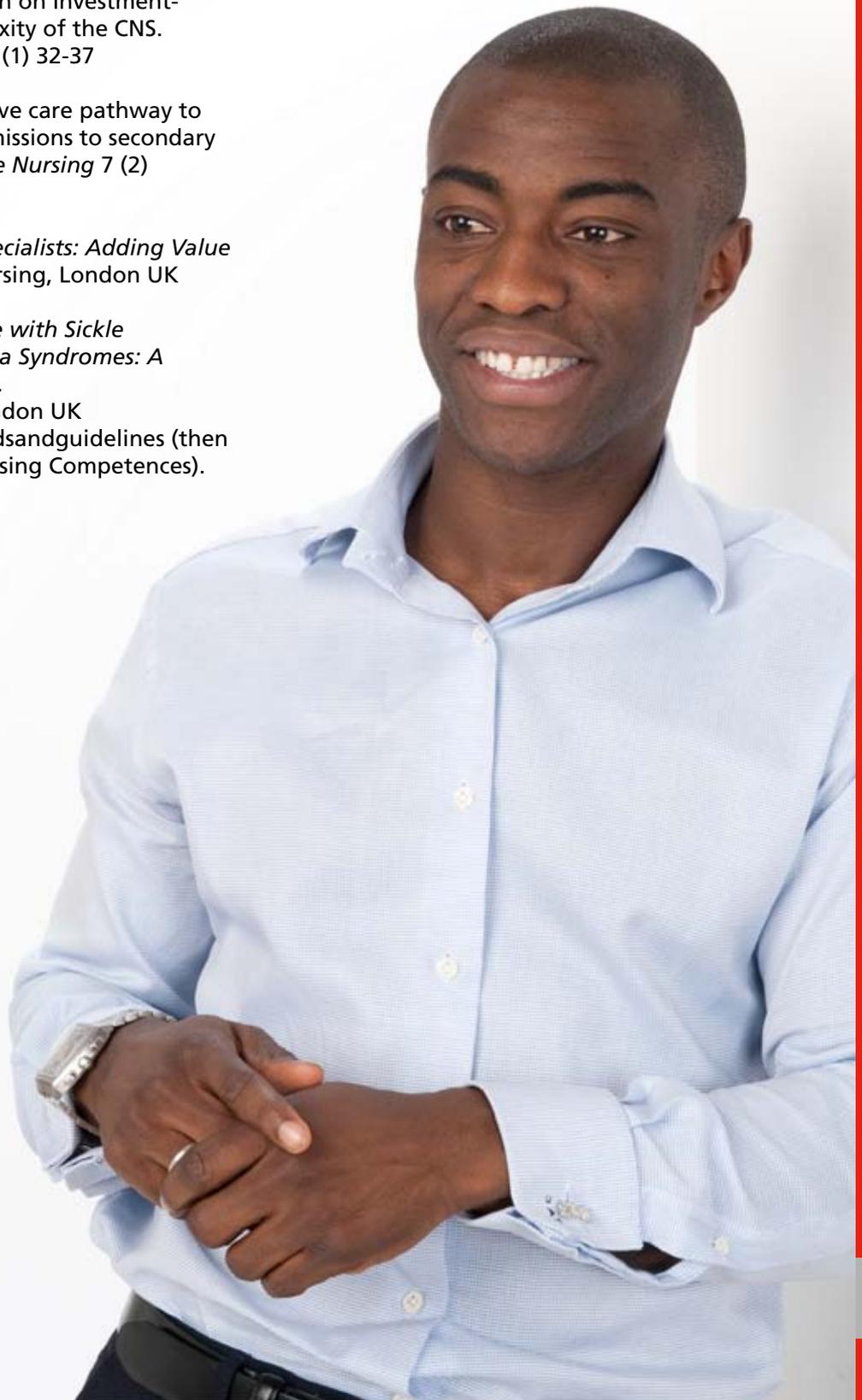
NHS Sickle Cell & Thalassaemia Screening Programme (2012) sct.screening.nhs.uk/publications accessed April 2012

Oliver S Leary A (2012) Return on Investment-Workload, value and complexity of the CNS. *British Journal of Nursing* 21 (1) 32-37

Quinn D (2011) A collaborative care pathway to reduce multiple sclerosis admissions to secondary care. *Journal of Neuroscience Nursing* 7 (2) 497 - 499

RCN (2010) *Clinical Nurse Specialists: Adding Value to Care*. Royal College of Nursing, London UK

RCN (2011) *Caring for People with Sickle Cell Disease and Thalassaemia Syndromes: A Framework for Nursing Staff*. Royal College of Nursing London UK sct.screening.nhs.uk/standardsandguidelines (then click on Royal College of Nursing Competences). Accessed April 2012



About the authors

Elizabeth Anionwu, CBE, FRCN

Elizabeth is Emeritus professor of nursing at the University of West London and Patron of the Sickle Cell Society.

Dr Alison Leary

Alison is an Independent researcher/analyst commissioned by statutory & charitable organizations. She is Reader in Advanced Practice at London South Bank University and holds several other honorary academic positions.

Versions of this report

This is a summary of a longer report which includes detailed references. You can download both versions from:

sct.screening.nhs.uk/publications

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The NHS Sickle Cell & Thalassaemia Screening Programme

7th Floor Capital House

42 Weston Street

London SE1 3QD

Tel: 020 7848 6634

sct.screening.nhs.uk