

A competence framework for those nursing people with sickle cell disease and thalassaemia has been launched to help address variations in standards of care

# A skills framework for sickle cell disease and thalassaemia

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**Abstract** Tangayi S et al (2011) A skills framework for sickle cell disease and thalassaemia. *Nursing Times*; 107: 41, 12-13. The standard care that people with sickle cell disease or thalassaemia experience varies widely. A specialist nurse was inspired to address this by developing a competency framework for these haemoglobin disorders, which has been accredited by the Royal College of Nursing.

This article looks at the rationale for creating competencies for nurses caring for people with these conditions, and outlines the standards required to achieve optimum patient care.

The care of people with sickle cell diseases and thalassaemia in the UK varies widely. Historically, people have tended to associate SCD and thalassaemia with particular ethnic groups, such as African, Caribbean or South Asian people. Some health professionals feel they are unlikely to treat patients with these conditions because of the geographic areas where they practise.

Evidence of discrepancies in care was demonstrated by the National Confidential Enquiry into Patient Outcome and Death (Lucas et al, 2008). The report highlighted the issues around mortality of SCD

patients because some nurses and doctors caring for them lacked knowledge.

One of the report's recommendations was that training for nurses and doctors caring for patients with SCD needed to improve, including the management of ongoing and acute pain. This should include in-service training and tailor-made courses for sickle cell pain management with regular updates.

To address the variations and ensure uniformly high-quality care, a competency framework has been developed specifically for nurses to help them care for and understand the needs of this patient group. *Caring for People with Sickle Cell Disease and Thalassaemia Syndromes: a Framework for Nursing Staff* is aimed at nurses at all levels and was developed through a collaboration between the Royal College of Nursing and the NHS Sickle Cell and Thalassaemia Screening Programme, with support from the Department of Health.

To ensure every aspect of care was explored, the framework was developed with input from patients and carers, haematologists, paediatricians and nurses. Other contributors included counsellors, psychologists, nurse education specialists, RCN members, the Sickle Cell Society UK and the UK Thalassaemia Society.

## What are sickle cell disease and thalassaemia?

SCD and thalassaemia are inherited blood disorders, passed from parents to children through altered haemoglobin genes. They affect around 13,500 people in England,

## 5 key points

**1** Sickle cell disease (SCD) and thalassaemia affect around 13,500 people in England

**2** SCD and thalassaemia are inherited blood disorders, which are passed on through altered haemoglobin genes

**3** People with SCD will usually need medical treatment and care throughout their lives

**4** Sickle cell anaemia is the most serious form of SCD

**5** With thalassaemia, the body does not produce enough red blood cells to transport oxygen around the body



Normal and sickle cells: sickle cell disease is inherited

making them the most common inherited conditions in the country (NHS Sickle Cell and Thalassaemia Screening Programme, 2011). The conditions are recessive, which means children can only inherit the conditions if they receive two faulty haemoglobin genes – one from each parent. The most serious SCD is sickle cell anaemia, while the most serious form of thalassaemia is beta-thalassaemia major.

In SCD, red blood cells can become rigid and shaped like a sickle, and can clog small blood vessels, leading to oxygen failing to reach all parts of the body. This can result in tissue death and cause severe pain, as well as other complications, including overwhelming infections, lung problems and stroke. People with the condition usually need medical treatment and care throughout their lives.

With thalassaemia major, the body does not produce enough red blood cells to

transport oxygen around the body. People with severe forms of the disorder need to have regular blood transfusions through-out their lives (usually every 3-4 weeks) to keep them alive. Possible complications of thalassaemia include diabetes and osteoporosis.

Children who inherit a faulty gene from one parent only are known as carriers. They do not have the condition and will never develop it but can pass on the faulty gene to their children. If two carriers have a baby together, there is a one in four chance with each pregnancy that their baby will inherit a blood disorder. The NHS Sickle Cell and Thalassaemia Screening Programme therefore identifies carriers of the sickle cell or thalassaemia so that carriers can receive information, support and advice to enable them to make informed reproductive choices.

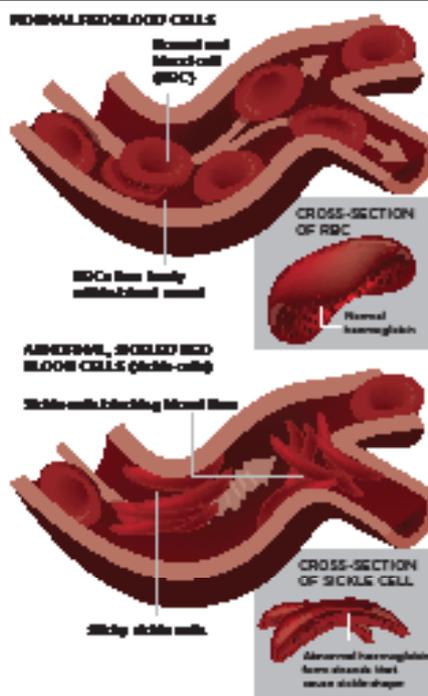
### Motivation to produce competencies

Sekayi Tangayi, the lead specialist nurse and service manager of the London-based Newham Sickle Cell and Thalassaemia Centre, was inspired to develop a competency framework for these conditions because of her patients' variable experiences of care. The aim of the nursing competencies are to:

- » Enhance nursing care for patients with SCD and thalassaemia to improve the lives of their carers and families;
- » Provide a training framework for nurses wishing to specialise in SCD and thalassaemia care so that they are encouraged to specialise and remain longer in this area.

As a haemo-oncology nurse, Ms Tangayi was required to refer to nursing competencies developed to provide optimum care for cancer patients. She realised that this model could easily be used for SCD and thalassaemia nursing. Implementing a framework would mean nurses could begin to appreciate fully the needs of SCD and thalassaemia patients and understand that having life-long conditions means many of them are experts in their care.

Many initiatives across the country aim to increase awareness among health professionals and the public about SCD and thalassaemia. For example, the UK Thalassaemia Society launched a suite of videos to educate people about thalassaemia and give patients' perspectives. The NHS Sickle Cell and Thalassaemia Screening Programme has produced *The Family Legacy* – a docudrama based on a story of a family whose newborn baby is diagnosed with SCD. Both these films are shown to



How sickle cell affects blood flow

community and support groups, adding to the awareness-raising campaign.

In 2010, the RCN Accreditation Board awarded the competency framework for SCD and thalassaemia full accreditation. The working group is focusing on working with higher education institutions and NHS organisations to incorporate the competencies into nurse education programmes, while the RCN Haematology Network is promoting the use of the nursing competencies in practice.

The next challenge is to ensure the framework is embedded within training and becomes part of nurses' everyday practice. **NT**

#### References

- Lucas SB et al (2008) *A Sickle Crisis. A Report of the National Confidential Enquiry into Patient Outcome and Death*. London: National Confidential Enquiry into Patient Outcome and Death. [tinyurl.com/sickle-report](http://tinyurl.com/sickle-report)
- NHS Sickle Cell and Thalassaemia Screening Programme (2011) *Frequently Asked Questions*. London. [tinyurl.com/screening-programme-FAQs](http://tinyurl.com/screening-programme-FAQs)

### FURTHER INFORMATION

- *RCN Competencies: Caring for People with Sickle Cell Disease and Thalassaemia Syndromes. A Framework for Nursing Staff.* [tinyurl.com/RCN-SCD-thalassaemia](http://tinyurl.com/RCN-SCD-thalassaemia)
- The NHS Sickle Cell and Thalassaemia Screening Programme. [www.sct.screening.nhs.uk](http://www.sct.screening.nhs.uk)

### THE TEN COMPETENCIES

- 1 Provides empathy and understanding, and works with the patient (and their family/carer) as an expert in their own condition
- 2 Assesses, in collaboration with the patient, their needs, taking into account the impact on their age and developmental stage, and their cultural and ethnic background
- 3 Undertakes comprehensive physical assessment and follows up with appropriate action, including referral to medical specialist, for relevant chronic healthcare conditions
- 4 Signposts and supports patients (and families/carers) in their understanding of their genetic conditions
- 5 Develops and evaluates a self-management plan with the patient
- 6 Works alongside and with the patient (and family/carers) to address the psychological and social impact of their condition
- 7 Works with the patient (and family/carers) to manage their pain (patients with SCD)
- 8 Provides specific interventions safely with regards to:
  - Undertaking phlebotomy and cannulations
  - Managing central venous access devices (CVAD), including portacaths
  - Transfusions and exchange blood transfusions
  - Fluid management/hydration
  - Pharmacological treatment and side-effects
  - Iron overload management including chelation therapy
- 9 Uses early warning tools/approaches (for example, red alert) to identify the patient's changing and deteriorating condition, and takes appropriate action
- 10 Actively improves and promotes services across the care pathway

- The RCN haematology network. Ana Champou: [ana.champou@rcn.org.uk](mailto:ana.champou@rcn.org.uk)
- *Sickle: A Sickle Crisis?* A report of the National Confidential Enquiry into Patient Outcome and Death. [tinyurl.com/sickle-cell-crisis](http://tinyurl.com/sickle-cell-crisis)
- *The Family Legacy.* [tinyurl.com/screening-family-legacy](http://tinyurl.com/screening-family-legacy)
- The UK Thalassaemia Society DVDs. [tinyurl.com/ukts-org-video](http://tinyurl.com/ukts-org-video)