MIND THE GAPS

SICKLE CELL & THALASSAEMIA ALL PARTY PARLIAMENTARY GROUP (SCTAPPG) REPORT INTO THE EDUCATION OF PRE-REGISTRATION NURSES AND MIDWIVES

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All-Party Parliamentary Group
Sickle Cell and Thalassaemia
SICKLE CELL SOCIETY
UNITED KINGDOM THALASSAEMIA SOCIETY
STANMAP

“I’m in crisis”

The Sickle Cell and Thalassaemia All-Party Parliamentary Group (SCTAPPG) in collaboration with the Sickle Cell and Thalassaemia Association of Nurses, Midwives & Allied Professionals (STANMAP), with contributions from the Sickle Cell Society, The UK Thalassaemia Society and The Chief Nursing Officer Black & Minority Ethnic Strategic Advisory Group report into the education of pre-registration nurses and midwives.

Executive summary

About the Stakeholders

The Sickle Cell & Thalassaemia Association of Nurses, Midwives and Allied Professionals (STANMAP)

Sickle Cell & Thalassaemia Association of Nurses, Midwives and Allied Professionals (STANMAP) was formally established in 2016, following the merger of STAC an acute and community nurses group that was established in 1987 and FAST-n an acute nurses group that was established in 2008. STANMAP continues both former organisations aims: to support members in their role as specialist and non-specialist care providers, maintaining continuing education and to support development of excellence in practice in caring for individuals, families and communities with and at-risk of sickle cell, thalassaemia and related conditions.

Sickle Cell and Thalassaemia All-Party Parliamentary Group (SCTAPPG)

The Sickle Cell and Thalassaemia All Party Parliamentary Group (SCTAPPG) *raison d’etre* is to reduce the health inequalities that are faced by sickle cell and thalassaemia patients in the UK. The SCTAPPG secures this by influencing and lobbying policymakers to raise the profile of sickle cell disease and thalassaemia on the political agenda. These efforts are ultimately intended to improve standards of care and address other critical issues, as recommended by the key stakeholders. Members achieve this aim by engaging with parliamentary colleagues, the government, health professionals, and community and patient groups to raise awareness relating to the conditions and needs of patients. The SCTAPPG works in collaboration with The Sickle Cell Society and the UK Thalassaemia Society striving to reflect the concerns of their membership and to act as a vehicle for change.

The Sickle Cell Society (SCS)

The Sickle Cell Society (SCS) is the only national charity in the UK that supports and represents people affected by sickle cell disease to improve their overall quality of life. The Society's mission is to enable and assist individuals with a sickle cell disorder to realise their full economic and social potential. The Sickle Cell Society (SCS) has been in existence since 1979 and has amassed a wealth of experience in sickle cell not only in the U.K. but through its network and association with other countries.

The United Kingdom Thalassaemia Society (UKTS)

The United Kingdom Thalassaemia Society (UKTS) is a national voluntary organisation established in 1978 and is dedicated to promoting an awareness of thalassaemia, education of health and allied professionals, community professionals and others. The UKTS is committed to development of resources to promote the interest of those with and at-risk of thalassaemia.

Chief Nursing Officer's BME Strategic Advisory Group (CNOBME)

The Chief Nursing Officer Minority Ethnic Strategic Advisory Group (CNOBME) was established in 2001 and is composed of a group of senior, highly acclaimed health professionals and academics working in the health service and academia to provide leadership, expert opinion and development of strategies for steering the nursing and midwifery education and practice agenda in England.

Background to the project

There are anecdotal reports that qualified nurses, doctors and other health care professionals lack knowledge of sickle cell and thalassaemia and that this affects their ability to provide skilled and effective care for those with and at risk of these life-long genetic, chronic and potentially fatal conditions.

The Sickle & Thalassaemia Association of Nurses, Midwives and Allied Professionals (STANMAP) established an advisory working group in May 2018 to discuss the possible lack of knowledge and skill among nurses and midwives caring for people with sickle cell and thalassaemia and to determine whether there is any credence in the opinion that a lack of inclusion in student nursing and midwifery curricula is a contributory factor for ineffective care and management of this patient group.

The advisory working group contained the following members:

- Dr. Lola Oni OBE Chair, Specialist Nurse Consultant/ Service Director / Lecturer Brent Sickle Cell & Thalassaemia Centre, Central Middlesex Hospital, London NW University Healthcare NHS Trust and Chair of STANMAP
- Professor Laura Serrant OBE, Chair of the Chief Nursing Officer Black and Minority Strategic Advisory Group, Professor of Nursing Sheffield Hallam University
- Associate Professor Stacy Johnson, University of Nottingham
- Joan Walters, Senior Practitioner Lecturer, King's College Hospital and King's College London University
- Romaine Maharaj, Operations Manager, UK Thalassaemia Society
- Adam Lloyd, Parliamentary Officer for Sickle Cell and Thalassaemia All Party Parliamentary Group (SCTAPPG)

The working group meetings were hosted by the Sickle Cell & Thalassaemia All Party Parliamentary Group (SCTAPPG) on behalf of the Sickle Cell Society and UK Thalassaemia Society. Following a number of roundtable discussions it was agreed to conduct a survey and examine the educational input about sickle cell and thalassaemia in nursing and midwifery pre-registration training.

Service Users voiced dismay at unnecessarily long waits in A&E during an acute illness, poor management of pain, prolonged stay in hospital is a recurring experience. Experienced clinicians are worried about the future of haemoglobinopathy nursing in a climate of decreasing specialist knowledge among generalist nurses and midwives. Patients and specialist clinicians are united in their passionate support to increase the level of understanding and knowledge of these two under recognised conditions.

The working group agreed to gather evidence of the training experiences of student nurses and midwives. The information gathered is to enable the group to explore how to best promote the preparation of future nurses, midwives and other healthcare professionals with the knowledge, skills and competencies to provide effective care for people with and at-risk of sickle cell and thalassaemia, two potentially fatal genetic conditions.

Since its inaugural meeting, the advisory group has convened three times to consider the following issues:

- Have student nurses & midwives had any formal teaching on the subject?
- How much teaching & clinical training have they had?
- If they have had any teaching do the students perceive that the input met their educational needs and development of skills to provide effective care?
- Do the students feel confident to care for clients/ patients with or at-risk of these conditions?

One of the first actions of the advisory group was to petition the Council of Deans in order to reach academics nationwide to garner support on the need for sickle cell disease and thalassaemia to warrant featuring on university curriculums. A survey was then undertaken and has resulted in the `I'm in crisis report'.

"I'm in crisis" are three words, we believe these three words can make an impact and a difference to those living with sickle cell disease and thalassaemia. These three words will act as a bridge from patient to nurse to enable the patient to start their journey to recovery. These three words will empower nurses to know instantly what care they can and should be providing. These three words will function as a guarantee to service users that they will have access to a quick response and the optimal care they need and deserve. Three words mean that nurses won't have to rely on other healthcare professionals or the patient to decide that the appropriate local and national patient care pathway and guideline should be used for immediate care provision.

"I'm in crisis" could be used as a starting point for any learning resources. This powerful three-word prompt will, we believe, act as a catalyst for health professionals to know they are not only dealing with a patient with one of these two genetic blood disorders, but more significantly how to deal with them. These three words can save lives.

About the Survey

A Google survey form was developed and circulated to all accredited education institutions (AEIs). The survey aimed to explore if nursing and midwifery students had received any training and education on sickle cell and thalassaemia and if they felt equipped to care for clients with or at risk of these conditions. An overwhelming response of 197 responses was received from self-selected students from 9 schools/faculties of nursing and midwifery: City, University of London, Kingston University and St. George's University of London, University of West London, University of Huddersfield, University of Salford, University of Surrey, Nottingham University, Kings College London University, who all welcomed the engagement and agreed to circulate it to their respective nursing and midwifery student cohorts. It was also circulated to students via the Royal College of Nursing (RCN), students on placement in individual hospitals including the London NW University Healthcare NHS Trust and specialist sickle cell & thalassaemia Centres.

We hope this report reflects the need to equip future nurses and midwives with the knowledge, skills and competence to care effectively and provide an equitable service for this client group with the aim of alleviating unnecessary suffering and to help reduce avoidable handicap, mortality and morbidity associated with these genetic conditions.

Survey findings

The survey evidence reveals:

- There was significant difference in the current awareness of sickle cell and thalassaemia, with 88.3% of respondents stating they were aware of sickle cell disease, whilst 37.1% were aware of thalassaemia. The poor level of knowledge and awareness of thalassaemia is worrying and needs urgent attention. Although there was greater awareness of sickle cell disease it is difficult to know whether this awareness is any more than any member of the general public or that the students know about the health and clinical implications of sickle cell disease. This will need to be explored in greater depth as part of a later project.
- In regards to the NHS Sickle Cell & Thalassaemia Screening Programme there was a rough 50:50 split between those who were and were not aware of the screening programme with slightly more (50.8%) being aware.
- It is of concern that for both sickle cell and thalassaemia there is a low number of respondents who have not nursed anyone with either thalassaemia or sickle cell disease. With 80.7% of participants, declaring they have not nursed a sickle cell patient and a larger 92.9% of respondents affirming they hadn't nursed a patient with thalassaemia. With so many trainee nurses and midwives not being exposed to this patient group during their training, it means they will not be well equipped to nurse these patients when they qualify. Access to the patient group is dependent on the region where training is taking place with those in major cities being more likely to encounter the patient population. However lack of educational input and basic knowledge about the subject is unfair to nurses who are an increasingly mobile national workforce looking after a mobile population of patients; it also poses dangers for patients who are at-risk of handicap, mortality and morbidity.
- There is lack of assurance from the respondents when asked to express their confidence if they were instructed to deal with patients with these two conditions: with only 26.4% of respondents claiming they would feel confident to look after a sickle cell patient and only 18.8% stating they would feel confident with caring for a patient with thalassaemia. It is acknowledged that some of these students have started their training recently and it is therefore anticipated that only a small proportion would feel confident in caring for this patient group. Although student nurses and midwives are supernumerary and not part of the workforce they contribute to caring for the patient group and their care and management of the patient group will create tensions and anxiety for the patients if their trainers also lack competence and the ability to convey and promote acquisition of the appropriate knowledge and skills these students need for caring in future.

- A substantial 71.1% of participants revealed that they had not had any formal teaching time on sickle cell throughout their education and of those that did only 16.7% percent of nurses in the sample had received a taught session of one hour or longer. Similarly, with thalassaemia 80.7% of nurses had not had any taught sessions on the subject and of those that did only 10.2% had received a session of an hour or longer.

See Appendix 1 for full data set and Methodology

Recommendations

The data suggests that student nurses and midwives are enthusiastic learners and provide a fertile ground for learning that ensures acquisition of life-long knowledge, skills and increasing competence to care for people with and at-risk of sickle cell and thalassaemia.

In view of the outcome of this survey, we make the following recommendations:

- Approved education institutions (AEIs) should incorporate the subject in all components of training. They can strive to achieve this by implementing pragmatic steps that could include:
  - AEIs should identify a subject champion within a group of universities who will support development of the subject within each institution
  - AEIs working with professional bodies and charities should develop a repository of learning resources and make them available for all institutions
  - AEIs should ensure the subject is incorporated into the assessment strategies nationally and in all areas of the curriculum
  - Increase the profile of the subject among university students generally for their own personal genetic needs
- AEI's should evaluate whether the existing omission of formal teaching time on these two conditions violates their respective equality and diversity policies. They should open this to consultation and work in conjunction with their student bodies.
- This is an opportune time to discuss these recommendations with the Council of Deans and individual schools of nursing and midwifery prior to their revalidation of nurse training programmes in 2019 and 2020. And the midwifery revalidation of training currently going through consultation, for publication in 2019 and implementation in 2020.
- The Nursing and Midwifery Council should use their new Standards for pre-registration nursing education (2018) when revalidating all nursing curricula in 2019 to ensure that they are committed to addressing key aspects of equality and diversity. We urge to them to monitor more closely and enforce existing standards to ensure there is commitment from educational providers to maintain equity in educational subject content and evaluation.
- The Department of Health (DoH) should look at ways in which they can support the inclusion of these two under recognised conditions in post registration healthcare professionals continuing education programmes.
- The Department of Education (DofE) equality and diversity policy "aims to set an example as an employer and is developing policies that make opportunity equal for

children and young people."[1] They do this by "making sure that meetings on different aspects of the curriculum involve partners from the various equality communities."[2] We consider this pledge offers an ideal platform in which to engage with this report's recommendations.

What is Sickle Cell & Thalassaemia?

Sickle Cell and Thalassaemia are two of the most common genetically inherited conditions worldwide. They are seen mainly in people from Africa, Asia, the Mediterranean Islands, South America, Middle and Far East. Because of migration and increasing intermarriage they are found increasingly in most parts of Northern Europe and worldwide. In the United Kingdom it is estimated approximately 15,000 people have some form of sickle cell disease (SCD) whilst 1,000 have thalassaemia.

The East Midlands Specialised Commissioning Group (2011) stated,

> "Sickle Cell Disease (SCD) is now the most common serious inherited genetic disorder in England affecting 1:2000 births. Approximately 350 newborn babies are detected with SCD annually by the linked Antenatal and Newborn Screening Programme. Approximately 20 – 30 babies are born each year with a significant thalassaemia condition" (2011:3).

These genetic conditions affect the red blood cell haemoglobin, which is the part of the blood that carries oxygen around the body. The type of haemoglobin an individual has is determined by the haemoglobin gene inherited from each parent.

The most common (normal) combination that the majority of people inherit from both parents is Haemoglobin AA commonly written (Hb AA). Some people inherit one normal haemoglobin A gene from one parent and a sickle haemoglobin S gene from the other parent and have sickle cell trait (Hb AS); these individuals are healthy carriers and do not experience any of the symptoms or clinical health problems associated with sickle cell disease. 1 in 4 (25%) West Africans, 1 in 10 (10%) Black Caribbean, 1 in 20 (5%) Asians and 1 in 100 Northern Greeks (1%) are carriers of the sickle gene.

A person who inherits one normal haemoglobin gene from one parent and, for example, a beta thalassaemia gene from the other (Hb Aβ^{Thal}) has beta thalassaemia trait, this is also a healthy carrier state and such individuals do not have the clinical health problems associated with thalassaemia major (disease). 1 in 7 Greek Cypriots, 1 in 12 Turks, 1 in 20-30 Asians, 1 in

[1] Equality and diversity https://www.gov.uk/government/organisations/department-for-education/about/equality-and-diversity
[2] Equality and diversity https://www.gov.uk/government/organisations/department-for-education/about/equality-and-diversity

30 -100 Black African & Black Caribbean and 1 in 1000 White English are carriers of the beta thalassaemia gene.

Those who have inherited two unusual haemoglobin gene have some form of disease for example, those who inherit sickle haemoglobin S gene from both parents have sickle cell anaemia (Hb SS); this combination is the most common and the most severe form of sickle cell disease.

Those who inherit a beta zero thalassaemia gene from both parents have beta thalassemia major (Hb $\beta^0\beta^{0Thal}$), this is the most severe form of beta thalassaemia disease.

What is sickle cell disease?

Normal red blood cells are round, soft, spongy, live 120 days, are able to manoeuvre through the blood vessels readily and carry oxygen efficiently; they maintain their shape and consistency even after giving up the oxygen to the body tissues.

As a result of the genetic mutation when red blood cells that contain mostly sickle haemoglobin give up the oxygen to the body tissues they become rigid, hard, brittle and inflexible; they break easily in the rigours of travelling though the blood vessels; they live about 10-20 days before becoming severely damaged and disposed of by the body. Because of their rigidity the sickled shaped red blood cells lose their flexibility and cannot flow easily through narrow blood vessels, they can "jam up," thereby causing obstruction in blood flow. When this happens, vital oxygen can no longer be carried to the affected part of the body, this causes a pain episode called a 'sickle cell pain crisis', this is the hallmark of sickle cell disease and the most common symptom associated with the condition.

During a moderate to severe sickle cell crisis episode an individual will require hospitalisation and specialist medical and nursing care.

A sickle cell crisis and associated complications can cause handicap, mortality and morbidity; hence it is essential that those looking after these patients have the necessary knowledge, skill and competence to care for them effectively.

What is beta thalassaemia major?

The red blood cells of those with beta thalassaemia major are fewer, smaller and paler, they lack the red pigmentation (haemoglobin) that allow the red blood cell to carry oxygen effectively because they do not mature sufficiently in the bone marrow before they are released into the blood circulation. These immature red blood cells are very fragile and get damaged in the blood circulation, hence they are not efficient at carrying oxygen around body, giving rise to a severe chronic anaemia which if untreated is life threatening.

Individuals with beta thalassaemia major are dependent on regular blood transfusion about every 3-4 weeks throughout their lives, without this blood transfusion they will not be able to carry oxygen or live beyond early childhood. The frequent blood transfusions cause excessive amounts of iron to build-up in the body therefore additional specialist treatment is required to prevent the complications associated with this iron overload.

Beta thalassaemia major is also associated with possible medical complications and individuals need to be cared for by health professionals who are knowledgeable and competent in managing this group of patients.

Why conduct a survey?

Following a number of damming reports about poor care and management of people with SCD and avoidable deaths occurring in the NHS the first specialist Centre was established in Brent in 1979. Since then there has been major improvement in scientific research, advances in treatment and knowledge of these conditions. Several specialist Centres have evolved in the last four decades.

Publications on care and management standards and their updates over the years have contributed to improving care, reducing handicap, morbidity and mortality, these include Children's Sickle Cell standards (2010) 2nd edition, Adult Sickle Cell Standards (2018) 2nd edition and Children and Adults Thalassaemia Standards (2016) 3rd edition. However in spite of these developments patients continue to report negative attitude from some members of staff and in some cases a lack of knowledge and skill in providing care. The National Confidential Enquiry into Patient Outcomes and Deaths (NCEPOD) (2008) highlighted that the lack of knowledge and skill among healthcare professionals is a major contributory factor to the mortality of these patient groups.

In a review of the workforce of specialist doctors and nurses Ryan suggested,

> *"The varied prevalence of haemoglobinopathies across England combined with the known difficulties in delivery of care to minority ethnic groups poses a challenge for access to specialist care with the risk for inequity across the country, particularly in low prevalence areas. This was recognised with the introduction of specialist commissioning for these disorders from 2013* (Ryan 2015: 5).

A transient student population and the recent phenomena of an increasing movement of people from high cost urban cities to rural areas of the UK has seen a major change in the demographic makeup of many rural towns and an increasing number of people with and at-risk of these genetic conditions.

Headline news reports have contributed to patients anxieties when encountering healthcare providers in community and acute hospital environments.


Sick boy, three, dies waiting for an appointment

Tony Bassett
15.11.10
Evening Standard

METRO NEWSPAPER
27 JANUARY 2011


Medics 'refused to aid dying student'

By Fred Attewill

TWO ambulance workers refused to take a dying student to hospital because she had soiled herself, it was claimed yesterday.

The pair have been suspended from frontline duty while the allegation by Sarah Mulega's landlady is investigated by ambulance bosses.

Ms Mulega suffered from sickle cell anaemia, a genetic condition which can restrict blood flow causing painful and potentially deadly episodes called crisis.

Her landlady, Chinwe Moneke, called for an ambulance after discovering the 21-year-old collapsed in her bedroom on January 9. When the first ambulance arrived, the crew 'did not touch Sarah at all', she claimed.

Collapse: Sarah Mulega

She said: "They did not check Sarah's temperature nor heartbeat. I heard one of them tell her, "If you want to be taken to hospital, then get up so we can take you."" Ms Moneke, from Barking, Essex, believes their refusal to help was because Ms Mulega had soiled herself.

She then called for another ambulance which took Ms Mulega to hospital but she died later. An inquest has opened into her death but a cause of death has not yet been established.

A spokesman for London Ambulance Service offered its condolences and said an investigation had begun.

Ms Mulega's cousin, Thomas Chisanga, said: "The people who were there to help her let her down in the last hour. I have so much anger towards them."

This has made it imperative that we have a healthcare workforce that is not totally reliant on specialists to care for this group of patients.

Secondly as Ryan (2015) has highlighted that the increasing difficulty to recruit specialist doctors and nurses compounds the situation and makes it even more urgent to develop a generalist workforce that is adequately prepared, skilled and competent to provide adequate care for patients with sickle cell disease and thalassaemia irrespective of where they live in the UK and indeed globally.

Discussion at a number of professional meetings highlighted the continuing poor experiences of patients with sickle cell disease and thalassaemia especially when presenting to A&E during an acute illness and when admitted to the hospital ward.

Some of these reported experiences relate specifically to medical and nursing staff's attitude to patients especially to those with sickle cell disease in acute care settings.

The outcome of a survey conducted by the Picker Institute among patients with sickle cell disease in 2015 validates patients' reports of staff's lack of knowledge, skill and competence and negative attitude to the patient group culminating in provision of inadequate and poor healthcare.

Patients interviewed stated,

> *"The help at [certain hospitals] has been good. I avoid some because of the lack of knowledge and care provided there." (Picker 2015:12)*
>
> *"Adults and parents appeared to have less confidence in the knowledge of the emergency healthcare staff, reporting that 21% (n=19) and 17% (n=13) respectively*

did not know enough about sickle cell disorder relative to only 5% (n=3) of children.." (Picker 2015:18)

"When comparing confidence in knowledge of staff by care settings, respondents were more likely to state that emergency healthcare staff lacked in-depth knowledge of sickle cell (15%; n=35) relative to planned care staff (4%, n=22)." (Picker 2015:20)

"The clinic she attends is very good, it's easy to attend too and the staff know my daughter which makes it easier to cope with. It's more the general health service or accident and emergency they have no idea about sickle cell." "More information and training for accident and emergency staff is needed." (Picker 2015:20)

"… respondents perceived healthcare staff in planned care settings (in particular, hospital outpatient or clinic appointments and day unit hospital admission) to have a much better understanding of sickle cell relative to urgent care staff, which could explain why patients report a better experience in these settings. Adults are less confident in the knowledge of emergency care staff than parents and children, as are Londoners relative to non-London residents." (Picker 2015:49)

The reason for a perpetuating negative attitude and avoidable deaths is perhaps a lack of knowledge, skill, competence and understanding of the potentially fatal nature of these genetic conditions. There is often a subjective view and a misconception that patients with sickle cell disease are merely 'drug seeking' and are difficult to manage during an acute illness; as a result nurses and doctors may fail to prescribe and administer adequate pain relief. Patients perceive an attitude of unwillingness to provide care and lose confidence in the care provider, thereby escalating an attitude of mistrust between patients and their Carers. Patients report increasing tension, anxiety, distress and an exacerbation of their symptoms and a worsening of their disease experience whilst they are in the health care setting.

In 2017 the NHS Sickle Cell & Thalassaemia Screening Programme (NHSSCTP) commissioned the Sickle Cell Society and the UK Thalassaemia Society to conduct a survey of women and couples at-risk of having an affected baby. Thirteen women and couples were interviewed, this culminated in an outcome report entitled, *Patients' Stories*.

The report outlines deficits in health care professionals' knowledge, and that this contributed to a delay in accessing services as well as receipt of a poor service.

The report stated,

"Several healthcare professionals lacked knowledge of the conditions and the screening pathway and did not recognise the need for prompt referral to counselling and PND" (NHSSCTP 2017:2).

"Once we were settled in England I went to register with a local GP and I remembered to ask him about thalassaemia, telling him that we are carriers… (the)

> *GP googled thalassaemia in front of me. He got very agitated and told me that this means the child has no chance of life and you have to terminate the pregnancy as soon as possible. I now know that the GP made a mistake and he was thinking of another kind of thalassaemia." (NHSSCTP 2017: 7)*
>
> *"A common theme in the interviews was an assumption on the part of the service users that the healthcare professionals would recognise the risks and act when carrier status was known. This was not the case even in families that already had a child or children affected by the condition." (NHSSCTP 2017: 13)*

A lack of knowledge and skill hinders the professional's ability to demonstrate knowledge, understanding and provide skilled, competent and empathetic care required by pregnant women and couples who were at risk of having a child with sickle cell disease or thalassaemia major. The Sickle Cell & Thalassaemia Screening Programme of the UK National Screening Committee (2013) provided a comprehensive tool to assist those involved in the antenatal and newborn screening pathway, one would expect practitioners to be familiar with the urgency of providing a timely service to those with and at risk of these genetic conditions but this does not appear to be the case.

In discussing the need for increasing diversity and removal of institutional racism in the NHS, Kline stated,

> "*The review by the West Midlands Quality Review Service looked at 34 services across England. Many patients described being made to feel like "second class" patients as they regularly lost out on beds on specialist wards to patients with white blood cell disorders such as leukaemia. Elaine Miller, national co-coordinator at the UK Thalassaemia Society, told* HSJ *red cell haematology was the "poor relation" to white cell haematology. She said: "If these conditions affected the white mainstream population rather than ethnic minority communities, maybe they wouldn't be so badly overlooked." Sickle Cell Society chief executive John James told* HSJ *race definitely had "a role to play" in the variability of services and the fact they appeared to be "neglected", even in high prevalence areas. About 80 per cent of sickle cell patients are in London. However, the review found the makeup of teams in the capital was "extremely variable." The review also found many emergency departments were not following National Institute for Health and Clinical Excellence guidelines on sickle cell crises, meaning patients were often left in agony awaiting pain relief (Roger 2014:45 citing Calkin 2013)*

It is possible to suggest that lack of training, knowledge and an awareness of these patients' health care needs causes healthcare professionals to lack the perception needed to provide optimal and timely interventions to alleviate pain and suffering especially for patients with sickle cell disease during an acute excruciatingly painful sickle cell crisis episode.

Pre-registration education of nurses and midwives

The impression is that pre-registration education about sickle cell and thalassaemia is not embedded in medical and nursing curricula in and its inclusion is reliant on the capricious will of an individual lecturer or school.

Although sickle cell disease is acknowledged as the most common genetic disease in the UK, and the majority of those affected live in major cities, students, including those who are training in major city universities, are more likely to learn about less common genetic diseases, such as cystic fibrosis, Phenylketonuria and Tay Sachs than about sickle cell disease or thalassaemia.

In order to ascertain whether this impression is accurate following a lengthy deliberation the working group agreed as a first step to gather information about the inclusion of the subject in pre- registration nursing and midwifery curricula.

Nursing and Midwifery code of practice and pre-registration nursing standards

The Nursing and Midwifery Council (NMC) is the regulator for the nursing and midwifery professions in the UK. The NMC maintains a register of all nurses, midwives and specialist community public health nurses eligible to practise within the UK. They set the standards of education, training, conduct and performance so that nurses and midwives can deliver high quality healthcare throughout their careers. The Code is central to all education programmes, and educators must enable students to develop knowledge, skill and competence to provide effective care in future. It is incumbent on the NMC to ensure that schools of nursing and midwifery provide the level of education required for practitioners to practice effectively when they qualify.

The new NMC pre-registration nursing standards (NMCb2018) outlines seven areas of proficiency (platforms) so that the public can be confident that all newly qualified nurses will practise safely and effectively. Five of the conditions that they need to meet are particularly pertinent to caring for those with sickle cell disease and thalassaemia, these are:

- **Proficiency 2 -** Promoting health and preventing ill health - support and enable people at all stages of life and in all care settings to make informed choices about how to manage health challenges in order to maximise their quality of life and improve health outcomes.
- **Proficiency 3** - Assessing needs and planning care – demonstrate and apply knowledge of all commonly encountered mental, physical, behavioural and cognitive health conditions, medication usage and treatments when taking full and accurate assessments of nursing care needs when developing, prioritising and evaluating person-centred care plans.
- **Proficiency 4** - Providing and evaluating care – take the lead in providing evidence based compassionate and safe nursing interventions

- **Proficiency 6 -** Improving safety and quality of care - work with people, their families, carers and colleagues to develop effective improvement strategies for quality and safety, sharing feedback and learning from positive outcomes and experiences, mistakes and adverse outcomes and experiences

(NMCb 2018)

Section 1 of The NMC code expects qualified nurses and midwives to:

> "…make their (patients) care and safety your main concern and make sure that their dignity is preserved and their needs are recognised, assessed and responded to… make sure that those receiving care are treated with respect, that their rights are upheld and that any discriminatory attitudes and behaviours towards those receiving care are challenged" (NMCa 2018: 6)

This survey suggests that nurses and midwives may graduate without an adequate level of knowledge or understanding of these two genetic blood disorders and will therefore have difficulty adhering to the principles outlined in this section 1 of the NMC code.

It is well established that due to the high cost of university tuition in major cities and exorbitant cost of property rental and or purchase many young people gravitate towards outer suburbs and rural areas for tertiary education, to work and raise a family.

The existing demography of the UK demands that on qualifying nurses and midwives must have sufficient knowledge and be equipped to deliver care to all service users which this report suggests may not be the case in some instances with regard to caring for those with sickle cell and thalassaemia. The constantly shifting demography of the UK population makes the need for inclusion in education curricular even more pertinent.

For nurses to graduate they have to satisfy "The Fitness for practice" criteria whereby students who are fit for practice are able to practise safely and effectively with minimal supervision, they are expected to have met the standards to practice safely, even as a novice nurse or midwife and have an understanding of the significant health issues that predominate in the area where they trained. Many of these students are training in major cities with a high minority ethnic population and numbers with these genetic conditions yet it appears they are not being enabled to access the knowledge and skill that should be readily available to them in their learning and placement environment.

The Standards for pre-registration nursing education (NMCb 2018) apply to 'pre-registration nursing education' these are the programmes that a nursing student in the United Kingdom undertakes in order to acquire the competencies needed to meet the criteria for registration with the Nursing and Midwifery Council (NMC). The standards aim to assist approved education institutions (AEIs) in understanding the standards and how to meet them. It reviews briefly the background and context, and the location, design and delivery of programmes.

It is clear in the competencies for entry to the register for adult nursing, mental health nursing, learning disabilities nursing and children's nursing that graduating nurses are not likely to satisfy the competencies required if they do not have sufficient input during their training. Registered nurses and midwives make an important contribution to the promotion of health, health protection and the prevention of ill health. They do this by empowering people, communities and populations to exercise choice, take control of their own health decisions and behaviours, and by supporting people to manage their own care where possible. Registered nurses provide leadership in the delivery of care for people of all ages and from different backgrounds, cultures and beliefs'

At present nurses cannot be expected to address these needs due to the lack of formal teaching time allocated to the subject by their respective institutions.

The standards for the pre-registration nursing education (NMCb 2018) stated two of the outcomes that is expected to be reflected in an accountable and proficient practitioner in all care settings and areas of practice, two of these are:

> *"demonstrate an understanding of, and the ability to challenge, discriminatory behaviour" (NMCb 2018: 8)*
>
> *"understand the demands of professional practice and demonstrate how to recognise signs of vulnerability in themselves or their colleagues and the action required to minimise risks to health (NMCb 2018:8)*

The survey participants suggested that they will be unable to meet this standard since they are not being given the means in which to do so.

What's more the NMC Education Standard also expects student education to ensure that when individuals are qualifying they are able to:

> *"…provide and promote non-discriminatory, person centred and sensitive care at all times, reflecting on people's values and beliefs, diverse backgrounds, cultural characteristics, language requirements, needs and preferences, taking account of any need for adjustments" (NMCb 2018:9)*
>
> "…understand the factors that may lead to inequalities in health outcomes"
> NMCb 2018:11)
>
> *"…facilitate equitable access to healthcare for people who are vulnerable or have a disability, demonstrate the ability to advocate on their behalf when required, and make necessary reasonable adjustments to the assessment, planning and delivery of their care" (NMCb 2018: 26)*

Due to the chronic life-long nature of the illness patients with sickle cell disease and thalassaemia develop co morbidities that add complexities to their disease experience and outcome, therefore, health care professionals must be able and competent to address the challenges that these comorbidities pose.

The education standard expects newly qualified nurses to:

> "…understand and recognise the need to respond to the challenges of providing safe, effective and person-centred nursing care for people who have co-morbidities and complex care needs" (NMCb:26)

This report suggest that nursing and midwifery education programme providers are not truly embracing the benefits of diversity and inclusion; the lack of sufficient input on the subject of sickle cell and thalassaemia teaching will contribute to lack of knowledge, skills and competence of qualified nurses and midwives and affect their ability to advocate for and on behalf of their patients, provide optimal care and support in the healthcare setting.

In order to advance through the accredited course students will have to demonstrate progression so they can ultimately qualify for "Entry on the register", they do this by achieving "progression points".

"I'm in crisis" are three words that would trigger a student and qualified nurse and midwife to carry out an assessment, provide effective treatment, evaluate the effectiveness of care given and where necessary revisit the care given.

However, the educational standards need to be translated in order to achieve demonstrable knowledge, skills and competence. Those three words should be enough to ensure effective care is given and handicap, mortality and morbidity reduced in this client group.

Recommendations

The data suggests that student nurses and midwives are enthusiastic learners and provide a fertile ground for learning that ensures acquisition of life-long knowledge, skills and increasing competence to care for people with and at-risk of sickle cell and thalassaemia.

In view of the outcome of this survey, we make the following recommendations:

- Approved education institutions (AEIs) should incorporate the subject in all components of training. They can strive to achieve this by implementing pragmatic steps that could include:
  - AEIs should identify a subject champion within a group of universities who will support development of the subject within each institution
  - AEIs working with professional bodies and charities should develop a repository of learning resources and make them available for all institutions
  - AEIs should ensure the subject is incorporated into the assessment strategies nationally and in all areas of the curriculum
  - Increase the profile of the subject among university students generally for their own personal genetic needs
- AEI's should evaluate whether the existing omission of formal teaching time on these two conditions violates their respective equality and diversity policies. They should open this to consultation and work in conjunction with their student bodies.
- This is an opportune time to discuss these recommendations with the Council of Deans and individual schools of nursing and midwifery prior to their revalidation of nurse training programmes in 2019 and 2020. And the midwifery revalidation of

training currently going through consultation, for publication in 2019 and implementation in 2020.

- The Nursing and Midwifery Council should use their new Standards for pre-registration nursing education (2018) when revalidating all nursing curricula in 2019 to ensure that they are committed to addressing key aspects of equality and diversity. We urge to them to monitor more closely and enforce existing standards to ensure there is commitment from educational providers to maintain equity in educational subject content and evaluation.
- The Department of Health (DoH) should look at ways in which they can support the inclusion of these two under recognised conditions in post registration healthcare professionals continuing education programmes.
- The Department of Education (DofE) equality and diversity policy "aims to set an example as an employer and is developing policies that make opportunity equal for children and young people."[3] They do this by "making sure that meetings on different aspects of the curriculum involve partners from the various equality communities."[4] We consider this pledge offers an ideal platform in which to engage with this report's recommendations.

Conclusion

This preliminary data gives credence to the subjective view that sickle cell and thalassaemia education is not embedded in pre-registration nursing and midwifery training and inclusion is dependent on individual lecturers or schools of nursing. Some of this can be attributed to changes in nursing and medical education where there has been a move to spiral or problem-based learning. The subject can be built up during the years of training e.g. the genetics component may cover how these conditions are inherited and epidemiology may look at the incidence and prevalence of these disorders. In problem-based learning students are given cases in which they present their approach to managing the clients' problems to a group facilitator.

This means that their learning will contain the general principles of care but unless the facilitator is a specialist in a given area they may not touch on specific health conditions or join the links from different courses in spiral learning. Thus they are likely to explore these conditions superficially and it is only if the students' interest is sparked by a particular condition and they choose to find out more about it through self-directed reading and in practice placements that their knowledge and skills are expanded.

Apart from specialist community and public health training programmes it would be unusual for a student to be required to complete a profile of a neighbourhood where they would collect information on demographics and common health problems in order to gain some insight into the communities that they serve. This aspect is often limited to post registration students doing community nursing training. Health educators should use local information and intelligence to shape their curricula with the expectation that many of their students

[3] Equality and diversity https://www.gov.uk/government/organisations/department-for-education/about/equality-and-diversity

[4] Equality and diversity https://www.gov.uk/government/organisations/department-for-education/about/equality-and-diversity

will stay within the neighbourhood on completion of their training and be better able to meet local needs.

One cannot rely on the knowledge base of many qualified nurses and midwives if they have not had any formal teaching on the specialist subject. Service users experience remain variable and this is contributing to their increasing anxiety when they visit the healthcare environment. Services users need to be assured that when they seek treatment they can be confident it will be administered with aptitude. If the current state of play continues the situation will only worsen. We see these recommendations, as a way in which to not only advance awareness and understanding of sickle cell disease and thalassaemia, but as root and branch for reforms that will benefit both the health professional and their patients.

Future survey or research

It will be useful to replicate this survey by:

- Analysing the data separately of 1st, 2nd and 3rd year students
- Separating direct entry student midwives and post nursing registered student midwives
- Doing a survey of registered nurses and midwives

Since publication

We are pleased to announce that since the reports publication there has seen an increase from 197 self-selected respondents to 216, with the University of Central Lancashire and Middlesex University participating in the study. The survey is still open and accepting responses and is available at (https://docs.google.com/forms/d/1TrdzYJOSliw3Yvc4saDYpU19E9viPKpflsuKcX4YYVU); we plan to follow up the report with a revised data set in the near future.

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Appendix 1

Survey Process and Outcome

Methodology

To determine the prevalence of nurses and midwives not being taught about sickle cell and thalassaemia the project working group commissioned a survey that was circulated to all approved education institutions (AEIs). We had an overwhelming response with 197 self-selected responses from participating students nurses and midwives from 10 nursing schools: City University, Kingston University and St. George's University of London; University of West London, University of Huddersfield, University of Salford, University of Surrey, Nottingham University, Kings College London University, University of North West London, who all welcomed the engagement and agreed to circulate it to their respective nursing and midwifery student cohorts. It was also circulated to students via the Royal College of Nursing (RCN), students on placement in individual hospitals including the London NW University Healthcare NHS Trust and specialist sickle cell & thalassaemia Centres.

The survey was designed to decipher how much knowledge student nurses and midwives had in understanding these two genetic disorders and more specifically if they have had any formal teaching on sickle cell or thalassaemia, whether they have cared for clients and patients with these genetic conditions and if they feel confident to provide the care required by these client and patient group.

The survey report is authored by Dr. Lola Oni OBE chair of the working group and Joan Walters and Adam Lloyd, members of the working group.

The data collected was secure, protected, and covered by the privacy statement outlined on the Sickle Cell Society website, which adheres to the latest GDPR regulations. All respondents' information is confidential and hence they will not be made identifiable in the analysis that follows.

Results of Survey

Participant Population

Pie chart 1 and 2 below illustrates the composition of the academic institutions that made up the data set. The respondents as discernible from the evidence are from a variety of institutions both in terms of location and in terms of size. This aids confidence in providing a broad picture on the nursing and midwifery curriculum input. The data size allows us to be sure the sample size is adequately representative of the national picture. With most universities situated in the south east and or in urban densely-populated areas, we presume that the data would look much bleaker if we'd had engagement from rural areas where the prevalence of SCD and thalassaemia would be much lower due to the smaller minority ethnic population in those areas.

Chart 1 – Region in UK where Participants' University is located

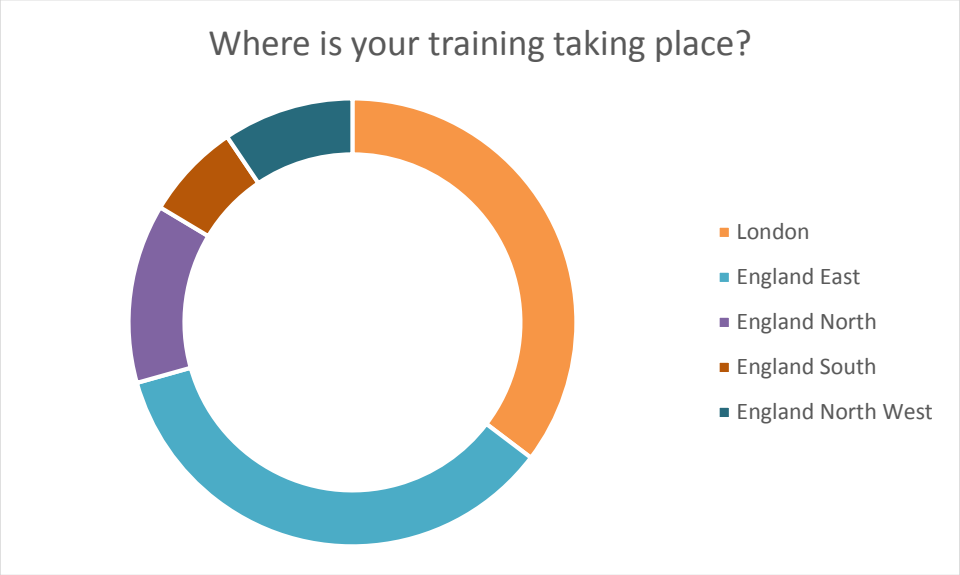
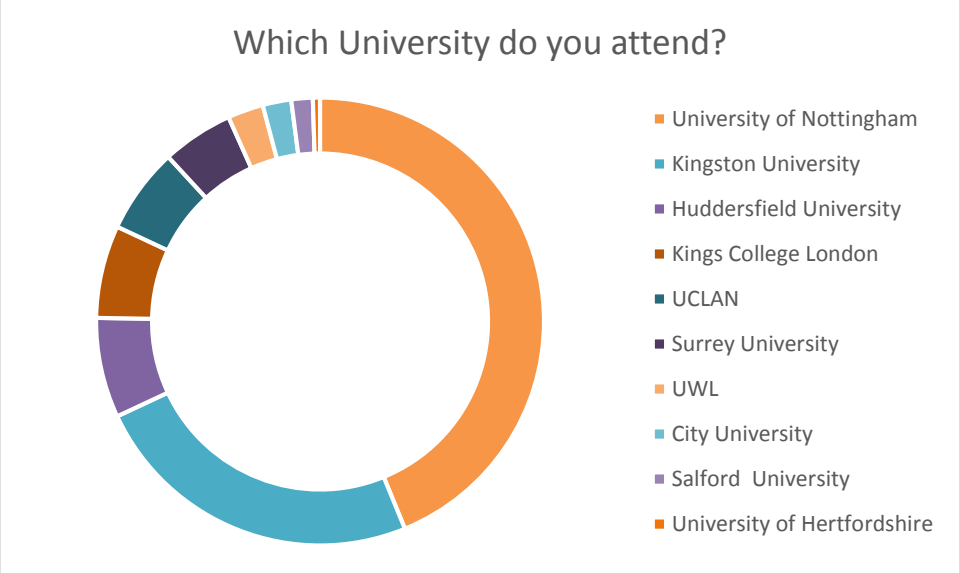


Chart 2 – University attended by participants



Not surprisingly there was greater response from students studying in London; this may have been influenced by the personal direct contact by the project team to lecturer colleagues in these universities and their encouraging their students to complete the survey.

Chart 3 – Composition of participants: Nursing Branch or Midwifery student?

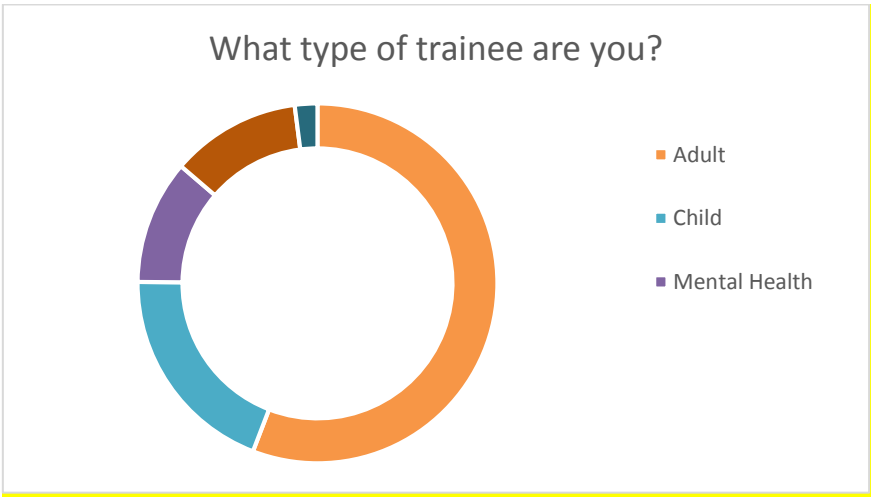


Chart 3 above shows the branch of nursing or if midwifery that the participants are pursuing with 75.1% in either adult or child nursing, whilst expectedly a smaller proportion are pursuing midwifery training. This is encouraging for the survey as these are traditionally the nurses who will be on the frontline dealing with people with sickle cell and thalassaemia.

Moreover, having participants in the cohort who specialise in mental health is equally significant as sickle cell disease and thalassaemia pose multiple and sometimes severe psychological challenges to patients. They are not only stigmatising medical conditions causing considerable socio-economic challenges but on a more individual level there can be severe psychological issues for some of these patients. It should be noted that many student midwives are qualified nurses, a further question may have been useful to separate direct entry student midwives from those with a nursing qualification already to see if there are differences in knowledge in these two groups.

Chart 4 – Did participants have an awareness of sickle cell?

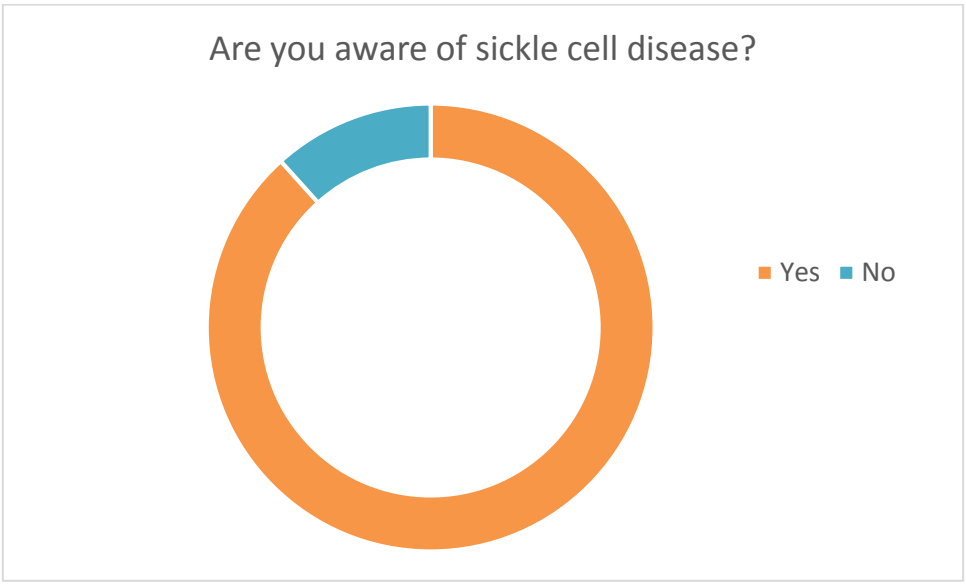
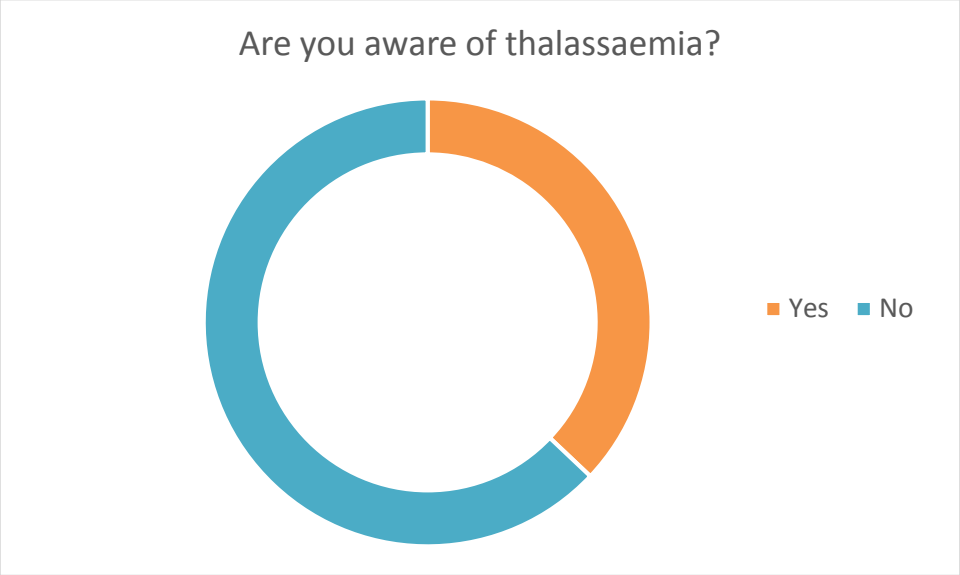


Chart 5 – Did participants have an awareness of thalassaemia?



The vast majority of respondents were aware of sickle cell but not thalassaemia. The chart above shows there is considerable disparity in awareness of sickle cell and thalassaemia, with 88.3% of respondents stating they were aware of sickle cell disease whilst 37.1% were aware of thalassaemia. This is a worrying gulf in knowledge around the two conditions and urgent attention need to be paid to increasing an awareness of thalassaemia within the nursing and midwifery community, especially in relation to genetics and the need for accessing timely antenatal screening, counselling and support services.

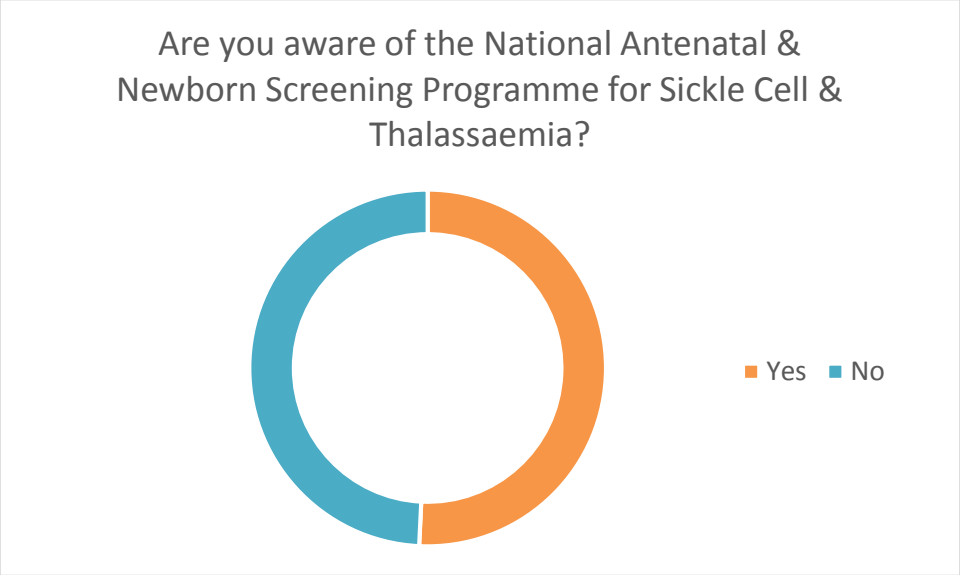
In free text responses, respondents expressed bewilderment at the lack of educational input in the subject and encouragingly expressed an appetite to learn about the two conditions:

> *"Needs to be more accessible training out there for student nurses and qualified staff nurses, as I am clueless on the matter"- Child nurse from the East of England*

> *"More training should be done to make students more aware of the condition" – Adult nurse from Greater London*

In regards to the National Antenatal & New-born Screening Programme for Sickle Cell & thalassaemia, there was a roughly 50:50 split between those who were and were not aware of the screening programme with slightly more (50.8%) being aware.

Chart 6 – Were participants aware of the national screening programme?



In order to ascertain the level of exposure to nursing and caring for people with and at-risk of sickle cell disease or thalassaemia students were asked to provide information on their level of experience to date. However it should be noted that some students were new to nursing and midwifery training and my not yet be in clinical practice hence exposure may be due to their stage of training. Secondly, some students are less likely to have exposure due to the small minority ethnic population in the region where they are receiving their training.

Chart 7 – Participants' teaching input on sickle cell disease

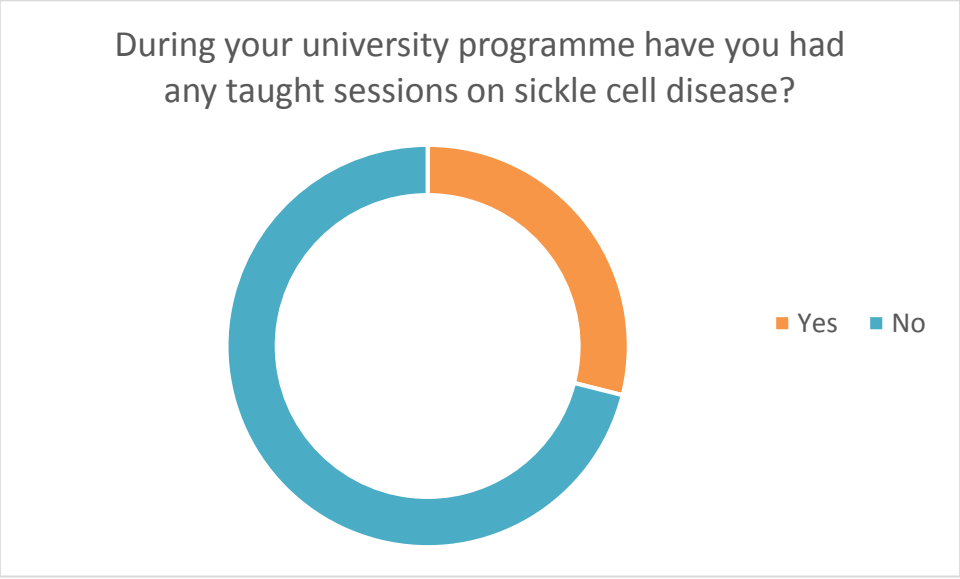
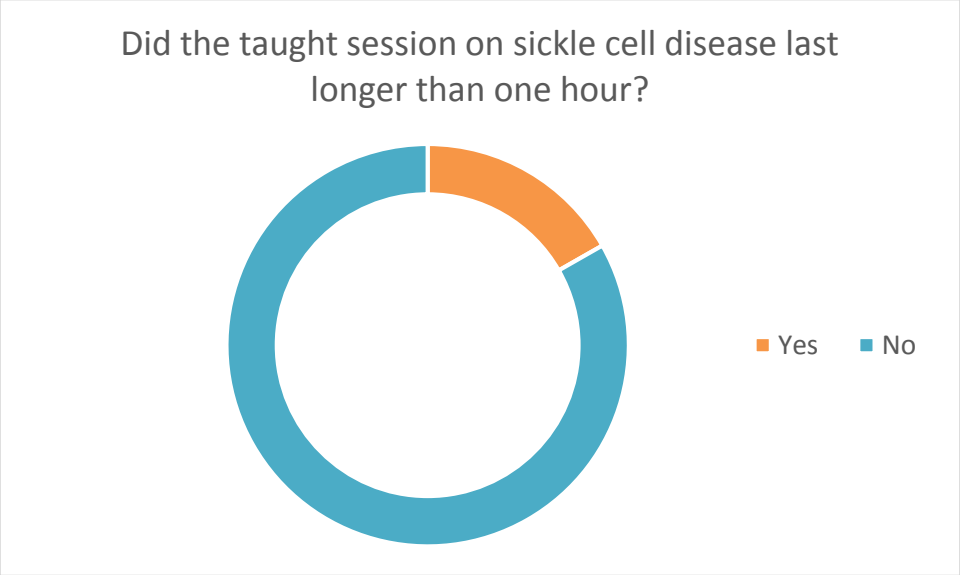
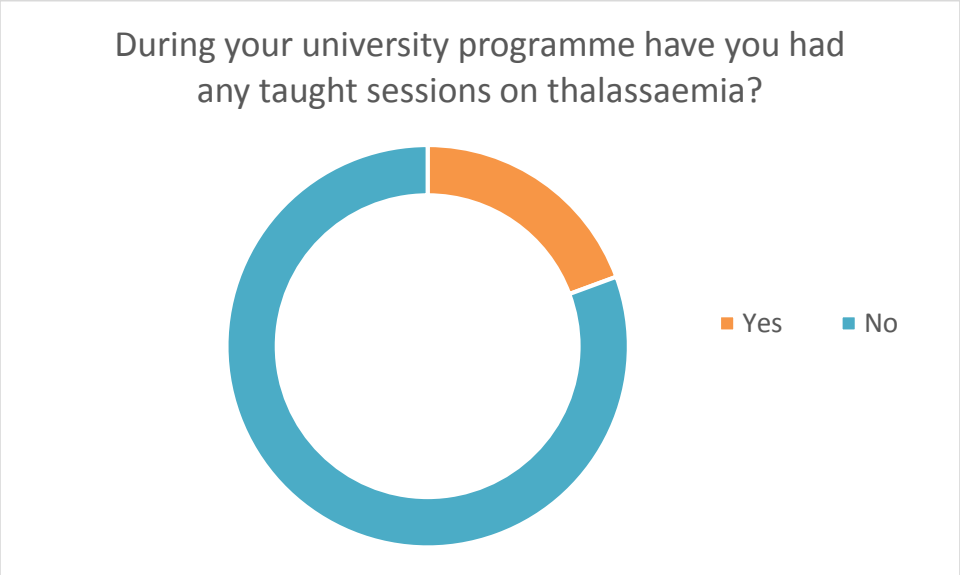


Chart 8 – Length of teaching session on sickle cell disease



A substantial 71.1% of respondents revealed that they had not had any formal teaching on sickle cell and of those that did only 16.7% of students had received a taught session of one hour or longer.

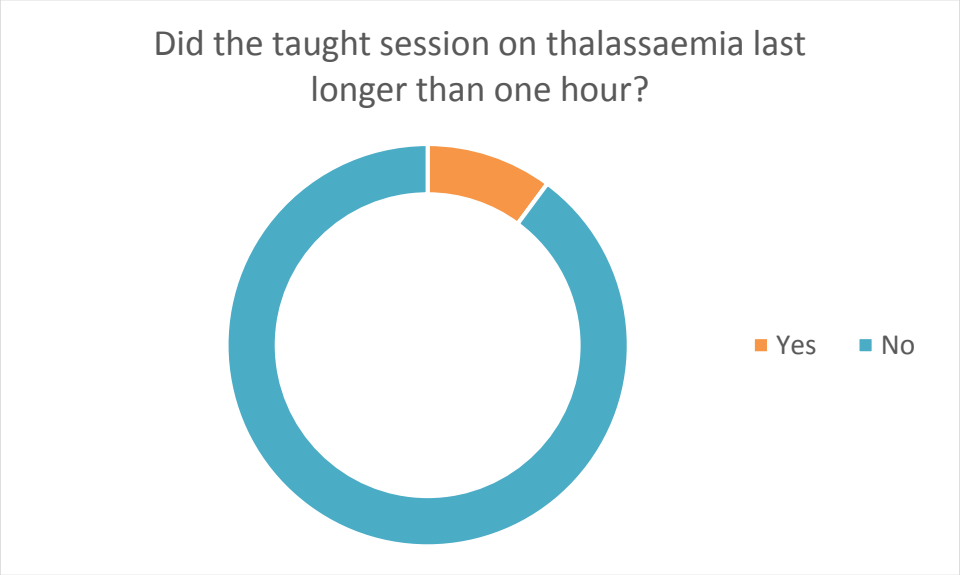
Chart 9 – Participants' teaching input on thalassaemia



Similarly, 80.7% of participants have not had any formal teaching session about thalassaemia, and of those that did only 10.2% had received a session of an hour or longer.

This clearly demonstrates either a complete absence of sickle cell and thalassaemia from the classroom or in many cases a very brief overview of less than one hour.

Chart 10 – Length of teaching session on thalassaemia



As stated previously this data is a mix of those starting their training and those nearing completion and it is possible that many of the students had not reached a stage in their training where the subject is due to be covered. None the less some of the students would be near completion and it will be useful to re-examine this cohort to ascertain the level of input to ensure nurses and midwives of the future are adequately prepared for their role as care givers.

When respondents were requested to offer free text responses they ranged from brief overviews offered by their respective institutions to a reliance on self-directed and independent research to attain knowledge:

> *"These conditions have not been mentioned in the course however most of the learning so far has focused on anatomy and we are only just starting to learn more about conditions in detail. The majority of the conditions addressed in the course have also tended to be those that can be linked to lifestyle or old age as opposed to those with more genetic causes" – Adult nursing student from the North of England*

> *"Sickle cell disease has been brought up briefly on occasions" – Adult nursing student from the South of England*

> *"What I learnt on sickle cell was from placement and my own research. The University did not touch upon this, when they did it was very brief and nothing substantial" – Child nursing student from Greater London*

Chart 7 – Nursing care of patients with sickle cell disease

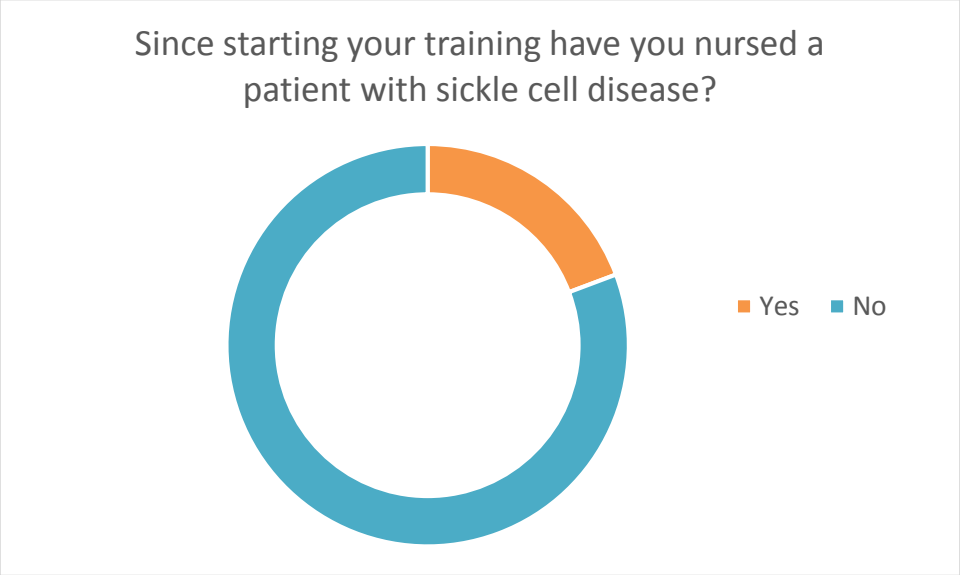
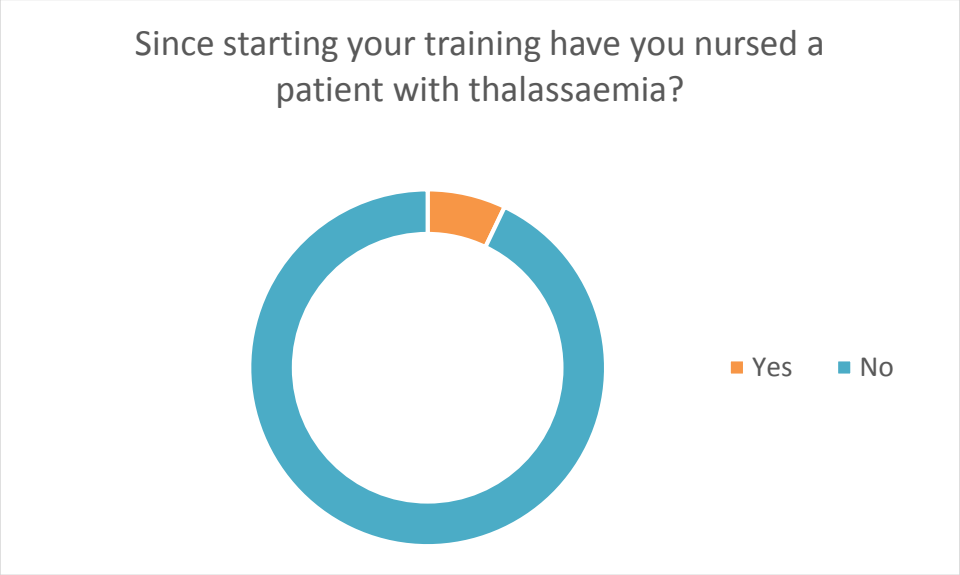


Chart 8 – Nursing care of patients with thalassaemia



Low number of respondents had nursed anyone with either thalassaemia or sickle cell disease; 80.7% of participants declared that they have not nursed a patient with sickle cell disease and 92.9% of respondents affirming they hadn't nursed a patient with thalassaemia. However, the caveat is that patients with thalassaemia are less likely to be admitted to hospital with an acute illness, the majority of their care is pre-planned and occur as routine Daycare admissions and they rarely have acute illness episodes however when they are acutely ill it can prove rapidly fatal. Secondly the lack of exposure could be due to the participants' stage of training and regional differences.

With an increasingly mobile population and so many trainees not having exposure to the subject in teaching sessions it means they will be less equipped to nurse these patients when they encounter them in a clinical environment post qualification. This places the

patients at-risk and exposes the nurse and midwife to making errors when required to care for the patient group especially in emergency situations. This is worrying especially in areas where there are large at risk populations and major cities where a significant proportion of these patients reside. The student nurses chance of encountering these patients post registration is relatively high.

In free text responses, one participant expressed her own awareness but was doubtful of fellow students, having any notion of this is as a specialist area:

> *"I did not receive any training in class about haemoglobinopathies, but am aware that they are a specialist area of Nursing, and that many of my classmates who may have trained may not have been in contact with any such patients during the entirety of their practice placements" – Adult nursing student training in London*

Chart 9 – Do participants feel confident to care for people with sickle cell disease?

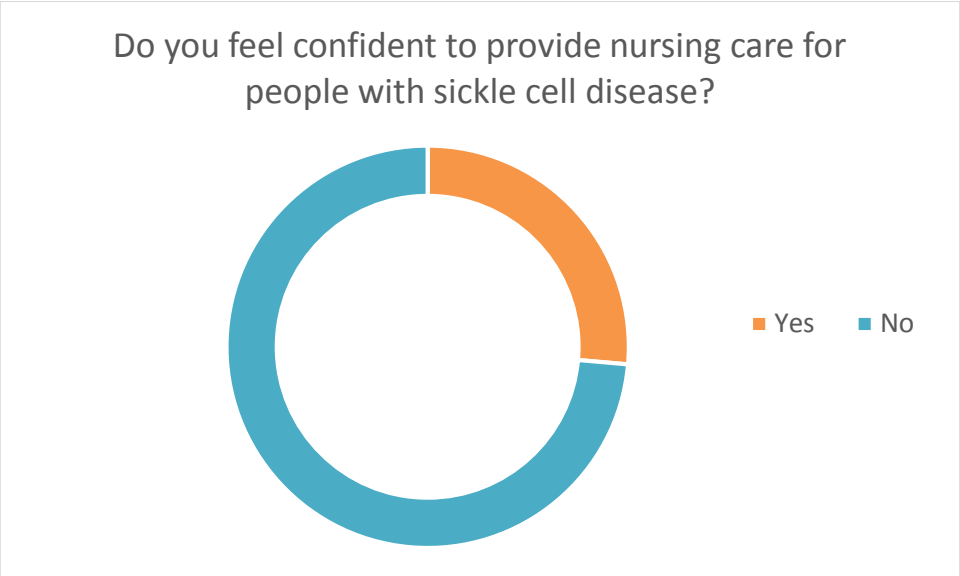
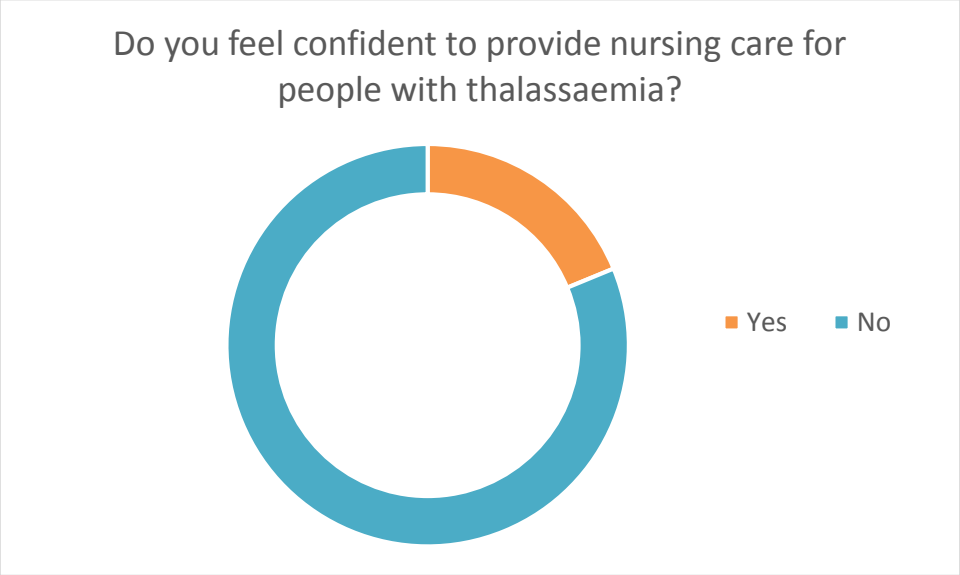


Chart 10 – Do participants feel confident to care for people with thalassaemia?



Again, we see a lack of assurance felt by respondents when asked to express their confidence if they were instructed to deal with these two conditions: with 26.4% of participants feeling confident to care for a patient with sickle cell disease and only 18.8% said they would feel confident to care for a patient with thalassaemia. As discussed previously level of exposure and development of competence is dependent on the stage of training and regional differences. However, educational input would ensure that students are prepared for an encounter with the patient group irrespective of the region in which they work in future. Exposure through teaching will also give them the confidence and ability to apply the principles of problem solving they have learnt in the classroom to the subject when they encounter this group of patients.

Patients need to be reassured that they will not be left vulnerable in a healthcare environment that purports to provide them with effective health care. There is evidence that many of these patients feel anxious at the lack of knowledge and the inability of nurses to care for them effectively. Student nurses and midwives become qualified nurses and midwives and it would be interesting to conduct a similar survey among qualified staff.

When participant were asked for personal experience to contextualise the findings they offered the following responses:

> *"I have heard of sickle cell anaemia but do not know what it is, how it affects an individual or the typical treatment offered" – Mental health nursing student from the East of England*
>
> *"I would feel more confident providing care for patients with this diseases if I had the training" – Adult nursing student from East of England*
>
> *"Although I have a bit of knowledge about both illness, I will not know what to do if I come across a patient with any of these diseases" – Mental health nursing student from Greater London*