Sickle Cell Community
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Summary

Sickle Cell disease introduction

- Sickle cell disease is the commonest genetic disorder in the UK. People affected cannot make enough normal haemoglobin (the protein that carries oxygen in the red blood cells), instead, an abnormal haemoglobin is produced, which changes its structure when oxygen levels are low.
- Sickle cell disease occurs when a person inherits two abnormal copies of the haemoglobin gene, one from each parent. A person with a single abnormal gene does not experience symptoms and is said to be a carrier with sickle-cell trait. The disease is more common in people descended from inhabitants of sub-Saharan Africa, Asia, the Middle East and the Eastern Mediterranean.

Sickle Cell disease key points

- Sickle cell disease is a serious and potentially life-threatening health problem.
- There are only limited means of preventing sickle cell disease. However, mothers are tested routinely during antenatal care for their sickle status. Infants are also screened for the condition.
- There is no widely available cure for sickle cell disease. However, there are several treatments which relieve symptoms and prevent or treat complications.
- There are fifteen people with sickle cell disease known to the NHS in Suffolk. There may be up to twelve further people with the diagnosis not known to the NHS.
- Sickle cell disease is stigmatised within some ethnic communities. One example is recent Portuguese migrants to Suffolk, who are affected by sickle cell disease but have only limited contact with services.
- There has been an apparent lack of awareness of the diagnosis in the local NHS.
- There are clear standards for NHS sickle cell services in England.
- Local screening services perform well.
- We found little information about primary care services.
- Hospital services for people with sickle cell disease are improving in Suffolk. However, there are apparent weaknesses in the clinical service for patients presenting to the emergency department.
- The numbers of children and adults in Suffolk admitted to hospital with sickle cell disease has been falling for the past three years.
Sickle Cell recommendations

1. Awareness of sickle cell disease in ethnic communities and in primary care needs improvement:

2. Suffolk’s clinical commissioning groups should address the issues in primary care, by leading and educating primary care staff to consider the diagnosis more often and perform the appropriate investigations.

3. The County Council should continue to support and foster the Thalassaemia and Sickle Cell Support Group. The Group should concentrate at first on promoting awareness of the disease, countering stigma and overcoming the apparent under-diagnosis of sickle cell disease.

4. Hospitals in Suffolk should continue their present efforts to improve sickle cell services. This should include consideration of a single tertiary hospital as a service partner, a clearer clinical pathway, better staff training and awareness, and clinical audit, including use of the National Haemoglobinopathy Register. They should signpost people to the Thalassaemia and Sickle Cell Support Group. The hospitals’ specialist clinicians should play a more active part in promoting awareness and understanding in the local community.

5. As the number of patients increases, it will become more appropriate for tertiary services to be provided locally, with specialist teams traveling to district hospitals rather than vice versa. This is more convenient for patients and families, and less expensive.

What is sickle cell disease and why is it important for Suffolk?
Sickle cell disease is the most common serious genetic disorder in the United Kingdom. It is an important serious and potentially life-threatening health problem, affecting an estimated 13,500 people, with an estimated 240,000 carriers (NHS Sickle Cell and Thalassaemia Screening Programme, 2015). People affected cannot make enough normal haemoglobin, the protein that carries oxygen in the red blood cells. Instead, an abnormal haemoglobin is produced, which changes its structure when oxygen levels are low. As a result, the red blood cells become distorted into a crescentic or sickle shape, are less malleable and have shorter life-spans (Figure 1).

Sickle cell disease occurs when a person inherits two abnormal copies of the haemoglobin gene, one from each parent. A person with a single abnormal gene almost never experiences symptoms and is said to be a carrier with sickle-cell trait. Carriers of sickle cell
disease have some resistance to malaria. For this reason, the disease is found in people descended from inhabitants of areas where malaria is common, such as sub-Saharan Africa. Sickle cell disease is however by no means confined to the Black population. It also occurs in people with Asian, Middle Eastern and Mediterranean ancestry. For example, in 2013/14, the condition was diagnosed in fifteen Asian infants and five White British infants (NHS Screening Programmes, 2014).

There are about 13,500 affected people in the United Kingdom. Sixty per cent of people in England with sickle cell disease live in London, and most of the others live in large conurbations, especially in Birmingham. In low-prevalence areas such as Suffolk, the needs of people with sickle cell disease may be inadvertently neglected because they are more easily overlooked.

Thalassaemia is a separate inherited disorder of haemoglobin structure which falls outside the scope of this needs assessment. Rarely, people are affected by both thalassaemia and sickle cell disease.
Figure 1: Blood flow in people with normal red cells and with sickle cell disease

A Normal red blood cells
- RBCs flow freely within blood vessel
- Normal red blood cell (RBC)

B Abnormal, sickled, red blood cells (sickle cells)
- Sickle cells blocking blood flow
- Sticky sickle cells
- Cross-section of RBC
  - Normal hemoglobin
- Cross-section of sickle cell
  - Abnormal hemoglobin form strands that cause sickle shape
Complications

- **Anaemia** is caused by the red cells’ short lifespan.

- **Sickle-cell crises** are sudden deteriorations in the health of a person with sickle cell disease. They occur when the blood supply to an organ is suddenly reduced by sickling. Crises are painful and often lead to organ damage.

- **Severe bacterial infections** arise because the disease leads to loss of spleen tissue and this reduces the body’s ability to clear bacteria from the bloodstream.

- **Stroke** is more common in people with sickle cell disease. Narrowing and blockage of blood vessels can prevent oxygen from reaching the brain. Cerebral haemorrhage can also occur. Strokes in people with sickle cell disease can occur at any age, are commoner in children and can be disabling or even fatal.

- **Gallstones** may result from the excessive rates of breakdown of sickled red blood cells.

- **Avascular necrosis** of the hip and other major joints may occur as a result of an inadequate blood supply.

- **Priapism** (prolonged and painful erections) occurs because of sickling of cells in the penis.

Prevention

There are only limited means of preventing sickle cell disease. Mothers are tested routinely during antenatal care for their sickle status. When a woman tests positive, the father is also tested. Couples at high risk of an affected baby are then offered pre-natal diagnosis. This provides early information to the parents, who may then choose to terminate the pregnancy. Couples can also be offered pre-implantation genetic diagnosis. Infants are screened for the condition so that appropriate treatment can be started promptly, and carriers identified.

Treatment

There is no widely available cure for sickle cell disease. However, there are several treatments which relieve symptoms and prevent or treat complications. The goals of treating sickle cell disease are to relieve pain, to prevent infections, organ damage, and strokes, and to control complications if they occur.

Infants who have been diagnosed with sickle cell disease through screening are treated with antibiotics to prevent infections, and are immunised against infection. Their parents are educated about the disease and how to manage it.
Many people with sickle cell disease take hydroxyurea, a drug which leads to the production of another type of haemoglobin. This helps prevent red blood cells from sickling and improves anaemia. People taking hydroxyurea therefore need fewer blood transfusions and have fewer hospital visits.

Blood transfusions are used to treat worsening anaemia and other sickle cell complications. They may also have a role in preventing complications such as stroke, spleen problems or acute chest syndrome. However, having routine blood transfusions can cause side effects, such as allergic reactions and an overload of iron in the body.

Adults who have sickle cell anaemia should be immunised against influenza every year, and also against pneumonia.

Sickle cell crises are treated with oxygen, blood transfusions, antibiotics, pain relief and rehydration.

**What is the local picture?**

**Results of screening**

Data on NHS screening services for sickle cell disease are published for each region (NHS Screening Programmes, 2014). Suffolk is part of the East of England region. In 2013/14, 81,303 blood samples from expectant women were processed in the region. There were 1120 (1.4%) positive tests, indicating that the woman was a carrier. Sixty-five high-risk couples were identified, in which both parents were sickle cell carriers. These results were similar to other parts of England with a comparable ethnic composition but lower than in London, where 5.2% of antenatal blood samples tested positive for sickle cell disease.

We obtained screening coverage results for Suffolk. Table 1 shows the proportion of pregnant women eligible for antenatal sickle and thalassaemia screening for whom a conclusive screening result is available. It shows good results, with improvement over the time period analysed.
This needs assessment was prepared in July 2015 by the Public Health Action Support Team on behalf of Suffolk County Council.

Table 1: Antenatal sickle cell and thalassaemia screening coverage, by Trust, 2012 to 2015

<table>
<thead>
<tr>
<th></th>
<th>2012/13 (%)</th>
<th>2013/14 (%)</th>
<th>2014/15 (%)</th>
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</thead>
<tbody>
<tr>
<td></td>
<td>Q1</td>
<td>Q2</td>
<td>Q3</td>
</tr>
<tr>
<td>Ipswich</td>
<td>100</td>
<td>96.5</td>
<td>100.2**</td>
</tr>
<tr>
<td>West Suffolk</td>
<td>97.7</td>
<td>98.7</td>
<td>98.1</td>
</tr>
</tbody>
</table>

Ipswich Hospital did not collect data.

** = There is a discrepancy recognised between the numerator and denominator, leading to a result exceeding 100%. The most likely explanation is that the initial sample cannot be analysed for technical reasons and a repeat sample is provided. This inflates the numerator but not the denominator.

Source: Public Health England

Table 2 shows the proportion of babies registered at birth, eligible for newborn blood spot screening and with a conclusive result recorded. It also shows high coverage, improving over time.

Table 2: Neonatal sickle cell and thalassaemia screening coverage, Suffolk PCT 2012 to 2014 and by Trust 2014 and 2015

<table>
<thead>
<tr>
<th></th>
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</thead>
<tbody>
<tr>
<td></td>
<td>Q1</td>
<td>Q2</td>
<td>Q3</td>
</tr>
<tr>
<td>Suffolk PCT</td>
<td>96.6</td>
<td>97.3</td>
<td>96.4</td>
</tr>
<tr>
<td>Ipswich</td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>West Suffolk</td>
<td>96.0</td>
<td>97.0</td>
<td>99.0</td>
</tr>
</tbody>
</table>

Source: Public Health England

Figures are not published on the results of screening for sub-regional populations. According to the 2011 Census, there were 117,400 people registered as Black in the East of England, of whom 6900 live in Suffolk (Office of National Statistics, 2014). Let us assume that the proportion of the Black population who have sickle cell trait, become pregnant and use antenatal services is equal across the region. If we then use Black ethnicity as an indicator of the overall population prevalence of sickle cell disease, we can expect about 65\textsuperscript{1} Suffolk

\[ \frac{6854}{117442} \times 1120 = 65.4. \]
women to have a positive result each year, and four\(^2\) high-risk couples to be identified. This is a crude approach, because the risk of sickle cell disease is not equal in all categories of Black people, nor is the disease confined to people of this ethnic group.

**Prevalence of sickle cell carriers and sickle cell disease**

The prevalence of sickle cell disease varies widely, depending on the proportion of people from specific ethnic groups and the prevalence in each of those groups.

There is no reliable means of obtaining accurate figures on the prevalence of sickle cell disease in Suffolk:

- We cannot rely on the numbers of people known to the NHS. Since the introduction of universal screening between September 2003 and July 2006, children born in the UK will almost certainly be diagnosed in infancy and can be enumerated. However, people born in the UK before then or born overseas may not have been diagnosed, or may not be in contact with services.

- We cannot readily apply information on the prevalence of sickle cell disease in specific ethnic groups to generate an overall prevalence estimate. Data on the ethnic composition of the County’s population and on the prevalence of sickle cell disease in specific ethnic groups are not sufficiently precise to enable us to calculate this reliably.

One approach is to base an estimate on the number of infants with the condition born in Suffolk and/or treated there. This number is the product of the ethnic composition of the population and the risk in each ethnic group, and so circumvents the problems with the approaches described above. However, it is still not straightforward to extrapolate from this to the number of people living in Suffolk with sickle cell disease. The total depends on the numbers of affected people born in, entering and leaving the County, and the longevity of people with sickle cell disease.

In 2014/15, two infants born in Suffolk screened positive for sickle cell disease. Both were born at the Ipswich Hospital. Importantly, of the fifteen people known to the hospital service in Suffolk with sickle cell disease, nine are under the age of sixteen years and at least two of the others are young adults who were diagnosed and treated as children. People with sickle cell disease often now live to around fifty years of age (NHS Choices, 2015).

\(^2\) \(\frac{6854}{117442} \times 65 = 3.79\)
The skewing of the age profile of diagnosed people towards a younger age suggests that there are adults with the condition as yet undiagnosed. Nine people with the diagnosis aged less than 16 years suggests perhaps another eighteen people above that age, of which only six are known to the NHS. However, this estimate should be treated with caution:

- Another explanation for the skewed distribution is that adults with sickle cell disease choose to live in higher prevalence areas, for cultural reasons or because health services are perceived to be of higher quality there.

- Some haematologists believe that it is unlikely that there are many people with untreated sickle cell disease. They argue that the disease is usually too symptomatic for this to be plausible. However, they suggest that there may be patients who attend hospital as emergencies for the treatment of sickle cell crises but are not seen regularly as outpatients.

- However, the local support group say that they are aware of twelve to fifteen adults in Suffolk who are not in contact with services.

Local support groups

There is a new support group for people in Suffolk with haemoglobinopathies, which received a grant from the County Council. Thalassemia Sickle Cell Support Suffolk first met in September 2014 with five members.

Members of the support group made these observations:

- Sickle cell disease is stigmatised within some ethnic communities. It is sometimes termed “bad blood”. Affected people and their families may ignore or suppress the existence of the diagnosis – even to the extent of denying it to a doctor – and not seek care because of fear of other people’s reactions.

- There are recent Portuguese migrants to Suffolk who are affected by sickle cell disease but are not seeking treatment.

- There has been an apparent lack of awareness of the diagnosis in the local NHS, particularly in primary care and the emergency department at Ipswich Hospital. Some people have received inappropriate care as a result.

In April 2015, members attended a meeting with executive directors of the Ipswich Hospital NHS Trust. The Trust agreed to add stickers to the notes of people with sickle cell disease to
ensure that staff were aware of the diagnosis, to design a care pathway for the condition and to implement training to improve staff awareness, especially in the emergency department.

What is the evidence base for interventions? What is best practice?

Services for children

*Sickle cell disease in childhood: standards and guidelines for clinical care* was published by the NHS in 2010 (NHS Screening Programmes, 2010). It sets out how specialist haemoglobinopathy teams should provide hospital services for sickle cell disease at:

- hospitals in urban areas with a large local population of children with sickle cell disease
- large district general or teaching hospitals in low prevalence areas with few local patients, but where geography dictates that they will need to provide most services for sickle cell patients. These hospitals act as a centre of expertise for patients in the surrounding areas.

The document sets standards for what should be available from a specialist haemoglobinopathy team. No hospitals in Suffolk fall into these categories.

All other district hospitals, such as those in Suffolk, should provide a local haemoglobinopathy service. This should:

- Have a named paediatrician to link to the specialist haemoglobinopathy team and neonatal screening laboratory
- Arrange initial contact with family and provide a paediatric clinic for routine outpatient management
- Promote and support management at home by parent, GP and community sickle cell and thalassaemia centre (if available)
- Manage acute pain and acute anaemia, and provide initial care for other complications before transfer to the specialist haemoglobinopathy team according to shared guidelines and protocols
- Liaise with the specialist haemoglobinopathy team for annual review
- Follow up children who fail to attend, reporting to specialist haemoglobinopathy team on annual activity as part of a network review meeting
- Liaise with local authorities, e.g. education and social services

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Manage transition to the adult service
Provide accurate, comprehensive and timely data to the NHS Sickle Cell and Thalassaemia Screening Programme to enable outcomes of newborn screening to be evaluated
Complete and update entries into National Haemoglobinopathy Registry when consent has been given.

The document makes these recommendations about the organisation of care for children with sickle cell disease:

- There should be a network of care based on local community care, including GPs, the local sickle cell and thalassaemia centre, health visitors, school nurses, the local hospital and specialised centre with links to paediatric intensive care where relevant. Parents should be put in touch with local and national voluntary organisations and local sickle cell and thalassaemia centres. There should be a named paediatrician responsible for follow-up in the local hospital.
- There should be a named paediatrician and/or paediatric haematologist in the specialist haemoglobinopathy team.
- General practitioners and community nurses should be regularly informed about patients’ progress.
- Parents should be encouraged to acquire knowledge about their child’s condition and should be informed about initiatives such as the Expert Patient Programme.
- There should be community paediatric services to coordinate the community needs of the child and to liaise with child and adolescent mental health services, local authority services and the voluntary sector as needed.
- Local authority services including education and social services should be aware of the specific needs of children with sickle cell disease and their families.
- Child and adolescent mental health services should be aware of the specific emotional and learning needs of children with sickle cell disease and their families.

These recommendations were based on clinical consensus in “the absence of directly applicable studies of good quality.” The document also makes specific recommendations about clinical practice.
Services for adults

*Standards for the Clinical Care of Adults with Sickle Cell Disease in the UK* was published by the Sickle Cell Society in 2008 (Sickle Cell Society, 2008). However, it appears to have official status, being published on the website of the NHS Sickle Cell and Thalassaemia Screening Programme and endorsed by the then Prime Minister and the Department of Health’s then Head of Blood Policy. The document identifies the following areas as most critical:

- acute pain
- acute complications
- detecting and managing chronic complications
- networks for care
- education and training
- adequate resources

The document then sets out standards for the care of adults with sickle cell disease under the following headings:

- organisation of care and commissioning sickle cell disease service
- managing acute complications
- managing chronic complications
- pregnancy, contraception and fertility
- blood transfusion
- surgery and specific therapies

These approaches are the basis for NHS England’s service specification for specialised services for haemoglobinopathy care.

The Royal College of Nursing (Royal College of Nursing, 2011) and the National Institute for Health and Care Excellence (National Institute for Health and Care Excellence, 2012) have published guidance on sickle cell disease.

**What is the pattern of services in Suffolk at present?**

Services for people with sickle cell disease are commissioned as follows:
Prevention

NHS screening services for sickle cell disease are commissioned by NHS England and described above.

Coverage of antenatal sickle cell screening and of newborn bloodspot screening form part of the public health outcomes framework, but neither data item is included in the most recent report on Suffolk, which covers 2013/14 (Public Health England, 2015). However, results at regional level are available (NHS Screening Programmes, 2014):

- Coverage at regional level is excellent, with 97.8% of eligible women tested.
- Fifty-eight per cent of samples are tested by ten weeks’ gestation. This is similar to other regions and above the acceptable threshold of 50%, but well below the screening programme’s achievable level of 75%.
- The proportion of eligible babies tested is 97.6%. This is similar to other regions and above the acceptable threshold of 95%, but below the achievable target of 100%.
- Two per cent of tests are avoidable repeats. This is low in comparison to other regions, on the acceptable threshold but above the achievable target of 0.5%.
- Results are available in timely fashion, with 99.7% communicated within six weeks, above the achievable target of 98% and one of the best results in England.

Primary care

This is also commissioned by NHS England, though some clinical commissioning groups are taking more responsibility for primary care services.

Primary care services include the provision of day-to-day support to people with sickle-cell disease, routine prescribing and the management of other health problems.

No data are available on the performance of primary care services for people with sickle cell disease. However, there are anecdotal reports that many general practitioners and other primary care staff lack experience in recognising and responding appropriately to the condition. While this is explained by the low prevalence of the condition in Suffolk, it has implications for the quality of care. For example, some of the Suffolk residents with undiagnosed sickle cell disease may present to primary care with symptoms attributable to the condition but without the diagnosis being considered. This may be more likely in people not obviously of Afro-Caribbean heritage. Greater awareness of the condition, and specifically of its occurrence in the non-Black population, would be valuable.
Secondary care

Services at district hospitals in Suffolk are commissioned by local clinical commissioning groups. They include emergency services and other care agreed with the specialist team on a shared care basis. Some patients may however make their way to their tertiary centre when they need emergency treatment.

In Suffolk, nine children and six adults are known to have sickle cell disease. Some of the children were born in Suffolk and diagnosed by neonatal screening, some were born and diagnosed elsewhere in the UK and moved to Suffolk and some were born and diagnosed overseas.

Children and adults can be seen at the Ipswich, West Suffolk and James Paget Hospitals with a shared care arrangement with either Addenbrooke’s Hospital in Cambridge or London hospitals including Central Middlesex Hospital in Acton, North Middlesex Hospital in Enfield and Kings College hospital. From July 2015, children treated in Ipswich will attend a specific haemoglobinopathy clinic at least twice a year.

No clinical audit information is available on the performance of these services.

Tertiary care

Specialist haemoglobinopathy services are commissioned by NHS England. The most important element of this is a multidisciplinary annual review. This may however be delegated or shared with a local hospital. There is a detailed service specification which describes how this service should operate (NHS England, 2013).

There are specialist haemoglobinopathy teams at Addenbrooke’s Hospital in Cambridge and at several hospitals in London.

Hospital activity analysis

We analysed hospital episode data about Suffolk residents admitted to hospital with main diagnosis of sickle cell disease.

Table 3 shows the number of Suffolk residents admitted to hospital at least once with a diagnosis of sickle cell disease. It shows a steep fall in admissions over the three years. This may indicate a declining prevalence of sickle cell disease, improving care with fewer
complications giving rise to admission, or a higher threshold for admission. The first of these explanations seems unlikely. Eighty percent of these admissions are for sickle cell crises.

Table 3: Patients admitted to hospital at least once with a diagnosis of sickle cell disease, Suffolk, 2012-13 to 2014-15, by age at admission

<table>
<thead>
<tr>
<th>Year</th>
<th>Unique patients admitted to hospital per year</th>
<th>Aged under 18 years</th>
<th>Aged 18 years or over</th>
</tr>
</thead>
<tbody>
<tr>
<td>2012-13</td>
<td>62</td>
<td>20</td>
<td>42</td>
</tr>
<tr>
<td>2013-14</td>
<td>37</td>
<td>8</td>
<td>29</td>
</tr>
<tr>
<td>2014-15</td>
<td>30</td>
<td>*</td>
<td>*</td>
</tr>
<tr>
<td>Total</td>
<td>129</td>
<td>*</td>
<td>*</td>
</tr>
</tbody>
</table>

Source: Hospital activity analysis

* Due to small numbers, these values have been suppressed.

2014/15 data are provisional and only include April 2014 to February 2015 inclusive.

This is corroborated by the data on admission numbers (Table 4). The fall in admissions affected children only and was continuous over all three years. For children, about two-thirds of the admissions were to Ipswich Hospital, with nearly all the rest being to Addenbrooke's Hospital in Cambridge. For adults, about half are to each hospital.

Table 4: Admissions to hospital with a diagnosis of sickle cell disease, Suffolk, 2012-13 to 2014-15, by age at admission

<table>
<thead>
<tr>
<th>Year</th>
<th>Admissions per year</th>
<th>Aged under 18 years</th>
<th>Aged 18 years or over</th>
</tr>
</thead>
<tbody>
<tr>
<td>2010/11</td>
<td>140</td>
<td>66</td>
<td>74</td>
</tr>
<tr>
<td>2011/12</td>
<td>123</td>
<td>50</td>
<td>73</td>
</tr>
<tr>
<td>2012/13</td>
<td>104</td>
<td>29</td>
<td>75</td>
</tr>
<tr>
<td>Total</td>
<td>367</td>
<td>145</td>
<td>222</td>
</tr>
</tbody>
</table>

Source: Hospital activity analysis

2014/15 data are provisional and only include April 2014 to February 2015 inclusive.
What additional information is needed?

- More complete information on the prevalence of sickle cell disease in Suffolk
- A clinical audit of paediatric and adult services at Ipswich Hospital, to check the extent to which they meet published standards.

What can be concluded?

The single most important finding from this needs assessment is the apparent under-diagnosis of sickle cell disease in Suffolk. Because of the previous absence of universal screening coupled with inward migration of affected people, there are probably about ten people with the condition in the County who are not known to the NHS. Some will be undiagnosed and others will be deterred by stigma from seeking help.

Some of these residents will have symptoms from their disease which treatment would reduce or abolish. All are at risk of complications which timely treatment could prevent. Antenatal and neonatal screening appears to function well in Suffolk. Although the performance of the screening programmes leaves some room for improvement, for example in the timeliness of testing, the programmes are successful.

We found little information about primary care services. The low prevalence of sickle cell disease in Suffolk means that members of primary health care teams have little experience of the condition. However, the likely presence of people with undiagnosed sickle cell disease in the County suggests that greater awareness would be valuable. It would make it more likely that people with symptoms possibly attributable to the diagnosis would be tested for it. It might also help reduce the stigma associated with the condition in some communities, by making it easier for affected people and their families to alert health care professionals about the possibility of the disease.

Hospital services for people with sickle cell disease are improving in Suffolk. Partly this is a result of rising numbers of diagnosed people, itself a consequence of the universal screening programmes and the increasing ethnic diversity of the County. There are however ways in which the service could be further enhanced:

- There are links to four tertiary centres. It might be more straightforward for patients and enhance professional working to have a partnership with a single centre.
There are apparent weaknesses in the clinical service for patients presenting to the emergency department. The Trust concerned has committed itself to resolving this, but this will need detailed attention.

The hospital is required to “promote and support management at home by parent, GP and community sickle cell and thalassaemia centre (if available)” (NHS Screening Programmes, 2010). At present, there is only limited engagement of hospital services with the wider community. In view of the stigma attached to the condition, the lack of public and professional awareness and the nascent state of the local support group, there is an opportunity for the hospital’s clinical staff to make an important contribution to improving awareness and understanding of sickle cell disease. This may assist in reaching the people as yet undiagnosed.

No clinical audit is apparent.

The support group for people with sickle cell disease is at an early stage of development. It has an important potential contribution to raising awareness and combating stigma, but will need continued investment of effort and resources to achieve its goals.

References