
SICKLE CELL AND THE BURDEN OF DISEASE IN ENGLAND



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ABSTRACT

Background

Sickle cell disorders (SCD) are the most common inherited blood disorders in England. Without prompt diagnosis and proper treatment, they can be a serious source of morbidity and mortality. Sickle cell diseases affect mainly black minority and ethnic populations, and have so far received relatively low priority from a health policy perspective. Antenatal and newborn screening, the development of minimum standards, antibiotic prophylaxis, comprehensive immunisations, and preventive diagnostic tests have positively influenced SCD management. There remains an unclear picture as to the trends and health care utilization of patients with SCD in England.

Aims

This study looks at the burden of disease in England by assessing hospital admissions, readmissions and related costs. It also aims to identify gaps in care and prevention which may identify possible contributors to avoidable admissions.

Findings

Using Hospital Episode Statistics (HES) data, trends for SCD hospital admissions in England showed a rise in 50% of hospital admissions over a 10 year period. The most deprived areas had a higher rate of readmission and in-patient mortality among those with SCD. Adolescents had a higher rate of readmission possibly identifying a gap in health care access. Local findings in a high prevalence area showed that the majority of admissions were for a short length of stay and 74% of patients accounted for multiple admissions. A patient focus group and questionnaire both identified potential gaps in care and prevention.

Conclusion

Through the use of 6 studies which showed the SCD admission rates in England, the readmission rates, local admissions, costs associated with admission and patient perspectives in both care and prevention, there is a clearer picture as to the trends and health care utilization of patients with SCD in England. The studies suggest that ascertaining the prevalence of at-risk groups in England as well as addressing inequalities in health care access among minority groups and areas of high disease prevalence can further aid in disease management. Shifting diagnostic and follow-up care from acute care facilities to primary care facilities and promoting preventive care measures and adherence to standards and guidelines may possibly decrease the cost burden, reduce avoidable hospital admissions and increase the timeliness and effectiveness of disease management. Investing in training and education of primary care physicians for sickle cell diseases may also improve quality of care.

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ABBREVIATIONS AND ACRONYMS

CCGs	Clinical Commissioning Groups
CFHI	Canadian Foundation for Healthcare Improvement
CLAHRC	Collaboration for Leadership in Applied Health Research and Care
DH	Department of Health
GP	General Practitioner
HBSS	Haemoglobin SS
HRG	Healthcare Resource Group
IOM	Institute of Medicine
LINKs	Local Involvement Networks
LOS	Length of Stay
NCEPOD	National Confidential Enquiry into Patient Outcome and Death
NHS	National Health System
NICE	National Institute for Health and Clinical Excellence
NIHR	National Institute for Health Research
PALS	Patient Advice and Liaison Service
PCTs	Primary Care Trusts
PCMH	Patient-centred Medical Home
PPGs	Patient Participation Groups
QOFs	Quality Outcomes Framework
RBC	Red Blood Cell
SCD	Sickle Cell Disease
SHA	Strategic Health Authority
WCC	World Class Commissioning
UK	United Kingdom
US	United States

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STATEMENT OF CONTRIBUTION

The work presented in this thesis is my own, but I received direction from my supervisor, Azeem Majeed. Anthony Lavery advised on the statistical analysis of the studies and supported all the analyses that used the Hospital Episode Statistics data. Publication outputs were contributed to by the CLAHRC SCD project team members.

1. RATIONALE FOR THESIS

At the start of this study, what was known is that Sickle Cell Disease is the most common genetic disorder in England; that two national standards have been published –paediatric and adult and that the National Confidential Enquiry into Patient Outcomes and Death was critical of the role of general practitioners and primary care trusts.

In May 2008, the National Confidential Enquiry into Patient Outcome and Death (NCEPOD) released a report entitled *A Sickle Crisis?* (Report) addressing the current challenges in Sickle Cell Disease (SCD) management in England. The NCEPOD investigates patient mortality and morbidity in the United Kingdom (UK), gathers baseline data and makes recommendations for practice for health care providers. The NCEPOD reviewed the lives of SCD patients and deaths due to SCD over two years and made recommendations on future practice. The review on deaths gathered information on why these deaths were occurring and whether addressing clinical care pathways could result in improved patient outcomes. The review on current clinical care practices in living patients found that the bridges linking these networks were fragile at best. The Report highlighted key findings and principal recommendations, including some aimed at health care providers and others at health policy makers.

The NCEPOD Report gives purpose behind the investigative nature of this thesis and has support from the Collaboration for Leadership in Applied Health Research and Care (CLAHRC) programme. CLAHRC was established by the National Institute for Health Research (NIHR) to undertake high-quality applied health research focused on the needs of patients and to support the translation of research evidence into practice in the NHS. One of the areas selected for improvement is SCD.

1.1. SICKLE CELL DISEASE

This first chapter gives a concise description of SCD, its history, discovery in the West, the consequences of the disease, and the ways in which the disease is currently being clinically managed. The objective is not to paint a clinical picture of SCD or its treatment options. The objective is to emphasise three things for the reader:

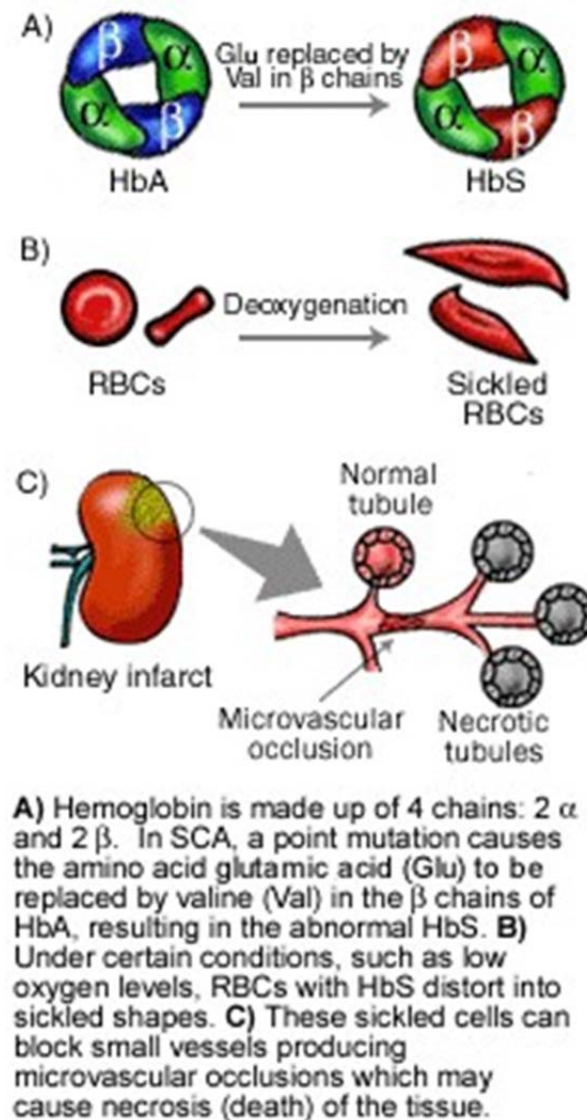
1. The importance of the disease from a public health perspective.
2. That the disease has the potential of being underserved from a public policy perspective.
3. That appropriate interventions could potentially reduce the burden of disease.

1.1.1. DEFINITION

Sickle cell disease, also known as sickle cell anaemia or sickle cell disorder, is an inherited blood disorder, also referred to as a haemoglobinopathy. The genetic disease is biochemically caused by a single amino acid substitution of valine for glutamic acid in the sixth position of the beta (β) chain of the haemoglobin tetramer (Pack-Mabien & Haynes, 2009; Pace & Zein, 2006). The disease damages and changes the shape of red blood cells (RBCs). The change in shape is a response to cell deoxygenation. When the oxygen uptake of the cell is low, cells change their shape from a healthy round disk to a crescent, holly leaf or other similarly distorted shape. This shape distortion is referred to as sickling. Hence, the disease is known as sickle cell disease. The sickled cells are rigid, less malleable and stickier than normal, healthy cells, so they consequently may stick to each other and obstruct blood vessels or are not sufficiently malleable and obstruct blood vessels. This obstruction causes harsh and painful complications. Often, these red blood cells will also break down and cause anaemia, so we also refer to the

disease as sickle cell anaemia. Consequent to the breakdown of red blood cells (haemolysis), cell survival may be reduced to as little as twenty days (Wilson, Krishnamurti & Kamat, 2003), whereas a normal red blood cell will last anywhere from 110-120 days (Allison, 1960). Haemolysis also causes jaundice, aplastic crisis (where the red blood cell does not mature so anaemia is worsened), and retarded growth and development in children.

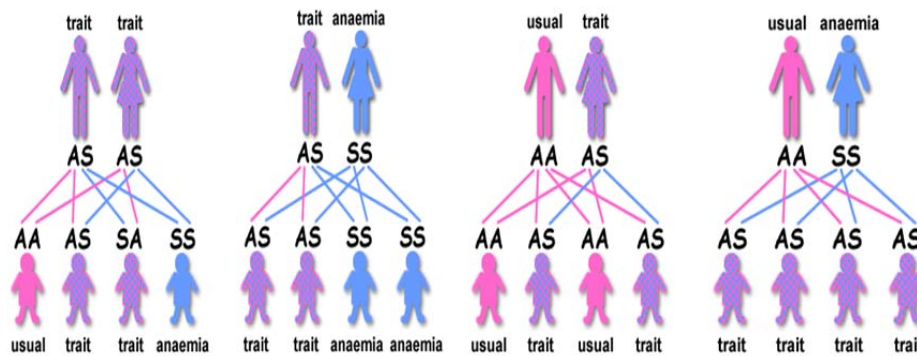
FIGURE 1: DEOXYGENATION OF THE RED BLOOD CELL



Source: <http://1.usa.gov/1ltZlPz>

Haemoglobinopathies, which consist of sickle cell and thalassaemia disorders, are passed to the next generation in an autosomal recessive manner (Pack-Mabien, Labbe, Herbert, *et al.*, 2001a). Those who are heterozygous for the haemoglobinopathies are trait carriers for sickle cell and, aside from some cases presenting with mild anaemia, show none of the clinical manifestations of the disease. Individuals who are homozygous for the haemoglobinopathies (designated in Figure 2 as with 'anaemia') will present with one of the sickle cell diseases (Wilson, Krishnamurti & Kamat, 2003).

FIGURE 2: GENE COMBINATIONS FOR SICKLE CELL DISEASE



AA = two normal genes • AS = one normal gene (A) and one sickle gene (S) SS = two sickle genes and with disease

Source: sickle cell society website (<http://bit.ly/1KAPcOg>)

As pictured in Figure 2, if both the mother and the father are sickle cell trait carriers ('trait'), there is a 1 in 4 chance (25%) of having a child born with the disease and a 2 in 4 chance (50%) of having a child born with the trait. If one parent has the trait and the other parent actually has the disease, there is a 2 in 4 chance (50%) of having a child born with the disease and a 2 in 4 chance (50%) of having a child born with the trait. If one parent is normal ('usual') and the other parent has the trait, there is a 2 in 4 chance (50%) of having a child born with the trait. If one parent is usual and the other parent has the disease, there is a 4 in 4 chance (100%) of having a child born with the trait.

There are several forms of SCD. The most common is haemoglobin SS (HbSS), also known as homozygous sickle cell anaemia. Patients with haemoglobin SS inherit one sickle cell gene from each parent. Other forms are heterozygous combinations of haemoglobins such as haemoglobin SC, haemoglobin SD, haemoglobin SO and haemoglobin S/β thalassaemia. The severity of sickling is proportional to the percentage of haemoglobin S present. Hence, HbSS is the most severe form of the disease (Standards for the Clinical Care of Adults with Sickle Cell Disease in the UK, 2008).

HbSS is the most clinically significant abnormal haemoglobin condition. It results when the gene for sickle haemoglobin is inherited from both parents. The predominant haemoglobin is Haemoglobin S (HbS). The infant appears normal at birth. The reason for this 'false normalcy' amongst newborn sickle babies is due to foetal Haemoglobin (Haemoglobin F). During the last seven months of pregnancy, Haemoglobin F is produced by all babies and is responsible for transporting oxygen around the baby's body. After birth, the baby continues for several weeks to produce Haemoglobin F as he/she starts to produce and build up its reservoir of adult Haemoglobin (Haemoglobin A). In a sickle cell baby, the baby also continues to produce Haemoglobin F but he/she also starts to produce the sickle Haemoglobin (Haemoglobin S).

Hence, anaemia develops within the first few months as Haemoglobin F decreases and Haemoglobin S increases (Orkin & Higgs, 2010a). The anaemia is usually moderate to severe. There are several other potential complications that will be discussed in further detail later. The topic of foetal haemoglobin will also be revisited again in the therapies section with regards to hydroxyurea as a treatment option for SCD (Richer & Chudley, 2005).

1.1.2. SICKLE CELL TRAIT

Sickle cell trait (HbAS) is a carrier state which, unlike HBSS, does not give rise to a significant clinical presentation. However, it has been reported that those with sickle cell trait are more prone to certain conditions and complications. Health care practitioner knowledge of these associations is critical for proper management. For example, renal medullary cancer, a rare but fatal form of kidney tumour, is seen almost exclusively in young individuals with sickle cell trait (Tsaras, Owusu-Ansah, Boateng, *et al.*, 2009). The trait is also linked to exercise related deaths from overexertion, splenic infarction at high altitudes such as in airplanes and mountains (Embury, 1986), and hyposthenuria, which is an inability to concentrate urine that often leads to dehydration (Tsaras, Owusu-Ansah, Boateng, *et al.*, 2009). There is also an increased rate of bacteriuria, pyelonephritis and haematuria, especially during pregnancy. Screening for these potential complications is essential and a positive results need to be treated with antibiotics promptly and rigorously (Serjeant & Serjeant, 2001). The good news about sickle trait is that there is a reason to believe that it came about due to genetic evolution as it offers some protection against contracting malaria in malarious regions of the world, which is discussed in more detail below.

1.2.HISTORY

1.2.1. GENETIC EVOLUTION OF SCD

At first, it was thought that the sickle cell gene mutation occurred during Neolithic times (also referred to as the Stone Age) around 10,000 BC in the Arabian Peninsula. During these times, humans started living in close proximity to each other and to a range of animal species, thus the period is attributed to a number of epidemics and the evolution of serious diseases (Khelil, Perrin, Lefranc, *et al.*, 2012). It was thought that with the migration of people, the sickle gene also migrated (Lehmann, 1954). However, while there is truth to the migration theory, there is also more scientific evidence that suggests the sickle cell mutation in fact occurred as several independent events giving rise to different types of the gene mutation (Desai & Dhanani, 2004).

The HbS gene is associated with at least four different chromosome structures (haplotypes) that represent independent sickle cell gene mutations, including the Benin haplotype, the Senegal haplotype, the Bantu haplotype and the Asian haplotype. Each haplotype is named based on the region where the original sickle cell gene mutation occurred (Pagnier, 1984). These different haplotypes explain why it is more common to see chromosomal heterogeneity in the West as compared to the homozygous conditions more likely to be seen in Africa, Arabia or Asia (Desai & Dhanani, 2004). The slave trade in the United States brought the Bantu haplotype found in the African region, but after the arrival of this population to the United States, this gene type mixed with gene types arriving from the Caribbean and Brazil, creating a mixture of African ethnic groups (Nagel, Erlingsson, Fabry, *et al.*, 1991).

FIGURE 3: ORIGINS OF THE SICKLE CELL GENE



Source: www.sicklecellinfo.net (accessed 29 July, 2010)

FIGURE 4: GLOBAL DISTRIBUTION OF THE SICKLE GENE



Source: (Oni, 2007 – Parent's Handbook)

1.2.2. THE ROLE OF MALARIA IN PERPETUATING THE SICKLE GENE

Plasmodium falciparum malaria is the most dangerous of the four species of human malaria and considered to be a killer. In places where *P. falciparum* malaria is endemic, populations have been wiped out. Studies have shown that malaria could have contributed to the fall of the Roman Empire by killing children and adults in villages during a time when the parasite made its way to parts of Europe. Much later, mosquito eradication programmes were carried out in Europe and the United States (Sallares, Bouwman & Anderung, 2004).

When the *falciparum* malaria parasite attacks an HbAS erythrocyte (Sickle Cell Trait), the cell sickles at a much higher rate than a non-parasitised cell. Once a parasitised cell is sickled, it is removed from circulation by **phagocytosis**, which is the main mechanism whereby a person

with sickle cell trait has a selective advantage against malaria (Luzzatto, Nwachuku-Jarrett & Reddy, 1970). Those who survive malaria are able to pass on their genes. Hence, it has acted as a selective factor by increasing the prevalence of the gene in areas where malaria exists. Over the generations, the sickle cell trait has therefore reached high frequencies in countries with endemic malaria. The factor in common to the distribution of the sickle cell gene is therefore malaria (Serjeant, 1997).

Although the sickle cell trait may protect against malaria, inheritance of two abnormal genes leading to SCD offers no such protection, and malaria is a major cause of ill-health and death in people with SCD (WHO Secretariat, 2006). This is thought to be partially due to the spleen, as the spleen plays an important role in malaria immunity. Therefore, increased morbidity and mortality from malaria in people with SCD increases as spleen function deteriorates with age in patients with SCD. A second reason is that SCD patients, even without malaria, are already anaemic. Malaria can cause anaemia, and a sudden decline in haemoglobin from an already low baseline value could be disastrous for a patient suffering from both SCD and malaria (McAuley, Webb, Makani, *et al.*, 2010).

FIGURE 5: MAP OF THE DISTRIBUTION OF MALARIA AND THE SICKLE CELL ALLELE



Map 1: Distribution of malaria in southern Europe, southwest Asia, and Africa around the year 1920 (green), prior to mosquito eradication programmes.

Map 2: Distribution of the sickle cell allele within the same area. The darker the blue, the greater the percentage of people carrying the allele.

Note the correlation between these maps.

Source: Understanding Evolution. 2010. University of California Museum of Paleontology. 22 August 2008
<http://bit.ly/1KXpkg8>

1.2.3. DISCOVERY IN THE WEST

The first record in the western world of a sickle cell disease blood sample was made by Chicago physician James Herrick in 1910. Herrick describes the patient's red blood cells as 'very irregular, but what especially attracted attention was the large number of thin, elongated, sickle shaped and crescent-shaped forms.' At the time, Herrick could not diagnose the patient, but rather just gave an account of his case (Herrick, 2001; Savitt & Goldberg, 1989). While this was the first published case in the west, there is evidence that suggests that many families in Africa were able to trace the disease through their family genealogy and gave the disease tribal names based on the symptoms of sickle cell disease. In the south of Nigeria, sickle cell disease was called 'arun romolegungun' which means 'disease that aches the child's bones' which is in essence a description of sickle cell crisis pain (Brent Sickle Cell & Thalassaemia Centre, <http://bit.ly/1J5Hz46>).

The next one hundred years (1910-2010) showed a vast improvement in the understanding and treatment of the disease with the discovery of the deoxygenation of the cells, gene mutations,

the protective nature of foetal haemoglobin, and the discovery of the chemotherapy drug hydroxyurea in the treatment of painful crisis.

FIGURE 6: SCD TIMELINE 1910-2010

- **1910**-Herrick provides the first formal description of sickle cell anaemia when he reports that the blood smear of a dental student at the Chicago College of Dental Surgery contains 'pear-shaped and elongated forms.'
- **1927**-Hahn and Gillespie associate the sickling of red blood cells with low oxygen conditions.
- **1940**-Sherman reports that the sickling of red blood cells in the absence of oxygen is caused by a change in the haemoglobin molecular structure.
- **1948**-Watson suggests that the presence of foetal haemoglobin in the red blood cells of sickle cell newborns is the reason they do not show disease symptoms.
- **1949**-Noted physical chemist Linus Pauling and associates publish 'Sickle Cell Anaemia, a Molecular Disease' in *Science*. This paper explains how protein electrophoresis was used to show that sickle cell haemoglobin differed in structure from normal haemoglobin. This was the first time that the cause of a disease was linked to a change in protein structure.
- **1956**-Vernon Ingram and J.A. Hunt sequence haemoglobin and discover that the change of a single amino acid in the protein sequence is the cause of sickle cell anaemia.
- **1978**-Flavell prepares maps of the human beta and delta globin genes.
- **1995**-Upon the completion of the Multicentre Study of Hydroxyurea in Sickle Cell Anaemia, Charache reports that the anticancer drug hydroxyurea is the first to reduce the frequent, painful complications that characterise sickle cell disease.

Source:

Online *Mendelian Inheritance in Man*, OMIM (TM). Johns Hopkins University, Baltimore, MD. MIM No. 141900 (6 November, 2001). <<http://goo.gl/qdoVoj>>

A Brief History of Sickle Cell Disease. Joint Center for Sickle Cell and Thalassemic Disorders (25 February, 2001). <<http://goo.gl/2AD9xH>>

Timeline taken from: <http://goo.gl/U15GH6>

1.3.INCIDENT AND PREVALENCE

1.3.1. GLOBAL INCIDENCE

There are an estimated 3 to 5 million people globally affected by SCD (Vichinsky, Neumayr, Gold, *et al.*, 2010; Steenhuisen, 2010), and there are 300,000 births per year of babies with SCD, with up to 70% of these births taking place in Africa (Makani, Komba, Cox, *et al.*, 2010). SCD is most common in people originating from Africa, the Mediterranean, the Middle East, India, Caribbean and Central and South America. All people of non-Northern European origin are therefore considered to be at greater risk of inheriting sickle cell disorders, but it is not exclusive to these populations and can affect people of any race, colour, or ethnic origin (Lehmann, 1954).

In the United States of America, SCD affects around 72,000 people, most of whose ancestors come from Africa. The disease occurs in about 1 in every 500 African-American births and 1 in every 1000 to 1400 Hispanic-American births (Genomic resource centre, <http://bit.ly/1ItZl0j>).

1.3.2. TRENDS IN THE INCIDENCE AND PREVALENCE OF SCD IN ENGLAND

SCD is the most common genetic condition in England. It is estimated that there are around 12,500 people with SCD living in England; 9,000 (3/4) of them live in London, and one baby in every 2,000 is born with the condition (<http://bit.ly/1cpGpDM>, Streetly, Latinovic, Hall, *et al.*, 2009). In London, SCD is now one of the most common reasons for admission to hospital (Streetly *et al.*, 2008, McColl Karen Pickworth & Raymond, 2006).

The significant migration of people from the West Indies, Africa and Asia during the 1950s and onwards may partially account for the high incidence of SCD in England. Figures also suggest that 6% of the population and approximately 10% of all births are at risk for haemoglobinopathy disorders (Anionwu & Atkin, 2001). With current trends in population migration into the country, these conditions are likely to become even more common over the next decade (Streetly, Clarke, Downing, *et al.*, 2008).

1.3.3. INCIDENCE OF SICKLE CELL TRAIT

It is estimated that 300 million people worldwide carry the sickle cell trait, with the highest concentration in Africa and the Mediterranean region. Approximately 1 in 3 persons in West Africa (33%) and 1 in 5 persons (20%) in the eastern province of Saudi Arabia are carriers (Tsaras, Owusu-Ansah, Boateng, *et al.*, 2009). Approximately 1 in 100 Greeks carry the sickle cell trait (Brent Sickle Cell & Thalassaemia Centre, <http://bit.ly/1J5Hz46>). In the United States, about 2 million people carry the trait (Genomic resource centre, <http://bit.ly/1ItZl0j>) and in England, around 9,723 carriers are born each year (NHS sickle and Thalassaemia Screening Programme, 2011/12).

1.3.4. PUBLIC HEALTH IMPLICATIONS OF GROWING PREVALENCE

There is a growing global public health concern regarding SCD due to a marked increase in prevalence over the past decade. The increase in sickle numbers is likely due to a number of factors including a rise in screening programmes, a move towards incorporating various haemoglobinopathy diagnosis codes, and population migration (WHO Secretariat, 2006). The increase is also due to an increase in life expectancy of patients living with sickle cell through improved secondary and tertiary prevention, including improving managing risk factors associated with haemoglobinopathies and earlier detection and management of disease-related complications.

The public health implications of SCD are great, particularly when health impact is measured by under-five mortality. With medical science advancements, an increasing proportion of affected children now survive past five years of age but still remain at risk of premature death. When health impact is measured by under-five mortality, SCD contributes the equivalent of 5% of under five deaths on the African continent, more than 9% of such deaths in West Africa, and up to 16% of under-five deaths in individual West African countries (WHO Secretariat, 2006).

In the USA, median survival was estimated in 1994 to be 42 years for men and 48 years for women, whereas comparable figures for Jamaica published in 2001 suggested 53 years for men and 58.5 years for women. In Jamaica, the greatest mortality occurs between 6 and 12 months of age when 10% of patients die despite considerable experience in the diagnosis and therapy of the condition and the absence of malaria. There are, however, no firm data on the survival of patients with sickle cell anaemia on the African continent. In sub-Saharan Africa mortality will be much higher than in Jamaica, and in some areas estimates derived from the age structure of populations attending clinics suggest that half of those with sickle cell anaemia have died by the age of five years, usually from infections including malaria, pneumococcal sepsis, and anaemia itself (WHO Secretariat, 2006).

Sickle cell disease can be detected from birth by a blood test. With health care practitioners and policy makers recognising that management of SCD can be achieved by early diagnosis, newborn screening programmes have rapidly developed internationally (Serjeant, 2000).

1.4. THE CONSEQUENCES OF SICKLE CELL DISEASE

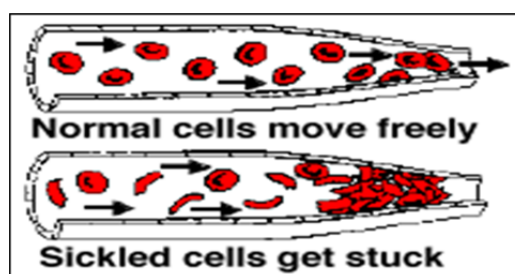
1.4.1. MORTALITY AND LIFE EXPECTANCY

Mortality rates have been published in the USA and the Caribbean, showing a decrease in mortality rates due to SCD in recent years. Sickle related deaths among African American children less than four years of age was shown to have fallen by 42% between 1999 and 2002 (Yanni, Grosse, Yang, *et al.*, 2009). As recent as the 1970s, people with SCD were not expected to survive into adulthood (A., J., A., *et al.*, 2009). These drops in mortality are attributed to timely diagnostic testing, educating patients, and advances in medical care, including neonatal screening, which allows for prophylactic treatment with antibiotics starting at two months of age up until the age of five years, thereby reducing premature deaths due to infection (Steinberg, 2002). Aside from two recorded studies, one from Brent and the other covering sickle centres from France and England that discuss mortality due to SCD, there is little information originating from the UK and the rest of Europe. Despite the increasing number of people in England with SCD or with the trait, England lacks a properly updated survey of how many people in England are dying from SCD or its complications (Lucas, Mason, Mason, *et al.*, 2008).

1.4.2. PATHOPHYSIOLOGY

The clinical features of SCD do not follow a single pattern; some patients have mild symptoms, and some have very severe symptoms. There are two pathological processes – vaso-occlusion and haemolysis. When sickled cells stick together and block blood vessels or get stuck in narrow blood vessels, blocking the flow of blood, this event is called vaso-occlusion. This can happen anywhere in the body, resulting in a vaso-occlusive crisis, or what is often referred to as painful crisis. Vaso-occlusion prevents blood flow into the surrounding tissues and organs, depriving them of oxygen and potentially resulting in localised tissue death and permanent organ damage (Serjeant & Serjeant, 2001; Frenette, 2002). Haemolysis results in anaemia and a functional deficiency of nitric oxide, which results in vascular endothelial damage and can be responsible for complications such as pulmonary hypertension and stroke (Stuart & Nagel, 2004). Some more common complications of the disease are discussed below (National Heart Lung and Blood Institute, 1996).

FIGURE 7: VASO-OCCLUSION



Source: Sickle Cell Society at www.sicklecellsociety.org

Hand-foot syndrome. This disease is also referred to as sickle cell dactylitis. Small blood vessels in hands or feet are blocked usually during a vaso-occlusive (pain) crisis and swelling can result, along with fever. This often results in a radiological change where small bones end up looking ‘moth eaten’ (Babhulkar, Pande & Babhulkar, 1995).

Anaemia. Symptoms of fatigue, paleness, and shortness of breath are present. These are all typical symptoms of a shortage of red blood cells (Anon, 2004).

Painful crisis. This is also referred to as a vaso-occlusive crisis. Pain occurs unpredictably in any body organ or joint wherever there is a block of oxygen flow due to sickled cells sticking together. Some patients have these episodes infrequently and some have them very frequently. The pain can last anywhere from a couple of hours to several weeks. Most hospitalisations for SCD patients are due to painful crisis. Pain is the principal symptom of SCD in both children and adults. Frequent painful episodes are associated with shorter lifespans (Castro, Brambilla, Thorington, *et al.*, 1994).

Sickle cell eye disease. The retina, i.e. the layer of cells at the back of the eye that receives and processes visual images, can deteriorate when it does not get enough nourishment from circulating red blood cells. Damage to the retina can be serious enough to cause blindness (Anon, n.d.). Clinically detectable retinal disease in sickle cell patients is found most commonly between 15 and 30 years of age (Condon & Serjeant, 1980).

Jaundice. Jaundice creates a yellowing of skin and eyes which are manifestations of the rapid breakdown of the red blood cells (Anon, n.d.).

Growth retardation. A shortage of red blood cells also gives rise to physical growth delays and a late onset of puberty (Anon, n.d.).

Infections. The most common cause of death for SCD is infection. In one study, 33-48% of all deaths with SCD as a diagnosis were reported due to infection (Manci, Culberson, Yang, *et al.*, 2003). In general, both children and adults with sickle cell anaemia are more vulnerable to infections and have a harder time fighting them off. A major contributor to this vulnerability is the result of spleen damage from sickled red cells, which prevents the spleen from destroying bacteria in the blood, also putting patients with SCD at higher risk for septicaemia (Magnus, Hambleton, Moosdeen, *et al.*, 1999). Infants and young children in particular are susceptible to bacterial infections that can kill them in as little as 9 hours from the onset of fever. Pneumococcal infections used to be the principal cause of death in children with sickle cell anaemia until physicians began routinely giving penicillin on a preventive basis to those who were diagnosed at birth or in early infancy (National Heart Lung and Blood Institute, 1996).

Stroke. In stroke, the sickled red blood cells stick to the blood vessel walls in the brain and result in narrow or blocked vessels that can lead to strokes, primarily in children (National Heart Lung and Blood Institute, 1996). About 10% of children with SCD suffer from stroke (Steinberg, 1999). Recurrence is common (Anionwu & Atkin, 2001). Another factor is anaemia. Anaemia increases the velocity of cerebral blood flow, and a high velocity of cerebral blood flow is strongly associated with stroke (Adams, McKie, Nichols, *et al.*, 1992).

Acute chest syndrome (ACS). ACS is similar to pneumonia and can be a life-threatening complication caused by infection or trapped sickled cells in the lung. It can show symptoms of chest pain, fever, and an abnormal chest X-ray (National Heart, Lung, and Blood Institute [NHLBI], 2002). After painful crisis, it is the most common reason for hospitalisation, morbidity, and mortality in SCD patients. It is most common in the winter months, and adults usually succumb to complications more frequently than children (Vichinsky, Styles, Colangelo, *et al.*, 1997).

Spleen complications. Acute splenic sequestration is a relatively common and potentially life-threatening event in young children with SCD (Aquino, Norvell & Buchanan, 1997). It is a condition that causes the spleen to fill with blood and become large and tender. This happens when the sickled red blood cells get trapped in the small blood vessels of the spleen, and the blood that normally flows through the spleen gets trapped and causes the spleen to enlarge. With more blood trapped in the spleen, there is less blood to flow through the rest of the body, which can cause the haemoglobin in the rest of the body to drop to a critically low level (Children's Mercy Hospitals and Clinics, 2010). Complications with the spleen are managed through red blood cell transfusions. Chronic hypersplenism (splenomegaly), also referred to as enlarged spleen, is often treated by removing the spleen either fully or partially (splenectomy) (NHLBI, 2002).

Leg ulcers. Leg ulcers occur in about 10-20% of persons with SCD (NIH, 2002). They increase in incidence after the second decade of life and are more commonly seen in men (Bonds, 2005). While the aetiology of leg ulcers is not entirely understood, decreased blood flow to a healed ulcer results in a higher chance of recurrence (Koshy, Entsuhah, Koranda, *et al.*, 1989).

Priapism. Sickling in the blood vessels of the penis can cause painful erections and result in impotence. This occurs in 30-45% of males with SCD. Little is known about the pathological processes and genetic risk factors that contribute to the occurrence of priapism (Elliott, Ashley-Koch, De Castro, *et al.*, 2007; Elliott, Ashley-Koch, De Castro, *et al.*, 2007; Hakim, Hashmat & Macchia, 1994). Treatment includes fluid delivery, surgery, drugs, transfusions, and hydroxyurea (Serjeant & Serjeant, 2001).

Pregnancy. It is only in the last half of the 20th century that women with SCD have survived to reproduce, and even with all the advances in modern medicine, pregnancy in sickle cell patients is regarded as high risk. Morbidity and mortality rates for pregnant women with SCD and their unborn babies are higher than that of the overall population. Maternal complications include an increased risk of all complications due to SCD such as painful crises and all those due to pregnancy such as pre-eclampsia. Risks to the unborn child are also increased with a 19% risk of spontaneous abortion, 21% risk of stillbirth, 32% risk of premature delivery and 42% risk of intrauterine growth restriction (Oteng-Ntim, Chase, Howard, et al., 2008). A recent study at the Department of Obstetrics and Gynaecology and the Department of Haematology at King's College Hospital in London showed that a combined obstetric and haematology clinic allows for a multidisciplinary team approach for pregnant women with SCD, and was found to be beneficial in the management of patients who are at risk for multiple complications (Yu, Stasiowska, Stephens, et al., 2009).

1.5. CLINICAL MANAGEMENT AND HEALTH IMPROVEMENT IN SICKLE CELL DISEASE

A combination of secondary and tertiary prevention programmes are required to reduce morbidity and mortality and improve health outcomes in sickle cell disease (Olney, 1999). There exists an evidence base for secondary and tertiary prevention measures to reduce complications of sickle cell disease, reduce hospital stays and decrease morbidity and mortality (Olney, 1999). Key secondary interventions include early detection, preclinical interventions and education; tertiary interventions include prophylactic transfusions to prevent the recurrence of stroke, folic acid supplementation, analgesics, hydration, and hydroxyurea therapy (Olney, 1999).

1.5.1. SECONDARY PREVENTION MEASURES

EARLY DETECTION

Newborn screening is important as a method of early disease detection and as a trigger for initiating prompt secondary preventive measures such as penicillin prophylaxis and targeted immunisations (Olney, 1999). In the United States, screening programmes are initiated through a programme enabling laws in each state. The lack of a national programme gives rise to programme differences in each state, but it also allows for a quicker response to necessary changes (Therrell & Hannon, 2006). In England, national newborn screening for sickle cell disease was implemented in 2004 (Streetly, Clarke, Downing, et al., 2008). National implementation of newborn screening in England has increased the number of people identified with sickle cell disease, and according to a study by Streetly et al., the national approach has allowed for the development of national materials and training programmes that make much more efficient use of resources, ensures consistency of information and drive further developments in screening methods (Streetly, Latinovic, Hall, et al., 2009). Screening programmes in Sub-Saharan Africa are now being designed after American and European models (Tshilolo, Kafando, Sawadogo, et al., 2008).

PRECLINICAL INTERVENTIONS

PROPHYLACTIC ANTIBIOTICS

Infection is a leading cause of death among patients with SCD with mortality rates due to infection reported as being 20-50%. Administering oral prophylactic penicillin in sickle patients reduces the risk of infection by 84% for patients under the age of three (Booth, Inusa & Obaro, 2010). This inexpensive and extremely effective intervention is a perfect example of how a simple intervention can create marked improvements in morbidity and mortality for SCD patients.

IMMUNISATIONS

It is well-known how important immunisations are for protecting against preventable life threatening diseases. This is particularly true for high risk groups such as those with SCD. That being said, immunisation uptake for those with SCD is not as one would hope or expect. An immunisation audit carried out against UK National Guidance (Salisbury, Ramsay & Noakes, 2006) of sickle cell patients living in Coventry revealed that only 56% of children age 16 and under and 12% of adults had complete immunisations, and that these rates were similar to those found in other UK studies (Howard-Jones, Randall, Bailey-Squire, et al., 2009). Immunisations in some cases have reduced mortality rates for children by 68%, as was the case for children aged three and under after the introduction of the 23-valent pneumococcal polysaccharide vaccine in 1984 and the Hib vaccine in 1988 (Yanni, Grosse, Yang, et al., 2009).

EDUCATION

Education can be a valuable tool in health improvement. Many primary care physicians were found to be unprepared to manage the follow-up care of children with a positive newborn screen, including initial counselling, diagnosis, and subspecialty referral (Kemper, Uren, Moseley, et al., 2006). Knowledge and confidence can be improved with educational interventions (Oyeku, Feldman, Ryan, et al., 2010). Education can also mean several things, including re-educating practice culture. In other words, changing the organizational culture of the hospital or GP practice where health care providers practice. For example, one study examined the influence of nurses' attitudes on the management of SCD and found that the practice culture was for nurses to believe that any patient presenting to hospital and looking for narcotic pain relief was a potential drug addict. The nurses would benefit from further education with a focus on attitudes towards pain and drug addiction in SCD (Pack-Mabien, Labbe, Herbert, et al., 2001b). Not only can health care providers be educated, but the patients, caregivers, and key members of the community can be educated to optimise health outcomes.

1.5.2. TERTIARY PREVENTION MEASURES

Doctors can do a great deal to help patients with SCD, and treatment is constantly being improved. The most common tertiary preventive measures for sickle cell patients are blood transfusions and hydroxyurea therapy.

Blood transfusions. Transfusions correct anaemia by increasing the number of normal red blood cells in the circulation. They can also be used to treat spleen enlargement in children before the condition becomes life-threatening. Regular transfusion therapy can help prevent recurring strokes in children at high risk (SCD Guidelines Adults).

Hydroxyurea. Basic treatment of painful crises relies heavily on painkilling drugs and oral and intravenous fluids to reduce pain and prevent complications. Painful crises account for 90% of all hospital episodes (Anon, 2008). Generally speaking, there is an enormous amount of sympathy for patients with cancer pain, postoperative pain, or pain due to trauma, but little sympathy or understanding for the pain of SCD (Pack-Mabien, Labbe, Herbert, et al., 2001b). Hydroxyurea is the first effective drug treatment for adults with severe sickle cell disease. It was discovered in 1995, when the Multicenter Study of Hydroxyurea in Sickle Cell Anemia (MSH) trial showed that this cancer therapy increases total and foetal haemoglobin and reduces the frequency of painful crises and acute chest syndrome. The patients taking the drug needed fewer blood transfusions (Orkin & Higgs, 2010b). Moreover, there was improved red blood cell hydration among other possible benefits (Dick, 2008). A follow-up study 10 years after the MSH trial showed that hydroxyurea is also effective for primary stroke prevention (Verduzco & Nathan, 2009).

Bone marrow transplantation. Haematopoietic transplantation, otherwise known as bone marrow transplantation (BMT), remains the only possible curative therapy for those with sickle cell disease (Mankad, 2001). BMT can change the phenotype of HbSS patients to HbAA or HbAS. The strongest cases for BMT are made when patients develop a more severe disease such as leukaemia or stroke, but the risks remain too high for this to be a common practice. Along with the extremely high cost of the procedure, risks include but are not limited to rejection of the transplant and neurological complications (Serjeant & Serjeant, 2001). BMT has recently given way to the developing idea of gene therapy.

Gene therapy. Gene therapy introduces a normal gene into a patient's cells and tissues either through viral vector or gene replacement in order to treat a genetic disorder such as SCD. If gene therapy is successful, it avoids the complications that are involved with BMT (Olowoyeye & Okwundu, 2010). So far, gene therapy has been used to correct SCD in mice (Frenette & Atweh, 2007) and it holds future promise as clinical trials are initiated (Romero, Urbinati, Geiger, et al., 2013).

TABLE 1: SCD PREVENTIVE CARE MEASURES

Secondary Prevention	Tertiary Prevention
Early Detection/Newborn Screening	Blood Transfusions
Antibiotic Prophylaxis	Hydroxyurea
Immunisations	Bone Marrow Transplant
Education	Gene Therapy

1.5.3. COMPLEMENTARY AND ALTERNATIVE THERAPIES

As with several other areas in medicine, SCD is not excluded from the exploration of alternative therapies as a compliment to allopathic therapy. In fact, two thirds of the world's population, primarily in developing countries, rely on what is referred to as traditional or herbal medicine as their primary form of health care (Sumner, 2000). Most developing countries have used medicinal plants in the treatment of painful crises associated with SCD either because they cannot afford the high cost of allopathic medicine or simply because herbal medicine is deeply rooted in the culture and traditionalists believe in its efficacy (Okpuzor, Adebessin, Ogbunugafor, et al., 2008). Most of the alternative therapies for SCD promote an anti-sickling effect on red blood cells. Extracts from dried fish (tilapia) and dried prawns (*Astacus red*) have been established to have the ability to stop the formation of HbS haemoglobin, increase foetal haemoglobin and reduce haemolysis in sickle cell blood (Nwaoguikpe & Uwakwe, 2005). Several

plants and seeds indigenous to Africa are used in the treatment and management of SCD, including the more commonly known aloe vera, Nigerian 'bitter kola' and unripened papaya (Okpuzor, Adebesein, Ogbunugafor, et al., 2008).

Some studies in the USA found that complementary and alternative medicine (CAM) can be useful (Post-White, Fitzgerald, Hageness, et al., 2009). One study measured the uptake of CAM for children with SCD, and found that 54% of the sample used CAM therapies for their children, 42% used bioenergetic therapies (prayer, spiritual and energy healing), 28% used lifestyle/mind-body therapies (relaxation techniques, exercise, imagery, and diet), 12% used biochemical therapies (herbal medicines, megavitamins, and folk remedies), and 5% used biomechanical therapy (massage) (Sibinga, Shindell, Casella, et al., 2006). More recently, the antioxidant properties of garlic and green tea have been touted as an important addition to sickle therapy (Ohnishi, Ohnishi & Ogunmola, 2001).

THE ROLE OF ANTIOXIDANTS

Some studies have shown that oxidant stress plays an important part in the pathophysiology of SCD (Hebbel, Eaton, Balasingam, et al., 1982; Rice-Evans, Omorphos & Baysal, 1986). Oxidative stress occurs when the body uses oxygen to make energy. The process creates molecules missing electrons, which are known as free radicals. The lack of an electron causes the molecule to be unstable. Oxidative stress is a normal side effect of body function; usually, nutrients from within the body known as antioxidants are used to control this process.

However, when these free radicals react with other molecules in the body and cause cell damage, this is seen as abnormal and the cause behind many illnesses (Nur, Biemond, Otten, et al., 2011). In the case of SCD, the oxidative stress can contribute to the sickling process. One of the flavour components in garlic (*Allium sativum*) known as AGE has been reported to have a significant antioxidant effect on sickled red blood cells, which can be exacerbated by oxidative stress (Takasu, Uykimpang, Sunga, et al., 2006). Green tea has been found to have a similar effect. The use of AGE and green tea as an antioxidant supplement could be a valuable and a relatively harmless compliment to managing SCD (Ohnishi, Ohnishi & Ogunmola, 2001).

1.6.KEY POINTS FROM CHAPTER 1

1. Sickle cell disease is an important global public health problem with a growing morbidity burden, and there is a need to assess what this burden is in England.
2. History and migration patterns of SCD can help to explain origins and evolution of disease including areas of high prevalence.
3. Presentation of clinical co-morbidities associated with SCD can illustrate that measurement of primary diagnosis for hospital admissions and readmissions of SCD will not capture the true burden unless we also account for the broad spectrum of associated complications.

2. AIMS AND OBJECTIVES

We do not know the burden of disease in England as can be measured through hospital admission, readmission and their related costs. The aims of this research were to:

1. Assess the burden of SCD in England by examining trends and admissions rates due to SCD; and
2. Suggest that some admissions could be preventable through better primary care.

The objective was to accomplish this by conducting one substantial study and five supporting studies:

1. Assessing trends in hospital admissions due to SCD and determining geographic variation using the HES national data set (chapter 3);
2. Characterizing emergency admissions in a PCT with high prevalence of SCD (chapter 3);
3. Measuring hospital readmission due to SCD (chapter 4);
4. Determining hospital costs due to SCD (chapter 5);
5. Viewing patient perspectives on disease management in primary care (chapter 6); and
6. Collecting patient views on improving SCD management in primary care (chapter 6).

Admission trends were surveyed both nationally and locally. We need to identify national trends but then be able to use a local benchmark that can recommend local policies which may inform or grow into national policies. This is the first study of hospital admission trends for SCD using HES data. Because SCD is a disease affecting primarily BME populations, the same HES data set was used to determine whether there was a link between socioeconomic deprivation and SCD hospital readmissions and also whether there was a link between socioeconomic deprivation and SCD hospital mortality. My research also revealed, that unlike in the US, there is very little published in England when it comes to cost and the burden of cost for the NHS from SCD. HES data were once again used to determine the hospital costs associated with SCD.

The research using the HES data drew a very colourful backdrop to the landscape of SCD in England, both nationally and by borough. A focus group of SCD patients and carers and a patient satisfaction questionnaire helped to identify gaps in the delivery of care.

3. NATIONAL TRENDS IN HOSPITAL ADMISSIONS FOR SCD

3.1. BACKGROUND AND AIM

Little data exists in England about the healthcare utilization of SCD patients. Hospital admission data could provide information about the impact of SCD on the NHS. It could highlight disease specific trends and it could be useful to clinicians, commissioners and health planners in prevention as well as disease management.

The aim of this particular study was to investigate trends in the rates of hospital admissions in England for SCD and to determine their geographic variations. The objective was to use data from the Hospital Episode Statistics (HES) database, which provides data on all NHS hospital admissions in England and to use the study period between April 2001 and April 2010. This study provided the foundation for which all other work regarding this thesis was built.

3.2. METHODS AND RESULTS

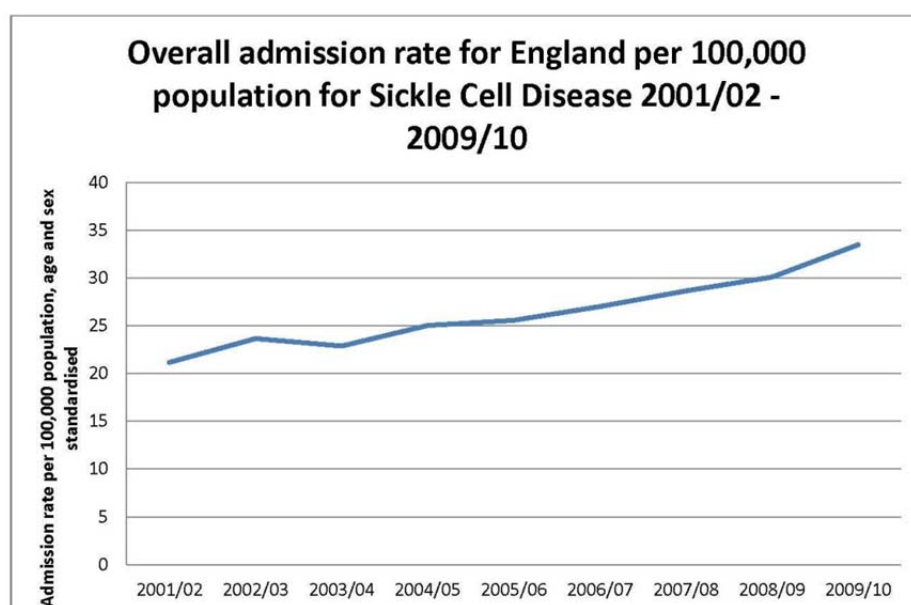
Hospital Episode Statistics (HES) data was used to show trends in hospital admissions for SCD, which I hoped could thereafter partially inform a larger policy recommendation on SCD management in England. As previously stated, sparse data exists in England about the healthcare utilisation of SCD patients. Data on hospital admissions in England for SCD can provide information about the impact of SCD on the National Health Service and would also be of value to clinicians, commissioners, and healthcare planners. Such data could also highlight disease-specific trends (Aljuburi, Laverty, Green, et al., 2012a).

Patients with a primary diagnosis of Sickle Cell Anaemia with Crisis (D57.0) or Sickle Cell Anaemia without Crisis (D57.1) were selected for this study. With Crisis suggests that the patient was admitted for an acute painful episode. Without Crisis suggests that the patient was admitted for something other than an acute painful episode. The overall admission rate per 100,000 population has risen from 21.2 per 100,000 in 2001/02 to 33.5 per 100,000 in 2009/10. Figure 8 and Table 2 show the overall admission rates for SCD as a primary diagnosis from 2001/02 to 2009/10. This shows a rise of over 50% in the admission rate from 2001/02 levels, with a consistent year-on-year rise except for 2002/03 and 2003/04 (Aljuburi, Laverty, Green, et al., 2012a).

TABLE 2: OVERALL ADMISSION RATES FOR SCD

Year									
Age (years)	2001/ 02	2002/ 03	2003/ 04	2004/ 05	2005/ 06	2006/ 07	2007/ 08	2008/ 09	2009/ 10
Men overall	20.7	24.1	24.1	25.4	25.9	28.5	31.3	32.7	35.1
<1	8.8	6.7	7.1	4.6	12.6	16.1	16.7	15.2	14.7
1 to 9	18.6	21.2	21.5	24.9	22.7	27.8	37.1	41.2	45.5
10 to 19	24.9	30.6	30.0	35.8	37.8	39.2	44.6	49.9	55.6
20 to 29	50.0	49.7	54.8	57.1	57.4	70.5	65.0	64.7	68.6
30 to 39	44.3	54.3	47.7	44.2	39.3	37.8	42.8	41.9	46.8
40 to 49	7.6	13.7	16.0	17.3	23.5	24.1	28.1	29.0	26.8
Over 50	1.6	1.4	1.6	2.3	2.3	2.1	2.7	2.8	3.2
Women overall	21.6	23.3	21.7	24.6	25.2	25.6	26.1	27.6	31.9
<1	7.3	7.0	7.9	5.2	12.8	11.9	10.9	13.2	10.8
1 to 9	12.6	18.5	17.7	20.3	23.0	23.5	26.1	36.5	42.2
10 to 19	29.1	36.3	31.9	40.3	41.8	47.3	48.5	51.2	56.4
20 to 29	32.4	35.4	38.0	44.6	51.9	50.9	52.1	52.4	57.9
30 to 39	52.4	50.1	41.2	41.7	41.4	36.6	35.7	37.0	45.3
40 to 49	25.4	24.5	25.9	28.0	22.1	25.7	26.0	24.7	29.2
Over 50	3.4	4.0	3.8	4.9	4.5	4.2	3.9	4.0	5.1
Total	21.2	23.7	22.9	25.0	25.6	27.0	28.7	30.1	33.5
(Aljuburi, Lavery, Green, <i>et al.</i> , 2012a)									

FIGURE 8: OVERALL ADMISSION RATES FOR SCD IN ENGLAND

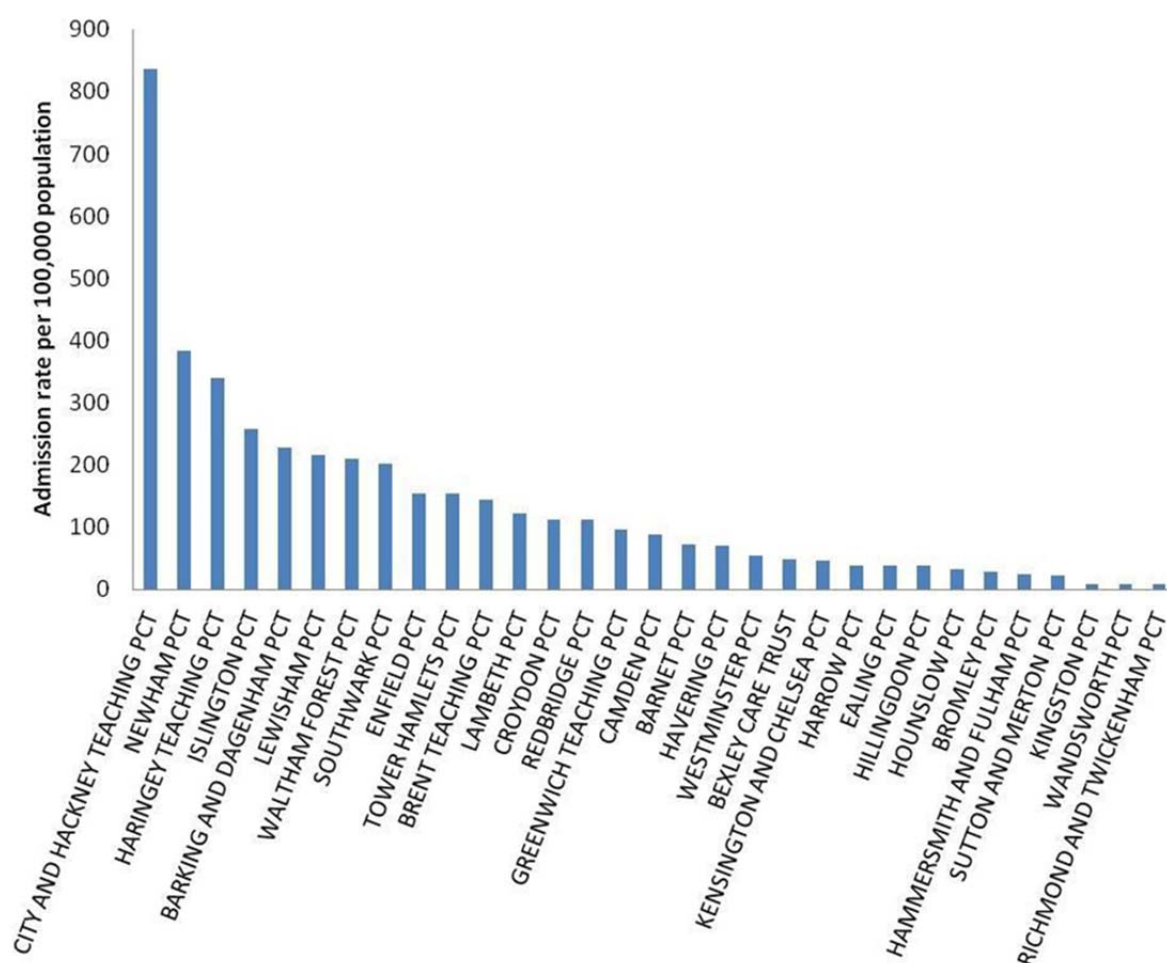


(Aljuburi, Laverty, Green, *et al.*, 2012a)

A rise in SCD admission rates can be seen in almost every age and sex group (Table 2). The largest rise in admission rates was seen amongst men aged between 40 and 49 years old, where admission rates per 100,000 population increased from 7.6 to 26.8 over this time period (Table 2) (Aljuburi, Laverty, Green, *et al.*, 2012a).

London accounts for almost three quarters of all SCD admissions in England (74.9% of all admissions nationally) (Aljuburi, Laverty, Green, *et al.*, 2012b; Green, Aljuburi, Majeed, *et al.*, 2012a). The highest admission rates can be seen in the City and Hackney Teaching PCT at 837.4 per 100,000 population (Figure 9).

FIGURE 9: ADMISSIONS PER 100,000 POPULATION



Source: (Aljuburi, Laverty, Green, et al., 2012b)

Table 3 shows the most common secondary and tertiary diagnoses for those admitted with SCD as the primary diagnosis. The most common additional diagnostic codes were for respiratory conditions including lower respiratory tract infection, lobar pneumonia, and asthma, followed by vaso-occlusive conditions such as occlusion and stenosis of the middle cerebral artery, Moyamoya disease, hemiplegia, and sequelae of stroke (not specified as haemorrhage or infarction).

TABLE 3: TOP SECONDARY AND TERTIARY DIAGNOSES

Top 10 Secondary Diagnoses	Number (% of 2009/10 total)
Acute Lower Respiratory Tract Infection	310 (1.9)
Chemotherapy	223 (1.3)
Asthma	187 (1.1)
Urinary Tract Infection	133 (0.8)
Lobar pneumonia	128 (0.8)
Naevi and melanomas	115 (0.7)
Occlusion and stenosis of middle cerebral artery	104 (0.6)
Moyamoya disease	96 (0.6)
Essential hypertension	91 (0.5)
Chest Pain	88 (0.5)
Top 10 Tertiary Diagnoses	Number (% of 2009/10 total)
Asthma	89 (0.5)
Sequelae of stroke, not specified as haemorrhage or infarction	64 (0.4)
Priapism	59 (0.4)
Dorsalgia	57 (0.3)
Naevi and melanomas	54 (0.3)
Acute Lower Respiratory Tract Infection	54 (0.3)
Hemiplegia	53 (0.3)
Essential hypertension	47 (0.3)
Vitamin D deficiency	42 (0.3)
Hyposplenism	41 (0.2)

Source: (Aljuburi, Lavery, Green, et al., 2012a)

Table 4 shows the breakdown of crisis and non-crisis admissions in 2009/10. Slightly over half of all admissions (50.3%) were for a sickle cell crisis, and those in the age group 20-29 had a higher proportion of crisis admissions than other groups (62.8% and 62.9% for men and women respectively). The lowest proportion of crisis admissions was in women aged over 50 (30.4%).

TABLE 4: BREAKDOWN OF CRISIS AND NON-CRISIS ADMISSIONS

Age (years)	Sickle Cell Crisis (%)	Sickle Cell non Crisis (%)	Total
Men (overall)	4509 (53.1)	3979 (46.9)	8488
<1	34 (68.0)	16 (32.0)	50
1 to 9	478 (38.1)	777 (61.9)	1255
10 to 19	855 (47.4)	948 (52.6)	1803
20 to 29	1554 (62.8)	919 (37.2)	2473
30 to 39	983 (60.7)	637 (39.3)	1620
40 to 49	481 (47.2)	539 (52.8)	1020
50+	124 (46.4)	143 (53.6)	267
Women (overall)	8572 (51.8)	7986 (48.2)	16558
<1	17 (48.6)	18 (51.4)	35
1 to 9	479 (43.0)	634 (57.0)	1113
10 to 19	891 (51.3)	847 (48.7)	1738
20 to 29	1260 (62.9)	744 (37.1)	2004
30 to 39	787 (50.2)	780 (49.8)	1567
40 to 49	482 (42.7)	648 (57.3)	1130
50+	147 (30.4)	336 (69.6)	483
Total Number Overall	4063 (50.3)	4007 (49.7)	8070

Source: (Aljuburi, Lavery, Green, et al., 2012a)

57.9% of all patients were discharged within 24 hours of being admitted to hospital, while only 15.4% of admissions stayed for longer than one week. The largest proportion of patients discharged on the same day was for those aged over 50 years old, at 72.1%.

3.3. DISCUSSION

This is the first national study on trends for hospital admission rates for SCD using HES data. The data has four main findings:

1. The number of admissions due to SCD has increased substantially over the study period (2001-2010);
2. The majority of admissions are in London;
3. Most admissions were discharged within 24 hours; and
4. There has been an increase in the number of admissions of middle-aged men.

The over 50% in SCD admissions, is probably due to a number of factors, the most important of which is likely to be an increased prevalence of SCD in England through domestic growth of the current at-risk population and population migration from countries where SCD is common (WHO Secretariat, 2006).

A previous study on the national implementation of newborn screening in England showed an increased number of children identified with SCD in some areas. The authors of this study concluded that past under-ascertainment of cases may have led to under-estimation of the

needs of people with SCD. Under-ascertainment of SCD cases may also have contributed to increased infant mortality in urban areas because babies were at risk of dying without a diagnosis of SCD being made and the initiation of appropriate management (Streetly, Latinovic, Hall, et al., 2009).

Aside from the reasons already mentioned in Chapter 1 regarding possible contributors to the increase of SCD prevalence in England, the increases in admissions for SCD may also be due to a drop in the threshold for admission of patients with SCD, through greater recognition of the importance of SCD crisis, or through patients being discharged earlier and being readmitted more frequently for the same episode of illness (Patel, Bell, Molokhia, et al., 2007).

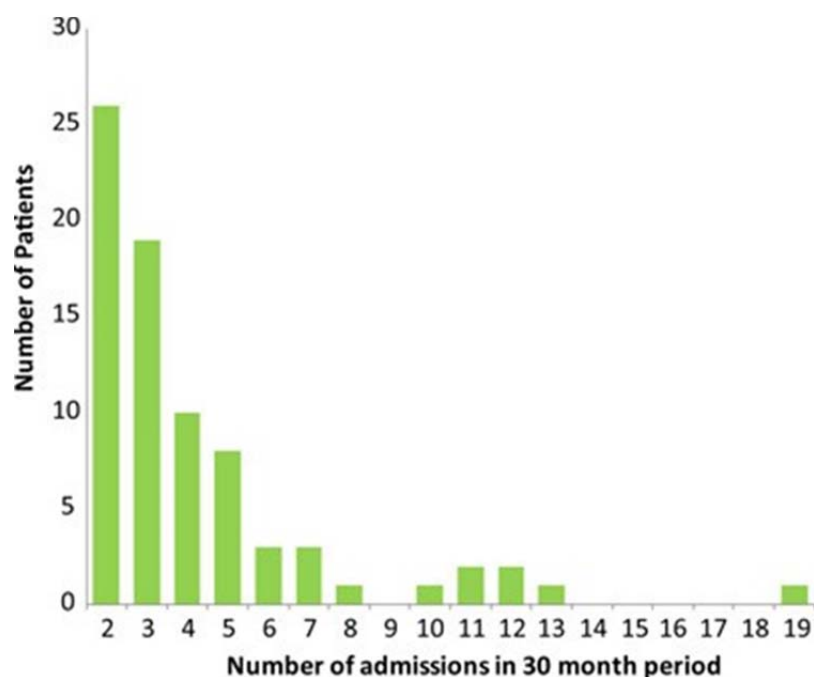
Studies from the United States have also shown readmission as an important contributor to rising rates of admissions in patients with SCD (Ballas & Lusardi, 2005a). This may also raise issues of how well hospitals are managing these patients (Maxwell, Streetly & Bevan, 1999a), and emphasises the importance of good discharge planning for these patients, which may minimise the risk of hospital readmission. Good discharge planning is a process that ensures strong and quality links between hospitals, primary and community care services, and patients and their carers. Effective discharge planning for those with SCD can support the continuity of health care for a patient as it links treatment received in the hospital to post-discharge care received in the community. One future study could be an audit of hospital discharge forms to show the number of hospital discharge forms being sent to GPs and the level of follow up from the GP.

The data from this study shows that SCD is a growing concern and provides a basis for further study. This study highlights that the majority of patients admitted with SCD were discharged within 24 hours. Further study could focus particularly on these admissions which may give a more precise indication of which ones could actually be addressed as preventable. Currently, our assumption is that the 24 hour and fewer admissions could potentially be prevented through better ambulatory and community care of patients, including better care in emergency departments.

3.4. BRENT STUDY

Our national finding that the majority of SCD admissions are discharged within 24 hours was also found in another study that we conducted in the London Borough of Brent which showed that 36% of SCD patient admissions in Brent resulted in a length of stay of less than 2 days and that 74% of total bed days are associated with patients with multiple admissions (Green, Aljuburi, Majeed, et al., 2012b).

FIGURE 10: EMERGENCY ADMISSIONS BETWEEN JANUARY 2008 AND JULY 2010 FOR SICKLE CELL CRISIS IN NHS BRENT BY THE FREQUENCY OF SPELLS FOR INDIVIDUAL PATIENTS ADMITTED (TOTAL NUMBER OF PATIENTS 77)



Source: (Green, Aljuburi, Majeed, et al., 2012a)

The objective of the local study was to characterise emergency admissions for patients with sickle cell crisis in NHS Brent. Brent was selected because it is considered one of the high prevalence areas for SCD (Hickman, Modell, Greengross, et al., 1999; Anon, 2005). This was an observational study and looked at emergency departments attended by residents of the London borough of Brent with the participants being patients with sickle cell disease registered with a general practitioner (GP) in the borough of Brent. We analysed admissions between January 2008 and July 2010 that included length of stay (average and <2 days versus ≥ 2 days) by age group and registered GP practice. In addition to finding that 36% of sickle cell disease admission spells resulted in a length of stay of less than two days, we also observed that 74% of total bed days are associated with patients with more than one admission during the period of analysis, i.e. multiple admissions (Green, Aljuburi, Majeed, et al., 2012b).

Both the national and local study suggested that a short length of stay and multiple admissions could suggest potentially avoidable hospital admissions. A 2013 Nuffield Trust observational study of hospital admission data showed that potentially avoidable hospital admissions make up one in every five emergency admissions in England (Blunt, n.d.) Potentially avoidable hospital admissions could benefit from better preventive and primary care interventions.

3.5. KEY POINTS FROM CHAPTER 3

We conducted the first national study on trends for hospital admission rates for SCD using HES data in England. The data provided four main findings:

1. The number of admissions due to SCD has increased substantially over the study period (2001-2010);
2. The majority of admissions are in London;
3. Most admissions were discharged within 24 hours; and
4. There has been an increase in the number of admissions of middle-aged men.

The Brent Observational study showed:

1. Thirty-six percent of sickle cell disease admission spells resulted in a length of stay of less than two days; and
2. Seventy-four percent of total bed days are associated with patients with more than one admission during the period of analysis, i.e. multiple admissions.

Both studies suggest that some of these admissions may be potentially avoidable hospital admissions and could benefit from better preventive or primary care.

4. EXAMINING THE HES DATA FOR THE INFLUENCE OF SOCIOECONOMIC CHARACTERISTICS ON THE RISK OF SCD HOSPITAL READMISSION

4.1. BACKGROUND AND AIM

The first analysis of the HES data showed that admission rates for SCD in England are increasing; wide variations exist in admission rates amongst primary care trusts, especially in London (Aljuburi, Laverty, Green, et al., 2012a). I noted in the previous chapter that there has been very little published about trends in SCD hospital admissions in England, but there has also been limited research which examined the influence of socioeconomic characteristics on the risk of SCD admission (Elandera, Beach & Haywood Jr, 2011). The aim of our next analysis was to investigate trends in the rates of emergency readmissions in England for patients with SCD, to determine inpatient mortality, and to assess whether there is an association between deprivation and comorbidity with risk of readmission and inpatient mortality (Aljuburi, Laverty, Green, et al., 2013a).

4.2. METHODS AND RESULTS

Data from a six-year period taken from the national HES database (April 2005 to March 2011) was analysed. The financial year 2005/06 was taken as the index year for this analysis, and all patients admitted with a primary or a secondary diagnosis of SCD in this year were identified and classed as SCD patients in our cohort. Secondary diagnoses are only meant to be coded if they are related to the reason for admission. For patients who were admitted more than once in the index year, their first admission was used as the index admission (Aljuburi, Laverty, Green, et al., 2013b).

Pseudonimised patient identifiers were used to identify subsequent hospital admissions for the patients identified in the index year. Outcome variables used were emergency hospital admissions, in-hospital mortality, and emergency readmissions to hospital. Predictor variables used in this analysis were age group, sex, national deprivation group (based on Lower Super Output Area Index of Multiple Deprivation ranks for the whole of England) (Department for Communities and Local Government, 2011) from 1 (most deprived) to 4 (least deprived), whether the index diagnosis was a primary or secondary diagnosis for SCD, whether the index diagnosis was for an SCD crisis or not (among those with a primary diagnosis of SCD), and the Charlson comorbidity index score. The Charlson score classifies patients into groups depending on the number and severity of their comorbidities on admission, with higher Charlson scores reflecting patients having a higher risk of mortality within ten years (Charlson, Pompei, Ales, et al., 1987). For multivariate analysis, a combination diagnosis variable was created to compare those admitted with SCD as a secondary diagnosis, those admitted with a primary diagnosis of non- crisis, and those admitted with a primary diagnosis with crisis (Aljuburi, Laverty, Green, et al., 2013b).

Descriptive statistics for the number of admissions and deaths in the index year were first produced and then summarised for study years individually. Cox proportional hazards models were used to examine the association between patient demographic variables and the likelihood of emergency admission over the time period as well as in-hospital mortality. For patients admitted more than once during the study period, only the first readmission was used. All of the

variables mentioned previously were used in these models. All statistical analyses were performed using the programme STATA® version 12. (Aljuburi, Laverty, Green, et al., 2013b)

The characteristics of the patients admitted for SCD in 2005/06 (the index year) can be seen in Table 5 (n=7679). The majority (58.1%) live in the most deprived quartile of Lower Super Output Areas nationally, with only 9.5% of admissions coming from patients living in the least deprived quartile. The majority (65%) involved SCD as a secondary diagnosis, and among those admitted with SCD as a primary diagnosis, the majority were for an SCD crisis (73%). Overall, the percentage of deaths in the index year was low at 0.4%. Deaths were higher among those with SCD as a secondary diagnosis compared to primary diagnosis (0.6% vs. 0.1%), and among those with a Charlson comorbidity score of two or more than a Charlson score of zero (2.7% vs. 0.1%) (Aljuburi, Laverty, Green, et al., 2013a).

Figure 1 shows the unadjusted cumulative time to readmission curves by national deprivation group among those with a primary admission for an SCD crisis (n = 1,964). Patients in the least deprived group were less likely to be readmitted over the whole time period compared to all other levels of deprivation. For example, 39.6% of the patients in the most deprived group were admitted at least once within two years, compared to 20.2% for the least deprived group (data not shown).

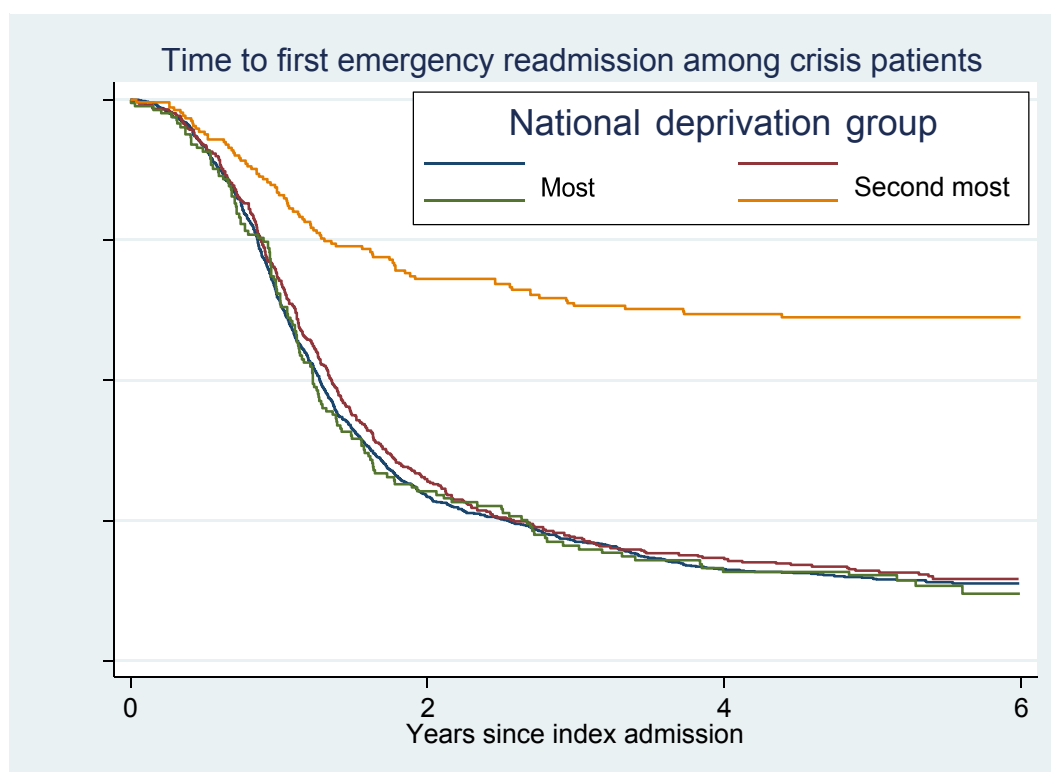
TABLE 5: CHARACTERISTICS OF PATIENTS ADMITTED AND INPATIENT MORTALITY WITH A PRIMARY OR SECONDARY DIAGNOSIS OF SICKLE CELL ANAEMIA WITH CRISIS OR SICKLE CELL ANAEMIA WITHOUT CRISIS IN THE INDEX YEAR (2005/06)

		Number	%	Deaths	%
Age group	Less than 1	142	1.8	0	0.0
	1 to 9	931	12.1	1	0.1
	10 to 19	1,122	14.6	1	0.1
	20 to 29	1,736	22.6	6	0.3
	30 to 39	1,739	22.6	6	0.3
	40 to 49	1,073	14.0	9	0.8
	50 plus	936	12.2	31	3.3
Sex	Male	3,049	39.7	32	1.0
	Female	4,630	60.3	22	0.5
National deprivation group	1 (Most deprived)	4,454	58.0	30	0.7
	2 (Second most deprived)	1,791	23.3	16	0.9
	3 (Second least deprived)	708	9.2	2	0.3
	4 (Least Deprived)	726	9.5	6	0.8
Initial diagnosis	Primary	2,681	34.9	9	0.3
	Secondary	4,998	65.1	45	0.9
Index admission for crisis*	Yes	1,955	25.5	6	0.3
	No	726	9.5	3	0.4
Charlson comorbidity index	0	6,265	81.6	10	0.2
	1	819	10.7	11	1.3
	>=2	595	7.7	33	5.5
Total		7679		54	0.7

*This includes only patients who had an initial primary diagnosis of SCD.

Source: (Aljuburi, Laverty, Green, et al., 2013a)

FIGURE 11: TIME TO FIRST EMERGENCY RE-ADMISSION ACROSS NATIONAL DEPRIVATION GROUPS, AMONG PATIENTS WITH AN INDEX ADMISSION FOR AN SCD CRISIS



Source: (Aljuburi, Laverty, Green, et al., 2013a)

Table 6 shows the annual percentages of readmissions (n=7679) by the demographic variables. Over the whole period, those patients aged 10-19 years had the highest risk of readmission (65.5%), as did those with SCD as a primary diagnosis (74%) and those with an index admission for an SCD crisis (77.5%). Those in the national deprivation group 1 (most deprived) were more likely to have a readmission over the whole period than those in national deprivation group 4, the least deprived group (54.2% vs. 28.0%) (Aljuburi, Laverty, Green, et al., 2013a).

TABLE 6: PERCENTAGE OF PATIENTS WITH EMERGENCY READMISSION AFTER ADMISSION IN INDEX YEAR (2005/6) BY PATIENT CHARACTERISTICS, (N=7679)

		2006/07	2007/08	2008/09	2009/10	2010/11	Any year
Age group	Less than 1	35.9	26.9	28.3	24.1	21.4	55.9
	1 to 9	53.0	46.4	45.9	44.9	43.5	65.2
	10 to 19	33.8	27.7	27.6	26.7	25.5	65.5
	20 to 29	12.4	10.7	12.0	11.3	12.8	49.3
	30 to 39	11.9	8.5	8.7	7.3	7.0	42.8
	40 to 49	30.7	27.7	26.1	27.8	25.5	45.3
	50 plus	42.8	41.9	43.9	42.6	45.8	45.3
Sex	Male	31.1	27.2	26.3	25.8	25.4	54.6
	Female	24.4	21.1	22.1	21.3	21.4	49.0
National deprivation group	1 Most deprived	28.9	25.1	25.2	24.5	24.9	54.2
	2 Second most deprived	28.3	25.3	24.7	24.2	24.2	54.0
	3 Second least deprived	25.0	20.3	23.2	22.5	18.2	49.0
	4 Least deprived	14.6	12.7	13.5	12.3	13.3	28.0
Initial diagnosis	Primary	24.3	22.1	22.8	21.9	22.2	73.8
	Secondary	32.0	26.0	25.6	25.2	24.4	38.9
Index admission for crisis	Yes	49.1	44.7	46.0	43.6	44.4	77.5
	No	33.0	29.9	30.9	31.1	31.4	63.7
Charlson comorbidity index	0	26.8	23.7	24.3	23.8	23.7	50.7
	1	26.5	23.7	21.6	21.0	21.9	53.5
	>=2	30.8	21.3	21.3	17.8	17.0	52.9
N admissions		2,076	1,805	1,826	1,772	1,767	3,932

Source: (Aljuburi, Laverty, Green, et al., 2012a)

Table 7 shows the annual in-hospital mortality by the demographic variables. Patients in the national deprivation group 1 were more likely to die in hospital over the whole period than those in national deprivation group 4 (2.7% vs 1.4%). Those with an index admission not for an SCD crisis were more likely to die than those with an index admission for a crisis (2.9% vs 1.9%), and those with a Charlson score of two or more were most likely to die in hospital over the whole period (12.6% vs 1.1 for those with a score of zero) (Aljuburi, Laverty, Green, et al., 2013a).

TABLE 7: PERCENTAGE OF PATIENTS WITH INPATIENT DEATH AFTER ADMISSION IN THE INDEX YEAR (2005/6) BY PATIENT CHARACTERISTICS, (N=7679)

		2006/07	2007/08	2008/09	2009/10	2010/11	Any year
Age group	Less than 1	0	0	0	0	0	0
	1 to 9	0	0.1	0.1	0.1	0.1	0.4
	10 to 19	0.2	0.2	0	0.1	0.2	0.6
	20 to 29	0.2	0.2	0.2	0.1	0.2	0.9
	30 to 39	0.2	0.3	0.3	0.3	0.2	1.4
	40 to 49	0.7	0.6	0.6	0.7	0.6	3
	50 plus	3.1	2.8	2.5	1.6	1.3	10.8
Sex	Male	0.8	0.6	0.5	0.5	0.5	2.9
	Female	0.5	0.5	0.5	0.3	0.3	2.1
National deprivation group	1 Most deprived	0.8	0.7	0.5	0.3	0.4	2.7
	2 Second most deprived	0.4	0.4	0.6	0.6	0.4	2.4
	3 Second least deprived	0.3	0.3	0.1	0.7	0.1	1.6
	4 Least deprived	0.1	0.3	0.4	0.3	0.3	1.4
Initial diagnosis	Primary	0.3	0.4	0.6	0.5	0.4	2.1
	Secondary	0.7	0.6	0.5	0.3	0.4	2.5
Index admission for crisis	Yes	0.3	0.5	0.3	0.4	0.4	1.9
	No	0.4	0.1	1.2	0.8	0.3	2.9
Charlson comorbidity index	0	0.2	0.2	0.2	0.3	0.2	1.1
	1	1.2	1.4	1.1	0.6	0.3	4.5
	>=2	3.5	3	3.4	1.3	2.1	12.6
N deaths		46	42	38	31	27	184

Source: (Aljuburi, Laverty, Green, et al., 2012a)

Results from the Cox proportional hazards models are given in Table 8. These show that, over the time period, those in national deprivation group 1 (most deprived) were more likely to be readmitted than those in national deprivation group 4 (least deprived) (hazard ratio (HR) 2.97, 95% confidence interval (CI) 2.57-3.43). Those with a Charlson comorbidity score of two or more were more likely to be readmitted than those with a score of zero (HR 1.71, 95% CI 1.51-1.94). The 30-39 year old age group was selected as the reference group because it was the mid-point of the age range of the patients with SCD

Those with a primary diagnosis of an SCD crisis were more likely to be readmitted than those with a secondary diagnosis of SCD (SCD as a comorbidity) (2.76, 95% CI 2.49-3.05), as were those with a primary diagnosis of a crisis (HR 2.99, 95% CI 2.78-3.21). For in-hospital mortality, those in national deprivation group 1 were more likely to die in hospital than those in national deprivation group 4 over the time period (Hazard Ratio (HR) 2.34, 95% CI 1.41-3.90), as were those in national deprivation group 3 (HR 2.00, 95% CI 1.16-3.46). Those with a primary diagnosis of SCD non-crisis were more likely to die in hospital than those with a secondary diagnosis of SCD (3.58, 2.25-5.68) as were those with a primary diagnosis of crisis (HR 3.04, 95% CI 2.11-4.39) (Aljuburi, Laverty, Green, et al., 2013a).

TABLE 8: MULTIVARIATE MODELS FOR RISK OF EMERGENCY RE-ADMISSION AND INPATIENT MORTALITY IN THE SUBSEQUENT FIVE YEARS (N=7679)

		Hazard ratio for admission	Confidence Interval	Hazard ratio for mortality	Confidence Interval
Age group	Less than 1	1.47*	1.17; 1.85	Missing	-
	1 to 9	1.42*	1.28; 1.59	0.33*	0.13; 0.85
	10 to 19	1.37*	1.24; 1.52	0.36*	0.16; 0.82
	20 to 29	1.1	1.00; 1.22	0.78	0.45; 1.35
	30 to 39	1	-	1	-
	40 to 49	1.1	0.98; 1.23	1.96*	1.22; 3.14
	50 plus	1.19*	1.05; 1.35	4.77*	3.12; 7.29
Sex	Male	1	-	1	-
	Female	1.07	1.00; 1.14	0.71*	0.55; 0.92
National deprivation group	1 Most deprived	1	-	1	-
	2 Second most deprived	0.96	0.89; 1.03	0.84	0.62; 1.14
	3 Second least deprived	0.84*	0.75; 0.94	0.49*	0.28; 0.86
	4 Least deprived	0.34*	0.29; 0.39	0.40*	0.24; 0.68
Charlson comorbidity index	0	1	-	1	-
	1	1.40*	1.26; 1.55	3.51*	2.40; 5.11
	>=2	1.72*	1.51; 1.94	8.20*	5.84; 11.51
Initial diagnosis	Secondary	1	-	1	-
	Primary - non-crisis	2.76*	2.49; 3.05	3.58*	2.25; 5.68
	Primary – crisis	2.99*	2.78; 3.21	3.04*	2.11; 4.39

* Statistically significant difference from reference category at p<0.05

4.3. DISCUSSION

4.3.1. HIGHER FREQUENCY OF SCD READMISSIONS SEEN IN AREAS OF INCREASED SOCIOECONOMIC DEPRIVATION

There is a higher frequency of SCD readmissions in areas of increased socioeconomic deprivation. Some of the readmissions we observed may be due to new hospital episodes, but we still observed lower rates in less deprived places. The majority of readmissions (over 58%) and highest inpatient mortality occurred in patients living in the most socioeconomically deprived areas; the highest risk of admission was seen in patients aged 10-19 years and the highest risk of death was in people aged >50. Death rates were higher in those with a Charlson score of 2 or more, suggesting death rates are higher in patients with increased comorbidities (who are also likely to be older) (Aljuburi, Lavery, Green, et al., 2013a). This is consistent with studies that have shown that patients who live in high poverty neighbourhoods are 24% more likely to be readmitted than others (Hu, Gonsahn & Nerenz, 2014).

4.3.2. RELATIONSHIP TO CURRENT LITERATURE

Much remains unclear about the influence of socioeconomic status on SCD. However, the wider socioeconomic status (SES) and chronic disease literature indicates a number of ways in which those coming from the most deprived sector may experience increased hospital readmissions, excess in-patient mortality, and consequent inequalities in health care (Saxena, Eliahoo & Majeed, 2002).

According to the 2001 United Kingdom census, 4.6 million or 7.9% of the total population of the UK belonged to ethnic minority groups. Indians were the largest minority group (1.8%), followed by Pakistanis (1.3%), those of mixed ethnic backgrounds (1.2%), Black Caribbeans (1.0%), Black Africans (0.8%) and people from Bangladesh (0.5%). The issue of health inequalities towards black, ethnic and minority (BME) populations has been recognised for some time, and in recent years, there has been an effort towards addressing such inequalities (Mackenbach, 2006).

4.3.3. MINORITY DISEASE AND CHRONIC DISEASE

Sickle cell disorders in England occur almost exclusively amongst BME populations, and these disorders have received relatively low priority from health policy makers because they are seen as minority diseases (Anionwu & Atkin, 2001). However, not only is SCD a minority disease, it is a chronic disease. Those suffering from chronic diseases such as diabetes, asthma, and cardiovascular disease also suffer from inequalities in health care treatment such as ineffective therapy and poor disease control (Wagner, 1997, 1998). Therefore, those with SCD as a chronic disease have the added stress of minority and socioeconomic disadvantage. Income levels for South Asians and African-Caribbean people working in the UK are lower than the UK average and their unemployment rates are about double those of white populations, which means that people with SCD are statistically more likely to be battling a combination of chronic disease, a disease that affects primarily minorities, and economic disadvantage (Anionwu & Atkin, 2001).

The Marmot Review stated that socioeconomic inequalities affect health outcomes and confirmed that those who are worse off financially also do worse on health outcomes. Socioeconomic influencers are the factors that have the greatest impact on health, i.e. 40% of all influences, as compared to health behaviours (30%) and the physical environment (10%) (Marmot & Bell, 2012). The 2013 Nuffield Trust observational study of hospital admission data showed that the level of deprivation in an area is also strongly linked to rates of potentially avoidable hospital admissions (Blunt, n.d.). I wanted to show a similar link between the influence of socioeconomic characteristics and the risk of SCD hospital admissions.

Studies have found that, in general, the causes of hospital readmission for patients with SCD include premature discharge, withdrawal syndrome, and the occurrence of new acute (sickle cell crisis) episodes (Ballas & Lusardi, 2005b). Another study found that low income is associated with greater emergency department use for SCD pain both in countries with and without universal health care coverage (Glassberg, Wang, Cohen, et al., 2012). Socioeconomic status may also influence the way health care providers manage chronic disease. A study by Struthers et al. (2000) showed an increased rate of hospital readmissions in those with lower socioeconomic status for congestive heart failure, independent of disease severity (Struthers, Anderson, Donnan, et al., 2000). The researchers suggest that this may be due to a number of reasons which include primary care providers in deprived areas having less time to invest in adequate disease management. Primary care providers working in deprived areas may also perceive their patients as having less capacity to understand and manage their own condition (Haque & Telfair, 2000), and patients living in deprived areas may perceive that their

community medical resources are insufficient to manage them and 'push' for hospital admission (Blair, Lloyd-Williams & Mair, 2002; Struthers, Anderson, Donnan, et al., 2000).

Another factor that may contribute to increased SCD readmissions, particularly amongst the most deprived patients, is patient self-discharge. A study in England found that 14% of the sample of SCD patients reported having self-discharged from hospital. The most common reasons given by the SCD patients for self-discharging were being 'tired of waiting for pain relief', potential conflicts related to suspected analgesics abuse, and because they just simply wanted to go home (Elander, Lusher, Bevan, et al., 2004). Ballas and Lusardi (2005) also suggest that the decision by the patient to leave the hospital may be due to family pressure, childcare, or fear of job loss if the patient is employed (Ballas & Lusardi, 2005b). A similar study published in the US found a much higher rate with 46.5% of their sample of SCD patients discharging themselves from hospital (Haywood Jr, Lanzkron & Ratanawongsa, 2011).

In this particular study, we were also unable to evaluate the impact of severity of SCD disease which may contribute to the risk of readmission and inpatient mortality, as information on this is not included in the HES. We also know that, in the UK, patients with SCD are a mobile population, especially in the young adult group who may be students temporarily resident in low prevalence/least deprived areas. Their readmissions may therefore be elsewhere in the UK (i.e. Wales, Scotland or Northern Ireland) or beyond and could have had an impact on reported readmissions. A future study may focus on what the secondary diagnoses were that led to patient deaths and if they are linked to primary diagnosis of SCD or unrelated. We also did not examine post-discharge factors that could influence the risk of admission. These factors include the characteristics of the home environment and the quality and continuity of health care of patients after they have been discharged from hospital, including use of community/primary care facilities. Incorporating the impact of post-discharge factors on readmission (Arbaje, Wolff, Yu, et al., 2008) may have provided further explanation into the pattern of SCD readmissions. (Aljuburi, Lavery, Green, et al., 2013a).

4.4. KEY POINTS FROM CHAPTER 4

1. The groups at highest risk of readmission were patients living in the most socioeconomically deprived areas (54.2%).
2. Inpatient mortality amongst readmissions was highest in patients living in the most deprived areas.
3. SCD patients from the most socioeconomically deprived areas and with comorbidities are at highest risk of both SCD readmission as well as in-hospital mortality, suggesting that there are inequalities in health care access and health outcomes amongst people with SCD.

5. HOSPITAL ADMISSION COSTS

5.1. BACKGROUND

Previous chapters showed trends in admissions and socioeconomic impact on admissions due to SCD in England. Both trends demonstrate a burden on the health care system from a disease perspective, but critical to any analysis of the burden of disease is the cost associated with such admissions. Globally, we are seeing a growing burden when it comes to SCD-associated costs. One rationale is that this is partially due to advances in research and science that have increased the life expectancy of patients living with SCD (Weatherall, 2010) and additionally because SCD patients receive care in acute care facilities, including emergency departments (Nietert, Silverstein & Abboud, 2002) under circumstances that could potentially be curbed through preventive care or treated at the primary care level. SCD care in the United States amounts to over \$1.1 billion in healthcare expenditures annually (Kauf, Coates, Huazhi, et al., 2009); paediatric medical costs from SCD each year are estimated to be at least \$335 million (Kauf, Coates, Huazhi, et al., 2009). Medical expenditures for children with SCD are up to eleven times that of a child without SCD (Amendah, Mvundura, Kavanagh, et al., 2010a). In my research on England, I noticed that we had very little published on what the cost is for a typical hospital visit for a patient with SCD. This became for me an important part of the full picture along with the 2013 Nuffield Trust observational study showing that hospital admissions for potentially avoidable conditions have been growing over the last ten years and account for an estimated £1.4 billion a year (Blunt, n.d.).

5.2. AIM AND OBJECTIVE

The aim was to assess the costs associated with hospital admissions in England for SCD. By doing so, we can more accurately link the impact of SCD on the NHS.

The objective is that expenditures associated with SCD can also be used to raise awareness of the severity of this condition and serve as a basis for exploring innovations in prevention or management strategies for SCD complication.

5.3. METHODS

Unlike NICE, our cost study used HES data, the same data set that we used to understand trends in SCD and its link to socioeconomic deprivation. We limited the cost study to the period from April 2010 to March 2011. The analysis was carried out at the national level for England and the results are reported for England, London and London PCTs. We used ICD-10 coding with the following primary diagnoses:

- Sickle cell anaemia with crisis (D57.0)
- Sickle cell anaemia without crisis (D57.1)
- Double heterozygous sickling disorders (D57.2)
- Sickle cell trait (D57.2)
- Other sickle cell disorders (D57.2-8)

HES data contain a number of administrative and demographic variables on patients admitted to hospitals and their healthcare providers. These variables, which can be seen in Table 9, include the patients' Primary Care Trust (PCT), their Strategic Health Authority (SHA), their sex,

ethnicity, age, length of hospital stay as well as whether the admission was emergency or elective (Pizzo, Lavery, Phekoo, et al., 2014). As in our other analyses of the HES data which helped identify trends in admissions, The ICD-10 codes above were used to split admissions into two groups:

- Sickle cell crisis (D57.0), or
- Sickle cell without crisis (D57.1-8)

As data regarding the cost and resource consumption for SCD admissions were not available, we used the Healthcare Resource Group (HRG) tariffs as a proxy of costs. HRGs form the basic currency for payment of NHS services by relying on an informed assumption that health care costs are exclusively driven by diagnosis or procedure. The English HRG tariffs provide cost data for elective and non-elective hospital episodes and additional per diem cost for very long stays that exceed the so-called 'trim point', which marks the expected length of stay (LOS) for each HRG. Estimated costs are reported as a national schedule of reference costs and are also aggregated by hospital to provide the reference cost index (Street & Dawson, 2002; Geue, Lewsey, Lorgelly, et al., 2012).

This was a first attempt to assess the cost of admissions due to SCD using the HES data. The available HRG tariffs for SCD are coded as (Table 10):

- PA47Z Sickle cell anaemia with crisis (for paediatric admissions);
- SA10D Sickle cell anaemia with complications (CC) (including SCD with crisis);
- SA10F Sickle cell anaemia without complications (CC) (Pizzo, Lavery, Phekoo, et al., 2014).

Because there are no specific tariffs for each ICD-10 code, we made certain assumptions:

- a. ICD-10 'Sickle cell anaemia with crisis' (D57.0) is linked to HRG PA47Z 'Sickle cell anaemia with crisis' for paediatric admissions (age between 0-19 years);
- b. ICD-10 'Sickle cell anaemia with crisis' (D57.0) is linked to HRG SA10D 'Sickle cell anaemia with CC' for adult admissions (age >20 years);
- c. ICD-10 'Sickle cell anaemia without crisis and other sickle cell disorders (D57.2-8) might be linked with HRG code SA10F 'Sickle cell anaemia without CC' (Pizzo, Lavery, Phekoo, et al., 2014).

All admissions where SCD did not appear as a primary diagnosis were eliminated from this analysis. We saw many episodes in which SCD appears as a secondary, tertiary or further underlying diagnosis. We understand that even if the main cause of admission is not an SCD crisis, the disease's underlying characteristics may have predisposed the patient to be admitted to hospital or increased the risk of comorbidity or complications. We excluded all of these episodes. This underestimated the cost of admission linked to SCD, but we elected to engage a conservative approach which excluded any costs that were not seen to be direct linked to SCD. Both emergency and elective admissions were analysed in isolation. They have different tariffs and by analysing elective and emergency separately, we were able to show the impact of emergency admissions due to SCD (Pizzo, Lavery, Phekoo, et al., 2014).

The LOS for each admission was also considered in order to assess hospital costs that exceeded the trim point of the HRG tariff. The trim point identifies the LOS covered by the tariff, after which an extra daily payment is required for each specific HRG. The trim points vary according to the type of HRG and type of admission (emergency versus elective admission) and extra days carry an extra cost. For HRG PA47Z Sickle cell anaemia with crisis, the trim point is 9 days for emergency admissions and 8 days for elective admissions; for HRG SA10D Sickle cell anaemia

with CC¹, the trim point is 30 days both for emergency and elective admissions (Appendix 1). Therefore, an LOS exceeding the trim point will increase the cost by £234 for each extra day for paediatric admissions and £204 for adult admissions. When we refer to the total cost of admissions for SCD, this amount is the combined sum of the cost of admissions in emergency and elective for both paediatrics and adults (Pizzo, Lavery, Phekoo, et al., 2014).

When we refer to the cost of extra LOS for admissions associated to SCD, this is the difference between the number of days spent in hospital (LOS) and the trim point from the HRG tariff. The total number of days in hospital exceeding the trim point is multiplied by the daily extra cost (£234 per day for paediatric admissions and £204 for adults). The total cost of extra LOS is the sum of the extra cost of LOS of emergency and elective admissions for both paediatrics and adults (Pizzo, Lavery, Phekoo, et al., 2014).

5.4. RESULTS

In 2010-2011, England had 6,077 admissions associated with SCD with crisis as a primary diagnosis. The total cost for these admissions was £18,798,255. Ninety-one percent (91%) of these costs were due to emergency admissions (Table 9). Adult admissions represent 84.5% of all admissions, representing a cost of £14,370,931 for emergency admissions. Admissions for children represent 15.5% of total admissions, and the cost of emergency admissions was £2,714,357. The NICE report estimated £16.2 million in costs (Anon, 2012) and our study estimated approximately £17.1 million in costs. Our study would have likely found even a greater cost burden had we taken into account all of the day case activities (which the NICE report did) or admissions that have SCD with crisis as a secondary, tertiary or further diagnosis which explains the cost discrepancy between our study and the NICE report. The approximately £1 million difference in cost estimation is likely due to the different data set used and/or the different time period. Neither study had a standard to go by (Pizzo, Lavery, Phekoo, et al., 2014).

In total, the LOS exceeded the trim point by 1,144 days, for an extra cost of £294,697 (93.1% for emergency admissions) (Table 9). For emergency admissions, the extra LOS is 438 days for children and 624 days for adults, for an added cost of £274,104. Because the tariff for children has a shorter trim-point compared to adults (9 days instead of 30 days) and an extra day costs more, long admissions for children represent a higher cost in terms of LOS (Pizzo, Lavery, Phekoo, et al., 2014).

Within England, London had the highest number of admissions associated with SCD, with 4,363 admissions for SCD crisis as the primary diagnosis at a total cost of £14,223,139, of which £13,984,884 were for emergency admissions (89.9%) (Table 9). In line with the national data, adults represent 87.1% of total admissions and children account for 12.9%. London accounts for 75.6% of all of England's costs for admissions associated to SCD and 80.8% of all of England's days beyond the trim point (extra days). This is consistent with our previously published analysis measuring trends for admissions related to SCD (Aljuburi, Lavery, Green, et al., 2012a). Table 10 shows a summary for all PCTs in England, with the cost of admissions for SCD and the rate of admissions every 100,000 (Pizzo, Lavery, Phekoo, et al., 2014).

¹ Haemoglobin C Disease

TABLE 9: COSTS FOR ADMISSIONS AND EXTRA LOS ASSOCIATED WITH SICKLE CELL ANAEMIA WITH CRISIS (ICD-10 D57.0) IN ENGLAND AND LONDON, 2010-11

Admissions (England)	Emergency	Elective	Total admissions
D57.0 primary diagnosis admissions (total)	5,514	563	6,077
D57.0 primary diagnosis admissions (paediatrics)	1,462	154	1,616
D57.0 primary diagnosis admissions (adults)	4,052	409	4,461
HRG tariff PA47Z (to be adjusted by MFF)	£1,534	£1,210	
HRG tariff SA10D (to be adjusted by MFF)	£2,959	£2,950	
Total cost for admissions	£17,085,288	£1,712,966	£18,798,255
Long stay trim point (days) for paediatric	9	8	
Long stay trim point (days) for adults	30	30	
Total days exceeding the trim point (paediatrics)	438	7	445
Total days exceeding the trim point (adults)	624	75	699
Per day long stay payment (for days exceeding trim point) paediatrics (£)	£234	£234	
Per day long stay payment (for days exceeding trim point) adults (£)	£204	£204	
Total cost for extra LOS	£274,104	£20,592	£294,697
Admissions (London)	Emergency	Elective	Total admissions
D57.0 primary diagnosis admissions (total)	3911	452	4363
D57.0 primary diagnosis admissions (paediatrics)	877	107	984
D57.0 primary diagnosis admissions (adults)	3034	345	3379
Total cost for admission in London	£12,788,339	£1,434,799	£14,223,139
Long stay trim point (days) for paediatric	9	8	
Long stay trim point (days) for adults	30	30	
Total days exceeding the trim point (paediatrics)	246	5	251
Total days exceeding the trim point (adults)	586	75	661
Total extra cost for extra LOS in London	£218,153	£20,101	£238,254

Source: our elaboration using HES data and HRG tariffs 2010-2011 (Pizzo, Laverty, Phekoo, et al., 2014)

There was high variability in the number of admissions across PCTs in England, but City and Hackney was the PCT with the highest number and the highest costs in 2010-2011, followed by Haringey and Brent (Appendix 1). In these PCTs, the rate of SCD admissions was the highest in England, i.e. 346.67 and 153.38 for every 100,000 people, respectively. City and Hackney also had the highest costs associated with LOS beyond the trim point, followed by Redbridge and Westminster (Appendix 1). Outside London, Luton was the PCT with the highest rate of SCD

admissions (39.41 every 100.000), followed by Nottingham (32.47), Manchester (20.01) and South Birmingham (20.10) (Appendix 1) (Pizzo, Lavery, Phekoo, et al., 2014).

TABLE 10: COST OF NON-ELECTIVE ADMISSIONS

Healthcare Resource Group	Number of Admissions	Cost of Non-Elective Admissions (£'000)
PA47Z Sickle cell anaemia with	2,489	4,198
SA10E Sickle cell anaemia with crisis or with co-morbidities	7,714	11,367
SA10F Sickle cell anaemia without co-morbidities	906	637
Total	11,109	16,202

The regression models performed to check whether age and gender are significantly associated with SCD costs showed that admissions in adults cost more than in children (Table 11). The total cost increases with age: admissions in age<9 and age 10-19 cost £1,740 and £1,686 (p<0.001) less compared to age 20-29; admissions costs £65 more in the group age 30-49 compared to 20-29. There was no statistically significant difference in the cost for patients over 50 years.

Admissions were more costly for males than females, but the differences are not statistically significant. The results for London are similar to those for the rest of England (Pizzo, Lavery, Phekoo, et al., 2014).

TABLE 11: REGRESSION OF TOTAL COSTS OF ADMISSION ON AGE AND SEX AT THE NATIONAL LEVEL AND LONDON LEVEL ONLY

National				
		Geometric mean cost (£)	Comparison (as % of reference group costs)	p-value
Age	<1	1758	50.26	<0.001
	1 to 9	1732	49.53	<0.001
	10 to 19	1812	51.83	<0.001
	20 to 29	3499	ref	ref
	30 to 39	3564	101.87	<0.001
	40 to 49	3565	101.94	<0.001
	50+	3535	101.10	0.194
Sex	Men	3021.	ref	ref
	Women	2854	99.59	0.195

London				
		Geometric mean cost (£)	Comparison (as % of reference group costs)	p-value
Age	<1	1828	50.27	<0.001
	1 to 9	1804	49.61	<0.001
	10 to 19	1881	51.71	<0.001
	20 to 29	3636	Ref	ref
	30 to 39	3640	100.11	<0.001
	40 to 49	3638	100.03	<0.001
	50+	3622	99.55	0.605
Sex	Men	3182	Ref	ref
	Women	3052	100.36	0.278

Source: (Pizzo, Lavery, Phekoo, et al., 2014)

The logistic regression to examine the association between age and gender on incurring extra bed days shows that patients 10-19 years old are much more likely to incur an extra bed days (odds ratio of 8.3 with $p < 0.001$) compared to patients aged 20-29 years old. The results for other age groups and for gender did not show statistically significant differences (Table 12) (Pizzo, Lavery, Phekoo, et al., 2014).

TABLE 12: LOGISTIC REGRESSION OF THE ASSOCIATION BETWEEN AGE AND GENDER ON INCURRING EXTRA BED DAYS AT THE NATIONAL LEVEL AND LONDON LEVEL

National				
		% staying extra days	Odds Ratio	p-value
Age	<1	0	-	-
	1 to 9	1.31	1.26	0.576
	10 to 19	8.08	8.38	<0.001
	20 to 29	1.04	Ref	ref
	30 to 39	1.11	1.07	0.834
	40 to 49	0.93	0.90	0.8
	50+	1.27	1.22	0.745
Sex	Men	1.98	Ref	ref
	Women	2.47	1.00	0.985

London				
		% staying extra days	Odds Ratio	p-value
Age	<1	0	-	-
	1 to 9	0.82	0.54	0.316
	10 to 19	8.1	5.65	<0.001
	20 to 29	1.47	ref	ref
	30 to 39	1.26	0.84	0.616
	40 to 49	0.85	0.56	0.24
	50+	1.7	1.09	0.895
Sex	Men	1.78	ref	ref
	Women	2.73	1.35	0.162

Source: (Pizzo, Laverty, Phekoo, et al., 2014)

5.5. DISCUSSION

My research showed that the studies that assess the cost of SCD are either primarily referring to children, not appropriately updated, or have been conducted in the United States (Amendah, Mvundura, Kavanagh, et al., 2010b; Ballas, 2009; Bilenker, Weller, Shaffer, et al., 1998; Kauf, Coates, Huazhi, et al., 2009; Nietert, Silverstein & Abboud, 2002). Studies conducted in the United States show that patients with SCD who are frequently admitted to emergency departments account for the majority of health care expenses associated with the disease (Carroll, Haywood & Lanzkron, 2011).

In England, the National Institute for Health and Clinical Excellence (NICE) has assessed, according to guidelines, the costs (and potential savings) attributed to managing an SCD acute painful episode (Anon, 2012). The cost savings may vary significantly depending on current practice and measuring the local prevalence of SCD. The NICE costing report for SCD provides an estimate for the cost of admission for 'sickle cell anaemia with crisis' in children and adults according to the National Schedule of Reference costs in 2010/11. The national schedules of reference costs show the national average unit cost for each service for which costs were collected in 2010-11. According to the report, the approximate total cost to the NHS of these admissions and some day-case activity was approximately £16.2 million. This total did not include elective admissions and showed a resulting estimate at £589 per day for children and £456 per day for adults. The costs were based on NHS Reference Costs for 2010/2011, but it was not clear if the period 2010-2011 was a calendar or fiscal year (Pizzo, Laverty, Phekoo, et al., 2014).

In our study, the admissions having SCD with crisis as a primary diagnosis represented 92.5% of all admissions for SCD with crisis (including also secondary, third, fourth and fifth diagnosis), so almost 7.5% of admissions are not included here. Assuming we can apply the same HRG tariffs for the excluded admissions (those after the primary diagnosis), the total cost for admissions linked to SCD with crisis will increase to £20,376,822, a difference of £1,578,567 (Pizzo, Laverty, Phekoo, et al., 2014). Our study also showed that the cost of admissions in adults cost more than in children but that those aged 10-19 years are more likely to incur longer hospital stays.

The other 13,589 admissions were linked to SCD without crisis (ICD-10 codes D57.1-8) as the primary or further diagnosis, but the cost for these admissions was not taken into account in this study. We cannot accurately estimate the cost of these admissions without knowing the

HRG tariff applied when the patient was discharged. However, the primary diagnosis of these admissions was mainly for SCD without complications, anaemia, respiratory infections and asthma. Taking into account that 25% of these admissions were in the emergency department, the cost of admissions for SCD is even higher (Pizzo, Lavery, Phekoo, et al., 2014).

The bottom line is that, in England, we are spending well over £16.1 million a year on hospital costs due to SCD and that approximately 90% of these costs are emergency admissions. Furthermore, over three-fourths of these admissions occur in London. This information can be of great value to health economists, policy makers and commissioners. From a policy perspective, the combination of trends and costs related to SCD should raise a red flag as to how we are managing this disease. It may be a disease minor in nature compared to other chronic diseases and one predominantly affecting minorities, but from a cost and efficiency perspective, SCD has a major impact on emergency hospital admissions and costs to the NHS.

5.6. KEY POINTS FROM CHAPTER 5

1. This is the first study in England to try to assess the cost of a hospital admission due to SCD using HES data.
2. In 2010-2011, England had 6,077 admissions associated with SCD with crisis as the primary diagnosis. The total cost for these admissions for commissioners was £18,798,255.
3. Over 90% of hospital admission costs were for emergency admissions.
4. London accounts for over 75% of all of England's costs for admissions associated with SCD.
5. SCD represents a significant cost for commissioners and the NHS. SCD patients must be managed better in order to potentially lead to a reduction in admissions and length of stay and associated costs.

6. TRANSLATING RESEARCH INTO PATIENT CENTRIC CARE

6.1. BACKGROUND AND OBJECTIVE

The first four studies within this thesis look at trends and costs of SCD management. The next two studies focus on patient input. One clear objective of patient participation is to help translate research into more patient-centric care. In fact, achieving patient-centric care has been a primary motive behind the majority of CLAHRC-funded projects (Renedo & Marston, 2011), including our project on SCD. (Parsons, 2010).

Based on the findings from the trends pieces, we are better able to see what the impact of disease is and what areas would be considered high prevalence. Using these high impact areas, I looked at the use of focus groups and patient questionnaires as methods for patient engagement in the design of health services that would be targeted to such areas.

6.2. FOCUS GROUPS AS A METHOD FOR PATIENT ENGAGEMENT IN THE DESIGN OF SERVICES

6.2.1. BACKGROUND

Focus group discussions are one of the methods that have been used to obtain patient involvement in design of health services and can be particularly useful for people with long term or chronic diseases. They provide a forum for a broad range of participants to express their concerns about the quality of care they receive from their care providers, their opinions on what the role of their care providers should be in managing their disease and to discuss their perceived barriers to utilising different level of care services (Underutilised, In & 1987;14:411–48, n.d.).

Furthermore, focus groups can provide a space for an open dialogue tailored to the level of the participants (Kitzinger, 1995) and allow the facilitator to clarify and push for more detailed responses to enrich the understanding of the issues (Wong, 2008). The use of a focus group as a data collection technique can be particularly useful with SCD to ensure sensitivity to cultural variables, which is why they are now so often used in cross-cultural research and work particularly well with ethnic minorities. SCD is a disease that disproportionately impacts people of African and Caribbean origin; the focus group setting allows the patients to reveal the frustration felt as an ethnic minority receiving health care services for a predominantly race-related disease.

Consequently, it makes them useful in studies examining why different sections of the population make differential use of health services (Underutilised, In & 1987;14:411–48, n.d.; Kitzinger, 1995). The group dynamic also works to facilitate the discussion of difficult or uncomfortable topics because the less reserved members of a focus group can ‘break the ice’ for the more reserved participants (Kitzinger, 1995). Some researchers have also noted that group discussions can generate more criticism of the health care system than interviews (Watts & Ebbutt, 1987). For example, Geis et al. found in their study that ‘there were more angry comments about the medical community’ in the group discussions than in the individual interviews: ‘perhaps the synergism of the group “kept the anger going” and allowed each participant to reinforce another's vented feelings of frustration and rage’ (Geis, Fuller & Rush, 1986). Some researchers argue that focus groups are limited in their ability to draw inferences for large groups or populations and their incapacity to test hypotheses in traditionally designed

experiments (Heary & Hennessy, 2002). Others have also claimed that focus groups can lead to 'tagging' which means that participants agree for the sake of the group's momentum and that with regards to more sensitive topics, interviews worked better because the participant feels a greater level of anonymity than when part of a group discussion (Heary & Hennessy, 2002). Such points show that the use of focus groups may be subject to bias and need to be individually evaluated (Wong, 2008). Focus groups may not add value to every study but can work well when combined with other forms of qualitative and quantitative methods (Morgan, 1997; Heary & Hennessy, 2002). In general, however, qualitative methods such as interviews and focus groups help to determine what matters to people so that services are designed to better meet these needs (NHS England, 2013).

6.2.2. METHOD AND RESULTS

We sought to engage SCD patients and carers in designing a local intervention aimed at better educating general practitioners on SCD through the use of a focus group discussion. On 12th August 2010, we held a focus group discussion at the Sickle Cell Society in the London Borough of Brent. We aimed to identify patient and carer perspectives of GP knowledge and treatment of SCD, and to identify areas in which improvements could be made in its coordination.

The discussion lasted for approximately two hours. The participants were recruited through the Sickle Cell Society in Northwest London and comprised people of a mixture of ages and genders. Specifically, the group was made up of ten patients; two males and eight females, aged 9-56 years, all of African/Afro-Caribbean heritage. The focus group was held at the Sickle Cell Society, which is conveniently located to where many people with SCD live. The location is also a familiar meeting point where all of the participants felt secure and at ease to voice their opinions. The focus group participants were recruited through a purposive sampling process which aimed to canvas the views of sickle patients and carers living within the specific locality in which the service improvement and evaluation work was being undertaken. Participants were recruited via the Sickle Cell Society, a national UK sickle cell charity, through a general mail out to members of the charity living within this area with information about the study. This was subsequently followed up by a telephone call (from Comfort Ndivo of the Sickle Cell Society) to assess interest in participating in the focus group. The discussion was facilitated by a clinical psychologist (Dr. Kofie Anie) from a local NHS Trust experienced in handling patients with SCD who explained the aims of the project to the participants; and who then asked the participants questions regarding their personal experience with primary health care in London. The questions were open-ended and focused on general practice utilisation, what patients do first when they are in an SCD crisis, and overall satisfaction with primary care. Eleven observers from the project group were present; two were scribes. As well as making written notes, the discussions were also video recorded (AlJuburi, Phekoo, Okoye, et al., 2013b).

The key themes used to explore the participants perceptions and aspirations regarding primary care services for SCD are listed in Table 13. The participants were also encouraged to raise their own concerns about SCD management. Following the focus group, the project team (Ghida AlJuburi and Dr. Karen Phekoo) read the transcripts independently and grouped the responses into key themes that would help inform planning of future SCD services. Each item in the data collected during the discussions was compared with the rest of the data to establish analytical groups. Consensus regarding categories and a final list of key themes identified during the discussions was achieved iteratively through discussion and re-reading of transcripts. As we aimed to present viewpoints, the data are not presented numerically (AlJuburi, Phekoo, Okoye, et al., 2013b).

TABLE 13: KEY THEMES USED TO GENERATE GROUP DISCUSSION

• Frequency in accessing GP care for managing a crisis
• Satisfaction level with primary health care services
• Current use of primary health care services
• What do you do first when in an SCD crisis?

The transcripts and recordings were reviewed to identify common perceptions and experiences among the participants.

Accessibility

GP access was an important issue. The participants highlighted that difficulty in obtaining an appointment with the GP and long waiting times led to patients' sidestepping their GP in the event of a painful crisis or other issues relating to SCD. Although GPs were seen as useful for prescribing antibiotics and other repeat prescriptions, they were seen to be unreliable during crises. Hospitals were seen to be more accessible. Participants proposed that GPs should offer more 'out of hours' and house calls for pain management, since many crises occur at night (AlJuburi, Phekoo, Okoye, *et al.*, 2013b).

I cannot walk in [to a General Practitioner's office] and I even if I could, I probably won't be able to see a GP immediately, so I go straight to the hospital if I am sick.

Doctor-Patient Relationships

Participants raised the issue of the lack of time to build up rapport and relationships with their GPs. This left patients with SCD with a feeling of dissatisfaction with the quality of the doctor-patient relationship. They felt that GPs were not actively interested or engaged in the progress and treatment of their condition. They encouraged GPs to take a proactive approach and interest in getting to know their SCD patients and to focus on preventive care, including preventive management of crises (AlJuburi, Phekoo, Okoye, *et al.*, 2013b).

If I take my son to hospital now, the doctors like to have a relationship and know how he is doing. If I take him to the GP there is no relationship; he is a stranger.

[You have a] five minute conversation with the GP. You are in and out. I prefer to go to the hospital, I know them there. I am safe. My nurse knows me.

I know my GP, but does he really know me? I go to the hospital. They know me.

GP Knowledge

There was a general feeling amongst the participants that GPs lack comprehensive knowledge of SCD, and the hospitals were viewed as more specialised in dealing with the condition. They expressed an interest in seeing GPs better informed about SCD, and its implications for management. One participant suggested a GP SCD 'champion' or specialist who could further inform their health professional colleagues about SCD. Another participant added that GPs who are not specialised in SCD need to be briefed by an expert before seeing an SCD patient in the surgery (AlJuburi, Phekoo, Okoye, *et al.*, 2013b).

They take time to know about other diseases; I feel my disease is not important, I feel I am not important.

Concerns about Pain Management & the Treatment Process

Because of the organisational barriers previously mentioned, many of the participants had difficulty in seeing the benefit of utilising a GP during an uncomplicated sickle crisis. They subscribed to a process that seemed to utilise the emergency department during a crisis and to utilise the GP for repeat prescriptions and immunisations. Patients are still waiting for facilitating factors highlighted by the below patient quote which would encourage them to use the GP as a means of support, prevention, and maintenance during crisis-free periods or during uncomplicated crisis pain episodes (AlJuburi, Phekoo, Okoye, *et al.*, 2013b).

It takes too long to see a GP. Sometimes something goes wrong while you are waiting for your appointment. Other than a prescription or a non-sickle illness, there is no need for the GP. It is a waste of time. When you have access to a seven day hospital, why go to a GP?

They [General Practitioners] don't take the time to find out about the patients they have on their books anyways.

Continuity of Care & Follow-Up

One discussion point that surfaced several times was the seeming lack of cohesion and communication between GPs, hospitals, and specialty centres. Participants emphasised that there was never follow-up from their GP after they were discharged from the emergency department, hospital or a specialty centre. Participants felt strongly that GPs should liaise with other healthcare professionals involved in the management of their sickle condition, and should follow-up with the patient soon after hospital discharge (AlJuburi, Phekoo, Okoye, *et al.*, 2013b). This Institutional and patient management is what we define in this paper as both integration and coordination of care, and I discuss this further in Chapter 10 where approaches to policy are summarised.

[There should be] review letters from the hospital. They [General Practitioners] should see how you are doing. That will be good.

[Following discharge from hospital] why don't they [GPs] ever call the patient and follow up?

Chronic Illness Management – Wellbeing Management

There was agreement amongst the focus group participants that many of the frustrations felt by patients with SCD are also shared by sufferers of other chronic illnesses. GPs were seen as treatment and medication prescribers rather than actively engaged in the management of their patients' well-being through prevention of disease and maintenance of health (AlJuburi, Phekoo, Okoye, *et al.*, 2013b).

[GP consultation] five minute conversation with the GP... [to get your] medication or antibiotics, or certain jabs, they don't take the time.

The key issues identified through the focus groups are summarised in Table 14.

TABLE 14: ISSUES IDENTIFIED BY FOCUS GROUP DISCUSSION

Access & difficulty in obtaining urgent appointments
Poor communication between health professionals
Poor follow-up and discharge planning processes
No proactive role in maintaining health and in preventive care
Limited knowledge about sickle cell disease
Limited knowledge about the standards and guidelines for disease management
Concerns about pain management

6.2.3. DISCUSSION ON FOCUS GROUP

Our focus group discussion results could have benefited by having more participants and more sessions. However, the discussion on SCD provided an opportunity to not only voice the frustrations of participants but created a venue whereby solutions could be proposed. The discussion was candid, clear and offered perspectives about the participants' individual and collective needs that could not otherwise be obtained from healthcare professionals. They offered constructive advice on what they needed to feel satisfied as health care consumers and gave a clear understanding about which areas we needed to focus on regarding the design of primary care practice intervention. They also earmarked issues that need to be addressed in future research, including auditing and improving communication between the different providers of health care and recommending quality improvement schemes in areas with a high prevalence of SCD (Aljuburi, Phekoo, Okoye, *et al.*, 2013b).

6.3. PATIENT QUESTIONNAIRE AS A METHOD FOR PATIENT ENGAGEMENT IN THE DESIGN OF SERVICES

6.3.1. BACKGROUND

In addition to focus group studies such as the one we conducted, other studies have shown that SCD patients are dissatisfied with the quality of care they receive from their GPs (Anie, Steptoe & Bevan, 2002). Primary care, however, is still a good place to deal with chronic illness as it offers continuity, coordination, and comprehensiveness (Rothman & Wagner, 2003). These studies also show that assessing satisfaction is an important part of evaluating comprehensive quality of care (Ireys & Perry, 1999). Patient satisfaction has been regarded as the 'ultimate outcome of the delivery of health care', and measuring and responding to satisfaction can be important with regards to treatment compliance and over or underutilising the health care system (Kelley, Alexander & Morris, 1991).

While properly constructed patient questionnaires can be useful in measuring patient satisfaction (Grogan, Conner, Norman, *et al.*, 2000), they are also considered another way in which we can engage patients in the design of health services. More specifically, we can measure patient satisfaction in primary care, the design of healthcare services in primary care, and the measurement and management in primary care of SCD.

One study showed that a questionnaire is a valid and valuable tool to use for assessing health-related quality of life in children with SCD and could serve as 'an important adjunct to determine the effect of SCD on the lives of children' (Panepinto, Mahar, DeBaun, *et al.*, 2004). Other studies have shown that questionnaires are a valuable tool in measuring quality of care or

patient satisfaction in primary care (Raaij, Landgraf, Bonsel, *et al.*, 2002), particularly where the questionnaire assesses a specific area that enables the GP or other health care provider to identify which aspects of service patients find unsatisfactory. This in turn allows for the opportunity to improve a specified area (Harris, Swindle, Mungai, *et al.*, 1999). A separate study discussing patient survey programmes showed that this method is a beneficial way of ‘monitoring performance across the system’, because they require providers to survey patient experiences and to publish these results (Coulter, 2005).

In a historical context, survey data can offer valuable insight as ‘they let us recapture what ordinary people really thought about critical issues’, like health services. Furthermore, regardless of whether people have a clear understanding of a particular health reform (or in our case, this can be held equally true for a particular health service), people always have a clear opinion about their experience which should not be dismissed or ignored because of a lack of detailed knowledge about a particular reform or service (Hayes, 2013).

Our analysis of emergency department admissions between January 2008 and July 2010 in the northwest London Borough of Brent showed that patients with SCD tend to use the emergency department rather than seek advice and support from their GP (Green, Aljuburi, Majeed, *et al.*, 2012a). Our focus group study aimed at obtaining patient perspectives held in Brent and showed that one of the reasons patients utilise the emergency department over their GP is because they perceive GPs as having limited knowledge of SCD (Aljuburi, Phekoo, Okoye, *et al.*, 2012). We also know that SCD clinical guidelines recommend that patients see a general practitioner (GP) for routine examination every six months, or more often if new problems arise or their treatment protocol changes (NHS Sickle Cell and Thalassaemia & Screening Programmes, 2010). In addition, immunisations, prescriptions, and other preventive care measures need to be delivered effectively by GPs to prevent recurring infections and pain crises (Wong, 2001).

We designed a pilot questionnaire to elicit the views of patients about the quality of care they have been receiving from their primary health care providers and what they thought about the role of primary care in SCD management (Aljuburi, Okoye, Majeed, *et al.*, 2012).

6.3.2. METHODS

The study consisted of the development and administration of a 14-item study-specific questionnaire devised by a sickle cell steering committee which examined patients’ perceptions towards SCD and key management issues in primary care, including the severity of disease, how many times patients visited the emergency department in the last year, how many times patients saw or called their GP for sickle and non-sickle related illnesses, and general questions about GP satisfaction (Figure 12). The validation process for the questionnaire followed several drafts reviewed by seven GPs practising across inner London, a professor of paediatric haematology specialising in SCD, a haematologist specialising in SCD in adults, an SCD specialist nurse, an SCD social worker, an SCD clinical psychologist, a number of quality improvement project managers, public health specialists, patient representatives with SCD, and directors of the Sickle Cell Society (a national UK sickle cell charity) (Aljuburi, Okoye, Majeed, *et al.*, 2012).

FIGURE 12: A SELECTION OF QUESTIONS FROM THE PATIENT QUESTIONNAIRE

Question

In the past year, how many times have you seen or called your GP for help with a sickle cell crisis?

.....

Question

Rank the three main things you do most often when in crisis:

() Do nothing () Go to GP

() Call Sickle Cell Specialist Nurse () Go to A&E

() Stay at home () Other (please state):.....

Question

How often do you utilise your GP to:

a) Manage a painful crisis

Never	Rarely	Sometimes	Fairly often	A lot
	(1-2/year)	(3-4/year)	(monthly)	(weekly)

b) Collect repeat prescription for antibiotics, folic acid, analgesia

Never	Rarely	Sometimes	Fairly often	A lot
	(1-2/year)	(3-4/year)	(monthly)	(weekly)

c) Get general advice about sickle cell disease

Never	Rarely	Sometimes	Fairly often	A lot
	(1-2/year)	(3-4/year)	(monthly)	(weekly)

d) Get help with non-sickle related illness (e.g. coughs, colds, etc.)

Never	Rarely	Sometimes	Fairly often	A lot
	(1-2/year)	(3-4/year)	(monthly)	(weekly)

Other reasons (please specify)

.....

Question

How satisfied are you with assistance given by your GP to help manage your sickle cell disease

0	1	2	3	4	5	6	7	8	9	10
---	---	---	---	---	---	---	---	---	---	----

(0=Not satisfied at all) (10=Very satisfied)

Question

List the kind of support you currently receive from your GP

Pain control

Collecting prescription

General SCD advice

Contraception

Other (please state)

Question

List the kind of support you would like to receive from your GP

* Full version of the questionnaire can be found in the Appendix.

Participants

One hundred questionnaires were distributed by post and in person over a nine-month period from November 2010 to July 2011 to members of the Sickle Cell Society and to patients that attended the Sickle Cell and Thalassaemia Centre, using a purposive sampling method. Forty questionnaires were properly completed and suitable for analysis (Aljuburi, Okoye, Majeed, *et al.*, 2012).

6.3.3. RESULTS

A response rate of 40% was achieved. Fifty percent (20) were completed by males and 50% (20) by females. The highest percentage of respondents (27%) was in the age range of 25- 39 years. Five percent of respondents were aged 65+years. The majority (62%) of the respondents were in good health and mobile. Nineteen (47%) respondents did not use their GP to manage a painful crisis and preferred to attend the emergency department. Twenty-four (59%) respondents went to their GP to collect repeat prescriptions. Twenty-two (55%) respondents did not visit their GP to get general advice about SCD (Table 16). Nine (23%) rarely visited their GP with four (10%) visiting sometimes or fairly often (Table 15). When asked how satisfied patients are with assistance given by their GP to help manage their SCD based on a scale of 0-10 (0 being not satisfied at all and 10 being very satisfied), the majority (54%) scored satisfaction with their GP as 5 or less while 43% scored a 6 or above and 3% did not answer the question at all.

Collecting prescriptions was the reason most cited for visiting the GP's office (Table 16). Some examples of comments around the services provided by GPs include:

'The GP should know about this disease.' (Patient Questionnaire 1) (Aljuburi, Okoye, Majeed, *et al.*, 2012)

'The GP does not know anything about SCD pain and crisis. I would rather manage ... at home or [go] to A+E where immediate action will be taken rather than call the GP who will ask us to book an appointment and more or less does not understand how to manage the pain or how severe or serious the pain is.' (Patient Questionnaire 2) (Aljuburi, Okoye, Majeed, *et al.*, 2012).

TABLE 15: HOW OFTEN DO YOU USE YOUR GP TO GET GENERAL ADVICE ABOUT SCD?

	Number of Respondents	% of Respondents
Never	22	55
Rarely	9	23
Sometimes	4	10
Fairly often	4	10
Not answered	1	3

Source: (Aljuburi, Okoye, Majeed, *et al.*, 2012)

TABLE 16

	Number of Respondents	% of Respondents
Pain control 1	4	35
Collecting prescription 3	7	93
General SCD advice	3	8
Contraception	2	5
Other (please state)	1	3

Source: (Aljuburi, Okoye, Majeed, *et al.*, 2012)

The results from the survey show that many patients are generally not satisfied with the quality of care that they are receiving from their primary health care providers for SCD. Thus, most do not make use of GP services for management of their SCD. More importantly, the majority of the group did wish for greater involvement from their GP services, even if it was just to refer them to a tertiary care facility or social support (Aljuburi, Okoye, Majeed, *et al.*, 2012).

6.3.4. DISCUSSION ON PATIENT QUESTIONNAIRE

The pilot study successfully identified issues with SCD management at the primary care level through the use of a questionnaire which could help inform the planning of a larger survey. Moreover, this study also identified a need to improve patient satisfaction and engagement with primary care. Raising awareness amongst GPs about SCD management was identified as a key method to improve GP-patient relationships and improve primary care clinic attendance (Aljuburi, Okoye, Majeed, *et al.*, 2012).

Although this pilot study increased our understanding of the barriers associated with SCD management in primary care, it has certain limitations which could limit its transferability to areas outside of northwest London. The questionnaire was based in an area with high prevalence of SCD and the sample size was small and consisted of a purposive or volunteer sample (Aljuburi, Okoye, Majeed, *et al.*, 2012). Despite its limitations, the study, along with the results of the focus group discussion which reinforced these findings (Green, Aljuburi, Majeed, *et al.*, 2012a), provides some valuable information that provide an opportunity to develop a disease specific intervention which aims to improve patient care and help to ensure that management in primary care is optimised through the establishment of a local policy initiative (Aljuburi, Okoye, Majeed, *et al.*, 2012).

Shared decision making between clinicians, health policy makers and patients is important in the design of health services, but how patient involvement is defined and the mechanism for involvement is not always clear and has been a source of debate between patients and health care providers (Renedo & Marston, 2011).

6.4. DISCUSSION

6.4.1. PATIENT SATISFACTION AND THE MERITS OF INVOLVING PATIENTS IN THE DESIGN OF SERVICES AND CARE PATHWAYS

There have been few data collected about SCD patients and their level of satisfaction with primary care services. However, one study showed a 46% non-adherence rate for routine primary care appointments for SCD patients during an eight-month period (Robinson N, Huber H & Jenkins P, 2006). One of the reasons cited for non-adherence was the patient-provider relationship. Those with a positive patient and GP relationship were more likely to attend their clinic appointments (Modi AC, Crosby LE, Guilfoyle SM, et al., 2009; Freed, Ellen, Irwin, et al., 1998). Other studies of chronic disease management showed that poor communication, multiple treating physicians, long waiting periods, and past negative experiences with health care providers all contributed to patient dissatisfaction with primary care and poor attendance at clinics (McCarthy, McGee & O'Boyle, 2000).

With patient satisfaction established as an important indicator of health outcomes, researchers and health care providers have become increasingly interested in measuring patient satisfaction as an indicator of quality of care. In fact, assessing patient satisfaction is one of the core requirements of contract for GPs in the UK (Grogan, Conner, Norman, et al., 2000). One study showed that assessing patient satisfaction allows GPs to investigate 'the extent to which their service meets the needs of their client group' (Avis, Bond & Arthur, 1995). Another study showed that satisfied patients are more likely to follow a treatment protocol because the patient is more likely to believe that the treatment will be effective (Hardy GE, West MA & Hill F, 1996). In order for satisfaction to be measured in a meaningful way, a valid and reliable measure should be applied. The use of patient questionnaires is one reliable way to assess patient satisfaction (Grogan, Conner, Norman, *et al.*, 2000) and also one of the Institute of Medicine's suggestions for achieving patient centred care (IOM).

The US-based Institute of Medicine has also included patient-centric care as one of the six aims of quality. Patient-centric primary care, in particular, has been defined to include seven attributes including 'access to care, patient engagement in care, information systems, care coordination, integrated and comprehensive team care, patient centred care surveys, and publicly available information' (Davis, Schoenbaum & Audet, 2005). In the US, a 2003 Commonwealth Fund National Survey found that only one in four primary care physicians incorporate one or more of the above named patient centred attributes in their practices. Some have suggested addressing this issue with a new system of primary care reimbursement that combines monthly per diems with a traditional fee for service payment alongside new incentives for patient-centred care (Davis, Schoenbaum & Audet, 2005).

The push for patient input relies on the premise that 'far more than clinicians, patients understand the realities of their condition, the impact of disease and its treatment on their lives, and how services could be better designed to help them' (Richards, Montori, Godlee, *et al.*, 2013; Young, 2013). In other words, in order to foster patient-centric health care services, patients need to be involved in the design of these services. As a response to this growing consensus, England has seen the enactment of the Health and Social Care Act of 2012, which legally enables patients and their carers to participate in the planning and management of their health care and health care services. It also calls for the participation of the public in commissioning of health care services in order for services to reflect local needs (Anon, n.d.). This was followed by the introduction of legislation mandating basic levels of quality of care following the Francis Report in 2013, which highlights the apparent lack of compassion within some parts of the health

service and the importance of working with patients (Francis, 2013). Both the legislation and inquiry create impact in strategy and shared values, almost providing a mandate to improve the quality of care patients receive and involving them directly in these initiatives.

Patient participation has also been shown to improve outcomes, provide value for money and improve quality of life (NHS England, 2013). One systematic review on involving patients in the planning and development of health care examined reports (which were written in English) between 1966 and 2000 to determine the effects of involving patients in the planning and development of health care. Changes to services (such as appointment simplification, extending opening times, improving access for people with disabilities, improving sources of information for patients) were seen as the most impactful. Organisational changes such as the attitudes of staff were also reported as being impactful (Crawford, 2002).

The King's Fund also studied engaging patients in the design of primary healthcare services and found that patients want:

- their health care professionals to have excellent interpersonal skills, as well as demonstrable up-to-date clinical knowledge and the willingness to provide timely and meaningful information;
- easy access to their clinicians;
- clean, accessible premises;
- continuity of care and smooth handovers between health care professionals;
- opportunities to take part in their own health care;
- a sense that the best possible care will be available when they need it; and
- to understand that this care will be equitably distributed, and that public money is being used efficiently (Parsons, 2010).

Engaging patients in research such as the study conducted by the King's Fund creates a baseline for policy makers to start addressing gaps and encourages systems of care that are bespoke to the service user – in our case, the patient with SCD and/or their caregiver.

6.4.2. MODELS OF PATIENT INVOLVEMENT AND ENGAGEMENT

While research in the UK has shown that patient input can lead to change in health provider access, continuity of care and patient self-care and empowerment, patient involvement and engagement is a growing health care service trend not only in the UK but also internationally. Canada, the US and Europe have all taken substantive steps in allotting for patient input in health service design, and low income countries such as Tanzania have gone as far as implementing initiatives that engage patients in the clinical delivery of health care.

6.4.2.1. INTERNATIONAL ENGAGEMENT

In 2010, the Canadian Foundation for Healthcare Improvement (CFHI) collaborated with local and national decision makers to design and deliver affordable patient centred healthcare for Canadians. CFHI since then has supported 17 teams across Canada whose aim is to involve patients and their families in decisions about service design and delivery. The teams used multiple strategies to engage patients in this exercise including advisory councils, surveys, and workshops. The results confirmed that by improving health care provider and patient

communication, the patient experience and care process efficiency also improved. The teams were able to share their lessons learned through the dissemination of papers and webinars² (Edgman-Levitan, Brady & Howitt, 2013b).

A study conducted in a large health care delivery system in Minnesota found that the least engaged patients from the perspective of understanding one's own role in the care process (what the researchers are naming patient activation levels) had an average cost 8% higher in the base year and 21 percent higher in the first half of the next year than the costs of patients with the highest engagement levels. Patient engagement was reported as a significant predictor of cost, even after adjustment (Hibbard, Greene & Overton, 2013).

Recent regulations in the United States have also encouraged the use of patients in decision making where patients and health care providers share the decision making process based on evidence based care pathways. One study found that barriers to shared decision making included overworked physicians, insufficient provider training, and clinical information systems incapable of prompting or tracking patients. Methods identified to improve shared decision making included a recommendation for 'substantial investments in provider training, information systems, and process reengineering' (Friedberg, Van Busum, Wexler, et al., 2013).

This is similar to our findings and the intervention approach we applied for SCD. We engaged in a patient informed intervention designed through the use of a focus group and a patient questionnaire and created outputs which included a GP educational intervention, an SCD template to be installed in primary health care practices and a policy recommendation to integrate primary secondary and tertiary delivery systems in the care of patients with SCD (see Chapters 7 and 8).

Patient engagement in a more developing health care system context can also be seen as growing in priority. Some examples can be found in the World Health Organization, which has set up initiatives including one global initiative for patient health and safety engaging patients in design and delivery (WHO, <http://goo.gl/rY8lFI>). In places like Tanzania, an organisation called Upendo Disadvantaged Group recruits former tuberculosis patients who help to organise community gatherings featuring music and poetry to educate their neighbours about tuberculosis and also help in the training and administration of drug therapy (Hockstein, 2009).

6.4.2.2. ENGAGEMENT IN THE UK

Government initiatives in the UK have been developed to facilitate patient engagement in the design of health care services. Local involvement networks (LiNs) were implemented in 2008 to identify what people like and dislike about their local services and then to work with local service providers and commissioners to help make them better (Parsons, 2010). LiNs provided an opportunity to improve services by investigating specific issues of concern, holding accountable services to get results, carrying out spot checks and drafting reports and recommendations (WCC PPI Steering Group, 2008).

Today, LiNs are being taken over by local healthwatch organisations. Healthwatch organisations are similar to LiNs in many of their strengths, but differ in that they take into account not only adult experiences, but also children's user experiences; they have a statutory place on the local health boards, and they are linked to a national network (Healthwatch England, <http://bit.ly/1GYWRmv>).

The World Class Commissioning (WCC) framework is a national programme launched by the Department of Health in 2007 based upon the intent to commission best practice and to engage

² <http://goo.gl/BH7Xqy>

patients in the commissioning of best practice (Parsons, 2010). The WCC requires that Clinical Commissioning Groups (CCGs) partner with clinicians, local authorities, communities and the providers of healthcare services in order to develop quality and integrated care that are aligned with the needs of the local population (WCC Introduction Publication found at <http://bit.ly/1HcKwk6>). The NHS has also developed the Smart Guides to Engagement series for those working in or with CCGs. The Smart Guides to Engagement have been written by experts to give user friendly advice on patient engagement (NHS Networks, <http://bit.ly/1F2aKSx>).

The Quality and Outcomes Framework (QOF) is a pay for performance scheme that financially rewards NHS GPs for implementing good practice in their clinics. The QOF scheme is voluntary, but it is also one way a GP can make a difference in their income. The criteria were grouped into four areas: clinical, organisational, patient experience and additional services. Patient engagement has been raised as an additional criteria area (Parsons, 2010) (National Institute for Health and Care Excellence, <http://goo.gl/AaU2MK>).

NHS England and CCGs encourage GP practices to operate patient participation groups (PPGs). PPGs are considered a partnership between the patient, the GP and the GP practice and a forum where suggestions can be made on quality improvement measurements and the way healthcare services are delivered. The suggestions are local, relevant and have the potential of creating more cost and process efficient ways forward. While PPGs have been around since 1972 (National Association for Patient Participation, <http://goo.gl/vWtb9R>), their importance and popularity have recently increased as the NHS becomes more focused on patient centred healthcare (Parsons, 2010).

There are other programmes that have been nationally initiated but locally implemented which help patients in accessing information and creating a culture of feedback on their health care such as Patient Advice and Liaison Service (PALS). Every NHS Trust must have a PALS, which is a service focused focal point (team or individual) who help patients solve problems, provide advice and information. PALS are also a good resource when it comes to understanding trends locally and can support in patient engagement activities. There is also a national system for responding to complaints and all NHS organisations are required to comply with the majority of organisations having a local and dedicated complaints team to both respond to complaints and monitor their trends (Renedo & Marston, 2011).

Several instruments have been used to measure the use of patient engagement and its impact including surveys, appraisal of GPs, and performance management reward structures. Some challenges to measuring patient engagement in primary care include knowing whether the samples being measured are representative and unbiased, what to do with the results of measuring patient engagement once they are collected, and the ability to benchmark the results against other practices. One of the issues is that without a proper framework or policy guideline on how patients should be engaged in service development, there is little in the way of creating sustainable impact (Shared decision making between clinicians, health policy makers and patients is important in the design of health services, but how patient involvement is defined and the mechanism for involvement is not always clear and has been a source of debate between patients and health care providers (Renedo & Marston, 2011)).

6.5. KEY POINTS FROM CHAPTER 6

1. Patient engagement in the design of health services has been shown to improve outcomes, provide value for money and improve patient satisfaction.
2. Focus groups are a means of engaging patients in satisfaction and design of services.
3. Patient questionnaires are a successful tool in measuring patient satisfaction.

7. POLICY IMPLICATIONS

7.1. SUMMARY OF FINDINGS

With the help of six studies, I was able to show SCD admission rates in England, the readmission rates, local admissions, costs associated with admission and patient perspectives in both care and prevention. Because of this work, there is a clearer picture in the trends and health care utilization of patients with SCD in England.

Using HES data, trends for SCD hospital admissions in England showed a rise in 50% of hospital admissions over a 10 year period. The most deprived areas had a higher rate of readmission and in-patient mortality among those with SCD. Adolescents had a higher rate of readmission possibly identifying a gap in health care access. Local findings in a high prevalence area showed that the majority of admissions were for short length of stay and 74% accounted for multiple admissions while the patient focus group and questionnaire both identified potential gaps in care and prevention.

Laying down these results allows us to look into the broader literature of chronic disease management and thereafter make policy recommendations bespoke to SCD that may help to reduce hospital admissions due to SCD.

7.2. CHRONIC DISEASE PREVALENCE IN ENGLAND

The Department of Health estimates that 17.5 million adults in the UK live with at least one chronic disease, and many of these people will experience more than one chronic condition at the same time (Department of Health, 2005). Chronic diseases account for 50% of general practice consultations and 70% of inpatient bed days per year within the UK (Langer, Chew-Graham, Hunter, et al., 2013; Department of Health, 2010). Two thirds of patients admitted as medical emergencies have an 'exacerbation of chronic disease' (Health, n.d.). They constitute a major challenge for healthcare services in general, and unscheduled care in particular (Langer, Chew-Graham, Hunter, et al., 2013).

'We want to give patients the most intensive care in the least intensive setting' (Department of Health, 2005).

Many interventions aimed at managing and preventing chronic disease are delivered in the primary and community care setting. Improving care and services for people with such conditions are also seen to have a beneficial impact on secondary and emergency care (Health, n.d.).

7.3. CHRONIC DISEASE MANAGEMENT FRAMEWORKS

Improving Care for People with Long-Term Conditions, a review of UK and international frameworks, identified four broad frameworks for chronic disease management: (1) the Chronic Care Model, (2) the Innovative Care for Chronic Conditions Model, (3) the Public Health Model, and (4) the Continuity of Care Model. ((Singh & Ham, 2006)).

The Chronic Care Model is the best known framework and suggests six elements to improve initiatives in chronic care: community resources, the healthcare system, patient self-

management, decision support, delivery system redesign, and clinical information systems. The report states that ‘while there is evidence that single or multiple components of the Chronic Care Model can improve quality of care, clinical outcomes, and healthcare resource use, it remains unclear whether all components of the model, and the conceptualisation of the model itself, is essential for improving chronic care.’ (Singh & Ham, 2006).

The Innovative Care for Chronic Conditions Model focuses on ‘improving care at three different levels: micro level (individual and family), meso level (healthcare organisation and community), and macro level (policy)’ (Singh & Ham, 2006).

The Public Health Model is a less-known version and focuses on a system wide perspective which ‘emphasises the determinants of disease as well as social, cultural, and economic factors’ that could affect quality and quantity of care (Singh & Ham, 2006).

The Continuity of Care Model outlines ‘how chronic conditions develop in response to risk factors in the community and suggests points at which to target prevention efforts, medical intervention, treatment, rehabilitation, and palliative care’ (Singh & Ham, 2006).

Based upon the mentioned frameworks and the models that have been developed which focus on how to deliver services, there is growing evidence that some of the essential components of good chronic disease management include:

- Using information systems to access key data on individuals and populations;
- Identifying patients with chronic disease;
- Involving patients in their own care (self-care);
- Coordinating care;
- Integrating care; and
- Aiming to minimise emergency visits and hospital admissions (Health, n.d.).

7.4. PATIENT ACCESS TO HEALTH INFORMATION IN PROMOTING CHRONIC DISEASE MANAGEMENT

Facilitating access to health information can ‘promote patient engagement in care’ (Woods, Schwartz, Tuepker, *et al.*, 2013). As a response, many organisations are creating programmes that enable their patients to access and download their own medical records. The Blue Button program, established by the US Veterans Administration, has branded a recognisable prompt on websites that allows electronic access to health information³. At the Mayo Clinic, downloading a smartphone application gives patients full access to their medical notes, pathology and radiology and is striving to deliver it in layman language (Richards, 2013). Other organisations allow patients to create personal health records that enable patients to create their own measures and share them with their providers. One such programme is called True Colours⁴, which is reported as being a ‘self-monitoring and management system’ that uses simple technology accessible to the patient such as a mobile phone or computer device and turns quantitative responses from patients into easy to read graphs for patient or provider use. Patients even have the option of creating their own questions and responses which may be unique to them. This programme was initially developed for mental health disorders but now has been expanded to include chronic



3

4 <http://bit.ly/1lwSGm8>

diseases such as diabetes and arthritis, which shows that the programme can be easily adaptable and transferrable to other disease categories (Edgman-Levitan, Brady & Howitt, 2013a).

One CLAHRC initiative was the creation of the 'My Medication Passport'⁵, which allows patients to carry a portable record of their medications and thereby improves communication across different care settings. Because it was originally developed for the elderly, it comes in both paper form and as a smartphone application (Edgman-Levitan, Brady & Howitt, 2013a). This application would be easily transferrable to patients with SCD, especially as part of their pain management protocol.

7.5. PRIMARY CARE INTERVENTION AS A MODEL OF CHRONIC DISEASE MANAGEMENT

A study by the BME Health Forum reported that when people with long-term conditions were asked why they went to emergency before trying to go to their GP first, 67% answered because they 'would not be seen quickly enough'. Similarly, when they were asked what change would make them go to the GP rather than the emergency, 59% of the respondents suggested same-day appointments, which according to the study 'suggests that changes in how quickly people could be seen in primary care would have the biggest impact in terms of reducing[emergency] attendances.' (BME Health Forum, 2014). Both our focus group study and our patient questionnaire found similar results with SCD patients. Another study showed that NHS hospital emergency department visits increased by 20% between 2007-2008 and between 2011- 2012 (Cowling, Cecil, Soljak, *et al.*, 2013), with a similar finding that (some) patients in the emergency attributed their visit to the inability to see a GP (Cowling, Cecil, Soljak, *et al.*, 2013).

A recently published study conducted an analysis that estimated the number of emergency department attendances in England during 2012-2013 which were preceded by the patient being unable to access an appointment at their general practice. The study found that approximately 5.8 million emergency department attendances were preceded by the patient being unable to obtain either an appointment or a convenient appointment. This accounted for approximately 27% of unplanned emergency attendances in England for 2012-2013, suggesting that a large number of emergency attendances each year are likely to be preceded by 'unsuccessful attempts to obtain convenient general practice appointments' (Cowling, Harris, Watt, *et al.*, 2014).

Such studies have triggered a response from the UK Department of Health (DH). In April 2014, the DH launched a scheme including 1147 general practices in England and 7.5 million registered patients offering appointments from 8:00-20:00, seven days a week (outside of the current GP opening hours), with priority to the most vulnerable populations (Department of Health. Millions to benefit from improved GP care [press release]⁶).

Studies have also found that the GPs who did provide more timely access to primary care had 'fewer self-referred discharged [emergency] visits per registered patient' (Cowling, Cecil, Soljak, *et al.*, 2013). Furthermore, those admissions with conditions that may have escalated over time (as is often seen in chronic and long-term conditions) and with the potential to be intercepted in primary care could also indicate 'suboptimal care (because the individual's health had deteriorated avoidably to the extent that hospitalisation was necessary)' (Blunt, n.d.). Hence, while access to primary care is a recurring challenge that may explain some unnecessary hospital visits and is repeatedly identified in the literature, we can also see that relocating some hospital services to primary care will not only reduce unnecessary hospital admissions

⁵ <http://goo.gl/Pk7mXB>

⁶ <https://goo.gl/DeIcTQ>

(Cowling, Cecil, Soljak, *et al.*, 2013), but may actually improve the equity (Sibbald, McDonald & Roland, 2007) and quality of services (Blunt, n.d.).

The intent is not to replace primary care health services for secondary health care services, but rather to allow secondary care to supplement primary care instead of the reverse.

Primary care is a major aspect of England's NHS, and responsible for 'approximately 300 million consultations per year with GPs in England, which represents 70-90% of all patient contacts with the NHS' (Gibbons, Bindman, Soljak, *et al.*, 2012).

Another necessary step is to encourage policy makers to link primary care and public health. Primary care is defined as 'providing medical services to individual patients with immediate health needs', and public health is defined as 'offering a broader array of services across communities and populations that collectively will help people to be healthy.' This union of primary care and public health can begin to offer a more holistic approach to the management of diseases that are considered chronic or long term (Primary Care and Public Health: Exploring Integration to Improve Population Health, released 28 March, 2012, Institute of Medicine).

7.6. SUPPORT FOR SELF-CARE FOR PATIENTS WITH CHRONIC DISEASE

For patients with long-term or chronic disease, there is a growing interest in self-care programmes that emphasise the patient's role in managing their illness. Policies and interventions that strengthen the role of the patient in managing their health care not only contribute to improved outcomes (Hibbard & Greene, 2013), but can also save the NHS substantial amounts of money. A 2013 study by Nesta estimated that £4.4 billion could be saved in the NHS through greater participation and self-management of long term conditions (Richards, 2013; Anon, n.d.).

An article in the BMJ suggests that support for self-care has the potential to improve the management of long-term conditions. Furthermore,

- Current interventions have only modest effects on patient health and use of health services;
- One possible explanation is that interventions tend to focus solely on the patient;
- Interventions need to reflect the ways in which patients and professionals respond to long-term illness; and
- Approaches need to target patients, professionals, and healthcare organisations (Kennedy, Rogers & Bower, 2007).

Supporting patient self-management is about 'acknowledging their central role in managing their own care and empowering them, their family and carers to handle their condition as effectively as possible' (Department of Health, 2005). This means ensuring the appropriate skills and knowledge to understand how to best handle their condition, providing information that is easily accessible, enabling and empowering patients and carers to manage the condition more effectively such as by implementing self-monitoring or applications that prompt or remind the patient when they should be doing something, providing a trusted and consistent person to contact and ensuring community network support or a patient mentor (Department of Health, 2005). There are several disease specific programmes currently being implemented such as the Diabetes Education and Self-Management for Ongoing and Newly Diagnosed (DESMOND) programme which focus on 'enhancing the patients skills and confidence in managing symptoms and medication' (Department of Health, 2005). Similar programmes could be implemented for a disease such as SCD.

One study measured the effects of a self-management programme on patients with chronic disease through a seven-week intervention attended by people with different chronic conditions; the authors reported that, after one year, the participants in the programme experienced a statistically significant improvement in health behaviour and had fewer visits to the emergency department. Although not statistically significant, they also found fewer outpatient visits to physicians and fewer days in hospital (Lorig, Sobel, Ritter, et al., 2001).

Another study created an intervention for a group of patients with a mix of chronic diseases, which included patients with comorbidities, and found that a chronic disease self- management programme was both feasible and beneficial with respect to improving health behaviour and status and resulted in few hospitalisations and days of hospitalisation (Lorig, Sobel & Stewart, 1999).

The Stanford School of Medicine has developed different chronic disease self- management programmes, including online programmes⁷ (Lorig & Holman, 2003). The School of Medicine engages patients in developing the self-management programmes in order to enhance the impact and sustainability of the programmes themselves. Topics include both physical and emotional aspects. Some focus on medication use, others on effective communication or nutrition. Those who complete the six-week course experience ‘fewer hospitalisations and report improved general health.’ Organisations in 22 countries have now been licensed to offer the programme (Edgman-Levitan, Brady & Howitt, 2013a).

The above programmes, along with research and policy reports are substantially leaning toward a holistic focus that supports patients and their carers to become more functional, independent and resilient. They encourage patients to manage their condition in the home environment as ‘preferable to a purely clinical focus on managing or treating medical symptoms’ (Goodwin, Sonola & Thiel, 2013).

7.7. MANAGING SCD AS A CHRONIC DISEASE

Historically, the management of SCD, particularly when it comes to pain management, has been through an acute care model. For the most part, SCD is still being treated under an acute care model (Maxwell, Streetly & Bevan, 1999b), despite studies that show the essential components of chronic disease management such as self-care (Jenerette & Murdaugh, 2008; Jenerette, Brewer & Leak, 2011), coordinated and integrated care (Mitchell, Lemanek, Palermo, *et al.*, 2007), and the use of technology (McClellan, Schatz, Puffer, *et al.*, 2009), all of which have successfully been used in the management of SCD. Additional evidence also suggests that patients with SCD who manage their disease at SCD-specific care clinics have a lower rate of emergency room visits and inpatient admissions than those who do not (Yang, Shah, Watson, *et al.*, 1995).

Self-care for SCD

One study on self-management models for sickle cell disease indicated the benefits of expanding self-management resources to improve health outcomes of sickle cell patients (Jenerette & Murdaugh, 2008). Another study further supports this by stating that ‘the current structure of pain management placed patients too dependent on the hospital setting for treatment, while a more empowering and self-management approach could be more beneficial’ (Maxwell, Streetly & Bevan, 1999b).

⁷ <http://goo.gl/QXgpWU>

Examining popular self-management programmes for chronic disease such as the one at Stanford Medical School can also benefit SCD self-management programmes, since the underlying objectives are similar. However, care needs to be taken in selecting the appropriate model. For example, the Expert Patient Programme⁸ is an NHS sponsored programme in the UK based on the Stanford model and is considered a major public health initiative. The self-management course is taught by laypersons who mostly suffer from chronic disease themselves.

The programme has received considerable promotion and positive press, but there are critics that argue that laypersons are not the best option for teaching self-management. Instead of creating efficiency and lowering costs, as touted by the programme, it could yield more frequent consultations with providers and higher associated costs (Griffiths, Foster, Ramsay, *et al.*, 2007).

SCD Interface

Health information technology is becoming more important in the management of SCD and its assessment. One study piloted an interactive web-based tool for SCD in paediatrics. The Take-Charge Programme assessed adherence to clinic visits and drug protocol along with their barriers and found that the tool offered an innovative approach to integrate newer technology to assess adherence and clinical outcomes for paediatric patients with SCD (Crosby, Barach, McGrady, *et al.*, 2012). Another study used a web-based assessment tool to determine barriers to SCD clinic attendance and to build problem-solving strategies around these barriers (Modi, Crosby, Hines, *et al.*, 2012).

In our study, the issues identified during the focus group discussion and the pilot questionnaires helped define key areas in need of improvement for SCD management. Some of the key themes and useful suggestions were used to develop a GP education intervention to improve GP management of SCD in primary care, and part of this education intervention was facilitated by developing an SCD template for data entry compatible with the main electronic patient record systems used by GPs in the UK. These electronic medical record systems are now widespread in UK primary care and have helped improve recording on patients with other chronic diseases (Majeed, 2004). In the longer term, these electronic patient records could be linked to disease recording templates to help improve the recording of information by primary care teams and the quality of primary care received by patients (Pappas, Anandan, Liu, *et al.*, 2011).

The SCD specific template⁹ was installed in 17 general practices across Brent with the intent to follow up with wider roll-out across northwest London. The template allowed GPs to register important aspects of management and co-morbidities associated with SCD. The template has the potential to ensure that key information about patients with SCD is recorded on their electronic medical record and GPs are better able to monitor the management of these patients both from the perspective of preventive care as well as managing comorbidities such as hypertension and kidney disease often found in SCD patients. The template can also ensure that the clinical investigations and treatment provided by hospital specialists are recorded. This can help improve communication with specialists and lead to the coordination of care of SCD patients.

⁸ <http://bit.ly/1J5RDdt> (accessed 2nd July 2014)

⁹ A screen shot of the template is available in the Appendix.

Smartphone applications or the use of wireless technology to implement pain management protocols have significant potential to improve symptom monitoring and improve communication between SCD patients and their healthcare providers (McClellan, Schatz, Puffer, et al., 2009). A recent search for SCD-related smartphone applications returned 10 results which included three games, one SCD resource locator, a newsletter application, an application that identified SCD-related conferences, a pain crisis application, and two others that appeared to be related more to other blood disorders such as haemophilia; one application is available that has been developed by Imperial College London called SiKL¹⁰. One of the areas that has previously been explored with SCD and patient access to health information is a patient-held record (attached as Appendix 2). The idea is that a patient-held record would enable patients with SCD to communicate better about their disease with health care professionals for prompt and effective treatment.

The SiKL smartphone application has been developed as a modern interpretation of the patient-held record¹¹. An electronic health records access programme could be another way to modernise this previously raised study area.

Primary Care Intervention and Sickle Cell as a Chronic Disease

Transferring what we know works for other chronic diseases, such as improving quality at the primary care level, may also increase the effectiveness and cost-efficiency of health care outcomes in SCD. However, at times, cost-efficiency may not be immediately evident. For example, many at-risk pregnant women and fathers are offered SCD screening too late in pregnancy to allow for reproductive choice. One study compared the costs of three screening approaches: 1) primary care screening with a test offered to the mother and father together, 2) primary care screening with a test offered to the mother and then the father only if the mother is a carrier, and 3) screening at the first midwife consultation. The study showed that screening at primary care had a higher cost per screening of £13, but it did not assess policy wise whether the higher cost per screening had a value of at least £13 GBP suggested a further study to investigate this point (Bryan, Dormandy, Roberts, *et al.*, 2011).

As stated previously, the National Confidential Enquiry into Patient Outcome and Death *A Sickle Crisis?* report criticised the lack of primary care involvement in healthcare for SCD while another study claimed that patients are being cared for by 'primary care providers with limited experience' (Raphael & Oyeku, 2013). We wanted to test a local intervention that could actively addresses the quality of primary care received by patients with SCD living in a high prevalence community such as northwest London by engaging both primary care providers and patients.

FIGURE 13: SiKL APP

Source: <http://bit.ly/1EtzVLt>



¹⁰ Apple Store search conducted on 28 May 2014.

¹¹ <http://apple.co/1F95bTM>

We had the following objectives:

- To improve management of SCD at the primary care level;
- To better educate GPs on SCD;
- To raise awareness about national SCD standards and guidelines;
- To increase GP confidence in managing SCD;
- To increase patient confidence in GPs with regards to SCD knowledge; and
- To increase patient satisfaction and well-being.

We conducted a three-month GP educational intervention as a pilot initiative. This intervention was designed based upon the two preliminary exercises discussed earlier – the focus group study and the patient questionnaire; both outputs informed the intervention.

The intervention was conducted with a group of GP practices in northwest London that have come together and refer to themselves as the Harness Group. The number of practices in the group is 15, plus one GP access centre¹². There are approximately 20 full-time and 9 part-time GPs and an estimated 50 registered patients with SCD within the Harness Group. This was our study group (Aljuburi, Okoye, Majeed, *et al.*, 2012).

The number of practices in northwest London that are not part of the Harness Group is 55.

There are approximately 122 full-time and 61 part-time GPs and an estimated 155 registered patients with SCD in the non-Harness group. The non-Harness Group of GP practices did not receive the intervention. This was our control group (Aljuburi, Okoye, Majeed, *et al.*, 2012).

A patient satisfaction survey of patients with SCD and a GP Questionnaire were used as tools to design the intervention (attached as Appendix 3). The patient satisfaction survey was conducted during a three-month *pre* and three month *post* intervention period. Patients and GPs were meant to complete the questionnaire twice to reveal any change in responses pre- and post-intervention. In the end, only the patient questionnaire was used. The survey was administered one-on-one by a researcher. The survey took approximately twenty minutes to complete. For outpatients, the researcher was the clinic nurse. For patients surveyed via support groups and for the targeted Harness Group patients, the researcher was the patient representative for that community (Aljuburi, Okoye, Majeed, *et al.*, 2012).

The intervention was based on two things:

1. Clinical educational sessions for GPs that were conducted over three months. The first was piloted on 2 December 2010 and further sessions were held on 13 January 2011 and 11 February 2011.
2. An education package on SCD management that was made available to the GPs.

Outcome

The desired outcome was to identify if there are any changes (expected improvements) in patient satisfaction, to identify whether GPs have made any changes in the way they manage SCD pre- and post-intervention, and whether they feel more confidence in managing SCD post-intervention.

¹² 1. Church End Medical Centre 2. Buckingham Road Surgery; 3. The Stonebridge Practice 4. Harness Harlesden Practice 5. Aksyr Medical Practice 6. Brentfield Medical Centre 7. Church Lane Surgery 8. Freuchen Medical Centre 9. Oxgate Gardens Surgery 10. Park Road Surgery 11. Harrow Road Practice 12. Hilltop Medical Practice 13. Acton Lane Surgery 14. Chaplin Road Surgery 15. Wembley Park Medical Centre and 16. Brent GP Access Centre

FIGURE 14: GP INTERVENTION CHART

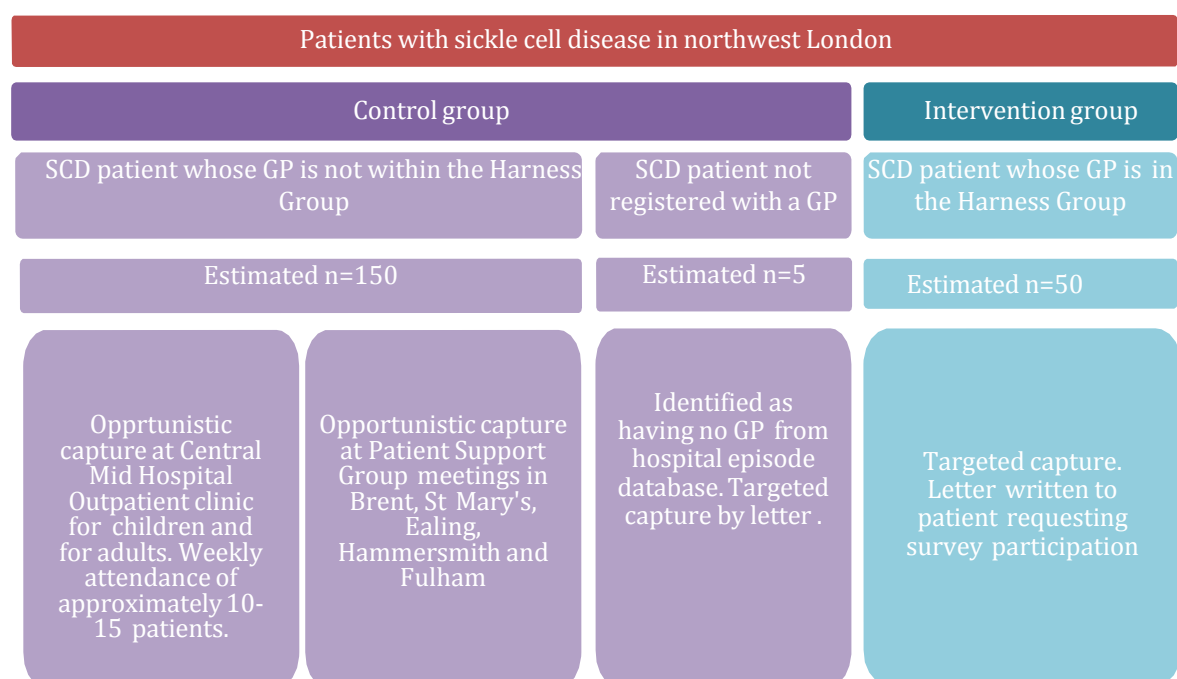


Chart Source: CLAHRC Study Design

Because the questionnaires were administered one-on-one, we were able to collect sufficient feedback and saw remarkable success in designing an intervention, i.e. the GP Educational Packet. The GP Educational Packet is attached as Appendix 4. With respect to identifying if there were any improvements in patient satisfaction or identifying whether GPs had made any changes in the way they manage SCD pre- and post-intervention and whether they felt more confidence in managing SCD post-intervention, we did not feel like we had sufficient participation to make any link between the intervention and patient satisfaction or to link the intervention and improved GP management of SCD. However, we were able with confidence to continue making available the educational intervention for GPs to engage at their option (Aljuburi, Okoye, Majeed, et al., 2012).

Through the implementation of GP educational intervention, we also learned that auditing and improving communication between different health care providers is also necessary. This could be facilitated by GPs receiving timely information on hospital admission and hospitals advising patients to book an appointment with their GP for review after discharge. Receiving proactive care from the GP could also be achieved by identifying areas with a relatively high prevalence of SCD and designing a local quality improvement scheme. Many patients with chronic illnesses do receive proactive care from their GPs; these are generally patients with diseases covered in the GP Quality and Outcomes Framework (HSCIC, 2013). At present, SCD is not a part of this framework, but could be considered for future inclusion in areas where there is a high prevalence of SCD (Aljuburi, Phekoo, Okoye, et al., 2013b).

Community Approach to SCD

A 'community hub' has also been proposed for SCD. The community hub can be defined as a patient centred approach which integrates non-clinical determinants of public health that can drive an innovative and collaborative solution to SCD challenges. The SCD community hub proposes creating a centre where all non-clinical services that could support sickle cell patients are centralised. Local members of the community 'should be seen as integral to the care-giving

process'. Community awareness and trust 'promotes legitimacy and engagement which can provide an essential resource within care coordination programmes' (Goodwin, Sonola & Thiel, 2013). The SCD community hub is one such community coordinated programme. It was first introduced by the Sickle Cell Society's Comprehensive Care Project. The SCD community hub supports an integrated sickle care pathway that incorporates mental, social, acute, and primary care needs. The view is that an all-encompassing model may increase user satisfaction and could assist in reducing unnecessary admissions to hospitals (Pizzo, Laverty, Phekoo, et al., 2014).

7.8. TRANSLATING RESEARCH INTO POLICY

The first part of this chapter has shown that many interventions aimed at managing and preventing chronic disease are delivered in the primary and community care setting, and that SCD should be included. It has also shown us that the use of information systems can be a critical tool in the support of SCD management, policies and interventions aimed at strengthening the role of patients in managing their own care (self-care) also contribute to improved outcomes. SCD should be included in such interventions and policies. These interventions are activated through policy reform.

The historical review of health policy reform in England shows a long and quite protracted approach. Both those proposing and opposing health reforms have put forward equally selective interpretations of history. These arguments often pose significant barriers to change or objectivity in health policy making, and reduce the space for policy innovations. Some of the health policies that pre-dated the NHS were effective in tackling health inequalities, and contemporary policy makers should not let public wariness of this imagined past blind them to its potential to inform policy today (Hayes, 2013).

In order for a change in health policy to have an impact and to be sustainable, such a change demands the cooperation and collaboration of governments, health authorities, and patients. A strategy with goals and objectives needs to be agreed with clear actions for implementation at the national, regional, and local levels. Regardless as to whether the decision is to improve quality, to reduce cost, or to engage patients, the process to affect that change has to have a holistic approach that assesses and intervenes at different entry points within the health care delivery system. Furthermore, local experiences should inform national strategies. This can be said to influence any health policy change, but in this case we are speaking specifically to SCD.

Policies specific to SCD in England have been few and far between. The NHS Sickle Cell and Thalassaemia Screening Programme was set up in 2001 in order to introduce an effective antenatal and neonatal screening programme for haemoglobinopathies (UK National Screening Committee, 2006). The programme gave way to universal neonatal screening for SCD which was introduced in England in 2006, with a primary aim of reducing mortality due to pneumococcal sepsis in the first few years of life by introducing antibiotic prophylaxis (Dick, 2008). National implementation of newborn screening in England has increased the number of children (Green, Aljuburi, Majeed, et al., 2012a) identified with SCD, and in many areas it has doubled the disease caseload. England is the first country in Western Europe to introduce a national screening programme (Streetly, Clarke, Downing, *et al.*, 2008). It now serves as model for other countries, including those in Africa where the WHO is supporting implementing newborn screening. Those who are not identified by newborn screenings are picked up through their GP or through hospital admission due to co-morbidity diagnoses (Streetly, Latinovic, Hall, *et al.*, 2009).

7.9. THREE POLICY RECOMMENDATIONS TO REDUCE HOSPITAL ADMISSIONS DUE TO SCD

The ability to control the rate of emergency admissions is a common indicator of success for a health system, particularly for conditions where ‘preventive management is possible in the community’ (Blunt, n.d.). Accordingly, this is achievable for SCD through three policy objectives that summarise our findings: the need to integrate and coordinate SCD management across health care sectors, the need to engage and educate primary care physicians on SCD, and the need to strengthen targeted and community based solutions for SCD (Figure 14).

TABLE 17: THREE POLICY OBJECTIVES

Integrate and coordinate SCD management across health care sectors

Engage and educate primary care physicians on SCD

Create and strengthen targeted and community based solutions for SCD

Chronic diseases require us to move away from acute care and reorient towards ‘prevention, self-care, more consistent standards of primary care, and care that is well coordinated and integrated.’ (Anon, n.d.) Hence, our policy approach to SCD should be multi- dimensional, in the same way that our clinical approach to a chronic disease is multi-dimensional. Policies for SCD should be not only coordinated and integrated but also commissioned and targeted. Previous studies have shown successful interventions that moved the management of SCD, particularly uncomplicated sickle cell pain episodes, from tertiary to primary care (Coleman, Irons & Nicholl, 2001; Benjamin, Swinson & Nagel, 2000). Our policy focus should continue to emphasise the importance of primary care in managing SCD, keeping in mind this multi-pronged approach.

We have been able to show that SCD is a chronic disease with a high prevalence in some parts of England, with a prevalence and cost impact warranting attention to policy. Patients with SCD who are admitted to emergency departments account for the majority of health care expenses associated with the disease (Carroll, Haywood, Fagan, et al., 2009; Pizzo, Lavery, Phekoo, et al., 2014). In 2010-2011, England had 6,077 admissions associated with SCD with crisis as a primary diagnosis. The total cost for these admissions was £18.8 million, of which approximately £17 million (91%) was for emergency admissions (Pizzo, Lavery, Phekoo, et al., 2014). In addition to the cost analysis study, both the study on admission trends using HES data and the NHS Brent local study suggest that the SCD admissions with a short length of stay and multiple admissions could be potentially avoidable hospital admissions (Aljuburi, Lavery, Green, et al., 2012a; Green, Aljuburi, Majeed, et al., 2012a).

Hospital readmission rates in England have been used as a measure of poor healthcare performance at a local level or when making comparisons over time between hospitals (Mason, Daly & Goldacre, 2000). However, there is a need to better understand the different factors that influence readmission rates for any disease-specific category (including SCD) to control for extenuating factors that would impact on readmissions by allowing a more thorough assessment of healthcare outcomes as well as more accurately defining potentially preventable readmissions (Nolte, Roland, Guthrie, et al., 2012). Our study shows that SCD patients coming from the most deprived areas nationally experience higher rates of SCD hospital readmissions as well as inpatient mortality related to SCD, suggesting that there may be inequalities in access to health care and health outcomes amongst people with SCD. This continued to be true after adjustment for age and comorbidities between these populations, which might commonly be thought to contribute to these differences in readmission rates (data not shown). Also, the age and comorbidity breakdowns were similar between the least and most deprived groups (data

not shown) (Aljuburi, Laverty, Green, et al., 2013a). All this suggests that better integration, coordination, engagement of both patient and provider, and targeted community based solutions could mitigate the challenges faced by the health system from SCD.

7.9.1. **OBJECTIVE 1: INTEGRATE AND COORDINATE SCD DISEASE MANAGEMENT ACROSS HEALTH CARE SECTORS**

There are no universal definitions for coordinated care or integrated care and, in fact, these terms are often used interchangeably, although the consensus seems to be that co-ordinated care refers specifically to a patient's clinical coordination of care whereas integrated care refers more to the system of health care organisation (Anon, n.d.). For our purposes, we will infer that coordination of care refers to the patient and integration of care refers to the system.

There is evidence that SCD admissions are increasing as an effect of readmissions of patients with SCD and that effective discharge planning with a link between hospitals, primary and community care might help prevent readmissions (Ballas & Lusardi, 2005a; Ballas & Lusardi, 2005a; Aljuburi, Phekoo, Okoye, et al., 2012).

According to the NICE guidelines, better coordination of SCD care, including training of health care professionals, pain management, administration of analgesia, and patient monitoring, may yield better results and reduce length of hospital stay (Anon, 2012).

In addition, one of the observations of the focus group that we held was that SCD care was neither coordinated nor integrated. The questionnaire pilot study also reinforced this observation (Aljuburi, Phekoo, Okoye, et al., 2012; Aljuburi, Okoye, Majeed, et al., 2012).

[There should be] review letters from the hospital. They [general practitioners] should see how you are doing. That will be good. [following discharge from hospital] why don't they [GPs] ever call the patient and follow-up?

(Aljuburi, Phekoo, Okoye, et al., 2013a)

One of the integrated and coordinated approaches to SCD care in the US that has recently received attention is the patient-centred medical home (PCMH). It is being touted as the 'cornerstone of efforts to reform chronic disease management in the US health care system and to transform primary care into a centrepiece for improving health care quality' (Raphael & Oyeku, 2013; Landon, Gill, Antonelli, et al., 2010). The PCMH functions like a multidisciplinary team of providers through the use of primary care that is focused on the specific patient's needs to ensure integration and coordination. PCMH implementation has shown improvements in quality and efficiency for both adults and children and has been advocated as the model to follow for every patient with SCD (Raphael & Oyeku, 2013).

A study conducted by the Hemoglobinopathy Learning Collaborative (HLC) in the US identified five key drivers of improved outcomes; included amongst them were reliable identification and follow-up and seamless co-management between primary and specialty care. The data was collected via a web-based system linked to a reporting portal. Participants included community organisations and primary and specialty care providers (Oyeku, Wang, Scoville, et al., 2012).

Coordination and integration of care for SCD can also follow successful pilot projects for other chronic diseases. Wales has initiated a shared care model for people with multiple chronic illnesses which coordinates care between primary, secondary, and social care. A reduction in emergency admissions for chronic illness of 27% was found between 2007 and 2009, which resulted in a cost savings of £2,224,201 (Goodwin, Sonola & Thiel, 2013).

Other international models using primary care as a basis to integrating and coordinating care could be studied as adaptable models. In Denmark, primary care practices are what policy makers would consider patient-centred. The reimbursement is a mix of per diem and fee for service. Primary care physicians are paid monthly to retain the patient in the practice and then through government negotiated fee schedules for additional individual services. They have electronic prescribing systems connected to local pharmacies and an off-hours service which includes a telephone service that is operated by physicians who have access to the patient's health registry information. The physicians providing off-hours services are paid for the telephone consultations and can either manage the problem by phone, fill a prescription electronically or ask the patient to see a physician in the clinic. After the telephone consultation, an email is sent to the patient's registered GP updating them of any off-hours services and facilitating any necessary handover to the GP (Davis, Schoenbaum & Audet, 2005).

There are some organisational and policy barriers as well as critics of integrated and coordinated care who claim that without a robust shared electronic patient record, integration is challenged. Integrated care is usually implemented through short-term pilot programmes, so there may be a lack of sustainability; currently, there is no single outcome framework that creates joint accountability. In order to address such barriers, one solution is a programme approach to a specific population group that includes good access to extended primary care services, supporting health promotion and primary prevention, and co-ordinating community-based packages for rehabilitation, re-enablement, and independent living (Ham, Imison, Goodwin, et al., 2011).

National Voices, a national coalition of health and social care charities in England suggested that commissioners track patients across the continuum of care (Goodwin, Smith, Davies, et al., 2012). A King's Fund study suggested that a significant investment in primary and community care services is needed in order to achieve well-coordinated care where general practices act as the hub of a 'wider system of care that takes direct responsibility for coordinating and signposting individuals to services within the NHS as well as beyond healthcare on a 24/7 basis' (Goodwin, Smith, Davies, et al., 2012). A Nuffield Trust analysis suggested encouraging integrated care through payment incentives (Bardsley, Smith & Car, 2013).

Another option is to use an accountable care model where providers are held jointly accountable for delivering outcomes for a certain population over a certain time at an agreed cost (McClellan, Kent, Beales, et al., 2013). The idea of moving from a supply-led health care system to a demand-led health care system speaks to the core of the accountable care model (McClellan, Kent, Beales, et al., 2013).

Five components deliver accountable care:

1. Jointly accountable designated population (such as an SCD population within a borough);
2. Outcomes that are targeted and respond to the patients' needs;
3. A measuring tool to monitor the performance of outcomes and to learn from the variation;
4. Payments and incentives aligned with target outcomes; and
5. Co-ordinated delivery across a range of providers organised in a way to achieve the desired outcome mentioned in the second point above (McClellan, Kent, Beales, *et al.*, 2013).

Accountable care models have been used successfully in emerging and even developing health care systems. In Singapore and the United States, these models have been used for the elderly, for people with diabetes, and for other chronic disease populations. Because reimbursement is

linked to patient outcomes, cost savings can be realised eventually, but what is realised more immediately is the quality and value for money in the actual care received. The approach also encourages patient engagement. The approach of accountable care takes a broader perspective than illness. Outcomes expand beyond the hospital and include primary and community care, public health and social and behavioural care. For policy makers, 'this may mean working across funding streams, agreeing on key outcomes, creating mechanisms to link datasets, and pushing for data transparency' (McClellan, Kent, Beales, et al., 2013). In other words, goals should be to adjust pay mechanisms to reward outcomes, collaborate across multiple providers, and enable information systems that allow for multiple providers and patients to share data.

Bundled care payments have seen success in dealing with patients with specific diseases such as diabetes. A particular episode of care is paid as a bundle across a range of services for that episode of care. This is particularly relevant for long-term or chronic diseases (Bloom, Graf, Anderer, et al., 2010).

The overlapping idea behind all of these approaches of care is the alignment of services and the providers of those services. There is no point in looking at the individual areas if the areas themselves are unable to form a full map of care.

Policy Recommendation: Coordinate and integrate care across primary secondary and tertiary care for better management of SCD.

7.9.2. **OBJECTIVE 2: ENGAGE AND EDUCATE PRIMARY CARE PHYSICIANS ON SCD**

If I take my son to hospital now, the doctors like to have a relationship and know how he is doing. If I take him to the GP there is no relationship; he is a stranger.

[You have a] five minute conversation with the GP. You are in and out. I prefer to go to the hospital, I know them there. I am safe. My nurse knows me. I know my GP, but does he really know me? I go to the hospital. They know me. (Aljuburi, Phekoo, Okoye, et al., 2012)

Our study assessing the views of patients with SCD showed that patients and carers often bypass their general practitioner for acute problems and head straight for the emergency department, suggesting that better primary care interventions could reduce emergency department attendances and emergency admissions (Aljuburi, Phekoo, Okoye, et al., 2012; Aljuburi, Okoye, Majeed, et al., 2012). A study in the East Midlands looked at 145 general practices and showed that as the proportion of patients able to consult their primary care physician increased, emergency admission rates decreased (Bankart, Baker, Rashid, et al., 2011). Another study conducted in London showed that overnight admissions decreased as a result of better pain management, use of analgesia, transfusions, community support, and improved education at home (Day, Thein, Drasar, et al.,

2011). Moving certain treatments from the hospital and into a primary, community, or home setting could help curb unnecessary hospital admissions. Elective transfusions, for example, are an essential treatment in SCD and can now be provided in an ambulatory setting instead of admitting the patients in hospital to receive the treatment (Day, Thein, Drasar, et al., 2011). Another study emphasised the need for 'a preventative and comprehensive model of care in addition to care management' (Kanter & Kruse-Jarres, 2013). Adults with SCD need coordinated care led by a primary care physician in coordination with a provider experienced in SCD (Inati, Chabtini, Mounayar, et al., 2009). However, patients and GPs need to collaboratively have the confidence to manage aspects of SCD at the primary care level; this can be better accomplished by engaging and educating primary care physicians on SCD. Our focus group and questionnaire

study showed a gap in this area. Our limited GP intervention which provided two outputs, an SCD database template and an educational packet aimed at primary care physicians, showed that there is both a need and a desire for educational support (Aljuburi, Phekoo, Okoye, et al., 2012; Aljuburi, Okoye, Majeed, et al., 2012).

Policy Recommendation: *Engage and educate primary health care providers for better management of SCD.*

7.9.3. **OBJECTIVE 3: CREATE AND STRENGTHEN TARGETED AND COMMUNITY BASED SOLUTIONS FOR SCD**

According to the King's Fund, coordinated care programmes flourish at the neighbourhood level where the 'benefits of engagement with local communities sit alongside the need to have close working relationships within multidisciplinary care teams' (Goodwin, Sonola & Thiel, 2013). Nationally and globally, there is growing evidence that this grassroots approach goes to the heart of public health policy.

In one study taking place in the thick forest-covered Indian mountains of Niligiris, an intervention for SCD care and immunisation was implemented. The intervention was very successful and the results of the study showed that even in a vastly disadvantaged area, people can overcome socioeconomic and cultural barriers related to health care at the community level. This community ownership is what the researchers credited the success of the programme to and what they claimed as being 'perhaps the most important lesson that could potentially be translated to other low-income settings' (Nimgaonkar, V, et al., 2014).

A study with a focus on reducing avoidable emergency admissions also boldly stated that the key to avoiding preventable hospital admissions is through 'preventive care delivered in the community, whether through improved technology or service design'; otherwise, we are likely to continue seeing potentially avoidable emergency admissions (Blunt, n.d.). This locally grown approach to reducing avoidable emergency admissions is just as applicable to SCD. SCD management must be targeted and local.

In our study of the HES data, we saw that the rate of hospital admissions in England has increased for SCD; London accounts for the majority of admissions in England and the majority of these hospitalisations are short-stay. These data show that SCD is a growing concern in England and provides a basis to continue further investigations into explaining such trends. Our study also highlighted that the majority of patients admitted with SCD were discharged within 24 hours, suggesting that these admissions could potentially be prevented through better ambulatory and community care of patients. This includes better care in emergency departments, which is an area that PCTs with a high prevalence of SCD should consider investigating (Aljuburi, Laverty, Green, et al., 2012a).

In two more of our studies – the one examining socioeconomic deprivation and the link to admission, and the one examining the costs associated with SCD admissions, we noted that adolescents are a high risk age group. The highest inpatient mortality for those with SCD occurred in patients living in the most socioeconomically deprived areas, with the highest risk of admission for patients aged 10-19 years. We also examined the association between age and gender on incurring extra bed days, showing that patients 10-19 years old are much more likely to incur extra bed days. In the study examining trends on admission using the HES data, we saw that those in the age group 20-29 years had a higher proportion of crisis admissions than the other groups (Aljuburi, Laverty, Green, et al., 2013a). This flags the issue of a gap in addressing the management of adolescents and young adults when it comes coordinating their care.

Adolescents need a bespoke policy that can provide them and their health care providers a bridge from the paediatric guidelines to the adult guideline, in what some researchers refer to as adolescent transitional care (Jordan, Swerdlow & Coates, 2013; Quinn, Rogers, McCavit, et al., 2010; Telfair, Ehiri, Loosier, et al., 2004). This supports a concept analogous to the inverse care law, where differentials in access to care vary inversely with the (SCD) population most at need (Lyrtatzopoulos, Havely, Gemmell, et al., 2005; Hart, 1971). Targeted intervention programmes that concentrate on a small number of high risk patients, such as improving access to appropriate care or adherence to medication protocols (Bloomberg, Trinkaus, Fisher, et al., 2003; Frei-jones, Field & DeBaun, 2009), provide genuine opportunities to prevent a large number of recurrent hospitalisations. Adolescents with SCD are one target group.

Some readmissions may also be prevented through policy initiatives such as the development and implementation of Local Enhanced Services for people with SCD. Local Enhanced Services are designed to meet local health needs and may therefore be particularly suitable for a condition such as SCD where the burden of disease is highest in a few urban areas such as London.

The assumption is that this local and targeted approach can come via a primary care intervention. Patients with short length of stay and multiple admissions may be potentially amenable to primary care intervention. The practices which have the highest numbers of sickle cell disease patients who frequently seek emergency care can be earmarked for educational intervention designed to help further engage general practitioners in the care and management of their sickle cell patients. This assumption is calculated in that it was built on the results from our three targeted exercises introduced in previous chapters.

1. A focus group to inform on patient feelings regarding SCD and how its care is handled.
2. A quantitative questionnaire to better assess the design of an intervention.
3. Installation of a disease specific primary care template that GPs can use when they receive SCD patients in their office.

Policy Recommendation: *Create and strengthen targeted and community based solutions for SCD.*

7.10. CONCLUSION

The results of this research have helped to inform a primary care educational intervention geared toward general practitioners. Supplementary to the educational intervention was the development of an SCD database template that could be uploaded into patient databases in GP surgeries, which could prompt GPs to ask necessary questions when an SCD patient visits his or her GP. The research opened an opportunity to both identify future areas of growth related to this study and to make policy recommendations specific to the management of SCD in England. We have been fortunate to realize several key outputs from this project. The research has resulted in several outputs that provide an opportunity for future research and policy implementation for SCD. It has produced the first national study on trends for hospital admission rates for SCD using HES data in England. It has produced the first study using HES data to assess the cost of admissions due to SCD, and it has delivered both a GP educational intervention guideline and a GP template with the potential for wider roll-out.

This study also identified adolescent care as an area that needs to be addressed. When studying hospital admissions that may be avoidable, counselling the adolescent age group may be an important way to teach how this disease is passed on and that responsible choices should be made. Counselling may also be relevant for not only disease carriers, but also for trait carriers as genetic counselling becomes a part of education for prevention of the next generation of disease.

HES data is a comprehensive database which allows us to see all NHS admissions even those that are carried out in private hospitals. Its limitation is that it does not allow one to understand the patient. For example, we are unable to see severity of disease; what was going on prior to admission or what happens after discharge which then may impact things such as readmission. The patient led studies, i.e., the patient focus group and questionnaire mitigated this, but future studies could further explore this as a broader picture or also examine the rise of hospital admissions due to SCD as compared to the rise in all hospital admissions. This will become more feasible in the future through the availability of new NHS data sets that can link data on individual patients across different parts of the health system and also with external data sets such as mortality records. But knowing this limitation also shows that not only do the findings of the studies give opportunity for future research and policy building, but so do the limitations of this study.

The outputs of the study have helped to inform the community hub project, the SiKL smartphone application, and also helped to secure second round funding for the NW London CLAHRC from the NIHR for continued research for the period 2013-2018. In the feedback from the application for CLAHRC funding, the work on sickle cell disease and the high level of patient participation were highly commended and a key factor in the successful application for funding.

Recently, SCD has also achieved specialised commissioning status, although the current specifications focus mainly on the provision of acute care (Pizzo, Laverty, Phekoo, et al., 2014). This research has provided the groundwork for policy recommendations related to SCD from a public health perspective, an economic perspective, and an equity perspective. Further research is needed for a detailed implementation plan and the absorption of any innovation.

Despite these outputs and the future goals of SCD policy, one thing to critically keep in mind is that the challenge and the reason for why many policies never actually succeed are not in the policies themselves but rather the execution of the policies. There is a disconnect between policy and implementation. In order to create policies that are actually successful and sustainable, including those mentioned above for SCD, there needs to be a bridge across this

divide. Future research will need to address this gap (Gauld, Blank, Burgers, et al., 2012). This will be taken forward in the new phase of CLARHC funding in work led by Professor John Warner and colleagues from Imperial College London and the NHS in NW London.

REFERENCES

- A., O., J., H., A., Y., L., B., et al. (2009) Standards of care for Adult patients with Sick Cell Disease in the United Kingdom. *British Journal of Haematology*. [Online]. 145 p.24. Available from: <http://goo.gl/RZLGxN>
- Adams, R., McKie, V., Nichols, F., Carl, E., et al. (1992) The use of transcranial ultrasonography to predict stroke in sickle cell disease. *The New England journal of medicine*. [Online] 326 (9), 605–610. Available from: <http://goo.gl/DZYq4i> [Accessed: 17 August 2013].
- Aljuburi, G., Laverty, A.A., Green, S.A., Phekoo, K.J., et al. (2013a) Socio-economic deprivation and risk of emergency readmission and inpatient mortality in people with sickle cell disease in England: observational study. *Journal of public health (Oxford, England)*. [Online] 35 (4), 510–517. Available from: doi:10.1093/pubmed/fdt100 [Accessed: 21 March 2014].
- Aljuburi, G., Laverty, A.A., Green, S.A., Phekoo, K.J., et al. (2013b) Socio-economic deprivation and risk of emergency readmission and inpatient mortality in people with sickle cell disease in England: observational study. *Journal of public health (Oxford, England)*. [Online] 35 (4), 510–517. Available from: <http://goo.gl/yT0oyh> [Accessed: 21 March 2014].
- Aljuburi, G., Laverty, A.A., Green, S.A., Phekoo, K.J., et al. (2012a) Trends in hospital admissions for sickle cell disease in England, 2001/02-2009/10. *Journal of public health (Oxford, England)*. [Online] 34 (4), 570–576. Available from: doi:10.1093/pubmed/fds035.
- Aljuburi, G., Laverty, A.A., Green, S.A., Phekoo, K.J., et al. (2012b) Trends in hospital admissions for sickle cell disease in England, 2001/02-2009/10. *Journal of public health Oxford England*. [Online] 1–7. Available from: doi:10.1093/pubmed/fds035.
- Aljuburi, G., Okoye, O., Majeed, A., Knight, Y., et al. (2012) Views of patients about sickle cell disease management in primary care: a questionnaire-based pilot study. *JRSM short reports*. [Online] 3 (11), 78. Available from: doi:10.1258/shorts.2012.011173.
- Aljuburi, G., Phekoo, K.J., Okoye, N.O., Anie, K., et al. (2013a) Patients' views on improving sickle cell disease management in primary care: focus group discussion. *JRSM Short Reports*. [Online] 3 (12), 84–84. Available from: doi:10.1258/shorts.2012.011153.
- Aljuburi, G., Phekoo, K.J., Okoye, N.O., Anie, K., et al. (2012) Patients' views on improving sickle cell disease management in primary care: focus group discussion. *JRSM short reports*. [Online] 3 (12), 84. Available from: doi:10.1258/shorts.2012.011153 [Accessed: 19 August 2013].
- Aljuburi, G., Phekoo, K.J., Okoye, N.O., Anie, K., et al. (2013b) Patients' views on improving sickle cell disease management in primary care: focus group discussion. *JRSM Short Reports*. [Online] 3 (12), 84–84. Available from: doi:10.1258/shorts.2012.011153 [Accessed: 11 March 2013].
- ALLISON, A.C. (1960) Turnovers of Erythrocytes and Plasma Proteins in Mammals. *Nature*. [Online] 188 (4744), 37–40. Available from: doi:10.1038/188037a0 [Accessed: 21 August 2013].

- Amendah, D.D., Mvundura, M., Kavanagh, P.L., Sprinz, P.G., et al. (2010a) Sick cell disease-related pediatric medical expenditures in the U.S. *American journal of preventive medicine*. [Online] 38 (4 Suppl), S550–6. Available from: <http://goo.gl/jWaeuc> [Accessed: 17 August 2013].
- Amendah, D.D., Mvundura, M., Kavanagh, P.L., Sprinz, P.G., et al. (2010b) Sick Cell Disease-Related Pediatric Medical Expenditures in the U.S. *American Journal of Preventive Medicine*. [Online]. 38. Available from: doi:10.1016/j.amepre.2010.01.004.
- Anie, K.A., Steptoe, A. & Bevan, D.H. (2002) Sick cell disease: Pain, coping and quality of life in a study of adults in the UK. *Br J Health Psychol*. [Online] 7331–344. Available from: doi:10.1348/135910702760213715.
- Anionwu, E.N. & Atkin, K. (2001) *The Politics Of Sick Cell*. Buckingham, Open University Press.
- Anon (n.d.) All Together Now: A Conceptual Exploration of Integrated Care :: *Longwoods.com*. [Online]. Available from: <http://goo.gl/DShkWY> [Accessed: 3 April 2014a].
- Anon (n.d.) *Health and Social Care Act 2012*. [Online] Available from: <http://goo.gl/GXToQD> [Accessed: 15 April 2014b].
- Anon (n.d.) *Integrated Care for Patients - NHS Reform / The Nuffield Trust*. [Online]. Available from: <http://goo.gl/JVR6gj> [Accessed: 3 April 2014c].
- Anon (2004) *Landmarks in Medical Genetics: Classic Papers with Commentaries*. [Online]. Oxford University Press. Available from: <http://goo.gl/GpJIU4> [Accessed: 13 September 2013].
- Anon (2012) *National costing report: sickle cell acute painful episode*. p.143.
- National Heart, Lung, and Blood Institute. (2002). *The Management of Sick Cell Disease*. (NIH Publication No. 02-2117).
- Anon (2005) *Northwest London Sick Cell & Thalassaemia Managed Clinical Network. Annual Report & Action Plan 2006–7*.
- Anon (n.d.) *People Powered Health could save the NHS £4.4bn a year / Nesta*. [Online]. Available from: <http://goo.gl/UPnqao> [Accessed: 16 April 2014e].
- Anon (2008) *Standards for the Clinical Care of Adults with Sick Cell Disease in the UK*.
- Aquino, V.M., Norvell, J.M. & Buchanan, G.R. (1997) Acute splenic complications in children with sickle cell-hemoglobin C disease. *The Journal of pediatrics*. [Online] 130 (6), 961– 965. Available from: <http://goo.gl/gXIH31> [Accessed: 17 August 2013].
- Arbaje, A.I., Wolff, J.L., Yu, Q., Powe, N.R., et al. (2008) Postdischarge environmental and socioeconomic factors and the likelihood of early hospital readmission among community-dwelling Medicare beneficiaries. *The Gerontologist*. [Online] 48 (4), 495–504. Available from: <http://goo.gl/Ocuafy>.
- Avis, M., Bond, M. & Arthur, A. (1995) Satisfying solutions? A review of some unresolved issues in the measurement of patient satisfaction. *Journal of advanced nursing*. [Online] 22316–322. Available from: doi:10.1046/j.1365-2648.1995.22020316.x.

- Babhulkar, S.S., Pande, K. & Babhulkar, S. (1995) The hand-foot syndrome in sickle-cell haemoglobinopathy. *The Journal of bone and joint surgery. British volume*. [Online] 77 (2), 310–312. Available from: <http://goo.gl/xRrwFj> [Accessed: 17 August 2013].
- Ballas, S.K. (2009) The cost of health care for patients with sickle cell disease. *Am. J. Hematol.* 84320–322.
- Ballas, S.K. & Lusardi, M. (2005a) Hospital readmission for adult acute sickle cell painful episodes: frequency, etiology, and prognostic significance. *American Journal of Hematology*. [Online] 79 (1), 17–25. Available from: doi:10.1002/ajh.20336 [Accessed: 3 March 2013].
- Ballas, S.K. & Lusardi, M. (2005b) Hospital readmission for adult acute sickle cell painful episodes: frequency, etiology, and prognostic significance. *American journal of hematology*. [Online] 79 (1), 17–25. Available from: doi:10.1002/ajh.20336 [Accessed: 3 March 2013].
- Bankart, M.J.G., Baker, R., Rashid, A., Habiba, M., et al. (2011) Characteristics of general practices associated with emergency admission rates to hospital: a cross-sectional study. *Emergency medicine journal : EMJ*. [Online] 28558–563. Available from: doi:10.1136/emj.2010.108548.
- Bardsley, M., Smith, J. & Car, J. (2013) *Evaluation of the first year of the Inner North West London Integrated Care Pilot*.
- Benjamin, L.J., Swinson, G.I. & Nagel, R.L. (2000) Sickle cell anemia day hospital : an approach for the management of uncomplicated painful crises Plenary paper Sickle cell anemia day hospital : an approach for the management of uncomplicated painful crises. *Blood*. [Online] 95 (4), 1130–1136. Available from: <http://goo.gl/VFqBKv> [Accessed: 17 August 2013].
- Bilenker, J.H., Weller, W.E., Shaffer, T.J., Dover, G.J., et al. (1998) The costs of children with sickle cell anemia: Preparing for managed care. *Journal of Pediatric Hematology/Oncology*. [Online] 20528–533. Available from: doi:10.1097/00043426-199811000-00003.
- Blair, A.S., Lloyd-Williams, F. & Mair, F.S. (2002) What do we know about socioeconomic status and congestive heart failure? A review of the literature. *The Journal of family practice*. [Online] 51 (2), 169. Available from: <http://goo.gl/27v11x>.
- Bloom, F.J., Graf, T., Anderer, T. & Stewart, W.F. (2010) Redesign of a Diabetes System of Care Using an All-or-None Diabetes Bundle to Build Teamwork and Improve Intermediate Outcomes. *Diabetes Spectrum*. 23 (3) pp.165–169.
- Bloomberg, G.R., Trinkaus, K.M., Fisher, E.B., Musick, J.R., et al. (2003) Hospital readmissions for childhood asthma: a 10-year metropolitan study. *American journal of respiratory and critical care medicine*. [Online] 167 (8), 1068–1076. Available from: doi:10.1164/rccm.2201015 [Accessed: 16 March 2013].
- Blunt, I. (n.d.) *Preventable Emergency Admissions – NHS Quality | The Nuffield Trust*. [Online]. Available from: <http://goo.gl/6u7sNj> [Accessed: 3 April 2014].
- BME Health Forum (2014) *Unscheduled Care Insight Project*.

- Bonds, D.R. (2005) Three decades of innovation in the management of sickle cell disease: the road to understanding the sickle cell disease clinical phenotype. *Blood reviews*. [Online] 19 (2), 99–110. Available from: <http://goo.gl/YjluR7> [Accessed: 17 August 2013].
- Booth, C., Inusa, B. & Obaro, S.K. (2010) Infection in sickle cell disease: a review. *International journal of infectious diseases : IJID : official publication of the International Society for Infectious Diseases*. [Online] 14 (1), e2–e12. Available from: <http://goo.gl/1hSKTy> [Accessed: 17 August 2013].
- Bryan, S., Dormandy, E., Roberts, T., Ades, A., et al. (2011) Screening for sickle cell and thalassaemia in primary care: a cost-effectiveness study. *The British journal of general practice : the journal of the Royal College of General Practitioners*. [Online] 61 (591), e620–7. Available from: [doi:10.3399/bjgp11X601325](https://doi.org/10.3399/bjgp11X601325) [Accessed: 4 May 2014].
- Carroll, C.P., Haywood, C., Fagan, P. & Lanzkron, S. (2009) The course and correlates of high hospital utilization in sickle cell disease: Evidence from a large, urban Medicaid managed care organization. *American journal of hematology*. 84 (10), 666–670.
- Carroll, C.P., Haywood, C. & Lanzkron, S. (2011) Prediction of onset and course of high hospital utilization in sickle cell disease. *Journal of Hospital Medicine*. [Online] 6248–255. Available from: [doi:10.1002/jhm.850](https://doi.org/10.1002/jhm.850).
- Castro, O., Brambilla, D.J., Thorington, B., Reindorf, C.A., et al. (1994) The acute chest syndrome in sickle cell disease: incidence and risk factors. The Cooperative Study of Sickle Cell Disease. *Blood*. [Online] 84 (2), 643–649. Available from: <http://goo.gl/MTwukQ> [Accessed: 12 August 2013].
- Charlson, M., Pompei, P., Ales, K. & MacKenzie, C. (1987) A new method of classifying prognostic comorbidity in longitudinal studies: development and validation. *Journal of Chronic Diseases*. 40 (5), 373–383.
- Children's Mercy Hospitals and Clinics (2010) *Splenic Sequestration*. [Online]. 2010. Available from: <http://goo.gl/VAGtuE> [Accessed: 7 August 2010].
- Coleman, P., Irons, R. & Nicholl, J. (2001) Will alternative immediate care services reduce demands for non-urgent treatment at accident and emergency? *Emergency medicine journal : EMJ*. [Online] 18 (6), 482–487. Available from: <http://goo.gl/Pp8kZ8>.
- Condon, P.I. & Serjeant, G.R. (1980) Photocoagulation in proliferative sickle retinopathy: results of a 5-year study. *The British journal of ophthalmology*. [Online] 64 (11), 832–840. Available from: <http://goo.gl/xhBeRn> [Accessed: 17 August 2013].
- Coulter, A. (2005) What do patients and the public want from primary care? *BMJ (Clinical research ed.)*. [Online] 331 (7526), 1199–1201. Available from: [doi:10.1136/bmj.331.7526.1199](https://doi.org/10.1136/bmj.331.7526.1199) [Accessed: 18 April 2014].
- Cowling, T.E., Cecil, E. V, Soljak, M.A., Lee, J.T., et al. (2013) Access to primary care and visits to emergency departments in England: a cross-sectional, population-based study. *PloS one*. [Online] 8 (6), e66699. Available from: [doi:10.1371/journal.pone.0066699](https://doi.org/10.1371/journal.pone.0066699) [Accessed: 21 March 2014].

- Cowling, T.E., Harris, M.J., Watt, H.C., Gibbons, D.C., et al. (2014) Access to general practice and visits to accident and emergency departments in England: cross-sectional analysis of a national patient survey. *British Journal of General Practice*. [Online] 64 (624), e434–e439. Available from: doi:10.3399/bjgp14X680533 [Accessed: 1 July 2014].
- Crawford, M.J. (2002) Systematic review of involving patients in the planning and development of health care. *BMJ*. [Online] 325 (7375), 1263–1263. Available from: doi:10.1136/bmj.325.7375.1263 [Accessed: 25 March 2014].
- Crosby, L.E., Barach, I., McGrady, M.E., Kalinyak, K.A., et al. (2012) Integrating interactive web-based technology to assess adherence and clinical outcomes in pediatric sickle cell disease. *Anemia*. [Online] 2012. Available from: doi:10.1155/2012/492428.
- Davis, K., Schoenbaum, S.C. & Audet, A.M. (2005) A 2020 vision of patient-centered primary care. *Journal of General Internal Medicine*. [Online]. 20 pp.953–957. Available from: doi:10.1111/j.1525-1497.2005.0178.x.
- Day, T.G., Thein, S.L., Drasar, E., Dick, M.C., et al. (2011) Changing Pattern of Hospital Admissions of Children With Sickle Cell Disease Over the Last 50 Years. *Journal of Pediatric Hematology/Oncology*. [Online]. 33 pp.491–495. Available from: doi:10.1097/MPH.0b013e31822543f4.
- Department for Communities and Local Government (2011) The English Indices of Deprivation 2010. *English*.
- Department of Health (2010) *Improving the Health and Well-Being of People with Long-Term Conditions*.
- Department of Health (2005) Supporting people with long term conditions: An NHS and social care model to support local innovation and integration. *Health Affairs*. [Online]. 28 p.48. Available from: <http://goo.gl/xNnTVh>.
- Desai, D. & Dhanani, H. (2004) Sickle Cell Disease: History And Origin. *The Internet Journal of Hematology*. [Online] 1 (2). Available from: doi:DOI: 10.5580/499.
- Dick, M.C. (2008) Standards for the management of sickle cell disease in children. *Archives of disease in childhood. Education and practice edition*. [Online] 93 (6), 169–176. Available from: <http://goo.gl/FP54eh> [Accessed: 17 August 2013].
- Edgman-Levitan, S., Brady, C. & Howitt, P. (2013a) *Partnering with Patients, Families, and Communities For Health: A Global Imperative*.
- Edgman-Levitan, S., Brady, C. & Howitt, P. (2013b) *Partnering with Patients, Families, and Communities For Health: A Global Imperative, Report of the Patient and Family Engagement Working Group 2013*.
- Elander, J., Lusher, J., Bevan, D., Telfer, P., et al. (2004) Understanding the causes of problematic pain management in sickle cell disease: evidence that pseudoaddiction plays a more important role than genuine analgesic dependence. *Journal of pain and symptom management*. [Online] 27 (2), 156–169. Available from: doi:10.1016/j.jpainsymman.2003.12.001 [Accessed: 16 March 2013].

- Elandera, J., Beach, M.C. & Haywood Jr, C. (2011) Respect, trust, and the management of sickle cell disease pain in hospital: comparative analysis of concern-raising behaviors, preliminary model, and agenda for international collaborative research to inform practice. *Ethnicity and Health*. [Online] 16 (4-5), 405–421. Available from: doi:10.1080/13557858.2011.555520. Respect.
- Elliott, L., Ashley-Koch, A.E., De Castro, L., Jonassaint, J., et al. (2007) Genetic polymorphisms associated with priapism in sickle cell disease. *British journal of haematology*. [Online] 137 (3), 262–267. Available from: doi:10.1111/j.1365-2141.2007.06560.x [Accessed: 19 August 2013].
- Embury, S.H. (1986) The clinical pathophysiology of sickle cell disease. *Annual review of medicine*. [Online] 37361–376. Available from: <http://goo.gl/e5KAjN> [Accessed: 17 August 2013].
- Francis, R. (2013) *Report of the Mid Staffordshire NHS Foundation Trust Public Inquiry: Executive summary*.
- Freed, L.H., Ellen, J.M., Irwin, C.E. & Millstein, S.G. (1998) Determinants of adolescents' satisfaction with health care providers and intentions to keep follow-up appointments. *Journal of Adolescent Health*. [Online]. 22 pp.475–479. Available from: doi:10.1016/S1054-139X(98)00002-0.
- Frei-jones, M.J., Field, J.J. & DeBaun, M.R. (2009) Risk Factors for Hospital Readmission Within 30 Days : A New Quality Measure for Children With Sickle Cell Disease. *Pediatric Blood Cancer*. [Online] 52481–485. Available from: doi:10.1002/pbc.
- Frenette, P.S. (2002) Sickle cell vaso-occlusion: multistep and multicellular paradigm. *Current opinion in hematology*. [Online] 9 (2), 101–106. Available from: <http://goo.gl/Tolhlo> [Accessed: 17 August 2013].
- Frenette, P.S. & Atweh, G.F. (2007) Sickle cell disease: old discoveries, new concepts, and future promise. *The Journal of clinical investigation*. [Online] 117 (4), 850–858. Available from: <http://goo.gl/YyPBuK> [Accessed: 17 August 2013].
- Friedberg, M.W., Van Busum, K., Wexler, R., Bowen, M., et al. (2013) A demonstration of shared decision making in primary care highlights barriers to adoption and potential remedies. *Health affairs (Project Hope)*. [Online] 32268–275. Available from: doi:10.1377/hlthaff.2012.1084.
- Gauld, R., Blank, R., Burgers, J., Cohen, A.B., et al. (2012) The World Health Report 2008 - Primary Healthcare: How Wide Is the Gap between Its Agenda and Implementation in 12 High-Income Health Systems? *Healthcare policy = Politiques de santé*. [Online] 7 (3), 38–58. Available from: <http://goo.gl/eUDkCh>.
- Geis, S.B., Fuller, R.L. & Rush, J. (1986) Lovers of aids victims: Psychosocial stresses and counseling needs. *Death Studies*. [Online]. 10 pp.43–53. Available from: doi:10.1080/07481188608252798.
- Geue, C., Lewsey, J., Lorgelly, P., Govan, L., et al. (2012) Spoilt for choice: Implications of using alternative methods of costing hospital episode statistics. *Health Economics (United Kingdom)*. [Online] 211201–1216. Available from: doi:10.1002/hec.1785.

- Gibbons, D.C., Bindman, A.B., Soljak, M.A., Millett, C., et al. (2012) Defining primary care sensitive conditions: a necessity for effective primary care delivery? *Journal of the Royal Society of Medicine*. [Online] 105 (10), 422–428. Available from: doi:10.1258/jrsm.2012.120178 [Accessed: 15 April 2014].
- Glassberg, J.A., Wang, J., Cohen, R., Richardson, L.D., et al. (2012) Risk factors for increased ED utilization in a multinational cohort of children with sickle cell disease. *Academic emergency medicine : official journal of the Society for Academic Emergency Medicine*. [Online] 19 (6), 664–672. Available from: doi:10.1111/j.1553-2712.2012.01364.x [Accessed: 13 September 2013].
- Goodwin, N., Smith, J., Davies, A., Perry, C., et al. (2012) *Integrated care for patients and populations: Improving outcomes by working together*.
- Goodwin, N., Sonola, L. & Thiel, V. (2013) *Co-ordinated care for people with complex chronic conditions: Key lessons and markers for success* . p.44.
- Green, S.A., Aljuburi, G., Majeed, A., Okoye, O., et al. (2012a) Characterizing emergency admissions of patients with sickle cell crisis in NHS Brent: observational study. *JRSM short reports*. [Online] 3 (6), 37. Available from: doi:10.1258/shorts.2012.011129.
- Green, S.A., Aljuburi, G., Majeed, A., Okoye, O., et al. (2012b) Characterizing emergency admissions of patients with sickle cell crisis in NHS Brent: observational study. *JRSM short reports*. [Online] 3 (6), 37. Available from: doi:10.1258/shorts.2012.011129 [Accessed: 11 March 2013].
- Griffiths, C., Foster, G., Ramsay, J., Eldridge, S., et al. (2007) How effective are expert patient (lay led) education programmes for chronic disease? *BMJ : British Medical Journal*. [Online] 3341254–1256. Available from: doi:10.1136/bmj.39227.698785.47.
- Grogan, S., Conner, M., Norman, P., Willits, D., et al. (2000) Validation of a questionnaire measuring patient satisfaction with general practitioner services. *Quality in health care : QHC*. [Online] 9210–215. Available from: doi:10.1136/qhc.9.4.210.
- Hakim, Lawrence S. Hashmat, A.I. & Macchia, R.J. (1994) Priapism. In: Stephen H. Embury, Robert P. Hebbel, Narla Mohandas, & Martin H Steinberg (eds.). *Sickle cell disease : basic principles and clinical practice*. New York, Raven Press.
- Ham, C., Imison, C., Goodwin, N., Dixon, A., et al. (2011) Where next for the NHS reforms? The case for integrated care. *The King's Fund*. [Online] 1–19. Available from: <http://goo.gl/E9s6gK>.
- Haque, A. & Telfair, J. (2000) Socioeconomic distress and health status: the urban-rural dichotomy of services utilization for people with sickle cell disorder in North Carolina. *The Journal of rural health : official journal of the American Rural Health Association and the National Rural Health Care Association*. 1643–55.
- Hardy GE, West MA & Hill F (1996) Components and predictors of patient satisfaction. *Br J Health Psychol*. 165–85.
- Harris, L.E., Swindle, R.W., Mungai, S.M., Weinberger, M., et al. (1999) Measuring patient satisfaction for quality improvement. *Medical care*. [Online] 371207–1213. Available from: doi:10.1097/00005650-199912000-00004.

- Hart, J.T. (1971) The Inverse Care Law. *The Lancet*. 297 (7696), 405–412.
- Hayes, N. (2013) *Health reforms, opinion polls and surveys: myths and realities*. [Online] Available from: <http://goo.gl/tcX4lo>.
- Haywood Jr, C., Lanzkron, S. & Ratanawongsa, N. (2011) Associations with Trust and Interpersonal Experiences with. *Journal of hospital medicine*. [Online] 5 (5), 289–294. Available from: doi:10.1002/jhm.643.Hospital.
- Health, D. of (n.d.) *Improving Chronic Disease Management*. [Online]. Available from: <http://goo.gl/rsZ2OS> [Accessed: 18 April 2014].
- Heary, C.M. & Hennessy, E. (2002) The use of focus group interviews in pediatric health care research. *Journal of Pediatric Psychology*. [Online] 2747–57. Available from: doi:10.1093/jpepsy/27.1.47.
- Hebbel, R.P., Eaton, J.W., Balasingam, M. & Steinberg, M.H. (1982) Spontaneous oxygen radical generation by sickle erythrocytes. *The Journal of clinical investigation*. [Online] 70 (6), 1253–1259. Available from: <http://goo.gl/v3Ushw> [Accessed: 17 August 2013].
- Herrick, J.B. (2001) Peculiar elongated and sickle-shaped red blood corpuscles in a case of severe anemia. 1910. *The Yale journal of biology and medicine*. [Online] 74 (3), 179–184. Available from: <http://goo.gl/uTkWdY> [Accessed: 11 August 2013].
- Hibbard, J.H. & Greene, J. (2013) What the evidence shows about patient activation: better health outcomes and care experiences; fewer data on costs. *Health affairs (Project Hope)*. [Online] 32207–214. Available from: doi:10.1377/hlthaff.2012.1061.
- Hibbard, J.H., Greene, J. & Overton, V. (2013) Patients with lower activation associated with higher costs; delivery systems should know their patients' "scores". *Health affairs (Project Hope)*. [Online] 32216–222. Available from: doi:10.1377/hlthaff.2012.1064.
- Hickman, M., Modell, B., Greengross, P., Chapman, C., et al. (1999) Mapping the prevalence of sickle cell and beta thalassaemia in England: estimating and validating ethnic-specific rates. *British journal of haematology*. 104 (4), 860–867.
- Hockstein, E. (2009) Cured patients help in TB treatment in Tanzania. *World Health Organization (WHO)*. [Online]. Available from: <http://goo.gl/rXJSmu>.
- Howard-Jones, M., Randall, L., Bailey-Squire, B., Clayton, J., et al. (2009) An audit of immunisation status of sickle cell patients in Coventry, UK. *Journal of clinical pathology*. [Online] 62 (1), 42–45. Available from: <http://goo.gl/SZzii> [Accessed: 17 August 2013].
- HSCIC (2013) *Quality and Outcomes Framework*. [Online]. 2013. Primary Care: Support and Guidance. Available from: <http://goo.gl/X6Oqse>.
- Hu, J., Gonsahn, M.D. & Nerenz, D.R. (2014) Socioeconomic Status And Readmissions: Evidence From An Urban Teaching Hospital. *Health Affairs*. [Online] 33778–785. Available from: doi:10.1377/hlthaff.2013.0816.
- Inati, A., Chabtini, L., Mounayar, M. & Taher, A. (2009) Current understanding in the management of sickle cell disease. *Hemoglobin*. [Online] 33 Suppl 1S107–S115. Available from: doi:10.3109/03630260903347682.

- Ireys, H.T. & Perry, J.J. (1999) Development and evaluation of a satisfaction scale for parents of children with special health care needs. *Pediatrics*. 1041182–1191.
- Jenerette, C.M., Brewer, C. & Leak, A.N. (2011) Self-Care Recommendations of Middle-Aged and Older Adults with Sickle Cell Disease. *Nursing Research and Practice*. [Online]. 2011 pp.1–5. Available from: doi:10.1155/2011/270594.
- Jenerette, C.M. & Murdaugh, C. (2008) Testing the theory of self-care management for sickle cell disease. *Research in Nursing and Health*. [Online] 31355–369. Available from: doi:10.1002/nur.20261.
- Jordan, L., Swerdlow, P. & Coates, T.D. (2013) Systematic review of transition from adolescent to adult care in patients with sickle cell disease. *Journal of pediatric hematology/oncology*. [Online] 35 (3), 165–169. Available from: doi:10.1097/MPH.0b013e3182847483.
- Kanter, J. & Kruse-Jarres, R. (2013) Management of sickle cell disease from childhood through adulthood. *Blood Reviews*. [Online] 27279–287. Available from: doi:10.1016/j.blre.2013.09.001.
- Kauf, T.L., Coates, T.D., Huazhi, L., Mody-Patel, N., et al. (2009) The cost of health care for children and adults with sickle cell disease. *American Journal of Hematology*. [Online] 84323–327. Available from: doi:10.1002/ajh.21408.
- Kelley, M.A., Alexander, C.S. & Morris, N.M. (1991) Maternal satisfaction with primary care for children with selected chronic conditions. *Journal of Community Health*. [Online] 16213–224. Available from: doi:10.1007/BF01324389.
- Kemper, A.R., Uren, R.L., Moseley, K.L. & Clark, S.J. (2006) Primary care physicians' attitudes regarding follow-up care for children with positive newborn screening results. *Pediatrics*. [Online] 118 (5), 1836–1841. Available from: doi:10.1542/peds.2006-1639 [Accessed: 11 October 2013].
- Kennedy, A., Rogers, A. & Bower, P. (2007) Support for self care for patients with chronic disease. *BMJ (Clinical research ed.)*. [Online] 335 (7627), 968–970. Available from: doi:10.1136/bmj.39372.540903.94 [Accessed: 22 March 2014].
- Khelil, A., Perrin, P., Lefranc, G. & Chibani, J. (2012) Synthetic review on the different anthropological aspects of hemoglobinopathies in Tunisia. *International Journal of Modern Anthropology*. [Online] 1 (5), 60–78. Available from: doi:10.4314/ijma.v1i5.4 [Accessed: 31 August 2013].
- Kitzinger, J. (1995) Introducing focus groups ' P. *BMJ: British Medical Journal*. 311299–302.
- Koshy, M., Entsuah, R., Koranda, A., Kraus, A.P., et al. (1989) Leg ulcers in patients with sickle cell disease. *Blood*. [Online] 74 (4), 1403–1408. Available from: http://goo.gl/6IfgcT [Accessed: 17 August 2013].
- Landon, B.E., Gill, J.M., Antonelli, R.C. & Rich, E.C. (2010) Prospects for rebuilding primary care using the patient-centered medical home. *Health affairs (Project Hope)*. [Online] 29827–834. Available from: doi:10.1377/hlthaff.2010.0016.

- Langer, S., Chew-Graham, C., Hunter, C., Guthrie, E.A., et al. (2013) Why do patients with long-term conditions use unscheduled care? A qualitative literature review. *Health & social care in the community*. [Online] 21 (4), 339–351. Available from: doi:10.1111/j.1365-2524.2012.01093.x [Accessed: 18 April 2014].
- Lehmann, H. (1954) Distribution of the sickle cell gene : A new light on the origin of the East Africans. *The Eugenics review*. [Online] 46 (2), 101–121. Available from: <http://goo.gl/YKiYVz> [Accessed: 17 August 2013].
- Lorig, K., Sobel, D. & Stewart, A. (1999) Evidence suggesting that a chronic disease self-management program can improve health status while reducing hospitalization: a randomized trial. *Medical care*. [Online] 375–14. Available from: <http://goo.gl/9zFswH>.
- Lorig, K.R. & Holman, H. (2003) Self-management education: history, definition, outcomes, and mechanisms. *Annals of behavioral medicine : a publication of the Society of Behavioral Medicine*. [Online]. 26 pp.1–7. Available from: doi:10.1207/S15324796ABM2601_01.
- Lorig, K.R., Sobel, D.S., Ritter, P.L., Laurent, D., et al. (2001) Effect of a self-management program on patients with chronic disease. *Effective clinical practice : ECP*. [Online] 4256–262. Available from: <http://goo.gl/EvP42i>.
- Lucas, S.B., Mason, D., Mason, M. & Weyman, D. (2008) *A Sickle Crisis? A Sickle Crisis? A Report of the National Confidential Enquiry into Patient Outcome and Death*.
- Luzzatto, L., Nwachuku-Jarrett, E.S. & Reddy, S. (1970) Increased sickling of parasitised erythrocytes as mechanism of resistance against malaria in the sickle-cell trait. *Lancet*. [Online] 1 (7642), 319–321. Available from: <http://goo.gl/0E9J4S> [Accessed: 17 August 2013].
- Lyratzopoulos, G., Havelly, D., Gemmell, I. & Cook, G. a (2005) Factors influencing emergency medical readmission risk in a UK district general hospital: A prospective study. *BMC emergency medicine*. [Online] 5 (1), 1. Available from: doi:10.1186/1471-227X-5-1 [Accessed: 11 March 2013].
- Mackenbach, J. (2006) *Socio-economic inequalities in health in Western Europe. From description to explanation to intervention*. Oxford, Oxford University Press.
- Magnus, S.A., Hambleton, I.R., Moosdeen, F. & Serjeant, G.R. (1999) Recurrent infections in homozygous sickle cell disease. *Archives of disease in childhood*. [Online] 80 (6), 537–541. Available from: <http://goo.gl/ijBDvG> [Accessed: 17 August 2013].
- Majeed, A. (2004) Sources, uses, strengths and limitations of data collected in primary care in England. *Health statistics quarterly / Office for National Statistics*. (21), 5–14.
- Makani, J., Komba, A.N., Cox, S.E., Oruo, J., et al. (2010) Malaria in patients with sickle cell anemia: burden, risk factors, and outcome at the outpatient clinic and during hospitalization. *Blood*. [Online] 115 (2), 215–220. Available from: doi:10.1182/blood-2009-07-233528 [Accessed: 17 August 2013].
- Manci, E.A., Culberson, D.E., Yang, Y.-M., Gardner, T.M., et al. (2003) Causes of death in sickle cell disease: an autopsy study. *British journal of haematology*. [Online] 123 (2), 359–365. Available from: <http://goo.gl/defJX4> [Accessed: 17 August 2013].

- Mankad, V.N. (2001) Exciting new treatment approaches for pathyphysiologic mechanisms of sickle cell disease. *Pediatric pathology & molecular medicine*. [Online] 20 (1), 1–13. Available from: <http://goo.gl/m3IQGm> [Accessed: 17 August 2013].
- Marmot, M. & Bell, R. (2012) Fair society, healthy lives. *Public Health*. 126 (SUPPL.1).
- Mason, A., Daly, E. & Goldacre, M. (2000) *Hospital re-admission rates: literature review – full report*.
- Maxwell, K., Streetly, a & Bevan, D. (1999a) Experiences of hospital care and treatment- seeking behavior for pain from sickle cell disease: qualitative study. *The Western journal of medicine*. [Online] 171 (5-6), 306–313. Available from: <http://goo.gl/tQvrSE>.
- Maxwell, K., Streetly, A. & Bevan, D. (1999b) Experiences of hospital care and treatment seeking for pain from sickle cell disease: qualitative study. *BMJ*. [Online]. 318 pp.1585– 1590. Available from: doi:10.1136/bmj.318.7198.1585.
- McAuley, C.F., Webb, C., Makani, J., Macharia, A., et al. (2010) High mortality from Plasmodium falciparum malaria in children living with sickle cell anemia on the coast of Kenya. *Blood*. [Online] 116 (10), 1663–1668. Available from: <http://goo.gl/80SeSh> [Accessed: 17 August 2013].
- McCarthy, K., McGee, H.M. & O’Boyle, C.A. (2000) Outpatient clinic waiting times and non-attendance as indicators of quality. *Psychology, Health & Medicine*. [Online]. 5 pp.287– 293. Available from: doi:10.1080/713690194.
- McClellan, C.B., Schatz, J.C., Puffer, E., Sanchez, C.E., et al. (2009) Use of handheld wireless technology for a home-based sickle cell pain management protocol. *Journal of pediatric psychology*. [Online] 34 (5), 564–573. Available from: doi:10.1093/jpepsy/jsn121 [Accessed: 7 June 2014].
- McClellan, M., Kent, J., Beales, S., Macdonnell, M., et al. (2013) *Focusing Accountability on the Outcomes that Matter*.
- McColl Karen Pickworth, S. & Raymond, I. (2006) Project: London : supporting vulnerable populations. *BMJ*. [Online] 331 (331 (7533):), 115–117. Available from: doi:10.1136/bmj.332.7533.115.
- Mitchell, M.J., Lemanek, K., Palermo, T.M., Crosby, L.E., et al. (2007) Parent perspectives on pain management, coping, and family functioning in pediatric sickle cell disease. *Clinical pediatrics*. [Online] 46311–319. Available from: doi:10.1177/0009922806293985.
- Modi, A.C., Crosby, L.E., Hines, J., Drotar, D., et al. (2012) Feasibility of web-based technology to assess adherence to clinic appointments in youth with sickle cell disease. *Journal of pediatric hematology/oncology*. [Online] 34e93–6. Available from: doi:10.1097/MPH.0b013e318240d531.
- Modi AC, Crosby LE, Guilfoyle SM, Lemanek KL, et al. (2009) Barriers to treatment adherence for pediatric patients with sickle cell disease and their families . *Child Health Care* . 38107– 122.
- Morgan, D.L. (1997) *Focus Groups as Qualitative Research*. [Online]. Available from: doi:10.1016/j.chroma.2010.04.020.

- Nagel, R.L., Erlingsson, S., Fabry, M.E., Croizat, H., et al. (1991) The Senegal DNA haplotype is associated with the amelioration of anemia in African-American sickle cell anemia patients. *Blood*. [Online] 77 (6), 1371–1375. Available from: <http://goo.gl/LkBVwK> [Accessed: 17 August 2013]. National Heart Lung and Blood Institute (1996) *Facts about sickle cell anemia*.
- NHS England (2013) *Transforming Participation in Health and Care “The NHS belongs to us all.”* [Online]. Available from: <http://bit.ly/1bKmJK4>.
- NHS Sickle Cell and Thalassaemia & Screening Programmes (2010) *Sickle Cell Disease in Childhood Standards and Guidelines for Clinical Care, 2nd edition*.
- Nietert, P.J., Silverstein, M.D. & Abboud, M.R. (2002) Sickle cell anaemia: epidemiology and cost of illness. *Pharmacoeconomics*. 20357–366.
- Nolte, E., Roland, M., Guthrie, S. & Brereton, L. (2012) *Preventing emergency readmissions to hospital: A scoping review*.
- Nur, E., Biemond, B.J., Otten, H.-M., Brandjes, D.P., et al. (2011) Oxidative stress in sickle cell disease; pathophysiology and potential implications for disease management. *American journal of hematology*. [Online] 86 (6), 484–489. Available from: doi:10.1002/ajh.22012 [Accessed: 21 March 2014].
- Nwaoguikpe, R.N. & Uwakwe, A.A. (2005) The antisickling effects of dried fish (tilapia) And dried prawn (Astacus red). *Journal of Applied Sciences and Environmental Management*. [Online] 9 (3), 115–119. Available from: <http://goo.gl/qTtF65> [Accessed: 17 August 2013].
- Ohnishi, S.T., Ohnishi, T. & Ogunmola, G.B. (2001) Green tea extract and aged garlic extract inhibit anion transport and sickle cell dehydration in vitro. *Blood cells, molecules & diseases*. [Online] 27 (1), 148–157. Available from: <http://goo.gl/C9UNKZ> [Accessed: 17 August 2013].
- Okpuzor, J., Adebisin, O., Ogbunugafor, H. & Amadi, I. (2008) The potential of medicinal plants in sickle cell disease control: A review. *International Journal of Biomedical and Health Sciences*. [Online] Available from: <http://goo.gl/Qh1OgM> [Accessed: 17 August 2013].
- Olney, R.S. (1999) Preventing morbidity and mortality from sickle cell disease: *American Journal of Preventive Medicine*. [Online] 16 (2), 116–121. Available from: doi:10.1016/S0749-3797(98)00140-8.
- Olowoyeye, A. & Okwundu, C. (2010) Gene therapy for sickle cell disease. *Cochrane Database Syst Rev*. [Online] (11). Available from: <http://goo.gl/PvNBBt> [Accessed: 11 October 2013].
- Orkin, S.H. & Higgs, D.R. (2010a) Medicine. Sickle cell disease at 100 years. *Science (New York, N.Y.)*. [Online] 329 (5989), 291–292. Available from: doi:10.1126/science.1194035 [Accessed: 12 August 2013].
- Orkin, S.H. & Higgs, D.R. (2010b) Medicine. Sickle cell disease at 100 years. *Science (New York, N.Y.)*. [Online] 329 (5989), 291–292. Available from: doi:10.1126/science.1194035.

- Oteng-Ntim, E., Chase, A.R., Howard, J., Khazaezadeh, N., et al. (2008) Sick cell disease in pregnancy. *Obstetrics, Gynaecology & Reproductive Medicine*. [Online] 18 (10), 272–278. Available from: doi:10.1016/j.ogrm.2008.08.001 [Accessed: 19 August 2013].
- Oyeku, S.O., Feldman, H.A., Ryan, K., Muret-Wagstaff, S., et al. (2010) Primary Care Clinicians' Knowledge and Confidence About Newborn Screening for Sick Cell Disease: Randomized Assessment of Educational Strategies. *Journal of the National Medical Association*. 102 (8), 676–682.
- Oyeku, S.O., Wang, C.J., Scoville, R., Vanderkruik, R., et al. (2012) Hemoglobinopathy Learning Collaborative: Using Quality Improvement (QI) to Achieve Equity in Health Care Quality, Coordination, and Outcomes for Sick Cell Disease. *Journal of Health Care for the Poor and Underserved*. [Online]. 23 pp.34–48. Available from: doi:10.1353/hpu.2012.0127.
- Pace, B.S. & Zein, S. (2006) Understanding mechanisms of gamma-globin gene regulation to develop strategies for pharmacological fetal hemoglobin induction. *Developmental dynamics : an official publication of the American Association of Anatomists*. [Online] 235 (7), 1727–1737. Available from: <http://goo.gl/ArppBU> [Accessed: 17 August 2013].
- Pack-Mabien, A. & Haynes, J. (2009) A primary care provider's guide to preventive and acute care management of adults and children with sickle cell disease. *Journal of the American Academy of Nurse Practitioners*. [Online] 21 (5), 250–257. Available from: <http://goo.gl/4xxWJI> [Accessed: 17 August 2013].
- Pack-Mabien, A., Labbe, E., Herbert, D. & Haynes, J. (2001a) Nurses' attitudes and practices in sickle cell pain management. *Applied nursing research : ANR*. [Online] 14 (4), 187–192. Available from: doi:10.1053/apnr.2001.26783 [Accessed: 17 August 2013].
- Pack-Mabien, A., Labbe, E., Herbert, D. & Haynes, J. (2001b) Nurses' attitudes and practices in sickle cell pain management. *Applied nursing research : ANR*. [Online] 14 (4), 187–192. Available from: <http://goo.gl/BxIS5b> [Accessed: 17 August 2013].
- Pagnier, J. (1984) Evidence for the Multicentric Origin of the Sick Cell Hemoglobin Gene in Africa. *Proceedings of the National Academy of Sciences*. [Online] 81 (6), 1771–1773. Available from: <http://goo.gl/ecI4Xu> [Accessed: 17 August 2013].
- Panepinto, J.A., O???Mahar, K.M., DeBaun, M.R., Rennie, K.M., et al. (2004) Validity of the Child Health Questionnaire for Use In Children With Sick Cell Disease. *Journal of Pediatric Hematology/Oncology*. [Online]. 26 pp.574–578. Available from: doi:10.1097/01.mph.0000136453.93704.2e.
- Pappas, Y., Anandan, C., Liu, J., Car, J., et al. (2011) Computer-assisted history-taking systems (CAHTS) in health care: benefits , risks and potential for further development. *Informatics in Primary Care*. 19155–161.
- Parsons, S. (2010) The quality of patient engagement and involvement in primary care. *Inquiry*.
- Patel, H., Bell, D., Molokhia, M., Srishanmuganathan, J., et al. (2007) Trends in hospital admissions for adverse drug reactions in England: analysis of national hospital episode statistics 1998-2005. *BMC clinical pharmacology*. [Online] 7 (1), 9. Available from: doi:10.1186/1472-6904-7-9 [Accessed: 21 March 2014].

- Pizzo, E., Lavery, a a, Phekoo, K.J., Aljuburi, G., et al. (2014) A retrospective analysis of the cost of hospitalizations for sickle cell disease with crisis in England, 2010/11. *Journal of public health (Oxford, England)*. [Online] 1–11. Available from: doi:10.1093/pubmed/fdu026.
- Post-White, J., Fitzgerald, M., Hageness, S. & Sencer, S.F. (2009) Complementary and alternative medicine use in children with cancer and general and specialty pediatrics. *Journal of pediatric oncology nursing : official journal of the Association of Pediatric Oncology Nurses*. [Online] 26 (1), 7–15. Available from: <http://goo.gl/UvOsVb> [Accessed: 8 August 2013].
- Quinn, C.T., Rogers, Z.R., McCavit, T.L. & Buchanan, G.R. (2010) Improved survival of children and adolescents with sickle cell disease. *Blood*. [Online] 115 (17), 3447–3452. Available from: doi:10.1182/blood-2009-07-233700 [Accessed: 15 July 2011].
- Raat, H., Landgraf, J.M., Bonsel, G.J., Gemke, R.J.B.J., et al. (2002) Reliability and validity of the child health questionnaire-child form (CHQ-CF87) in a Dutch adolescent population. *Quality of Life Research*. [Online] 11575–581. Available from: doi:10.1023/A:1016393311799.
- Raphael, J.L. & Oyeku, S.O. (2013) Sickle cell disease pain management and the medical home. *ASH Education Program Book*. [Online] 2013433–438. Available from: doi:10.1182/asheducation-2013.1.433.
- Renedo, A. & Marston, C. (2011) Healthcare professionals' representations of "patient and public involvement" and creation of "public participant" identities: Implications for the development of inclusive and bottom-up community participation initiatives. *Journal of Community and Applied Social Psychology*. [Online] 21268–280. Available from: doi:10.1002/casp.1092.
- Rice-Evans, C., Omorphos, S.C. & Baysal, E. (1986) Sickle cell membranes and oxidative damage. *The Biochemical journal*. [Online] 237 (1), 265–269. Available from: <http://goo.gl/T9xPkn> [Accessed: 17 August 2013].
- Richards, T. (2013) Services for patients with long term conditions must be reconfigured, says meeting. *BMJ (Clinical research ed.)*. [Online] 346 (apr12_1), f2316. Available from: doi:10.1136/bmj.f2316 [Accessed: 15 April 2014].
- Richards, T., Montori, V.M., Godlee, F., Lapsley, P., et al. (2013) Let the patient revolution begin. *BMJ*. [Online] 346 (may14_1), f2614–f2614. Available from: doi:10.1136/bmj.f2614 [Accessed: 23 March 2014].
- Richer, J. & Chudley, A.E. (2005) The hemoglobinopathies and malaria. *Clinical genetics*. [Online] 68 (4), 332–336. Available from: doi:10.1111/j.1399-0004.2005.00503.x [Accessed: 17 August 2013].
- Robinson N, Huber H & Jenkins P (2006) *Improving Access to Medical Care for Children with Sickle Cell Disease*. In: 2006 Memphis, 29th Annual Meeting of the National Sickle Cell Disease Program.
- Romero, Z., Urbinati, F., Geiger, S., Cooper, A.R., et al. (2013) β -globin gene transfer to human bone marrow for sickle cell disease. *The Journal of clinical investigation*. [Online] 123 (8), 3317–3330. Available from: <http://goo.gl/1CZBsc> [Accessed: 26 February 2014].

- Rothman, A.A. & Wagner, E.H. (2003) Chronic illness management: what is the role of primary care?. *Annals of internal medicine*. 138256–261.
- Salisbury, D., Ramsay, M. & Noakes, K. (2006) *Immunisation against infectious diseases*. 3rd Editio. [Online]. The Stationery Office. Available from: <http://goo.gl/YACg8G> [Accessed: 17 August 2013].
- Sallares, R., Bouwman, A. & Anderung, C. (2004) The spread of malaria to Southern Europe in antiquity: new approaches to old problems. *Medical history*. [Online] 48 (3), 311–328. Available from: <http://goo.gl/YdK9CC> [Accessed: 17 August 2013].
- Savitt, T.L. & Goldberg, M.F. (1989) Herrick's 1910 case report of sickle cell anemia. The rest of the story. *JAMA : the journal of the American Medical Association*. [Online] 261 (2), 266–271. Available from: <http://goo.gl/l3G87U> [Accessed: 17 August 2013].
- Saxena, S., Eliahoo, J. & Majeed, A. (2002) Socioeconomic and ethnic group differences in self study. *British Medical Journal*. 3251–6.
- Serjeant, G.R. (2000) Screening for sickle-cell disease in Brazil. *Lancet*. [Online] 356 (9224), 168–169. Available from: doi:10.1016/S0140-6736(05)73186-X [Accessed: 13 September 2013].
- Serjeant, G.R. (1997) Sickle-cell disease. *Lancet*. [Online] 350 (9079), 725–730. Available from: <http://goo.gl/056gC0> [Accessed: 17 August 2013].
- Serjeant, G.R. & Serjeant, B.E. (2001) *Sickle cell disease*. [Online]. Oxford University Press. Available from: <http://goo.gl/FQmgbm> [Accessed: 17 August 2013].
- Sibbald, B., McDonald, R. & Roland, M. (2007) Shifting care from hospitals to the community: a review of the evidence on quality and efficiency. *Journal of health services research & policy*. [Online] 12 (2), 110–117. Available from: doi:10.1258/135581907780279611 [Accessed: 2 April 2014].
- Sibinga, E.M.S., Shindell, D.L., Casella, J.F., Duggan, A.K., et al. (2006) Pediatric patients with sickle cell disease: use of complementary and alternative therapies. *Journal of alternative and complementary medicine (New York, N.Y.)*. [Online] 12 (3), 291–298. Available from: <http://goo.gl/TcGBqj> [Accessed: 17 August 2013].
- Singh, D. & Ham, C. (2006) *Improving care for people with long term conditions: a review of UK and international frameworks*.
- Steenhuysen, J. (2010) *Sickle cell disease affects brain function*. 2010. Reuters.
- Steinberg, M.H. (1999) Management of sickle cell disease. *The New England journal of medicine*. 340 (13), 1021–1030.
- Steinberg, M.H. (2002) The Management of Sickle Cell Disease. *The New England journal of medicine*. [Online]. 340 (13) pp.1021–1030. Available from: <http://goo.gl/wMPJ3F> [Accessed: 17 August 2013].
- Street, A. & Dawson, D. (2002) Costing hospital activity: The experience with healthcare resource groups in England. *European Journal of Health Economics*. [Online] 33–9. Available from: doi:10.1007/s10198-001-0086-1.

- Streetly, a, Latinovic, R., Hall, K. & Henthorn, J. (2009) Implementation of universal newborn bloodspot screening for sickle cell disease and other clinically significant haemoglobinopathies in England: screening results for 2005-7. *Journal of clinical pathology*. [Online] 62 (1), 26–30. Available from: doi:10.1136/jcp.2008.058859 [Accessed: 11 March 2013].
- Streetly, A., Clarke, M., Downing, M., Farrar, L., et al. (2008) Implementation of the newborn screening programme for sickle cell disease in England: results for 2003-2005. *Journal of medical screening*. [Online] 15 (1), 9–13. Available from: doi:10.1258/jms.2008.007063 [Accessed: 15 July 2011].
- Struthers, A.D., Anderson, G., Donnan, P.T. & MacDonald, T. (2000) Social deprivation increases cardiac hospitalisations in chronic heart failure independent of disease severity and diuretic non-adherence. *Heart (British Cardiac Society)*. [Online] 83 (1), 12–16. Available from: <http://goo.gl/z1o9AP>.
- Stuart, M.J. & Nagel, R.L. (2004) Sickle-cell disease. *Lancet*. [Online] 364 (9442), 1343–1360. Available from: <http://goo.gl/W8XzDL> [Accessed: 17 August 2013].
- Sumner, J. (2000) *The Natural History of Medicinal Plants*. [Online]. Timber Press. Available from: <http://goo.gl/FnIgtq> [Accessed: 17 August 2013].
- Takasu, J., Uykimpong, R., Sunga, M.A., Amagase, H., et al. (2006) Aged garlic extract is a potential therapy for sickle-cell anemia. *The Journal of nutrition*. [Online] 136 (3 Suppl), 803S–805S. Available from: <http://goo.gl/r1Tkmd> [Accessed: 17 August 2013].
- Telfair, J., Ehiri, J.E., Loosier, P.S. & Baskin, M.L. (2004) Transition to adult care for adolescents with sickle cell disease: Results of a national survey. *International Journal of Adolescent Medicine and Health*. 16 (1), 47–64.
- Therrell, B.L. & Hannon, W.H. (2006) National evaluation of US newborn screening system components. *Mental retardation and developmental disabilities research reviews*. [Online] 12 (4), 236–245. Available from: <http://goo.gl/YeQVLe> [Accessed: 11 October 2013].
- Tsaras, G., Owusu-Ansah, A., Boateng, F.O. & Amoateng-Adjepong, Y. (2009) Complications associated with sickle cell trait: a brief narrative review. *The American journal of medicine*. [Online] 122 (6), 507–512. Available from: <http://goo.gl/LNbdxW> [Accessed: 17 August 2013].
- Tshilolo, L., Kafando, E., Sawadogo, M., Cotton, F., et al. (2008) Neonatal screening and clinical care programmes for sickle cell disorders in sub-Saharan Africa: lessons from pilot studies. *Public health*. [Online] 122 (9), 933–941. Available from: doi:10.1016/j.puhe.2007.12.005 [Accessed: 11 October 2013].
- UK National Screening Committee (2006) *NHS Sickle Cell and Thalassaemia Screening Programme*. [Online]. 2006. Available from: <http://bit.ly/1Ffjof7> [Accessed: 10 July 2010].
- Underutilised, B.C.F. group interview: an, In, research technique for improving theory and practice & 1987;14:411–48, health education. H.E.Q. (n.d.) *No Title*.

- Nimgaonkar, V., Krishnamurti, L., Prabhakar, H., & Menon, N. (2014). Comprehensive integrated care for patients with sickle cell disease in a remote aboriginal tribal population in southern India. *Pediatric blood & cancer*, 61(4), 702-705. Available from: <http://goo.gl/rCDtRL>.
- Verduzco, L.A. & Nathan, D.G. (2009) Sickle cell disease and stroke. *Blood*. [Online] 114 (25), 5117–5125. Available from: doi:10.1182/blood-2009-05-220921. [Accessed: 17 August 2013].
- Vichinsky, B.E.P., Styles, L.A., Colangelo, L.H., Wright, E.C., et al. (1997) *Acute Chest Syndrome in Sickle Cell Disease: Clinical Presentation and Course*.
- Vichinsky, E.P., Neumayr, L.D., Gold, J.I., Weiner, M.W., et al. (2010) Neuropsychological dysfunction and neuroimaging abnormalities in neurologically intact adults with sickle cell anemia. *JAMA : the journal of the American Medical Association*. [Online] 303 (18), 1823–1831. Available from: <http://goo.gl/UDwSK0> [Accessed: 17 August 2013].
- Wagner, E.H. (1997) Managed care and chronic illness: health services research needs. *Health services research*. [Online] 32 (5), 702–714. Available from: <http://goo.gl/W1tbLk> [Accessed: 17 August 2013].
- Wagner, E.H. (1998) *Management : What Will It Take To Improve*. 2–4.
- Watts, M. & Ebbutt, D. (1987) More than the Sum of the Parts: research methods in group interviewing. *British Educational Research Journal*. [Online]. 13 pp.25–34. Available from: doi:10.1080/0141192870130103.
- WCC PPI Steering Group (2008) *Patient and Public Engagement Toolkit for World Class Commissioning*.
- Weatherall, D.J. (2010) The inherited diseases of hemoglobin are an emerging global health burden. *Blood*. [Online] 115 (22), 4331–4336. Available from: <http://goo.gl/jlr80w> [Accessed: 6 August 2013].
- WHO Secretariat (2006) *Sickle-cell anaemia*.
- Wilson, R.E., Krishnamurti, L. & Kamat, D. (2003) Management of sickle cell disease in primary care. *Clinical pediatrics*. [Online] 42 (9), 753–761. Available from: <http://goo.gl/XqC9q7> [Accessed: 19 August 2013].
- Wong, L.P. (2008) Focus group discussion: a tool for health and medical research. *Singapore medical journal*. [Online] 49256–260; quiz 261. Available from: <http://goo.gl/LPjLGg>
- Wong, W.Y. (2001) Prevention and management of infection in children with sickle cell anaemia. *Paediatric drugs*. 3793–801.
- Woods, S.S., Schwartz, E., Tuepker, A., Press, N.A., et al. (2013) Patient experiences with full electronic access to health records and clinical notes through the My HealtheVet Personal Health Record Pilot: qualitative study. *Journal of medical Internet research*. [Online] 15 (3), e65. Available from: doi:10.2196/jmir.2356 [Accessed: 16 May 2014].
- Yang, Y.M., Shah, A.K., Watson, M. & Mankad, V.N. (1995) Comparison of costs to the health sector of comprehensive and episodic health care for sickle cell disease patients. *Public health reports*. 110 (1), 80–86.

- Yanni, E., Grosse, S.D., Yang, Q. & Olney, R.S. (2009) Trends in pediatric sickle cell disease-related mortality in the United States, 1983-2002. *The Journal of pediatrics*. [Online] 154 (4), 541–545. Available from: doi:10.1016/j.jpeds.2008.09.052 [Accessed: 17 August 2013].
- Young, K. (2013) Doctors' understanding of rheumatoid disease does not align with patients' experiences. *BMJ (Clinical research ed.)*. [Online] 346 (may14_1), f2901. Available from: doi:10.1136/bmj.f2901 [Accessed: 16 April 2014].
- Yu, C.K.H., Stasiowska, E., Stephens, A., Awogbade, M., et al. (2009) Outcome of pregnancy in sickle cell disease patients attending a combined obstetric and haematology clinic. *Journal of obstetrics and gynaecology : the journal of the Institute of Obstetrics and Gynaecology*. [Online] 29 (6), 512–516. Available from: <http://goo.gl/KqQJ7U> [Accessed: 17 August 2013].
- Health watchdogs and authorities. (n.d.). Retrieved May 24, 2014, from: <http://bit.ly/1GYWRmv>
- Smart guides. (n.d.). Retrieved July 1, 2014, from: <http://bit.ly/1JC5FkZ>
- An Introduction. (n.d.). Retrieved May 24, 2014, from: <http://goo.gl/sc5ofU>

APPENDICES

Appendix 1: Costs for admissions by SHA and PCT 2010-11

Appendix 2: Patient Questionnaire

Appendix 3: Patient Comments Snapshot

Appendix 4: GP Questionnaire

Appendix 5: SCD Template installed in GP Surgeries

APPENDIX 1: COSTS FOR ADMISSIONS BY SHA AND PCT 2010-11

SHA	Co de	PCT	SCD adm. / 100,0 00 pop. (1)	COST NON- ELECTIVE (A) £	COST ELECTIVE (B) £	COST ADMISS . (A+B) £	COST EXTRA LOS (c) £	TOTAL COST (A+B+C) £
Q.30 North	5K F	Gateshead	1.54	7,693.03		7,693.03	241.91	7,934.94
Q.30 North	5K M	Middlesbrough	6.11	18,439.87	1,241.50	19,681.37	240.09	19,921.46
Q.30 North	5D 7	Newcastle	6.89	36,195.98	1,250.92	37,446.90		37,446.90
Q.30 North	5D 8	North Tyneside	0.48	3,059.07		3,059.07		3,059.07
Q.30 North	5E 1	Stockton-on-Tees	3.11	18,151.57		18,151.57		18,151.57
Q.31 North	5H G	Ashton, Leigh and Wigan	0.66	3,226.53		3,226.53		3,226.53
Q.31 North	TA P	Blackburn with Darwen	5.93	27,478.61		27,478.61		27,478.61
Q.31 North	5H Q	Bolton	1.88	11,156.80		11,156.80	992.66	12,149.45
Q.31 North	5J X	Bury	2.74	10,981.34	3,138.11	14,119.45		14,119.45
Q.31 North	5N G	Central Lancashire	1.99	16,008.56		16,008.56		16,008.56
Q.31 North	5N H	East Lancashire Teaching	0.27	3,053.18		3,053.18		3,053.18
Q.31 North	5N Q	Heywood, Middletton	5.88	31,440.38		31,440.38	738.27	32,178.65
Q.31 North	5N L	Liverpool	3.94	28,514.06	2,515.16	31,029.22		31,029.22
Q.31 North	5N T	Manchester	20.01	264,153.72	4,787.14	268,940.86	240.89	269,181.75
Q.31 North	5N F	North Lancashire T.	0.30	3,028.49		3,028.49		3,028.49
Q.31 North	5J5	Oldham	1.79	6,338.40	3,111.88	9,450.28		9,450.28
Q.31 North	5F5	Salford	4.68	17,895.42		17,895.42		17,895.42
Q.31 North	5N J	Sefton	0.37	3,087.41		3,087.41		3,087.41
Q.31 North	5F7	Stockport	1.40	6,502.53		6,502.53		6,502.53
Q.31 North	5N R	Trafford	0.93	6,620.55		6,620.55		6,620.55
Q.31 North	5J2	Warrington	1.01	6,184.42		6,184.42		6,184.42
Q.31 North	5N K	Wirral	0.33	1,594.32		1,594.32	1,945.61	3,539.93
Q.32 Yorkshire	5JE	Barnsley	0.84	3,043.71	3,043.71	6,087.42		6,087.42
Q.32 Yorkshire	5N Y	Bradford & Airedale	6.92	89,905.22	5,590.79	95,496.01		95,496.01
Q.32 Yorkshire	5J6	Calderdale	1.46	4,799.20		4,799.20		4,799.20
Q.32 Yorkshire	5N 5	Doncaster	2.02	18,349.76		18,349.76		18,349.76
Q.32 Yorkshire	5N X	Hull Teaching	4.08	30,279.89		3,090.16		3,090.16
Q.32 Yorkshire	5N 2	Kirklees	4.94	61,715.86		61,715.86		61,715.86
Q.32 Yorkshire	5N 1	Leeds	7.30	153,191.20	3,119.09	156,310.29	1,233.30	157,543.59
Q.32 Yorkshire	TA N	North East Lincolnshire	0.61	1,565.62		1,565.62		1,565.62
Q.32 Yorkshire	5N V	North Yorkshire and York	0.38	9,118.57		9,118.57		9,118.57
Q.32 Yorkshire	5H 8	Rotherham	1.22	7,670.81		7,670.81		7,670.81
Q.32 Yorkshire	5N 4	Sheffield	4.62	56,370.18	19,511.22	75,881.40		75,881.40
Q.32 Yorkshire	5N	Wakefield District	0.88	3,208.70	1,265.49	4,474.19		4,474.19

and H.	3							
Q. 33 East	5N 7	Derby City	5.64	28,385.27	3,077.73	31,463.00		31,463.00
Q. 33 East	5N 6	Derbyshire County	0.57	7,705.67	1,255.50	8,961.17		8,961.17
Q. 33 East	5P C	Leicester City	2.71	221,819.63	3,107.22	224,926.85	8,108.81	233,035.66
Q. 33 East	5P A	Leicestershire County R	3.16	38,297.65		38,297.65	1,965.77	40,263.42
Q. 33 East	5N 9	Lincolnshire Teaching	0.69	10,679.28		10,679.28	237.54	10,916.83
Q. 33 East	5E M	Nottingham City	32.47	283,754.21	8,092.28	291,846.49		291,846.49
Q. 33 East	5N 8	Nottinghamshire C.T	0.15	3,070.27		3,070.27		3,070.27
Q.34 West	5P G	Birmingham East and N.	10.73	104,037.47	1,257.54	105,295.01	243.19	105,538.21
Q.34 West	5M D	Coventry Teaching	3.57	37,170.42	1,270.55	38,440.97		38,440.97
Q.34 West	5P E	Dudley PCT	4.29	22,268.98	1,270.55	23,539.52		23,539.52
Q.34 West	5M X	Heart of Birmingham T	5.40	262,470.11	9,225.77	271,695.88	3,931.35	275,627.23
Q.34 West	5P H	North Staffordshire	0.97	3,133.15		3,133.15		3,133.15
Q.34 West	5P F	Sandwell	10.39	68,804.28	1,270.55	70,074.82	491.42	70,566.24
Q.34 West	5M 2	Shropshire County	0.69	6,041.01		6,041.01		6,041.01
Q.34 West	5Q W	Solihull	1.42	7,819.73		7,819.73		7,819.73
Q.34 West	5M 1	South Birmingham	20.10	190,790.53	4,345.80	195,136.34	491.42	195,627.76
Q.34 West	5P K	South Staffordshire	0.50	6,271.39		6,271.39		6,271.39
Q.34 West	5PJ	Stoke on Trent	3.44	25,741.22		25,741.22		25,741.22
Q.34 West	5M K	Telford and Wrekin	4.29	13,759.16	3,020.50	16,779.66		16,779.66
Q.34 West	5M 3	Walsall Teaching	4.76	33,876.12		33,876.12		33,876.12
Q.34 West	5M V	Wolverhampton City	1.24	9,316.41		9,316.41		9,316.41
Q.34 West	5P L	Worcestershire	1.45	14,133.78	1,257.54	15,391.32		15,391.32
Q.35 East of England	5P2	Bedfordshire	3.29	31,726.42	10,217.96	41,944.38	1,317.60	43,261.98
Q.35 East of England	5P P	Cambridgeshire	1.98	32,019.07	3,908.06	35,927.13		35,927.13
Q.35 East of England	5P R	Great Yarmouth and W	0.44		1,218.32	1,218.32		1,218.32
Q.35 East of England	5Q V	Hertfordshire	8.92	263,068.06	55,312.47	318,380.53	1,211.88	319,592.41
Q.35 East of England	5G C	Luton	39.41	203,152.33	3,332.28	206,484.61		206,484.61
Q.35 East of England	5P X	Mid Essex	2.42	25,239.97		25,239.97		25,239.97
Q.35 East of England	5P Q	Norfolk	0.26	4,764.59		4,764.59		4,764.59
Q.35 East of England	5P W	North East Essex	0.30	3,082.23		3,082.23		3,082.23
Q.35 East of England	5P1	South East Essex	0.30	9,672.14		9,672.14		9,672.14
Q.35 East of England	5P Y	South West Essex	3.56	41,255.26	8,715.70	49,970.96		49,970.96
Q.35 East of England	5P T	Suffolk	2.72	30,894.60	1,260.39	32,154.99	1,306.27	33,461.26
Q.35 East of England	5P V	West Essex	3.32	26,353.86		26,353.86		26,353.86
Q.36 London	5C 2	Barking and Dagenham	68.56	364,400.50	64,994.92	429,395.42	8,953.02	438,348.44
Q.36 London	5A 9	Barnet	23.06	230,222.55	12,512.72	242,735.27	2,977.07	245,712.35
Q.36 London	TA K	Bexley Care Trust	30.65	163,582.69	14,821.25	178,403.94	570.66	178,974.61
Q.36 London	5K 5	Brent Teaching	102.7 1	816,513.50	12,279.81	828,793.31	14,464.82	843,258.13
Q.36 London	5A 7	Bromley	17.00	112,356.40	15,740.26	128,096.66	290.30	128,386.95
Q.36 London	5K 7	Camden	18.20	132,341.63	28,954.52	161,296.15	6,676.22	167,972.36
Q.36 London	5C 3	City and Hackney Teach.	346.6 7	2,263,929.38	534,160.71	2,798,090.0 8	63,582.41	2,861,672.49

Q.36 London	5K9	Croydon	77.91	745,439.97	19,714.91	765,154.88	9,894.25	775,049.14
Q.36 London	5HX	Ealing	34.82	303,962.39	20,635.42	324,597.81	2,557.92	327,155.73
Q.36 London	5C1	Enfield	68.49	592,207.17	29,422.94	621,630.11		621,630.11
Q.36 London	5A8	Greenwich Teaching	70.42	419,118.03	26,249.18	445,367.22		445,367.22
Q.36 London	5H1	Hammersmith and Fulh.	48.24	238,238.83		238,238.83	4,193.96	242,432.79
Q.36 London	5C9	Haringey Teaching PCT	153.38	1,203,689.40	42,598.08	1,246,287.48	5,895.77	1,252,183.26
Q.36 London	5K6	Harrow	33.19	196,649.77	3,700.03	200,349.81		200,349.81
Q.36 London	5A4	Havering	10.94	92,387.46	1,437.84	93,825.30		93,825.30
Q.36 London	5AT	Hillingdon	14.02	89,094.73		89,094.73		89,094.73
Q.36 London	5HY	Hounslow	5.06	36,578.71	3,670.89	40,249.60		40,249.60
Q.36 London	5K8	Islington	31.80	165,137.17	50,453.93	215,591.10	14,279.81	229,870.91
Q.36 London	5LA	Kensington and Chelsea	34.38	193,403.20	34,192.20	227,595.40	532.01	228,127.41
Q.36 London	5A5	Kingston	14.89	65,249.58	7,280.35	72,529.93		72,529.93
Q.36 London	5LD	Lambeth	78.54	701,592.68	67,198.84	768,791.52	10,316.11	77,514.96
Q.36 London	5LF	Lewisham	92.44	667,443.96	57,368.46	724,812.42	13,694.84	738,507.26
Q.36 London	5C5	Newham	62.55	498,489.48	51,103.68	549,593.16	10,189.18	559,782.34
Q.36 London	5NA	Redbridge	67.95	602,773.58	32,508.31	635,281.89	23,964.23	659,246.12
Q.36 London	5M6	Richmond and Twicken.	0.53	1,918.17		1,918.17		1,918.17
Q.36 London	5LE	Southwark	60.62	461,880.46	61,307.34	523,187.80		523,187.80
Q.36 London	5M7	Sutton and Merton	28.74	252,620.20	57,982.86	310,603.05	2,548.21	313,151.26
Q.36 London	5C4	Tower Hamlets	8.90	69,996.30	7,816.12	77,812.42		77,812.42
Q.36 London	5NC	Waltham Forest	51.25	315,550.09	97,761.35	413,311.44	5,391.55	418,702.99
Q.36 London	5LG	Wandsworth	35.78	309,506.58	35,640.86	345,147.45	17,807.38	362,954.83
Q.36 London	5LC	Westminster	32.25	263,911.91	23,190.68	287,102.59	19,475.12	306,577.70
Q.37 South	5LQ	Brighton and Hove City	11.73	91,234.47	6,851.82	98,086.29		98,086.29
Q.37 South	5P7	East Sussex Downs W.	0.60	3,214.69		3,214.69		3,214.69
Q.37 South	5QA	Eastern and Coastal Kent	0.54	50,565.20	6,932.11	57,497.31		57,497.31
Q.37 South	5P8	Hastings and Rother	1.71	7,808.30		7,808.30		7,808.30
Q.37 South	5L3	Medway	9.93	62,161.74	1,333.59	63,495.33		63,495.33
Q.37 South	5P5	Surrey	2.35	88,227.56		8,227.56		88,227.56
Q.37 South	5P9	West Kent	5.69	71,170.18	4,035.28	75,205.46	580.59	75,786.05
Q.37 South	5P6	West Sussex	2.36	,060.35	7,816.12	54,876.47	251.55	55,128.02
Q. 38 South	5QG	Berkshire East	1.98	19,666.46	3,196.18	22,862.64		22,862.64
Q. 38 South	5QF	Berkshire West	6.85	94,427.94		94,427.94	1,082.14	95,510.08
Q. 38 South	5QD	Buckinghamshire	3.72	49,435.48	7,051.63	56,487.11		56,487.11
Q. 38 South	5QC	Hampshire	0.47	14,625.91		14,625.91		14,625.91
Q. 38 South	5QT	Isle of Wight NHS	0.70	3,075.99		3,075.99		3,075.99
Q. 38 South	5CQ	Milton Keynes	0.41	3,279.96		3,279.96		3,279.96
Q. 38 South	5QE	Oxfordshire	4.73	67,009.44	8,047.49	75,056.93		75,056.93
Q. 38 South	5FE	Portsmouth City Teaching	3.27	20,859.09		20,859.09		20,859.09
Q. 38 South	5L1	Southampton City	9.71	66,712.88	6,326.39	73,039.27		73,039.27
Q.39 South	5FL	Bath and North East Som.	7.36	36,330.77	9,567.15	45,897.92		45,897.92

Q.39 South	5Q J	Bristol	17.02	180,505.09	33,917.74	214,422.84	8,768.54	223,191.38
Q.39 South	5Q Q	Devon	0.53	12,038.89		12,038.89		12,038.89
Q.39 South	5Q H	Gloucestershire	2.85	38,148.60	3,189.05	41,337.65		1,337.65
Q.39 South	5M 8	North Somerset	0.47	3,189.05		3,189.05		3,189.05
Q.39 South	5F1	Plymouth Teaching	4.44	20,585.88	2,608.15	23,194.02		3,194.02
Q.39 South	5A 3	South Gloucestershire	2.34	15,945.25	1,304.07	17,249.32		17,249.32
Q.39 South	5K 3	Swindon	0.48	1,594.40		1,594.40		1,594.40
Q.39 South	5Q K	Wiltshire	0.22	1,589.00		1,589.00		1,589.00

(1) The rate is given by the number of admissions in emergency and elective over the number of population in the PCT for 2011.

(2) Source of population at PCT level: NHS 2011 <https://goo.gl/hPkQ67> (Pizzo, Lavery, Phekoo, et al., 2014)

SECTION 1: DEMOGRAPHIC INFORMATION

- What year did you qualify as a physician?
- Did you qualify in the UK? Yes ☐ No ☐
- How many full-time equivalent GPs work in your practice?
- How many full-time equivalent nurses work in your practice?
- What is your practice list size?
- Do you hold any of the qualifications listed (tick all that apply):
 - MRCGP/FRCGP ☐
 - MRCP/FRCP ☐
- Is your practice:
 - Postgraduate training practice ☐
 - Undergraduate teaching practice ☐
- Which, if any, clinical computer system or electronic patient record (EPR) system does your practice use?
 - EMIS ☐
 - Vision ☐
 - System One ☐
 - Other ☐
 - None ☐
- Does your practice use an electronic disease register with specific read codes for haemoglobinopathies?
 - Yes ☐
 - No ☐

- Does your practice use a computer driven call/recall system to identify patients with long-term illness due for a periodic review?

Yes ☐

No ☐

- GP name and practice address

.....
.....
..... Post Code

SECTION 2: SCREENING and IMMUNISATIONS

Question 1.

Do you arrange for high risk patients to be screened for haemoglobinopathies if there is no record of prior testing?

Yes ☐

No ☐

Question 2.

- a. For children with SCD, do you keep a record in the EPR system of whether they have undergone Transcranial Doppler Ultrasonography (TCD) screening?

Yes ☐

No ☐

- b. If there is no record, would you seek advice from a paediatric haematologist to ensure they have received a TCD?

Yes ☐

No ☐

Question 3.

For your patients with SCD, do you liaise with a haematologist to ensure they receive regular comprehensive medical exams including screening for renal, pulmonary, and liver disease?

Yes ☐

No ☐

Question 4.

- a. Do you carry out an annual general health review of your SCD patients or keep a record of the results of their annual review with a haematologist?

Yes ☐

No ☐

- b. If YES, please tick what items are measured at these annual reviews:

() height () weight () blood pressure () urinalysis () liver function
() cholesterol () glucose () renal function () smoking status ()
() alcohol intake

Question 5.

Which of the following vaccines do you administer as a standard practice to your SCD patients?

☐ Influenza Vaccine for above age 6 months ☐ pneumococcal conjugate vaccine (PREVNAR)

☐ Pneumovax vaccine at 2 years ☐ Meningococcal vaccine at age 2 year + with follow-up dose at 7 and 12 yrs

☐ Hepatitis B ☐ Other (please state).....

SECTION 3: INTERVENTIONS

Question 6.

How often do you prescribe:

- a) For children 5 and under, twice Daily Penicillin V or an equal antibiotic as prophylaxis

Never	Rarely (1-2/year)	Sometimes (3-4/year)	Regularly (Monthly)
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- b) Folic acid, zinc and/or other supplements

Never	Rarely (1-2/year)	Sometimes (3-4/year)	Regularly (Monthly)
-------	----------------------	-------------------------	------------------------

- c) Analgesia or other pain control protocol

Never	Rarely (1-2/year)	Sometimes (3-4/year)	Regularly (Monthly)
-------	----------------------	-------------------------	------------------------

- d) Other (please specify)

.....

Question 7.

For patients who experience frequent vaso-occlusive painful episodes (3 or more per year), would you consider referring to a haematologist to see if they would benefit from hydroxyurea therapy?

Yes ☐

No ☐

Question 8.

How confident are you with your clinical knowledge of SCD and the treatment options given to your patient to manage their disease?

0 1 2 3 4 5 6 7 8 9 10

(0=Not confident at all) (10=Very confident)

Question 9.

List the kind of support you **currently** give to your patients or ensure they receive this from a specialist clinic

- | | |
|--|--------------------------|
| Pain Control | ☐ |
| Prescriptions | ☐ |
| General SCD advice | ☐ |
| General health and lifestyle advice | ☐ |
| Contraception advice | ☐ |
| Genetic and pre-conceptual counselling | ☐ |
| Immunisation | ☐ |
| Specialist Referral | ☐ |
| Other (please state) | ☐ |

Question 10.

List the kind of support or intervention you would **like** to receive from the NHS in order to better manage SCD at the primary care level or any other comments you may have

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End of questionnaire

APPENDIX 3: PATIENT COMMENTS SNAPSHOT

Question 3

Any other comments

The GP should know
more about sickle cell.

Question 3

Any other comments

I would like my GP & haematology
to help more with my SCA

SECTION 1: DEMOGRAPHIC INFORMATION

- What year did you qualify as a physician?
- Did you qualify in the UK? Yes ☐ No ☐
- How many full-time equivalent GPs work in your practice?
- How many full-time equivalent nurses work in your practice?
- What is your practice list size?
- Do you hold any of the qualifications listed (tick all that apply):
 - MRCGP/FRCGP ☐
 - MRCP/FRCP ☐
- Is your practice:
 - Postgraduate training practice ☐
 - Undergraduate teaching practice ☐
- Which, if any, clinical computer system or electronic patient record (EPR) system does your practice use?
 - EMIS ☐
 - Vision ☐
 - System One ☐
 - Other ☐
 - None ☐
- Does your practice use an electronic disease register with specific read codes for haemoglobinopathies?
 - Yes ☐
 - No ☐
- Does your practice use a computer driven call/recall system to identify patients with long-term illness due for a periodic review?
 - Yes ☐
 - No ☐

- GP name and practice address

.....
.....

Post Code.....

SECTION 2: SCREENING and IMMUNISATIONS

Question 1.

Do you arrange for high risk patients to be screened for haemoglobinopathies if there is no record of prior testing?

Yes ☐

No ☐

Question 2.

- a. For children with SCD, do you keep a record in the EPR system of whether they have undergone Transcranial Doppler Ultrasonography (TCD) screening?

Yes ☐

No ☐

- b. If there is no record, would you seek advice from a paediatric haematologist to ensure they have received a TCD?

Yes ☐

No ☐

Question 3.

For your patients with SCD, do you liaise with a haematologist to ensure they receive regular comprehensive medical exams including screening for renal, pulmonary, and liver disease?

Yes ☐

No ☐

Question 4.

- a. Do you carry out an annual general health review of your SCD patients or keep a record of the results of their annual review with a haematologist?

Yes ☐

No ☐

- b. If YES, please tick what items are measured at these annual reviews:

() height () weight () blood pressure () urinalysis () liver function
() cholesterol () glucose () renal function () smoking status () alcohol intake

Question 5.

Which of the following vaccines do you administer as a standard practice to your SCD patients?

☐ Influenza Vaccine for above age 6 months ☐ pneumococcal conjugate vaccine (PREVNAR)

☐ Pneumovax vaccine at 2 years ☐ Meningococcal vaccine at age 2 year + with follow-up dose at 7 and 12 yrs ☐ Hepatitis B ☐ Other (please state).....

SECTION 3: INTERVENTIONS

Question 6.

How often do you prescribe:

- a) For children 5 and under, twice Daily Penicillin V or an equal antibiotic as prophylaxis

Never	Rarely (1-2/year)	Sometimes (3-4/year)	Regularly (Monthly)
-------	----------------------	-------------------------	------------------------

- b) Folic acid, zinc and/or other supplements

Never	Rarely (1-2/year)	Sometimes (3-4/year)	Regularly (Monthly)
-------	----------------------	-------------------------	------------------------

- c) Analgesia or other pain control protocol

Never	Rarely (1-2/year)	Sometimes (3-4/year)	Regularly (Monthly)
-------	----------------------	-------------------------	------------------------

- d) Other (please specify)

.....

Question 7.

For patients who experience frequent vaso-occlusive painful episodes (3 or more per year), would you consider referring to a haematologist to see if they would benefit from hydroxyurea therapy?

Yes ☐

No ☐

Question 8.

How confident are you with your clinical knowledge of SCD and the treatment options given to your patient to manage their disease?

0 1 2 3 4 5 6 7 8 9 10

(0=Not confident at all) (10=Very confident)

Question 9.

List the kind of support you **currently** give to your patients or ensure they receive this from a specialist clinic

- | | |
|--|--------------------------|
| Pain Control | ☐ |
| Prescriptions | ☐ |
| General SCD advice | ☐ |
| General health and lifestyle advice | ☐ |
| Contraception advice | ☐ |
| Genetic and pre-conceptual counselling | ☐ |
| Immunisation | ☐ |
| Specialist Referral | ☐ |
| Other (please state) | ☐ |

Question 10.

List the kind of support or intervention you would **like** to receive from the NHS in order to better manage SCD at the primary care level or any other comments you may have

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End of questionnaire

APPENDIX 5: SCD TEMPLATE INSTALLED IN GP SURGERIES

Inbox - Microsoft Outlook

LV for Windows (C) 2001 EMIS

File Edit View Macros Settings Favourites Help

Modules

nGMS Registers : 0 Alerts: 0 EDI: 43 PN: 3 Email: 0 Repeats Req: 0

No.3357. Mr Test Edi, Acton Lane London Age 63 years PC

Template For Sickle-Cell Anaemia

Prompt	Result	Date	Last Recorded Entry
Sickle-cell anae	(Y or N)		Sickle-cell anae -----
Pain control			Pain control -----
Text			Text -----
Folic acid proph			Folic acid proph -----
Prophylactic ant			Prophylactic ant -----
Antibiotic			Antibiotic -----
Contraception			Contraception -----
Type			Type -----
Priapism			Priapism -----
Erectile dysfunc			Erectile dysfunc -----

<Page Up> for all past data , <File> to file <Shift Arrow> to page

Network link NUM Fri 08 Oct 2010 16:25 PC-Dr P T Chatlani PDS-2.3 WS-N Line-708 Job-19 Tv-3

Start Inbox - Microsoft Outlook LV5.2 EMIS Server LV5.2 EMIS Server 16:25

Document3 - Microsoft Word

LV for Windows (C) 2001 EMIS

File Edit View Macros Settings Favourites Help

Modules

nGMS Registers : 0 Alerts: 0 EDI: 43 PN: 3 Email: 0 Repeats Req: 0

No.3357. Mr Test Edi, Acton Lane London Age 63 years PC

Template For Sickle-Cell Anaemia

Prompt	Result	Date	Last Recorded Entry
O/E - height	cm		250 cm 21.5.2009
Ideal weight			143.8 Kg 21.5.2009
O/E - weight			80 Kg 26.10.2002
Body Mass Index			26.1 26.10.2002
Systolic BP			160 11.10.2007
Diastolic BP			20 mm Hg 11.10.2007
Cardiovasc exam			Cardiovasc exam -----
Text			
Respiratory exam			Respiratory exam -----
Text			

Enter Yes No

Network link NUM Fri 08 Oct 2010 16:25 PC-Dr P T Chatlani PDS-2.3 WS-N Line-708 Job-19 Tv-3

Start Inbox - Microsoft Outlook LV5.2 EMIS Server LV5.2 EMIS Server Document3 - Microsoft W... 16:25

Document3 - Microsoft Word

LV for Windows (C) 2001 EMIS

File Edit View Macros Settings Favourites Help

Modules

nGMS Registers: 0 Alerts: 0 EDI: 43 PN: 3 Email: 0 Repeats Req: 0

No.3357. Mr Test Edi, Acton Lane London Age 63 years PC

Template For Sickle-Cell Anaemia

Prompt	Result	Date	Last Recorded Entry
SaO2 %	(Y or N)		SaO2 -----
GI/Abdo exam Text			GI/Abdo exam -----
Non-pressure ulc Text			Non-pressure ulc -----

Network link NUM Fri 08 Oct 2010 16:25 PC-Dr P T Chatlani PDS-2.3 WS-N Line-708 Job-19 Tv-3

Start Inbox - Microsoft Outlook LV5.2 EMIS Server LV5.2 EMIS Server Document3 - Microsoft W... 16:25

Document3 - Microsoft Word

LV for Windows (C) 2001 EMIS

File Edit View Macros Settings Favourites Help

Modules

nGMS Registers: 0 Alerts: 0 EDI: 43 PN: 3 Email: 0 Repeats Req: 0

No.3357. Mr Test Edi, Acton Lane London Age 63 years PC

Template For Sickle-Cell Anaemia

Prompt	Result	Date	Last Recorded Entry
Ophth referral	(Y or N)		Ophth referral -----
Consultants			
Hospitals			
Place of Procedu			
Consultants			
Department			
Hospital			

Network link NUM Fri 08 Oct 2010 16:26 PC-Dr P T Chatlani PDS-2.3 WS-N Line-708 Job-19 Tv-3

Start Inbox - Microsoft Outlook LV5.2 EMIS Server LV5.2 EMIS Server Document3 - Microsoft W... 16:26

Document3 - Microsoft Word

LV for Windows (C) 2001 EMIS

File Edit View Macros Settings Favourites Help

Modules

nGMS Registers : 0 Alerts: 0 EDI: 43 PN: 3 Email: 0 Repeats Req: 0

No.3357. Mr Test Edi, Acton Lane London Age 63 years PC

Template For Sickle-Cell Anaemia

Prompt	Result	Date	Last Recorded Entry
Orthopaed referr	(Y or N)		Orthopaed referr -----
Consultants			
Hospitals			
Place of Procedu			
Social Serv Ref			Social Serv Ref -----
Place of Procedu			
Influenza vaccin			Influenza vaccin -----
Place of Procedu			
Text			

Network link NUM Fri 08 Oct 2010 16:26 PC-Dr P T Chatlani PDS-2.3 WS-N Line-708 Job-19 Tv-3

Start Inbox - Microsoft Outlook LV5.2 EMIS Server LV5.2 EMIS Server Document3 - Microsoft W... 16:26

Document3 - Microsoft Word

LV for Windows (C) 2001 EMIS

File Edit View Macros Settings Favourites Help

Modules

nGMS Registers : 0 Alerts: 0 EDI: 43 PN: 3 Email: 0 Repeats Req: 0

No.3357. Mr Test Edi, Acton Lane London Age 63 years PC

Template For Sickle-Cell Anaemia

Prompt	Result	Date	Last Recorded Entry
1st PCV	(Y or N)		1st PCV -----
Place of Procedu			
Text			
2nd PCV			2nd PCV -----
Place of Procedu			
Text			
3rd PCV			3rd PCV -----
Place of Procedu			
Text			

Network link NUM Fri 08 Oct 2010 16:26 PC-Dr P T Chatlani PDS-2.3 WS-N Line-708 Job-19 Tv-3

Start Inbox - Microsoft Outlook LV5.2 EMIS Server LV5.2 EMIS Server Document3 - Microsoft W... 16:26

Document3 - Microsoft Word

LV for Windows (C) 2001 EMIS

File Edit View Macros Settings Favourites Help

Modules

nGMS Registers : 0 Alerts: 0 EDI: 43 PN: 3 Email: 0 Repeats Req: 0

No.3357. Mr Test Edi, Acton Lane London Age 63 years PC

Template For Sickle-Cell Anaemia

Prompt	Result	Date	Last Recorded Entry
Boost hep B vac	(Y or N)		Boost hep B vac -----
Place of Procedure Text			
Pneumococcc Vac			Pneumococcc Vac -----
Place of Procedure Text			
First men C vacc			First men C vacc -----
Place of Procedure Text			

Network link NUM Fri 08 Oct 2010 16:27 PC-Dr P T Chatlani PDS-2.3 WS-N Line-708 Job-19 Tv-3

Start Inbox - Microsoft Outlook LV5.2 EMIS Server LV5.2 EMIS Server Document3 - Microsoft W... 16:27

Document3 - Microsoft Word

LV for Windows (C) 2001 EMIS

File Edit View Macros Settings Favourites Help

Modules

nGMS Registers : 0 Alerts: 0 EDI: 43 PN: 3 Email: 0 Repeats Req: 0

No.3357. Mr Test Edi, Acton Lane London Age 63 years PC

Template For Sickle-Cell Anaemia

Prompt	Result	Date	Last Recorded Entry
Second menC vacc	(Y or N)		Second menC vacc -----
Place of Procedure Text			Place of Procedure -----
Hib/menC vacc			Hib/meningitis 13.5.2008
Place of Procedure Text			G.P.Surgery
1st HiB vacc			1st HiB vacc -----
Place of Procedure Text			

Network link NUM Fri 08 Oct 2010 16:27 PC-Dr P T Chatlani PDS-2.3 WS-N Line-708 Job-19 Tv-3

Start Inbox - Microsoft Outlook LV5.2 EMIS Server LV5.2 EMIS Server Document3 - Microsoft W... 16:27

Document3 - Microsoft Word

LV for Windows (C) 2001 EMIS

File Edit View Macros Settings Favourites Help

Modules

nGMS Registers : 0 Alerts: 0

EDI: 43 PN: 3 Email: 0 Repeats Req: 0

No.3357. Mr Test Edi, Acton Lane London

Age 63 years PC

MR

Template For Sickle-Cell Anaemia

Prompt	Result	Date	Last Recorded Entry
2nd HiB vacc	(Y or N)		2nd HiB vacc -----
Place of Procedu Text			
3rd HiB vacc			3rd HiB vacc -----
Place of Procedu Text			
1st hepB vacc			1st hepB vacc -----
Place of Procedu Text			

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Yes No

Network link NUM Fri 08 Oct 2010 16:27 PC-Dr P T Chatlani PDS-2.3 WS-N Line-708 Job-19 Tv-3

Start

Inbox - Microsoft Outlook

LV5.2 EMIS Server

LV5.2 EMIS Server

Document3 - Microsoft W...

16:27

Poster presentations resulting from this work

Development and Implementation of Sickle Cell Disease Template in Primary Care Services in Northwest London

Okoye O, Amobi C, Chatlani P, Ansari S, Oni L, Aljuburi G, Green SA, Cho G, Phekoo K.J, Khan I, Nkohkwo A, Ojeer P, Ndive C, Majeed A, Knight Y, Anie K, Banarsee R

The study aims to improve the quality of care for patients with SCD in primary care through:

- Improving GPs knowledge about SCD
- Raising awareness of new UK care standards
- Engagement and empowerment of patients with SCD

Background

Sickle Cell Disease (SCD) is the most common and fastest growing genetic blood disorder in England, predominantly affecting Black and ethnic minority (BME) groups. SCD is a most common reason to be admitted to hospital for this patient group and has a high rate of multiple admissions for individual patients.

Problem

National confidential enquiry into patient outcomes and deaths (2008) was critical of the role of GPs in increasing vaccination and antibiotic prophylaxis uptake to prevent sepsis from hypo-splenism and recommended that management in primary care needs to be improved. In addition it had been observed that many patients with SCD were using the emergency departments of local hospitals rather than consult with their GPs.

Intervention

This study was led by NHS Brent and consisted of a multidisciplinary team from the NHS, academia and the voluntary sector including consultant haematologists, GPs, a specialist nurse, public health specialists and researchers. A multi-component intervention was designed which comprised :

- A GP training programme
- A GP SCD clinical management handbook
- Primary Care electronic template
- Awareness raising of primary care services with patients and carers

The intervention was designed following engagement with patients and carers, through the voluntary sector and primary and secondary care staff.

The Development and implementation of SCD template was championed by GPs, the template captures key information on UK paediatric and adult standards.



Measuring for improvement

The template was reviewed by primary care staff and users at training sessions, and was piloted on 3 GP practices prior to wider roll out. All target practices 100% had templates installed and received training on usage. 100% identified SCD patients on their system. The baseline of ED admissions has been established for each practice this will be used to evaluate the effect of the intervention of practices that have engaged.

Effects of change

Sickle Cell leads within practices championing audits and review of services for sickle cell patients on their list.

A GP conducted an audit of SCD patients in her surgery, identifying at-risk childbearing population, within a day of attending. The GP also liaised with a specialist nurse consultant to obtain health promotion materials and devised template letter sent to patients to improve uptake of pre-conception screening and specialist care.

Conclusion

The SCD template provided an effective tool for recording patient information in a structured and consistent way.

In addition the project facilitated joint working between primary & secondary care and patients; identified practical approaches for joint working to improve user experience.

It gives users and primary care staff confidence and clarity in navigating the system.

Lessons learnt

Securing GP champions is key for peer leadership and ongoing engagement. It is important to involve practice managers early on in the project. They play a crucial in implementing data systems. Incentives are beneficial to

enable staff to focus on the additional work required.

Acknowledgements: Dr Mabel Alli North West London Haemoglobinopathy Network Coordinator (deceased), Harness GPs, sickle cell patients and carers and the Sickle Cell Steering Committee for their dedication to a project that focuses on improving patient quality of care at the primary care level. **Contact:** Dr Ogo Okoye, NHS Brent – ogo.okoye@brent-harrowpcts.nhs.uk

Engaging Sickle Cell Patients and Carers in Improving Primary Care Services in Northwest London

Okoye O, Oni L, Alli M, Aljuburi G, Green SA, Cho G, Phekoo KI, Nkohkwo A, Ojeer P, Ndive C, Amobi C, Majeed A, Chatlani P, Knight Y, Nortey P, Anie K, Ansai S, Banarsee R

The study aims to improve the quality of care for patients with SCD in primary care through:

- Improving GPs knowledge about SCD
- Raising awareness of new UK care standards
- Engagement and empowerment of patients with SCD

Background

Sickle Cell Disease (SCD) is the most common and fastest growing genetic blood disorder in England, predominantly affecting Black and ethnic minority (BME) groups. SCD is a most common reason to be admitted to hospital for this patient group and has a high rate of multiple admissions for individual patients.

Problem

National confidential enquiry into patient outcomes and deaths (2008) was critical of the role of GPs in increasing vaccination and antibiotic prophylaxis uptake to prevent sepsis from hypo-splenism and recommended that management in primary care needs to be improved. In addition it had been observed that many patients with SCD were using the emergency departments of local hospitals rather than consult with their GPs.

Intervention

This study was led by NHS Brent and consisted of a multidisciplinary team from the NHS, academia and the voluntary sector including consultant haematologists, GPs, a specialist nurse, public health specialists and researchers. A multi-component intervention was designed which comprised of:

- A GP training programme
- A GP SCD clinical management handbook
- Primary Care electronic template
- Awareness raising of primary care services with patients and carers

The intervention was designed following engagement with patients and carers, through the voluntary sector. In addition primary and secondary care health professionals were involved in developing and delivering the handbook and training programme.

The delivery of an educational package to primary care staff with hospital doctors and users as trainers tailored to improve clinical knowledge as well as holistic care. Development and implementation of SCD template led by GPs, the template captures key information on UK paediatric and adult standards.



Measuring for improvement

The training programme, handbook and the electronic patient template, which sits within the GPs current system, were delivered to a group of GP practices within Brent. The baseline of ED admissions has been established for each practice for the 2 years preceding the delivery of the intervention, this will be used to evaluate the effect of the intervention of practices that have engaged with the training and materials.

Effects of change

Immediate change in clinical practice was demonstrated as one GP conducted an audit of SCD patients in her surgery, identifying at-risk childbearing population, within a day of attending. The GP also liaised with a specialist nurse consultant to obtain health promotion materials and devised template letter sent to patients to improve uptake of pre-conception screening and specialist care. Sickle Cell leads within practices championing audits and review of services for sickle cell patients on their list.

Conclusion

The project facilitated joint working between primary & secondary care and patients; identified practical approaches for joint working to improve user experience.

It gives users confidence and clarity in navigating the system. One user's views about being part of the training team for primary care staff: "I have not really seen my GP for ages; it's good my doctors are talking together, now I can see when and why I need to see my GP."

Lessons learnt

Engagement of multidisciplinary staff takes time, important to build in time and enough flexibility to respond to competing priorities/changes in staff roles. Securing GP champions will be key for peer leadership and on-going engagement.

Acknowledgements: We thank the Sickle Cell Steering Committee for their dedication to a project that focuses on improving patient quality of care at the primary care level for an under served population. Contact: Dr Ogo Okoye, Head of Public Health Delivery, NHS Brent - ogo.okoye@brentpct.nhs.uk

Publications resulting from this work

- Aljuburi, G., Lavery, A.A., Green, S.A., Phekoo, K.J., et al. (2013b) Socio-economic deprivation and risk of emergency readmission and inpatient mortality in people with sickle cell disease in England: observational study. *Journal of public health (Oxford, England)*. [Online] 35 (4), 510–517. Available from: <http://goo.gl/R55Ve7> [Accessed: 21 March 2014].
- Aljuburi, G., Lavery, A.A., Green, S.A., Phekoo, K.J., et al. (2012a) Trends in hospital admissions for sickle cell disease in England, 2001/02-2009/10. *Journal of public health (Oxford, England)*. [Online] 34 (4), 570–576. Available from: doi:10.1093/pubmed/fds035.
- Aljuburi, G., Okoye, O., Majeed, A., Knight, Y., et al. (2012) Views of patients about sickle cell disease management in primary care: a questionnaire-based pilot study. *JRSM short reports*. [Online] 3 (11), 78. Available from: doi:10.1258/shorts.2012.011173.
- Aljuburi, G., Phekoo, K.J., Okoye, N.O., Anie, K., et al. (2013a) Patients' views on improving sickle cell disease management in primary care: focus group discussion. *JRSM Short Reports*. [Online] 3 (12), 84–84. Available from: doi:10.1258/shorts.2012.011153.
- Green, S.A., Aljuburi, G., Majeed, A., Okoye, O., et al. (2012a) Characterizing emergency admissions of patients with sickle cell crisis in NHS Brent: observational study. *JRSM short reports*. [Online] 3 (6), 37. Available from: doi:10.1258/shorts.2012.011129.
- Pizzo, E., Lavery, a a, Phekoo, K.J., Aljuburi, G., et al. (2014) A retrospective analysis of the cost of hospitalizations for sickle cell disease with crisis in England, 2010/11. *Journal of public health (Oxford, England)*. [Online] 1–11. Available from: doi:10.1093/pubmed/fdu026.

Socio-economic deprivation and risk of emergency readmission and inpatient mortality in people with sickle cell disease in England: observational study

Ghida AlJuburi¹, Anthony A. Lavery¹, Stuart A. Green², Karen J. Phekoo², Derek Bell², Azeem Majeed¹

¹Department of Primary Care and Public Health, Imperial College London, London W6 8RP, UK

²NIHR CLAHRC for Northwest London, Imperial College London, London SW10 9NH, UK

Address correspondence to Ghida AlJuburi, E-mail: g.aljuburi07@imperial.ac.uk

ABSTRACT

Background Sickle cell disease (SCD) is a cause of frequent emergency readmissions. We examined trends in SCD emergency readmissions and inpatient mortality in England in relation to socio-economic status.

Methods Data from Hospital Episode Statistics were extracted for all SCD patients admitted in 2005/06. The financial year 2005/06 was taken as the index year for analysis. We calculated readmission rates and inpatient mortality for patients admitted with a primary or secondary diagnosis of sickle cell anaemia with crisis and without crisis in the index year during the subsequent 5 years (2006/07–2010/11). Charlson Score was used to measure comorbidity. Using Cox proportional hazards models, we also examined the relationship between patient characteristics and both emergency readmissions and inpatient mortality.

Results In 2005/06, there were 7679 SCD index admissions. Over the subsequent 5-year period, patients living in the most socio-economically deprived areas were at highest risk of readmission (54.2% readmitted over the study period compared with 28% of the least deprived group). Inpatient mortality amongst readmissions was highest in patients living in the most deprived areas [hazard ratio (HR) 2.34, 95% CI 1.41–3.90].

Conclusion SCD patients from the most socio-economically deprived areas and with comorbidities are at highest risk of both SCD readmissions and in-hospital mortality, suggesting that there are inequalities in healthcare access and health outcomes amongst people with SCD.

Keywords chronic disease, management and policy, socioeconomics factors

Introduction

Sickle cell disease (SCD) is a group of inherited blood disorders in which recurring attacks of acute painful episodes (also known as a sickle crisis or vaso-occlusive crisis) are the most common reason for hospital admissions^{1–4} and the cause of frequent hospitalizations.^{5,6} SCD patients with high rates of painful episodes and frequent hospitalizations have also been found to have a higher mortality than those with low rates.⁷

Hospital admissions for SCD in England have increased substantially in recent years, particularly in London.⁸ Although health outcomes have improved with more patients now surviving into adult life, many patients still experience emergency admissions for complications of SCD, with many patients also experiencing readmissions.^{5,6} Identifying factors that predict

risk of readmission in SCD patients could help in targeting groups particularly at risk, thereby improving their health outcomes, quality of life and reducing the burden on the National Health Service (NHS) from SCD.

Few prior studies have examined the healthcare utilization of SCD patients in England. In a recent study, we showed that

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admission rates for SCD in England are increasing; and wide variations in admission rates amongst primary care trusts, especially in London exist.⁸ There has been limited research in England examining the influence of socio-economic characteristics on the risk of SCD admission.¹⁰ The aim of this study was to investigate trends in the rates of emergency readmissions in England for patients with SCD, to determine inpatient mortality and to assess whether there is an association between deprivation and comorbidity with risk of readmission and inpatient mortality.

Methods

We obtained data over a period of 6 years from the national Hospital Episode Statistics (HES) database (April 2005 – March 2011). HES is the national administrative database for

hospital activity in England and contains data on all admissions and outpatient appointments in the NHS, including patients whose treatment is funded by the NHS but performed in private hospitals.¹¹ HES data contain details on the diagnoses of every patient admitted to an NHS hospital in England; diagnoses are coded using the International Classification of Diseases version 10 (ICD 10). Patients can be coded as having up to 20 different diagnoses. This analysis included patients with a primary diagnosis or secondary diagnosis of *sickle cell anaemia with crisis* or *sickle cell anaemia without crisis*. The financial year 2005/06 was taken as the index year for this analysis, and all patients admitted with a primary or a secondary diagnosis of SCD in this year were identified and classed as SCD patients in our cohort. Secondary diagnoses are only meant to be coded if they are related to the reason for admission. For patients who were admitted more than once in the index year, their first admission was used as the index admission.

Pseudonymized patient identifiers were used to identify subsequent hospital admissions for the patients identified in the index year. Outcome variables used were emergency hospital admissions, in-hospital mortality and emergency readmissions to hospital. Predictor variables used in this analysis were age group, sex, national deprivation group (based on lower super output area index of multiple deprivation ranks for the whole of England)¹² from 1 (most deprived) to 4 (least deprived), whether the index diagnosis was a primary or secondary diagnosis for SCD, whether the index diagnosis was for an SCD crisis or not (among those with a primary diagnosis of SCD) and Charlson comorbidity index score. The Charlson score classifies patients into groups depending on the number and severity of their comorbidities on admission, with higher Charlson scores reflecting patients having a higher risk of mortality within 10 years.¹³ For multivariate analysis, a combination diagnosis variable was created to compare those admitted with SCD as a secondary diagnosis; those admitted with a primary diagnosis of non-crisis and those admitted with a primary diagnosis of a crisis.

Descriptive statistics for the number of admissions and deaths in an index year were first produced and then summarized for study years individually. Cox proportional hazards models were used to examine the association between patient demographic variables and the likelihood of emergency admission over the time period as well as in-hospital mortality. For patients admitted more than once during the study period, only the first readmission was used. All of the variables mentioned previously were used in these models. All statistical analyses were performed using the programme STATA^W version 12.

Table 1 Characteristics of patients admitted and inpatient mortality with a primary or secondary diagnosis of *sickle cell anaemia with crisis* or *sickle cell anaemia without crisis* in index year (2005/06)

	Number	%	Deaths	%
Age group				
<1	142	1.8	0	0.0
1–9	931	12.1	1	0.1
10–19	1122	14.6	1	0.1
20–29	1736	22.6	6	0.3
30–39	1739	22.6	6	0.3
40–49	1073	14.0	9	0.8
50 plus	936	12.2	31	3.3
Sex				
Male	3049	39.7	32	1.0
Female	4630	60.3	22	0.5
National deprivation group				
1 (most deprived)	4454	58.0	30	0.7
2 (second most deprived)	1791	23.3	16	0.9
3 (second least deprived)	708	9.2	2	0.3
4 (least deprived)	726	9.5	6	0.8
Initial diagnosis				
Primary	2681	34.9	9	0.3
Secondary	4998	65.1	45	0.9
Index admission for crisis ^a				
Yes	1955	25.5	6	0.3
No	726	9.5	3	0.4
Charlson comorbidity index				
0	6265	81.6	10	0.2
1	819	10.7	11	1.3
2::2	595	7.7	33	5.5
Total	7679		54	0.7

^aThis includes only patients who had an initial primary diagnosis of SCD.

Results

The characteristics of the patients admitted for SCD in 2005/06 (the index year) can be seen in Table 1 ($n = 7679$). The majority (58.1%) live in the most deprived quartile of lower super output areas nationally, with only 9.5% of admissions coming from patients living in the least deprived quartile. The majority (65%) involved SCD as a secondary diagnosis, and among those admitted with SCD as a primary diagnosis, the majority were for an SCD crisis (73%). Overall, the percentage of deaths in the index year was low at 0.4%. Deaths were higher among those with SCD as a secondary diagnosis compared with primary diagnosis (0.6 versus 0.1%), and among those with a Charlson comorbidity score of 2 or more than a Charlson score of 0 (2.7 versus 0.1%).

Figure 1 shows the unadjusted cumulative time to readmission curves by national deprivation group among those with a primary admission for an SCD crisis ($n = 1964$). Patients in the least deprived group were less likely to be readmitted over the whole time period compared with all other levels of deprivation. For example, 39.6% of the patients in the most deprived group were admitted at least once within 2 years, compared with 20.2% for the least deprived group (data not shown).

Table 2 shows the annual percentages of readmissions ($n = 7679$) by the demographic variables. Over the whole period, those patients aged 10–19 had the highest risk of readmission (65.5%), as did those with SCD as a primary

diagnosis (74%) and those with an index admission for an SCD crisis (77.5%). Those in the national deprivation group 1 (most deprived) were more likely to have a readmission over the whole period than those in national deprivation group 4, the least deprived group (54.2 versus 28.0%).

Table 3 shows the annual in-hospital mortality by the demographic variables. Patients in the national deprivation group 1 were more likely to die in hospital over the whole period than those in national deprivation group 4 (2.7 versus 1.4%). Those with an index admission not for an SCD crisis were more likely to die than those with an index admission for a crisis (2.9 versus 1.9%), and those with a Charlson score of 2 or more were most likely to die in hospital over the whole period (12.6 versus 1.1 for those with a score of zero).

Results from Cox proportional hazards models are given in Table 4. These show that over the time period, those in national deprivation group 1 (most deprived) were more likely to be readmitted than those in national deprivation group 4 (least deprived) [hazard ratio (HR) 2.97, 95% CI 2.57–3.43]. Those with a Charlson comorbidity score of two or more were more likely to be readmitted than those with a score of 0 (HR 1.71, 95% CI 1.51–1.94). Those with a primary diagnosis of SCD crisis were more likely to be readmitted than those with a secondary diagnosis of SCD (SCD as a comorbidity) (2.76, 95% CI 2.49–3.05), as were those with a primary diagnosis of crisis (HR 2.99, 95% CI 2.78–3.21). For in-hospital mortality, those in national deprivation group 1 were more likely to die in hospital than those in national deprivation

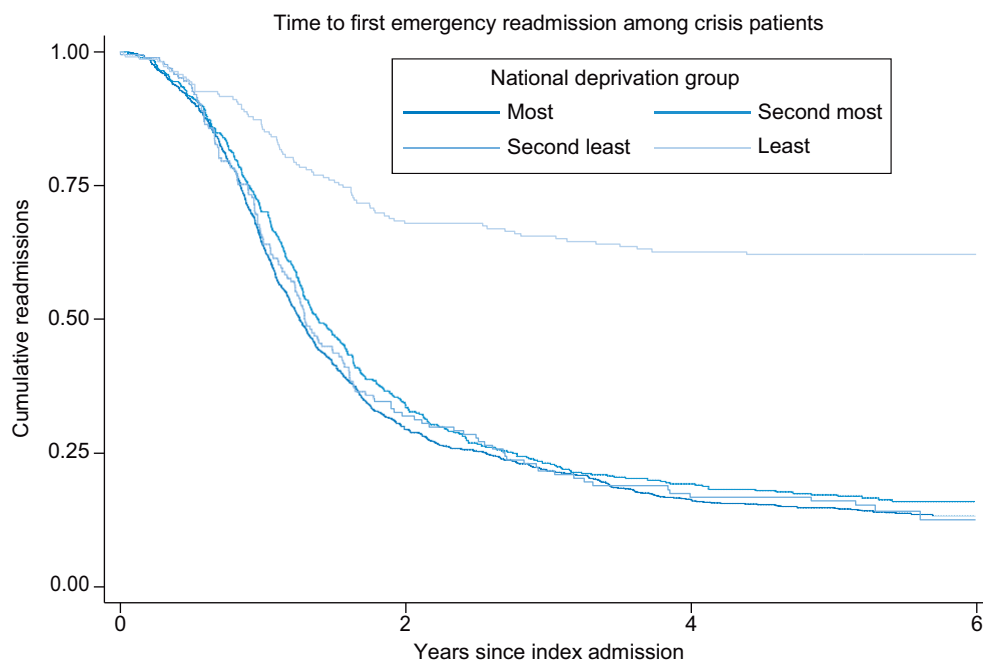


Fig. 1 Time to first emergency readmission across national deprivation groups, among patients with an index admission for an SCD crisis.

Table 2 Percentage of patients with emergency readmissions after admission in index year (2005/06) by patient characteristics (n=7697)

	2006	2007	2008	2009	2010	Any year
Age group						
<1	32.4	28.9	27.5	24.6	21.1	57.0
1–9	34.8	31.5	30.4	31.8	29.6	64.4
10–19	35.9	34.9	36.4	35.7	38.4	65.1
20–29	29.1	25.2	25.2	23.4	23.6	49.3
30–39	20.6	17.5	18.1	17.4	17.0	43.0
40–49	20.6	17.5	18.9	18.6	19.7	46.7
50 plus	19.8	16.2	15.4	13.6	12.3	44.2
Sex						
Male	30.8	27.3	26.4	25.6	25.3	74.3
Female	23.9	21.1	22.1	21.3	21.5	36.0
National deprivation group						
Most deprived	29.0	25.1	25.1	24.4	24.6	54.4
Second most deprived	27.1	25.3	25.2	24.5	24.7	53.8
Second least deprived	24.0	19.9	22.6	21.8	19.4	48.2
Least Deprived	13.5	13.1	13.6	12.1	12.5	27.8
Initial diagnosis						
Primary	44.5	41.2	41.3	39.9	41.3	73.7
Secondary	17.0	14.1	14.4	14.0	13.2	39.1
Index admission for crisis						
Yes	49.6	45.1	45.3	43.6	44.2	77.2
No	30.7	30.7	30.6	29.8	33.2	64.3
Charlson comorbidity index						
0	26.6	23.8	24.5	23.8	23.5	50.8
1	26.6	23.2	21.6	21.5	23.4	54.5
2::2	26.4	21.5	20.0	17.3	17.1	51.4
N admissions	2044	1807	1829	1767	1767	3392

Percentages relate to patients admitted at least in a year

group 4 over the time period (HR 2.34, 95% CI 1.41–3.90), as were those in national deprivation group 3 (HR 2.00, 95% CI 1.16–3.46). Those with a primary diagnosis of SCD non-crisis were more likely to die in hospital than those with a secondary diagnosis of SCD (3.58, 2.25–5.68) as were those with a primary diagnosis of crisis (HR 3.04, 95% CI 2.11–4.39).

Discussion

Main finding of this study

There is a higher frequency of SCD readmissions in areas of increased socio-economic deprivation. Some of the readmissions we observe may be due to new hospital episodes, but we still observe lower rates in less deprived places. The majority of readmissions (over 58%) and highest inpatient mortality occurred in patients living in the most socio-economically

deprived areas, with the highest risk of admission for patients aged 10–19 and highest risk of death in people aged ≥50. The highest risk age group highlights the importance of adolescent transitional care in patients with SCD^{14–16} and supports a concept analogous to the ‘inverse care law’ where differentials in access to care vary inversely with the (SCD) population most at need.^{17,18} Death rates are higher in those with Charlson of 2 or more suggesting death rates are higher in patients with increased comorbidities (who are also likely to be older).

Much remains unclear about the influence of socio-economic status (SES) on SCD. However, the wider SES and chronic disease literature indicates a number of ways in which those coming from the most deprived sector may experience increased hospital readmissions, excess in-patient mortality and consequent inequalities in health care.¹⁹

Table 3 Percentage of in hospital mortality after admission in index year (2005/06) by patient characteristics (n = 7679)

	2006	2007	2008	2009	2010	Any year
Age group						
<1	0.0	0.0	0.0	0.0	0.0	0.0
1–9	0.0	0.1	0.1	0.1	0.1	0.4
10–19	0.2	0.2	0.0	0.1	0.2	0.6
20–29	0.2	0.2	0.2	0.1	0.2	0.9
30–39	0.2	0.3	0.3	0.3	0.2	1.4
40–49	0.7	0.6	0.6	0.7	0.6	3.1
50 plus	3.1	2.8	2.5	1.6	1.3	11.8
Sex						
Male	0.8	0.6	0.5	0.5	0.5	3.0
Female	0.5	0.5	0.5	0.3	0.3	2.1
National deprivation group						
Most deprived	0.8	0.7	0.5	0.3	0.4	2.7
Second most deprived	0.4	0.4	0.6	0.6	0.4	2.5
Second least deprived	0.3	0.3	0.1	0.7	0.1	1.6
Least Deprived	0.1	0.3	0.6	0.3	0.4	1.7
Initial diagnosis						
Primary	0.3	0.3	0.6	0.5	0.5	2.3
Secondary	0.8	0.7	0.4	0.3	0.3	2.5
Index admission for crisis						
Yes	0.2	0.3	0.5	0.5	0.5	2.0
No	0.6	0.4	1.0	0.6	0.4	3.0
Charlson comorbidity index						
0	0.2	0.2	0.2	0.3	0.2	1.2
1	1.2	1.4	1.1	0.6	0.3	4.7
2::2	3.5	3.0	3.4	1.3	2.1	14.2
N admissions	46	42	38	31	27	184

What is already known on this topic

Within London, around 80% of all SCD admissions are from people living in the most deprived areas.¹ A study in the London Borough of Brent showed that that 74% of total bed days are associated with patients with multiple admissions.²⁰ Studies have found that, in general, causes of hospital readmission for patients with SCD included premature discharge, withdrawal syndrome and the occurrence of new acute [sickle cell crisis] episodes.⁵ SES may also influence the way healthcare providers manage chronic disease. A study by Struthers *et al.*²¹ showed an increased rate of hospital readmissions in those with lower SES for congestive heart failure independent of disease severity. The researchers suggest that this may be due to a number of reasons which included primary care providers in deprived areas having less time to invest in adequate disease management. Primary care providers working in deprived areas may also perceive their patients as having less capacity to understand and

manage their own condition,¹⁶ and patients living in a deprived areas may perceive that their community medical resources are insufficient to manage them and ‘push’ for hospital admission.^{21,22}

Another factor that may contribute to increased SCD readmissions, particularly amongst the most deprived patients, is patient self-discharge. A study in England found that 14% of their sample of SCD patients reported having self-discharged from hospital. The most common reasons given by the SCD patients for self-discharging were being ‘tired of waiting for pain relief’; potential conflicts related to suspected analgesics abuse and because they just simply wanted to go home.²³ Ballas and Lusardi⁵ also suggest that the decision by the patient to leave the hospital may be due to family pressure, childcare or fear of loss of job if the patient is employed. A similar study published in the USA found a much higher rate with 46.5% of their sample of SCD patients discharging themselves from hospital.²⁴

Table 4 Multivariate models for risk of emergency re-admission and inpatient mortality in 5 years after at least one admission in the index year (2005/06) ($n = 7697$)

	<i>Hazard ratio for admission</i>	<i>Confidence interval</i>	<i>Hazard ratio for mortality</i>	<i>Confidence interval</i>
Age group				
<1	1.42*	1.12; 1.78	Missing	—
1–9	1.46*	1.31; 1.63	0.33*	0.13; 0.84
10–19	1.38*	1.25; 1.54	0.41*	0.19; 0.89
20–29	1.12*	1.01; 1.23	0.76	0.44; 1.33
30–39	1.00	—	1.00	—
40–49	1.10	0.98; 1.23	2.03*	1.27; 3.26
50 plus	1.15*	1.02; 1.31	5.11*	3.34; 7.81
Sex				
Male	1.00	—	1.00	—
Female	1.04	0.97; 1.11	0.71*	0.55; 0.92
National deprivation group				
4 (Least deprived)	1.00	—	1.00	—
3 (Second least deprived)	2.54	2.13; 3.03	1.12	0.54; 2.32
2 (Second most deprived)	2.83	2.44; 3.31	2.00	1.16; 3.46
1 (Most deprived)	2.97	2.57; 3.43	2.34	1.41; 3.90
Charlson comorbidity index				
0	1.00	—	1.00	—
1	1.40*	1.26; 1.55	3.40*	2.33; 4.95
2::2	1.68*	1.48; 1.90	8.15*	5.81; 11.42
Initial diagnosis				
Secondary	1.00	—	1.00	—
Primary, non-crisis	2.68*	2.42; 2.97	3.46*	2.17; 5.52
Primary, crisis	2.92*	2.72; 3.14	3.31*	2.30; 4.77

*Statistically significant difference from reference category at $P < 0.05$.

Limitations of this study

The data may be subject to missing, inaccurate or incomplete data not uncommon to the data collection process. However, the advent of diagnosis-based payment of hospitals has accelerated improvement of data quality in HES.²⁵ We were also unable to evaluate the impact of severity of SCD disease which may contribute to the risk of readmission and inpatient mortality, as information on this is not included in HES. Additionally, we know that the UK has a mobile population, especially in the young adult group who may be students temporarily resident in the low prevalence/least deprived areas. Their readmissions may therefore be elsewhere in the UK (i.e. Wales, Scotland or Northern Ireland) or beyond and could have had an impact on reported readmissions. Future papers could also focus on what the secondary diagnoses were that led to patient deaths and if they are linked to primary diagnosis of SCD or unrelated. We also did not examine post-discharge factors that could influence risk of admission. These factors include characteristics of the home environment; and the quality and continuity of health care

of patients after they have been discharged from hospital including use of community/primary care facilities. Including post-discharge factors impact on readmission²⁶ could have provided further explanation into the pattern of SCD readmissions.

What this study adds

Hospital readmission rates in England have been used as a measure of poor healthcare performance at a local level or when making comparisons over time between hospitals.²⁷ However, there is a need to better understand the different factors that influence readmission rates for any disease specific category (including SCD) to control for extenuating factors that would impact on readmissions allowing a more thorough assessment of healthcare outcomes as well as more accurately defining potentially preventable readmissions.²⁸ Our study shows that SCD patients coming from the most deprived areas nationally experience higher rates of SCD hospital readmissions as well as inpatient mortality related to SCD, suggesting that there may be inequalities in access to health care

and health outcomes amongst people from this population. This continued to be true after adjustment for age and comorbidities between these populations, which might commonly be thought to contribute to these differences in readmission rates (data not shown). Also, the age and comorbidity breakdowns were similar between the least and most deprived groups (data not shown). Targeted intervention programs^{29,30} that concentrate on a small number of 'high-risk' patients, such as improving access to appropriate care or adherence to medication protocols^{31,32} provide genuine opportunities to prevent a large number of recurrent hospitalizations. Some readmissions may also be prevented through policy initiatives such as the development and implementation of local enhanced services for people with SCD. Local enhanced services are designed to meet local health needs and may therefore be particularly suitable for a condition such as SCD where the burden of disease is highest in a few urban areas such as London.

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Ethical approval

Not needed as this study is based on an analysis of routinely available data. Permission to use HES data was given by the NHS Information Centre.

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Conflict of interest

This article presents independent research commissioned by the National Institute for Health Research (NIHR) under the Collaborations for Leadership in Applied Health Research and Care (CLAHRC) programme for North West London. The views expressed in this publication are those of the author(s) and not necessarily those of the NHS, the NIHR or the Department of Health.

References

- 1 Lucas SB, Mason D, Mason M *et al*. A sickle crisis? A Report of the National Confidential Enquiry into Patient Outcome and Death. London: National Confidential Enquiry into Patient Outcome and Death, 2008.
- 2 Aisiku IP, Smith WR, McClish DK *et al*. Comparisons of high versus low emergency department utilizers in sickle cell disease. *Annals of emergency medicine*. *Am Coll Emerg Phys* 2009;53(5):587–93.
- 3 Olney RS. Preventing morbidity and mortality from sickle cell disease. *Am J Prev Med* 1999;16(2):116–21.
- 4 Murtaza LN, Stroud CE, Davis LR *et al*. Admissions to hospital of children with sickle-cell anaemia: a study in south London. *BMJ* 1981;282(6269):1048–51.
- 5 Ballas SK, Lusardi M. Hospital readmission for adult acute sickle cell painful episodes: frequency, etiology, and prognostic significance. *Am J Hematol* 2005;79(1):17–25.
- 6 Brousseau DC, Owens PL, Mosso AL *et al*. Acute care utilization and rehospitalizations for sickle cell disease. *JAMA* 2010;303(13):1288–94.
- 7 Platt O, Brambilla D. Mortality in sickle cell disease—life expectancy and risk factors for early death. *N Engl J Med* 1994;330:1639–44.
- 8 Aljuburi G, Laverty AA, Green SA *et al*. Trends in hospital admissions for sickle cell disease in England, 2001/02–2009/10. *J Public Health (Oxf)* 2012;34(4):570–6.
- 9 Anie KA, Steptoe A, Ball S *et al*. Coping and health service utilisation in a UK study of paediatric sickle cell pain. *Arch Dis Child* 2002;86(5):325–9.
- 10 Elandera J, Beach MC, Haywood C Jr. Respect, trust, and the management of sickle cell disease pain in hospital: comparative analysis of concern-raising behaviors, preliminary model, and agenda for international collaborative research to inform practice. *Ethn Health* 2011;16(4–5):405–21.
- 11 The Health and Social Care Information Centre. *Hospital Episode Statistics Online [Internet]*. 2013. <http://www.hesonline.nhs.uk/Ease/servlet/ContentServer?SiteID=1937&categoryID=87> (11 March 2013, date last accessed).
- 12 Department for Communities and Local Government. *The English Indices of Deprivation 2010*. English. London: Department for Communities and Local Government, 2011.
- 13 Charlson M, Pompei P, Ales K *et al*. A new method of classifying prognostic comorbidity in longitudinal studies: development and validation. *J Chron Dis* 1987;40(5):373–83.
- 14 Jordan L, Swerdlow P, Coates TD. Systematic review of transition from adolescent to adult care in patients with sickle cell disease. *J Pediatr Hematol Oncol* 2013;35(3):165–9.
- 15 Quinn CI, Rogers ZR, McCavit TL *et al*. Improved survival of children and adolescents with sickle cell disease. *Blood* 2010;115(17):3447–52.
- 16 Telfair J, Ehiri J, Loosier PS *et al*. Transition to adult care for adolescents with sickle cell disease: Results of a national survey. *Int J Adolesc Med Health* 2004;16(1):47–64.
- 17 Lyratzopoulos G, Havelly D, Gemmell I *et al*. Factors influencing emergency medical readmission risk in a UK district general hospital: a prospective study. *BMC Emerg Med* 2005;5(1):1.

- 18 Hart JT. The inverse care law. *Lancet* 1971;297(7696):405–12.
- 19 Saxena S, Eliahoo J, Majeed A. Socioeconomic and ethnic group differences in self study. *BMJ* 2002;325:1–6.
- 20 Green SA, Aljuburi G, Majeed A *et al.* Characterizing emergency admissions of patients with sickle cell crisis in NHS Brent: observational study. *JRSM short reports* 2012;3(6):37.
- 21 Struthers AD, Anderson G, Donnan PT *et al.* Social deprivation increases cardiac hospitalisations in chronic heart failure independent of disease severity and diuretic non-adherence. *Heart* 2000;83(1):12–6.
- 22 Blair AS, Lloyd-Williams F, Mair FS. What do we know about socioeconomic status and congestive heart failure? A review of the literature. *J Fam Pract* 2002;51(2):169.
- 23 Elander J, Lusher J, Bevan D *et al.* Understanding the causes of problematic pain management in sickle cell disease: evidence that pseudoaddiction plays a more important role than genuine analgesic dependence. *J Pain Symptom Manage* 2004;27(2):156–69.
- 24 Haywood Jr C, Lanzkron S, Ratanawongsa N. Associations with trust and interpersonal experiences with. *J Hosp Med* 2011;5(5):289–94.
- 25 Audit Commission. *Payment by Results Data Assurance Framework [Internet]*. PbR data assurance framework 2012/13. London, 2012. <http://www.audit-commission.gov.uk/information-and-analysis/data-assurance-framework/> (29 August 2012, date last accessed).
- 26 Arbaje AI, Wolff JL, Yu Q *et al.* Postdischarge environmental and socioeconomic factors and the likelihood of early hospital readmission among community-dwelling Medicare beneficiaries. *Gerontologist* 2008;48(4):495–504.
- 27 Mason A, Daly E, Goldacre M. Using admission rates as health outcome indicators: literature review—full report. Oxford: National Centre for Health Outcomes Development, 2000.
- 28 Nolte E, Roland M, Guthrie S *et al.* *Preventing Emergency Readmissions to Hospital: A Scoping Review*. Santa Monica, CA: RAND Corporation, 2012.
- 29 Aljuburi G, Okoye O, Majeed A *et al.* Views of patients about sickle cell disease management in primary care: a questionnaire-based pilot study. *JRSM Short Rep* 2012;3(11):78.
- 30 Aljuburi G, Phekoo KJ, Okoye NO *et al.* Patients' views on improving sickle cell disease management in primary care: focus group discussion. *JRSM Short Rep* 2013;3(12):84–90.
- 31 Bloomberg GR, Trinkaus KM, Fisher EB *et al.* Hospital readmissions for childhood asthma: a 10-year metropolitan study. *Am J Respir Crit Care Med* 2003;167(8):1068–76.
- 32 Frei-jones MJ, Field JJ, DeBaun MR. Risk factors for hospital readmission within 30 Days: a new quality measure for children with sickle cell disease. *Pediatric Blood Cancer* 2009;52:481–5.

Trends in hospital admissions for sickle cell disease in England, 2001/02–2009/10

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ABSTRACT

Background Sickle cell disease (SCD) is a rising cause of mortality and morbidity in England and consequently an important policy issue for the National Health Service. There has been no previous study that has examined SCD admission rates in England.

Methods Data from Hospital Episode Statistics were analysed for all hospital episodes (2001/10) in England with a primary diagnosis of sickle cell anaemia with crisis (D57.0) or without crisis (D57.1). Secondary and tertiary diagnoses were examined among those patients admitted with either of these codes as their primary diagnosis.

Results The overall SCD admission rate per 100 000 has risen from 21.2 in 2001/02 to 33.5 in 2009/10, a rise of over 50%. London accounts for 74.9% of all SCD admissions in England. 57.9% of patients admitted are discharged within 24 h. The largest rise in admission rates was seen among males aged 40–49 years where admission rates per 100 000 increased from 7.6 to 26.8 over the study period.

Conclusions Our data show that SCD admissions are rising in England, particularly in London. Over half of patients admitted with SCD were discharged within 24 h, suggesting that some of these admissions could be prevented through better ambulatory care of patients.

Keywords chronic disease, management and policy, public health

Introduction

Sickle cell disease (SCD) is a group of inherited blood disorders in which red blood cells are abnormally formed. The abnormal cells break down causing anaemia and can also cause severe complications requiring frequent hospitalization, as well as reducing the quality of life and life expectancy. SCD affects millions of people around the world and is particularly common amongst populations originating from Africa and the Caribbean.¹ SCD is now the fastest growing and most common genetic disorder in England² with "12 500 people living with the condition in the UK and 240 000 SCD trait carriers.³ SCD is also a common reason for hospital admissions.^{4,5}

Although the majority of health care for patients with SCD should be occurring in ambulatory and primary care settings, the hallmark of SCD, acute painful episodes also known as vaso-occlusive crisis, is often the reason for repeat

admissions into hospital.⁶ SCD is less common than chronic diseases such as cardiovascular disease, diabetes and cancer, but its high hospital utilization make the disease important from a health policy perspective.^{7–9} Thus, the National Health Service (NHS) should be closely

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monitoring pathways of care including hospital utilization of patients with SCD.

Little data exist in England about the healthcare utilization of SCD patients. Data on hospital admissions in England for SCD can provide information about the impact of SCD on the NHS and would also be of value to clinicians, commissioners and healthcare planners. Such data could also highlight disease-specific trends. The aim of this study was to investigate trends in the rates of hospital admissions in England for SCD and to determine geographic variations. The study period covered 2001/02–2009/10. We used data from the Hospital Episodes Statistics (HES) database, which provides data on all NHS hospital admissions in England.¹⁰ HES have been collected since 1987 by the Department of Health.¹¹

Methods

We obtained data from the national HES database from April 2001 to April 2010. HES is the national administrative database for hospital activity in England and contains data on all admissions and outpatient appointments in the NHS, including patients whose treatment is funded by the NHS but performed in private hospitals.¹² HES data contains details on the diagnoses of every patient admitted to an NHS hospital in England, which are coded using the International Classification of Diseases version 10. Patients can be coded as having up to 20 different diagnoses. This analysis included patients with a primary diagnosis of ‘sickle cell anaemia with crisis’ (D57.0) or ‘sickle cell anaemia without crisis’ (D57.1). Primary diagnoses correspond to the main reason the patient was admitted to hospital. Secondary and tertiary diagnoses were examined among those patients admitted with SCD as their primary reason for admission.

Patients may be under the care of different consultants during their stay in hospital, and each of these is known as a ‘finished consultant episode’. All data were collected related to hospital admissions. Different hospital episodes were aggregated into hospital spells (i.e. admissions to hospital) using a method recommended by the NHS Information Centre, based on patient identifying information, their dates of admission and discharge.¹³

Data on admissions were broken down by age (1-year old, 1–9, 10–19, 20–29, 30–39, 40–49 years and .50 years old), sex and Strategic Health Authority and Primary Care Trust (PCT). The admission rates were sex- and age standardized. Less than 1-year old was included as a category in its own right to reflect concern over potentially high admission rates in this group and in line with previous work.⁷ As London accounts for the majority of the SCD cases in England, admission rates by PCT are presented only for London due to small numbers of admission in other PCTs. Denominator data for population estimates which were used to calculate rates were obtained from the Office for National Statistics.¹⁴ All analyses were conducted using Stata, v. 10.0.

Results

Figure 1 and Table 1 show the overall admission rates for SCD as a primary diagnosis from 2001/02 to 2009/10. From Table 1 we can see that the overall admission rate per 100 000 population has risen from 21.2 per 100 000 in 2001/02 to 33.5 per 100 000 in 2009/10. This represents a rise of .50% in the admission rate from 2001/02 levels, with a consistent year-on-year rise with the exception of between 2002/03 and 2003/04.

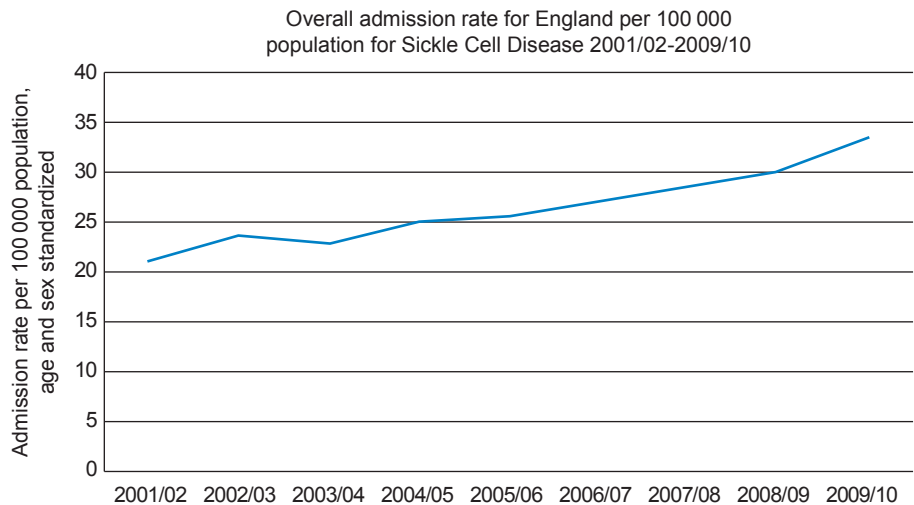


Fig. 1 Trends in admissions for sickle cell disease in England, 2001/02–2009/10.

Table 1 Age- and sex-specific admission rates per 100 000 population in England for SCD as a primary diagnosis 2001/02–2009/10

Age (years)	Year								
	2001/02	2002/03	2003/04	2004/05	2005/06	2006/07	2007/08	2008/09	2009/10
Men overall	20.7	24.1	24.1	25.4	25.9	28.5	31.3	32.7	35.1
<1	8.8	6.7	7.1	4.6	12.6	16.1	16.7	15.2	14.7
1–9	18.6	21.2	21.5	24.9	22.7	27.8	37.1	41.2	45.5
10–19	24.9	30.6	30.0	35.8	37.8	39.2	44.6	49.9	55.6
20–29	50.0	49.7	54.8	57.1	57.4	70.5	65.0	64.7	68.6
30–39	44.3	54.3	47.7	44.2	39.3	37.8	42.8	41.9	46.8
40–49	7.6	13.7	16.0	17.3	23.5	24.1	28.1	29.0	26.8
≥50	1.6	1.4	1.6	2.3	2.3	2.1	2.7	2.8	3.2
Women overall	21.6	23.3	21.7	24.6	25.2	25.6	26.1	27.6	31.9
<1	7.3	7.0	7.9	5.2	12.8	11.9	10.9	13.2	10.8
1–9	12.6	18.5	17.7	20.3	23.0	23.5	26.1	36.5	42.2
10–19	29.1	36.3	31.9	40.3	41.8	47.3	48.5	51.2	56.4
20–29	32.4	35.4	38.0	44.6	51.9	50.9	52.1	52.4	57.9
30–39	52.4	50.1	41.2	41.7	41.4	36.6	35.7	37.0	45.3
40–49	25.4	24.5	25.9	28.0	22.1	25.7	26.0	24.7	29.2
≥50	3.4	4.0	3.8	4.9	4.5	4.2	3.9	4.0	5.1
Total	21.2	23.7	22.9	25.0	25.6	27.0	28.7	30.1	33.5

This rise in admission rates has occurred in every age and sex group with the exception of women aged 30–39 years, whose admission rate per 100 000 population declined from 52.4 to 45.3 over the time period. (Table 1) The largest rise in admission rates was seen amongst men aged between 40 and 49 years old, where admission rates per 100 000 population increased from 7.6 to 26.8 over the time period (Table 1).

There were 16 558 admissions for SCD in England in 2009/10 and 8488 (51.3%) of these were men (Table 2). The admission rate per 100 000 population in this period was slightly higher in men than for women, 35.1 compared with 31.9 (Table 1). The highest admission rates in 2009/10 were amongst men aged 20–29 at 68.6 per 100 000 population (Table 1). The lowest rates were amongst people over 50 years old at 3.2 per 100 000 population for men and 5.1 per 100 000 for women (Table 1).

London accounted for almost three quarters of all SCD admissions in England (74.9% of all admissions nationally, data not shown). The highest admission rate was in City and Hackney Teaching PCT at 837.4 per 100 000 population. This rate is over twice that of the next highest PCT, Newham at 388.1 per 100 000 population (Supplementary data, Figure A). The map demonstrates geographical heterogeneity between boroughs with a clustering of boroughs with some of the highest admissions occurring in City, Hackney, Newham, Haringey, Waltham Forest and Islington (Fig. 2).

Table 2 Age and sex breakdown of crisis and non-crisis primary diagnosis, England 2009/10

Age (years)	Sickle cell crisis (%)	Sickle cell non-crisis (%)	Total
Men (overall)	4509 (53.1)	3979 (46.9)	8488
<1	34 (68.0)	16 (32.0)	50
1–9	478 (38.1)	777 (61.9)	1255
10–19	855 (47.4)	948 (52.6)	1803
20–29	1554 (62.8)	919 (37.2)	2473
30–39	983 (60.7)	637 (39.3)	1620
40–49	481 (47.2)	539 (52.8)	1020
≥50	124 (46.4)	143 (53.6)	267
Women (overall)	8572 (51.8)	7986 (48.2)	16558
<1	17 (48.6)	18 (51.4)	35
1–9	479 (43.0)	634 (57.0)	1113
10–19	891 (51.3)	847 (48.7)	1738
20–29	1260 (62.9)	744 (37.1)	2004
30–39	787 (50.2)	780 (49.8)	1567
40–49	482 (42.7)	648 (57.3)	1130
≥50	147 (30.4)	336 (69.6)	483
Total number overall	4063 (50.3)	4007 (49.7)	8070

Table 3 shows the most common secondary and tertiary diagnoses for those admitted with SCD as the primary diagnosis. The most common additional diagnostic codes were for respiratory conditions including lower respiratory tract

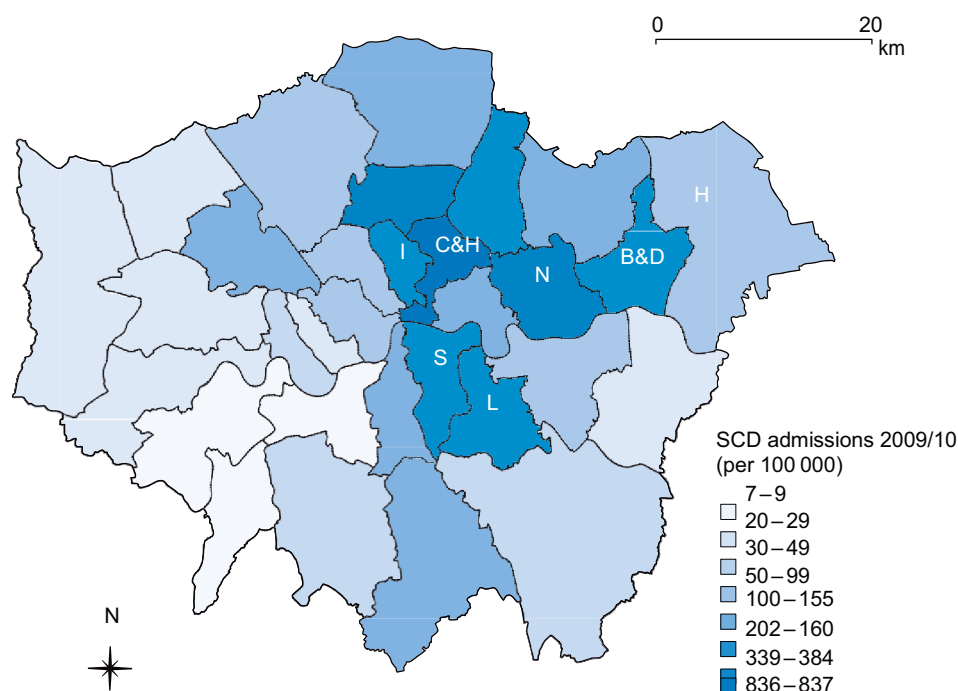


Fig. 2 Admission rate for SCD per 100 000 by PCTs, London, 2009/10; Top 8 boroughs indicated: City and Hackney (C&H), Newham (N), Haringey (H) Islington (I), Barking and Dagenham (B&D), Lewisham (L), Waltham Forest (WF), Southwark (S) in descending order of admissions.

infection, lobar pneumonia and asthma, followed by vaso-occlusive conditions such as occlusion and stenosis of middle cerebral artery, Moyamoya disease, hemiplegia and sequelae of stroke (not specified as haemorrhage or infarction).

Table 2 shows the breakdown of crisis and non-crises admissions in 2009/10. Slightly over half of all admissions (50.3%) were for a sickle cell crisis, and those in the age group of 20–29 years had a higher proportion of crisis admissions than other groups (62.8 and 62.9% for men and women, respectively). The lowest proportion of crisis admissions was in women aged over 50 (30.4%).

57.9% of all patients were discharged within 24 h of being admitted to hospital, while only 15.4% of admissions stayed for longer than a week. The largest proportion of patients discharged on the same day was for those aged over 50 years old, at 72.1% (data not shown).

Discussion

Main finding of this study

This is the first national study on trends for hospital admissions rates for SCD using HES data in England. Our data have four main findings: (i) the number of admissions due to SCD has increased substantially over the study period (2001/10); (ii) the majority of admissions are from London; (iii) most admissions were discharged within 24 h and

(iv) there has been an increase in the number of admissions of middle-aged men.

What is already known on this topic

Our data show an increase of .50% in SCD admissions. This will be due to a number of factors, the most important of which is likely to be an increased prevalence of SCD in England through domestic growth of the current at-risk population and population migration from countries where SCD is common.¹⁵

A previous study on the national implementation of newborn screening in England showed an increased number of children identified with SCD in some areas. The authors of this study concluded that past under-ascertainment of cases may have led to under-estimation of the needs of people with SCD. Under-ascertainment of SCD cases may also have contributed to increased infant mortality in urban areas because babies were at risk of dying without a diagnosis of SCD being made and appropriate management started.¹⁶

The significant migration of people from the West Indies, Africa and Asia also partially accounts for the increasing incidence and prevalence of SCD in England. Some studies suggest that 6% of the population (in England) and "10% of all births are at risk for haemoglobinopathy disorders.¹⁷ With current trends in at-risk population migration into the

Table 3 Secondary and tertiary diagnoses among those with SCD as a primary diagnosis in NHS Hospitals, England, 2009/10

	Number (% of 2009/10 total)
Top 10 secondary diagnoses	
Acute lower respiratory tract infection	310 (1.9)
Chemotherapy	223 (1.3)
Asthma	187 (1.1)
Urinary tract infection	133 (0.8)
Lobar pneumonia	128 (0.8)
Naevi and melanomas	115 (0.7)
Occlusion and stenosis of middle cerebral artery	104 (0.6)
Moyamoya disease	96 (0.6)
Essential hypertension	91 (0.5)
Chest pain	88 (0.5)
Top 10 tertiary diagnoses	
Asthma	89 (0.5)
Sequelae of stroke, not specified as haemorrhage or infarction	64 (0.4)
Priapism	59 (0.4)
Dorsalgia	57 (0.3)
Naevi and melanomas	54 (0.3)
Acute lower respiratory tract infection	54 (0.3)
Hemiplegia	53 (0.3)
Essential hypertension	47 (0.3)
Vitamin D deficiency	42 (0.3)
Hyposplenism	41 (0.2)

country and continuing growth in the domestic at-risk population, these conditions will become even more common over the next decade.¹⁸ Platt *et al.*¹⁹ also explained that increases in prevalence are due to an increase in life expectancy of patients living with sickle cell through improved secondary and tertiary prevention, including improving the management of risk factors associated with haemoglobinopathies; and through earlier detection and management of disease-related complications.

The increases in admissions for SCD may also be due to a drop in the threshold for admission of patients with SCD, through greater recognition of the importance of SCD crisis, or through patients being discharged earlier and being readmitted more frequently for the same episode of illness.²⁰

Studies from the USA have also shown readmission as an important contributor to rising rates of admissions in patients with SCD.²¹ This may also raise issues of how well hospitals are managing these patients,²² and emphasizes the importance of good discharge planning for these patients which may minimize the risk of hospital readmission. Good discharge planning is the process that ensures strong and

quality links between hospitals, primary and community care services and patients and their carers. Effective discharge planning for those with SCD can support the continuity of health care for a patient as it links treatment received in the hospital to post-discharge care received in the community.

A study conducted at King's College Hospital showed that there has been a decrease in overnight admission rates for King's patients with SCD over the last 50 years. This is due to better disease management and better discharge planning. The number of at-risk people served at King's College Hospital has not decreased but the admissions have dropped from 111 admissions per 100 SCD patient-years in 1960–1979 to 41 in 2008–2009. The study reported that this decrease is particularly accounted for by better acute pain management and transfusions. This includes the use of hydroxyurea (a chemotherapeutic agent), increased community support, increased use of oral analgesia and improved parental education about the management of acute pain at home.^{23–26} The study also reported that admission to hospital for elective blood transfusion was formerly the second most common reason for SCD admission but fell from 213 admissions to 1 over the study period. Elective transfusions are still an important aspect of care for SCD but these have been largely moved to ambulatory settings.^{23,27}

SCD is still one of the most common reasons to be admitted to hospital and SCD also has one of the highest rates of multiple admissions for individual patients. The distribution of SCD in London also shows that 80% of all admissions are from people living in the most deprived areas of city.^{4,5,28} Our finding that the majority of SCD admissions are discharged within 24 h was also found in another study that was conducted in the London Borough of Brent which showed that 36% of SCD patient admissions in Brent resulted in a length of stay of <2 days and that 74% of total bed days are associated with patients with multiple admissions.²⁸

Limitations of this study

The strength of our study is in its comprehensiveness in that it covers all NHS hospital admissions in England. Our data have the weaknesses associated with missing, inaccurate or incomplete routine data, such as incorrect hospital diagnoses and coding. However, the advent of diagnosis-based payment of hospitals has accelerated improvement in the data quality. We used primary diagnosis to identify admissions from SCD, and 87.2% of primary diagnoses for hospital admissions in England are now coded correctly.²⁹ We did not examine readmissions and multiple admissions in the same patient, which could have provided further valuable insight into patterns of SCD admissions.

We also did not examine the mortality data linked to hospital episodes,³ which would have given information of deaths associated with SCD hospital admissions. However, as this was a study about the burden of admissions, re-admissions and mortality data were not relevant to this study.

What this study adds

We examined trends in hospital admissions associated with SCD in England. We found that between 1999/2000 and 2009/10, the rate of hospital admissions in England has increased for SCD; that London accounts for the majority of admissions in England and that majority of these hospitalizations are short stay. These data show that SCD is a growing concern in England and provides a basis to continue further investigation into explaining such trends. The study also highlighted that the majority of patients admitted with SCD were discharged within 24 h. These admissions could potentially be prevented through better ambulatory and community care of patients, including better care in emergency departments, and this is an area that PCTs with a high prevalence of SCD should consider investigating.

Supplementary data

Supplementary data are available at the *Journal of Public Health* online.

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Conflict of interest

This article presents independent research commissioned by the National Institute for Health Research (NIHR) under the Collaborations for Leadership in Applied Health Research and Care (CLAHRC) programme for North West London. The views expressed in this publication are those of the author(s) and not necessarily those of the NHS, the NIHR or the Department of Health.

References

- 1 Weatherall D, Hofman K, Rodgers G *et al* A case for developing north-south partnerships for research in sickle cell disease. *Blood* 2005;105:921–3.
- 2 NHS Sickle Cell and Thalassaemia Screening Programmes. *Sickle Cell Disease in Childhood Standards and Guidelines for Clinical Care*, 2nd edn. Sickle Cell Society, 2010.
- 3 Department of Health. National service framework for long-term conditions. Department of Health, 2005.
- 4 London Health Observatory. Analysis of frequent hospital users, by PCT 2003/4 [Online]. 2006. <http://www.lho.org.uk/viewResource.aspx?id=10286> (September 2011, date last accessed).
- 5 Lucas SB, Mason DG, Mason M *et al* A sickle crisis? A report of the National Confidential Enquiry into Patient Outcome and Death, 2008. http://www.ncepod.org.uk/2008report1/Downloads/Sickle_report.pdf#search=sicklecell.
- 6 Aisiku IP, Smith WR, McClish DK *et al* Comparisons of high versus low emergency department utilizers in sickle cell disease. *Ann Emerg Med* 2009;53(5):587–93.
- 7 Epstein K, Yuen E, Riggio JM *et al* Utilization of the office, hospital and emergency department for adult sickle cell patients: a five year study. *J Natl Med Assoc* 2006;98(7):1109–13.
- 8 Bilenker JH, Weller WE, Shaffer TJ *et al* The costs of children who have sickle cell anemia: preparing for managed care. *J Pediatr Hematol Oncol* 1998;20:528–33.
- 9 Davis H, Moore RM Jr, Gergen PJ. Cost of hospitalizations associated with sickle cell disease in the United States. *Public Health Rep* 1997;112:40–3.
- 10 Department of Health. *An Introduction to HES, Overview of the Hospital Episode Statistics System*. London: Department of Health, 1999.
- 11 Majeed A. Hospital admissions data: why are they collected? *Clin Manag* 1998;7:160–6.
- 12 Hospital Episode Statistics Online. <http://www.hesonline.nhs.uk/Ease/servlet/ContentServer?siteID=1937&categoryID=87> (12 September 2011, date last accessed).
- 13 The Health and Social Care Information Centre. The information centre: methods for construction of provider spells. <http://www.hesonline.nhs.uk/Ease/servlet/ContentServer?siteID=1937&categoryID=1072> (21 September 2011, date last accessed).
- 14 ONS population estimates. <http://www.ons.gov.uk/ons/publications/re-reference-tables.html?edition=tcn%3A77-50032> (28 September 2011, date last accessed).
- 15 World Health Organization. Sickle cell anaemia report by the secretariat, 2006, A59/9.
- 16 Streetly A, Latinovic R, Hall K *et al* Implementation of universal newborn bloodspot screening for sickle cell disease and other clinically significant haemoglobinopathies in England: screening results for 2005 to 2007. *J Clin Pathol* 2009;62:26–30.
- 17 Anionwu EN, Atkin K. *The Politics of Sickle Cell and Thalassaemia*. Buckingham: Open University Press, 2001.
- 18 Streetly A, Clarke M, Downing M *et al* Implementation of the newborn screening programme for sickle cell disease in England: results for 2003–2005. *J Med Screen* 2008;15(1):9–13.

- 19 Platt OS, Brambilla DJ, Rosse WF *et al* Mortality in sickle cell disease: life expectancy and risk factors for early death. *N Engl J Med* 2008;330:1639–44.
- 20 Patel H, Bell D, Molokhia M *et al* Trends in hospital admissions for adverse drug reactions in England: analysis of national hospital episode statistics 1998–2005. *BMC Clin Pharmacol* 2007;25(7):9.
- 21 Ballas SK, Lusardi M. Hospital readmission for adult acute sickle cell painful episodes: frequency, etiology, and prognostic significance. *Am J Hematol* 2005;79:17–25.
- 22 Maxwell K, Streetly A, Bevan D. Experiences of hospital care and treatment seeking for pain from sickle cell disease: qualitative study. *BMJ* 1999;318:1585.
- 23 Day TG, Thein SW, Drasar E *et al* Changing pattern of hospital admissions of children with sickle cell disease over the last 50 years. *J Pediatr Hematol Oncol* 2011;33(7):491–495.
- 24 Halsey C, Roberts IA. The role of hydroxyurea in sickle cell disease. *Br J Hematol* 2003;120:177–86.
- 25 Jacobson SJ, Kopecky EA, Joshi P *et al* Randomised trial of oral morphine for painful episodes of sickle-cell disease in children. *Lancet* 1997;350:1358–61.
- 26 Mahat G, Scoloveno MA, Donnelly CB. Written educational materials for families of chronically ill children. *J Am Acad Nurse Pract* 2007;19:471–6.
- 27 Murtaza LN, Stroud CE, Davis LR *et al* Admissions to hospital of children with sickle-cell anaemia: a study in south London. *Br Med J* 1981;282:1048–51.
- 28 Green S, AlJuburi G, Amobi C *et al* Characterising emergency admissions of patients with sickle cell crisis in NHS Brent: observational study, 2011.
- 29 Audit Commission. PbR data assurance framework, 2009. <http://www.audit-commission.gov.uk/SiteCollectionDocuments/AuditCommission-Reports/NationalStudies/20090827pbrdataassuranceframework0809-rep.pdf>.



Views of patients about sickle cell disease management in primary care: a questionnaire-based pilot study

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DECLARATIONS

Competing interests

None declared

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Summary

Objectives To determine how patients with sickle cell disease (SCD) perceive the quality of care that they receive from their primary healthcare providers.

Design A questionnaire-based pilot study was used to elicit the views of patients about the quality of care they have been receiving from their primary healthcare providers and what they thought was the role of primary care in SCD management.

Setting Sickle Cell Society and Sickle Cell and Thalassaemia Centre, in the London Borough of Brent.

Participants One hundred questionnaires were distributed to potential participants with SCD between November 2010 and July 2011 of which 40 participants responded.

Main outcome measures Analysis of 40 patient questionnaires collected over a nine-month period.

Results Most patients are generally not satisfied with the quality of care that they are receiving from their primary healthcare providers for SCD. Most do not make use of general practitioner (GP) services for management of their SCD. Collecting prescriptions was the reason most cited for visiting the GP.

Conclusion GPs could help improve the day-to-day management of patients with SCD. This could be facilitated by local quality improvement schemes in areas with high disease prevalence. The results of the survey have been used to help develop a GP education intervention and a local enhanced service to support primary healthcare clinicians with SCD's ongoing management.

Ethical approval

Ethical approval was not required for this work as it is part of a service evaluation and improvement project

Guarantor

GA

Contributorship

KJP, YK and AM originally conceived the study; OO and KJP conducted the data analysis; GA wrote the first draft; all authors contributed in the revision of the manuscript

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Reviewer

Kamran Abbasi

Introduction

Sickle cell disease (SCD) is the most common inherited blood disorder in England. Without prompt diagnosis and proper treatment, it can be a serious source of morbidity and mortality. SCD is caused by a single amino acid substitution of valine for glutamic acid in the sixth position of the beta (β)-chain of the haemoglobin tetramer.^{1,2} The disease damages and changes the shape of red blood cells. The change in shape is a response to cell deoxygenation. When the oxygen uptake of the cell is low, the cells change their shape from a healthy round disk to a crescent (sickle shape), holly leaf or other similarly distorted shape. The sickled cells are rigid, less malleable and stickier than a normal cell; consequently, they may stick to each other and obstruct blood vessels. This obstruction causes harsh and painful complications. The complications can lead to frequent hospital visits and proper management of SCD is needed to minimize the risk of developing such complications.³

SCD clinical guidelines recommend that patients see a general practitioner (GP) for routine examination every six months and more often if new problems arise or their treatment protocol changes.⁴ In addition, immunizations, prescriptions and other preventive care measures need to be delivered effectively by GPs to prevent recurring infections and pain crisis.⁵

An analysis of emergency department admissions between January 2008 and July 2010 in the Northwest London Borough of Brent showed that patients with SCD tend to use the emergency department rather than seek advice and support from their GP.⁶ A focus group aimed at obtaining patient perspectives held in Brent showed that one of the reasons patients utilize the emergency department over their GP is because they perceive GPs as having limited knowledge of SCD.⁷ A primary care educational intervention has been designed, informed by these studies. To further triangulate the experience of patients, this pilot study was designed to elicit the views of patients about the quality of care they have been receiving from their primary healthcare providers and what they thought was the role of primary care in SCD management.

Methods

Study design and questionnaire

The study consisted of the development and administration of a 14-item study-specific questionnaire devised by a sickle cell steering committee which examined patients' perceptions towards SCD and key management issues in primary care including severity of disease, how many times patients visited the Emergency Department in the last year, how many times patients saw or called their GP for sickle and non-sickle-related illnesses and general questions about GP satisfaction (Box 1). The validation process for the questionnaire followed several drafts reviewed by seven GPs practising across inner London, a professor of paediatric haematology specializing in SCD, a haematologist specializing in SCD in adults, a SCD specialist nurse, a SCD social worker, a SCD clinical psychologist, a number of quality improvement project managers, public health specialists, patient representatives with SCD and directors of the Sickle Cell Society (a national UK sickle cell charity). The pilot study is in itself part of the mechanism of validation for a larger questionnaire study.

Participants

One hundred questionnaires were distributed by post and in person (CN and PO) over a nine-month period from November 2010 to July 2011 to members of the Sickle Cell Society and to patients who attended the Sickle Cell and Thalassemia Centre, using a purposive sampling method. Forty questionnaires were completed and suitable for analysis (OO and KJP).

Results

A response rate of 40% was achieved. Fifty percent (20) were completed by men and 50% (20) by women. The highest percentage of respondents (27%) was in the age range of 25–39 years. Five percent of respondents were aged 65+ years. The majority (62%) of the respondents were in good health and mobile. Nineteen (47%) respondents did not use their GP to manage a painful crisis and preferred to attend the emergency department. Twenty-four (59%) respondents went to

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Box 1

A selection of questions from the patient questionnaire

Question

In the past year, how many times have you seen or called your GP for help with a sickle cell crisis?

Question

Rank the three main things you do most often when in crisis:

- | | |
|--|---|
| ☐ Do nothing | ☐ Go to GP |
| ☐ Call Sickle Cell Specialist Nurse | ☐ Go to A&E |
| ☐ Stay at home | ☐ Other (please state):..... |

Question

How often do you utilise your GP to:

- | | | | | |
|---|----------------------|-------------------------|---------------------------|-------------------|
| a) Manage a painful crisis | | | | |
| Never | Rarely
(1-2/year) | Sometimes
(3-4/year) | Fairly often
(monthly) | A lot
(weekly) |
| b) Collect repeat prescription for antibiotics, folic acid, analgesia | | | | |
| Never | Rarely
(1-2/year) | Sometimes
(3-4/year) | Fairly often
(monthly) | A lot
(weekly) |
| c) Get general advice about sickle cell disease | | | | |
| Never | Rarely
(1-2/year) | Sometimes
(3-4/year) | Fairly often
(monthly) | A lot
(weekly) |
| d) Get help with non-sickle related illness (e.g. coughs, colds, etc) | | | | |
| Never | Rarely | Sometimes | Fairly often | A lot |

their GP to collect repeat prescriptions. Twenty-two (55%) respondents did not visit their GP to get general advice about SCD (Table 2). Nine (23%) rarely visited their GP with four (10%) visiting sometimes or fairly often (Table 1). When asked how satisfied patients are with assistance given by their GP to help manage their SCD based on a scale of 0–10 (0 being not satisfied at all and 10 being very satisfied), the majority (54%) scored satisfaction with their GP as 5 or less while 43% scored a 6 or above and 3% did not answer the question at all. Collecting prescriptions was the reason most cited for visiting the GP's office (Table 2). Some examples of comments around the services provided by GPs include:

'The GP should know about this disease'.
(Patient Questionnaire 1)

'The GP does not know anything about SCD pain and crisis. I would rather manage

... at home or [go] to A + E where immediate action will be taken rather than call the GP who will ask us to book an appointment and more or less does not understand how to manage the pain or how severe or serious the pain is'. (Patient Questionnaire 2)

Discussion

Key findings

The results from the survey show that many patients are generally not satisfied with the quality of care that they are receiving from their primary healthcare providers for SCD. Thus, most do not make use of GP services for management of their SCD. More importantly, the majority of the group did wish for greater involvement from their GP services, even if it was just to refer them to a tertiary care facility or social support.

Table 1
How often do you use your general practitioner to get general advice about sickle cell disease?

	<i>Number of respondents</i>	<i>% of respondents</i>
Never	22	55
Rarely	9	23
Sometimes	4	10
Fairly often	4	10
Not answered	1	3

Comparison with existing literature

Primary care satisfaction and SCD

There are few data collected about SCD patients and their level of satisfaction with primary care services. However, one study showed a 46% non-adherence rate for routine primary care appointments for SCD patients during an eight-month period.⁸ One of the reasons cited for the non-adherence was patient-provider relationships. Those with a positive patient and GP relationship were more likely to attend their clinic appointments.^{9,10} Other studies of chronic disease management showed that poor communication, multiple treating physicians, long waiting periods and past negative experiences with healthcare providers all contributed to patient dissatisfaction with primary care and poor attendance at clinics.¹¹

Patient satisfaction as an indicator of health outcomes

Patient satisfaction is an important indicator of health outcomes. Researchers and healthcare providers have become increasingly interested in measuring patient satisfaction as an indicator of quality of care. Assessing patient satisfaction is a core requirement of contract for GPs in the UK.¹² One study showed that assessing patient satisfaction allows GPs to investigate 'the extent to which their service meets the needs of their client group'.¹³ Another study showed that satisfied patients are more likely to follow treatment protocol because the patient is more likely to believe that the treatment will be effective.¹⁴ In order for satisfaction to be measured in a meaningful way, a valid and reliable measure should

Table 2
Support currently received from general practitioner

	<i>Number of respondents</i>	<i>% of respondents</i>
Pain control	14	35
Collecting prescription	37	93
General SCD advice	3	8
Contraception	2	5
Other (please state)	1	3

SCD, sickle cell disease

be applied. The use of patient questionnaires has been one reliable way to assess patient satisfaction.¹²

The use of questionnaires to measure patient satisfaction

Properly constructed patient questionnaires have been found to be useful in measuring patient satisfaction,¹² but they may also be valuable more specifically for measuring patient satisfaction in primary care and for primary care management of SCD. One study showed that a questionnaire was a valid and valuable tool to use in assessing the health-related quality of life in children with SCD and could serve as 'an important adjunct to determine the effect of SCD on the lives of children'.¹⁵ Other studies showed questionnaires as a valuable tool in measuring quality of care or patient satisfaction in primary care particularly where the questionnaire assesses a specific area that enables the GP or other healthcare provider to identify with which aspects of service patients are less satisfied. This allows¹⁷ for the opportunity to improve a specified area.

Strengths and limitations of this study

Our pilot study successfully identified issues with SCD management at the primary care level through the use of a questionnaire which could help inform the planning of a larger survey. Moreover, this study also identified a need to improve patient satisfaction and engagement

with primary care. Raising awareness among GPs about SCD management was identified as a key method to improve GP–patient relationships and improve primary care clinic attendance.

Although this pilot study increases our understanding of the barriers associated with SCD management in primary care, it has certain limitations which could limit its transferability to areas outside of Northwest London. The questionnaire was based in an area with high prevalence of SCD and the sample size was small and consisted of purposive or volunteer sample. In addition, it is unclear how the identification of barriers in SCD management in primary care relates to the actual uptake of primary versus secondary care in SCD. Future studies would need to examine this link.

Conclusions

Despite its limitations, the study, along with the results of the focus group discussion which reinforced these findings,⁶ provides some valuable information that will give an opportunity to develop a disease-specific intervention which aims to improve patient care and help to ensure that management in primary care is optimized through the establishment of a local enhanced service. In the next stage of this project, we will implement this intervention and evaluate its impact on the management of patients with SCD by general practices in Northwest London.

References

- 1 Pack-Mabien A, Haynes J. A primary care provider's guide to preventive and acute care management of adults and children with sickle cell disease. *J Am Acad Nurse Pract* 2009;21:250–7
- 2 Pace BS, Zein S. Understanding mechanisms of γ -globin gene regulation to develop strategies for pharmacological fetal hemoglobin induction. *Dev Dyn* 2006;235:1727–37
- 3 AlJuburi G, Lavery A, Green SA, *et al.* Trends in hospital admissions for sickle cell disease in England, 2001/02–2009/10. *J Public Health (Oxf)* first published online 31 May 2012:1–7
- 4 NHS Sickle Cell and Thalassaemia Screening Programmes. *Sickle Cell Disease in Childhood Standards and Guidelines for Clinical Care*, 2nd edn. NHS Sickle Cell and Thalassaemia Screening Programmes, London, 2010
- 5 Wong WY. Prevention and management of infection in children with sickle cell anaemia. *Paediatr Drugs* 2001;3:793–801
- 6 Green SA, AlJuburi G, Amobi C, *et al.* Characterising emergency admissions of patients with sickle cell crisis in NHS Brent: observational study. *J R Soc Med Sh Rep* 2012;3:3737
- 7 AlJuburi G, Phekoo KJ, Okoye O, *et al.* Patients' views on improving sickle cell disease management in primary care: focus group discussion. *J R Soc Med Sh Rep* 2012 (submitted)
- 8 Robinson N, Huber H, Jenkins P, *et al.* *Improving Access to Medical Care for Children with Sickle Cell Disease*. The 29th Annual Meeting of the National Sickle Cell Disease Program, Memphis, TN, April 2006
- 9 Freed LH, Ellen JM, Irwin CE Jr, *et al.*, Determinants of adolescents' satisfaction with health care providers and intentions to keep follow-up appointments. *J Adolesc Health* 1998;22:475–9
- 10 Modi AC, Crosby LE, Guilfoyle SM, Lemanek KL, Witherspoon D, Mitchell MJ. Barriers to treatment adherence for pediatric patients with sickle cell disease and their families. *Child Health Care* 2009;38:107–22
- 11 McCarthy K, McGee HM, O'Boyle CA. Outpatient clinic waiting times and non-attendance as indicators of quality. *Psychol Health Med* 2000;5:287–93
- 12 Grogan S, Conner M, Norman P, Willits D, Porter I. *Qual Health Care* 2000;9:210–5
- 13 Avis M, Bond M, Arthur A. Satisfying solutions? A review of some unresolved issues in the measurement of patient satisfaction. *J Adv Nurs* 1995;22:316–22
- 14 Hardy GE, West MA, Hill F. Components and predictors of patient satisfaction. *Br J Health Psychol* 1996;1:65–85
- 15 Panepinto JA, O'Mahar KM, DeBaun MR, Rennie KM, Scott JP. Child Health Care Questionnaire in sickle cell disease. *J Pediatr Hematol Oncol* 2004; 26:574–8
- 16 Raat H, Landgraf JM, Bonsel GJ, *et al.* Reliability and validity of the Child Health Questionnaire – Child Form (CHQ-CF87) in a Dutch adolescent population. *Qual Life Res* 2002;11:575–81
- 17 Harris LE, Swindle RW, Mungai SM, *et al.* Measuring patient satisfaction for quality improvement. *Med Care* 1999;37:1207–13

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Patients' views on improving sickle cell disease management in primary care: focus group discussion

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DECLARATIONS

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Summary

Objectives To assess sickle cell disease (SCD) patient and carer perspectives on the primary care services related to SCD that they receive from their general practitioner (GP).

Design A focus group discussion was used to elicit the views of patients about the quality of care they receive from their primary health-care providers and what they thought was the role of primary care in SCD management. The focus group discussion was video recorded. The recording was then examined by the project team and recurring themes were identified. A comparison was made with notes made by two scribes also present at the discussion.

Setting Sickle Cell Society in Brent, UK.

Participants Ten participants with SCD or caring for someone with SCD from Northwest London, UK.

Main outcome measures Patients' perceptions about the primary care services they received, and a list of key themes and suggestions.

Results Patients and carers often bypassed GPs for acute problems but felt that GPs had an important role to play around repeat prescriptions and general health care. These service users believed SCD is often ignored and deemed unimportant by GPs.

Conclusion Participants wanted the health service to support primary health-care providers to improve their knowledge and understanding of SCD. Key themes and suggestions from this focus group have been used to help develop an educational intervention for general practice services that will be used to improve SCD management in primary care.

Contributorship

KJP, OO and AM

originally conceived the study; GA and KJP undertook the analysis; GA wrote the first draft; and all authors contributed in the revision of the manuscript.

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Introduction

For people with sickle cell disease (SCD), a single point mutation on the haemoglobin molecule creates a lifetime of episodic and illness, until recently, dramatically reduced life-expectancy. A better understanding of the disease and improvements in science, technology, drug therapy and health policy have all contributed to patients with SCD surviving well into adulthood.

SCD is the most common and fastest growing genetic disorder in England. About 350 babies are born each year with SCD and a further 9500 babies are found to be carriers of the disease.¹ England could be dealing with a sickle cell crisis as immigration from Africa and the Caribbean increases disease prevalence, while primary health-care providers still struggle with the disease's multidisciplinary management.² SCD is an illness in which recurrent pain, also referred to as vaso-occlusive crisis, is a chronic concern. Primary care professionals therefore play a key role during the initial presentation of symptoms, as well as the management of long-term complications including such recurrent pain.

In England, London residents account for approximately three quarters of all SCD admissions to hospitals,^{2,3} with the London Borough of Brent being one of the highest risk areas for SCD.^{4,5} Many aspects of SCD can be effectively and efficiently managed in primary care.⁶ Previous studies have shown successful interventions that move management of SCD, particularly uncomplicated sickle cell pain episodes, from tertiary towards primary care.^{7,8}

Focus groups may be useful for obtaining developmental input in managing SCD in primary care. Focus groups may be defined as 'thoughtful planned discussions among participants with similar experiences that allow the moderator to obtain the individuals' cognitive and emotional perceptions in a non-threatening and relaxed environment'.^{9,10} In this paper, we report the findings of a focus group study held at the Sickle Cell Society in the London Borough of Brent. We aimed to identify patient and carer perspectives of general practitioner (GP) knowledge and treatment of SCD, and to identify areas in which improvements could be made. This information helped to design an educational intervention which could help further engage primary health-care professionals in the

care and management of their sickle cell patients. The study was part of a wider service improvement programme to improve the management of SCD in the London Borough of Brent.

Methods

As this study was hypothesis generating, we used qualitative methods – specifically, a focus group design – to learn about the experiences and perception of SCD patients and their carers. The discussion lasted for approximately two hours. The participants were recruited through the Sickle Cell Society in Northwest London and comprised a mixture of ages and genders. Specifically, the group was made up of 10 patients: two men and eight women, aged 9–56 years, all of African/Afro-Caribbean heritage. The focus group was held at the Sickle Cell Society which is conveniently located to where many people with SCD live. The location is also a familiar meeting point where all of the participants felt secure and at ease to voice their opinions. The focus group participants were recruited through a purposive sampling process which aimed to canvas the views of sickle patients and carers living within the specific locality in which the service improvement and evaluation work was being undertaken. Participants were recruited via the Sickle Cell Society, a national UK sickle cell charity, through a general mail out to members of the charity living within this area with information about the study. This was subsequently followed up by a telephone call (CN) to assess interest in participating in the focus group. The discussion was facilitated by a clinical psychologist (KA) from a local NHS Trust experienced in handling patients with SCD who explained the aims of the project to the participants; and who then asked the participants questions regarding their personal experience with primary health care in London. The questions were open-ended and focused on general practice utilization, what patients do first when they are in a SCD crisis, and overall satisfaction with primary care. Eleven observers from the project group were present; two were scribes. As well as making written notes, the discussions were also video recorded.

The key themes used to explore the participants' perceptions and aspirations regarding primary care

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services for SCD are listed in Table 1. The participants were also encouraged to raise their own concerns about SCD management. Following the focus group, the project team (GA and KP) read the transcripts independently and grouped the responses into key themes that would help inform planning of future SCD services. Each item in the data collected during the discussions was compared with the rest of the data to establish analytical groups. Consensus of categories and a final list of key themes identified during the discussions were achieved iteratively through discussion and re-reading of transcripts. As we aimed to present viewpoints, the data are not presented numerically.

Results

The transcripts and recordings were reviewed to identify common perceptions and experiences among the participants.

Accessibility

GP access was an important issue. The participants highlighted that difficulty in obtaining an appointment with the GP, and that long waiting times lead to patients' sidestepping their GP in the event of a painful crisis or other issues relating to SCD. Although GPs are seen as useful for prescribing antibiotics and other repeat prescriptions, they were seen to be unreliable during crises. Hospitals were seen to be more accessible. Participants proposed that GPs should offer more 'out of hours' and house calls for pain management, since many crises occur at night:

I cannot walk in [to a General Practitioner's office] and even if I could, I probably won't be able to see a GP immediately, so I go straight to the hospital if I am sick.

Table 1

Key themes used to generate group discussion

- Frequency in accessing GP care for managing crisis
- Satisfaction level with primary health-care services
- Current use of primary health-care services
- What do you do first when in a SCD crisis?

GP, general practitioner; SCD, sickle cell disease

Doctor-patient relationships

Participants raised the issue of the lack of time to build up rapport and relationships with their GPs. This left patients with SCD with a feeling of dissatisfaction with the quality of the doctor-patient relationship. They felt that GPs were not actively interested or engaged in the progress and treatment of their condition. They encouraged GPs to take a proactive approach and interest in getting to know their SCD patients and to focus on preventive care including preventive management of crisis:

If I take my son to hospital now, the doctors like to have a relationship and know how he is doing. If I take him to the GP there is no relationship; he is a stranger.

[You have a] five minute conversation with the GP. You are in and out. I prefer to go to the hospital, I know them there. I am safe. My nurse knows me.

I know my GP, but does he really know me? I go to the hospital. They know me.

GP knowledge

There was a general feeling among the participants that GPs lack comprehensive knowledge of SCD, and the hospitals were viewed as more specialized in dealing with the condition. They expressed an interest in seeing GPs better informed about SCD, and its implications for management. One participant suggested a GP SCD 'champion(s)' or specialists who could further inform their health professional colleagues about SCD. Another participant added that GPs who are not specialized in SCD need to be briefed by an expert before seeing a SCD patient in the surgery.

They take time to know about other diseases; I feel my disease is not important, I feel I am not important.

Concerns about pain management and treatment process

Because of the organizational barriers previously mentioned, many of the participants had difficulty in seeing the benefit of utilizing a GP during an uncomplicated sickle crisis. They subscribed to a

process that seemed to utilize the emergency department during a crisis and to utilize the GP for repeat prescriptions and immunizations. Patients are still waiting for facilitating factors which would encourage them to use the GP as a means of support, prevention and maintenance during crisis-free periods or during uncomplicated crisis pain episodes.

It takes too long to see a GP. Sometimes something goes wrong while you are waiting for your appointment. Other than a prescription or a non-sickle illness, there is no need for the GP. It is a waste of time. When you have access to a seven-day hospital, why go to a GP?

They [General Practitioners] don't take the time to find out about the patients they have on their books anyways.

Continuity of care and follow-up

One discussion point that surfaced several times was the seeming lack of cohesion and communication between GPs, hospitals and specialty centres. Participants emphasized that there was never follow-up from their GP after they were discharged from the emergency department, hospital or a specialty centre. Participants felt strongly that GPs should liaise with other health-care professionals involved in the management of their sickle condition, and should follow-up with the patient soon after hospital discharge.

[There should be] review letters from the hospital. They [General Practitioners] should see how you are doing. That will be good.

[Following discharge from hospital] why don't they [GPs] ever call the patient and follow-up?

Chronic illness management – wellbeing management

There was a general consensus that many of the frustrations felt by patients with SCD are also shared by sufferers of other chronic illnesses. GPs were seen as treatment and medication prescribers rather than actively engaged in the management of their patients' wellbeing

through prevention of disease and maintenance of health.

The GP consultation is a five minute conversation with the GP... [to get your] medication or antibiotics, or certain jabs, they don't take the time.

The key issues identified through the focus groups are summarized in Table 2.

Discussion

Key findings

The focus group study identified a number of barriers to patients with SCD and their carers engaging fully with primary care services. These ranged from practical difficulties accessing services when required, poor communication between primary and secondary care professionals and a perceived lack of confidence in the ability of primary care professionals to provide chronic disease management specific to SCD.

Education, communication and quality improvement

The issues identified during the focus group discussion have helped define key areas in need of improvement for SCD management. Some of the key themes and useful suggestions have been used to develop a GP education intervention to improve GP management of SCD in primary care. Part of the education intervention has been facilitated by developing a SCD template for data entry compatible with a main electronic patient record system used by GPs. These electronic medical

Table 2

Issues identified by focus group discussion

- Access and difficulty in obtaining urgent appointments
- Poor communication between health professionals
- Poor follow-up and discharge planning processes
- No proactive role in maintaining health and in preventive care
- Limited knowledge about sickle cell disease
- Limited knowledge about the standards and guidelines for disease management
- Concerns about pain management

record systems are now widespread in UK primary care and have helped improve recording on patients with other chronic diseases.¹¹ In the longer term, these electronic patient records could be linked to clinical decision support systems or disease recording templates to help improve recording of information by primary care teams and the quality of primary care received by patients.¹²

Future studies could focus on other issues identified such as auditing and improving communication between different health-care providers. This could be facilitated by GPs receiving timely information on hospital admission and hospitals advising patients to book an appointment with their GP for review after discharge. Receiving proactive care from the GP could also be achieved by identifying areas with a relatively high prevalence of SCD and designing a local quality improvement scheme. Many patients with chronic illnesses do receive proactive care from their GPs but these are generally patients with the diseases covered in the GP Quality and Outcomes Framework.¹³ At present, SCD is not a part of this framework but could be considered for future inclusion in areas where there is a high prevalence of SCD.

Comparison with existing literature

Previous studies have shown that SCD patients are dissatisfied with the quality of care that they receive from their GPs and at the primary care level.¹⁴ They also show that primary care is still a strong place to deal with chronic illness as it offers continuity, coordination and comprehensiveness¹⁵ and that assessing satisfaction is an important part of evaluating comprehensive quality of care.¹⁶ Patient satisfaction is regarded as the 'ultimate outcome of the delivery of health care' and measuring and responding to satisfaction can be important with regard to treatment compliance and over or underutilization of the health-care system.¹⁷ The strengths and limitations of the current focus group discussion also support the findings of similar studies that have identified focus group discussions as a means for qualitative data collection.

Strengths of the focus group discussion

The focus group discussion provided a forum for a broad range of participants to express their

concerns about the quality of care they received from their primary health-care providers, their opinions on what the role of primary care in managing SCD should be and to discuss their perceived barriers to utilizing primary care services. This method has been demonstrated as an appropriate approach to investigate these issues.¹⁸ Furthermore, focus groups can provide a space for an open dialogue tailored to the level of the participants¹⁹ and allows the facilitator to clarify and push for more detailed responses to enrich the understanding of the issues.²⁰

The use of a focus group as a data collection technique was particularly useful with SCD to ensure sensitivity to cultural variables, which is why they are now so often used in cross-cultural research and work particularly well with ethnic minorities. SCD is a disease that disproportionately impacts people of African and Caribbean origin; the focus group setting allowed us to reveal the frustration felt as an ethnic minority receiving health-care services for a predominantly race-related disease. Consequently, it makes them useful in studies examining why different sections of the population make differential use of health services.^{18,19}

The group dynamic also worked to facilitate the discussion of difficult or uncomfortable topics because the less reserved members of the group 'broke the ice' for the more reserved participants.¹⁹ Some researchers have also noted that group discussions can generate more criticism of the health-care system than interviews.²¹ For example, Geis *et al.*, in their study found that 'there were more angry comments about the medical community'¹⁹ in the group discussions than in the individual interviews: 'perhaps the synergism of the group "kept the anger going" and allowed each participant to reinforce another's vented feelings of frustration and rage'.²²

Limitations of the focus group discussion

Our focus group discussion results could have benefited by having more participants and more sessions. Some researchers also argue that focus groups are limited in their ability to draw inferences for large groups or populations and their incapacity to test hypotheses in traditionally designed experiments.¹⁰ Others have also claimed that focus groups can lead to 'tagging' which

means that participants agree for the sake of the group's momentum and that, with regard to more sensitive topics, interviews work better because the participant feels a greater level of anonymity than when part of a group discussion.¹⁰ Such points show that the use of focus groups may be subject to bias and need to be individually evaluated.²⁰ Focus groups may not add value to every study but can work well when combined with other forms of qualitative and quantitative methods.^{10,23}

Implications for future research and clinical practice

The focus group discussion on SCD provided an opportunity to not only voice the frustrations of participants but created a venue whereby solutions could be proposed. The discussion was candid, clear and offered perspectives about the participants' individual and collective needs that could not otherwise be obtained from health-care professionals. They offered constructive advice on what they needed to feel satisfied as health-care consumers and gave a clear understanding about which areas we needed to focus the design of the primary care practice intervention. They also earmarked issues that need to be addressed in future research including auditing and improving communication between the different providers of health care and recommending quality improvement schemes in areas with a high prevalence of SCD.

Including patients in the development and implementation of a GP educational intervention may be an efficient and effective way to help pilot a new programme aimed at meeting the needs of patients with SCD. Future focus groups may explore with what areas in particular the participants are not satisfied and how to incorporate into future programme planning some of the preliminary suggestions made at the first focus group.

Conclusions

This study has highlighted that SCD patients and carers have many criticisms about the current quality of services for managing patients with SCD in primary care. The study has helped provide key themes and useful suggestions that

have been used to develop an educational intervention for general practices that will be used to improve SCD management in primary care.

References

- 1 NHS Sickle Cell and Thalassaemia Screening Programmes. *Sickle Cell Disease in Childhood Standards and Guidelines for Clinical Care*. 2nd edn. England: The NHS Sickle Cell and Thalassaemia Screening Programme in partnership with Sickle Cell Society London, 2010
- 2 NCEPOD. A Report of the National Confidential Enquiry into Patient Outcome and Death: A Sickle Crisis? England: NCEPOD London, 2008
- 3 AlJuburi G, Laverty A, Green SA, *et al*. Trends in hospital admissions for sickle cell disease in England, 2001/02–2009/10. *J Public Health (Oxf)* (first published online), 31 May 2012, doi:10.1093/pubmed/fds035 and pp 1–7
- 4 Green SA, AlJuburi G, Amobi C, *et al*. Characterising emergency admissions of patients with sickle cell crisis in NHS Brent: observational study. *J R Soc Med Sh Rep* 2012;3:37
- 5 Hickman M, Modell B, Greengross P, *et al*. Mapping the prevalence of sickle cell and beta thalassaemia in England: estimating and validating ethnic-specific rates. *Br J Haematol* 1999;104:860–7
- 6 Lenfant C, *et al*. *The Management of Sickle Cell*. NHLBI, NIH Publication No. 02-2117 [Internet]. 2002. See http://www.nhlbi.nih.gov/health/prof/blood/sickle/sc_mngt.pdf
- 7 Coleman P, Irons R, Nicholl J. Will alternative immediate care services reduce demands for non-urgent treatment at accident and emergency? *Emerg Med J* 2001;18:482–7
- 8 Benjamin LJ, Swinson GI, Nagel RL. Sickle cell anemia day hospital: an approach for the management of uncomplicated painful crisis. *Blood* 2000;95:1130–6
- 9 Wyatt TH, Krauskopf PB, Davidson R. Using focus groups for program planning and evaluation. *J School Nurs* 2008;24:71–7
- 10 Heary CM, Hennessy E. The use of focus group interviews in pediatric health care research. *J Pediatr Psychol* 2002;27:47–57
- 11 Majeed A. Sources, uses, strengths and limitations of data collected in primary care in England. *Health Stat Q* 2004;21:5–14. See <http://goo.gl/hW0Do>
- 12 Pappas Y, Anandan C, Liu J, Car J, Sheikh A, Majeed A. Computer-assisted history-taking systems (CAHTS) in health care: benefits, risks and potential for further development. *Inform Prim Care* 2011;19:155–60
- 13 GP Quality and Outcomes Framework [Internet]. [Cited 17 July 2011]. See <http://www.qof.ic.nhs.uk/>
- 14 Anie KA, Steptoe A, Bevan DH. Sickle cell disease: pain, coping and quality of life in a study of adults in the UK. *Br J Health Psychol* 2002;7:331–44
- 15 Rothman AA, Wagner EH. Chronic illness management: what is the role of primary care? *Ann Intern Med* 2003;138:256–62
- 16 Ireys HT, Perry JJ. Development and evaluation of a satisfaction scale for parents of children with special health care needs. *Pediatrics* 1999;104:1182–91
- 17 Kelley MA, Alexander CS, Morris NM. Maternal satisfaction with primary care for children with selected chronic conditions. *J Community Health* 1991;16:213–24

- 18 Basch CE. Focus group interview: an underutilised research technique for improving theory and practice in health education. *Health Educ Q* 1987;14:411–48
- 19 Kitzinger J. Introducing focus groups. *BMJ* 1995;311:299–302
- 20 Wong LP. Focus group discussion: a tool for health and medical research. *Singapore Med J* 2008;49:256–61
- 21 Watts M, Ebburt D. More than the sum of the parts: research methods in group interviewing. *Br Educ Res J* 1987;13:25–34
- 22 Geis S, Fuller R, Rush J. Lovers of AIDS victims: psychosocial stresses and counseling needs. *Death Stud* 1986;10:43–53
- 23 Morgan DL. *Focus Groups as Qualitative Research* 1997. London: Sage, 1997

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Characterizing emergency admissions of patients with sickle cell crisis in NHS Brent: observational study

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SG, RB and CA
conceived of the
study; SG performed
all analyses, GA
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Summary

Objectives To characterize emergency admissions for patients with sickle cell crisis in NHS Brent and to determine which patients and practices may benefit most from primary care intervention.

Design Observational study

Setting Emergency departments attended by residents of the London borough of Brent

Participants Patients with sickle cell disease registered with a general practitioner (GP) in the borough of Brent

Main outcome measures Analysis of admissions between January 2008 and July 2010 that included length of stay (average and <2 days versus ≥2 days) by age group and registered GP practice.

Results Thirty six percent of sickle cell disease admission spells resulted in a length of stay of less than two days. Seventy four percent of total bed days are associated with patients with more than one admission during the period of analysis, i.e. multiple admissions. Two general practices in Brent were identified as having the highest number of patients admitted to the emergency department for sickle cell crisis and may benefit most from primary care intervention.

Discussion Patients with short length of stay and multiple admissions may be potentially amenable to primary care intervention. The practices which have the highest numbers of sickle cell disease patients who frequently seek emergency care will be earmarked for an education intervention designed to help further engage general practitioners in the care and management of their sickle cell patients.

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Introduction

Sickle cell disease (SCD) is the most common genetic disorder worldwide, predominantly affecting Black and ethnic minorities (BME) groups. Incidence and prevalence in England has increased significantly in recent decades, particularly in urban areas.¹ The growth in affected populations and the associated rising costs have attracted the attention of the healthcare community to SCD as an important public health issue. While just twenty years ago, infants born with SCD often did not survive childhood, a better understanding of the disease and improvements in medical technology and drug therapy have all contributed to patients with SCD surviving well into adulthood. However, positive health and life-expectancy outcomes are dependent on disease management and continuity of care throughout childhood, adolescence and adulthood.²

SCD is a genetic blood disorder characterized by abnormal, crescent-moon shaped red blood cells. These so called 'sickled' cells become lodged inside blood vessels, thereby obstructing blood flow and impeding efficient oxygen delivery to tissues throughout the body. Severe blockages cause episodes of acute pain, referred to as 'crisis,' which may be triggered by a range of physical and psychological stresses, including but not limited to infection, pregnancy, surgery, anxiety, or depression. If not treated promptly, crises can result in internal organ and tissue damage, particularly to the lungs, kidneys, liver and bones. The frequent recurrence of crisis can lead to chronic complications such as leg ulcers, blindness, and stroke. Acute chest syndrome, or chest crisis, is a common and particularly dangerous complication that is currently the leading cause of death among SCD patients.³ Ambulatory care strategies such as nutritional counselling, folic acid supplementation, pain medication protocols, vaccinations and antibiotics for the prevention and treatment of infection, are essential to SCD management.⁴

In England, approximately three quarters of all SCD admissions to hospitals are by London residents. SCD is also one of the most common reasons to be admitted to hospital and has one of the highest rate of multiple admissions for individual patients. The distribution of SCD in London also shows 80% of all admissions are in people living in the most deprived areas.^{5,6}

The London Borough of Brent is one high prevalence area for SCD.^{7,8} A pre-study focus group held at the Sickle Cell Society in Brent revealed low patient confidence in general practitioner (GP) knowledge of management strategies for SCD. The perception that GPs lack the skills to properly treat and control SCD may help to explain why SCD patients and their caregivers expressed a preference for attending the hospital emergency department (ED) during a crisis.^{9,10}

The rising cost of healthcare places the health system in England under increasing pressure to reduce such inappropriate hospital admissions.^{11,12} The national confidential enquiry into patient outcome and death reports that a multidisciplinary and multi-agency approach is needed in the ongoing pain management of patients with sickle cell disease, and that this should largely be taking place outside of hospitals.⁶ The purpose of our analysis of emergency admissions of residents in Brent with SCD aims to provide a picture of admissions presenting at the ED and to identify areas for improving primary care intervention to improve the care of patients with SCD and reduce such admissions.

Methods

Data source

SUS (Secondary Uses Service) data were provided by NHS Brent for all admissions through the Emergency Department using temporal, admission and diagnostic parameters for the period January 2008 and July 2010 (Figure 1). Before analysis, data quality was checked by ensuring that episodes were commensurate with the admission and discharge dates of spells. Whilst in hospital patients may be under the care of a number of different consultants, each known as a finished consultant episode (FCE) and a spell is comprised of one or more continuous FCE. The FCE data was converted to spells by combining contiguous FCEs through cross-referencing a combination of postcode, age and registered practice. (Figure 2).

Analysis and outcome measures

Analysis was undertaken to compare admissions across the three diagnostic groups: sickle

Figure 1

Conversion of full consultant episode (FCE) data to admission spell

Postcode	Age	Primary ICD-10	Primary ICD-10 Name	GP Practice Name	Episode start date	Episode end date
SS1 2SS	29	D571	Sickle Cell Anaemia without crisis	Medical Centre A	18-May-08	20-May-08
SS1 2SS	29	D571	Sickle Cell Anaemia without crisis	Medical Centre A	20-May-08	24-May-08

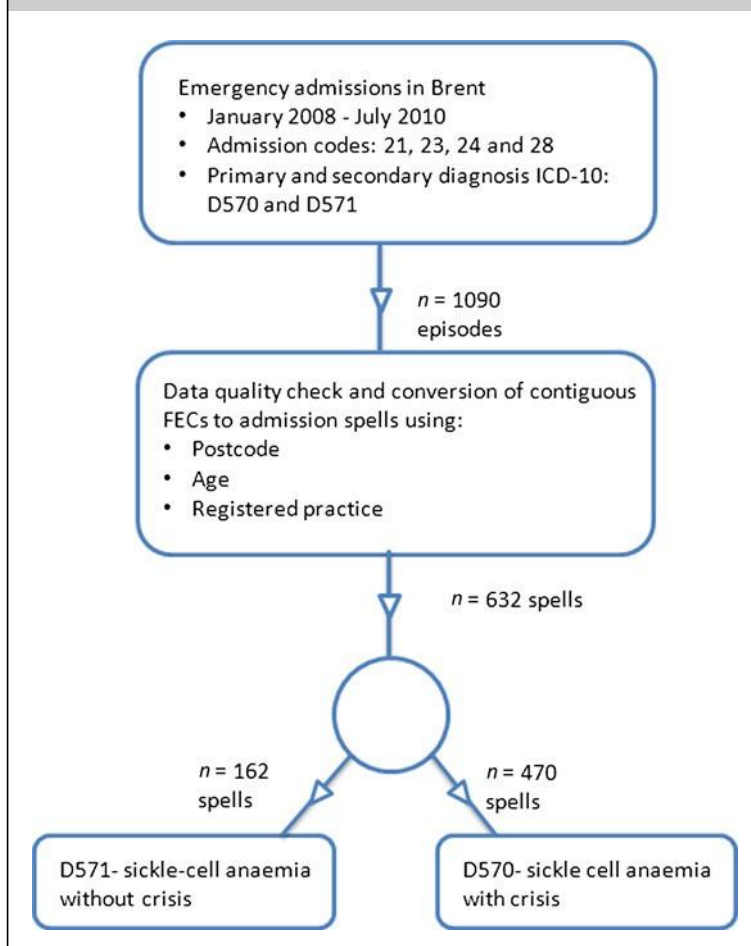


Postcode	Age	Primary ICD-10	Primary ICD-10 Name	GP Practice Name	Admission date	Discharge date
SS1 2SS	29	D571	Sickle Cell Anaemia without crisis	Medical Centre A	18-May-08	24-May-08

A single spell can be made from a number of episodes and is generated by matching contiguous admission episodes using postcode, age and GP practice name.

Figure 2

Schematic demonstrating the extraction, filtering, conversion and analysis of emergency admission data from NHS Brent



cell anaemia with crisis, sickle cell anaemia without crisis and sickle cell trait. Descriptive analysis of patients admitted with a diagnosis of sickle cell anaemia with crisis (D570) was undertaken; the main outcome measure was length of stay (average and <2 days versus ≥2 days) by age group and registered GP practice. Additional analysis of the frequency of individual patient admission was completed by linking patient spells over the period of analysis.

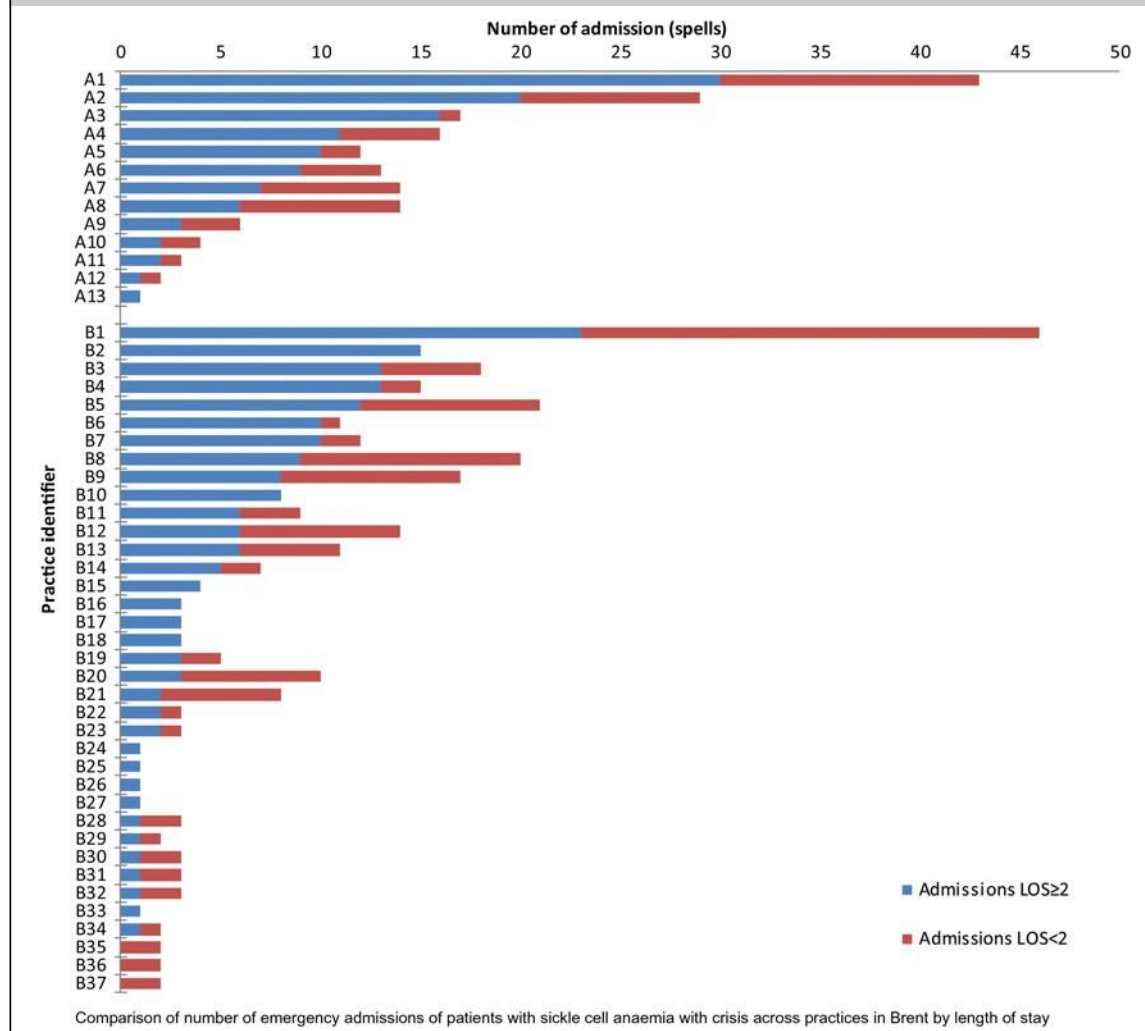
Results

Hospital emergency admissions

A total of 1090 episodes were identified, during the period January 2008 and July 2010, as emergency admissions, with either a primary or secondary diagnosis of sickle cell anaemia with crisis (D570), sickle-cell anaemia without crisis (D571) or sickle cell trait (D573). The conversion of episode data to admissions spells resulted in 632 unique spells, of which 470 spells included a diagnostic code for sickle cell anaemia with crisis (74.4 percent), 97 spells with sickle-cell anaemia without crisis (15.4 percent) and 65 spells with sickle cell trait (10.2 percent) (Figure 3). Only patients admitted with a primary or secondary diagnosis of sickle cell anaemia with crisis were included in subsequent analysis.

Figure 3

Emergency admissions of patients registered at practices in the London borough of Brent



Length of stay and age

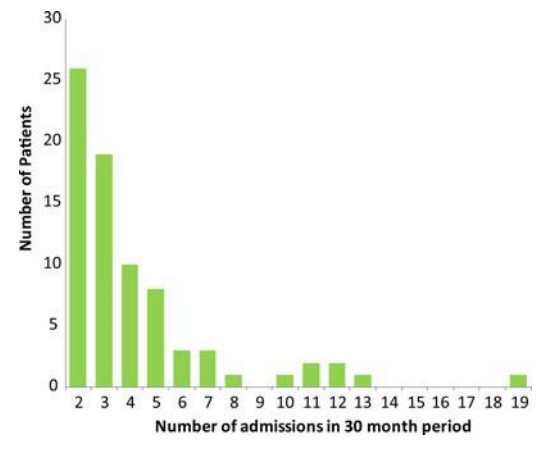
The most frequently admitted age group were patients between 21 – 25 years (85/470 spells). Thirty six percent of admissions resulted in a length of stay of less than two days (163/470 spells) (Figure 4).

Practice level analysis

The practice level analysis included all practices in NHS Brent where registered patients were admitted over the 30 month study period. Practices were split into two groups, those involved

in a current Sickle Cell Disease educational programme (Group A) and those not involved (Group B). The educational programme started after the current study end date (July 2010). Group A comprised of 15 GP practices plus one GP access centre with approximately 20 full-time and 9 part-time GPs and an estimated 50 registered patients with SCD. Group B is comprised of 55 practices with approximately 122 full-time and 61 part-time GPs, and an estimated 155 registered patients with SCD. Two practices were identified in Group A with the highest numbers of admissions A7 (43 spells) and A6 (29 spells), and have some of the highest rates of emergency

Figure 4
Emergency admissions between January 2008 and July 2010 for sickle cell crisis in NHS Brent by frequency of spells for individual patients admitted (total number of patients 77)



admissions for Sickle Cell crisis. Within Group B, practice B3 had the highest number (47 spells) (Table 1).

Multiple admissions

Seventy-seven patients accounted for multiple admissions (327 spells) and 143 patients were single admissions (143 spells). Multiple admissions accounted for 1530 bed days and 74% of the total number of admissions (Table 2).

Discussion

Main findings

This analysis showed that 36% of SCD patient admissions resulted in a length of stay of less than two days, and 74% of total bed days are inpatients with multiple admissions. Patients with short length of stay and multiple admissions may be good candidates for primary care interventions to reduce their risk of SCD-related complications and admissions. They may also benefit from more supportive management when they attend emergency departments, sickle cell centres and outpatient clinics, as many aspects of the management of SCD patients are outside the remit of primary care teams. The study also

Table 1
Parameters for data extraction

Parameter	Details
Time period	January 2008 to July 2010 (30 months)
Method of admission codes	21- Accident and emergency or dental casualty department of the Health Care Provider 23- Bed bureau 24- Consultant Clinic of this or another Health Care Provider 28- Other means
Primary and Secondary diagnosis ICD- 10 codes	- D570- sickle cell anaemia with crisis - D571- sickle-cell anaemia without crisis - D573- sickle cell trait

SUS data were provided by NHS Brent for all emergency hospital admissions of patients living in the London borough of Brent with one of three ICD-10 codes for the period January 2008 until July 2010

Table 2

Comparison of patients admitted with sickle cell crisis by length of stay (LoS) for various age groups (0–75 years)

Age Group	Number of admissions		Average LoS (days)
	LoS $\geq$ 2 days	LoS < 2 days	
0-5	16	4	3.1
6-10	9	11	3.3
11-15	26	13	2.9
16-20	36	17	4.0
21-25	55	30	4.8
26-30	44	24	3.5
31-35	39	20	3.5
36-40	38	19	7.7
41-45	4	6	4.4
46-50	26	14	3.2
51-55	4	3	5.6
56-60	3	1	22.0
61-65	3	1	8.5
66-70	0	0	0.0
71-75	1	0	2.0

Most patients admitted were between the ages of 21-25 years and admissions with a length of stay less than 2 days form a significant proportion of all admissions.

identified two GP practices, currently part of a GP education programme, that have patients who frequently seek emergency care for SCD-related problems. The Group A Practices will be assessed to identify any benefits realized from an educational intervention designed to help further engage GPs in the care and management of their sickle cell patients. Such benefits may include increasing the quality of care for patients with SCD and reducing the inappropriate use of the ED.

Limitations of the analysis

The analysis has certain limitations which could limit its transferability to areas outside of north-west London. The analysis is based on an area

with a high prevalence of SCD. GPs in such areas may be more open to the idea of an educational intervention and more committed to its implementation. In addition, the data did not include those patients that attended the ED but were not admitted into hospital. Future studies may need to account for this subset of patients, as they may also have unmet needs and make high use of NHS resources. A planned GP educational intervention may reduce the number of patients admitted for two days or less or with multiple admissions, but we do not yet know that these patients could be managed in primary care.

Comparison with Existing Literature

This analysis is consistent with other studies that have identified potentially inappropriate use of acute inpatient care as a growing concern, and earmarked the important benefits of GP involvement and primary care in limiting this utilization.^{13–15} Inappropriate ED attendance distracts ED staff attention and resources from severe and acute cases that need urgent hospital care; adds to the waiting times in the ED, compromises the quality of care a patient receives and increases cost.¹⁶ While severe crises require the resources of secondary care and tertiary care, the long-term management of uncomplicated sickle cell pain episodes may be more effectively and efficiently managed in a primary care setting.¹⁷ Interventions that move management of SCD, particularly uncomplicated sickle cell pain episodes from tertiary towards primary care, have been successfully implemented elsewhere.^{15,18}

One study showed an average annual reduction of 804 bed days over a three year period, with the average daily cost of a specialist haematology inpatient bed estimated at £356.^{12,19} Another study saw a 40% reduction in sickle cell ED attendance by introducing a local day hospital concept where SCD patients could bypass the ED and seek primary care plus within a hospital setting.¹⁸

Implications for future research and clinical practice

This analysis of Sickle Cell crisis admissions provided an opportunity to not only characterize the admissions but to identify practices with

higher levels of admissions especially related to multiple admissions or short length of stays. It also establishes a baseline to monitor progress and validate associations in relation to improving primary care support for patients and reducing ED admissions. The strategic engagement of a small number of GPs followed by further rollout across the London Borough of Brent, will ensure that the delivery of the GP education package is targeted where needed. This programme will make use of the joint work that Brent is carrying out with voluntary groups in its locality with the aim of improving patient experience and addressing health inequalities.²⁰ The data analysis can also facilitate discussions between primary and secondary care as part of the multi-disciplinary, multi-professional project team, as well as encourage ongoing work to explore patient satisfaction through the enhanced care delivered by GPs and with the aim of improving the patient experience.

Conclusions

Patients with SCD make relatively high use of urgent care services, including inpatient care. More appropriate management of these patients in the community may limit this demand and contribute to better health outcomes for patients, as well as economic benefits for the local health economy.

References

- 1 Streetly A, Clarke M, Downing M, *et al.* Implementation of the newborn screening programme for sickle cell disease in England: results for 2003–2005. *Journal of Medical Screening* 2008;15:9–13
- 2 Quinn C, Rogers Z, McCavit T, Buchanan G. Improved survival of children and adolescents with sickle cell disease. *Blood* 2010;115:3447–52
- 3 Moore F, Mortimer P, Wooller J. *Updated Sickle Cell Crisis Guidelines*. Joint Royal Colleges Ambulance Liaison Committee. 2009
- 4 Sickle-cell disease and other haemoglobin disorders. Fact sheet N°308. WHO, January 2011. Online <http://www.who.int/mediacentre/factsheets/fs308/en/index.html> (last accessed 18 August 2011)
- 5 London Health Observatory. 2006. Analysis of Frequent Hospital Users, by PCT 2003/4. [Online]. <http://www.lho.org.uk/viewResource.aspx?id=10286> (last accessed July 2011)
- 6 Lucas SB, Mason DG, Mason M, Weyman D. *A Sickle Crisis?* National Confidential Enquiry into Patient Outcome and Death. 2008
- 7 Northwest London Sickle Cell & Thalassaemia Managed Clinical Network. *Annual Report & Action Plan 2006–7*. 2005
- 8 Hickman M, Modell B, Greengross P, *et al.* Mapping the Prevalence of Sickle Cell and Beta Thalassaemia in England: estimating and validating ethnic-specific rates. *British Journal of Haematology* 1999;104:860–7
- 9 Okoye O, Aljuburi G. Patient Perspective of Primary Care Management of Sickle Cell Disease. The Society for Academic Primary Care: London and South-East Region Annual Scientific Meeting 2011
- 10 Lega F, Mengoni A. Why non-urgent patients choose emergency over primary care services? Empirical evidence and managerial implications. *Health Policy* 2008;88:326–38
- 11 Carret ML, Fassa AG, Domingues MR. Inappropriate use of emergency services: a systematic review of prevalence and associated factors. *Cad. Saúde Pública* 2009;25:7–28
- 12 Wright J, Bareford D, Wright C, *et al.* Day case management of sickle pain: 3 years experience in a UK sickle cell unit. *British Journal of Haematology* 2004;126:878–80
- 13 Boeke JP, van Randwijck-Jacobze ME, de Lange-Klerk EMS, Grol SM, Kramer M, van der Horst HE. Effectiveness of GPs in accident and Emergency Departments. *British Journal of General Practice* 2010;60:378–84
- 14 Dale J, Lang H, Roberts JA, Green J, Glucksman E. Cost effectiveness of treating primary care patients in accident and emergency: a comparison between general practitioners, senior house officers and registrars. *BMJ* 1996;312:1340–4
- 15 Coleman P, Irons R, Nicholl J. Will alternative immediate care services reduce demands for non-urgent treatment at accident and emergency? *Emergency Medical Journal* 2001;18:482–7
- 16 Martin A. 'Inappropriate' attendance at an accident and emergency department by adults registered in local general practices: how is it related to their use of primary care? *Journal of Health Service Research and Policy* 2002;7:160–5
- 17 Lenfant C (ed.). *The Management of Sickle Cell*. NIH publication No. 02-2117, 4th Edition. [Online] Bethesda: National Institute of Health; 2002. Available from: http://www.nhlbi.nih.gov/health/prof/blood/sickle/sc_mngt.pdf (last accessed July 2011)
- 18 Benjamin LJ, Swinson GI, Nagel RL. Sickle cell anemia day hospital: an approach for the management of uncomplicated painful crisis. *Blood* 2000;95:1130–6
- 19 Netten A, Curtis I. *Unit Costs of Health and Social Care 2002*. Personal Social Services Unit, University of Kent, 2002
- 20 Majeed A, Banarsee R, Molokhia M. Health Disparities and Community Participation in England. *Journal of Ambulatory Care Management* 2009;32:297–301

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A retrospective analysis of the cost of hospitalizations for sickle cell disease with crisis in England, 2010/11

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ABSTRACT

Background Sickle cell disease (SCD) is an inherited blood disorder which may result in a broad range of complications including recurring and severe episodes of pain—sickle 'crises'—which require frequent hospitalizations. We assessed the cost of hospitalizations associated with SCD with crisis in England.

Methods Hospital Episodes Statistics data for all hospital episodes in England between 2010 and 2011 recording Sickle Cell Anaemia with Crisis as primary diagnosis were used. The total cost of admissions and exceeded length of stay due to SCD were assessed using Healthcare Resource Group tariffs. The impact of patients' characteristics on SCD admissions costs and the likelihood of incurring extra bed days were also examined.

Results In 2010–11, England had 6077 admissions associated with SCD with crisis as primary diagnosis. The total cost for these admissions for commissioners was £18 798 255. The cost of admissions increases with age (children admissions costs 50% less than adults). Patients between 10 and 19 years old are more likely to stay longer in hospital compared with others.

Conclusion SCD represents a significant cost for commissioners and the NHS. Further work is required to assess how best to manage patients in the community, which could potentially lead to a reduction in hospital admissions and length of stay, and their associated costs.

Keywords chronic disease, costs, hospitalization, management and policy, public health, sickle cell

Introduction

Sickle cell disease (SCD) is an inherited blood disorder that dominantly affects people of African and Caribbean origin.¹ The disease is known for its painful episodes (also known as vaso-occlusive crisis or sickle crisis) which lead to a significant number of hospital admissions and readmissions each year.^{2–4}

A recent study in England showed a 58% increase in the overall SCD admission rate between 2001 and 2010 using Hospital Episodes Statistics (HES) data for England.⁵

SCD is the fastest growing and most frequent inherited disorder in England; "12 500 people are living with the condition⁶ and 240 000 have SCD trait.⁷ SCD may be less common than other known chronic diseases^{8–10} but its growth and high impact on hospital admissions poses a potential cost burden for the National Health Service, in the addition to the impact it has on patients and their families, particularly in areas with a high prevalence such as London.

With the exception of a recent National Institute of Clinical Excellence (NICE) report,¹¹ there is little data about the

overall cost of health-care utilization associated with SCD and hospital admissions in England. In contrast, several studies have been conducted in the USA where patients with SCD frequently attend emergency departments or are admitted as inpatients and account for the majority of health-care expenses associated with the disease.^{12–16} Expenditure associated with SCD can be used to raise awareness of the severity of this condition and to explore innovations in prevention or management of disease complications. Given this is the most common inherited disorder in the UK, data on costs associated

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with hospital admissions can provide information about the impact of SCD on the National Health Service (NHS) and be of value to health economists, policy-makers and commissioners. The aim of this study was to estimate the costs for commissioners associated with hospital admissions due to SCD in England.

Methods

HES provide data on all NHS hospital admissions, emergency department attendances and outpatient visits in England, including treatment performed in private hospitals but funded by the NHS. This study uses HES data on hospital admissions for the period from April 2010 to March 2011. HES data contain details on both the reason that patients are admitted to hospital (known as the 'primary diagnosis') and any other conditions they have, which are coded using ICD-10.¹ This analysis uses patients with a primary diagnosis of 'sickle cell anaemia with crisis' (D57.0).

As data regarding the cost and resource consumption for hospital admissions, including SCD admissions, were not available, we used the Healthcare Resource Groups (HRG) tariffs as a proxy to assess the cost for commissioners. HRGs form the basic currency for payment of NHS services by relying on an informed assumption that health-care costs are exclusively driven by diagnosis or procedure. The English HRG tariffs provide cost data for elective and non-elective hospital episodes and additional per diem cost for very long stays that exceed the so-called 'trim point', which marks the expected LOS for each HRG. Estimated costs are reported as a national schedule of reference costs and are also aggregated by hospital to provide the reference cost index.^{17,18}

Despite the tariffs, the real costs of SCD borne by the NHS appear not to be reflected; this is a first attempt to assess the cost of admissions due to SCD adopting the commissioners perspective. There are only three available HRG tariffs for SCD which are coded as:

- (1) PA47Z Sickle cell anaemia with crisis (for children admissions).
- (2) SA10D Sickle cell anaemia with complications (CC) (including SCD with crisis).
- (3) SA10F Sickle cell anaemia without complications (CC).

Because there are no specific tariffs for each ICD-10 code, we made certain assumptions which are consistent with those of

the NICE report.¹¹ The tariffs were linked only to primary diagnosis:

- (a) ICD-10 'Sickle cell anaemia with Crisis' (D57.0) is linked to HRG PA47Z 'Sickle cell anaemia with crisis' (age between 0 and 19 years);
- (b) ICD-10 'Sickle cell anaemia with Crisis' (D57.0) is linked to HRG SA10D 'Sickle cell anaemia with CC' for adult admissions (age ≥20 years);
- (c) ICD-10 'Sickle cell anaemia without crisis and other sickle cell disorders (D57.2 – 8) might be linked with HRG code SA10F 'Sickle cell anaemia without CC;

All other admissions, in which the SCD with crisis appeared in any other of the diagnoses, but not as a primary diagnosis, were excluded from our analysis. This will underestimate the cost of admissions linked to SCD, but we have adopted a conservative approach and not included extra costs that we are not certain have been appropriately coded for SCD. Both emergency and elective admissions have been analysed separately, as they have different tariffs, and to show the impact of emergency admissions due to SCD.

The length of stay (LOS) for each admission was considered in the analysis to assess the cost due to hospitalizations that exceed the trim point of the HRG tariff. The trim point identifies the LOS covered by the tariff after which an extra daily payment is required for each specific HRG. The trim points vary according to the type of HRG and type of admission (emergency versus elective admission) and extra days carry an extra cost. For HRG PA47Z Sickle Cell Anaemia with Crisis, the trim point is 9 days for emergency admissions and 8 days for elective admissions; for HRG SA10D Sickle Cell Anaemia with CC, the trim point is 30 days both for emergency and elective admissions. Therefore, an LOS exceeding the trim point will increase the cost by £234 for each extra day for children admissions and £204 for adult admissions.

HRGs tariffs and extra LOS payments have been adjusted for the Market Forces Factor (MFF) to take into account of specific differences in the costs of providing services across providers.¹⁹ For example, admissions in Central London would typically be more expensive as labour and building costs are higher and this has been reflected applying a specific MFF to each provider.¹⁹

Analysis

The cost of admissions associated with SCD was measured as follows:

- (1) The total number of episodes with a primary diagnosis containing ICD-10 code 'Sickle cell anaemia with Crisis' (D57.0.) data was taken from HES with a distinction between emergency and elective admissions.

¹The ICD-10 is a system of coding created by the World Health Organization that notes various medical records including diseases, symptoms, abnormal findings and external causes of injury.

- (2) The total number of admissions for patients between 0 and 19 years has been multiplied by the correspondent HRG tariff PA47Z '*Sickle cell anaemia with crisis*', respectively in emergency and elective admission, adjusted for MFF for each provider.
- (3) The total number of admissions for patients 20 years or older (adults) has been multiplied by the correspondent HRG tariff SA10D '*Sickle Cell Anaemia with CC*' respectively in emergency and elective admission, adjusted for MFF for each provider.
- (4) The total cost of admissions for SCD is the sum of the total cost of admissions in emergency and elective for children and adults.

The cost of extra LOS for admissions associated with SCD has been assessed as follows:

- (1) For each episode presenting as primary diagnosis containing the ICD-10 code '*Sickle cell anaemia with Crisis*' (D57.0.) the extra LOS has been assessed as the difference between the number of days spent in hospital (LOS) and the trim point from the HRG tariff.
- (2) The total number of days in hospital exceeding the trim point has been multiplied by the daily extra cost (£234 per day for children admissions and £204 for adults), adjusted for the MFF for each provider.
- (3) The total cost of extra LOS is the sum of the extra cost of LOS of emergency and elective admissions for adults and children admissions.

The analysis was carried out at a national level for England and the results are reported for England, London and English Primary Care Trusts (PCTs)².

A linear regression model was used to analyse the impact of age and gender on SCD cost. Here we have log transformed the costs and present our results as the percentage difference from the reference group. We analysed the data separately for London and the rest of the country to examine whether any associations differ between locations. We also used logistic regression to examine the association between age and gender on incurring extra bed days.

Results

In 2010–11, England had 6077 admissions associated with SCD with crisis as a primary diagnosis. The total cost for

these admissions was £18 798 255 of which £17 085 288 (91%) was for emergency admissions (Table 1). Adult admissions represent 84.5% of all admissions, representing a cost of £14 370 931 for emergency admissions. Children admissions represent 15.5% of total admissions, and the cost of emergency admissions was £2714 357.

In total, the LOS exceeded the trim point by 1144 days, for an extra cost of £294 697 (93.1% for emergency admissions) (Table 1). For emergency admissions, the extra LOS is 438 days for children and 624 days for adults, for an extra total cost of £274 104. Because the tariff for children has a shorter trim point compared with adults (9 days instead of 30 days) and an extra day costs more, long admissions for children represent a higher cost in terms of LOS.

Within England London had the highest number of admissions associated with SCD, with 4363 admissions for SCD crisis as primary diagnosis at a total cost of £14 223 139 of which £13 984 884 were for emergency admissions (89.9%) (Table 1). In line with the national data, adults represent 87.1% of total admissions and children account for 12.9%.

London accounts for 75.6% of all of England's costs for admissions associated with SCD and 80.8% of all of England's days beyond the trim point (extra days). In Table 2 we provide a summary for all PCTs in England, with the cost of admissions for SCD and the rate of admissions every 100 000 (Table 2).

There is a high variability in the number of admissions across PCTs in England. City and Hackney is the PCT with the highest number and the highest costs in 2010–11, followed by Haringey and Brent (Table 2). In these PCTs, the rates of SCD admissions are the highest in England, 347, 153 and 102 every 100 000, respectively. City and Hackney also had the highest costs associated with LOS beyond the trim point, followed by Redbridge and Westminster (Table 2). Outside London, Luton is the PCT with the highest rate of SCD admissions (39.41 every 100 000), followed by Nottingham (32.47), Manchester (20.01) and South Birmingham (20.10) (Table 2).

The regression models performed to check whether age and gender are significantly associated with SCD costs show that admissions in adults cost more than in children (Table 3). The total cost increases with age: admissions in age 9 and age 10–19 cost £1740 and £1686 ($P > 0.001$), less, respectively, compared with the age 20–29; admissions costs £65 more in the group age 30–49 compared with the age 20–29. There is no statistically significant difference in cost for patients over 50 years. Admissions were more costly for males than females, but the differences are not statistically significant. The results for London are similar to those for the rest of England.

²An NHS Primary Care Trust (PCT) was a type of NHS Trust, part of the NHS in England. PCTs were largely administrative bodies, responsible for commissioning primary, community and secondary health services from providers. Primary Care Trusts were abolished on 31 March 2013, but existed while this study was conducted.

Table 1 Costs for admissions and extra LOS associated with sickle cell anaemia with crisis (ICD-10 D57.0) in England and London, 2010–11

<i>Admissions (England)</i>	<i>Emergency</i>	<i>Elective</i>	<i>Total admissions</i>
D57.0 primary diagnosis admissions (total)	5514	563	6077
D57.0 primary diagnosis admissions (paediatrics)	1462	154	1616
D57.0 primary diagnosis admissions (adults)	4052	409	4461
HRG tariff PA47Z (to be adjusted by MFF)	£1534	£1210	
HRG tariff SA10D (to be adjusted by MFF)	£2959	£2950	
Total cost for admissions	£17 085 288	£1 712 966	£18 798 255
Long stay trim point (days) for paediatric	9	8	
Long stay trim point (days) for adults	30	30	
Total days exceeding the trim point (paediatrics)	438	7	445
Total days exceeding the trim point (adults)	624	75	699
Per day long stay payment (for days exceeding trim point) paediatrics (£)	£234	£234	
Per day long stay payment (for days exceeding trim point) adults (£)	£204	£204	
Total cost for extra LOS	£274 104	£20 592	£294 697
<i>Admissions (London)</i>	<i>Emergency</i>	<i>Elective</i>	<i>Total admissions</i>
D57.0 primary diagnosis admissions (total)	3911	452	4363
D57.0 primary diagnosis admissions (paediatrics)	877	107	984
D57.0 primary diagnosis admissions (adults)	3034	345	3379
Total cost for admission in London	£12 788 339	£1 434 799	£14 223 139
Long stay trim point (days) for paediatric	9	8	
Long stay trim point (days) for adults	30	30	
Total days exceeding the trim point (paediatrics)	246	5	251
Total days exceeding the trim point (adults)	586	75	661
Total extra cost for extra LOS in London	£218 153	£20 101	£238 254

Source: our elaboration using HES data and HRG tariffs 2010–11.

The logistic regression to examine the association between age and gender on incurring extra bed days shows that patients 10–19 years old are much more likely to incur an extra bed days (an odds ratio of 8.3 with $P > 0.001$) compared with 20–29 years old. The results for other age groups and for gender do not show statistically significant differences (Table 4).

Discussions

What is already known on this topic

Studies that assess the cost of SCD are either primarily referring to children, not appropriately updated or have been conducted in the USA.^{12–15} Studies conducted in the USA show that patients with SCD who are frequently admitted as an emergency account for the majority of health-care expenses associated with the disease.¹⁶

NICE has assessed the costs (and potential savings) attributed to managing an adult SCD acute painful episode compared with their guidelines.¹¹ The costs (and savings) may

vary significantly depending on current practice and variation in local prevalence of SCD.

The same document provides an estimate for the cost of admission for ‘sickle cell anaemia with crisis’ in children and adults according to the National Schedule of Reference costs³ in 2010/11. According to the report, the approximate total cost to the NHS of these admissions and some day-case activity was “£16.2 million, not including elective admissions (which is similar to our estimated costs £17 085 288). Moreover, the costs were not based on HES data and it is not clear if the period 2010–11 was starting in January or in April, with the financial year.

Better clinical management of SCD based on the NICE guidelines (health-care professionals training, pain management, administration of analgesia and patients monitoring) may positively impact on LOS.¹¹ However, the savings cannot be accurately estimated because the variation in current

³The national schedules of reference costs show the national average unit cost for each service for which costs were collected in 2010–11 reference costs.

Table 2 Costs for admissions and extra LOS associated with sickle cell anaemia with crisis (ICD-10 D57.0) in England PCTs 2010–11

SHA	Code	PCT	SCD adm./ 100 000 pop. ^a	Cost non-elective (A) (£)	Cost elective (B) (£)	Cost adm. (A + B) (£)	Cost extra LOS (c) (£)	Total cost (A + B + C) (£)
Q.30 North East	5D7	Newcastle	6.89	36 195.98	1250.92	37 446.90		37 446.90
Q.30 North East	5KM	Middlesbrough	6.11	18 439.87	1241.50	19 681.37	240.09	19 921.46
Q.30 North East	5E1	Stockton-on-Tees	3.11	18 151.57		18 151.57		18 151.57
Q.30 North East	5KF	Gateshead	1.54	7693.03		7693.03	241.91	7934.94
Q.30 North East	5D8	North Tyneside	0.48	3059.07		3059.07		3059.07
Q.30 North East	Total		1.45	83 539.52	2492.42	86 031.94	482.00	86 513.94
Q.31 North West	5NT	Manchester	20.01	264 153.72	4 787.14	268 940.86	240.89	269 181.75
Q.31 North West	TAP	Blackburn with Darwen	5.93	27 478.61		27 478.61		27 478.61
Q.31 North West	5NQ	Heywood, Middleton R	5.88	31 440.38		31 440.38	738.27	32 178.65
Q.31 North West	5F5	Salford	4.68	17 895.42		17 895.42		17 895.42
Q.31 North West	5NL	Liverpool	3.94	28 514.06	2515.16	31 029.22		31 029.22
Q.31 North West	5JX	Bury	2.74	10 981.34	3138.11	14 119.45		14 119.45
Q.31 North West	5NG	Central Lancashire	1.99	16 008.56		16 008.56		16 008.56
Q.31 North West	5HQ	Bolton	1.88	11 156.80		11 156.80	992.66	12 149.45
Q.31 North West	5J5	Oldham	1.79	6338.40	3111.88	9450.28		9450.28
Q.31 North West	5F7	Stockport	1.4	6502.53		6502.53		6502.53
Q.31 North West	5J2	Warrington	1.01	6184.42		6184.42		6184.42
Q.31 North West	5NR	Trafford	0.93	6620.55		6620.55		6620.55
Q.31 North West	5HG	Ashton, Leigh and Wigan	0.66	3226.53		3226.53		3226.53
Q.31 North West	5NJ	Sefton	0.37	3087.41		3087.41		3087.41
Q.31 North West	5NK	Wirral	0.33	1594.32		1594.32	1945.61	3539.93
Q.31 North West	5NF	North Lancashire T.	0.3	3028.49		3028.49		3028.49
Q.31 North West	5NH	East Lancashire Teaching	0.27	3053.18		3053.18		3053.18
Q.31 North West	Total		2.74	447 264.72	13 552.29	460 817.01	3917.43	464 734.43
Q.32 Yorkshire and H.	5N1	Leeds	7.3	153 191.20	3119.09	156 310.29	1233.30	157 543.59
Q.32 Yorkshire and H.	5NY	Bradford & Airedale	6.92	89 905.22	5590.79	95 496.01		95 496.01
Q.32 Yorkshire and H.	5N2	Kirklees	4.94	61 715.86		61 715.86		61 715.86
Q.32 Yorkshire and H.	5N4	Sheffield	4.62	56 370.18	19 511.22	75 881.40		75 881.40
Q.32 Yorkshire and H.	5NX	Hull Teaching	4.08	30 279.89		3090.16	3090.16	33 370.05
Q.32 Yorkshire and H.	5N5	Doncaster	2.02	18 349.76		18 349.76		18 349.76
Q.32 Yorkshire and H.	5J6	Calderdale	1.46	4799.20		4799.20		4799.20
Q.32 Yorkshire and H.	5H8	Rotherham	1.22	7670.81		7670.81		7670.81
Q.32 Yorkshire and H.	5N3	Wakefield District	0.88	3208.70	1265.49	4474.19		4474.19
Q.32 Yorkshire and H.	5JE	Barnsley	0.84	3043.71	3043.71	6087.42		6087.42
Q.32 Yorkshire and H.	TAN	North East Lincolnshire	0.61	1565.62		1565.62		1565.62
Q.32 Yorkshire and H.	5NV	North Yorkshire and York	0.38	9118.57		9118.57		9118.57
Q.32 Yorkshire and H.	Total		3.25	439 218.72	32 530.30	444 559.29	4323.46	476 072.48
Q.33 East Midlands	5EM	Nottingham City	32.47	283 754.21	8092.28	291 846.49		291 846.49
Q.33 East Midlands	5N7	Derby City	5.64	28 385.27	3077.73	31 463.00		31 463.00
Q.33 East Midlands	5PA	Leicestershire County R	3.16	38 297.65		38 297.65	1965.77	40 263.42
Q.33 East Midlands	5PC	Leicester City	2.71	221 819.63	3107.22	224 926.85	8108.81	233 035.66
Q.33 East Midlands	5N9	Lincolnshire Teaching	0.69	10 679.28		10 679.28	237.54	10 916.83
Q.33 East Midlands	5N6	Derbyshire County	0.57	7705.67	1255.50	8961.17		8961.17
Q.33 East Midlands	5N8	Nottinghamshire C.T	0.15	3070.27		3070.27		3070.27
Q.33 East Midlands	Total		5.25	593 711.98	15 532.73	609 244.71	10 312.12	619 556.84
Q.34 West Midlands	5M1	South Birmingham	20.1	190 790.53	4345.80	195 136.34	491.42	195 627.76
Q.34 West Midlands	5PG	Birmingham East and N.	10.73	104 037.47	1257.54	105 295.01	243.19	105 538.21
Q.34 West Midlands	5PF	Sandwell	10.39	68 804.28	1270.55	70 074.82	491.42	70 566.24

Continued

Table 2 Continued

SHA	Code	PCT	SCD adm./ 100 000 pop. ^a	Cost non-elective (A) (£)	Cost elective (B) (£)	Cost admss. (A + B) (£)	Cost extra LOS (c) (£)	Total cost (A + B + C) (£)
Q.34 West Midlands	5MX	Heart of Birmingham T	5.4	262 470.11	9225.77	271 695.88	3931.35	275 627.23
Q.34 West Midlands	5M3	Walsall Teaching	4.76	33 876.12		33 876.12		33 876.12
Q.34 West Midlands	5PE	Dudley PCT	4.29	22 268.98	1270.55	23 539.52		23 539.52
Q.34 West Midlands	5MK	Telford and Wrekin	4.29	13 759.16	3020.50	16 779.66		16 779.66
Q.34 West Midlands	5MD	Coventry Teaching	3.57	37 170.42	1270.55	38 440.97		38 440.97
Q.34 West Midlands	5PJ	Stoke on Trent	3.44	25 741.22		25 741.22		25 741.22
Q.34 West Midlands	5PL	Worcestershire	1.45	14 133.78	1257.54	15 391.32		15 391.32
Q.34 West Midlands	5QW	Solihull	1.42	7819.73		7819.73		7819.73
Q.34 West Midlands	5MV	Wolverhampton City	1.24	9316.41		9316.41		9316.41
Q.34 West Midlands	5PH	North Staffordshire	0.97	3133.15		3133.15		3133.15
Q.34 West Midlands	5M2	Shropshire County	0.69	6041.01		6041.01		6041.01
Q.34 West Midlands	5PK	South Staffordshire	0.5	6271.39		6271.39		6271.39
Q.34 West Midlands	Total		6.14	805 633.76	22 918.80	828 552.55	5157.38	833 709.94
Q.35 East of England	5GC	Luton	39.41	203 152.33	3332.28	206 484.61		206 484.61
Q.35 East of England	5QV	Hertfordshire	8.92	263 068.06	55 312.47	318 380.53	1211.88	319 592.41
Q.35 East of England	5PY	South West Essex	3.56	41 255.26	8715.70	49 970.96		49 970.96
Q.35 East of England	5PV	West Essex	3.32	26 353.86		26 353.86		26 353.86
Q.35 East of England	5P2	Bedfordshire	3.29	31 726.42	10 217.96	41 944.38	1317.60	43 261.98
Q.35 East of England	5PT	Suffolk	2.72	30 894.60	1260.39	32 154.99	1306.27	33 461.26
Q.35 East of England	5PX	Mid Essex	2.42	25 239.97		25 239.97		25 239.97
Q.35 East of England	5PP	Cambridgeshire	1.98	32 019.07	3908.06	35 927.13		35 927.13
Q.35 East of England	5PR	Great Yarmouth and W	0.44		1218.32	1218.32		1218.32
Q.35 East of England	5PW	North East Essex	0.3	3082.23		3082.23		3082.23
Q.35 East of England	5P1	South East Essex	0.3	9672.14		9672.14		9672.14
Q.35 East of England	5PQ	Norfolk	0.26	4764.59		4764.59		4764.59
Q.35 East of England	Total		4.42	671 228.53	83 965.18	755 193.71	3835.75	759 029.46
Q.36 London	5C3	City and Hackney Teach.	346.67	2 263 929.38	534 160.71	2 798 090.08	63 582.41	2 861 672.49
Q.36 London	5C9	Haringey Teaching PCT	153.38	1 203 689.40	42 598.08	1 246 287.48	5895.77	1 252 183.26
Q.36 London	5K5	Brent Teaching	102.71	816 513.50	12 279.81	828 793.31	14 464.82	843 258.13
Q.36 London	5LF	Lewisham	92.44	667 443.96	57 368.46	724 812.42	13 694.84	738 507.26
Q.36 London	5LD	Lambeth	78.54	701 592.68	67 198.84	768 791.52	10 316.11	779 107.63
Q.36 London	5K9	Croydon	77.91	745 439.97	19 714.91	765 154.88	9894.25	775 049.14
Q.36 London	5A8	Greenwich Teaching	70.42	419 118.03	26 249.18	445 367.22		445 367.22
Q.36 London	5C2	Barking and Dagenham	68.56	364 400.50	64 994.92	429 395.42	8953.02	438 348.44
Q.36 London	5C1	Enfield	68.49	592 207.17	29 422.94	621 630.11		621 630.11
Q.36 London	5NA	Redbridge	67.95	602 773.58	32 508.31	635 281.89	23 964.23	659 246.12
Q.36 London	5C5	Newham	62.55	498 489.48	51 103.68	549 593.16	10 189.18	559 782.34
Q.36 London	5LE	Southwark	60.62	461 880.46	61 307.34	523 187.80		523 187.80
Q.36 London	5NC	Waltham Forest	51.25	315 550.09	97 761.35	413 311.44	5391.55	418 702.99
Q.36 London	5H1	Hammersmith and Fulh.	48.24	238 238.83		238 238.83	4193.96	242 432.79
Q.36 London	5LG	Wandsworth	35.78	309 506.58	35 640.86	345 147.45	17 807.38	362 954.83
Q.36 London	5HX	Ealing	34.82	303 962.39	20 635.42	324 597.81	2557.92	327 155.73
Q.36 London	5LA	Kensington and Chelsea	34.38	193 403.20	34 192.20	227 595.40	532.01	228 127.41
Q.36 London	5K6	Harrow	33.19	196 649.77	3700.03	200 349.81		200 349.81
Q.36 London	5LC	Westminster	32.25	263 911.91	23 190.68	287 102.59	19 475.12	306 577.70
Q.36 London	5K8	Islington	31.8	165 137.17	50 453.93	215 591.10	14 279.81	229 870.91
Q.36 London	TAK	Bexley Care Trust	30.65	163 582.69	14 821.25	178 403.94	570.66	178 974.61
Q.36 London	5M7	Sutton and Merton	28.74	252 620.20	57 982.86	310 603.05	2548.21	313 151.26

Continued

Table 2 Continued

SHA	Code	PCT	SCD adm./ 100 000 pop. ^a	Cost non-elective (A) (£)	Cost elective (B) (£)	Cost admss. (A + B) (£)	Cost extra LOS (c) (£)	Total cost (A + B + C) (£)
Q.36 London	5A9	Barnet	23.06	230 222.55	12 512.72	242 735.27	2977.07	245 712.35
Q.36 London	5K7	Camden	18.2	132 341.63	28 954.52	161 296.15	6676.22	167 972.36
Q.36 London	5A7	Bromley	17	112 356.40	15 740.26	128 096.66	290.3	128 386.95
Q.36 London	5A5	Kingston	14.89	65 249.58	7280.35	72 529.93		72 529.93
Q.36 London	5AT	Hillingdon	14.02	89 094.73		89 094.73		89 094.73
Q.36 London	5A4	Havering	10.94	92 387.46	1437.84	93 825.30		93 825.30
Q.36 London	5C4	Tower Hamlets	8.9	69 996.30	7816.12	77 812.42		77 812.42
Q.36 London	5HY	Hounslow	5.06	36 578.71	3670.89	40 249.60		40 249.60
Q.36 London	5M6	Richmond and Twicken.	0.53	1918.17		1918.17		1918.17
Q.36 London	Total		55.28	12 570 186	1 414 698	13 984 884	238 254	1 422 313
Q.37 South East Coast	5LQ	Brighton and Hove City	11.73	91 234.47	6851.82	98 086.29		98 086.29
Q.37 South East Coast	5L3	Medway	9.93	62 161.74	1333.59	63 495.33		63 495.33
Q.37 South East Coast	5P9	West Kent	5.69	71 170.18	4035.28	75 205.46	580.59	75 786.05
Q.37 South East Coast	5P6	West Sussex	2.36	47 060.35	7816.12	54 876.47	251.55	55 128.02
Q.37 South East Coast	5P5	Surrey	2.35	88 227.56		8227.56		88 227.56
Q.37 South East Coast	5P8	Hastings and Rother	1.71	7808.30		7808.30		7808.30
Q.37 South East Coast	5P7	East Sussex Downs W.	0.6	3214.69		3214.69		3214.69
Q.37 South East Coast	5QA	Eastern and Coastal Kent	0.54	50 565.20	6932.11	57 497.31		57 497.31
Q.37 South East Coa.	Total		3.81	421 442.49	26 968.92	368 411.41	832.14	449 243.55
Q.38 South Central	5L1	Southampton City	9.71	66 712.88	6326.39	73 039.27		73 039.27
Q.38 South Central	5QF	Berkshire West	6.85	94 427.94		94 427.94	1082.14	95 510.08
Q.38 South Central	5QE	Oxfordshire	4.73	67 009.44	8047.49	75 056.93		75 056.93
Q.38 South Central	5QD	Buckinghamshire	3.72	49 435.48	7051.63	56 487.11		56 487.11
Q.38 South Central	5FE	Portsmouth City Teaching	3.27	20 859.09		20 859.09		20 859.09
Q.38 South Central	5QG	Berkshire East	1.98	19 666.46	3196.18	22 862.64		22 862.64
Q.38 South Central	5QT	Isle of Wight NHS	0.7	3075.99		3075.99		3075.99
Q.38 South Central	5QC	Hampshire	0.47	14 625.91		14 625.91	2800.32	17 426.23
Q.38 South Central	5CQ	Milton Keynes	0.41	3279.96		3279.96		3279.96
Q.38 South Central	Total		3.11	339 093.15	24 621.69	363 714.84	3882.46	367 597.30
Q.39 South West	5QJ	Bristol	17.02	180 505.09	33 917.74	214 422.84	8768.54	223 191.38
Q.39 South West	5FL	Bath and North East Som.	7.36	36 330.77	9567.15	45 897.92		45 897.92
Q.39 South West	5F1	Plymouth Teaching	4.44	20 585.88	2608.15	23 194.02	4306.24	27 500.27
Q.39 South West	5QH	Gloucestershire	2.85	38 148.60	3189.05	41 337.65	10 624.99	51 962.64
Q.39 South West	5A3	South Gloucestershire	2.34	15 945.25	1304.07	17 249.32		17 249.32
Q.39 South West	5QQ	Devon	0.53	12 038.89		12 038.89		12 038.89
Q.39 South West	5K3	Swindon	0.48	1594.40		1594.40		1594.40
Q.39 South West	5M8	North Somerset	0.47	3189.05		3189.05		3189.05
Q.39 South West	5QK	Wiltshire	0.22	1589.00		1589.00		1589.00
Q.39 South West	Total		2.52	309 926.93	50 586.16	360 513.09	23 699.77	384 212.87

^aThe rate is given by the number of admissions in emergency and elective over the number of population in the PCT for 2011.

(2) Source of population at PCT level: NHS 2011. <https://www.gov.uk/government/publications/exposition-book-2011-2012>.

practice and lack of baseline data in several areas make it difficult to assess the cost impact of implementing the guideline.¹¹

One study assessing the views of patients with SCD showed that patients and carers often bypass their general practitioner for acute problems and attend directly at accident

and emergency (A&E) suggesting better primary care interventions could reduce emergency admissions.^{20,21} One study looked at 145 general practices in the East Midlands and showed that as the proportion of patients able to consult their primary care physician increased, emergency admission rates decreased.²²

Table 3 Regression of total costs of admission on age and sex at national level and London level only

	<i>National</i>			<i>London</i>		
	<i>Geometric mean cost (£)</i>	<i>Comparison (as % of reference group costs)</i>	<i>P value</i>	<i>Geometric mean cost (£)</i>	<i>Comparison (as % of reference group costs)</i>	<i>P value</i>
Age						
<1	1758	50.26	<0.001	1828	50.27	<0.001
1–9	1732	49.53	<0.001	1804	49.61	<0.001
10–19	1812	51.83	<0.001	1881	51.71	<0.001
20–29	3499	Ref.	Ref.	3636	Ref.	Ref.
30–39	3564	101.87	<0.001	3640	100.11	<0.001
40–49	3565	101.94	<0.001	3638	100.03	<0.001
50+	3535	101.10	0.194	3622	99.55	0.605
Sex						
Men	3021	Ref.	Ref.	3182	Ref.	Ref.
Women	2854	99.59	0.195	3052	100.36	0.278

Source: our elaboration.

Table 4 Logistic regression of the association between age and gender on incurring extra bed days at national level and London level

	<i>National</i>			<i>London</i>		
	<i>% Staying extra days</i>	<i>Odds ratio</i>	<i>P value</i>	<i>% Staying extra days</i>	<i>Odds ratio</i>	<i>P value</i>
Age						
<1	0	—	—	0	—	—
1–9	1.31	1.26	0.576	0.82	0.54	0.316
10–19	8.08	8.38	<0.001	8.1	5.65	<0.001
20–29	1.04	Ref.	Ref.	1.47	Ref.	Ref.
30–39	1.11	1.07	0.834	1.26	0.84	0.616
40–49	0.93	0.90	0.8	0.85	0.56	0.24
50+	1.27	1.22	0.745	1.7	1.09	0.895
Sex						
Men	1.98	Ref.	Ref.	1.78	Ref.	Ref.
Women	2.47	1.00	0.985	2.73	1.35	0.162

Source: our elaboration.

A study conducted in London has shown that overnight admissions decreased as a result of better pain management, use of analgesia, transfusions, community support and improved education at home.^{23–26} Moving treatments from hospital to community or ambulatory setting could contribute to avoid patients being admitted to hospital to be treated, with savings in terms of resources. Elective transfusions for example are an essential treatment in SCD and can now be provided in ambulatory setting instead of admitting the patients in hospital to receive the treatment.^{23–25} There is

evidence that admissions are increasing as an effect of readmissions of patients with SCD and that effective discharge planning with a link between hospitals, primary and community care might help prevent readmissions.^{22,27}

What this study adds

This study is the first in England to use HES data to assess the cost of admissions for SCD, applying the HRGs tariffs to those admissions presenting SCD as primary diagnosis only.

We adopted a conservative approach to ensure the costs are linked to SCD with crisis to provide an accurate estimate of the costs for hospitalization linked to SCD.

According to a report published by the Audit Commission, clinical coding errors continue to affect Healthcare Resource Coding (HRG) assignment in England.²⁰ Nevertheless, this study relies on good quality coding (in 2009 primary diagnoses codes were validated as accurate the 87% of the time).²⁰

Limitations of this study

The analysis underestimates the real costs of admissions associated with SCD, as it takes into account only the cases of SCD with crisis as primary diagnosis, but there are many episodes in which SCD appears as a secondary, tertiary or further underlying diagnosis. Even if the main cause of admission is not an SCD crisis, the disease's underlying characteristics may have predisposed the patient to be admitted to hospital or increased the risk of comorbidity or complications.

In our study, the admissions having SCD with crisis as a primary diagnosis represent 92.5% of all the admissions for SCD with crisis (including also secondary, third, fourth and fifth diagnosis) and so almost 7.5% of admissions are not included here. Assuming we can apply the same HRG tariffs for the excluded admissions (those after the primary diagnosis), the total cost for admissions linked to SCD with crisis will increase to £20 376 822, a difference of £1 578 567.

The other 13 589 admissions are linked to SCD without crisis (ICD-10 codes D57.1–8) as primary or further diagnosis, but the cost for these admissions has not been taken into account in this study. It is very difficult to estimate the cost of these admissions without knowing the HRG tariff applied when the patient is discharged. However, the primary diagnosis of these admissions is mainly for SCD without complications, anaemia, respiratory infections and asthma. Considered that 25% of these admissions are in emergency, the cost of admissions for SCD is even higher. Further research can help gain a more detailed estimate of the costs borne by the NHS and also for unaccounted expenditures such as those spent by patients and their families to treat and manage this condition at home or for other sectors, providing a closer estimate of the entire societal cost.

Finally, we used the episode as unit of analysis instead of spells or continuous inpatient stay as we excluded from the analysis patients who have been transferred. We are aware that this might have an impact on excess payments for extra LOS and might underestimate or overestimate the cost for some providers. However, it would have been difficult to attribute in a reliable way the cost of extra LOS to each Trust. We therefore decided to adopt a conservative approach.

Conclusions

According to our study, in 2010–11, the cost of emergency admissions in England due to SCD was £17.1 million which compares with the £16.2 million estimated in the NICE report.¹¹ In this study we did not take into account all of the day-case activities or admissions that have SCD with crisis as a secondary, tertiary or further diagnosis which explains the cost discrepancy between our study and the NICE report.

London accounts for 75.7% of all of England's costs for admissions associated with SCD.

The results show that emergency admissions represent 90.9% of the entire admissions cost. Shifting the balance of care from A&E to primary care is possible²⁸ and better clinical management of patients with SCD may positively impact emergency admissions.

Management of SCD needs to take a more modern approach.²⁹ Proper management is essential in reducing crises and episodes that require hospital admissions. In order to provide effective and safe care, primary care physicians, community nurse specialists and community paediatricians should have a better awareness of warning symptoms and signs which require assessment in hospital.³⁰

This study shows that patients 10–19 years old are much more likely to incur extra bed days and that the cost of an extra day for them is higher than for adults. Therefore, a better management of SCD in children to prevent hospital admissions or at least reduce the LOS could be cost saving.

A recent study emphasizes the need for 'a preventative and comprehensive model of care in addition to care management'.³¹ Adults with SCD need coordinated care led by a primary care physician in coordination with a provider experienced in SCD. Patients and parents of children affected by SCD should also have a good awareness of symptoms and when and how to seek help.³²

Recently, SCD has achieved specialized commissioning status but the current specifications focus mainly on provision of acute care.³³ Links between primary care and community care could be improved through proper discharge planning, multi-disciplinary teams and better communication with primary care.³⁴ The 'community hub model' was first introduced by the Sickle Cell Society's Comprehensive Care Project and in contrast shows that the patients and patient groups prioritize social determinants of health. The model advocates a patient-centred approach where an integrated sickle care pathway encompassing all public health-care needs are addressed specifically mental, social (employment and schooling), acute and primary. This in turn may increase user satisfaction and could assist in reducing unnecessary admissions to hospitals.^{34–36}

Further research which better understands hospital utilization of patients with SCD could also explain allocation of health-care costs due to SCD. HRG tariffs could also be revisited to link disease severity with scalable HRGs which could give a clearer picture as to whom the majority of the SCD patients consuming resources are.

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Conflict of interest

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References

- Weatherall D, Hofman K, Rodgers G *et al.* A case for developing North-South partnerships for research in sickle cell disease. *Blood* 2005;105(3):921–3.
- Analysis of frequent hospital users, by PCT 2003/4 [Online] [database on the Internet] 2006. <http://www.lho.org.uk/viewResource.aspx?id=10286> (May 2013, date last accessed).
- Aisiku IP, Smith WR, McClish DK *et al.* Comparisons of high versus low emergency department utilizers in sickle cell disease. *Ann Emerg Med* 2009;53(5):587–93.
- Lucas SB, Mason DG, Mason M *et al.* A sickle crisis? A report of the National Confidential Enquiry into Patient Outcome and Death, 2008. <http://www.ncepod.org.uk/2008report1/Downloads/>.
- Aljuburi G, Lavery AA, Green SA *et al.* Trends in hospital admissions for sickle cell disease in England, 2001/02–2009/10. *J Public Health* 2012;34(4):570–6.
- NHS sickle cell and thalassaemia screening programmes. Sickle Cell Disease in Childhood Standards and Guidelines for Clinical Care, 2nd edn. London: Sickle Cell Society, 2010.
- Department of Health. National service framework for long-term conditions. London: Department of Health, 2005.
- Bilenker JH, Weller WE, Shaffer TJ *et al.* The costs of children with sickle cell anemia: preparing for managed care. *J Pediatr Hematol Oncol* 1998;20(6):528–33.
- Davis H, Moore RM Jr, Gergen PJ. Cost of hospitalizations associated with sickle cell disease in the United States. *Public Health Rep* 1997;112(1):40–3.
- Epstein K, Yuen E, Riggio JM *et al.* Utilization of the office, hospital and emergency department for adult sickle cell patients: a five-year study. *J Natl Med Assoc* 2006;98(7):1109–13.
- NICE. National costing report: sickle cell acute painful episode. Report No.: 143. Manchester: NICE, 2012.
- Amendah DD, Mvundura M, Kavanagh PL *et al.* Sickle cell disease-related pediatric medical expenditures in the U.S. *Am J Prev Med* 2010;38(4 Suppl):S550–6.
- Ballas SK. The cost of health care for patients with sickle cell disease. *Am J Hematol* 2009;84(6):320–2.
- Kauf TL, Coates TD, Huazhi L *et al.* The cost of health care for children and adults with sickle cell disease. *Am J Hematol* 2009;84(6):323–7.
- Nietert PJ, Abboud MR, Zoller JS *et al.* Costs, charges, and reimbursements for persons with sickle cell disease. *J Pediatr Hematol/Oncol* 1999;21(5):389–96.
- Carroll CP, Haywood C Jr, Fagan P *et al.* The course and correlates of high hospital utilization in sickle cell disease: Evidence from a large, urban Medicaid managed care organization. *Am J Hematol* 2009;84(10):666–70.
- Street A, Dawson D. Costing hospital activity: the experience with healthcare resource groups in England. *Eur J Health Econ* 2002;3(1):3–9.
- Geue C, Lewsey J, Lorgelly P *et al.* Spoilt for choice: implications of using alternative methods of costing hospital episode statistics. *Health Econ* 2012;21(10):1201–16.
- Department of Health Payment by Results Team (2012). Step-by-step guide: calculating the 2012–13 National Tariff. Leeds: Department of Health.
- Audit Commission. PbR data assurance framework 2008/09. Key messages from Year 2 of the national clinical coding audit programme. London: Audit Commission, 2009.
- Aljuburi G, Okoye O, Majeed A *et al.* Views of patients about sickle cell disease management in primary care: a questionnaire-based pilot study. *JRSM Short Rep* 2012;3(11):78.
- Aljuburi G, Phekoo KJ, Okoye NO *et al.* Patients' views on improving sickle cell disease management in primary care: focus group discussion. *JRSM Short Rep* 2012;3(12):84.
- Day TG, Thein SW, Drasar E *et al.* Changing pattern of hospital admissions of children with sickle cell disease over the last 50 years. *J Pediatr Hematol Oncol* 2011;33(7):491–5.
- Halsey C, Roberts IA. The role of hydroxyurea in sickle cell disease. *Br J Hematol* 2003;120:177–86.
- Jacobson SJ, Kopecky EA, Joshi P *et al.* Randomised trial of oral morphine for painful episodes of sickle-cell disease in children. *Lancet* 1997;350:1358–61.
- Mahat G, Scoloveno MA, Donnelly CB. Written educational materials for families of chronically ill children. *J Am Acad Nurse Pract* 2007;19:471–6.

- 27 Ballas SK, Lusardi M. Hospital readmission for adult acute sickle cell painful episodes: frequency, etiology, and prognostic significance. *Am J Hematol* 2005;79:17–25.
- 28 Bankart MJ, Baker R, Rashid A *et al.* Characteristics of general practices associated with emergency admission rates to hospital: a cross-sectional study. *Emerg Med J* 2011;28(7):558–63.
- 29 Roberts E, Mays N. Can primary care and community-based models of emergency care substitute for the hospital accident and emergency (A & E) department? *Health Policy* 1998;44(3):191–214.
- 30 Telfer PT. Management of sickle cell disease: out-patient and community aspect. *Paediatr Child Health* 2011;21(8):357–62.
- 31 Kanter J, Kruse-Jarres R. Management of sickle cell disease from childhood through adulthood. *Blood Rev* 2013;27:279–87.
- 32 Inati A, Chabtni L, Mounayar M *et al.* Current understanding in the management of sickle cell disease. *Hemoglobin* 2009;33(Suppl 1): S107–15.
- 33 Telfer P. Management of sickle cell disease: acute episodes in the community and in hospital. *Paediatr Child Health* 2011;21(8): 363–8.
- 34 SSNDS. Definition No. 38: Specialised haemoglobinopathy services. *Specialised Services National Definitions Set*. 3rd edn. London: Department of Health, 2010.
- 35 Matthew L. Enhancing communication between GPs and hospital consultants. *Health Serv J* 2012. http://www.hsj.co.uk/resource-centre/best-practice/it-e-health-and-e-medicine-resources/enhancing-communication-between-gps-and-hospital-consultants/5044381.article#.U0Ql8_kdV8E.
- 36 Cooke M, Nkohkwo A, Bennett L. Report of an Independent Evaluation by Health and Wellbeing Impact Assessment of the Society's Regional Support Care Advocacy Project (2004–07). The sickle cell society, London, 2007. http://www.sicklecellsociety.org/app/webroot/files/files/RCP_HIAevaluatJuneD.pdf.