Sickle Cell Disease: Patient Care, Management and Reduction of Hospital Admission

Lynne Jalalian and Ogechi Anokwuru

Department of Science and Technology
Middlesex University of London (2017)

DOI: 10.29322/IJSRP.10.05.2020.p101106
http://dx.doi.org/10.29322/IJSRP.10.05.2020.p101106

Abstract- Background: Sickle cell disease (SCD) is a common genetic blood disorder affecting primarily the Black African/Caribbean and ethnic minority population. In Britain, there has been a sharp rise in individuals being diagnosed with sickle cell as well as a rapid increase in the number of sickle cell patients being admitted into hospital regularly. Pain management is effective in reducing hospital admission and delays in being discharged.

The aims of this research were to define sickle cell disease, the sickle cell crisis and the preventive measures in place to reduce the frequency of hospital admission.

Methods: Literature searches were conducted between December 2016 and July 2017. This comprised of searching of utilising searches electronically e.g. websites, online publications and other literature search engines. A quantitative survey was handed out to patients to fill out at their discretion and to sickle cell support groups. In total, 65 participants were involved in this study, after which the results were collected and analysed using statistical analysis through Excel. The null hypothesis was preventive measures do not improve sickle cell patients’ quality of life, and did not to the rate of hospital admission. The alternative hypothesis was that preventive measures helped reduce hospital admission. The level of significance was set to 0.05.

Results: The results showed that many felt that they were not getting the care they require. Furthermore, many patients believed that a care plan would benefit them and reduce the rate of hospital admission. Others suggested that community care should be introduced to curb the frequent hospital admission.

Conclusions: This research showed that more needs to be done regarding providing healthcare plans for sickle cell patients to reduce hospital admission and reduce costs for patients who stay in hospital for extended periods of time. In addition to this, further investigations based on the achieved findings should demonstrate an inclusion of more preventive therapies e.g. Bone marrow transplantation (BMT), use of traditional medicine and utilising community care.

Index Terms: Sickle Cell Anaemia, Sickle Cell Disease, Hospital Admission, Therapy, Pain Management

I. INTRODUCTION

Sickle cell disease (SCD) is one of the top leading diseases affecting the black and ethnic minority population. In the UK, sickle cell disease is found amongst the African and Caribbean population (NHS, 2016). Sickle cell disease belongs to a set of genetic blood disorders whereby erythrocytes (red blood cells) are malformed (AlJubri et al, 2012).

In healthy individuals, normal red blood cells are biconcave shape that bypass blood vessels to transport oxygen within the body (CDC, 2016). In affected individuals with SCD, the erythrocytes become “crescent shaped”, ‘hard and sticky’, these cells die and there’s a lack of supply of erythrocytes (CDC, 2016). These malformed erythrocytes also become trapped in the blood vessels, thereby restricting blood flow causing anaemia (CDC, 2016).

This often results in painful crises, stroke, cardiac problems, acute chest syndrome to name a few (CDC, 2016) or in some cases fatality if not treated on time. Other complications include short and long-term damage to organs, and musculoskeletal complications (Soe et al, 2017). These symptoms and problems often impair quality of life and result in constant admission into hospital (AlJubri et al, 2012).

According to the WHO (2011), approximately, 5% of the world’s population carry the trait, as well as 300,000 babies born each year with severe haemoglobin disorders. Due to the heavy health burden, of sickle cell and other haemoglobin disorders, this can be reduced through management and prevention programmes (WHO, 2011).

Evidence has shown that SCD is rapidly growing to be one of the top common genetic disorders within England (AlJubri et al, 2012). It is roughly estimated that 12,500 persons live with SCD in the UK and roughly 240,000 persons have the trait (AlJubri et al, 2012). In addition to this, AlJubri et al, (2012) had suggested that information on SCD hospital admission is necessary in caring and undergoing better management for patients with sickle cell disease. In can also be stated that AlJubri et al (2012), had covered the rates of admission for nearly 10 years (2001/02-2009/10). Compared to 2001, hospital admission was reduced to 30% (BBC, 2001).

On the other hand, patients’ numbers still doubled since 1998 according to Guy’s and St Thomas (BBC, 2001). Furthermore, ‘unnecessary hospital admission has been reported to result in having a negative impact on emotional and physical health’ (BBC, 2001). This report also highlighted how to manage sickle cell crises better since it accounts for 90% of hospital admission in London (BBC, 2001).

Moreover, recent studies have highlighted the need to reduce the hospital admission rate (Latnovic and Streetly 2013; Pizzo, 2014; NICE, 2011). This can also be compared to rate of admission from older studies of sickle cell patients in Britain.
where both recent and older publications (Witnicki et al, 2015; Murtaza et al 1981; Thomas et al 1998; Pizzo, 2014; Thompson et al 2014) suggested that ambulatory care and community care would reduce the cost and hospital admission and duration.

The aims of this research were to define sickle cell disease, the sickle cell crisis and the preventive measures in place to reduce the frequency of hospital admission in London and across London boroughs. This research was conducted to evaluate selected measure currently in place. The selected research questions posed: are these selected preventive measures helping sickle cell patients improve their quality of life and are these selected preventive measures assist in reducing hospital admission, and is better management required to reduce hospital admission.

This research used quantitative surveys in a sample population of 65 participants affected with sickle cell. The null hypothesis was that preventive treatment did not make a difference to those with sickle cell, and did not reduce or increase hospital admission rate. The alternative hypothesis was, that preventive treatment reduced hospital admission.

**Sickle Cell Disease explained**

Sickle cell disease has been reported to be the second most frequent type of sickle cell disease. This occurs when an individual inherits the Hb C gene from one parent and the Hb S (Herman and Chaudhury, 2010). Individuals with Hb SC tend to have similar symptoms to people with Hb SS; on the other hand, the level of anaemia they experience has been reported to be less severe (NIHNLBI, 2002; Herman and Chaudhury, 2010). Another type of sickle cell disease is SB+ (Beta,β), thalassaemia, which primarily affects beta glob in gene expression and production (NIHNLBI, 2002; Herman and Chaudhury, 2010).

In this type, individuals normally have red blood cells seem much smaller due to less beta protein produced (NIHNLBI, 2002; Herman and Chaudhury, 2010). When inherited and combined with the Hb S gene, it codes for haemoglobin S beta thalassaemia. It has been reported that symptoms with this type are normally not as severe as the other types (CDC, 2015; WHO 2014; Herman and Chaudhury, 2010).

Haemoglobin SB 0 (β beta-zero) thalassaemia, is the fourth type of sickle cell disease, like SB+, the beta globin gene is dysfunctional. On the other hand, beta zero thalassaemia shares similarities with Hb SS anaemia (Herman and Chaudhury, 2010). Although, beta zero thalassaemia has in some cases proven to be more severe, it has been reported to be found to have a poorer prognosis (Herman and Chaudhury, 2010).

There are other types of sickle cell disease: haemoglobin SD, SE and SO, which have been reported to be very rare, the severity often varies from low severity to far severity (Herman and Chaudhury, 2010). Lastly, there are those who are rarely affected by sickle cell disease, individuals who inherit a mutated gene only known as the sickle cell trait (AS) (CDC, 2015; Herman and Chaudhury, 2010). This arises when one parent is known to have the trait (NHS, 2016).

**Pathophysiology of Sickle Cell Disease explained**

As a result of sickle cell disease, there are many complications that arise from it. Its pathophysiology and complications will be explained (AlJubri et al, 2012).Due to its sickle ‘crescent’ shape, HbS causes extensive damage to the red blood cell membrane, as a result, excessive calcium ions enters the cell (Herman and Chadhury, 2010; CDC, 2015; Rogers, 2017; Bridges, 2000).This results in an influx of calcium ions often results in membrane proteins being cross linked, activating specific channel proteins for potassium ions and water to leave the cell (Rogers, 2017). Therefore, red blood cells become dehydrated e.g. “sickling”.This further causes a complication known as vasoocclusive crisis (Akar and Akile, 2008). As most of the sickling red blood cells are blocking and reducing blood flow to crucial organs, necrosis, excruciating pain, ischaemia occur (Herman and Chadhury, 2010).

Repetitive sickle episodes often cause bone infarction and bone marrow degeneration over long periods of time (Manwani et al., 2014). The bones affected by this are: long bones, on the other hand, the pain can extend to other bones such as the sternum, ribs, vertebrae and the cranium (Herman and Chaudhury, 2010; Okpuzor et al, 2008). Pulmonary fat embolisms also occur often in patients with sickle cell disease (Bridges, 2000). This often fatal or life threatening (Herman and Chadhury, 2010).

Other complications of sickle cell disease include haemolysis, where sickle cell patients whose red blood cells are very weak are subject to intravascular haemolysis. Furthermore, extravascular haemolysis is most damaging to sickle patients, as this mechanism often leads to a reduced red blood cell survival time (Bridges, 2000; Herman and Chadhury, 2010).

This process happens when these inflexible red blood cells become stuck within the spleen and undergo phagocytosis, by the reticuloendothelial system (Bridges, 2000). Within sickle patients, their bone marrow struggles to counters this by producing a higher rate of red blood cells, but is counterproductive as their system cannot keep up or match the rate of red blood cell destruction (CDC, 2015; Manwani et al., 2014).

For instance, a normal red blood cell’s life span is about 120 days compared to a sickle red blood cell’s life span is up to 20 days (Herman and Chaudhury, 2010; Kirenska et al., 2014). In cases of liver damage, increased haemolytic production often results in cholelithiasis (excessive bilirubin production) (Herman and Chaudhury 2010; Kirenska et al., 2014).

Hyposplenism and bacterial infections are common complications of sickle cell disease. Hyposplenism occurs as a result of compromised immune response against infections and its infectious agents (Kirenska et al., 2014). The spleen is reported to be involved in making and maturing B white blood cells, and opsonins for fighting infections (Kirkineska et al., 2014). Due to the nature of SC, hyposplenism undergoes this following process:

Splenic sequestration of sickled cells causes blockage in the spleen (splenomeagly), as a consequence, this reduces immune activity (Herman and Chadhury, 2014; WHO, 2015). It is reported that the spleen plays an important role in destroying bacteria (encapsulated) via phagocytosis (Herman and Chaudhury, 2014). However, people with sickle cell disease are subject to getting bacteraemia e.g. *Streptococcus pneumonia*, *Haemophilus Influenzae*, *Neisseria meningitides* (Herman and Chaudhury, 2014). If left untreated, can cause sepsis and can be life threatening and fatal (Bridges, 2000).

**Bacterial infections associated with Sick Cell**
Mycoplasma pneumoniae, Chlamydia pneumoniae and Legionella are some of the bacteria associated with bacterial infection found in sickle cell sufferers (Herman and Chaudhury, 2010; Kirenska et al., 2014). The bacteria previously mentioned are known for pulmonary infections (Heran and Chaudhury, 2010). On the other hand, S. Pnuemoniae and H. influenza type b are rare (Wright et al., 2004). Other diseases caused as part of SCD are osteomyelitis and septic arthritis; which often affects the smaller portion of patients with SCD (Wright et al., 2004; Nolan et al., 2008). This happens due to extensive damage done to the bone and very poor splenic function, so pathogens that are gram negative (Salmonella and S. Aureus) seize opportunity to invade (Shiel, 2017; Herman and Chadhury, 2010). Due to poor splenic function, this causes infarction (functional asplenia) (AlJubri et al., 2012).

As for the neurologic complications, it shares a relationship with the vasocclusive crisis e.g. strokes and silent strokes (Herman and Chadhury, 2010; CDC, 2015).

**Acute Chest Syndrome**

Continuous episodes also cause acute chest syndrome (ACS), Hb that is effluxed from haemolyzed cells has a higher affinity and binds easily and causes a reduction of nitrous oxide (Herman and Chadhury, 2010; Rogers, 2017). This allows smooth muscle cell contraction and causes an increased rate of platelet aggregation. Often at times, occlusion is frequent which causes most patients to be very hypoxic (Okpuzor et al., 2008; AlJubri et al., 2012).

ACS, occurs when there is pyrexia, hypoxia, and chest pain (mild to severe) which varies from patient to patient (Herman and Chadhury, 2010). Other contributing factors to ACS include, infarction or pulmonary embolism. In addition to this, there are two cardiac complications that arise, chamber enlargement (an increased output due to life-long anaemia, including decreased oxygen desaturations) (Bridges, 2000; Herman and Chadhury, 2010). The second cardiac problem is myocardial infarction due to vasocclusive crisis occurring in the arteries (Herman and Chaudhury, 2010; WHO, 2016).

**Hospital Admission**

Hospital admission England in particular, 2005-2006 admission rates for sickle cell disease rapidly increased along with age (AlJubri et al., 2012). For those aged between 0-18 years old, and 18 years old plus had the highest rate of admission, approximately 38 per 100,000 (AlJubri et al., 2012). Furthermore, London especially has the been recorded to have the highest level of blood disorder patients within the UK for hospital admission (AlJubri et al., 2012). Roughly 75% of sickle cell disease admission are found specifically in London, the most common reason for admission (AlJubri et al., 2012). During 2005-2006, 13,000 episodes had occurred compared to 3,200 (2000-2001) and 4,600 (2005/2006) (Latinovic et al., 2013; AlJubri et al., 2012).

It is also reported that hospital admission rates vary across London with 50% coming from the most deprived areas (AlJubri et al., 2012). For 25-34 year olds, the admission rate tends to peak and decrease rapidly after 45 years old (AlJubri et al., 2012).

**Current management of SCD**

Management for sickle cell disease varies depending on the type of sickle cell an individual may have. However, the management of the disease has rapidly increased the past few decades resulting in better life outcome and longer life (AlJubri et al., 2012). As with any disease, there are guidelines put in place by clinicians for patients to abide by, whether it is acute related pain resulting from a crisis or from chronic sickle cell disease (Yawn et al., 2014). This is in support of Department of Health, who endeavour that all patients receive upmost care and treatment. The current UK guidelines (NICE, 2011) regarding long term management of sickle cell consist of prevention and intervention (Table 1).
Pain management

Opioids are the most common form of pain relief for acute crisis, especially when the crisis is full blown (Bridges, 2000). It can be very excruciating and unbearable for patients (Bridges, 2000). Since the pain can last for a few minutes to a couple of days (Bridges, 2000; Sanders, 1992). This often changes over lifetime from childhood to adulthood (Bridges, 2000; Platt, 1994). Patients often take opioids via oral consumption and the remaining with fluids to enable the episode to phase out especially in a home setting (Bridges, 2000).

Management of oral analgesics are carried out well by sickle cell patients, as they would normally have a supply at home should a crisis should occur (Wright et al., 1992). This often changes over lifetime from childhood to adulthood (Bridges, 2000; Platt, 1994). Patients often take opioids via oral consumption and the remaining with fluids to enable the episode to phase out especially in a home setting (Bridges, 2000).

<table>
<thead>
<tr>
<th>Table 1 shows clinical presentation of SCD, with prevention, intervention and the reason to manage sickle cell (NICE, 2011).</th>
</tr>
</thead>
<tbody>
<tr>
<td><strong>Clinical Presentation</strong></td>
</tr>
<tr>
<td>Pregnancy</td>
</tr>
<tr>
<td></td>
</tr>
<tr>
<td>Bacterial infection</td>
</tr>
<tr>
<td>Anaemia</td>
</tr>
<tr>
<td></td>
</tr>
<tr>
<td></td>
</tr>
<tr>
<td>Acute Chest syndrome</td>
</tr>
<tr>
<td>Renal function</td>
</tr>
<tr>
<td></td>
</tr>
<tr>
<td>Painful crises</td>
</tr>
<tr>
<td>Priapism</td>
</tr>
<tr>
<td>Gall bladder</td>
</tr>
<tr>
<td>Necrosis (hip)</td>
</tr>
<tr>
<td>Brain injury</td>
</tr>
<tr>
<td>Chronic leg ulcers</td>
</tr>
</tbody>
</table>

On the other hand, pain often varies at different sites of the body and frequently happens in the extremities: the thorax, abdomen and back (Ballas and Delengowski, 1993). Due to the nature of painkillers being opioids (Bridges, 2000), it can be controlled. But often leaves patients drowsy, as well as combating the fatigue experienced from the lack of rest from dealing with the crisis at home (Bridges, 2000; Pegelow, 1992).

Even with the drowsiness patients’ experience, there is a misconception that patients do not experience pain while sleeping (Bridges, 2000). Often at times, pain interrupts their sleeping pattern even when they have had analgesics (Robieux, et al., 1992). On the other hand, the half-life of meripidine (morphine) can cause complication for patients (Bridges, 2000; Tobias, 1993). For instance, the half-life of the drug in circulation is roughly 4 hours. This is because the liver converts it to another derivative (analgesic) but still toxic (Bridges, 2000). At times, a grand mal seizure can occur when large doses of meperidine are administered (Sanders, et al., 1992). However, according to the American Pain Association (APA), meperidine is no longer used for long term treatment (Bridges, 2000).

According to Tobias (1993), epidural analgesia has been for pain control for some sickle cell patients, which has proven to be effective in the chest where patients receive most pain. However, other patients require systemic analgesics but with a lower dose (Bridges, 2000). Furthermore, some of these patients may have a phobia of needles (psychological) entering their back (Bridges, 2000).
NSAIDs, are normally taken during acute sickle cell pain (Sanders et al. 1992). However, while taking NSAIDs, it can disrupt kidney function which further impacts renal injury due to sickle cell disease being present already (Bridges, 2000). For some cases, healthca re professiona ls tend to avoid giving patients NSAIDs (Bridges, 2000).

Blood transfusion therapy, is one of the most common preventive therapies in managing any sickle cell crisis (Thompson et al., 2014). With blood transfusions it helps where tissues are very badly perfused due to the vaso-occlusion crisis (Clark et al., 1980). On the other hand, the transfused red blood cells will not have any effect or improve blood flow where microcirculation is occluded due to the sickle red blood cells (Bridges, 2000). In essence, simple transfusion is not effective for long term management (Bridges, 2000). In addition to this, exchange transfusion has been reported to be better relieving pain (Davies and Brozovic, 1989).

Hydroxurea a newer drug, tends to inhibit ribonucleotide reductase, which blocks new DNA being synthesised and cell division, and increases fatal haemoglobin by creating erythroid cells (Platt et al., 1994). This is efficient as fatal Hb stops sickling, it also increases the level of the red cell volume and reduce cell density (Bridges, 2000; Platt et al., 1994).

For sickle cell patients, vitamin supplementation is necessary as they are regularly anaemic, and need to take daily folic acid (Harvard, 2000). However, their folate is often consumed rapidly so the intake is at times less and imbalanced (Harvard, 2000).

**Psychosocial support for individuals with Sickle Cell**

Psychiatric support/support groups are necessary in helping managing sickle cell disease as the psychosocial aspect is already fairly complex (Bridges, 2000; Thomspon et al., 2014). Bouts of loneliness, low self-esteem, anger, even isolation are experienced in sickle cell patients (Bridges, 2000; Whitten and Fischoff, 1974) which often, results in them unable to manage pain efficiently, support groups as well as counselling have often helped with better patient care and understanding in how to cope with pain (Gil et al., 1994).

II. METHODS

The research design used a quantitative method. The survey was designed to accomplish three main things universally that will assist in achieving the aims of this research by collecting information from participants based on their experience with hospital admission. This provided an idea what needs to be done to improve current services in place

To identify what areas need to be changed to have a positive impact on reducing hospital admission. This provided sample data to make comparisons against current published data

To establish whether there is a relationship between pain management, taking preventive treatment in relation to hospital admission.

As this was carried out, each participant was given a participant information form and a consent form to complete. Sickle Cell Society agreed to partake in this research as well as Ealing Hospital, Fulham Hammersmith Sickel Cell and Thalaessemia support group. SickleKan and Sickle Cell Cause also agreed to take part in this research.

The questionnaire initially was piloted with the head of Sicklekane, who suggested some adjustments to the questions initially asked (see Appendix I). Therefore, adjustments were made to the initial piloted questions; which needed to be answered. Further research on the type of questions was necessary as data obtained from these questions need to be accurate as possible. In addition to this, some of the questions will be influenced by previous studies (AlJubri et al 2012; Maxwell and Bevan; 1999; Murtaza et al; 1981; Latnovic and Streetly, 2013; Brozovic et al 1987; Wititiwicki et al, 2015).

This allowed clarity and the aims to be properly defined and the hypotheses to be correctly justified. Primary data collection was obtained through surveys given to sickle cell patients addressing questions relating to pain management, lifestyle and frequency of hospital admission will be explained and critically analyzed. This was achieved through online surveys, online support groups, and individual support groups within London.

When this was achieved, using their responses, data collected underwent statistical analysis via descriptive measures. Secondary data was obtained with permission from the Sickle Cell Society and other data sets from published articles, which was used to discuss in the research.

This descriptive research investigated adults in London with sickle cell disease who attended local support groups in London, who attended for different reasons during April 2017 until the end of August 2017. A total of 65 patients were recruited; with their ages ranging from 18 to 55 years old.

Therefore, this research aimed to determine whether the null hypothesis was accepted/rejected or whether or not the alternative hypothesis was accepted or rejected.

Ethic approval was sought from the Ethics governing body, NHS ethics approval was not needed from sickle cell support groups as these filled out anonymously without asking for hospital details.

**Setting**

The research took place in London, amongst different support groups: Ealing, Fulham and Hammersmith Sickel cell support group, Sickle Cell Society, Sickle Cell Cause and online Sickle Cell support groups who are solely based in London and the UK. As with any geographical setting, participants will be mainly from different ethnic minority backgrounds as well as belonging to different religious/cultural beliefs.

**Participants, sampling strategy and sampling size**

The sample population will come from sickle cell support groups from the London Borough of Brent and Ealing, Tottehnham, and across London boroughs. This was compared to data obtained via a literature review which was critically discussed. This was achieved by literature searches, analyzing, comparing and contrasting different publications.

The participants in this research are individuals who currently have sickle cell living in different boroughs in London. Part of the strategy was attending these support groups and explaining to these patients what the questionnaire consists of and the need to obtain results. In addition to this, these questions were distributed online. Therefore, the method used was not random,
Data collection methods and tools

The data used in this research was retrieved from an online survey from the support groups mentioned. The survey consisted of questions: identifying what type of sickle cell, ethnic background, disease management, pain management, how frequent hospital admission occurs, patient, staff interaction and adequate care based on their experience during hospital stay. This was measured using Microsoft Excel. Numerical variables such as frequency of admission rate, how regular medication is required will be measured.

Also whether pain and treatment management are effective and what actions used were measured. Since this survey will be completed online, participants can fill out these surveys within their own time so that the survey is filled out properly to avoid any mistakes.

Literature searches were conducted online databases: Pubmed, Springer, Nature and Science Direct, Google Scholar, Wiley Online, Pubmed, NCBI, Middlesex University Library. Keywords were used in search terms to attain results from relevant literature.

The search terms used:

Sickle cell disease, demographics, hospital admission, drug treatment, genetics, alternative treatment, staff and patient care.

After relevant searches were made, more stringent searches were conducted to produce a more coherent understanding from a general perspective. This included, referencing reports from literature already in use. Other pieces of information such as abstracts were also read and analysed.

Primary data was sourced from surveys completed, this encompassed "raw data". Extensive through checks were carried out to make there were no incomplete sections. This showed which sickle cell variant is more problematic among the population and whether it correlates to hospital admission rate, while comparing it to previous available and data obtained with the permission by the Sickle Cell Society and other publications. Furthermore, more patients handed out brand new surveys to family members and other local support groups to get more people involved. Secondary data was obtained from Sickle Cell Society online publications and AIJubri et al (2012), regarding hospital admissions within 2001-2010. This data set had data from admissions within England and London. Other secondary data were extracted from studies conducted by Sickle Cell Society PREM study and data from Green et al (2012) for hospital admission of sickle cell patients in the London Borough of Brent. Before any analysis was carried out, data was thoroughly checked to ensure data was accurate and there were no discrepancies.

Data analysis

Data analysis achieved in this research was conducted using Excel. Excel will be used effectively by dividing the variables into the appropriate categories, e.g. whether descriptive, analytical. In turn, statistical measures such as the mode, mean, median, percentage and other forms of statistical metric will be applied and further analyzed. For instance, the dependent variable is the pain and management of sickle cell disease and the independent variables are gender or age.

Alongside statistical analysis, a correlation coefficient will show whether there was any possibility that pain and management relate to hospital admission. Any unanswered questions will be included in the analysis and use of Excel will be critically analyzed in the discussion.

Analysis was conducted by comparing results attained with results from different local support groups who had different types of sickle cell: sickle cell trait (AS), SS, SC, SE, and beta thalassaemia. Descriptive analysis of patients who were admitted into hospital were admitted on either a weekly, monthly or yearly basis.

III. RESULTS

The total number of patients who took part in this study were based in London, and attended different hospitals and centres was 65. Out if these patients, 65 had sickle cell disease with most 95% of them being hospitalized (this is not including those who have had subsequent readmission and hospitalization within the type period this research was collected). Of the 65 participants, 54% were female and 46% were male (Table 2).
Table 2 illustrates the percentage ratio of participants involved in this study. The majority of participants were female (62.4%), whereas the remaining minority of participants male (37.3%).

<table>
<thead>
<tr>
<th>Gender</th>
<th>Percentage</th>
</tr>
</thead>
<tbody>
<tr>
<td>Male</td>
<td>37.3%</td>
</tr>
<tr>
<td>Female</td>
<td>62.4%</td>
</tr>
<tr>
<td>Total</td>
<td>100%</td>
</tr>
</tbody>
</table>

Table 3a illustrates regression statistics obtained using ANOVA. Multiple R had the highest total (0.94), R squared co-efficient (0.89) and adjusted R square (0.86). With a standard error of 8.63. The p-value achieved was at 0.04, there is a level of statistical significance.

<table>
<thead>
<tr>
<th>Regression Type</th>
<th>Regression Value</th>
</tr>
</thead>
<tbody>
<tr>
<td>Multiple R</td>
<td>0.944486</td>
</tr>
<tr>
<td>R Square</td>
<td>0.892054</td>
</tr>
<tr>
<td>Adjusted R Square</td>
<td>0.865068</td>
</tr>
<tr>
<td>Standard Error</td>
<td>8.63867</td>
</tr>
<tr>
<td>P-value</td>
<td>0.004537</td>
</tr>
</tbody>
</table>

Table 4 shows whether hospital admission improves sickle cell condition. Majority of participants voted 'No' (37), while the minority voted yes (28).

<table>
<thead>
<tr>
<th>Hospital admission condition?</th>
<th>improve sickle</th>
<th>Frequency</th>
</tr>
</thead>
<tbody>
<tr>
<td>Yes</td>
<td></td>
<td>28</td>
</tr>
<tr>
<td>No</td>
<td></td>
<td>37</td>
</tr>
<tr>
<td>Total</td>
<td></td>
<td>65</td>
</tr>
</tbody>
</table>

Table 5 illustrates whether hospital admissions whether introduction of treatment plans can reduce hospital admission. Majority of participants said 'Yes' (36), while the minority said 'No' (29).

<table>
<thead>
<tr>
<th>Do you think treatment plans can prevent hospital admission?</th>
<th>frequency</th>
</tr>
</thead>
<tbody>
<tr>
<td>Yes</td>
<td>36</td>
</tr>
<tr>
<td>No</td>
<td>29</td>
</tr>
<tr>
<td>Total</td>
<td>65</td>
</tr>
</tbody>
</table>

*Table 6a (see Appendix 1)
Table 7a illustrates votes made by 65 participants with sickle cell on whether specialized community care prevents hospital admission. The highest votes were those who voted 'Yes' (30), followed by those who voted 'No' (15). The lowest votes were participants who voted 'Don't Know' (15).

<table>
<thead>
<tr>
<th>Can specialized community care prevent hospital admission?</th>
<th>Frequency</th>
</tr>
</thead>
<tbody>
<tr>
<td>Yes</td>
<td>30</td>
</tr>
<tr>
<td>No</td>
<td>20</td>
</tr>
<tr>
<td>Don't Know</td>
<td>15</td>
</tr>
<tr>
<td>Total</td>
<td>65</td>
</tr>
</tbody>
</table>

Table 7b shows suggestions made by participants with sickle cell. Two of the highest votes were seen with more care in the community, (20) and home care and visits (20). Followed by participants who voted for day care centres (14). The least improvement suggested was rehab centres (11). (N=65, p=0.05).

<table>
<thead>
<tr>
<th>How can NHS improve in reducing hospital admission</th>
<th>Frequency</th>
</tr>
</thead>
<tbody>
<tr>
<td>More care in the community</td>
<td>20</td>
</tr>
<tr>
<td>Day Care centres with beds</td>
<td>14</td>
</tr>
<tr>
<td>Rehab centres</td>
<td>11</td>
</tr>
<tr>
<td>Home care and visits</td>
<td>20</td>
</tr>
<tr>
<td>Total</td>
<td>65</td>
</tr>
</tbody>
</table>
Table 8a shows votes made by 65 participants with sickle cell about their hospital experience. Majority of participants voted that the efficiency of medical staff was not satisfactory (38), whereas the minority of participants voted efficiency of medical staff was satisfactory (27).

<table>
<thead>
<tr>
<th>Efficiency of medical staff satisfactory</th>
<th>Frequency</th>
</tr>
</thead>
<tbody>
<tr>
<td>Yes</td>
<td>27</td>
</tr>
<tr>
<td>No</td>
<td>38</td>
</tr>
<tr>
<td>Total</td>
<td>65</td>
</tr>
</tbody>
</table>

Table 9b shows some of the most common types of pain participants experience during a sickle cell crisis. Majority of patients felt their pain they experienced was chronic (25), followed by acute (21). Shortness of breath was experienced among participants (10), whereas a few experienced muscle stiffness (8). The lowest classified their pain as other.

<table>
<thead>
<tr>
<th>Type of pain</th>
<th>Frequency</th>
</tr>
</thead>
<tbody>
<tr>
<td>Acute</td>
<td>21</td>
</tr>
<tr>
<td>Chronic</td>
<td>25</td>
</tr>
<tr>
<td>Muscle stiffness</td>
<td>8</td>
</tr>
<tr>
<td>Shortness of breath</td>
<td>10</td>
</tr>
<tr>
<td>Other</td>
<td>1</td>
</tr>
<tr>
<td>Total</td>
<td>65</td>
</tr>
</tbody>
</table>
Figure 6 illustrates the different type of sickle cell from 65 participants. The most frequent type of sickle cell was SS (46%), while the least frequent type of sickle cell was SE (3%). While the AS trait was the second most common type of sickle cell (36%). Followed by participants with the SC trait (15%).

Different age groups of participants with sickle cell

Figure 7 illustrates differing age groups of 65 participants with sickle cell. The mode was found in the 26-29 age group (25), followed by 30-34 years (15), followed by 22-25 years old (13), followed by 35 years and above (8). 18-21 years was observed to be the lowest age group with the lowest number of participants (3).
Rate of hospital admission vs number of patients admitted into hospital.

Figure 8 illustrates the rate of hospital admission amongst 65 participants. The highest rate of admission was seen at a monthly rate (35), followed by yearly, followed by twice a week (10), followed by 3-4 times a week (5). The lowest rate was seen at once a week (2).

Figure 9a illustrates the type of preventive treatments SC patients take on a regular basis to manage their condition to lower their chances to be admitted into hospital. Pain relief was the most common used treatment (40), followed by oxygen therapy (10), followed by Iron supplements (9), and followed by exercise, followed by antibiotics. The least type of preventive treatment was herbal medicine (1).
This was very crucial and important in helping to define and decide whether preventive treatment is effective in reducing hospital admission amongst sickle cell patients and whether better pain management and plans need to be put in place. For this reason, the null hypothesis (H0) was, that preventive treatments do not reduce hospital admission. While the alternative hypothesis (H1) was, to determine whether preventive therapies reduce hospital admission. Using the p-value, the level of significance was set to 0.05.

Based on the results, the null hypothesis was rejected as the level of significance is less than 0.05 (Table 4). Therefore, the alternative hypothesis was accepted. The most frequent amount of times those admitted to hospital are monthly admissions (Figure 8).

The mode also suggested that most patients would refer to use pain relief, iron supplements, oxygen therapy, herbal medicine and pain relief as a means of managing their sickle cell crisis (Figure 9a). Whereas few believed that herbal medicine made a significant difference in managing their symptoms (Figure 9b).

Regression analysis suggested that the mean and data fell within the data (Table 3a). This suggested that the data has a good fit, showing statistical significance.

The results also suggested that patients with sickle cell would like treatment plans (Table 5). The results also suggested that the hospital stay did not improve sickle cell condition (Table 4). Many patients believed that alternative therapy e.g. herbal medicine, heat therapy, regular exercise, helped alleviate some of their symptoms, which made them less likely to be admitted into hospital (Figure 9a, 9b).

Pain management also suggested that regular NSAIDs, was satisfactory in managing pain, whereas hospital admission was only necessary if the pain was too severe and they required morphine (Figure 10a, 10b). Even in this instance, many felt that nurses and healthcare professionals were not supporting and caring towards them (Table 7b, 8). These were the key findings identified from the questionnaire (Table 5). Having said this, these issues will not simply go away.

The results also suggested that patients can manage their sickle cell more effectively without relying on a third party or go into hospital and stay confined to a bed for a set number of hours, days, weeks or even months due to the nature of the disease (Table 7b, 8).

This is supported by AlJubri et al (2012), where it has been mentioned that general practitioners (GPs) have collected information on their patients who are registered with sickle cell who have been admitted into hospital. This allows room for improvement of care that is necessary as sickle cell patients often get admitted into hospital on a regular basis.

As mentioned previously, the results suggested (Figure 10) that there is lack of efficiency, satisfaction and care they receive in hospital. But AlJubri et al (2012) and Ane et al (2002), stated that many were unhappy with the GP care and primary care.

On the other hand, this must be in line with diseases that are part of the GP Quality and Outcomes Framework, which sickle cell disease is not included (AlJubri et al 2012). Unfortunately, this is problematic, as sickle cell disease continues to rise (AlJubri et al., 2012).

The results suggested in this research, that those who were admitted into hospital once (Figure 8) had reduced chances of being readmitted into hospital due to receiving the appropriate medication. This is supported by Ane et al (2012), who stated that short term stay and regular hospital admission (74%) were in-patients who were admitted regularly.

In addition to this, Benjamin et al (2000), also support the argument that sickle cell crisis episodes are frequently treated inappropriately with respect to time and quality of care. This also adds to unnecessary waiting time, delayed treatment, unhappy patient care and ultimately an extended stay in hospital (Benjamin et al, 2000). Based on this argument, the null hypothesis was rejected and the alternative hypothesis was accepted.

On this basis, it can be suggested that these type of sickle cell patients are suitable for primary care to reduce admission (Ane et al 2012). The results from this research also support the idea of increased supportive management (Table 10) when admitted into hospital. The results also suggested that home care visit, day centres would benefit sickle cell sufferers and reduce hospital admission (Table 7b).

This is supported by Ane et al. (2012), who reported that there was a decrease in hospital admission due to sickle cell patients being directed to outpatients, sickle cell centres and day care centres.

Existing literature has reported that day case management, centres and primary care settings are essential to curb hospital admission as well as the cost needed to tackle sickle cell disease and crises that they endure on a daily basis (Benjamin et al, 2000).

Day centres have been set up to curtail and increase better management of sickle cell disease regarding the “uncomplicated” sickle pain crises (Wright et al., 2004). The results from Wright et al (2004), suggested that introduction of day centres had reduced hospital admission by 43% and a further 49% decline in beds being occupied. A 3 year study documented in its final year, approximately 84% of patients were not admitted into hospital (Wright et al 2004).

This supports the results achieved in this research. However, Wright et al. (2004), further suggests that day case management regarding excruciating painful crises are effective and reduce hospital admissions for sickle cell patients who may not need to be admitted (Elhasid, 2017).

Wright et al (2004), also suggested that this method of approach is also cost effective and also safe for sickle cell patients. AlJubri et al (2012), and existing literature also suggest that interventions and preventive therapies are necessary to reduce hospital admission.

This is important as the spectrum of those who experience any disease varies from mild to severe and need to be attended to with the best quality of care that suits and covers all their needs. In addition to this, studies have shown that GP practices are now part of a GP education programme regarding patients who frequently attend A and E/hospital admission due to SCD (Ane et al, 2012).

As reported by Ane et al (2012), this benefits not only healthcare professionals in providing ‘educational intervention’, all medical staff such as GPs, nursing, medical, and management are helping sickle cell patients manage their disease better thereby reducing hospital admission and hospital stay (Ane et al 2012).
This also contributes to better and effective efficiency in medical and hospital staff. Even when compared with existing literature, it has been confirmed that there is an inappropriate use of acute inpatient care and it is increasing (Ane et al. 2012). This is due to hospital staff and resources, are distracted from those who need such care immediately thus increasing hospital stay and waiting times unnecessarily (Ane et al. 2012). This is similar to the results achieved in the research (Table 6b).

It has also been suggested in other studies that preventive treatments e.g. immunization for pneumococcal and meningitis, taking antibiotics (penicillin), blood transfusion etc are able to reduce the likelihood of being admitted into hospital on a regular basis (McGann et al., 2013). However, with all these preventive measures in place, secondary conditions associated with sickle cell disease e.g. leg ulcers, chronic lung disease and excessive cardiac damage due to iron overload has increased hospital stay (McGann et al., 2013).

As the results show that painful crisis is the most frequent problem for hospital (Table 9b). This is supported by the Salman and Hassan (2015), and results from Brown et al in Nigeria (61.5%), Akar and Adekile (Kuwait).

With the results achieved from this research and the current publications (AlJubri et al, 2012) had suggested that the NHS need to do better with their policy in regards to sickle cell hospital admission. This can be combated using care plans e.g. through education intervention as many have suggested to be more effective (Green et al 2012). As their results suggested that indicated that having local day hospitals recued hospital admissions by 40%, in addition to having seen a heavy declined of hospital stay (804 bed days) within a three-year period as a result of using a haematology inpatient bed a mere cost of £356 (Green et al, 2012; Wright et al 2004; Netten et al 2002).

Regarding pain management in general, the results suggested that alternative treatment such as herbal therapy and healthy exercise helps to alleviate their pain and sickle cell related symptoms (Figure 9a, 9b). Existing literature regarding herbal remedies have mentioned and measured the effectiveness of controlling sickle cell disease (Okpuzor et al., 2008). It has been suggested that crude extracts from plants manages sickle cell, as well assattending to traditional medicine have been effective (Okpuzor et al. 2008). It was also suggested that eating Tiliap ia (dried fish) and dried prawns assisted in inhibiting polymerization of HbS and improving iron levels (Okpuzor et al., 2008).

Olujobungbe and Yardumian (2014) reported that other treatments such as magnesium salts, or oral medication (Clotrimazole) are effective in reducing sickling in sickle cell patients. However, it is not available in UK compared to the USA (Olujobungbe and Yardumian, 2014).

Furthermore, it can be suggested that major changes can occur on a practical level. This is supported by Green et al., (2012), where having a baseline to watch progression and to allow for improvements in primary care for sickle cell patients and also reducing hospital admission. This answers the research question proposed "is better management in place required to reduce hospital admission?" As studies are in support of better management will reduce hospital admission.

On the other hand, if such changes were to be implemented as shown in the studies carried out, then this would also form as an integral part of ensuring that primary care settings, e.g. GPs that already are providing education packages that will essentially address improving patient experience as well as monitoring the health inequalities (Green et al, 2012). On the other hand, with such data, this can be analyzed further to make sure that primary and secondary care is embedded into the multidisciplinary teams to attain patient satisfaction (Green et al, 2012).

In addition to this, sickle cell patients tend to be frequent users of hospital care, in particular in-patient care (Green et al, 2012). That then leaves this research question open for more research to be repeated for future comparison. However, this doesn’t take away from the fact that more effective management of patients within the community will inevitably decrease hospital admission (Green et al., 2012). Thereby improving local health economy and funds that can be re allocated to sickle cell centres and lessen the cost the NHS is undergoing (Green et al, 2012).

This argument is also supported by the NICE guidelines (2011), where it states that "patients should be listened to and discuss their treatment plan, treatment received during previous episodes, any concerns they may have about their current episode and psychological/social support they need". These recommendations are necessary and crucial for hospitals and organizations to implement these recommendations.

Even with the better management observed in patients with sickle cell, there are still incidences of patients still being admitted into hospital and require specialist treatment and hospitalization (Salman and Hassan, 2015). On the other hand, sickle cell patients are also still being admitted into hospital even with the decline in morbidity and mortality (Salman and Hassan, 2015).

However, having said this, interventions aimed at preventing further sickle cell complications can reduce the negative economic impact (Salman and Hassan, 2015). On the other hand, even as the results suggested that many patients required pain relief, (Table 7b), studies have shown that those with painful crisis and require pain relief have a greater chance of being readmitted into hospital (Salman and Hassan, 2015). Whereas, in another study (Loureiro et al, 2009), such a relationship didn't exist, rather readmissions occurred due to pre-existing and recurrent vasocclusive crises and kidney failure (Salman and Hassan, 2015; Loureiro et al, 2009).

Future Recommendations

Based on NCEPOD (2008), guidelines in place have advised patients to be aware of the importance of monitoring and consistent review, which will lead to effective management of their condition. This is supported by primary and secondary care Trusts (NCEPOD, 2008). Furthermore, sufferers of sickle cell tend to be admitted into hospital more regularly, which poses a lot of problems when ensuring management is carried out (NCEPOD, 2008). In addition to this, results achieved in this research suggested that healthcare professionals were not given them the treatment they deserved (Table 6b).

This is in support with studies by NCEPOD (2008), who reported that medical staff were lax in managing patients with sickle cell. Recommendations in place by NCEPOD (2008), were to inform and advise patients to know they are aware of patterns that lead to an impending crisis.
Another recommendation that medical staff should be wary and understanding that complications associated with sickle cell can be detrimental (NCEPOD, 2008).

Alternative and herbal medicine for treating sickle cell

Introduction of using medicinal plants such as Aloe Vera, Piper guineensis, etc. has shown in vitro, it inhibits sickling (Okpuzor et al., 2008). In addition to this, medicinal plants have its benefits which have been overlooked, as its holistic use can work as painkillers. So for future studies, implementing alternative medicine in care plans to reduce hospital admission. Ameh et al. (2012) and Okpuzor et al. (2008), concluded that anti-sickling herbs that can be used in future studies. However, current strategies must be implemented for this to be effective (Ameh et al., 2012).

Currently, NISPRAN (made from 4 plant extracts) has been used successfully in Nigeria to treat sufferers with sickle cell and has been used as treatment since 2006 (Ameh et al., 2012). It has passed phase II clinical trials and the United States Federal Drug Agency (USFDA) have approved of its safety and efficacy to initiate phase III trial (Ameh et al., 2012).

Exercise and physiotherapy in reducing sickling conditions

As well as using herbal treatment, exercise and physiotherapy have shown to decrease the occurrence of acute chest syndrome, pain and associated infections e.g. chest infections (Rees et al., 2003). But as with all recommendations, it cannot account for all known complications of sickle cell disease as it is beyond the scope of this research. Future studies would need to factor more on management of current treatments that have been previously mentioned (Rees et al., 2003).

The last recommendation suggested is that hydroxyurea is the only drug that has decreased acute and painful crises for sickle cell patients by 50% (Agarwal et al., 2014). It has also been advised to treat patients with hydroxyurea more frequently (Agarwal et al., 2014) to reduce and avoid sickle cell patients developing chronic diseases (Table 9b).

IV. LIMITATIONS

There were a number of limitations when conducting this research. Firstly, when carrying out this research, getting support groups to fill out all completed forms was difficult. Initially due to the nature of the disease, many had been in and out of hospital. This greatly affected the number of participants involved as some were unable to complete it. Another limitation, many were committed to being on board with participating with the study, who were all provided with the background information of the research, the questionnaire and what was required. However, some were unable to continue due to miscellaneous reasons. Having said that, they were informed that is optional to be part of the research and that their contributions still mattered.

Some of the participants who were initially involved may have had issues with the number of forms they were required to fill out, hence why the initial hesitation to complete such forms. On the other hand, others who completed all forms stated that the instructions were easy to follow and had passed on the questionnaire to other groups. This encouraged more to get involved to participate outside their support groups. Furthermore, the Sickle Cell Society were unable to go through with questionnaire as the forms to fill out were too much even in electronic form. In addition to this, they preferred to be in an electronic format e.g. Survey Monkey.

However, given the time frame and the late notice, lack of communication and feedback from them, it was not possible to continue with the Sickle Cell Society. To counter this for future purposes, online contributions from other online support groups and individuals with sickle cell were able to participate and complete the forms necessary. To amend this research for repetition, using Survey Monkey would solve this problem.

As mentioned previously, talking about the research to different groups and individuals with sickle cell made many interested. At the same time, even when it was explained that it's confidential, many still felt stigmatised and did not want others to know of their condition. In addition to this, the numbers of participants involved do not represent a specific representation of the population at risk. Even when applying the appropriate sampling size, it cannot account for the setbacks that will affect the aims and objectives set out initially during the initial conduct of research.

Another limitation seen in this investigation was, there were limited outcomes when using questionnaires as they lead to closed questions. This often leads to limited answers and options that participants may not necessarily expand on. For future purposes, one must either make questions broader and may utilise focus groups and interviews to get more information from participants. It can also be time consuming to shift through accurate and reliable data while disposing unnecessary data. Quantitative data needs a lot of time and requires patience to perform necessary data analysis. This can be challenging for the researcher in question when trying to make sure this research can be refined and retested for future purposes.

Due to the nature of this research and its short duration, this one of the limitations. Maybe an extended period would have yielded more participants and additional factors such as associated morbidity. As well as attaining data, since the standard of mean is so large and the regression is high, it does not necessarily mean that this study was an accurate study as the sample size was smaller than expected. With the initial sample size being 100, many along the course of the research, had either not completed the survey or simply ‘forgot’ even with reminders given on a basis at intervals. This reduced the sample size to 65. However, some initial participants who dropped out took longer to complete the survey due to hospitalization. Furthermore, as this study was voluntary, participants and respondents were not obliged to complete it as they were invited to know about this study and were given the option when to consent or partake in the study. Having a larger sample size would have demonstrated a better and better fit of data points (Table 3a). Plotting a regression graph would have illustrated the relationship achieved in the results (Table 3a).

V. CONCLUSIONS

The findings of the research undertaken, suggested that more needs to be done to tackle the ongoing battle with reducing hospital admission for sickle cell patients. As discussed and carried out with this research, it is evident that more specialised staff and training is needed.
As the results suggested, better management, care, access treatments to these patients are necessary to ensure their needs and wants are met at a very high standard. As the results suggested more community care e.g. homecare visits would benefit long term. Moving forward, suggestions on a more efficient care plan, framework (albeit an updated one), be in place for patients to feel safe and responded to.

As well as more community and home visits for sickle cell patients to manage their crisis better. It is the opinion of the author and based on the evidence obtained, that more research and awareness needs to be addressed to healthcare professionals across the NHS to endeavour that emotional, physical support is given to sickle cell patients.

Sickle cell disease is growing concern in the community making it a public health concern, which can reduce hospital burden if managed correctly. Preventive therapies combined with excellent patient care will inevitably reduce hospital admission.

ACKNOWLEDGEMENT
I would like to thank Middlesex University of London and my supervisor Lynne Jalalian for assisting me on this project during my Master’s in 2016-2017. Ethics approval was provided by Ethics governing body at Middlesex University of London

REFERENCES

[54] Welcome Trust,YourGenome (2017). Figure 3What is sickle cell anaemia?. [online] Available at: https://www.yourgenome.org/facts/what-is-sickle-cell-anaemia/ [Accessed 17 Aug. 2017].

AUTHORS

First Author – Lynne Jalalian, MSc, Middlessex University of London, L.Jalalian@mdx.ac.uk

Second Author – Ogechi Anokwuru, PhD candidate, MSc, BSc (Hons), Middlessex, University of London,