Patient Perspectives of Sickle Cell Disease Care in Emergency Centres of South African Hospitals

By

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Declaration

I, Nabeelah Peerbhai, hereby declare that the work on which this dissertation is based is my original work (except where acknowledgments indicate otherwise) and that neither the whole work, nor any part of it has been, is being, or is to be submitted for another degree at this or any other university. I authorise the university to reproduce, for research purposes, either the whole or any portion of the contents in any manner whatsoever.

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Abstract

Introduction:

Sickle Cell Disease (SCD) is an inherited, chronic blood disorder. Patients affected by SCD experience systemic, life-threatening complications that requires frequent emergency care. Western studies have reported poorer experiences by individuals with SCD when seeking emergency care compared to other chronic conditions.

Aim:

This study aimed to investigate the perceived experiences of adult patients with SCD who presented for care to the emergency centres of South African hospitals.

Methods:

A mixed methods approach was used to conduct this research. Participants were recruited from the Sickle Cell South Africa online support group and Groote Schuur Hospital in Cape Town. Surveys were completed by 51 participants and then analysed using both descriptive and comparative statistics. Open-ended interviews were conducted with eight of the participants, which were analysed using thematic analysis.

Results:

In the last five years, 47.5% of participants visited the emergency centres between two and five times, 29.4% attended more than five times and the remainder attended only once. Most respondents (45.1%) described their recent experience in the emergency centre as average, while 31.4% described their experience as good and 23.5% as poor. Exploring participant confidence levels identified 21.6% of participants felt very confident in the nurses treating them while more participants (33.3%) felt very confident in the doctor treating them.

Five themes emerged from the data 1) Knowledge and understanding of SCD amongst healthcare professionals and patients, 2) Treatment in emergency centres, 3) Patient advocacy and support, 4) Emotional and psychological impact and 5) Access to healthcare and care coordination. Participants identified a lack of knowledge and understanding amongst HCPs relating to SCD, which they believe, contributed to delayed care, misunderstanding of the condition, disbelief in their symptoms and ignorance regarding their pain. Notably, participants expressed a higher confidence in doctors' understanding of SCD than nurses', highlighting the need for targeted education interventions. Participants further emphasized the importance of comprehending their own condition and being able to articulate the nature of SCD pain, which may or may not be visible. The treatment received in the emergency centre,

such as the urgency of receiving medical care, the essential need for patient-centred pain management, challenges with the triage system and the positive influence of strong physician-patient interactions, played a role in the type of experience they had. Self-advocacy and family/social support were important factors when receiving emergency care. Patients developed various coping mechanisms yet expressed the challenge of navigating pain symptoms, anxiety, fear and uncertainty. Lastly, patients described transport and financial challenges when accessing healthcare services, as well as the need for coordinated healthcare services inclusive of genetic counselling. These results were consistent across both public and private facilities, with no statistically significant difference being noted between the two.

Conclusion:

Insight into patient experiences can actively involve patients in managing their health to ensure the patient's voice is heard. This research identified factors contributing to patient perceptions, the strengths of emergency care while suggesting potential solutions to better patient satisfaction.

In the name of Allah, Most Gracious and Most Merciful.

The source of grounding and understanding that our profession is sacred, dealing with the most precious gifts of life and intellect.

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List of Abbreviations

$\alpha_2\beta_2$	Two Alpha and Two Beta Proteins
A&E	Accident and Emergency
β6Glu→Val	Substitution in the Sixth Position of the β-globin Chain Valine for Glutamic Acid
CT	Cape Town
COVID	Coronavirus
DRC	Democratic Republic of Congo
EC	Emergency Centre
EMSSA	Emergency Medicine Society of South Africa
EDSC3	Emergency Centre Department Sickle Cell Care Coalition
ER	Emergency Room
GCs	Genetic Counsellors
GSH	Groote Schuur Hospital
Hb	Haemoglobin
HbA	Adult haemoglobin
HBB	Haemoglobin β
HbE	Haemoglobin E
HbF	Foetal Haemoglobin
HbS	Haemoglobin S
HbSS	Haemoglobin SS
HCPs	Healthcare Providers
KZN	Kwa-Zulu Natal
MMR	Mixed Methods Research
NBS	Newborn Screening
NC	Northern Cape
SA	South Africa
SCA	Sickle Cell Anaemia
SCD	Sickle Cell Disease
SCT	Sickle Cell Trait
SCSA	Sickle Cell South Africa
SSA	Sub-Saharan Africa
UK	United Kingdom
USA	United States of America
VOC	Vaso-occlusive Crises
WC	Western Cape
WHO	World Health Organization

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Chapter One: Introduction

1.1. Chapter Introduction

The study investigated perceptions and experiences of patients living with Sickle Cell Disease (SCD) who presented for care to emergency centres (ECs) in South African hospitals. The Emergency Medicine Society of South Africa (EMSSA) defines the EC as *"the dedicated area within a health facility that is organised and administered to provide a high standard of emergency care to those in the community who perceive the need or are in need of acute or urgent care. Emergency cases are those patients who present to the EC seeking non-scheduled care* (Twomey *et al*., 2012:2). Globally, the EC is also referred to as the Trauma Unit, Casualty, Emergency Room (ER) or Accident and Emergency (A&E) Department (Hardcastle, 2008).

The first chapter consists of a literature review relating to SCD, focusing on the phenotype, genetics, epidemiology and global research on the topic. In addition, the review will discuss SCD in South Africa (SA) and the role of genetic counselling. This chapter will serve as a background for the aim and objectives of this study, which will be introduced in more detail towards the end of the chapter. Chapter two will describe the methodological approach that was implemented in this study. The results and discussion will be explored in chapter three. The last chapter will provide a summary of the key findings, limitations of this study and future recommendations.

1.2. Literature Review

1.2.1. Definition and Genetic Aetiology of Sickle Cell Disease

Adult haemoglobin (HbA) molecules are tetramers comprising four subunits of proteins; two alpha and two beta proteins ($\alpha_2\beta_2$), each forming a globin chain (Weatherall & Clegg, 2001; Williams & Thein, 2018). The alpha globin proteins are produced by two alpha globin genes on chromosome 16 while the beta proteins are produced by one (*HBB)* gene on chromosome 11. This composition allows haemoglobin β the haemoglobin molecule to fold into a specific shape, allowing the protein to fulfil its major role of carrying oxygen (Williams & Thein, 2018). Variants in these genes lead to chronic disorders affecting haemoglobin (Hb) structure, function and synthesis, known as haemoglobinopathies (Modell & Darlison, 2008). Haemoglobinopathies can be further described as quantitative changes in the *HBB* gene such

as thalassemias, which lowers the amount of Hb and qualitative changes in the *HBB* gene which produce an abnormal Hb structure, as in the case of SCD (Modell & Darlison, 2008).

SCD was mentioned in Western literature in 1910 by Dr J Herrick while investigating a patient with severe anaemia (Herrick, 1910). Dr Herrick documented irregularly shaped erythrocytes, describing them as thin, elongated and sickle-shaped. SCD is an umbrella term used when the primary type of haemoglobin present in an individual with SCD is sickled haemoglobin or haemoglobin S (HbS). However, other forms of SCD can also occur in combination with other haemoglobin gene variants such as Haemoglobin C, D, E and β-thalassemia. Although SCD is often used interchangeably with the term Sickle cell anaemia (SCA), SCA refers to a homozygous HbSS state (Modell & Darlison, 2008; Williams & Thein, 2018).

In 1949, Linus Pauling and his colleagues discovered that abnormal production of a protein could be the cause of a genetic condition (Pauling et al., 1949). Thereafter, in 1956 Vernon Ingram described a point mutation within the *HBB* gene to be the cause of SCD (Ingram, 1956). HbS aggregate into rigid polymers due to a substitution of valine in place of glutamic acid in the sixth position of the amino acid sequence in the β-globin chain (β6Glu→Val) (Odièvre et al., 2011). This substitution produces abnormal β-globin chains which polymerise during deoxygenation and dehydration.

The genes encoding haemoglobin are developmentally regulated, producing specific types of globin during different stages of human development (Figure 1.1) (Forget, 2011). Foetal haemoglobin (HbF) consists of two alpha proteins and two gamma proteins ($\alpha_2\gamma_2$). HbF is the predominant type present in the blood during foetal life. However, at birth, gene expression switches the type of haemoglobin produced, gradually replacing HbF with HbA by 6 months of age. This phenomenon is termed the haemoglobin switch (Forget, 2011). A healthy adult has between 95-98% of HbA and only 1-2% HbF. As the production of HbF decreases, HbA production increases. In individuals with SCD, HbS replaces HbA, leading to the phenotypic expression of the condition arising after 6 months of age (Williams & Thein, 2018).

SCD is inherited in an autosomal recessive manner (Modell & Darlison, 2008). Carriers of SCD are heterozygous for the *HBB* β6Glu→Val variant (Williams & Thein, 2018). These individuals are said to have the sickle cell trait (SCT) (Weatherall & Clegg, 2001; Williams & Thein, 2018). Individuals with SCT need to understand the reproductive outcomes, as they are at risk of passing the variant on to their children. Furthermore, if their partners are also carriers, they have a 25% chance of having an affected child during each pregnancy. SCD can also occur in a compound heterozygote state, due to the inheritance of HbS in combination with a different HBB variant (Chakravorty & Williams, 2015; Williams & Thein, 2018).

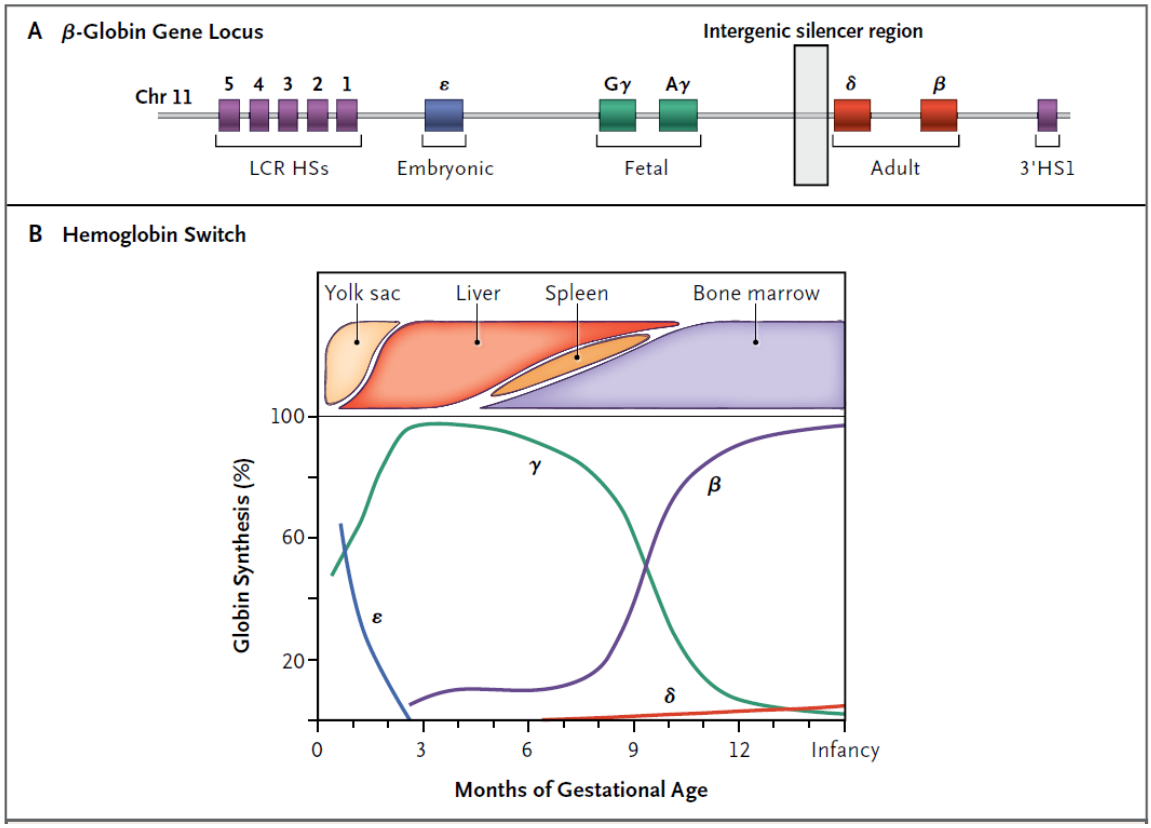

Figure 1.1: The HbF to HbA Switch. Section A Illustrates the Developmental Genes While Section B Illustrates the Timing of the Switch (Forget, 2011)

1.2.2. Epidemiology Sickle Cell Disease

Haemoglobinopathies are reported to be the most common monogenic disorders with 300 000 to 400 000 babies born each year worldwide. Approximately, 90% of these births are reported to occur in developing countries (Williams & Weatherall, 2012). According to the World Health Organization (WHO), 20-25 million people worldwide have SCD and of these affected individuals, 12-15 million people affected with SCD reside in Sub-Saharan Africa (SSA), of which 25%-30% of SCD births occur in Nigeria (Oron et al., 2020). It is estimated that 5%-40% of individuals carry SCT in Sub-Saharan Africa (Weatherall & Clegg, 2001).

The distribution of SCD and SCT globally demonstrates the positive selection for this condition in malaria endemic regions as seen in figure 1.2 (Williams & Thein, 2018). Individuals with SCT have lower mortality rates from malaria conferring an evolutionary advantage (Weatherall & Clegg, 2001; Williams & Thein, 2018). While individuals with SCD have a certain degree of immunity to malaria, they can still get malaria infections which exacerbate symptoms of SCD. The protective mechanism is not fully understood yet, but it is thought to involve immune-mediated mechanisms conferring some level of protection against *Plasmodium falciparum* (Weatherall & Clegg, 2001; Williams & Thein, 2018). A paper by Chackravorty & Williams

(2014) describes the HbS mutation to be the archetypal example of natural selection due to the HbS carrier frequency of 40% in malaria endemic populations. Apart from these populations, the HbS variant has spread globally due to human migration.

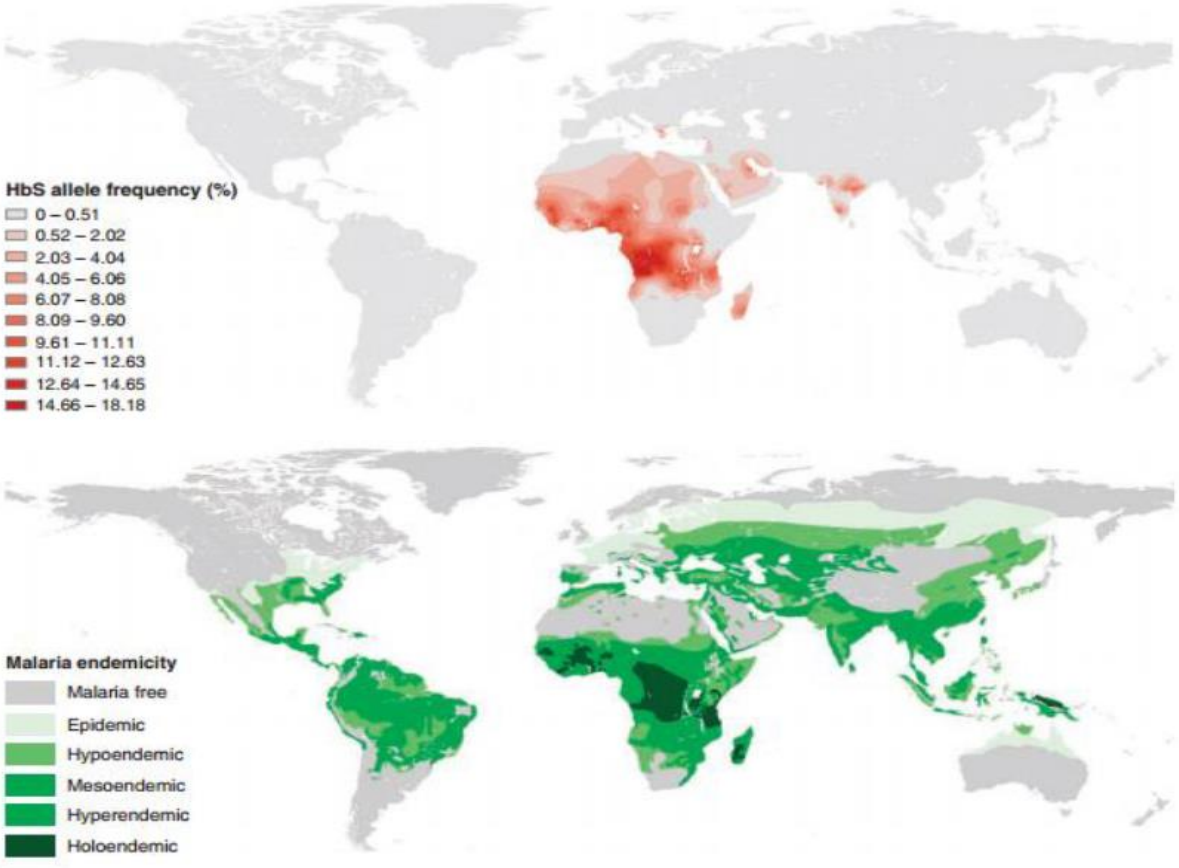

Figure 1.2: The Distribution of the Sickle Cell Gene Corresponding to Malaria Endemic Regions in the World (Williams & Thein, 2018)

Human migration due to colonization, globalisation and economic opportunities has led to a diverse distribution of genetic traits (Wonkam et al., 2012). This has caused the emergence of SCD in areas of the world that were not historically associated with the condition. Recognizing that the spread of HbS is not limited to specific ethnic groups or regions and tracing the global spread is important for public health efforts to provide appropriate healthcare services for individuals affected by SCD (Snyder et al., 2022).

In SA, SCD was originally reported to have a low prevalence rate attributed to the region having few occurrences of malaria (Beighton & Botha, 1986). Literature regarding the prevalence in SA is rare, however, a study done in 1986 investigated the prevalence of inherited haematological conditions in the Black SA population. The study mentioned several different haemoglobin variants occurring in the mixed ancestry population (Beighton & Botha,

1986). The most common variants seen are HbS and HbE with a prevalence of 1%. In addition, SCD was also reported in the SA Indian and White populations. In the SA Black population, individuals were found to carry SCT; however, the frequency was only 0.2% (Wonkam et al., 2012).

The immigration of individuals from neighbouring countries to SA brought a 2% rise in the number of SCD cases from 2002 (Wonkam et al., 2012). While differences in prevalence have been seen in different ethnic groups in SA, the increase has been attributed to the rising political instability in neighbouring countries, causing individuals to seek refuge in SA (Wonkam et al., 2012). Furthermore, the study reported that 93.1% of individuals with SCD seen in a paediatric hospital in Cape Town (CT) were from neighbouring countries.

Literature detailing the known SCD genotypes, characteristics and prevalence worldwide was published in 2010 (Rees, Williams & Gladwin, 2010). HbSS is the most common and severe type accounting for 70% of cases in Black Africans. Haemoglobin SC is the second most common type with 25-35% also affecting Black African individuals. HbS with β-thalassemia is reported as the third most common type, highly prevalent in the Eastern Mediterranean and Indian populations with varying disease severity. Table 1.1 displays the other genotypes seen globally in lower prevalence.

Table 1.1: SCD Genotypes, Distribution and Characteristics (Rees, Williams & Gladwin, 2010)

	Characteristics
Severe sickle-cell disease	
HbS/S (β6Glu>Val/β6Glu>Val); sickle-cell anaemia	The most common form of sickle-cell disease
HbS/β^0 thalassaemia	Most prevalent in the eastern Mediterranean region and India[14]
Severe HbS/β^+ thalassaemia	Most prevalent in the eastern Mediterranean region and India; 1–5% HbA present[14]
HbS/OArab (β6Glu>Val/β121Glu>Lys)	Reported in north Africa, the Middle East, and the Balkans; relatively rare[14]
HbS/D Punjab (β6Glu>Val/β121Glu>Gln)	Predominant in northern India but occurs worldwide[14]
HbS/C Harlem (β6Glu>Val/β6Glu>Val/β, β73Asp>Asn)	Electrophoretically resembles HbSC, but clinically severe; double mutation in β-globin gene; very rare[15]
HbC/S Antilles (β6Glu>Lys/β6Glu>Val, β23Val-Ile)	Double mutation in β-globin gene results in severe sickle-cell disease when co-inherited with HbC; very rare[16]
HbS/Quebec-CHORI (β6Glu>Val/β87Thr>Ile)	Two cases described; resembles sickle-cell trait with standard analytical techniques[17]
Moderate sickle-cell disease	
HbS/C (β6Glu>Val/β6Glu>Lys)	25–30% cases of sickle-cell disease in populations of African origin[13]
Moderate HbS/β^+ thalassaemia	Most cases in the eastern Mediterranean region; 6–15% HbA present[14]
HbA/S Oman (β^A/β6Glu>Val, β121Glu>Lys)	Dominant form of sickle-cell disease caused by double mutation in β-globin gene; very rare[18]
Mild sickle-cell disease	
Mild HbS/β^{++} thalassaemia	Mostly in populations of African origin; 16–30% HbA present[14]
HbS/E (β6Glu>Val/β26Glu>Lys)	HbE predominates in southeast Asia and so HbSE uncommon, although frequency is increasing with population migration[19]
HbA/Jamaica Plain (β^A/β6Glu>Val, β68Leu/Phe)	Dominant form of sickle-cell disease; double mutation results in Hb with low oxygen affinity; one case described[20]
Very mild sickle-cell disease	
HbS/HPFH	Group of disorders caused by large deletions of the β-globin gene complex; typically 30% fetal haemoglobin[14]
HbS/other Hb variants	HbS is co-inherited with many other Hb variants, and symptoms develop only in extreme hypoxia

1.2.3. Pathophysiology and Clinical Characteristics

The polymerization of beta-globin chains alters the structure of erythrocytes from a biconcave shape to a deoxygenated, sickle shape. The level and frequency of polymerization lead to the vascular pathophysiology of SCD and is *inter alia* dependent on HbS and HbF concentration, oxygen levels and pH (Lovett, Sule & Lopez, 2014; Odièvre et al., 2011). The pathophysiological mechanism of SCD was detailed in the 1960s to 1970s which led to a better understanding of the condition (Odièvre et al., 2011). The haemoglobin switch, as described earlier, switches the production of embryonic haemoglobin to HbF in the first 3 months of conception. After birth, HbF is replaced by HbA with lower amounts of HbF being produced after 6 months of age (Forget, 2011).

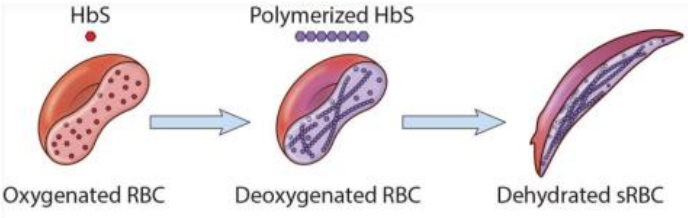

Figure 1.3: Pathophysiology of Sickle-Shaped Blood Cell (Williams & Thein, 2018)

Symptoms of SCD are therefore present after 6 months of age as HbS is produced instead of HbA. HbS produces long, insoluble polymers that when deoxygenated, form the characteristic sickle-shaped red blood cells (Figure 1.3) (Williams & Thein, 2018). Haemolysis and haematological complications arise due to the short lifespan of sickle-shaped blood cells. Research has estimated the average lifespan of these abnormally shaped cells to be between 6-9 times shorter than a normal red blood cell (Rees, Williams & Gladwin, 2010). SCD is a debilitating, multisystem disorder. Individuals with SCD experience a myriad of health problems that can be grouped into four categories: haemolysis and haematological complications, vaso-occlusive crises (VOC), end-organ dysfunction and infection (Lovett, Sule & Lopez, 2014).

An acute anaemic crisis is characterized by a rapid reduction in haemoglobin, increased reticulocytes and increased levels of bilirubin may occur (Makani et al., 2013). Patients typically present with jaundice, nosebleeds, and symptoms of anaemia. Chronic haemolysis can lead to the development of gallstones. Viral infections may precipitate an aplastic crisis. A splenic sequestration crisis is a life-threatening complication characterised by red cell sequestration in the spleen and a sudden drop in haemoglobin in patients where the spleen is not yet fibrotic after previous splenic infarctions (Makani et al., 2013).

VOC are a hallmark feature of SCD occurring when sickle-shaped blood cells adhere to the walls of blood vessels preventing the normal flow of blood (Lovett, Sule & Lopez, 2014; Makani et al., 2013). The resulting decrease in blood flow can lead to tissue ischemia causing acute and chronic pain episodes as well as lead to organ damage. VOC can be life-threatening when associated with chronic anaemia and hypovolemic shock. Patients with SCD experience complex pain that can be multifactorial; however, the primary aetiology of acute, episodic pain is reported to be from VOC and most frequently involves marrow-rich areas of the body like the legs and hands (Glassberg, 2017). Factors such as an increase in the percentage of sickle-shaped cells present, nutritional, stress factors and increase in barometric pressure have been suggested to trigger pain crises in patients (Chakravorty & Williams, 2015). Individuals with SCT can display symptoms such as respiratory complications at high altitudes or due to dehydration, kidney problems, blood clots and leg ulcers (Hulsizer et al., 2022).

Haemolytic events and VOC lead to tissue damage throughout the body. Patients with SCD often experience acute or chronic complications such as acute chest syndrome, delayed growth and sexual maturity, SCD retinopathy, liver damage and cerebrovascular complications arising from obstructed blood flow (Chakravorty & Williams, 2015). Ischemic strokes, haemorrhages and seizures have also been reported in the literature. Organ damage is reported to be irreversible and can be an important risk factor affecting a patient's prognosis. In addition, infection is also a contributing factor to morbidity and mortality in patients (Chakravorty & Williams, 2015; Makani et al., 2013). SCD increases one's susceptibility to infections due to immune deficiencies e.g. reduced splenic function, complications related to SCD such as end organ failure and chronic haemolysis resulting in a higher bone marrow turnover (Makani et al., 2013).

1.2.4. Prognosis and Management of Sickle Cell Disease

SCD prognosis ranges from mild to severe, impacting quality of life and expectancy. Studies note that the median age of survival is 38 years old for males and 42 years old for females (Jiao et al., 2023). Main causes of death include cardiopulmonary disease, chronic organ dysfunction, infections and iron overload (Manci et al., 2003). While countries like the United States of America (USA) and United Kingdom (UK) have reduced mortality through the implementation of newborn screening (NBS) and comprehensive care strategies such as antibiotic prophylaxis, vaccination and hydroxyurea, a recent USA study determined the life expectancy of individuals with SCD to be ~2 decades shorter than those without SCD (Jiao et al., 2023). In comparison, the under-5 mortality of individuals with SCD in SSA has been estimated to be ≥ 50% (Green et al., 2022). Factors contributing to mortality include the lack

of early infant NBS programs and failure of early entry into comprehensive care programs (Green et al., 2022).

SCD requires complex management, including regular clinical follow-ups with specialists and strategies to prevent infections, manage complications and provide psychosocial support (Acharya et al., 2023). Medical interventions involve blood transfusions for maintaining haemoglobin levels, controlling acute anaemia and managing pregnancy, surgery and stroke risk. Furthermore, long term blood transfusion therapy is indicated for patients with cerebrovascular disease or are experiencing frequent VOC. In terms of oral medication, hydroxyurea was the first approved treatment in 1998 and to date, the gold standard of SCD treatment, shown to increase levels of HbF. Between 2017 and 2021, three additional drugs were approved, L-glutamine (reduces sickling), Crizanlizumab (reduces VOC) and Voxelotor (increases oxygen affinity). Lifetime standard care costs from the age of 24 years is $1.2 million in the USA. In SSA the majority of individuals cannot afford medication to manage the condition (Acharya et al., 2023; Tanhehco, Nathu & Vasovic, 2022).

Currently curative approaches to SCD include a hematopoietic stem cell transplantation (Tanhehco, Nathu & Vasovic, 2022). However, a limited number of patients are able to find a matched donor and complications such as rejection or graft versus host disease are considerable. Autologous hematopoietic stem cell gene therapy showed some promise in clinical trials and was mentioned to be successful in a single SCA patient. Although initial results were encouraging, comprehensive data is needed regarding the long-term success of this approach. Lastly, gene therapies to modify the dysfunctional gene through gene editing either by preventing sickling or inducing HbF are being explored (Maitta, Reeves & Fontaine, 2023; Tanhehco, Nathu & Vasovic, 2022).

1.2.5. Sickle Cell Patients' Emergency Centre Experiences

Patients affected by SCD can experience multiple life-threatening complications, resulting in the need for frequent emergency care. Western studies have reported poorer experiences by individuals with SCD when seeking EC care, compared to other chronic conditions. Disparities in empathy, courtesy, information giving as well as the quality of pain management delivered were noted (Lattimer et al., 2010). However, in Africa, research measuring patients' experiences in ECs is limited (Oyegbile & Brysiewicz, 2020).

Some of the clinical presentations requiring acute intervention are; VOC, acute flares of chronic pain, neuropathic pain, acute chest syndrome, splenic sequestration, aplastic crises, fever and infection, pulmonary hypertension or embolism and priapism in males. Data collected in the USA between 1999 and 2007 list the reasons for individuals with SCD seeking

emergency care as follows; 5% of patients mentioned breathing problems, 11% chest pain and 6% fever. Unspecified pain is the most common reason for seeking emergency care, accounting for 67% of patient's visits to the EC which correlated with a study done in Tanzania, where pain was also the most common complaint from patients presenting to the EC (Sawe et al., 2018; Yusuf et al., 2010). The various presentations of SCD increases the risk of misdiagnosis and mismanagement of symptoms (Lovett, Sule & Lopez, 2014; Williams & Thein, 2018). ECs are important in providing acute urgent care and are one of the major modes of healthcare utilization in patients with SCD (Yusuf et al., 2010).

Historically, there have been concerns raised about narcotic abuse in patients with SCD; however, literature has proved that the rate of narcotic dependency is low in patients with SCD (Manci et al., 2003; Todd et al., 2006). Management guidelines emphasize early intervention and aggressive pain management to control SCD and for healthcare providers (HCPs) to differentiate between SCD and other painful conditions when treating patients in emergency settings (Tanabe et al., 2021).

There are US studies demonstrating negative attitudes by HCPs toward individuals with SCD creating barriers in the management of SCD (Haywood et al., 2014). In a study evaluating emergency physician's attitudes towards SCD patients' pain levels, 46% of emergency physicians were reported to believe that more than 10% of individuals with SCD have an opioid addiction (Glassberg, 2017). In addition, a study done in 2014 reported nurses' beliefs were significantly more negative than physicians exhibiting greater levels of frustration when caring for individuals with SCD and viewing these patients as addicted to opioids (Freiermuth et al., 2014).

Patients in the US have reported that they delay seeking care from the EC even during times of necessity due to previous negative experiences. Factors relating to negative experiences included facing stigma due to their disease, low healthcare satisfaction and a lack of belief about the pain they are in by HCPs (Abdallah et al., 2020). Furthermore, care in the EC have been reported by individuals with SCD to be in greatest need of improvement in the US (Glassberg, 2017). EC experiences, presentations, and complications of SCD are well studied in developed countries, however, there is limited information reporting on these topics in Africa (Oyegbile & Brysiewicz, 2020; Sawe et al., 2018).

1.2.6. Genetic Services and the Significance of Genetic Counselling in South Africa

Genetic services in SA remain limited even though research and new developments in Africa are expanding (Malherbe et al., 2017). Limited genetic services are available in 4 out of 9 provinces, thus resulting in a majority of the population not having access to genetic services (Kromberg, Sizer & Christianson, 2013). In provinces where genetic services are available, accessibility is still a barrier and rural outreach clinics are needed to overcome this challenge (Malherbe et al., 2017). There are only 8 genetic counsellors (GCs) practicing in the state sector, 1 per 7 million which falls under the national recommendations of 1 per 580 000 (Malherbe et al., 2017).

GCs play an important role in assisting affected or at-risk individuals in understanding medical information related to a disease condition (Kromberg, Wessels & Krause, 2013). GCs can better understand how patients perceive and understand medical information, enhancing communication strategies that are clearer and more empathetic. Furthermore, they serve to assist individuals and families in understanding the medical information related to the genetic condition, the course and management of the condition as well as offer psychosocial support (Aneke & Okocha, 2016).

1.3. Rationale, Aims and Objectives

1.3.1. Rationale

SCD contributes to a high health burden globally and in SSA where it is associated with high morbidity and mortality rates. Organizations such as WHO, listed SCD as a global health issue and a public health priority in 2006 (Thein & Thein, 2016). In 2022, WHO Africa, launched a campaign to increase advocacy on SCD and called upon health institutions to form stronger health systems that ensure quality, uninterrupted, sustainable and accessible services to individuals with SCD (Zapfel et al., 2023). Patient experiences within a healthcare service describe how they interact and perceive the care they receive (Oyegbile & Brysiewicz, 2020). Investigating these experiences can be a means of ensuring the patient's voice is heard as well as enabling patients to be knowledgeable about their healthcare, which promotes increased adherence to treatment (Koch, 1998). Gaining insight into SCD patient experiences when presenting to ECs in SA, can assist in identifying possible strengths and shortfalls in emergency care. Enhancing patient experiences in ECs can not only add to improved clinical outcomes but also improve their satisfaction, reduce patient dropout and the risk of patients seeking alternative therapies (Oyegbile & Brysiewicz, 2020).

This study aligns with global health imperatives and was carried out by a genetic counselling student and supervised by a GC, medical geneticist and clinical haematologist. In SCD, genetic counselling can help adults better navigate the stigma and misconceptions that they encounter about the aetiology of the condition as well as educate patients about the treatment options available (Headings & Fielding, 1975). In addition, the results of this research could potentially provide GCs with a better understanding of the challenges individuals with SCD face when seeking emergency care. This knowledge can allow genetic counselling services to be tailored towards preparing individuals with SCD on what to expect when presenting to the EC, education related to the internal functioning of ECs, effective coping strategies for patients and their caregivers and offer psychosocial support.

1.3.2. Aims

To investigate the perceived experiences of individuals with SCD who present for care to hospital ECs in South African hospitals.

1.3.3. Objectives

- To determine their perceptions of care received in the EC
- To identify common barriers faced by participants that can facilitate the understanding of acute care needs of patients with SCD
- To explore possible suggestions that individuals with SCD have regarding aspects that can improve their experience in the EC

Chapter Two: Methodology

2.1. Chapter Introduction

This chapter discusses the framework underpinning the methodology of this study. The focus will centre on the rationale and suitability of using a mixed-method research (MMR) approach detailing the interplay between the quantitative and qualitative sections. It will expand upon the recruitment process, data collection and analysis. Furthermore, the ethical considerations through which the research was conducted will be discussed concluding with the reliability and validity of the study.

2.2. Research Design

2.2.1. Rationale for Employing Mixed Methods Research

"*Good medical research recognizes the complementarity and interpretation of quantitative and qualitative methods of inquiry*" (Holman, 1993:34). A MMR design is defined as a procedure for collecting and analysing both quantitative and qualitative research in one study. This methodology dates back to the late 1980s and is reported to be advantageous when studying a combination of perspectives found in managing a chronic illness. Integrating both quantitative and qualitative data is thought to provide greater insights than incorporating one method alone (Creswell & Creswell, 2017).

This approach is suited to answering the research question as it allows for multiple viewpoints to be considered as well as an exploration of the medical, psychosocial and psychological facets that contribute to patient experiences. Quantitative surveys provide statistical insights and identify trends in the data (Östlund et al., 2011), such as the frequency of emergency visits and the common challenges faced by patients. While qualitative methods, such as open-ended interview questions, capture the personal narratives of patients, revealing their emotional and psychological experiences during emergency visits. By combining patient stories with quantitative evidence, we can tailor support services that are both evidence-based and patient-centered, addressing specific needs identified through the data. In the context of this study, this method provides a comprehensive and balanced understanding of the research problem by obtaining detailed information, comparing different perspectives and allowing an explanation of quantitative results through qualitative investigation (Östlund et al., 2011).

2.2.2. Type of Mixed Methods Study Design

An explanatory sequential design was used involving a two-phase data collection procedure. In the first phase, the quantitative data was used to generate a broad overview of patient experiences and identify key areas of interest that could be expanded upon during interviews. The quantitative interviews included the following topics; demographic details, healthcare access, frequency of EC visits, experiences with provider knowledge, treatment and quality. In the second phase, results from the first phase were expanded upon via the qualitative interviews. Responses that were common amongst survey participants were investigated in the interview to understand why participants responded the way they did. This design allows for more detailed explanation of the quantitative data through the incorporation of a qualitative approach (Creswell & Creswell, 2017). The results from this sequential exploration method will be presented in chapter three with sociodemographic data displayed quantitatively, followed by the integration of quantitative and qualitative findings, thematically capturing patient experiences and discussing them in the context of relevant literature.

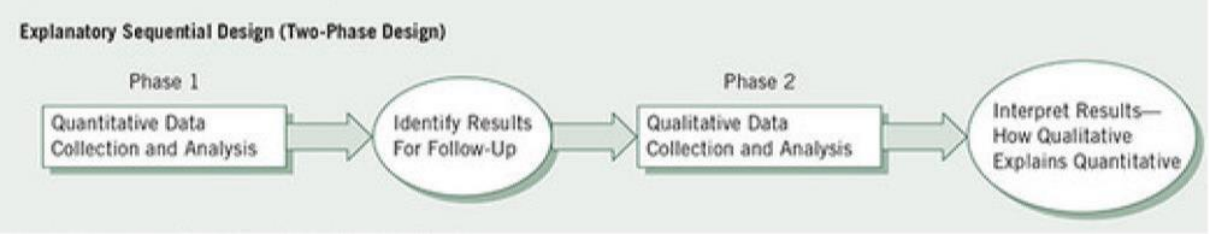

Figure 2.1: A Diagrammatic Representation of the Core Design Method to be Used (Cresswell & Cresswell, 2018)

2.3. Study Site, Population and Sample Size

This study aimed at recruiting 50 participants for the quantitative section and estimated 8 participants in the qualitative section or until data saturation was achieved. Gathering a larger sample size for quantitative research is needed to infer meaningful results, literature advises on sample sizes of 10-30 for descriptive studies and a minimum sample size of 50 for comparative studies (Creswell & Creswell, 2017). Smaller sample sizes are used for qualitative data, literature suggests a sample size of between 5 to 10 participants to obtain more extensive information (Creswell & Creswell, 2017). The discrepancy in sample sizes will not invalidate the study as each method serves a different purpose i.e. to allow generalizations to be made for the population and to gain further insights (Creswell & Creswell, 2017).

A multistage procedure was used for the sample selection. Multistage sampling is a means of data collection suitable when the researcher aims to have a geographically spread-out sample (Creswell & Creswell, 2017). Two recruitment platforms were utilized simultaneously, the Sickle Cell SA (SCSA) online support group, one support group body that has multiple social media pages and a Haematology unit in the Western Cape (WC) province of SA to increase the reach of participants and ensure our quantitative sample size was met. Figure 2.2 summarizes the recruitment process.

Snowball sampling was an added measure used to ensure an adequate sample size, by requesting initial participants to identify other known participants that would like to participate or may benefit from the study. Snowball sampling is mentioned to be advantageous when no sampling frame is available and as SCD is considered a rare disease in SA, it was therefore beneficial to request participants to recommend other people in their SCD network who would like to contribute to the study (Creswell & Creswell, 2017; Wonkam et al., 2012).

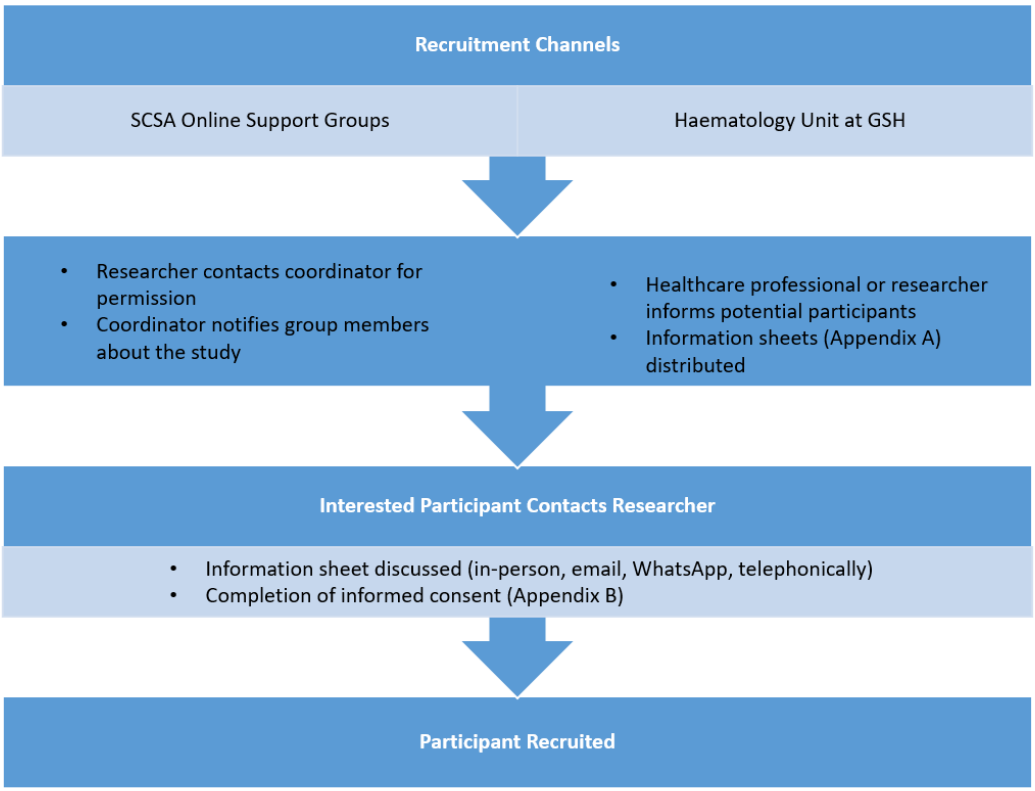

Figure 2.2: Summary of the Recruitment Process

2.3.1. Inclusion and Exclusion Criteria

The inclusion and exclusion criteria are displayed in Table 2.1. The primary sampling criteria was individuals in SA living with SCD >18 years. An adult is anyone 18 years or older who is legally allowed to make independent decisions in SA (Legal Aid, 2018). A second sampling criterion was adult patients with SCD who presented to the EC seeking care in the last five years i.e. 2017-2023, as patients may struggle to recall their experiences prior to five years ago.

English-speaking participants were chosen, despite most of the patients affected with SCD in SA coming from countries where French is their main language (Pule et al., 2017). As French is a non-official South African language, translational support was considered a potential challenge and in addition, these individuals receive their healthcare correspondence in English in SA. If individuals could speak English but were not literate in the language, provision for the written survey to be read out by the researcher was arranged. Individuals with cognitive impairment have been excluded from this study due to their vulnerability to coercion and may lack the mental capacity to provide informed consent (Oruche, 2009).

2.4. Research Instrumentation

The main research instrument was a structured questionnaire. Structured questionnaires have the advantage of generating a larger number of responses while remaining cost and time effective (Creswell & Creswell, 2017). A limitation of surveys is that not all factors influencing the answer may be known for each individual. This study attempted to balance this limitation by incorporating a comment section after each close ended question and inviting participants to take part in an open-ended interview, conducted via a semi-structured interview guide. The content of the survey (Appendix C) and open-ended interviews (Appendix D) was adapted from previous studies by Freiermuth et al*.,* (2014), Crego et al*.,* (2021) and SA triage guidelines by Cheema & Twomey (2012).

Table 2.1: Inclusion and Exclusion Criteria used in the Study

Inclusion Criteria	Exclusion Criteria
Adult patients with SCD	Individuals with cognitive impairment
Patients who sought care from hospital EC in SA in the past five years	Individuals not proficient in speaking English
English speaking individuals	

2.4.1. Pilot Study

A pilot of the survey was sent out to five staff members at the UCT Division of Human Genetics and two pilot interviews were conducted. Feedback pertaining to typesetting errors, grouping of questions pertaining to the same topic and rephrasing certain survey questions for clarity purposes was actioned. None of the data generated from the pilot study was included in the analysis.

2.4.2. Research Survey

A 30-minute electronic survey (Appendix C) was created on Google forms which also allowed it to be printed for hardcopy completion. The researcher made provisions to allow in-person, written or telephonic completion for participants who had no internet access and were attending hospitals in the same geographical location as the researcher (in CT) by meeting them in person on the same day of their clinic appointment. At the end of the survey, there was a prompt, requesting whether participants would like to share more information on their experience in the EC. Participants who ticked *"yes"* were interviewed in-depth.

2.4.3. Research Interview

Open-ended interviews (Appendix D) were conducted by the researcher to elaborate on participants' experiences in the EC. These interviews were conducted either in person at the GSH hospital or through Microsoft Teams. The interviews were approximately 45 minutes and recorded to allow the researcher to transcribe the interview. This method allowed for the flexibility of responses in comparison to responses being standardised in the questionnaire.

The researcher was able to adjust the questions based on the participant's responses and note down experiences important to them.

2.5. Ethical Considerations

This research study was guided by the principles outlined in the declaration of Helsinki, maintaining respect towards the research participants, maintaining their rights to autonomy and their rights to informed consent. The participant's well-being as well as ethical considerations took priority over the interests of science and society (World Medical Association, 2014).

2.5.1. Ethics Approval

Ethics approval was secured from the University of Cape Town (UCT) Human Research Ethics Committee (742/2022, Appendix F) prior to commencing data collection. Participants were first provided with an information sheet and consent form before partaking in the study. Participants were assured of data anonymity, provided the opportunity to clarify any points of concern and given the choice to withdraw at any time.

In addition, hospital approval was formally requested from three hospitals affiliated to UCT: Groote Schuur Hospital (GSH), New Somerset Hospital and Victoria Hospital, through the National Department of Health website. Approval was granted from GSH (Appendix F), while no responses were received from the remaining medical facilities, therefore, the latter two hospitals were excluded from the research process.

2.5.2. Data Safety and Monitoring

Survey data generated from Google Forms were anonymous unless participants opted to provide their name and contact number for the research interviews. In that case, the researcher and supervisors were the only parties with access to the data. Once the interviews were conducted and transcribed, the names were deleted. Each transcript had a number assigned to it and all personal identifiable information was removed. All data was stored on a password protected laptop for the duration of the research project and will be disposed of upon conclusion of the research study.

2.5.3. Risks and Benefits to Participants

There were no foreseeable risks for the participants partaking in this research study. If at any time the questions asked evoked an emotionally charged response, participants had the choice not to answer. Furthermore, participants had the option of being referred to a counsellor.

A potential benefit of this research study included gaining insight into SCD patients' perceived experiences in ECs in SA. The output of this research may help patient stories to be heard; this has been seen to be therapeutic to patient groups (Koch, 1998) and improve patient satisfaction.

2.6. Data Collection

All participants included in this study had adequate Internet access and chose to participate using the online form. Participant responses were automatically captured from Google forms to an online spreadsheet. Fifty-one (n=51) responses were obtained from the 65-survey links distributed to participants: 26 males and 39 females. One male participant passed away from SCD related complications before completing the survey. A once-off reminder was sent out to participants to complete the survey, participants who didn't follow through were not questioned. Participants could only submit a response once. The survey did not collect data from participants who dropped out mid-completion. Participants' gender, age, geographical location within SA, country of origin and whether they immigrated to SA during their childhood or adulthood were among the demographic data collected. This data enabled the researcher to identify trends and draw comparisons regarding the factors that contribute to how adult patients with SCD experience the ECs in SA. The researcher stopped recruiting participants once the desired number of surveys were completed.

From the 51 participants who completed the survey, 19 participants (three males and 16 females) were willing to be interviewed. Two participants ticked "*yes*" to share more information but entered "*no*" when they needed to fill out their names and contact numbers. The researcher interviewed participants at random until data saturation was achieved. The cohort consisted of 8 participants and included 1 male and 7 females.

2.7. Data Analysis

2.7.1. Quantitative Analysis

Survey data was coded and analysed using SPSS® software which was freely available from the researchers' institution. Descriptive statistics were used to summarize the data numerically allowing graphical representations to be made. Comparison analysis involves examining similarities and differences between data sets in order to gain insights into relationships and patterns in the data. This analytical approach was utilized to identify similarities and differences among participant responses. The conventional statistical significance level of 0.05 was employed throughout. Fisher's exact test was used to determine whether participant satisfaction levels differed between healthcare sectors. Fisher's test was chosen due to the small sample sizes involved and the lower counts seen (<10) in the categorical variables compared in this study. Fisher's exact test was also used to compare referrals to genetic counselling services between the WC and Gauteng province, as the data could be categorized in a 2x2 contingency table. A T-Test was used when comparing responses of participants from two different South African provinces, Gauteng and WC to the different healthcare sectors. These two provinces were chosen for comparison as they consisted of the highest number of participants (n=18 and n=25 respectively). This study compared means derived from survey responses quantified on a Likert scale, rather than the distribution of categorical data. Survey responses following a Likert scale were coded to allow for comparison of average numerical values. This approach assumes that the Likert scale data, although ordinal, can be treated as continuous for the purposes of the T-test. This assumption is common in social sciences, but it is acknowledged that it introduces some limitations. Normality of the data distribution was assessed using normality tests (Field, 2013; Pallant, 2020). From the quantitative data, specific aspects of participant responses that warranted further investigation through qualitative exploration were identified.

The responses from the close-ended and open-ended questions in the survey were analysed as part of the qualitative analysis to identify the underlying theme they represent. This approach provided the researcher with an understanding of the underlying factors underpinning the statistical observations and contributed to gaining comprehensive insights.

2.7.2. Qualitative Analysis

Interviews conducted with each participant were audio recorded and manually transcribed. Participant identities were only known by the researcher and kept anonymous on the

recordings and transcriptions. Survey data was organized into open-ended responses and closed-ended responses, the open-ended questions were coded and with the transcript data, separated into different themes using thematic analysis. The close-ended questions were analysed using descriptive statistics to identify the key areas that were qualitatively explored. Throughout this research, discussions were held with the researcher and her supervisors to establish consensus on interpreting themes derived from the qualitative data (Östlund et al., 2011). The researcher followed a stepwise approach summarised in figure 2.3. During the data familiarization stage, the researcher initially familiarized herself with the data by reading through all the survey responses multiple times to get a sense of the overall content. The code generation phase involved breaking down the data into discrete parts and coding these parts with labels that describe their content. These codes were discussed with the research supervisors to allow for transparency and the refining of codes. This method of analysis allowed the data obtained to be organised into meaningful sets of categories (Creswell & Creswell, 2017).

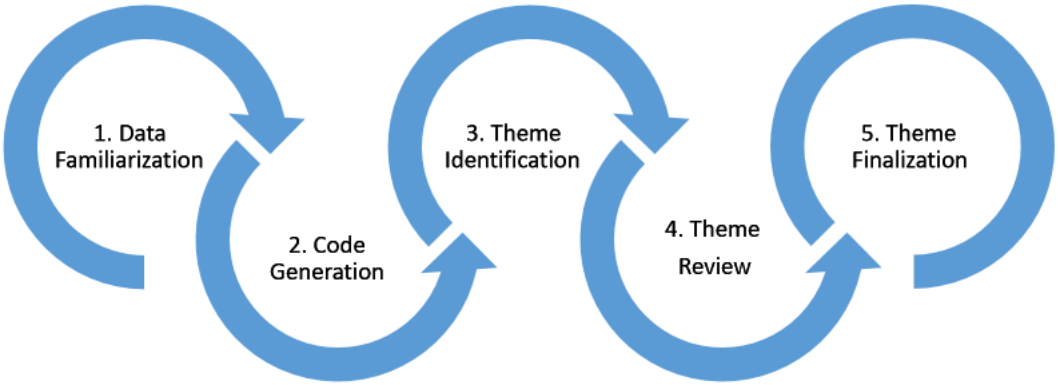

Figure 2.3: Summary of the Stepwise Approach to Thematic Analysis

2.8. Reliability and Validity

Reliability refers to the consistency and reproducibility of the research study while validity refers to accurately measuring the intended aspects of the study while drawing meaningful conclusions (Creswell & Creswell, 2017). In this study, initial measures to strengthen the validity and reliability of the study were taken by accurately documenting data collection and analysis processes, ensuring accountability and reproducibility. In addition, triangulation was considered an important factor; participants were recruited from multiple sources making the findings of the study more broadly applicable.

The practice of reflexivity was incorporated through various stages of the research involving the researcher and her supervisors. Reflexivity is defined as a process where researchers examine their own perspectives, biases, assumptions, and potential influence on the research (Creswell & Creswell, 2017). This integration increased the researcher's self-awareness, managed researcher bias and contributed to inter-rater reliability.

Chapter Three: Results and Discussion

3.1. Chapter Introduction

This chapter presents the results of a sequential exploration method with the sociodemographic data displayed quantitatively, presenting the demographic information in separate sections provides context of the study population and allows for discussion of the demographic data observed. The contributing findings from both sections will be shown thematically, using data triangulation and weaving of the quantitative and qualitative sections, holistically capturing the patient experiences within the EC and discussed against the relevant findings from literature.

The quantitative component offers statistical insights into the trends in patient demographics, preferences, quality of care, pain management and waiting times. While the majority of quantitative data was analysed statistically, participants had the option of commenting after each question. These comments will also be shared as quotes supplementing the quantitative findings. The qualitative component captures patient narratives, sharing the emotions and contextual factors that shape patients perceptions.

3.2. Sample Demographics

3.2.1. Participants' Gender and Age

Out of the 51 responses received, 72.5% were female and 27.5% were male. The higher number of female participants is in keeping with other studies (Cull et al., 2005). This tendency is attributed to females being more inclined to social communication and expression (Cull et al., 2005), possibly explaining the higher female participation in the survey interviews.

The average age of participants was 31.6 years (range 18-45 years, SD = 6.6). Figure 3.1 displays the age of individuals who participated in this study.

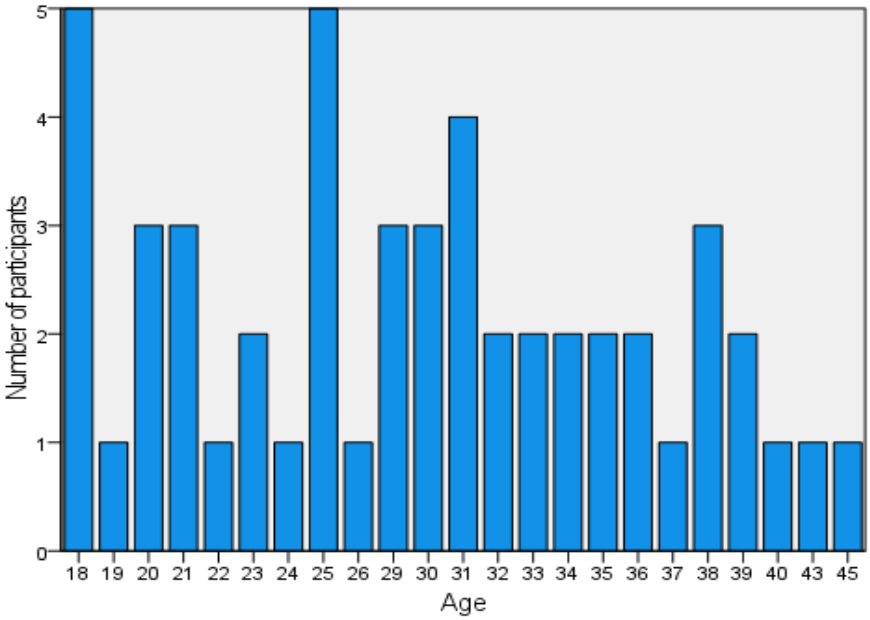

Figure 3.1: Age of Participants in this Study

3.2.2. Participants' Country of Origin

More than half of the participants (52.9%) were not originally from SA with 47.1% immigrating during childhood and 25.5% in their adulthood. This data was consistent with the observed increase in individuals with SCD being seen in SA (Wonkam et al., 2012). Figure 3.2 depicts the number of participants and their respective countries of origin with 37.3% of participants immigrating from the Democratic Republic of Congo (DRC), followed by 7.8% from Nigeria and Zimbabwe, 3.9% from Uganda and Tanzania. One participant (2%) was recorded from each of the following countries: Ghana, Kenya, Malawi, Rwanda, Yemen and Zambia. Only one participant in this study was from outside the African continent.

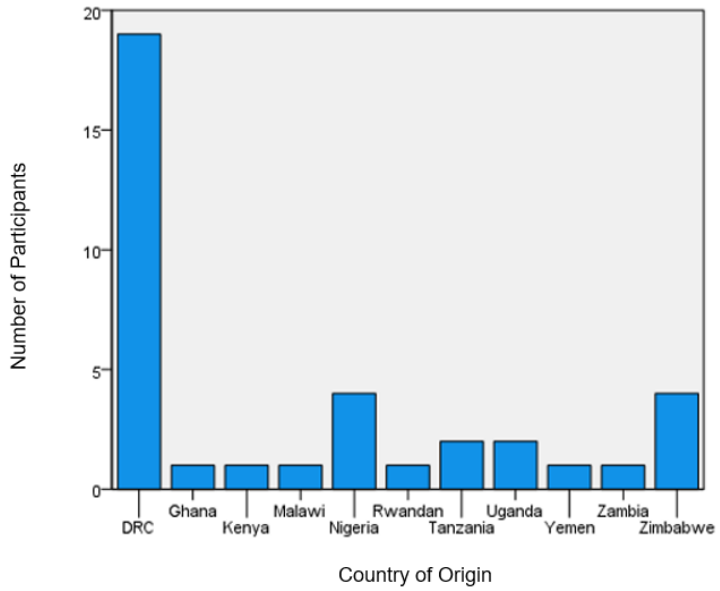

Figure 3.2: Cohorts' Country Representation in Numbers

3.2.3. Provinces Participants Sought Healthcare in

Out of the 51 responses, most of the participants 49% sought healthcare in the WC province of SA, likely due to the founding location of the online SCD support group and the researchers' recruitment efforts at the tertiary healthcare institution in the same region. Gauteng was the second highest (35.3%) province represented, potentially due to the second highest number of members contributing to the online support group. There were 13.7% who sought healthcare in KwaZulu-Natal (KZN), 11.8% in the Eastern Cape and 3.9% in the Northern Cape (NC). A greater number of participants were observed from the provinces (WC, Gauteng and KZN) with established genetic services which refer individuals to the online support group. There were no participants who sought healthcare in 4 out of the 9 provinces. Figure 3.3 graphically represents the number of participants seeking healthcare in the respected provinces.

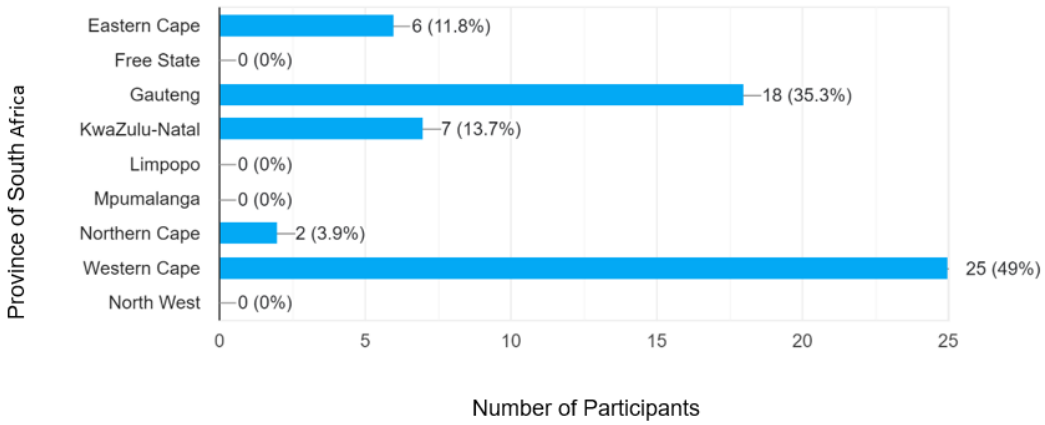

Figure 3.3: Participants' Healthcare Distribution by Province

3.2.4. Healthcare Sector Utilization by Participants

Of the 51 participant responses, 47.1% utilized healthcare in the public sector. While 31.4% reported attending private healthcare facilities and 21.6% made use of both the public and private sector. Figure 3.4. summarizes the healthcare sector utilization per province with participants in the NC using both sectors, one participant from each sector in the Eastern Cape, the private sector being mainly used by participants in KZN and Gauteng while the public sector was mainly utilised by participants in the WC. The slant towards the public sector could also be explained by the founding sector of the online support group and the public institution where the researcher's recruitment efforts where directed.

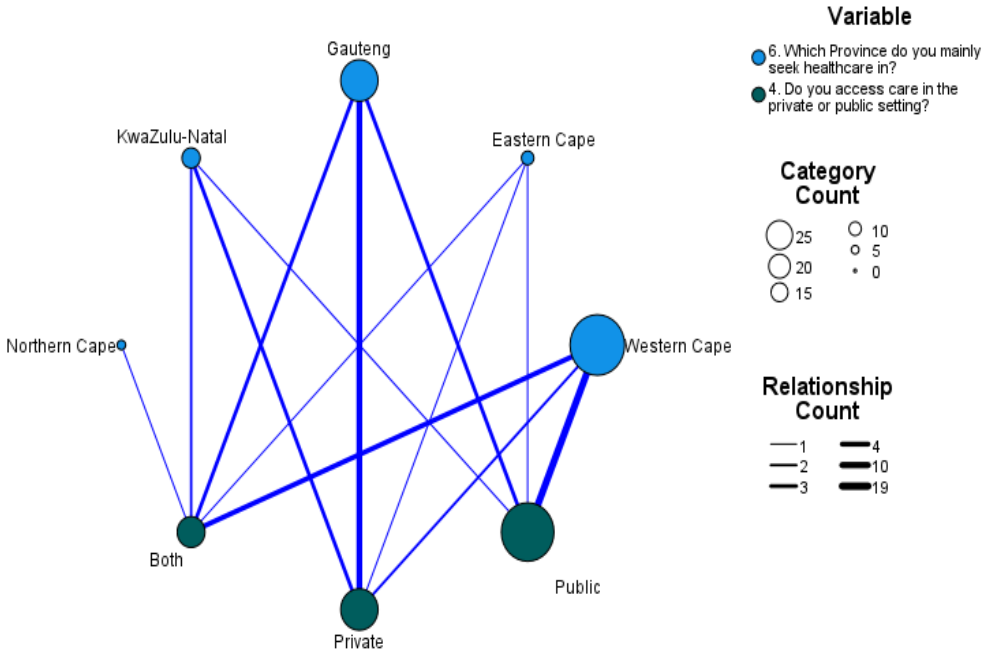

Figure 3.4: Relationship Map: Healthcare Sector Utilization by Province

3.2.5. Frequency of Emergency Centre Visits

From 2017-2022, out of the 51 responses, 47.1% of participants visited the EC between 2-5 times while 29.4% attended more than 5 times. The remainder (23.5%) attended only once. The mean frequency of visits were 2.2 times with a mode of 3 times. In comparison, in the last year, 37.3% of participants attended 2-5 times, 13.7% attended more than 5 times, 21.6% attended once and 27.5% had not attended. The mean was 2.8 while the mode remained at 3.

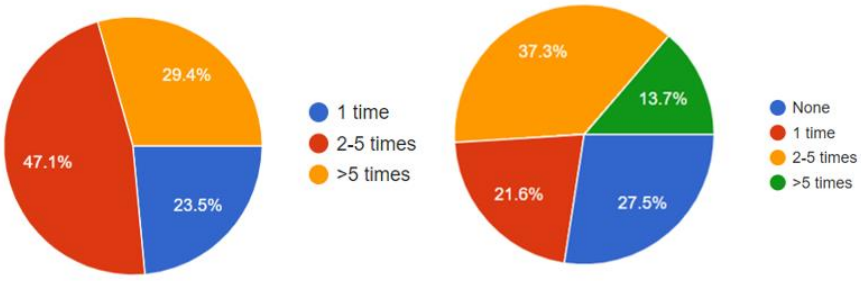

Figure 3.5: Frequency of EC visits in the last 5 years (left) and last year (right)

The annual mean frequency rates of participant visits to the EC aligns with research done in the US and Canada which report the annual mean EC visits for adult patients to be between 2.6 to 14.2 visits per patient (Chan et al., 2020; Loeffler et al., 2015). Interestingly, the frequency of EC visits in the present study didn't reach a high of 14.2 annual visits. This observation could potentially be influenced by factors such as climate conditions favouring the severity of disease, or it may be linked to cultural beliefs and practices where individuals rely on traditional healing methods; future research could investigate the observed differences. Reasons participants delayed seeking emergency care that form part of the themes of this study include: challenges in accessing healthcare, previous negative experiences in the EC and prolonged waiting times which could factor into the lower number observed.

3.3. Assessing Patient Satisfaction in the Emergency Centre

In this study (n=51), 45.1% of respondents described their recent EC visit as *"average"* while 31.4% described it as above average (19.6% *"good"* and 11.8% *"very good").* In comparison, below average comprised of 23.5% of responses (17.6% *"poor"* and 5.9% *"very poor)".*

Participants' experiences upon arrival to the EC was recorded as *"neutral"* in most cases (52.9%). In the waiting area, most (56.9%) stated *"poor"* experiences while in the treatment

room, most responses (43.1%) indicated their experiences as *"good".* These results are summarised in table 3.1 and 3.2.

Table 3.1: Summary of Participant Satisfaction in EC from their Recent Visit

Description of Recent EC Visit	Number of Participants (n=51)	Percentage of Respondents
Very Good	6	11.8
Good	10	19.6
Average	23	45.1
Poor	9	17.6
Very Poor	3	5.9

Table 3.2: Summary of Participant Satisfaction in Different Areas of the EC

Participant's Experience in Different Areas of the EC	Number of Participants (n=51)	Percentage of Respondents
Arrival Experience		
Good	16	31.4
Neutral	15	52.9
Poor	29	15.7
Waiting Area Experience		
Good	7	13.7
Neutral	15	29.4
Poor	29	56.9
Treatment Room Experience		
Good	22	43.1
Neutral	18	35.3
Poor	11	21.6

Participant satisfaction levels during their last EC visit aligned to literature from a US study (Kanter et al., 2020) where the majority (39%) reported their care as *"neutral",* 32% reported

most *"positive experience"* and 29% indicated it as "most negative". The study highlighted that positive EC experiences were higher in younger patients treated in paediatric facilities, patients of our study reflected that they felt their care was better when they were younger. An important differentiating factor of the other study is that a large number of the participants (68%) were on a private medical insurance (Kanter et al., 2020).

No significant association was found between the participants experience at their last visit to the EC and the healthcare sector that was utilized (Fisher, p=0.50). No significant association was found when comparing participant satisfaction levels in the different areas of the EC to healthcare sector utilization either (Fisher, p=0.27). The p-values comparing satisfaction levels upon arrival to the EC, in waiting area and treatment room compared to the health sector utilization was 0.14, 0.47 and 0.53 respectively. These results suggest that factors influencing EC experiences do not show significant correlation with the broader healthcare sector utilization or the variables examined in our study are not strong predictors of the factors representing satisfaction levels. Additional investigations into the potential factors influencing healthcare utilization and satisfaction can be beneficial in understanding real-world relationships.

3.4. Exploring Themes Impacting Patient Experiences in the Emergency Centre

Codes identified from the quantitative and qualitative sections were grouped into 20 categories and 11 sub-themes which converged into 5 main overarching themes displayed in figure 3.6 below. In addition, the themes were often seen to interconnect and reinforce one another.

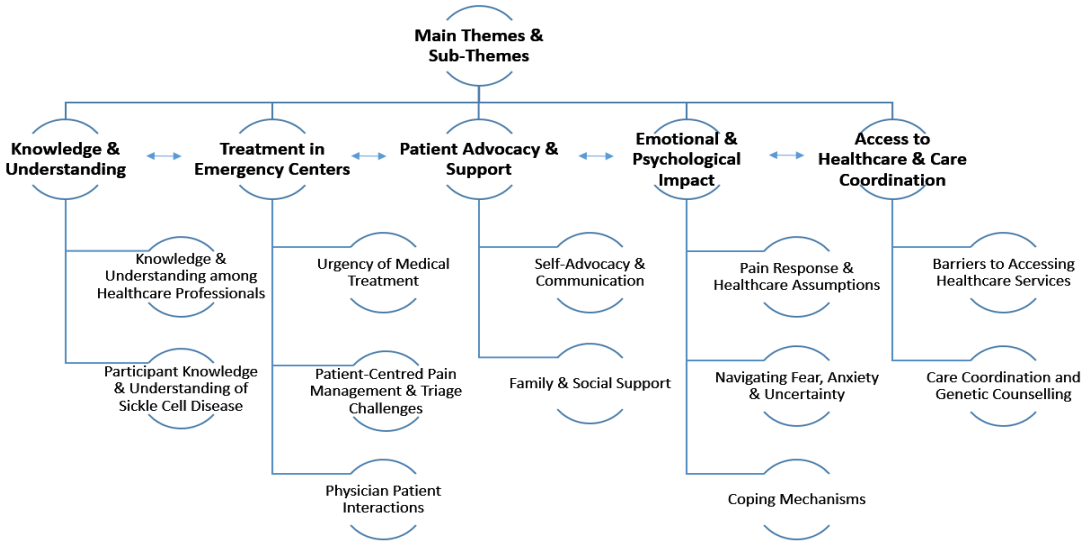

Figure 3.6: Diagrammatic Representation of the Themes and Sub-Themes Identified

3.5. Theme 1: Knowledge and Understanding

Participants identified a lack of knowledge and understanding amongst HCPs surrounding SCD which they believe contributed to delayed care, misunderstanding of the condition, disbelief in their symptoms and ignorance regarding their pain. Furthermore, participants emphasized the importance of comprehending their own condition and being able to articulate the nature of SCD pain which may or may not involve visibility. The influence of knowledge and understanding will be further described under two subthemes.

3.5.1. Subtheme 1: Knowledge and Understanding among Healthcare Providers

In this study, out of 51 participants, 64.7% reported the nearest hospital as their first point of care. Private care was sought in 29.4% of participants while only 5.9% went to their local clinic. These findings differed from literature reporting that most patients with SCD first report to their primary care physician (Mainous et al., 2018; Mehta et al., 2006). The mentioned reasons for attending the hospital and not their closest clinic was related to knowledge and understanding of SCD by HCPs.

"The nearest hospital, it's difficult when the pain is high and you need quick treatment. The clinic would be faster to get to but then they take too long to understand my condition and know how to treat me" ~ P7, survey

"I wish I could go to my nearest doctor for help because emergency visits happen unexpectedly and it would be closer but my pain is dismissed there. So I rather go to the hospital where I know they have the knowledge to manage it" ~ P38, survey

Exploring participants' (n=51) confidence level in the nurse's knowledge that treated them, identified that 39.2% felt *"not confident at all"* that the nurses knew how to treat SCD while 21.6% felt *"very confident"* (figure 3.7).

"*I was disappointed because they were asking ME what the hell SCD was" ~ P11, survey*

"They couldn't even spell it and kept saying its anaemia and anaemia doesn't cause pain like this" ~ P48, survey

Conversely, when asked how confident participants (n=51) were, that the doctor treating them knew how to treat SCD, 33.3% (n=51) of participants were *"very confident"* and only 11.4% felt *"not confident at all"* (figure 3.4). Various cases where nurses consulted the patients'

specialist before doing further examinations and where HCPs knew what SCD is, fostered some confidence in participants.

"I know the nurse was ensuring that she was doing her work and making the necessary arrangements for my treatment, she did her job by first consulting my doctor before doing any examinations. I would say that I was somewhat confident that she knew how to treat SCD" P14, survey

"The doctors can read about my condition from my file and understand, but the nurses look more confused after reading my file" P41, survey

Figure 3.7: Participant Confidence in the Nurse (left) and Doctor (right) Treating Them

A T-Test showed a statistical difference (p=<0.01) between the participants' confidence levels in both nurses and doctors treating them in Gauteng (n=18) and WC (n=25) Provinces. The T-Test compared the average confidence levels (quantified from Likert scale responses) between the two provinces. For nurses, the average confidence level in WC was higher, with 28% of individuals selecting *"very confident".* In contrast, 24% of participants selected *"neither confident nor unconfident"* and another 24% selected *"not confident at all".* In Gauteng, 50% of participants selected *"not confident at all"* regarding the nurses treating them. Responses indicating *"somewhat confident"* and *"somewhat unconfident"* were in the minority for both provinces. Regional variations in healthcare infrastructure, resources, geographical accessibility, and HCP training in SCD can contribute to the differences observed between provinces.

Participant confidence levels in nurses or doctors treating them were analysed separately for the private (n=10) and public sectors (n=18) to compare the mean confidence levels between the healthcare sectors. The results showed no significant difference between the private and public sector with p-values from the T-Test of 0.32 and 0.25 respectively. Notably, 55.6% of Gauteng participants sought private healthcare while 72% from the WC sought public

healthcare. This data suggests comparable confidence levels irrespective of the healthcare sector. Healthcare sector preferences of participants could potentially be due to other factors such as accessibility or perceived quality of care. The majority of participants expressed *"not confident at all"* in the nurses treating them, both in the private (37.5%) and public sectors (63.6%). Among those who used both sectors (n=11), 63.6% selected *"not confident at all"*. These findings highlight a potential area of improvement in confidence levels in nurses across healthcare sectors.

Confidence levels in the doctor treating them was selected as *"very confident"* in the majority of participants in both healthcare sectors (50% in private and 43.5% in public). Majority (45.5%) of participants who made use of both sectors selected *"neither confident nor unconfident"* as their response. The positive perception of doctors' treatment irrespective of the healthcare sector can be influenced by perceived levels of understanding of SCD by doctors or doctors' communication skills. Factors like personal preferences and past experiences can contribute to the nuance perspective of participants.

When asked whether participants (n=51) ever left the EC to seek alternative help, 52.9% participants responded *"yes".* A breakdown of their responses from a multi-response format revealed that multiple factors influenced their decision-making process. Majority (75%) of participants selected *"lack of confidence in the HCPs' knowledge"*. An exploration of the comments left by these participants suggest they feel anxious and unsafe when the nurses lack knowledge and understanding about SCD causing them to leave and seek care elsewhere.

"When the nurses don't know what my condition is, I feel very scared to stay there, so I go to another one even if it's further away from home" ~P13, survey

"The nurses don't know what my condition is, then I have to travel further to go to another hospital" ~ P34, survey

"The nurse asked me to spell my condition out to her while I was in crisis and then told me, oh it's just anaemia, so I left and went to a different EC" ~P37, survey

Other contributing factors that led participants to leave the EC and seek alternative help were; "*prolonged waiting times to see the doctor*" selected in 50% of responses, 33% indicated "*prolonged waiting time to see the nurse"*, *"lack of HCPs empathy"* accounted for 25% of selected responses while *"poor communication and language barriers"* contributed 12.5% and 8.3% respectively. The option of *"other"* was selected by two participants, however upon analysis the responses linked to *"lack of HCPs knowledge".*

These findings reveal the complex factors influencing participants' decision to seek alternative help. The predominant concern being HCP knowledge of SCD which aligns with previous literature identifying HCP knowledge of SCD as the main barrier towards treating patients with SCD (Kaur et al., 2020). Reasons provided from the literature list the availability of a limited number of haematologists that specialize in SCD care as well as the presence of very few comprehensive multidisciplinary clinical centres, notably a challenge affecting SA as well (Kaur et al., 2020, Malherbe et al., 2017; Wonkam et al., 2012).

From the interview data, knowledge and understanding among HCPs was further expressed by participants as they emphasized the unique challenges and pain associated with SCD which they feel some doctors lack awareness and understanding of. They believe that the lack of knowledge and understanding leads to doctors questioning the validity and extent of their symptoms leading to the inability to treat it urgently and appropriately.

"The doctors only open my file once they go and check their textbooks, because you first have to tell them I have SCD then they go and check their books, they read it, then they look at you and then they'll ask you what treatment do you want. They go back to their books and they'll be like, no, let's put less" ~ P6, interview

Participants also spoke about nurses who lack understanding of SCD and the communication challenges that arise from the unfamiliarity of the condition. In some instances, participants felt that doctors' are knowledgeable about the condition and therefore believe when they are in pain compared to nurses who question them and lack awareness.

"The nurses don't understand what SCD is, when I tell them it's SCD, they like what is that when you explain to them, then they more confused but the doctors they're okay, because they know these things, they really help me a lot and explain to me the things very clearly when I don't understand" P3, interview

In addition, participants described their experience with paramedics when needing emergency care.

"Maybe the paramedics, if you can also help tell them what SCD is. When I tried calling them, I had to explain for an hour what it SCD is. You can't get ambulances quickly because you have to explain and it's really hard. So many times I had to end up cancelling and taking public transport and it's really hard for me in pain" ~P4, interview

These findings align to studies by Crego et al., (2021) where individuals with SCD identified SCD knowledge gaps among EC providers as a barrier to medical treatment. However, the literature didn't differentiate on patient perspectives of knowledge gaps between doctors and nurses. Our study identified that, at times, patients feel more confident in the doctors treating them compared to nurses due to their experiences where certain doctors understood what SCD is.

A number of studies, both in the US and Ghana, have investigated gaps in HCP knowledge and understanding regarding SCD, resulting in the poor health outcomes of individuals with SCD (Dennis-Antwi & Ohene-Frempong, 2008; Haywood et al., 2014; Kaur et al., 2020). Furthermore Glassberg (2017) identified that EC providers consistently went against the recommended evidence-based guidelines while treating SCD due to poor understanding of SCD. However, these studies did not distinguish between the knowledge gaps between nurses and doctors. Addressing the knowledge gaps, improving communication and minimizing waiting times could potentially enhance the EC experience.

3.5.2. Subtheme 2: Participant Knowledge and Understanding of Sickle Cell Disease

The survey further revealed that out of the 51 participants, 31.4% felt they had an excellent understanding of SCD, 35.3% indicated that although they have a good understanding, they still have questions and 29.4% felt that they knew enough to manage their condition. The percentage of participants with questions suggest opportunities for additional education on certain aspects of SCD.

> *"I would like to learn more, just with less medical jargon in the way" ~P32, survey*

Out of 51 participants, majority (56.9%) remember receiving information about their condition, 29.4% have never received information and 13.7% indicated they don't remember. When asked who gave them the information, 58.8% indicated their *"doctor"*, 3.9% *"a nurse"*, 7.8% indicated *"both a nurse and doctor"* while 29.4% selected *"other".* Other sources that were listed were, their parents (14%), the genetics team (1%) self-searching through books and internet (7.2%) and through the support group (7.2%). Although the primary sources were HCPs, the diversity of information sources highlights the many ways individuals access knowledge. In addition, the inclusion of support groups as a source emphasizes the importance of peer support and shared experiences in understanding their condition. Some participants indicated they don't remember receiving information which raises awareness on the need for clear, memorable and repetitive educational interventions.

When comparing the responses from participants in Gauteng (n=18) to the WC (n=25) regarding how they would describe their level of knowledge about SCD, a T-Test demonstrated there was a significant difference between the two groups (p=0.01). Most participants from WC (40%) responded *"excellent"* while only 20% responded *"fair".* Whereas half of the participants from Gauteng (50%) responded "*fair"* and only 16.7 responded *"excellent".* A minority of responses from both provinces selected *"poor".* Variations in healthcare infrastructure, access to educational resources, or cultural attitudes toward seeking information about SCD might influence participants' perceptions of their knowledge. This data identifies the need for targeted, accessible educational activities in specific regions to address gaps in understanding.

In response to whether participants (n=51) received an explanation of what to expect before going to the EC, 72.5% hadn't received an explanation and 27.5% of participants responded that they knew what to expect. The majority of respondents (80.4%) selected that they would like to know how the EC works as it may better their experience. This observation did not differ significantly across provinces or different health sectors. The p-value from the T-Test was p=0.23 and p=0.27 respectively. A consistent desire for information was identified among participants regardless of their geographical location or healthcare sector utilization, believing that more information could enhance the overall EC experience.

From the interviews, participants felt their own knowledge and understanding of the condition impacts their experiences in the EC. They find themselves in a unique position compared to other patients seeking care in the EC as their lived experiences and personal knowledge of their treatment becomes an important factor in the care they receive.

"It takes special people to be able to do this work but healthcare workers and patients must listen to each other and communicate because yes, the textbooks can teach, but we here living everyday with this disease we know from experience" ~P7, interview

Participants emphasized the importance of having knowledge and understanding about their own condition, which increased their awareness of symptoms and facilitated communication with HCPs, resulting in prompt treatment in the EC. They felt they could be proactive in advocating for themselves when visiting the EC as they are more aware of their symptoms, understood when they needed urgent care and were better equipped to communicate their needs to the HCP.

"I have to manage my condition everyday so I educated myself and I learnt from experience what works for me. Over time I learnt to communicate to the HCPs treating me, telling them the medication I need" ~P3, interview

However, some participants felt that they lacked knowledge and understanding of their own condition. They expressed their desire to know more about the condition, how their symptoms should be handled in such a setting and what their patient rights are in the EC.

"I still struggle with this disease, yho, I have been living with it for years but I don't understand. When the doctors say it's because of the SCD, I get so confused I don't understand how" ~P2, interview

"(Laughs), I would like to know more, but who would have the time to explain to me? but with the times I been now I already know with experience" ~ P4, interview

The importance of knowledge acquisition wasn't unique to our study. Patients in European countries have described a need for increasing their knowledge to allow them better control of their illness and negotiate for the hospital care they need (Asnani et al., 2017). The study found that patients with higher knowledge scores made more sense of their condition and perceived greater personal and treatment control (Asnani et al., 2017).

However, there was limited literature related to disease specific knowledge of African SCD patients. Considering participants' request for disease specific knowledge with less medical jargon and the pivotal role of knowledge in positive patient experiences, the incorporation of GC's into the healthcare team of patients with SCD can be a valuable resource, providing specialized, targeted information and support, this aspect will further be discussed in section 3.9.2. Care Coordination and Genetic Counselling.

3.6. Theme 2: Treatment in the Emergency Centre

Drawing from patient accounts, the theme of "Treatment in the emergency centre" will be explored. This theme was divided into the sub-themes of "Treatment Urgency", "Patient-Centred Pain Management and Triage Challenges" and "Physician-Patient Interactions". Among the factors participants (n=51) considered important in the EC, *"relief of symptoms"* was ranked the highest (56.9%) highlighting the importance patients place on effective symptom management, followed by "*efficacy*" (29.4%) addressing the specific needs of

patients in the EC. Smaller percentages were seen in *"care and compassion"* (3.9%), *"dignity and respect"* (3.9%) and lastly, *"information and communication"* (3.9%) reflecting the broader dimensions of patient care that can contribute to positive experiences. Participants (2%) who selected *"other"* stated that all these factors are important indicating the holistic aspect of patient care.

3.6.1. Subtheme 1: Urgency of Medical Treatment

Participants expressed their desire to move from public hospital care into private where they could experience quicker medical treatment because the lack of urgency that their crisis is treated with in the EC is the biggest challenge. From the results of a multi-response question enquiring how participants felt before visiting the EC, those that answered *"negative"* selected *"worried they will wait too long"* in 55.2% of cases. The 20.7% that selected *"other"* noted being called names (n=3) and anxiety from previous experiences in the EC (n=3). While *"positive"* feelings were associated with *"confident receiving medical care will provide symptom relief"* in 71.4% of responses. The results can be seen in figure 3.8 and 3.9.

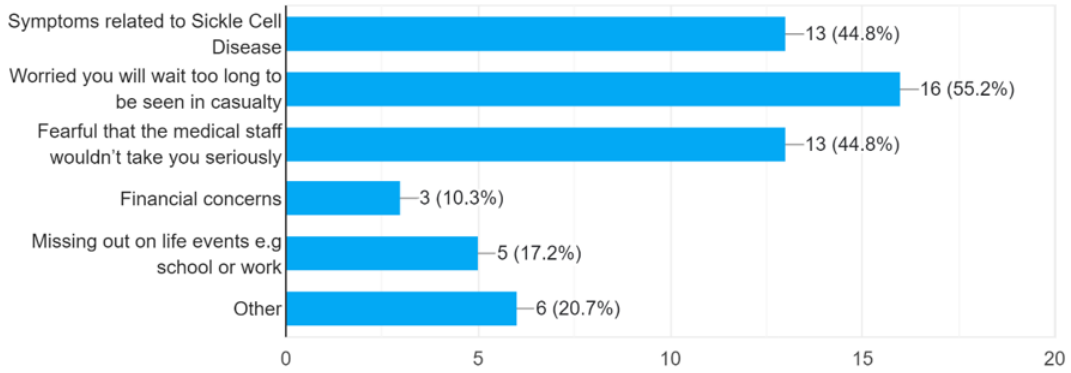

Figure 3.8: Participant's Negative Feelings Pre-Emergency Care

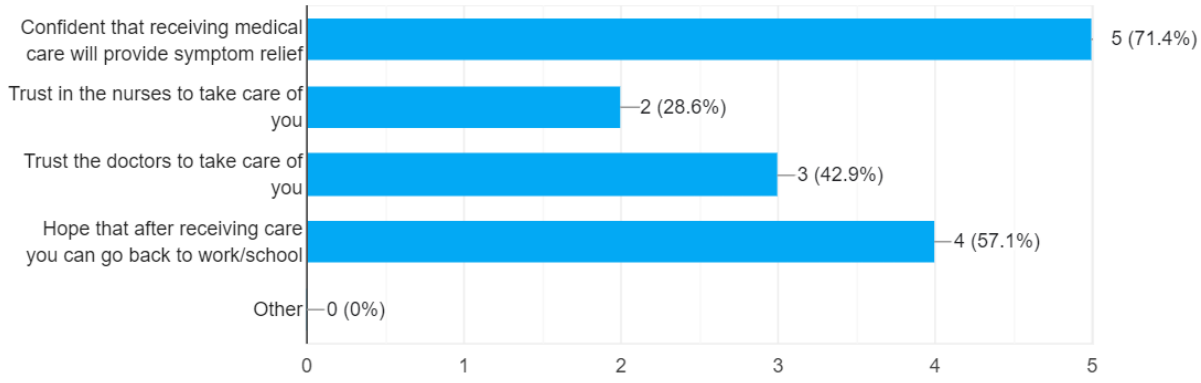

Figure 3.9: Participants' Positive Feelings Pre-Emergency Care

When comparing participant waiting times for administration of medication; out of the 51 responses, one hour was the mode recorded in 31.4% of cases while four hours was recorded in 19.6% of cases. Interestingly, the three participants who recorded waiting 10 minutes to receive medication received their care in the private sector in Gauteng, suggesting a level of efficacy and expediency in operational processes. However, participants who received care in private from the WC and KZN, recorded one hour (n=6) and four hours (n=3) respectively noting the variability in the waiting time experiences.

South African guidelines for managing acute presentations indicate that pain management should be individualised, due to patients having varying threshold of pain and pain tolerance from past exposures, rapid clinical assessment should be performed, and pain relief should also be administered within 30 minutes of arrival (Alli et al., 2014). Majority (51%) of patients who participated in this study (n=51), reported waiting from 30 minutes to 3.5 hours longer than the recommended guidelines to receive medication. A study comparing the pain management for SCD between EC and infusions centres found that when patients received pain medication within 30 minutes of arrival, they were less likely to be admitted into hospital and reported feeling more satisfied with their care and felt safer in the healthcare setting (Lanzkron et al., 2021).

The interviewed participants consistently emphasized the urgency of treating their condition. The impact of delayed care caused concern in patients who are aware that prolonged waiting times can be detrimental, leading to irreversible organ damage. Participants also reported knowing other patients with SCD who were not treated urgently and lost their lives.

"It's very painful. It's excruciatingly painful. It's lifelong and uhm, it can kill. It can kill very quickly if not addressed urgently and that's something they forget"~ P1, interview

"When someone's coming into the EC especially with SCD and complaining of pain, it should be taken seriously. The effects of low oxygen or low blood levels can have is lifelong and as you know our organs aren't the strongest to begin with" ~ P3, interview

The participant attributed the delayed care to be due to HCPs not taking their complaints about pain and distress seriously and dismissing their condition, which can escalate rapidly without having visible symptoms.

"Not being treated in time because you feel it getting worse and worse" ~ P4, interview

“I would say getting someone to see me. The pain just gets worse and no one attends to you, like you invisible, so the biggest challenge for me is getting someone to attend” ~ P5, interview

In section 3.5, lack of knowledge and understanding amongst HCPs was discussed. Participants attributed the lack of urgency to the nurses not knowing what SCD is.

"They didn't even know what SCD is. I waited so long. The nurses, they didn't know." ~P7, interview

Delays in receiving medical treatment was consistent in literature where individuals with SCD reported concerns in not receiving prompt medical attention (Crego et al., 2021). A study by Abdallah et al. (2020) found that 67% of patients felt that their EC care was delayed leading to increased severity of pain, lower healthcare satisfaction and more frequent visits to the EC. Data from a USA study identified that individuals with SCD waited 50% longer compared to patients who arrived with broken legs or arms despite medical guidelines also recommending patients to receive their first dose of pain medication no longer than 30 minutes after arriving at the EC (Haywood et al., 2014). The study by Crego et al., (2021) in the USA mentions difficult intravenous access, lack of HCP education about management guidelines and hospital overcrowding as reasons for delayed care. Solutions put forward by the author includes the use of existing online SCD training resources that HCPs can use. A possible solution that can be adopted and tailored to the healthcare setting in SA where hospitals without Internet access could be provided with a hardcopy of a SCD resource kit to be kept in the EC. Additionally, participants in this study suggested that maintaining a documented list of medications that have proven effective for them in their hospital file would be beneficial.

3.6.2. Subtheme 2: Patient-Centred Pain Management and Triage Challenges

In this study cohort (n=51), pain was the most frequent symptom for participants seeking emergency care recorded by 84.3% of participants. These findings corroborate the data from other studies reporting pain as the most common reasons for visits to the EC (Crego et al., 2021; Lanzkron et al., 2021; Yusuf et al., 2010).

Participants felt that in 66.7% of EC visits the doctors believed that they were in pain. This number differed by 25.5% when asked if they felt the nurses believed they were in pain. This data is graphically represented in figure 3.10.

"SCD is listed as 1 of 20 most painful conditions but people and hospital staff think your pain is for attention" ~ P38, survey

"Some doctors believe your pain, others think it's a joke. The nurse's don't take you seriously and believe you at all" ~ P21, survey

"It's traumatic. I often thought I would die there and I often felt that I'd rather die than be lying there. It's excruciating pain and then you have to still remain in your senses to help the nurses believe and understand what you have" ~P38, survey

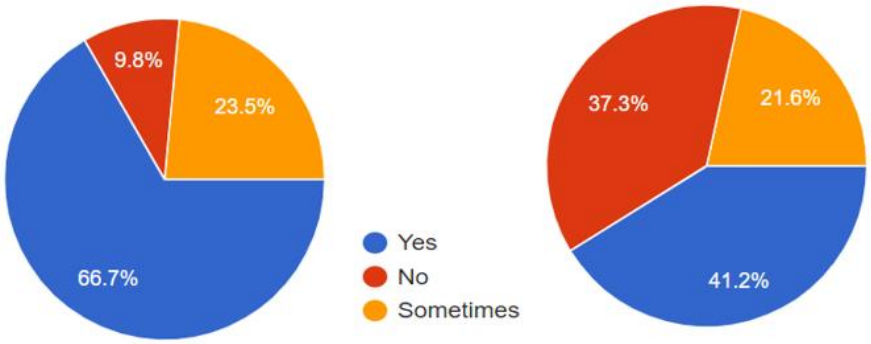

Figure 3.10: Participants' perceptions of pain belief: doctors (left) vs nurses (right)

Literature reported by Crego et al., (2021) and Kanter et al., (2020) highlights patients' perceptions of disbelief in the level of pain they express. This disbelief was attributed to the stigma associated with SCD patients, who are sometimes unfairly labelled as drug addicts and drug seekers (Crego et al., 2021). Interestingly, findings of our study attributed the disbelief to lack of understanding and only three out of 51 participants expressed feeling labelled as drug seekers.

Participants felt that HCPs should go on a pain management course to learn how to manage SC pain and administer appropriate pain medication while waiting.

"HCPs need to go for a pain management course. They need to learn about SCD and how to administer faster care to us. The lack of knowledge is extremely frustrating" ~P5, survey

"Some medication to ease the pain while waiting SCD pain is bone crushing there's nothing like it, it makes you wish you were dead instead"~P43, survey

Interviewed participants narrated challenges with medical treatment and treatment protocols which influenced their experiences.

"I'll go in, give my complaint and wait and wait, I literally waited for up to four hours just to be given pain meds and the pain meds they give is literally such a slap to the face, I don't understand how you going to treat SCD pain with Panado" ~P6, interview

"Start with A, and if A doesn't work, give it two to three hours. Start with B, Just a small process that caters for SCD so that the person is not in pain while you're trying to figure out" ~ P8, interview

Patients explained that the triage system's standard pain protocol may not adequately capture the acute pain episodes experienced by SCD patients, leading to a significant discrepancy between perceived and actual pain levels resulting in delays and inadequacies in the treatment they receive.

"I also feel like that triaging system is flawed, you assigned with a number or a colour, that's not true. Like, that's not true to my pain I feel in that moment, so I feel like the way they triage patients that's where the fault starts" ~ P3, interview

This delay in treatment not only exacerbates their pain but also affects their satisfaction as well as health outcomes.

"Because the pain is too much to keep waiting and then you getting worse because it isn't treated with urgency, sometimes while waiting, I black out from the pain" ~ P4, interview

The limitations of pain scales for patients with SCD was evaluated by Collins, Renedo and Marston (2022) who emphasized significant drawbacks, including the subjective meaning of the scale, its relationship to social, statistical, and psychological factors, as well as mistrust between physicians and patients. Proposed strategies for improving pain management which includes incorporating opioids, nonsteroidal anti-inflammatory drugs, hydration and oxygenation. The use of distractions, local heat options such as hot water bottles, incentive spirometry and early ambulation was suggested (Matthie & Jenerette, 2015).

The need for individualized pain management is a crucial factor in SCD patients' experiences in the EC, echoed consistent findings in the literature (Crego et al., 2021; Kanter et al., 2020). Freiermuth et al. (2014) identified treatment delays and ineffective pain medication in the EC, prompting the development of an evidence-based EC care tool. Section 3.5.1 discussed findings supporting the need for additional educational resources on managing SCD. The Emergency Centre Department Sickle Cell Care Coalition (EDSC3) crafted a point-of-care tool, focusing on six areas: triage, communication, history, evaluation, treatment and disposition. The primary focus of the tool is evaluation of pain indicating that "patient report of pain is the gold standard" (Freiermuth & Ramsey, 2022). This tool offers guidance for EC

providers to enhance patient care, aligning with factors contributing to patient experiences in our study. Its potential incorporation into ECs in SA could improve patient outcomes.

3.6.3. Subtheme 3: Physician-Patient Interactions

Apart from the urgency of medical treatment and management, patients valued being treated humanely, with understanding, empathy, care, kindness and being listened to.

"We just need caring and compassionate healthcare workers" ~P27, survey

"It's very scary to have SCD and to go to the EC with so much pain and then be scared how the medical staff will treat you, will they see you human to, will they understand because sometimes the crisis put me in a coma and I wake up all blue and bruised as well, so how did I get those bruises, how were they treating me"~P48, survey

"Living with SCD is a painful and lonely life. Sometimes we don't know what part of the condition is and what a separate problem is. I wish the hospital staff would listen to understand instead of hearing just to give you a reply quickly"~ P38, survey

These aspects were mentioned by Freiermuth & Ramsey, 2022 and factored into the EC care tool mentioned earlier to include; building trust by believing the patient is in pain, emphasizing that past experiences in the EC might make them guarded, empathetic verbal and nonverbal communication is essential and to be patient as pain can make anyone irritable, impatient, upset and experience difficulty communicating.

In response to whether participants (n=51) delayed seeking emergency care, only 21.6% (n=11) of participants indicated that they don't delay going to the EC. These findings are 18.4% higher than the number of individuals (60%) found to delay going to the EC in the study by Crego et al., (2021). Participants in our study, expressed that EC care is the last resort and figure 3.11 displays the reasons for participants sharing this sentiment. Majority of participants stated "*fearful the medical staff won't take them seriously*" as the main reason. Interestingly, this response differs from the study by Crego et al., (2021) and Abdallah et al., (2020) where prior bad experiences in the EC was listed as the main reason participants avoided going to the EC.

"It's like they don't believe the degree of pain you're in so rather suffer at home until you cannot take the pain no more"~P52, survey

Literature providing recommendations for improving care relate SCD pain in EC to the "iceberg phenomenon" stating that the most pain experienced by patients occurs at home and by the time they present to the EC it reaches the tip of the iceberg which then gets underestimated by HCPs (Matthie & Jenerette, 2015). This aligned with our research findings of patients delaying presenting to the EC and seeing it as the last resort to manage their pain.

Participants who listed *"other"* shared comments relating to knowledge and understanding in HCPs, treatment urgency and transport challenges which will be discussed in section 3.6 and 3.9.

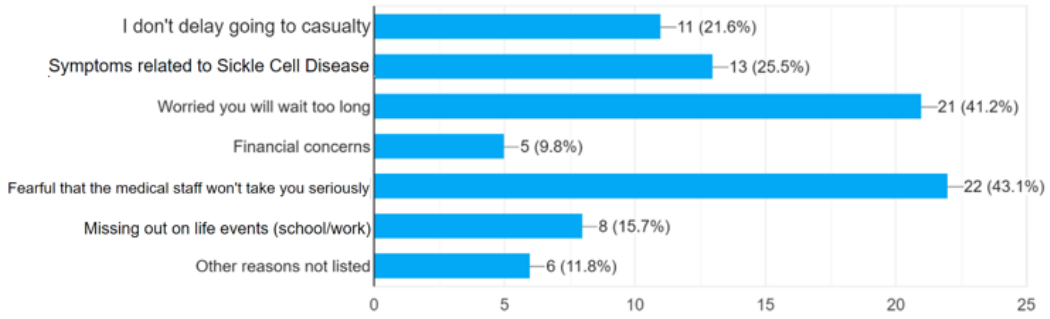

Figure 3.11: Participant Reasons for delaying Emergency Care

Exploring participant (n=51) satisfaction with the communication they received about their health in the EC revealed that the majority (27.5%) felt *"not satisfied at all"* with the communication they received from nurses while only 11.8% felt *"not satisfied at all"* with the communication they received from the doctors. In comparison, 13.7% felt *"very satisfied"* with the communication received from nurses whereas 37.5% felt "*very satisfied"* with the communication from doctors. A significant amount of participants (33.3%) felt "*somewhat satisfied*" with the communication from doctors while only 17.6% were *"somewhat confident"* with the communication from nurses. These findings are detailed in figure 3.12.

When comparing responses from Gauteng (n=18) to the WC (n=25), an equal number of participants from both provinces (33%) responded they were *"neither satisfied nor dissatisfied"* with the communication received from the nurses, however 20% in WC responded *"very satisfied"* compared to only 5.6% from Gauteng. Contrasting to the communication received from doctors, 44% in WC responded *"very satisfied"* and only 27.8% from Gauteng. Two separate T-Tests were conducted to compare the average satisfaction levels with communication from nurses and doctors between the two provinces. The T-Test ($p<0.001$) showed a significant difference in the communication patients received from nurses and doctors between the two provinces. This study found that patients in the WC are more satisfied

with the communication received from both doctors and nurses compared to patients in the Gauteng province.

Figure 3.12: Participant Satisfaction with Communication from Nurses (left) and Doctors (right)

"The nurses don't understand, explain more and only then they take it seriously and communicate with you, the doctors were able to communicate more" ~P32, survey

"The nurse's judge you more than treat you by saying things like oh if you know your condition why can't you manage it and or that oh it can't be that bad. The doctors at least they talk to you and know that the pain can be as severe as you saying it is"~ P48, survey

One participant acknowledged that they understand the nurses are busy but would appreciate better communication as they have to live with the condition and don't want to end up back in hospital.

"I understand the nurses are busy but they can communicate better, I mean, I have to live with my condition and recover on my own. At least talk to me properly about my follow-up care so I don't end up back in hospital" ~ P41, survey

Participants expressed relief when a doctor who was familiar with SCD was involved in their care, this made them feel understood and receive prompt treatment.

"The doctors read my file and kindly understood about sickle cell and what I needed"~ P7, interview

Participants also described their need for communication from doctors in the EC to help better their experiences.

“Also for the doctors to learn how to treat the patients better, learn how to work with patients and I think as a patient as well, I need to work with doctors as well to help other patients learn how to deal with doctors as well learn how to speak out” ~ P1, interview

One of the other six focus areas in the care tool included communication (Freiermuth & Ramsey, 2022). Literature previously indicated that patient satisfaction in the EC is the highest when patients feel that there is a two-way communication flow between themselves and HCPs. Furthermore, patients expressed that they care less about the waiting period if they are kept informed about the waiting times (Feuerwerker et al., 2019). Listening and understanding was narrated in the interviews, participants elaborated on the need to feel heard while taking their medical history and nature of their condition in account.

“Do you know what, I don't want to seem too positive because there’s still work and learning to be done, but honestly speaking where I've been, I've been lucky in a sense of they're willing to listen” ~ P8, interview

"I’ve never experienced someone just being kind, listening, believing that you're in pain and acting in haste when hearing you're in pain." ~ P4, interview

Interview data highlighted positive experiences when HCPs were able to support them, provide urgent care and appropriate pain management.

“In private it’s been good. The nurses understand me, they try to cheer me on, make sure I’ve eaten, make sure I’m drinking something, try to de-stress me, calm my triggers down and get me to calm down as quick as possible so they can treat the pain and hopefully get me home”~ P1, interview

The acknowledgement of supportive HCPs who can help de-escalate the crisis contributes to the impact of patients’ overall satisfaction in the EC.

“If you are kind to someone, that really helps the anxiety and the pain level. Like you taught how to deescalate the pain in nursing and med school and just like being soft with someone can help them, distract them from the feeling, so for me it’s like they don’t care about kind, they don’t care about listening” ~ P3, interview

A supportive healthcare environment influenced participant experiences and was seen in literature with the "champion" role of oncology nurses (Matthie & Jenerette, 2015). This

emphasized the crucial role HCPs in other healthcare sectors can play in caring for individuals with SCD by being aware of their needs. Fostering open communication, empathy, belief in patients' pain reports, and respecting individualized coping behaviours (Matthie & Jenerette, 2015).

Negative experiences were attributed to feeling ignored, misbelieved, misunderstood and mistreated. Instances where participants felt they were treated with discrimination or being stigmatized contributed to negative experiences.

"*At times they will call you a drug seeker because how do you know the dose of the pethidine or morphine you need to control your pain. They forget that we've been dealing especially if you a 30 something year old adult, you've been dealing with that pain for 30 something years, you know your history"~ P1, interview*

"That makes me feel judged, and for me it's.
I don't understand why I'm being judged because I never asked for this"~P3, interview

"They will abruptly tell me I'm not the only patient there and there are others that have priority, so I must wait my turn and get shoved aside"~ P5, interview

Literature emphasizes that language and communication have the potential to stigmatize, cause patients' to feel judged and affect the continuity of care (Speight et al., 2021). This data converged with the results of our study where patients reported feeling judged, being stigmatized as a drug seeker and feeling anxious due to past experiences in the EC impacting patients emotionally and psychologically which links into the theme of Emotional and Psychological Impact (Section 3.8). Empathetic language was demonstrated to influence patient's experience in conditions such as Type 1 Diabetes (Speight et al., 2021), studies emphasized seeing the person first before the condition, highlighting that words create a culture that can be inclusive, reflect support, understanding and respect.

3.7. Theme 3: Patient Advocacy and Support

The theme "Patient Advocacy and Support" was prominent from the data obtained. Patients highlighted the important role that advocacy, either by themselves, family members, friends or society at large plays in navigating the EC. The themes could be split into "Self-Advocacy and Communication" and "Family and Social Support" to provide a deeper understanding of how these aspects contribute to patient experiences in the EC as well as maintain their well-being.

The T-Test found no significant differences between provinces or healthcare sectors (p=0.30 and p=0.23 respectively) suggesting that this theme is relevant across different regions and healthcare settings.

3.7.1. Subtheme 1: Self-Advocacy and Communication

Participants narrated the challenges they experienced due to the lack of knowledge and understanding among HCPs, which was discussed in section 3.5.1. Participants further explained the need for themselves to play an active role in their healthcare by advocating for effective medical management effective. Patient self-advocacy is a skill defined as the direct communication of one's health needs (Bondal, Villanueva & Gleason, 2020).

"You need to ask for strong pain medication and say that we might need blood or need antibiotics and need IV fluid or other treatment because they don't know, so you must know" ~P12, survey

"I know about my disease a lot and most of the time we advocate to the doctors and nurses about our condition, and I wish I can do more but unfortunately we not able because we don't have the tools and the support" ~ P3, survey

Participants emphasized that their lived experience with the condition helps them to advocate for themselves and their desire to use it to help advocate for others. Participants felt that advocacy efforts could overcome treatment challenges.

"I have the lived experience of 25 years with SCD, I don't think the amount or level of pain can ever be comprehended if you don't have the illness yourself. I have to use my lived experience to advocate for myself but also to help others" ~P36, survey

"If we as SCD patients can help educate the EC doctors and relevant departments and even advocate to the community to be aware and know more about our disease, maybe we get treated better" ~P14, survey

Data from the interviews identified the need for patients to advocate for themselves and participants took initiative to learn more about their condition so that they could advocate for themselves.

"They (patients) must always be active in their own health like me I'm even reading a book by a doctor now on treating SCD" ~P7, interview

A patient-centred approach has been recognized in healthcare as the most effective care occurring when patients work together with their providers to make decisions and develop care plans specifically tailored to their needs (Bondal, Villanueva & Gleason, 2020).

Interview data further showed that by feeling empowered patients can confidently communicate their pain management needs and self-advocate for a sense of urgency in receiving care.

> *"I was a teenager and because I was with my guardian, she could advocate for them to move faster and give me the required painkillers and so but then again they would not handle it fast enough. As I got older and I learnt to advocate for myself" ~P1, interview*

A study by Ramos Salazar (2018) indicated that patient self-advocacy has a positive influence on physician-patient relationships as this skill allows patients to confidently approach physicians with less uncertainty and better adaptability. The same study mentions that patients' willingness to seek information and request healthcare education, positively predicted their perceived level of satisfaction with the healthcare they received (Ramos Salazar, 2018).

Although in section 3.5.2 participants expressed the need to have the knowledge and understanding of their condition to advocate for themselves in the EC, they questioned their role as an educator in the setting where they are seeking help, highlighting the challenge of self-advocacy in the midst of a SCD crisis.

> *"How do I have to educate the doctor about my symptoms yet I am seeking care from them. It's somewhat alarming" ~ P4, interview*

A participant narrated an instance where her visit to the EC felt like an interview session with HCPs questioning her about SCD. Furthermore, the participant compared the knowledge HCPs have about SCD to other conditions seen in the EC and that those conditions are seemingly treated without needing explanations.

> *"What stands out to me the most is, there are people willing to learn what SCD is, but we need to have the time and place. When I'm in pain, I don't have time to educate you about it. Like when other people go in with other conditions, you don't hear them explaining, how long have they had it? What treatments they usually take, they get treated. It's just a lot at the point when you are sick, you don't have the energy" ~ P8, interview*

Another participant used the analogy of childbirth to emphasize the difficulty in articulating her medical needs during her symptoms and another emphasized the importance of having support during EC interactions.

"Because the pain is too much how must I be able to educate the nurse how to treat me, imagine you in labour and then you must explain to the nurses what is labour pain, how much pain it is and what needs to be done, its inhumane" ~P1, interview

"If I go alone it's hard for me to advocate for myself"~ P3, interview

Participant frustration about feeling like educators during their EC visits identifies tension in EC interactions, highlighting the need for achieving patient centred care without the burden placed on patients to be educators. Patient centred care transforms the traditional patient-doctor dynamic into a partnership where both parties contribute to the knowledge base. Healthcare professionals can foster patient engagement and feedback, encourage interdisciplinary collaboration and a practical understanding of the disease, ultimately leading to better patient outcomes and more effective management strategies (Crego et al., 2021).

3.7.2. Subtheme 2: Family and Social Support

Transitioning from an individual level of self-advocacy, participants further highlighted that their EC experiences are better when accompanied by a support person. Section 3.5.2 identified that parents and support groups played a role in empowering participants about their condition. In addition, out of 51 responses, 68.6% indicated that their experience in ECs is better when accompanied by a family member or friend.

"At least they will be checking how far your folder is and at times they can advocate for you when you are in pain and can't speak" ~P49, survey

One participant related his experience during the Coronavirus pandemic (COVID) where they noticed the difference it made.

"I went when it was still COVID and they didn't let my dad come in and I was unable to speak or walk so it was so annoying and such a struggle" ~P11, survey

Participants interviewed also expressed their need for someone else to speak on their behalf when they are in pain indicating that when people around them know and understand their condition, they can get help efficiently.

"I've trained my friends and family, so they go with me and start telling the doctors what to do when I'm in severe pain, you need somebody to be there, to speak up for you" ~P8, interview

"I always found that hospital staff was always willing to help when you had someone there to advocate for you." ~P1, interview

"Like people around me know about it and they can help me when I need without wasting time because they understand"~P6, interview

One participant indicated that their experience was better in the EC if she has a letter from her doctor providing information on SCD, tying into the aspect of coordinated healthcare discussed in section 3.9.2.

"It's better when I have company in the EC then I'm not so depressed and also when my doctor gives me letter explaining about the SCD" ~P4, interview

Studies report that family members and social networks can serve as a strong support system, advocating for patients in the EC, their involvement contributes to a comprehensive understanding of patient's needs, preferences and medical history (Nori et al., 2023). In addition, participants of our study expressed relief being accompanied by someone else as they didn't have to worry about advocating for themselves while experiencing pain. Research in rare diseases mention that patients and their caregivers serve as advocates, educating and energizing the public around the rare disease they are affected by. The article further states that with over 7000 rare diseases and the scarcity of expert physicians, patients and caregivers experience a huge burden of accessing knowledgeable providers, and therefore have to take it upon themselves to act as educators, advocating to a variety of audiences (Nori et al., 2023).

One of the participants emphasized the emotional and physical assistance that was needed from his support system in order to help ease both physical manifestation as well as the psychological distress that accompanies an emergency medical crisis.

"My wife, it was very difficult for me to show her when I'm in pain it can bring you down to your knees but now she can help ease the pain with massages and warmth"~ P7, interview

This data reveals the importance of family and social support in patient's experiences in the EC. Patient's support systems are able to act as advocates, caregivers and bring emotional strength. The communication between patients, their support networks and HCPs contributes to positive perceptions of EC care in individuals with SCD and is said to align with the principle of patient-centred care. By providing an effective healthcare experience and creating a positive effect on the physician-patient relationship, patients can feel that they are supported in the healthcare environment (Nori et al., 2023).

Interestingly, out of the 51 responses of our study, 21.6% responded that it *"sometimes"* makes a difference when accompanied and five felt that it made no difference on their experience. Only one participant who selected *"no difference"* shared more information indicating that they still waited long to receive medical attention.

"It doesn't matter who takes you to the hospital you still wait a long time before receiving something for the pain" ~P16, survey

The nuanced insight highlights the interplay of various factors influencing patient experiences within the EC setting and the complex relationship these factors share with patient support systems.

3.8. Theme 4: Emotional and Psychological Impact

The experiences of individuals attending the EC has significant emotional and psychological impact. Subthemes of "Pain Response and Healthcare Assumptions", "Navigating Anxiety, Fear and Uncertainty" and "Coping Mechanisms" will be explored in this section.

3.8.1. Subtheme 1: Pain Response and Healthcare Assumptions

Regarding SCD pain, participants mentioned the exhaustion and emotional strain a SCD crisis has on them apart from the symptom of pain. It creates feelings of frustration and worry as they never know what effect the intensity of pain will have on them.

"I feel different ways with pain but if I go to the EC, it means the pain is unbearable, that pain makes me want to die"~P13, survey

"I worry about not looking sick enough even though the pain is crushing inside the staff won't believe it enough to treat me quickly"~ P48, survey

Participant interview responses highlighted the dual burden of handling their pain in a certain way in order to be treated.

"You have to also learn to behave in a certain way to get treated better so my role now is to be a bridge between the two worlds the medical world and the SCD world" ~P1, interview

"It can make you cry its uh horrible amounts of pain sometimes, it can make you a bit aggressive you don't want to be touched you don't want to talk" ~P3, interview

Patients felt that this physical pain combined with HCP expectations of what pain intensity should look like, creates a stressful environment for them in which they have to worry about balancing pain and maintaining composure in order for their pain to be taken seriously.

"A lot of people with SCD who've had natural labour have said that SCD crisis is worse than giving birth. So you can imagine, how bad SCD is and how a person may act dealing with a lot of pain, at times doctors don't want to deal with someone making that much noise or being aggressive, because again, there's no evidence of pain"~P5, interview

"If you in a lot pain but calm and collected than you can't be in that much pain to warrant you being in hospital (laughs) you have to find your balance (laughs), you have to find a way of dealing with the nurses and doctors to make sure they give you the proper care and respect which can be quite exhausting when you not in that proper mental and emotional space because of the pain"~ P1, interview

This sub-theme also tied in with the theme of physician-patient relationship where kindness displayed by HCPs towards participants provided a positive experience in the EC and eased their anxiety and pain.

"I told this nurse I was in so much pain like I was delirious and I felt like my mind was not my own and I was asking her can I please get another dose of medication and she's like yho in another language "Uyangisokolisa"…and that means you are troubling me or you annoying me and I was like, I don't believe this, so for me it's just like kindness. If you kind to someone that really helps the anxiety the pain level"~P3, interview

Zouki et al (2018) highlighted that patients with SCD report being undertreated for pain in the EC due to providers lacking understanding and compassion. He added that although VOC pain can appear exaggerated for HCPs and necessitates higher doses of narcotics which most

EC providers are not accustomed to administering. HCPs need to keep in mind that every VOC crisis presents differently, every patient has a unique sensation, perception and expression of pain (Zouki et al, 2018). Studies emphasized the need for HCPs to be able to distinguish between tolerance, physical dependence and addiction in individuals with SCD (Matthie & Jenerette, 2015).

Collins, Renedo, and Marston (2022) critically assessed the limitations of pain scales for patients with SCD, highlighting drawbacks such as the subjective nature of the scale, its ties to social, statistical, psychological factors and the presence of mistrust between physicians and patients. This evaluation is particularly more relevant with the findings in section 3.6.3 indicating that patients view the EC as the last resort when seeking medical attention. By the time patients present to the EC their pain is compared to being at the top of the iceberg, medically referred to as the established phase. This phase and extent of pain is known to cause emotional and psychological impact, often resulting in frustration and depression. The complexities identified in pain assessment scales contribute to the need for patient-centred pain management strategies discussed in section 3.6.2.

3.8.2. Subtheme 2: Navigating Anxiety, Fear and Uncertainty

Participants' of this study described the anxiety caused by a continuous cycle of uncertainties. They not only face daily uncertainties inherent to living with a chronic illness, but the anxiety also influences their experiences in the EC. Being already anxious about their condition amplifies their distress during EC encounters, which lead to negative experiences. The negative experiences, in turn, heighten their overall anxiety, creating a cycle where the heightened anxiety leads to poorer EC experiences, further exacerbating their anxiety and influencing their health outcomes.

While there is limited research regarding illness uncertainty in SCD patients, studies exploring the influence of illness uncertainty in cancer has shown to be an additional psychosocial stressor on patients and families (Blake et al., 2018). In section 3.6, among the reasons listed as *"other"* when asked what contributed to negative feelings before visiting the EC, three participants mentioned anxiety as expressed by the quotes below.

"It's anxiety inducing like a double edged sword I can wait at home and get worse or go to casualty wait and worsen" ~ P9, survey

"Is it always going to be like this with the pain and fatigue and little options, every day is a struggle, coming to emergency piles on" ~P48, survey

During the interviews participants mentioned the same factors relating to the uncertainty EC visits can have on their everyday life which causes fear and anxiety.

"We here living everyday with this disease we know from experience and I think it also just reminds me that life is hard but yes life problems are there and now over and above life problems you have this health burden to worry about and it's a challenge because now I can't go to work some days you never know when that's going to happen and yeah it just shows you it's hard out there for people with diseases"~P3, interview

"I never know how a crisis is going to be and how bad a crisis is going to be for me sometimes it's really bad and I lose control of myself and sometimes I am in control Sometimes the pain is also its bad enough for me not to be at home to go to the hospital" ~P1, interview

"I feel scared I don't know what will happen and will the staff be nice and then I question myself like is the pain really bad is it that bad or can I just wait it out" ~P6, interview

A portion of individuals shared that their negative experiences in the EC contribute to traumatic life experiences. In one instance, the treatment they received in the EC reminded them of the bullying they faced in school. While in another, the EC causes them to relive previous traumatic EC experiences causing fear and anxiety.

"Like I get reminded why I was bullied at school, and they would laugh at me coming everyday so it brought like old memories of how I was bullied" ~P2, interview

"It has never been a pleasant experience for me and for me when I get sick I have never been afraid of getting sick I'm afraid of going there because I know what awaits me I know the trauma that I will experience" ~P3, interview

Illness uncertainty refers to the cognitive and emotional challenges experienced by patients due to the unpredictable and ambiguous nature of their health condition (Blake et al., 2018). Illness uncertainty has been viewed as one of the biggest challenges to successful adaptation to a health condition as it affects patients' ability to cope and increases their level of stress. Illness uncertainty plays an important role in the patient's experience and may influence the quest for a cure, decision-making about treatment options and discussing prognosis (Blake et al., 2018).

Reducing the uncertainty can have an impact on reducing adverse psychological outcomes (Blake et al., 2018). One way of alleviating illness uncertainty is by incorporating GC's into SCD patients' healthcare team (Kromberg, Wessels & Krause, 2013). GC's play an important role in helping individuals navigate uncertainties associated with genetic conditions by explaining medical information, empowering patients, facilitating open communication, providing psychosocial support, clarifying disease misconceptions as well as assisting in developing effective coping mechanisms and ensuring coordinated care with other HCPs. These aspects are discussed in the subthemes that follow, Coping Mechanisms and Care Coordination.

3.8.3. Subtheme 3: Coping Mechanisms

Participants' of this study described various coping mechanisms shaping their experiences in the EC. Understanding how patients cope with the physical, emotional and psychological challenges can enrich our understanding influencing patient centred care and the quality of care received in the EC. The quotes below illustrate how participants employ various means of coping to assist their emergency experiences. Participant 4 mentioned her faith in God that the pain didn't take her life.

"It's only through the grace of God that nothing bad has happened to me and that He gave me a second chance in life" ~P4, Survey

A participant described coping through releasing their emotions, followed by withdrawal, becoming less communicative. She expressed the reliance on her support structure to advocate, ensuring she received the appropriate care which was discussed in section 3.7.2. Lastly, her quote mentioned her realization of not affording better care, touching on the financial burden of healthcare faced by patients.

"When I'm in pain I'm literally crying, when I go into a crisis I don't know what it is, is it like the pain and the disappointment of being sick is it the realization that I can't even afford proper care, health care so that's why I'm in this situation I can't really speak for myself. I'm just crying so yeah I'm not really vocal, in terms of telling people like please attend to me so my mum does that" ~P3, interview

The participants also explained how they mentally prepare themselves for EC visits by seeking social support and empowering themselves to cope with the length of hospital stay, potential

work disruptions and interactions with HCPs in the EC. Other means of patient coping mechanisms used during EC visits included; usage of painkillers, and effective communication with the healthcare staff.

"Let me take more painkillers. It's going to help me. I only go when I can see that here there's nothing more I can do."~P8, interview

“The pain from SCD is like nothing you will wish on your enemy. It makes you into another person but you have to be strong mentally and remain calm and not act on the pain. So you talk nicely to the staff and explain to them you have this condition inside your blood and you need treatment quickly and what helps you like the fluids and morphine.” ~P7, interview

There is a scarcity of research pertaining to the utilization of coping mechanisms in healthcare settings by SCD patients. However, a study exploring coping strategies of students with SCD found that 53.9% of students preferred avoidant coping strategies, including self-distraction, denial, substance use and self-blame (Bowmik et al., 2021. In comparison, 46.09% used coping mechanisms like active coping, emotional support and positive reframing. Literature exploring factors influencing patient’s experiences during magnetic resonance imaging examinations (Madl et al., 2022) complemented the findings of this research study. The study found tailoring information on what to expect and how to prepare according to patient coping styles was seen to empower patients.

Bowmik et al., (2021) and Madl et al., (2022) highlights the effect of psychological preparation which can enhance patients’ coping abilities and contribute to a more positive healthcare experience. The authors further emphasized the importance of developing individual coping mechanisms and tailoring interventions to suit patient coping styles towards achieving better patient experiences. Lastly, these aspects link into the last theme discussing Access to Healthcare and Care Coordination.

3.9. Theme 5: Access to Healthcare and Care Coordination

This section discusses the theme “Access to Healthcare and Care Coordination” by breaking it down into two sub-themes. Sub-theme one “Barriers to Accessing Healthcare Services” sheds light on some of the challenges participants faced when accessing healthcare services. Subtheme two “Care Coordination and Genetic Counselling” explores the impact coordinated care has on patient experiences in the EC as well as the value of genetic counselling.

3.9.1. Subtheme 1: Barriers to Accessing Healthcare Services

Among the reasons participants (n=51) delayed going to the EC (section 3.6.3) involved challenges relating to accessing healthcare services. These included transportation challenges (7%) and financial strain and employment issues (25%). In addition, accessibility to specialized care and navigating transitions in healthcare were contributing factors to patient experiences in the EC. When asked where participants (n=51) go for follow-up care, 49% responded to their *"haematologist"* while 23.5% responded to their *"general practitioner",* 19.6% responded *"both"* depending where they could afford to go whereas 7.8% responded *"other"* and when asked to elaborate they indicated they cannot afford follow-up.

"If I have money privately or to government if I have transportation or I don't go at all" ~P9, survey

"The delays it has to be done for family responsibility because you know what it's like with no work no pay" ~P2, survey

Participants interviewed also described the challenges of trying to sustain a job and earn an income while being sick.

"It's hard to get and keep a job when you didn't ask for this, to be sick. How are you supposed to pay your medical bills and look after your family?"~ P4, interview

"I think just something the government should like help with patients who miss work frequently, like I saw my last employers gave me less and less hours to clock and then they got rid of me. I became more sick with stress and couldn't seek care"~P6, interview

The data illustrates the financial challenges and economic barriers to healthcare which impacts both EC care and follow-up medical care. Studies in the US have found that SCD receives minimal resources and less clinical and research funding when compared to other genetic conditions that are deemed equally devastating (Kanter et al., 2021). The authors attribute this disparity to lack of SCD awareness and a political climate that deprioritise diseases affecting vulnerable under-represented populations. The mentioned reasons can hold true for SA, in that the burden of SCD is disproportionately felt by disadvantaged communities in SA (Wonkam et al., 2012) and there is lack of population based data, surveillance and policies prioritizing rare diseases (David et al.; Jacobson, 2022).

A preference for follow up care with their haematologist was noted amongst participants indicating a strong reliance on specialised care. However, a significant portion consult their general practitioner, emphasizing the importance of a multi-disciplinary approach to health care and coordinating care between HCPs.

3.9.2. Subtheme 2: Care Coordination and Genetic Counselling

Care coordination services and tools were assessed in the surveys. Participants (n=51) were asked whether they were referred for genetic counselling, 45.1% responded *"No"*, 31.4% responded *"Yes*". Although an encouraging number of participants have received genetic counselling, the notable portion of individuals reported not being referred for genetic counselling, highlights a gap in provision of this service. The portion of individuals (23.5%) who responded *"I don't know what genetic counselling is"* emphasizes the need for increased awareness and education regarding the benefits of the service.

Majority of participants (58.8%) would like to be referred for genetic counselling whereas the remainder (41.2%) declined a referral. The high number of participants who expressed a desire to be referred for genetic counselling, indicate a need among participants for additional support and information regarding their condition. It would be important to understand the reasons behind individuals who declined a referral and investigate how healthcare services can be tailored to their preferences ensuring that they are adequately informed and supported in their healthcare. This could suggest a future area for research.

A participant from the survey described relief at the opportunity to speak to someone who understands the condition and knowing that the resource is available.

"When I was attending Red Cross Hospital, two GCs from GSH came to explain what SCD is. I was relieved at the chance to speak to someone who knows the condition. Again this year they came, different GCs and answered other questions I had. It helps, you know, like having that chance and not only the internet" ~P10, survey

From the interviews, a participant highlighted how genetic counselling helped in understanding the effects of the SCD.

"When I received education from the GCs that came to explain the condition and how it's formed and the effects of it, I understood better my symptoms and why I had to visit the EC so much" ~P4, interview

Participant testimonies regarding the relief at the opportunity to speak to a GC and have the resource available, indicates the positive impact of genetic counselling. The emotional and informational support that genetic counselling can provide emphasizes its importance in the overall care coordination. These findings align with a study exploring the views of genetic counselling in single young adults (18-28 years) in Ghana (Appiah et al., 2020) which revealed that although participants had some knowledge on SCD there were still gaps in the management of their condition. The mentioned study showed positive perceptions of genetic counselling (Appiah et al., 2020).

A significant difference was observed (Fisher's, *p=0.03*) between the genetic counselling referrals between the Gauteng (n=18) and WC (n=25) provinces. More participants (48% were referred for the service in WC compared to only 12.5% who were referred in Gauteng. Furthermore, 31.3% of participants in Gauteng indicated they didn't know what genetic counselling is while only 12% participants in WC reported not knowing what genetic counselling is. These findings highlight the variation among provinces in access to genetic counselling services as well a disparity in awareness about the service.

Participants (n=51) were asked whether they have a medic alert bracelet, 76.2% responded *"No"* and only 23.8% said *"Yes"*. Participants who answered *"No"* were asked to provide a reason, the majority (75%) answered that they were never offered one, 14.6% responded it was a personal choice, 6.8% answered that they don't think it will help and 4.5% chose *"Other"*. The reason mentioned as *"other"* was that it can't help because HCPs don't know what the condition is. Participants who answered *"Yes"* to having a medic alert bracelet were further asked whether it helped them in the EC where 80% responded *"No"* and 20% *"Yes".* Participants from the survey agreed with the sentiment that the medic alert bracelet didn't help because the HCPs didn't know what the condition was.

"I have one yes but it can't help because I go to EC and the nurses see it but don't know what the condition is" ~P35, survey

"The ambulance they don't know what it means and then the nurses also don't know SC on the emergency tag" ~P38, survey

Interview data was consistent with these findings, expressed in the quote bellow.

"I had one (medic alert bracelet) but they didn't know what the condition means so it didn't help but if they can learn together with the bracelet and what it is maybe it can help" ~P4, interview

The small number of participants reported having a medic alert bracelet indicates a relatively low adoption of this accessory among individuals with SCD. This raises questions about awareness, accessibility and communication between HCPs and patients regarding the importance of such identification.

There is limited literature on patients' perspectives regarding the utility of medic alert bracelets. However studies exploring the awareness of medical identification (ID) tags amongst HCPs highlighted a lack of awareness of medical IDs and their importance, with majority (60-65%) of HCPs in the studies not advising their chronically ill patients to use them (Al-Alwan et al., 2020; Rahman, Walker & Sultan, 2017). The authors recommend that awareness campaigns be run to promote the use of medical IDs among both patients and HCPs as well as campaigns for HCPs directed at recognizing a medical ID tag to provide prompt and appropriate medical treatment. Similar interventions in SA may assist to effectively utilize the resource (Al-Alwan et al., 2020; Rahman, Walker & Sultan, 2017).

Apart from genetic counselling and medic alert bracelets, one participant described a positive experience when he attended an EC in a different province of SA. The doctors treating him in the EC were able to communicate with his specialist physician who manages his condition in his hometown. This emphasizes the positive influence care coordination between healthcare facilities can have on patient experiences.

"I had a good experience the one time in Pretoria, even though I waited a long time, the doctor yeah he actually called my specialist here in Cape Town before treating me but once he treated me my pain resolved" ~P7, interview

Another participant described a challenge with the lack of coordinated care within the hospital, due to poor administrative processes.

"*They needed my yellow folder and it took a long time, they couldn't find it" ~P5, interview*

Challenges due to lack of care coordination was observed when participants described their transition from a paediatric hospital to an adult hospital. A participant highlighted that as a child they received specialized EC in a paediatric hospital, which they found to be effective with short waiting times. These aspects reinforce the importance of effective communication and collaboration among HCPs through administrative processes of maintaining patient records. Coordinated care efforts should extend across different healthcare settings, ensuring communication and timely access to information to better patient experiences.

"Red Cross Hospital because they've got their own like Oncology/Haematology unit so you don't go to EC you go to the ward so there's no waiting time that was the only time as soon as you come you seen and as soon as I left Red Cross as a teenager and adult I've never had a positive experience in a public hospital" ~P3, Interview

Another participant mentioned transitioning into adult care is challenging due to HCPs not taking her symptoms seriously, while as a child she felt she was always believed.

"I think maybe when you a child people are more likely to listen and be more open and understanding. So as a child I don't really remember coming across staff that didn't believe me, like that wasn't something I worried about when I was a child even going into my teens. Now as an adult it's oh my gosh these nurses will not believe me, now you have this fear of being believed, you know will people take me seriously?" ~P8, interview

The challenges experienced by individuals with SCD when transitioning from paediatric to adult care aligns with literature from other countries. Literature reported that individuals with SCD experience challenges with receiving timely care, appropriate pain relief medications, felt that non-specialist staff didn't know enough to treat them effectively and HCPs seemed not prepared to listen, believe them or act on what the patients requested (Renedo et al., 2019). The authors identified a need for making healthcare services user friendly for young adults by offering compassionate care, training staff in SCD and enhancing HCP staff communication skills (Renedo et al., 2019). The study also suggested young adults be actively involved in shaping their healthcare by self-management strategies and self-advocacy (Renedo et al., 2019). These aspects tie into the broader findings of this study echoing the interconnected nature of each theme and how their interplay significantly shapes the perceptions of EC care by SCD patients.

Chapter Four: Conclusions, Strengths and Limitations of this Study and Recommendations for Future Research

4.1. Chapter Introduction

The concluding chapter of this study summarises the relevant findings from this research. The strengths and limitations will also be outlined along with suggestions for future research.

4.2. Conclusion

This study aimed to investigate the perceived experiences of individuals with SCD who present for care at ECs in South African hospitals. Gaining insight into SCD patient experiences when presenting to ECs in SA can assist in identifying possible strengths and shortfalls in emergency care. Enhancing patient experiences in the EC may not only add to improved clinical outcomes but also improve patient satisfaction, reduce patient dropout and the risk of patients seeking alternative therapies.

The findings firstly revealed that participants felt a lack of awareness and understanding among HCPs impacted patient care and led to delayed treatment. Concerns about inadequate HCP knowledge prompted some participants to seek alternative care. Additionally, provincial variations in participant confidence levels suggest potential disparities in healthcare infrastructure, resources and training. Participants expressed a higher confidence in doctors' understanding of SCD than nurses', highlighting the need for targeted education interventions. Participants recognized the impact of their own knowledge on EC experiences. Participants who felt well-informed about their condition demonstrated increased awareness of symptoms leading to effective communication with HCPs and prompt treatment. This contrasts with individuals who expressed confusion and frustration due to inadequate knowledge, emphasizing the importance of patient education to empower other individuals in managing their condition.

Secondly, the urgency of receiving medical treatment was a concerning factor affecting patient experiences in the EC, with delays causing increased pain and distress. The waiting times for medication administration often exceeded the recommended guidelines, indicating a need for more efficient processes in the EC as well as patient centred pain management strategies. In addition, positive patient experiences correlated with effective communication, compassion and timely symptom relief.

Thirdly, the study illustrated the crucial role of patients actively communicating their health needs to HCPs. Despite challenges, participants believed that their lived experiences with the condition empowered them to advocate not only for themselves but also for others. This theme also identified the challenge patients felt with the need to take on an educator role during their visits to the EC, emphasizing the need for achieving a balance between patient-centred care without placing a burden on patients. Family and social support shed light on the positive impact of having a support person during emergency visits. The emotional and physical assistance provided by the support system was crucial in easing both the physical and psychological distress experienced in the EC.

Fourth, the emotional strain and frustration experienced by participants during a SCD crisis and visit to the EC was discussed. The challenge of expressing pain and conforming to HCP expectations was narrated by patients. Participants described the anxiety stemming from illness uncertainty that are linked to negative feelings before and during EC visits. The impact of previous negative experiences on mental health contributed to experiences in the EC with the use of various coping mechanisms to better manage their experiences in the EC. These findings highlighted the significance of psychosocial interventions tailored to individual coping styles.

Lastly, barriers to accessing healthcare services, including transportation challenges and financial strain was identified. The importance of care coordination and genetic counselling was noted as positively contributing towards more positive experiences in the EC. There was a notable gap in the provision of genetic counselling, indicating a need for increased awareness and education. Moreover, the adoption and effectiveness of medic alert bracelets was explored. The findings suggest a relatively low adoption of this accessory, with participants narrating a lack of awareness amongst HCPs regarding its importance.

4.3. Practical Implications of this Research

The study emphasizes the interconnected nature of the identified themes and their impact on the perceptions of EC care by individuals with SCD. While we acknowledge certain aspects are reflective of resource constrained settings and more challenging to address, patients have shared valuable insights from their experiences, suggesting practical areas for improvement which may be easier to implement. To facilitate the discussion, these suggestions will be categorized into two sections: those suggestive of easier implementation and those addressing more challenging aspects.

Simpler suggestions that may contribute towards a significant difference:

- Participant suggestions were made for having a documented list of medications placed in their files that have been shown to be effective from their medical history. This may facilitate more efficient treatment.
- Participants requested interim conservative pain relief measures while they wait for more comprehensive treatment, this included options like hot water bottles.
- Participants narrated more positive experiences when accompanied by a support person in the EC, allowing family members or friends to be present during the EC visit, if not already allowed in, might provide patients with comfort and facilitate communication between EC providers.
- Promoting the use of empathetic language in healthcare settings merits consideration towards building trust with patients, enhancing communication, providing emotional support while reducing anxiety and can improve patient satisfaction.
- Encouraging and empowering patients to advocate for themselves by educating patients about their condition, fostering a sense of agency in managing their healthcare and including initiatives to support patients in navigating the EC. This may be achieved by referring patients for genetic counselling and to a support group.
- GCs could tailor information to SCD patients, assisting in educating patients on the genetic, medical and psychosocial aspects of their condition as well as preparing them on what to expect in the EC, the impact of pain crises on their overall wellbeing and assisting with the development of coping mechanisms. Incorporating GCs in the development of educational materials, resources and toolkits for individuals affected by SCD could potentially provide participants with knowledge without the use of complex medical jargon.
- Given the emotional and psychological impact reported by participants, healthcare settings could provide pamphlets with the details for social worker services and mental health resources should patients need assistance.

Suggestions that are more challenging to address:

- The study identifies the need for continuous, targeted education programs for HCPs to bridge knowledge gaps related to SCD.
- Efficient access to medical records: participants mentioned the need for quicker access to patients' medical records, possibly through a dedicated folder or system for emergencies. This could help streamline the treatment process, especially in situations where time is critical.

- Addressing delays in medical treatment, particularly in pain relief. While this aspect is challenging due the high patient to low staff ratio, individualized, effective pain relief strategies can be implemented for patients as well as the implementation of a more streamlined triage system for haematology patients.
- The study emphasizes the importance of individualized and patient-centered pain management approaches. SCD treatment protocols should be adopted so HCPs can adequately address the unique needs of patients.
- The need for an improved transport system or alternative options such as telemedicine which can help HCPs to assess the need for pain relief medication, prescription renewals without the need for in-person visits, adjusting of pain medication and allowing more timely interventions before patients' symptoms reach the tip of the iceberg.
- Encourage collaboration between GC's, HCPs, and support structures to ensure comprehensive care. Examples of this is a haematologist providing the initial diagnosis and referring to a GC to expand more on the medical and genetic information, the GC can inform about risks involved with the condition when need arises refer to a social worker to help navigate the emotional and socio-economic challenges patients might face or to a mental health provider to assist with coping mechanisms.
- Participants' of this study showed limited adoption of medic alert bracelets. To enhance their effectiveness during EC visits, alternative identification tools can be suggested. Additionally, exploring the potential of an extended support tool linked to the medic alert bracelet, providing detailed information about SCD or the patient's medical history, might be a valuable avenue for improvement.
- Financial considerations for patients with chronic illness such as financial assistance programs.
- Policymakers could implement policies addressing the specific needs of individuals with SCD in the EC like updating existing guidelines, allocating resources to provide pamphlets, hot water bottles and promoting research to continually enhance the quality of care.

4.4. Recommendations for Future Research

- The exploration of gaps in HCP knowledge to develop targeted education programs.
- Intervention studies for reducing waiting times for individuals with SCD in the EC. Evaluate the effectiveness of these interventions in improving patient satisfaction, health outcomes and adherence to recommended treatment guidelines.

- Further qualitative research to gather insights into the specific factors influencing confidence levels among individuals with SCD between provinces. This can contribute to targeted interventions to improve patient confidence and overall healthcare experiences.
- Explore and develop patient-centred pain management strategies specifically tailored to the needs of SCD patients.
- Explore the impact of psychosocial support services and mental health education on patient experiences in healthcare settings
- Comparative and comprehensive studies across healthcare settings in SA comparing the experiences of SCD patients. Investigating the variations in care quality, waiting time and patient outcomes to identify other factors affecting patient experiences and potential areas for improvement in both sectors.
- Further explore the challenges of self-advocacy faced by individuals with SCD during EC visits. Investigate the factors that hinder patients from effectively communicating their needs and explore interventions to support self-advocacy.
- Investigate and explore the option of community-based interventions in supporting individuals with SCD during emergencies. Community resources, peer support networks and educational programs could play a crucial role in improving overall patient well-being
- Evaluate the impact of existing healthcare policies on the care provided to individuals with SCD in emergency settings. Assess whether policy changes or updates are needed to better address the unique needs of this patient population.
- Investigate strategies to make healthcare services more user-friendly for young adults with SCD, addressing the challenges reported during transitions from paediatric to adult care. Assess the impact of compassionate care, staff training in SCD and enhanced communication skills on patient satisfaction and healthcare outcomes.
- Compare the experiences of individuals with SCD to those with other chronic conditions to identify common challenges and effective strategies for improving healthcare delivery. This can contribute to a broader understanding of best practices in managing chronic diseases.
- Conduct international comparative studies to assess how emergency care experiences for individuals with SCD vary across different countries and healthcare systems. Identify best practices and lessons that can be shared globally.

4.5. Strengths of this Study

- To the best of our knowledge, this is the first study in Africa to explore the perceptions of adult patients with SCD when attending the EC.
- The study used a mixed method approach to provide a comprehensive understanding of the factors influencing patient experiences in the EC.
    - The quantitative section gathered numerical data that provided a broader view of the prevalence of specific perceptions as well as allowed comparisons to be made across different provinces and healthcare sectors.
    - Qualitative methods, such as interviews or focus groups, provided in-depth insights into the lived experiences and perceptions of adult patients with SCD. This allowed for a comprehensive understanding of their feelings, attitudes and behaviours related to EC visits.
- Triangulation was used to recruit participants from different settings as well as to compare and contrast both quantitative and qualitative findings.

4.6. Limitations of this Study

- The study met the minimum sample size requirement (n=51) for the quantitative section limiting the generalizability of the results and compromising statistical precision.
- All participants had internet access.
- Participants were mostly under the age of 30.
- Bias in sampling as only those attending a tertiary institution or had access to social media were recruited.
- Under representation of males.
- Terminology used in the questionnaire. The questionnaire had terms such as “poor, average and good” which were later noted that these terms could have ambiguous meanings and the question could be phrased to ask what was good and what was bad in participants EC visits.
- The sample consisted of a skewed representation of participants from the different provinces with Gauteng and WC being the most represented.

- The researcher tried to recruit participants from other hospitals within her vicinity, however ethics was only granted for one hospital. The hospital the researcher was granted approval from is one of the better staffed and resourced settings.
- All participants had to be English speaking to be included in the study. English was not necessarily the native language of all participants therefore the quality of data may have been affected.

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Appendices

Appendix A: Information Sheet

MMedSci Genetic Counselling Research Information Sheet

The University of Cape Town, Faculty of Health Sciences, Department of Human Genetics

For any questions about the research project, please contact Ms Nabeelah Peerbhai on 0847050392 or at Prbnab001@gmail.com, Ms Kalinka Popel at Kalinka.popel@uct.ac.za, Dr Careni Spencer at Careni.spencer@uct.ac.za, Prof Vernon Louw at Vernon.louw@uct.ac.za or the UCT ethics committee at 021 650 1236 or hrec-enquiries@uct.ac.za.

You are Invited to Participate in a Research Project Investigating the Perceived Experiences of Adult Patients with Sickle Cell Disease who Present for Care to the Emergency Centre's of South African Hospitals

The research will add to the body of knowledge in caring for Sickle Cell Disease patients and contribute to health care professionals having a better understanding of how adult patients with Sickle Cell Disease perceive their care in Emergency Centres (ECs) of South African hospitals. This information will assist health care professionals working in EC's to be more aware of patients' experiences. Genetic counsellors will be able to use the information from this study to help individuals with SCD understand and better prepare for emergency visits.

The survey can be conducted either electronically, telephonically at a time convenient to you, over video call or in person if you attending a clinic affiliated with the University of Cape Town. The interaction will be in English and will take a maximum of 30 minutes. There will be an option to further participate in an interview with the researcher after the survey is completed. Should you wish to be interviewed, you will be asked about your experience in the ECs in more detail. The interviews will take approximately 40 minutes and be audio recorded for research purposes with all information being kept confidential and anonymous.

Informed consent will be required before participating in the interview. Your participation is voluntary and you have the option of withdrawing at any point. Kindly note that there will be no financial gain from your participation in this research project.

Please contact the researcher, Nabeelah Peerbhai, on 0847050392 via SMS, Whatsapp or via email at prbnab001@myuct.ac.za should you wish to participate.

Many thanks,

Nabeelah Peerbhai (Prbnab001@myuct.ac.za)

Dr C Spencer (Careni.spencer@uct.ac.za)

Ms K Popel (Kalinka.popel@uct.ac.za)

Prof V Louw (Vernon.louw@uct.ac.za)

Appendix B: Consent Form

Statement by participant

I, _____ confirm that:

1) I have been invited to participate in the above mentioned research project through the Division of Human Genetics at the University of Cape Town. I understand that other adult participants will be involved in the study and that my name and other personal information will not be shared.

2) I understand that the objective of the study is to explore adult patients with Sickle Cell Disease experiences in Emergency Centre's of South African hospitals.

3) I understand that the interview will take place in the hospital on the day of my appointment at Haematology or alternatively over the phone.

4) I understand that I voluntarily choose to participate in this study and if I choose to no longer continue, my decision will not in any way affect the health care services I receive.

5) I understand that I may choose not to answer any questions if I do not wish to do so. Should I wish to stop with the interview process at any point, it will not impact my medical care in any way.

6) All information collected will remain confidential and will only be used for research purposes.

7) I understand that the interview will be recorded for research purposes. Audio recordings will be safely stored on a computer will only be accessible via a password. The researcher and her supervisors will have access to the data.

8) I understand that the interview will take place in English.

9) I understand that this study has been approved by the registered Human Research Ethics Committee at the Faculty of Health Sciences at the University of Cape Town. I have been given contact details should I wish to contact the committee about how I was treated as a research participant

10) The researcher/HCP has explained the information of this study in English and I understand this information.

Participant name and surname:

_____

Contact details of participant:

_____

Participant`s signature:

_____

Researcher's signature:

_____

Date: _____

Human Research Ethics Committee contact information: 021 650 1236 or hrec-enquiries@uct.ac.za

Appendix C: Quantitative Survey

Patient Perspectives of Sickle Cell Disease Care in Emergency Centres of South African Hospitals

Kindly complete the survey until it gives you the option to submit.

Where applicable you may tick more than one appropriate answer.

Below each question there is a space for any additional comments should you wish to add anything extra relating to the question.

*Required

1. Gender*

Mark only one.

☐ Male

☐ Female

☐ Non-binary

☐ Prefer not to say

2. How old are you? *

_____

3. Are you originally from South Africa?*

Mark only one.

☐ Yes Skip to question 4

☐ No Go to question 3.1

3.1. If no, when did you move to South Africa?

Mark only one.

☐ Adult (18 years or older)

☐ Child (under 18 years of age)

3.2. What is your country of origin?

_____

4. Do you access care in the private or public setting?*

Mark only one.

☐ Public

☐ Private

☐ Both

5. Which province/s do you seek healthcare in?*

Please select all that apply

☐ Eastern Cape

☐ Free State

☐ Gauteng

☐ KwaZulu-Natal

☐ Limpopo

☐ Mpumalanga

☐ Northern Cape

☐ Western Cape

☐ North West

6. Which province do you **mainly** seek healthcare in?*

Mark only one.

☐ Eastern Cape

☐ Free State

☐ Gauteng

☐ KwaZulu-Natal

☐ Limpopo

☐ Mpumalanga

☐ Northern Cape

☐ Western Cape

☐ North West

7. Where do you go first when feeling unwell?*

Mark only one.

☐ Private doctor

☐ To the local clinic

☐ To the nearest hospital

Any additional comments:

_____

8.1. Has anyone ever explained to you what to expect when visiting the Emergency Centre?*

Mark only one.

☐ Yes

☐ No

Any additional comments:

_____

8.2. Would you like to know more about how the Emergency Centre functions?*

Mark only one.

☐ Yes

☐ No

Any additional comments:

_____

9. Which factor is the most important to you when visiting the Emergency Centre?*

Mark only one.

☐ Relief of symptoms (to feel better than you did before you came to the EC)

☐ Care & compassion (Feeling heard or understood)

☐ Dignity & respect (Being treated positively, with belief and trust)

☐ Efficiency (how quickly you were attended to)

☐ Information & communication (whether the staff spoke to you and explained things to you)

☐ Other

If other, please specify

_____

Any additional comments:

_____

10. Which factor is the second most important when visiting the Emergency Centre?*

Mark only one.

☐ Relief of symptoms (to feel better than you did before you came to the EC)
☐ Care & compassion (Feeling heard or understood)
☐ Dignity & respect (Being treated positively, with belief and trust)
☐ Efficiency (how quickly you were attended to)
☐ Information & communication (Whether the staff spoke to you & explained things)
☐ Other

If other, please specify

_____

Any additional comments:

_____

11. Generally, how would you describe visits to the Emergency Centre?*

Mark only one.

☐ Very good
☐ Good
☐ Average
☐ Poor
☐ Very poor

Any additional comments:

_____

12. In the last 5 years, how often have you been to the Emergency Centre due to complications related to Sickle Cell Disease?*

Mark only one.

☐ 1 time

☐ 2-5 times

☐ >5 times

Any additional comments:

_____

13. In the last year, how often have you been to the Emergency Centre due to complications related to Sickle Cell Disease?*

Mark only one.

☐ None

☐ 1 time

☐ 2-5 times

☐ >5 times

Any additional comments:

_____

14. How would you describe your last visit to the Emergency Centre?*

Mark only one.

☐ Very good

☐ Good

☐ Average

☐ Poor

☐ Very poor

Any additional comments:

_____

15. How would you describe your experience upon arrival in Emergency Centre?*

Mark only one.

☐ Good

☐ Neutral

☐ Poor

16. How would you describe your experience in the waiting area?*

Mark only one.

☐ Good

☐ Neutral

☐ Poor

17. How would you describe your experience in the treatment room?*

Mark only one.

☐ Good

☐ Neutral

☐ Poor

Any additional comments:

_____

18. How do you feel before needing to visit the Emergency Centre?*

Mark only one.

☐ Positive Skip to question 18.1

☐ Neutral Skip to question 19

☐ Negative Skip to question 18.2

18.1. If you answered positive, what makes you feel this way?

Tick all that apply.

☐ Confident that receiving medical care will provide symptom relief

☐ Trust in the nurses to take care of you

☐ Trust the doctors to take care of you

☐ Hope that after receiving care you can go back to work/school

☐ Other

If other, please specify

_____

18.2. If you answered negative, what were the reasons for feeling this way?

Tick all that apply.

☐ Symptoms related to Sickle Cell Disease

☐ Worried you will wait too long to be seen in Emergency

☐ Fearful that the medical staff wouldn't take you seriously

☐ Financial concerns

☐ Missing out on life events e.g. school or work

☐ Other

If other, please specify

_____

Any additional comments:

_____

19. Do you ever delay going to the Emergency Centre because of reasons listed below?*

Tick all that apply.

☐ I don't delay going to the Emergency Centre

☐ Symptoms related to Sickle Cell Disease

☐ Worried you will wait too long

☐ Financial concerns

☐ Fearful that the medical staff won't take you seriously

☐ Missing out on life events (school/work)

☐ Other reasons not listed

If other reasons, please specify

_____

Any additional comments:

_____

20. How satisfied were you with the communication you received regarding your health from the nurses in the Emergency Centre?*

Mark only one.

☐ Very satisfied

☐ Somewhat satisfied

☐ Neither satisfied nor dissatisfied

☐ Somewhat dissatisfied

☐ Not satisfied at all

Any additional comments:

_____

21. How satisfied were you with the communication you received regarding your health from the doctor's in the Emergency Centre?*

Mark only one.

☐ Very satisfied

☐ Somewhat satisfied

☐ Neither satisfied nor dissatisfied

☐ Somewhat dissatisfied

☐ Not satisfied at all

Any additional comments:

_____

22. During your last visit to Emergency, how confident were you that the nurses knew how to treat Sickle Cell Disease?*

Mark only one.

☐ Very confident

☐ Somewhat confident

☐ Neither confident nor unconfident

☐ Somewhat unconfident

☐ Not confident at all

Any additional comments:

_____

23. During your last visit to Emergency, how confident were you that the doctors knew how to treat Sickle Cell Disease?*

Mark only one.

☐ Very confident

☐ Somewhat confident

☐ Neither confident nor unconfident

☐ Somewhat unconfident

☐ Not confident at all

Any additional comments:

_____

24. Were any of your visits to the Emergency centre due to pain?*

Mark only one.

☐ Yes

☐ No

Any additional comments:

_____

25. In your opinion, did the doctors believe that you were in pain?*

Mark only one.

☐ Yes

☐ No

☐ Sometimes

Any additional comments:

_____

26. In your opinion, did the nurses believe that you were in pain?*

Mark only one.

☐ Yes

☐ No

☐ Sometimes

Any additional comments:

_____

27. How would you grade your pain after your Emergency Centre visit?*

Mark only one.

☐ Resolved

☐ Improved

☐ Unchanged

☐ Worsened

Any additional comments:

_____

28. Upon discharge from the Emergency Centre, did you feel?*

Mark only one.

☐ Your symptoms improved

☐ Your symptoms remained the same

☐ Your symptoms felt worse

☐ Other

If other, please specify

_____

Any additional comments:

_____

29. Have you ever had to return to the Emergency centre within a day or two of your last visit?*

Mark only one.

☐ Yes

☐ No

Any additional comments:

_____

30. Have you ever left the Emergency centre to seek alternative help?*

Mark only one.

☐ Yes Skip to question 30.1

☐ No Skip to question 31

30.1. If yes, what was the reason for leaving?

Tick all that apply.

☐ Prolonged waiting time to see the nurses

☐ Prolonged waiting time to see the doctors

☐ Lack of confidence in the healthcare practitioner's knowledge

☐ Lack of healthcare practitioner's empathy

☐ Poor communication regarding my management plan

☐ Language barriers

☐ Other

If other, please specify

_____

Any additional comments:

_____

31. What would you describe as prolonged waiting time?*

Mark only one*

☐ 10 minutes

☐ 30 minutes

☐ 1 hour

☐ 2 hours

☐ 4 hours

☐ Over 4 hours

Any additional comments:

_____

32. How long do you think is an appropriate time to wait to have your observations (blood pressure, temperature etc.) taken?*

Mark only one.

☐ 10 minutes

☐ 15- 30 minutes

☐ 1 hour

☐ 2 hours

☐ 4 hours

☐ Over 4 hours

Any additional comments:

_____

33. How long do you think is an appropriate time to wait to see a nurse?*

Mark only one.

☐ 10 minutes

☐ 30 minutes

☐ 1 hour

☐ 2 hours

☐ 4 hours

☐ Over 4 hours

Any additional comments:

_____

34. How long do you think is an appropriate time to wait to see a doctor?

Mark only one.

☐ 10 minutes

☐ 30 minutes

☐ 1 hour

☐ 2 hours

☐ 4 hours

☐ Over 4 hours

Any additional comments:

_____

35. How long do you think is an appropriate time to wait before medication is administered?*

Mark only one.

☐ 10 minutes

☐ 30 minutes

☐ 1 hour

☐ 2 hours

☐ 4 hours

☐ Over 4 hours

Any additional comments:

_____

36. How long do you wait before medication is administered?*

Mark only one.

☐ 10 minutes

☐ 30 minutes

☐ 1 hour

☐ 2 hours

☐ 4 hours

☐ Over 4 hours

Any additional comments:

_____

37. Do you felt like your experience in the Emergency centre is better when accompanied by a family member or friend?*

Mark only one.

☐ Yes

☐ No

☐ Sometimes

Any additional comments:

_____

38. Where do you go for follow-up care?*

Tick all that apply.

☐ To my General Practitioner

☐ To my Haematologist

☐ Both

☐ Other

If other, please specify

_____

Any additional comments:

_____

39. What else do you think could improve your experience as a patient?*

_____

40. Have you ever received education (oral or in writing) about your condition?*

Mark only one.

☐ Yes

☐ No

☐ Maybe, I don't remember

Any additional comments:

_____

41. Who gave you the information?*

Mark only one.

☐ My doctor

☐ A nurse

☐ Both

☐ Other

Any additional comments:

_____

42. How would you describe your level of knowledge about your condition?*

Mark only one.

☐ Excellent, I understand all aspects of my condition

☐ Good, I understand most aspects but still have questions

☐ Fair, I know enough to manage my condition

☐ Poor, there are aspects I am unclear about

Any additional comments:

_____

43. Would you like to learn more about your condition?*

Mark only one.

☐ Yes

☐ No

☐ Maybe

Any additional comments:

_____

44. Have you ever been referred for Genetic Counselling?*

Mark only one.

☐ Yes Skip to question 45

☐ No Skip to question 44.1

☐ I dont know what Genetic Counselling is Skip to question 45

Any additional comments:

_____

44.1. Would you like to be referred for Genetic Counselling?*

Mark only one.

☐ Yes If yes, please enter your name and number

☐ No

45. Do you have a medic alert bracelet?

Mark only one.

☐ Yes Skip to question 45.1

☐ No Skip to question 45.2

45.1. If yes, has this helped you when visiting the Emergency centre?

Mark only one.

☐ Yes

☐ No

45.2. If no, please choose the most appropriate reason:

Mark only one.

☐ Personal choice

☐ I was never offered one

☐ I don't think it will help

☐ Other

If other, please specify

_____

Any additional comments:

_____

46. Would you like to share more details about your experience in the Emergency centre in an interview?*

Mark only one.

☐ Yes If yes, please enter your name and number

☐ No

Appendix D: Qualitative Interview Guide

The questions will focus on more detailed experiences and challenges patients with SCD face when going to the ECs in South African hospitals. Questions will draw from the quantitative answers of participants and will also include how patients think the care and service levels can be improved.

Describe to me what happens when you visit the EC?

What aspects stand out for you from your last visit?

Tell me about a positive experience that you had in the EC?

Tell me about a time when you had a negative experience in the EC?

What would you say is the biggest challenge that you experience when visiting the EC?

How has your experiences in the EC impacted your overall view of living with SCD?

What is your biggest concern when visiting the EC?

What has your experience been with the nurses and doctors in the EC?

What would be one thing that you would like the doctors or nurses treating you in the EC to know about SCD?

Have you needed to remain alert in the EC?

What was the service like for SCD in the country that you from? (If originally from a different country than SA).

What would you like to know more about regarding the EC?

What suggestions do you have to help better the care you receive in the EC?

Appendix E: Research Reflection

Before starting this research project, I felt overwhelmed with doubt, questioning whether I had the capacity to hold space for the experiences shared with me, and if I would be able to do it justice. I once read a book by a Rare Disease Patient titled *"Rare".* Two quotes stood out for me.

"Disease distracts. It fades out everything in the background and shines a light on the centre stage: the will to live, the will to survive, or the will to do whatever the moment begs for. I'm on a journey I don't know how to be on".

This second quote resonated with me during the writing process:

"The real story is spread widely across different people, places and pages, in my mind and deeply in my heart. I have pieced it together here, the best way I know how".

Eventually the feelings of doubt were overwhelmed by purpose and gratitude. I've been a part of the NGO Rare Diseases SA and the Sickle Cell South Africa support group for a few years now. It is through these platforms that I heard fragments of patient experiences that sparked curiosity. The gratitude stems from this opportunity, to be guided by my supervisors whose insights and contributions have been so valuable. They are the most knowledgeable, qualified and yet, humble and kind individuals I got to meet. The interactions with the participants and my supervisors during this research process has helped me to actively listen, craft compassionate connections and elevate the level of empathy. I've grown as a person, genetic counsellor and researcher in ways that I'm still processing. The last sentiment of gratitude is a teaching from the Qur'an that says that a man should leave three things behind, and one of them is knowledge that is beneficial.

Engaging in a mixed methods research blended the inevitable challenges dealing with a large amount of data, the thrill of discovery when data converged, and then, everything finally making sense. I appreciated the versatility of a mixed methods approach, by being able to triangulate data through both a quantitative and qualitative lens. One phase bringing structure and statistical rigor, and the other providing rich narratives that required a significant investment of time and energy that I wasn't anticipating. I appreciate the synthesis more so in hindsight, neither method could achieve such comprehensive understanding in isolation.

In the course of my interactions with participants, examples emerged about the challenges faced when seeking emergency care. One participant recounted an instance where the necessity of administering medication was doubted due to having no visible symptoms, contributing to a sense of judgment and frustration. This example emphasizes the gap in

understanding the unique needs of sickle cell patients and the judgement they face. The participant questioned why she is being judged when she has no control over it and didn't ask for it. In that moment, I realized the depth of patients' encounters and how it affects them psychologically.

I hope that these findings will serve as a catalyst for positive change, inspire targeted interventions and education initiatives to enhance the overall quality of care for sickle cell patients in emergency settings. I hope that these findings can be connected to wider context, add value to stakeholders like Rare Diseases SA and the Sickle Cell South Africa Support Group. I hope that one day, it leads to systemic changes which can positively affect the lives of individuals with sickle cell disease.

It pleases me to know that the understanding of patient experiences can be used to better their experiences, that there are simple solutions that can be implemented. Even though patients are on a journey they don't know how to be on as the quote described, patients and health care providers can work together to help make the journey slightly more pleasant.

Completing this research thesis marks the end of a process that has challenged me mentally, physically and emotionally. Yet, leaving me with newfound knowledge that I hope to leverage in various ways. I hope this research not only has academic merit, but also has a meaningful impact on society.

Appendix F: Research Approval Documents

UNIVERSITY OF CAPE TOWN
Faculty of Health Sciences
Human Research Ethics Committee

Room 45 E-52-E-Floor- Old Main Building
Groote Schuur Hospital
Observatory 7925
Telephone [021] 406 6492
Email: hrec-submissions@uct.ac.za
Website: www.health.uct.ac.za/home/human-research-ethics

28 November 2022

HREC REF: 742/2022

Mrs K Popel
Division of Human Genetics
Department of Pathology
Email: Kalinka.popel@uct.ac.za
Student: Nabeelahpeerbhai@gmail.com

Dear Mrs Popel

PROJECT TITLE: PATIENT PERSPECTIVE OF SICKLE CELL DISEASE CARE IN CASUALTY UNITS OF SOUTH AFRICAN HOSPITALS
(MMEDSCI -CANDIDATE- MS NABEELAH PEERBHAI)

Thank you for submitting your study to the Faculty of Health Sciences Human Research Ethics Committee (HREC) for review.

It is a pleasure to inform you that the HREC has **formally approved** the above-mentioned study.

Approval is granted for one year until the 30 November 2023.

- The student should consider having less prompts for the interview as a larger number of questions may result in having predetermined themes.

Please submit a progress form, using the standardised Annual Report Form (FHS016) if the study continues beyond the approval period. Please submit a Standard Closure form if the study is completed within the approval period.
(Forms can be found on our website: www.health.uct.ac.za/fhs/research/humanethics/forms)

The HREC acknowledge that the student: Ms Nabeelah Peerbhai will also be involved in this study.

Please quote the HREC REF 742/2022 in all your correspondence.

Please note that the ongoing ethical conduct of the study remains the responsibility of the principal investigator.

Please note that for all studies approved by the HREC, the principal investigator **must** obtain appropriate institutional approval, where necessary, before the research may occur.

Yours sincerely

Signed by candidate

PROFESSOR M BLOCKMAN
CHAIRPERSON, FACULTY OF HEALTH SCIENCES HUMAN RESEARCH ETHICS COMMITTEE

Federal Wide Assurance Number: FWA00001637. Institutional Review Board (IRB) number: IRB00001938 NHREC-registration number: REC-210208-007
This serves to confirm that the University of Cape Town Human Research Ethics Committee complies to the Ethics Standards for Clinical Research with a new drug in patients, based on the Medical Research Council (MRC-SA), Food and Drug Administration (FDA-USA), International Council for Harmonisation of Technical Requirements for Pharmaceuticals for Human Use: Good Clinical Practice (ICH GCP), South African Good Clinical Practice Guidelines (DoH 2020), based on the Association of the British Pharmaceutical Industry Guidelines (ABPI), and Declaration of Helsinki (2013) guidelines. The Human Research Ethics Committee granting this approval is in compliance with the ICH Harmonised Tripartite Guidelines E6: Note for Guidance on Good Clinical Practice (CPMP/ICH/135/95) and FDA Code Federal Regulation Part 50, 56 and 312.

UNIVERSITY OF CAPE TOWN

FACULTY OF HEALTH SCIENCES
Human Research Ethics Committee

FHS016: Annual Progress Report / Renewal

HREC office use only (FWA00001637; IRB00001938) This serves as notification of annual approval, including any documentation described below.			
☑ Approved	Annual progress report	Approved until/next renewal date	30.11.2024
☐ Not approved	See attached comments		
Signature Chairperson of the HREC/ Designee	Signed by candidate	Date Signed	18/9/2023

Note: Please email this form and supporting documents (if applicable) in a combined pdf-file to hrec-enquiries@uct.ac.za.
Please clarify your plan for research-related activities during COVID-19 lockdown.
Please use the latest form found on our website:
http://www.health.uct.ac.za/fhs/research/humanethics/forms

Comments to PI from the HREC

Principal Investigator to complete the following:

1. Protocol information

Date (when submitting this form)	15/09/2023		
HREC REF Number	742/2022	Current Ethics Approval was granted until	30/11/2023
Protocol title	Patient Perspectives of Sickle Cell Disease Care in Casualty Units of South African Hospitals		
Protocol number (if applicable)			
Are there any sub-studies linked to this study?	☐ Yes	✓ No	
If yes, could you please provide the HREC Reference number for all sub-studies? **Note:** A separate FHS016 must be submitted for each sub-study.			

(Note: Please complete the Closure form (FHS010) if the study is completed within the approval period)

Western Cape Government

GROOTE SCHUUR HOSPITAL

Enquiries: Dr Bernadette Eick
e-mail: GSHResearch.Request@westerncape.gov.za

Mrs K. Popel
Division of Human Genetics

E-mail: kalinka.popel@uct.ac.za

Dear Mrs Popel

RESEARCH PROJECT: Patient Perspectives of Sickle Cell Disease Care in Casualty Units of South

Your recent letter to the hospital refers.

You are granted permission to proceed with your research, which is valid until **30 November 2023**

Please note the following:

a) Your research may not interfere with normal patient care.
b) Hospital staff may not be asked to assist with the research.
c) Confidentiality must always be maintained.
d) No additional costs to the hospital should be incurred as indicated in your Annexure 2 i.e. Lab, consumables or stationery. **If access to TRACK Care/NHLS is required, kindly attach our letter of approval to the application form and approach Information Management to assist with data.**
e) **No patient folders may be removed from the premises or be inaccessible**
f) Please provide the research assistant/field worker with a copy of this letter as verification of approval.
g) **Should you at any time require photographs of your subjects, please obtain the necessary indemnity forms from our Public Relations Office (E45 OMB or ext. 2187/2188).**
h) Should you require additional research time beyond the stipulated expiry date, please apply for an extension.
i) Please discuss the study with the HOD before commencing.
j) Please introduce yourself to the person in charge of an area before commencing.
k) On completion of your research, please forward any recommendations/findings that can be beneficial to use to take further action that may inform redevelopment of future policy / review guidelines.
l) If the researcher is not GSH staff member, a supernumerary contract is required before commencement of the research.
m) Please contact Michelle Riley (Patient Fees) at ext. 2276 to ascertain if there will be charges for conducting the Research and to obtain a quote or to discuss charges
n) **Kindly submit a copy of the publication or report to this office on completion of the research.**
o) At no time should any posters encouraging patients to partake in research, be displayed within a clinical area.
p) Please adhere to ALL COVID-19 regulations and Groote Schuur Hospital policies.

I would like to wish you every success with the project.

Yours sincerely

Signed by candidate

DR BERNADETTE EICK
CHIEF OPERATIONAL OFFICER
Date: 19 January 2023
C.C. Mr. L. Naidoo, Mr. A. Mohamed, Dr N. Khumalo, Professor N. Ntusi

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