VOICES OF SICKLE CELL DISEASE: STRATEGIES FOR RAISING AWARENESS, EDUCATION, AND STIGMA REDUCTION IN THE BRITISH ISLES, THE US, AND SUB-SAHARAN AFRICA – A THEMATIC AND COMPARATIVE STUDY

Student: Sune Poulsen 20192801

Supervisor: John McClellan

Censor: Nina Trige Andersen

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Abstract

Sickle Cell Disease (SCD) is a genetic disease affecting 20 million people across the world, with 300 million living with Sickle Cell Trait. SCD can be treated but needs awareness from healthcare professionals (HCPs) to help SCD patients receive correct treatment. People affected by SCD live with many challenges in poor environments, facing socioeconomic burdens, stigmas, discrimination and high treatment costs. Previous research has focused on the lack of awareness and education, combined with low interest in rare diseases (RDs) in general, making it difficult to create greater attention and funding for awareness projects. Stigmas and behaviours towards people with RDs are present in the healthcare system and needs attention to improve across different parts of the world. The theories Theory of Planned Behaviour (TPB) and Reasoned Action Approach (RAA) should guide how both SCD patients and HCPs can change their behaviour towards living with SCD, and how both patients with SCD and HCPs can alter their attitudes and behaviour in their behavioural, informational and inheritable beliefs of SCD patients, changing their behavioural, normative and controllable beliefs. Self-care and self-efficacy become important for SCD patients to be aware of, helping them manage potential SCD crisis. This research makes use of an interpretivist paradigm, utilizing the understanding of different realities created by people living similar lives. By being interculturally focused and understanding the lived experiences of five interviewees from British Isles, the US and Sub-Saharan Africa (SSA), different viewpoints on how awareness in cultural awareness can help reduce SCD prevalence are highlighted. Campaigns such as 'Believe It' by Sickle Cell Disease Association of America Inc. (SCDAA) and Forma Therapeutics' increased information advertisement aim to reduce stigmas and highlight the burdens and issues associated with SCD. The research will be a thematic and comparative study, with a qualitative mixed method approach being used. The aim of this research is to have people believe SCD-affected individuals when they share about their lived experiences and how their situation of living is difficult compared to others. Comparing these lived experiences can help future research, campaigns, and projects to aid SCD patients in becoming more aware of the changes from receiving paediatric care to adult care, focusing on mental health and other burdens they might have. Lastly, a discussion focusing on the future research strategies for SCD, along with additional suggestions on reducing the Sustainable Development Goal (SDG) under-5 mortality rate, broadening the availability of care and treatment in SSA, and improving care solutions should be enforced to ensure a brighter future for SCD patients.

Keywords: Sickle Cell Disease, Awareness, Sickle Cell Trait, United States, British Isles, sub-Saharan Africa, stigmatization, education, behaviour, attitude

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1. Introduction

Sickle Cell Disease (SCD) is a genetic, inherited disease (Esoh et al. 2021; Rees et al. 2010), which causes abnormality in the oxygen-carrying protein haemoglobin (Hb) found in red blood cells (NHLBI 2024). SCD is an autosomal recessive disorder (Sedrak and Kondamudi 2023), causing significant morbidity and mortality in people of African and Mediterranean ancestry.

Approx. 300,000 – 400,000 infants are born annually worldwide with SCD (Aygun and Odame 2012; Kato et al. 2018; Williams 2016), and approx. 20 million worldwide affected by the disease (NHLBI 2024) and 300 million living with Sickle Cell Trait (SCT). Sub-Saharan Africa (SSA), Middle East and Caribbean are some of the places where SCD is a common rare disease (RD), and with SCD being a genetic disorder of Hb (Ally and Balandya 2023; Nelson et al. 2023; Manfo et al. 2023), it is easy to contain and mitigate if creating awareness and detect the disease early with the use of newborn screenings (Cortés-Castell et al. 2017). Newborn screening for Hb tests was implemented in the US on May 16, 1972, with the enactment of the Sickle Cell Anaemia Control Act (Benson and Bradford 2010), but was lacking interest in implementation, although being acknowledged as a significant public health issue. Although National Institute of Health (NIH) made a recommendation in 1987 to utilize newborn screenings in all states in the US, it did take 20 years before every state required newborn screenings.

Countries and unions have different perception of when a disease is considered rare. In United States a disease is rare if it affects fewer than 1 in 1,250 people, whereas European Union (EU), United Kingdom and Africa define it when a disease affects fewer than 5 in 10,000 people (equivalent to 1 in 2,000 people) (Ferreira 2019; Beacon 2024; Remuzzi and Garattini 2008; Hristova-Atanasova et al. 2023; Jonas et al. 2017; Eurodis 2024; Groft et al. 2019).

Awareness spreading can be crucial to RD treatments. RDs have a large deficit in experts, often creating uncertainty, ambiguity, and unpredictability when it comes to routine care for RD patients, herein SCD patients. Distrust and conflict in SCD patients has previously been shared to cause issues in patient-healthcare professional (HCP) relationship (Tsyvkin, Riessman and Mathew 2015), due to lack of knowledge, stigmas, and trust in patients' description. SCD patients are experiencing discrimination (Haywood et al. 2014), stigmatization, and mistrust from the medical system, societies and HCPs. With the majority people affected by SCD being of African descent, hypotheses about systematic racism towards these descents are creating a lack of awareness and interest in learning about SCD and the issues in correlation with the disease. HCPs and societies create attitudes and

behavioural reactions on the ethnicity, and the growing racial discrimination can lead to misinterpretation and wrongful medication, lack in pain management, creating a larger burden on the patient coping with the disease themselves (Fain 2021; Knoebel et al. 2021).

Geographic location and socio-economic status of patients are common and potential dangers for rightful access to correct diagnosis and care (Groft et al. 2021). Focusing on differences in cultural, ethical, legal, and social issues should be considered when creating awareness about the correct diagnosis for the families and caregivers of a rare disease, together with the psychological, sociological, and financial/economic burdens that may be part of being affected with SCD (Groft et al. 2021).

People affected by a RD usually form communities for those affected by a RD. These Patient Advocacy Groups (PAGs) consists mainly of one specific RD group, meaning SCD patients and affected, together with a few HCPs and/or specialists in the field of the SCD would be part of one specific group (Groft et al. 2021). PAG collaborates with scientific communities, governments, and pharma in terms of detection, optimal care and research (Stoller 2018). Advocates educate people affected by RDs and fuels understanding and knowledge about their disease of expertise (Nori et al. 2022). With SCD being a common RD, multiple advocacy organizations are founded across the world, but are limited in size and resources. Umbrella organizations such as National Organization for Rare Diseases (NORD) and Global Genes and European Organizations for Rare Diseases (EURODIS) contributes to all RDs through fundraising, clinical trials participation and advisory boards. Furthermore, these organizations oppose policy changes and are regulators or running local support groups (Nori et al. 2022). Smaller PAG groups have also shown impact on these topics (Stoller 2018), but due to limitation might not be able to help in every case regarding their RD.

This research aims towards showing how SCD awareness is depicted by people living with SCD and how the awareness concerning SCD can become better in the future to lower the prevalence of people born with SCD. The research also aims towards sharing the stigmas, burdens and discrimination that is associated with people affected by SCD, and how these differentiate in markets of British Isles, the US and SSA. A discussion focusing more specifically on future aspects of SCD will be performed, utilizing the findings in the research and previous research into understanding how education and awareness can lower the under-5 mortality for people living with SCD.

1.1 Problem formulation

Looking into awareness of Sickle Cell Disease (SCD), how can SCD awareness be raised in British Isles, US and SSA to lower the prevalence and under-5 mortality of SCD patients, together with educate the public and HCPs in SCD education to lower stigmas and discriminations?

1.2 Terminology

The following paragraph will elucidate the concepts connected to the problem formulation and its research, clarifying the understanding of the relevancy of the concepts which will be used in this paper. These have been chosen as being relevant to understand the reason for this research.

1.3 Challenges

Low prevalence in RDs create challenges such as lack of knowledge, scarcity, and expertise, together with their chronic, degenerative, and life-threatening nature (Wakap et al. 2019: Beacon 2024). Because of the lack of knowledge, on average, patients consult 5-10 doctors and physicians, in which they receive three misdiagnoses, additionally receiving the correct diagnosis long time after first symptoms. Most research suggests the time before receiving the correct diagnosis is after 5 years (Beacon 2024; Schieppati et al. 2008), with some researchers indicating under one year after symptoms, some researchers mentioning up to 7-14 years in developed countries. In developing countries, RDs does sometimes never get diagnosed due to the lack of access of care (Choudhury and Saberwal. 2019; Hristova-Atanasova et al. 2023; Krajnovic 2012). This is quite possibly due to physicians' lack of RD awareness, but also the different difficulties giving correct diagnosis to less known rare diseases compared to common rare diseases (Beacon 2024; Schieppati et al. 2008; Li et al. 2021; Choudhury and Saberwal 2019).

1.3.1 Stigmas, discrimination and burdens

When people know about SCD but have only heard about it or experienced others living with the disease, social, cultural, religious, and racial stigmas, associated with the societal context of where people live, and their ethnicity might be created towards these stigmas. The individual encounter between SCD patient and HCP can differentiate considering whether the HCP has stigmas associated with the people who are affected by SCD. Symptoms might be interpreted differently, explanations of pain might not be taken serious etc., potentially given poor clinical device, herein faking the pain that they are experiencing, which can be present in the US with the opioid problems (Pokhrel et al. 2023), showcasing the lack of pain management. Systemic racism and stereotyping African Americans as some with drug-seeking behaviour, disease- and race-based discrimination is

experienced (Bediako et al. 2016). The misunderstandings and perceived race-based bias and discrimination, together with mistrust from HCPs not being the expert in a disease, causes poorer health and mental health, a decrease in adherence from HCPs towards medical advice, and an overall increased mistrust to HCPs (Brody et al. 2006; Haywood et al. 2014). Black individuals with SCD experience can become susceptible to discrimination and racism in their childhood (Blakey et al. 2022).

Understanding the cultural and societal experience is far more interesting for pharmaceutical companies to understand in order to build their awareness towards the disease. Diving into stigmas, societal impacts, discrimination, challenges patients are facing, all based on their location of living with the disease can differentiate due to cultures and communities. SCD is not a typical white man disease and with more people of African descent in Europe and US being affected by SCD, with stigmatisation and racism from HCPs have been mentioned as causes to less help provided to SCD patients (Pellegrini et al. 2023).

1.3.1.1 Immigration and religions

Many infectious and genetic diseases are spread at a regional, national, and global scale due to immigration (Guo and Li 2012), and genetic diseases are mainly clustered due to inbreeding and eventually consanguinity, which is a problem due to larger risk of receiving recessive genes inherited in ancestors (Anwar et al. 2014). The increased immigration to developed countries in Europe, Oceania amongst some should be more concerned about the disease as these typically carry this gene malfunction with them (Kunz et al. 2017; Cortés-Castell et al. 2017). Immigrants migrating to a developed Western World countries, bringing local genetic diseases such as SCD might be limited in awareness and understanding of this disease for HCPs. Immigrants coming to a new country might not know they have SCD or SCT, which can cause higher prevalence of SCT outside its usual locations.

There is no straightforward answer to what religion in Africa is (Meyer 2021). In African societies there is a specific way of understanding God and understanding the World, compared to other ethnicities and cultures, which, although bringing medicine and healthcare from the Western world, might be overlooked compared to traditional African healthcare and rituals (White 2015). Continued use of historical knowledge and practice in order to prevalent or eliminate physical, mental, and social diseases (White 2015) is still present to this day by using ritual and past urban treatments to treat diseases. African religions have several ways of understanding the causes of disease, and a common

contact is the view of the disease being caused by evil or bad spirits (White 2015). Not treating ancestors well is also mentioned as a potential punish to people with disease (White 2015; Magesa 1997; Westerlund 2006), with the balance between the living world and spiritual world being the essential for higher quality of life. Witchcraft is also associated with diseases and being sick in African religions, traditionally those defying scientific pain and treatment. With the SCD symptoms being different from other genetic diseases, not understanding the scientific reasons and treatments to these, portraying families as witches is imminent.

The African traditional health care system has been acknowledged as important for the African countries as their healing processes has advantages for their cultural living experiences, with WHO recognising their importance of continuing these productions of local, traditional medicines (White 2015). With religion and trust in God being present in specifically African cultures, getting exact numbers and newborn screenings, together with SCD management of people in the country carrying the disease, the understanding and interpretation towards SCD can be scarce, difficult, and problematic to comprehend. With scepticism towards the cultural aspect of immigrating to new societies (Edo et al. 2018), religious perspectives and systemic racism can also be presented depending on the situation in new countries Africans settle in.

1.4 Sickle Cell Disease

Humans have six genotypes based on their type of haemoglobin (Hb): AA, AC, AS, CC, SC, and SS. SCD has many genotypes, herein homozygous SCD and compound heterozygous β-globin S and β-globin C being the most common ones. A sickle cell is when a red blood cell takes shape as a sickle, making them easier destroyable, causing anaemia (Kato et al. 2018). If a child receives one sickle mutation, this child will have SCT, and if the child receives both, the child will have SCD (Kassebaum et al. 2023).

SCD can affect multiple organs in the body, potentially causing long term complications, as not enough blood and oxygen can reach organs. These are vaso-occlusion (VOC), which is when red blood cells get stuck in capillaries, attacking the bones. When attacking bones of hands and feet it causes digits or inflammation, other bones will create pain crisis or avascular necrosis of the bones. Sickled cells can get stuck in the cerebral vasculature causing vessel damage such as stroke, acute chest syndrome when clogging lungs, and when affecting the renal papillae, necrosis can be caused (NORD 2024).

Acute complications of SCD includes hepatic, splenic sequestration, stroke, acute ocular conditions, acute chest syndrome, acute anaemia, aplastic crisis, ocular disease, priapism, fever, pain, and multisystem organ failure (Onimoe and Rotz 2020; Adewoyin 2015). Few chronic morbidities, herein leg ulcers are also possible to occur for SCD patients. Acute chest syndrome (ACS) is the most life-threatening complication for people living with SCD.

SCD is predominant in Africa, more specifically SSA, where every 500-2000/100,000 are SCD births, covering approx. 75 % of new SCD cases every year. It is estimated 300,000 births each year are born with SCD on a global perspective, with 240,000 of them being in SSA (Paintsil et al. 2022). An estimated ~100,000 and in United States live with SCD (CDC 2023, EMA 2024), with most of these being African Americans (Hassell 2010; Brousseau et al. 2010; Paulukonis et al. 2014). Furthermore an approx. estimate shows that about every 1 in 365 African American is born with SCD, together with 1 in 13 African American having a SCT (Onimoe and Rotz 2020; Pokhrel et al. 2023). Approx. 300 million people worldwide carries a SCT (GBD 2021).

By 2050, the number is estimated to increase to 404,200 births yearly with SCD, with sub-Saharan Africa carrying this burden with 85% of the cases (Piel et al. 2013). With immigrations happening across the world, more countries are about to getting used to having more SCD patients, spreading to other countries and areas in the world (Nelson et al. 2023).

Patients with SCD are recipients of emergency transfusions and countries without or limited opportunities to provide this, mortality can become inevitable. Patients with HbSS are more at risk at having anaemia crises compared to other SCD patients, as their blood count level are lower than the others. Because of this, the severity of the pain crises does therefore differentiate whether having HbSS or HbSC. Those who have HbSS can be bed sickened for multiple months, having long-lasting complications, with other SCD patients being less affected, only having larger risk of complications when being more active than usually or staying in cold weather conditions. The experiences of symptoms and crises are different from SCD patients and not on the specific type of SCD genotype they have. This is the reason early SCD awareness and tests is necessary to understand and prepare for the eventual future of living with SCD.

1.4.1. Sustainable Development Goals (SDG) and under-5 mortality

Sustainable Development Goals (SDG) were created in 2015 by United Nations consisting of 17 world development goals. SCD monitoring is relevant for SDG 3.1 maternal mortality, 3.2 neonatal and under-5 mortality and 3.4 premature mortality due to non-communicable disease, as the SCD

burden in malaria-endemic regions such as Africa, Middle East, Caribbean, and South Asia is larger (GBD 2021 Sickle Cell Disease Collaborators 2023). Grosse et al. (2011) concluded through their research that under-5 mortality among children born with SCA in sub-Saharan Africa remained between 50% and 90%. In developed countries, more than 95% of children with SCD reach adulthood, which is due to better infection controls, vaccination, and screening programs, together with an intensive disease management, something that developing countries are lacking (Gardner et al. 2016). United Nations and World Health Organization (63rd World Health Assembly in 2010) recognises SCD as an important but neglected problem and have taken an increasingly active interest in the detection and management of the condition (Williams 2016; Ally and Balandya 2023). In 2010 WHO created a strategy addressing SCD as a prioritization in Africa, developing the Newborn screening programs to these areas (Ally and Balandya 2023).

1.5 Treatment and costs

Selfcare is a priority when living with SCD. Hydration and not overextending the volume of exercise, tiring the body, is important to control. A general strategy for SCD affected is to have regular check-ups, staying hydrated, eating a healthy diet and getting a fine amount of rest. Avoiding extreme temperatures is also essential. If these are upheld, pain crisis can be prevented to a larger degree.

Most therapies and treatments for SCD are symptom focused, preventative or disease modifying (Abraham and Tisdale 2021). There are medicines that prevent and/or reduce different symptoms which SCD patients can experience. Penicillin and hydroxyurea prevent and/or reduce respectively sickling of red blood cells, VOC and pain crises. Specifically, hydroxyurea is an oral medicine which experts recommend on a daily use for children and adults if they are haemoglobin SS (NIH 2023). Penicillin has been suggested in the past to be taken twice a day until 5 years of age to prevent sickle crisis (Brousse et al. 2014).

Medications for SCD are in many countries covered by the government, but with governments covering the medications, supply of demand might not be present, and the potential risk of having to buy it themselves after visiting the doctor might in some cases be needed. This can cause major issues in countries with poor communities where there is a lack of care centres for SCD patients, together with limited governmental paid medication.

SCD is one of the few RDs who has a cure. The cure for SCD is a either a stem cell or bone marrow transplant (NHS 2022, NHLBI 2023). These cures are rare, as they also involve significant risks to backfire, potentially causing death. The transplanted cells can end up attacking the other cells in the

body and the cure of bone marrow transplant is only considered for children in extreme circumstances, where other treatments have not helped, with the risk being greater creating overall larger long-term benefits, outweighing the possible risks (NHS 2022). With these being costly and with the high risk, people are still avoiding these cure options unless necessary for survival.

These are all some concerns and issues that is present towards the awareness of SCD. In the next section, my literature review, I will dive more into the stigmas and burdens associated with SCD, together with understanding the reason to why RD and SCD awareness might be limited compared to public health awareness.

2. Literature Review

In the following section, literature regarding awareness within public health and rare diseases (RDs), together with stigmas and burdens associated with Sickle Cell Disease (SCD) will be described. To understand the differences of attention and awareness of both public health and RDs should be understood before diving into what this research can provide. The literature of distinctions between markets and how to utilize the different platforms to create awareness, diving into the burdens, education opportunities and more will be gone through. Living circumstances of cultures, economies and societies is different and should be explained in advance to understand the differences people with SCD live with, considering their "market". After reading this literature review, the reader should understand the differences of markets and focuses on the world, before going further into this research.

2.1 Stigmas and burdens

In the following section, I will be diving into research of stigmas and burdens for SCD affected. These can be crucial to understand for the reader, as having an unknown connection with SCD can portray different perspective to those living with it.

Whenever people living with SCD enter the emergency room (ER) with a pain crisis or other issue they cannot control, there is a greater risk that they are in serious situation of dying. Therefore, delays in receiving care in emergency departments for adults living with SCD is one of the major issues for SCD patients, as delays happens because of lack of knowledge in the seriousness of SCD attacks and the issues they enter the ER with. Not being able to see a crisis, having to objectively interpret a pain crisis can be difficult, although genetics of SCD are well defined. With objectivity and experiences being relevant to prescribe drugs, the difference in prescription of drugs between black and white patients are presented to be stigmatizing (Pletcher et al. 2008), creating potential mistrust, racial stereotyping, and difficult HCP-Patient relationship (Todd et al. 2006). Past negative pain management experiences can indicate SCD patients to delay care-seeking treatment by avoiding ER due to past treatment experiences (Jenerette et al. 2014), relating to stigmatization of patients, with HCPs labelling SCD patients as drug-seekers and similar. Young adults living with SCD have the highest rate of acute care encounters and frequent hospitalizations, being categorized as those with the highest risk of being stigmatized by HCPs (Brousseau et al. 2010).

Burdens of SCD have been studied a lot lately which can be due to the focused awareness of SCD becoming relevant for developed societies. Local research on SCD management, clinical, economic,

and political burdens (Brousse et al. 2023; Adigwe et al. 2023; Nelson et al. 2023; Silva-Pinto et al. 2022; Jenerette et al. 2014), how to control the prevention and treatment of SCD complications and other issues related to SCD management have been focused on more frequently. Diving into understanding the burdens from a personal level on those affected by SCD seems to lack, and with many recent studies focusing on initiatives in their respective country or continent (Russo et al. 2019; García-Morín et al. 2020; Brousse et al. 2023), highlighting the difficulties in personal experiences in different cultures should be investigated more in-depth in the future, as management has been the focus at first, containing and managing those already affected. The lack of awareness and attention towards extended help towards Africa does also mean that they carry a larger burden on under-5 mortality rates (Wastnedge et al. 2018), although clear numbers are difficult to make due to limited data, bad system management and more. Many great examples of clustering prevalence to certain areas are made (Houweling et al. 2005; Burke et al. 2016; Chao et al. 2018) but controlling these and being 100% precise is inevitable due to aforementioned lack of data and bad system managements in districts.

2.2 Public health awareness

Public health awareness can change in a short amount of time depending on crises, both on a personal, social, and/or global level. Global crises and common diseases are more well-known for the public, as these affect more people, having larger attention due to their severity. Health awareness days, focusing on one specific disease has also been around for decades. Although having weeks of raising charity money towards cancer treatment research (just to mention one), health awareness days have been criticized with having issues, herein costs, effect, and usefulness (Purtle and Roman 2015; Budnick 2015). These researchers casted scepticism towards the use of awareness in campaigns concerning charities, with health charities using the word ‘awareness’ twice as much (Budnick 2015). There are more than 200 health awareness days, weeks, or months in the calendar, and is interpreted as an intervention strategy, stimulating thinking and discussion of certain health risks and issues (Purtle and Roman 2015). Searching for ‘Public health awareness’ shows over 150,000 articles on PubMed today, with having record years every year (except 2023), empathizing the potential irrelevancy of public health awareness days.

With the limitations of informational and product advertisements, a medical campaign targeting public health awareness has a high cost, with the risk of the campaign not being effective (Seymour 2018). Seymour (2018) and Masiuliene (2015) defines public awareness campaigns as (a) organized

communication activities designed to raise awareness, (b) induce behaviour change, and (c) improve quality outcomes for individuals and populations. A problem that has been created in terms of public awareness campaigns is that they change people's behaviour when it has a high potential in affecting them or others mental or physical health. There are many information source channels which can help provide information about public health. These can be both beneficial but also manipulative. Apps have been highly appreciated for those living with conditions such as diabetes, being able to perform self-care behaviour (Kebede and Pischke 2019). Video-based learnings and overall electronic resources in terms of publishers such as Elsevier, Oxford etc. adding medical search bases as PubMed with more advanced public health understanding available. Individuals also utilizes social media platforms to share information in regard to health issues (Lopez et al. 2016; Chen et al. 2018). The lack of quality of information on these information might create scepticism in not containing enough evidence (Lavorgna et al. 2018), creating potential fake news, which is also an issue which were highly experienced during COVID-19 pandemic (Naeem et al. 2021; Ali and Bhatti 2020). A different type of awareness which Bragazzi et al. (2017) focused on is the public health awareness towards a disease through a celebrity's death if a specific disease was the reason. They discovered that larger number of searches for the disease and its symptoms was made to understand the disease and reason of death by a famous person raising the awareness of a disease, at least for a short to longer time, compared to years before the death.

Having these limitations, SCD and other RDs which are genetic, are struggling to raise the awareness it needs for patients, the doctors, haematologists and more to understand the disease. Larger scales of physical and mental health in death, illness, and disabilities are far superior in having an impact and awareness of society (Seymour 2018). Financial supports for RDs in both research and development (R&D) and public awareness are limited (Schieppati et al. 2008). The first public awareness concerning the difficulties RD patients went through was raised in 1989 in a report by US Government, six years after the passing of the Orphan Drug Act 1983. Although SCD is concerned as a RD in most parts of the world, it remains a public health challenge in places with high prevalence per person, for instance in a hotspot like SSA (Adigwe et al. 2023) and with limited knowledge and interest from developed countries into RD not affecting their societies, their public health awareness towards prevalence of SCD becomes difficult to change.

2.3 Rare disease awareness and knowledge – education

RDs are a fundamental issue when regarding the diversity and individuality, as RDs might not be perceived as a public health problem, even though RDs in some locations are considered a public health issue due to the number of people with the disease per capita. Most RDs are serious chronic diseases which usually begins in childhood, and might be life-threatening as people indirectly can be caregivers of a RD. People with RD form a sub-population of large dimensions will have limitations on the social level. The economic impact on health systems might also be present, if RDs are not taken seriously. Those who have a RD usually also affects those who are related to them, family, friends etc.

Previous research have shown the need for education about RD in the healthcare system (Groft et al. 2019; Hristova-Atanasova et al. 2023). Evolving their professional growth and skillset by engaging with patient advocacy groups (PAGs) can be essential for future HCPs and their communities for pharmaceutical companies to invest in their RD research and development (R&D) strategies. Fantini and Vaccaro (2019) identify that knowledge of most RDs are insufficient, and even though knowledge on RDs and their epidemiology is increasing (Posada et al. 2017), there is still a lack of interest in RDs and low R&D in this area (Krajnovic 2012). The need of educating HCPs and pharmacists regarding awareness towards RDs, together with stigmas, systemic racism, discrimination, and inequity is presented as evident and needed to eliminate these. Patient experiences and knowledge of needs can be interpreted by HCPs and pharmacists as being either too specific or vocal, creating scepticism towards the patient. Past experiences, attitudes, and beliefs influence how individuals' approach towards new information, learning and decision-making (Catz et al. 2005) will be when visiting an HCP, and with lack of education, combined with people being sceptical on tests, there is a gap in the knowledge RDs and trait status when regarding SCD (Gustafson et al. 2007; Treadwell et al. 2006; Long et al. 2011).

RDs has received much more attention lately and becoming better. National research' has been made but does share the international experiences of the lack of awareness and knowledge on RDs (Hristova-Atanasova et al. 2023; Jonas et al. 2017; Walkowiak and Domaradzki 2021). The burden of future HCPs taking care of RD patients is challenging, as their possess of knowledge around RDs is insufficient or poor, not feeling prepared to take care of RD patients (Li et al. 2021; Groft et al. 2019). Furthermore, lack of interest in taking part in these burden is an issue in terms of creating knowledge about RDs, not developing the awareness towards younger HCPs.

Publications of RDs in high impact journals is difficult, as the lack of awareness about the significance of these diseases are present at editors and reviewers of these publications, making limitations on publications due to fewer specialists that can confirm the publication (Cismondi et al. 2015). HCPs and physicians are mostly aware of the normal diseases, and lack knowledge and understanding of RDs, leading to a higher possibility of giving misdiagnosis to patients with a RDs (Vashishta et al. 2023). Research show both national and international issues towards the lack of access to modern diagnostic methods in knowledge learning on RDs (Walkowiak and Domaradzki 2021).

Li et al. (2021) shows the worrying situations of physicians and doctors' interest in RDs. Most of their respondents only wanted to become aware of RDs that have a cure, situating a larger issue there is within rare diseases, with doctors and physicians not being interested in rare diseases, having patients being the expert (Walkowiak and Domaradzki 2021). Lack of awareness on RDs can be many, both cultural, language, locational, and educational reasons (Hristova-Atanasova et al. 2023; Cismondi et al. 2015; Li et al. 2021). Undergraduate medical educations do not place attention on RDs, creating unknown understanding of these. This can be seen worldwide, with RD education lacking in medical training with no to limited specialized courses on clinical picture, models of disease, therapeutic interventions, and the social and ethical issues around RDs (Cismondi et al. 2015). Campaigns of RDs, creating awareness towards a specific RDs can be beneficial in providing necessary information to target groups (Li et al. 2021; Abdo 2023) but also for experts and HCPs interested in RDs, with exception of those not interested (Vashishta et al. 2023). Previous research results have showcased the limited understanding and knowledge about RD in medical students, both on a national and international level (Hristova-Atanasova et al. 2023), with education on clinical research and experimental medicine, herein its role in understanding and development of novel therapies are not taught (Cismondi et al. 2015). Researchers have found results describing interest from students towards RD elective courses to boost their education, but limited opportunities due to few RD experts make these elective courses to the curriculums difficult to create. Collaboration with stakeholders in terms of PAG advocates and organisations has therefore been considered as a solution in sharing knowledge and expertise, at least on a national basis, as not all RDs are necessary to learn taking the prevalence numbers into consideration with a RD which is not in circulation in a country would be irrelevant to learn about (Hristova-Atanasova et al. 2023).

Solutions and possibilities of creating a better system for RD patients has been proposed by researchers in the past, although critique on lacking initiatives on how to improve the knowledge of health professionals on RDs has been presented (Cismondi et al. 2015). RDs are receiving more

attention from universities but has only happened recently, and only with the most common RDs. Pre-diagnosis centres and centres specializing in RDs, herein gathering clinical and medical genetic experts in specific locations across countries has been raised as a solution, especially for larger countries with further distances to hospitals with access to such experts (Li et al. 2021). This will require multidisciplinary and interdisciplinary clinical experts in order to lower the misdiagnoses. Researchers and HCPs are the key in sharing and increasing RD awareness, herein designing strategies for the specific countries (Vashishta et al. 2023).

Defining quality of health care can be difficult, comparing to general population with individuals expecting high-quality to be the best health care available, which can differ (Williams and Smith-Whitley 2016). With the target being to figure out a way to improve diagnosis and management, research focusing on understanding the natural history, epidemiology, prevalence among the local population, elucidating etiological factors, and drug discovery and development of RDs are necessary. Both researchers and HCPs will have a higher possibility to influence government and policymakers, together with pharma industries if they raise the concern of lack on awareness. They also play a larger role in generating awareness among other stakeholders with their knowledge (Vashishta et al. 2023). A vast majority of patients with RD experience poor or fair understanding and knowledge of symptoms connected to a RD from HCPs (Engel et al. 2013). Creating awareness towards RDs by beginning introducing common diseases in undergraduate medical trainings can improve the core understanding and care of RD patients, both in a sustainable and long-term improvement for both parts and the patient's families (Hristova-Atanasova et al. 2023). Suggestion of implementing RD education for psychologists and psychiatrists involving families should also be considered (Walkowiak and Domaradzki 2021). In previous papers (García-Perales et al. 2022; Verger et al. 2021), better research on the RD has been considered an essential for affected people to improve their well-being, focusing on typology, prevalence, and warning signs of RDs in training processes. Although these focus on teacher's inclusion to have children feel included, it should also be a focus point for HCPs. SCD care have also been presented in earlier research to favor children more than adults when focusing on care and prevention of diseases, saving lives of children with SCD (Williams and Smith-Whitley 2016). Self-care and self-efficacy are presented as critical concepts in association with SCD management (Matthie et al. 2015). Self-care resources mediate the relationship between vulnerability factors and health outcomes. Jenerette et al. (2014) mentions vulnerability factors as lack of sickle cell crisis cue recognition and response, number of complications, number of acute pain episodes per year, and overprotection, and aligns these to have a negative effect to self-care resources,

herein coping behaviour, self-efficacy, which ends up resulting in negative affect on health outcomes such as pain management experience, perceived health-related stigma and similar. The transition from paediatric care to adult care should be prioritized for SCD affected and HCPs, as multiple factors such as insurance (Jordan et al. 2013), financial independence, decision-making experience, future education preparation (Laurance et al. 2006), and interest (Sobota et al. 2011) changes when becoming an adult. Interest and education in low-income areas have also been shared to be present (Sobota et al. 2011).

As mentioned earlier in this literature review, media and society have been argued as a factor in awareness of RD, being more proactive towards RDs (Hristova-Atanasova et al. 2023). The socio-economic burden has been pointed as one of the main issues in awareness, as engagement in young medical doctors' interest in RDs would improve diagnosis, treatment, and follow-up (Hristova-Atanasova et al. 2023; Angelis et al. 2014). Lack of interest in RDs has been demonstrated in earlier research to be highly present in elder and experienced HCPs. Limitations towards accessibility and possibility to introduce RD as a topic is present in many countries and cultures across the world (Hristova-Atanasova et al. 2023; Jonas et al. 2017). For the best futuristic approach in terms of understanding RD, Groft et al. (2019) emphasises, together with many other researchers, that PAGs should be prioritised, collaborating with them so insights and further awareness and knowledge can be shared in healthcare communities. This collaboration also involves university studies which can have RD courses in their curriculum, although elective courses might also be beneficial in the longer perspective to find those who wants to specialize in RDs. Groft et al. (2019) are the first researchers to give concrete examples of organizations that can help in creating awareness towards RDs, both on a practitioner level and a patient level, how a collaboration between these two would be fruitful for the RD and medical societies. If this could be executed, this would create the awareness needed, but with small steps been made so far, and the awareness is still limited. Collaborative positioning of having HCPs join PAGs with medical, scientific, and patient advisory boards, Angelis et al. (2015) mentions these could benefit from each other. PAGs could identify and share patient-driven needs assessments in areas of research, as their key outcomes are important to patients themselves. PAGs can also share awareness from a personal level, creating better overall understanding for publics and other interested outside the medical sector.

With these research focused in the past on opportunities problems in RDs such as SCD, I will share how Theory of Planned behaviour and the Reasoned Action Approach can help provide future insights

into how SCD awareness and education can become better in the future, limiting stigmas and burdens, together with creating better education for medical students.

3. Theoretical Framework

In this research, guiding this study are Theory of Planned Behaviour (TPB) by Ajzen (1978, 2020) and Reasoned Action Approach by Fishbein and Ajzen (2009). With these theories being adding to each other's work, they should be understood as helping and guiding towards a clearer understanding of behavioural factors for this research.

3.1 Theory of planned behaviour

Icek Ajzen was the founder of Theory of Planned Behaviour (TPB) (Ajzen 1978). TPB is a theory which is applied to predict and change behaviour, herein physical activity or drug use to mention some. TPB is used for the proximal determinants of behaviour with behavioural intentions representing a person's motivation towards the conscious plan, decision, or self-instruction to perform a targeted behaviour (Conner 2020). Proximal determinants of behaviour are factors that are connected to the performed behaviour, herein having socioeconomic conditions shaping the personal development, and individual factors like biological characteristics, sex etc. influencing their health and development. The degree of actual control over behaviour depends on the ability to overcome such conditions together with past experiences (Ajzen 2020).

TPB consists of three determinants of behavioural intentions: (1) attitude toward the behaviour, (2) subjective norm concerning the behaviour, and (3) perceived behavioural control (Ajzen 2020).

The first factor, attitude toward the behaviour, consists of accessible belief and subjective evaluation. Attitude toward the behaviour accounts the accessible beliefs of the behaviour's likely consequences. The person's subjective probability that performing a behaviour of interest will lead to a certain outcome or provide a certain experience, past experience can be a factor to consider before making the outcome. When the person has considered whether past experiences make it worth the outcome to perform the attitude toward the behaviour the execution of the first factor has been "made" (Ajzen 2020).

The second factor, subjective norm concerning the behaviour can be distinguished between two types of normative beliefs: injunctive and descriptive. These subjective norms are associated with the social pressures in which an individual feel to perform or not perform a particular behaviour (Conner 2020). Injunctive normative belief is with the expectation/subjective probability that a given referent individual, or group approves/disapproves of performing the behaviour which is under consideration. Descriptive normative beliefs are when others perform the behaviour. They contribute to the

perceived social pressure to engage in the behaviour (Ajzen 2020). The normative belief, whether injunctive or descriptive, contributes to the overall subjective norm in the interaction with the referent's importance or significance to the individual. This subjective norm is therefore directly proportionate to the sum of the products of the normative belief(s) and the importance or significance.

Thirdly, and lastly, perceived behavioural control is based on accessible control beliefs, which are concerned with factors which can facilitate or impede performance of behaviour. Perceived behavioural control has been conceptualized as being a direct predictor of behaviour alongside intentions but have since been conceptualized as moderating the impact of intentions on behaviour. The probability of a given facilitating or inhibiting factor being present in the situation of interest is the control belief. Perceived behavioural control accounts the strength of control and the accessible control factors. The actual behavioural control moderates the influence of attitude and subjective norm on intention, together with moderating the effect of intention on behaviour (Ajzen 2020). Additionally, if there is limited knowledge about the actual behavioural control, the perceived behavioural control can be used as a proxy to predict the behaviour through without knowing the intention.

The behavioural beliefs produce a positive or negative attitude toward the behaviour, herein specifically the valence of each anticipated outcome or experience. These contribute to the overall attitude, producing the outcome or experience in question. A person's attitude should be directly proportional to the strength of each accessible belief multiplied by the subjective evaluation of the outcome or experience. Furthermore, attitudes are normally tapped using semantic differential measures (Ajzen 2002; Conner and Sparks 2015). Conner (2020) critiques the semantic differential measures, as they often focus more on instrumental or cognitive rather than experiential or affective aspects of attitude. Conner (2020) does by the help of previous studies (Lawton, Conner, and Parker 2007; Lawton, Conner, and McEachan 2009) report that experiential attitudes are more closely linked to intentions and behaviour, compared to instrumental attitudes. Belief systems, different functions, experimental manipulations, and empirical differences can be discriminated in these two components of attitude. The importance of the factors is expected to vary across behaviours, populations, and situations, with different understandings and previous experiences can vary (Conner 2020).

Perceived behavioural control and self-efficacy share no differences except the operational difference. Items such as 'rated ability to perform the behaviour' and 'extent to which the behaviour is under my control' are respectively linked to self-efficacy and perceived behavioural control (Ajzen 2020).

TPB can be used to change behaviour in people. Ajzen (2015) mentions five key stages in developing an intervention to change beliefs: (1) eliciting easily accessible behavioural, normative and control beliefs in a representative sample of the target population; (2) selecting specific existing accessible beliefs or perhaps novel beliefs to target in the intervention; (3) designing an intervention that changes the selected beliefs; (4) testing that the intervention produces large changes in targeted belief and does not produce any negative effects on other beliefs; and (5) demonstrating that the intervention had significant effects on the aggregate of behavioural, normative, and /or control beliefs (p. 134). These intervention intentions should in addition use formative research to develop the aforementioned determinants if it regards lack of motivation. Ajzen argues that if the intervention is needed to establish positive intentions but failing to act on them it involves making sure that behavioural and accessible beliefs does not differ, having the skills to perform the behaviour of interest, potential barriers are removed, and no unexpected events or new information leads to change in intervention. If all these can be met, positive and favourable determinants is expected in the changes in beliefs (Ajzen 2015).

Unconscious attitudes can also cause behaviour, which might not have been wanted to be shared. Racist behaviour can occur towards other races without intend, having stigmatized viewpoints (Levy 2013). These consciousness actions made by people are to a slight degree expressed as direct responsible. By acting on implicit attitudes, people express their values, expressing who they are, being justified and able to be held responsible for our actions. People can have values in which they are unaware of, creating actions without the meant intend. Values can be manifested as dispositions, herein asserting claims, emotional responses etc. (Levy 2013). Levy argue that conscious attitudes claim to generate the dispositions which create the stereotype associated with the correlative value of a person, with implicit attitudes activating a different kind of stereotype. Conscious attitudes activate the (normative) dispositional stereotype, while unconscious attitudes activate the everyday stereotype (Levy 2013).

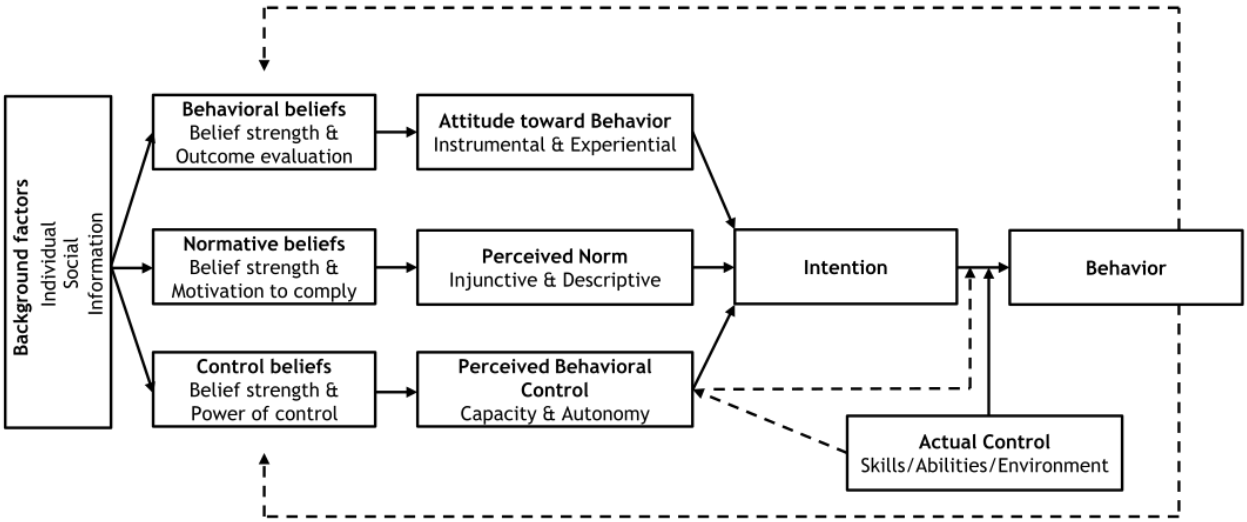
3.2 Reasoned Action Approach

Reasoned Action Approach is the latest version of the theoretical framework of Fishbein and Ajzen (2009). They depict that once behaviour of interest is identified, determinants can start being examined, herein indicating that human social behaviour follows from the information or beliefs people possess about the behaviour that is under consideration (Fishbein and Ajzen 2009). These beliefs can be personal experiences, formal education, informational advertisement, interactions with

friends and family and more. Certain individual differences, herein demography and personality, can shape their behaviour through experiences, influencing this behaviour, creating different beliefs. These background factors can serve as a guide towards understanding relevancy towards the origins of behavioural, normative, and control beliefs concerning a specific given behaviour. Minority groups amongst some who can be influenced by personal prejudice towards a group of people can be affected by reasoned behavioural decision (Dovidio and Gaertner 2000). Prejudice and specific behaviours do although have little correlation between each other, with little to no relation in findings indicating this having a role (Fishbein and Ajzen 2009).

Beliefs about personal and environmental factors can become a factor when attempting to carry out a behaviour. The combination of the attitudes, perceived norms, and perceived behavioural control can guide and determine the intentions and behaviour made by the person. They align to either lead towards a behavioural intention or readiness to perform the behaviour (Fishbein and Ajzen 2009). Although expected to target the same group, different behaviour in terms of population, cultural and religious reasoning makes these three determinants of intention to be expected behaviour. Fishbein and Azjen (2009) adds that: “lack of requisite skills and abilities, or presence of environmental constraints, can prevent people from acting on their intentions. That is, they may lack actual control over performance of the behaviour” (p. 21). This indicates that even if they want to perform a behaviour, the actual control can in specific circumstances be uncontrollable, therefore not possible performing by the individual. The behaviour also depends on the amount of weight placed on the different predictors of intention. The performed behaviour depends on a combination of attitudinal, normative and controllable considerations, which can be the indicator for differences in behaviour, although there is similar attitude, perceived norms and perceptions of control.

A theory with same overall structure as TPB, Reasoned Action Approach suggests that the aforementioned three factors of TPB can be broken down into two sub-components (Figure 2). Attitude towards the behaviour consists of experiential and instrumental attitude, perceived norm consists as the same as with TPB, and Perceived Behavioural Control consists of capacity and autonomy. All of these are intentions, which then leads to behaviour. The capacity and autonomy can besides intentions also be behaviour without any intention. Each of these sub-components have been theorised as independent predictors of intention, but previous research (McEachan et al. 2016) has shown that these can be manipulated individually in order to showcase which one is the most important predictor.



Four elements exist within behaviour: action, target, context, and time. Behaviour consists of an action/behaviour, which is performed on or toward a target/object, in a particular context, at a specified time/occasion. Defining the behaviour depends on these elements as taking (action) medication (target) at home (context) for the rest of their life (time frame) creates a picture on behaviour.

Following background factors, Fishbein and Ajzen (2009) define beliefs as subjective probabilities. Considering a background factor that is handled differently by two people should not be interpreted that other beliefs are different then between these two. Performing a behaviour involving subjective probability leads to a certain outcome. Injunctive normative beliefs are subjective probabilities, which is referred to pre- or proscribe a performance of a behaviour. Descriptive normative beliefs are subjective probabilities towards referents and are or are not performing the behaviour. Control beliefs involves that particular factors that can facilitate or impede performance of the behaviour being present (Fishbein and Ajzen 2009). Knowledge of a certain topic create more informed decisions in line with one's personal preferences, meaning the more we know about, in this case, the treatment of patients with SCD in a given location and situation, choosing the behaviour becomes more accountable.

Fishbein and Ajzen (2009) mention three types of beliefs: (1) observational, (2) informational, and (3) inferential.

The direct observation has an action when behavioural, with a certain outcome follows. Injunctive and descriptive normative belief is respectively directly observing a referent telling someone what

they should do, or what the referent actually does. Control factors involves barriers or facilitating factors. In the case of this research, the encounter with treatment/drugs could result in different observational beliefs.

Informational beliefs include accepting information provided by an outside source, usually from television and radio, online platforms, newspapers, friends, relatives and so forth. Here, the different factors will present themselves differently in terms of how the information is provided and understood.

Inferential beliefs are mainly through observational beliefs. Observing an outcome produced by someone's behaviour, the same outcome will be inferred to occur to the person observing this behaviour. Observing that one's own behaviour does produce a certain different outcome, this may be inferred as another related outcome likely to occur. If a drug/treatment is discovered to help a problem, it might be inferred as providing a better and healthier life.

In addition to predicting and changing behaviour, Fishbein and Ajzen (2009) mention moods and emotions as indicators of attitudes and behaviour. Experiences can make people feel happy or sad, anxious or excited, fearful or frightened. If needed to perform a behaviour in a specific mood can create lack of control, with anger or fear being problematic for HCPs dealing with patients can make it difficult for them to operate with these individuals. Once an unpleasant behaviour and attitude has been created, it might influence future beliefs as negative. If the reasoned action perspective is anticipated to make use of negative affective consequences, unfavourable attitude should be expected and vice versa. Going into a situation knowing your emotions will be negative or positive will also influence the determinants. Emotions can change during a performed behaviour, in which the logic of reasoned action is argued to no longer be applied (Fishbein and Ajzen 2009).

3.3 Final comments

TPB and Reasoned Action Approach are being paid considerable attention, and criticism has been shared previously by many different theorists (Conner 2020; Sheeran et al. 2013; Sniehotta et al. 2014). TPB and Reasoned Action Approach are critiqued as being too rational and deliberative, failing into taking other affective/emotional, non-conscious, or irrational determinants of human behaviour into account. TPB does not propose people as rational/having a rational manner, making no assumptions about the objectivity or veridicality of behavioural, normative, or control beliefs. TPB assumes that non-cognitive, non-conscious, or irrational factors indeed may influence the formation of beliefs, but only assumes that people's attitudes, subjective norms, and perceptions of control

follow these beliefs, creating the way of influence on their intentions and behaviour (Fishbein and Ajzen 2009; Conner 2020).

The theories will be used into understanding how different behaviour across different societies can change and understand how and why the differences from those part of the data is behaving as they are. Furthermore, it will provide help to a greater knowledge on how to depict awareness on SCD in the future, and how change of behaviour, attitude and action can help educate HCPs and medical students in the future, creating better future experiences to be considered.

4. Methodological Framework

In the following section, I will explain my methodological framework, herein philosophy of science, who I am as a researcher, and ethical considerations. Furthermore, data collection methods for interviews and choice of campaign, why these specific interviewees and campaign were of interest will be shared. At the end of my methodological framework, my data analysis method will be described and explained why these have been used for this research.

4.1 Philosophy of Science

My philosophy of science is a combination of my ontology, epistemology, and philosophical orientation. This paradigm will shape my thesis paper's foundations, methods and implications of science.

4.1.1 Ontology

Ontology considers the nature of the acclaimed meanings/realities in relation to the existence of objective reality (Kavanagh et al. 1994). Ontology is the study of being. It concerns about what actually exists in the world, which is available for researchers to acquire knowledge. It's the critical aspect of certainty to existence of objects when researching and the social ontology concerns the nature of social entities (Bryman 2016). The social entities can be considered objective or constructed.

My ontological perspective consists of understanding that people create multiple realities and that there is not one specific reality which creates how it is perceived by other equals. This means for this research that lived and presented experiences of data is not interpretated as being the only constructed world in their social societies. I perceive the reality of lived experiences to be their reality, in which I will respect these as depicted as their own precepted reality. I recognize that the nature and existence of people living with sickle cell disease (SCD) and the realities of them created can differentiate and that the realities shared in this research differentiate and are personal realities. Thereby said, I dive into this analysis with the interpretation that what is told is their reality and might not be true for all realities of people living with SCD.

4.1.2 Epistemology

Epistemology is the theory of knowledge. I myself am concerned with all aspects of validity, scope, and methods of acquiring knowledge. These can be what constitutes a knowledge claim, how can knowledge be acquired or produced, and how the extent of its transferability can be assessed (Moon and Blackman 2014).

An interpretivist paradigm will be used during this research. I understand that people living with SCD have different lived experiences, and that multiple realities exist, hence my reason to analyse interviews with SCD affected and an information campaign created about SCD.

Learning about the individual's feelings, perceptions and experiences of reality, together with believing the individual's interpretations of reality affecting how they interpret their social world to be the cause, I will interpret them as experts of their own experiences and therefore understanding their individual realities of their lived experiences is the goal of this research. Without knowing how SCD issues might relate on a personal level, equipping knowledge through research papers, interpreting social realities, the interviewees are interpreted by myself as experts of known knowledge about SCD. Utilizing their voice in the work is important in reflecting their words, understanding the subjective meanings of individuals and how these attach to their lived experiences. Therefore, understanding their social worlds and not changing them is the purpose. Considering differences in terms of cultures, circumstances, together with timing is leading to different social realities. In addition, analysing the information campaign and comparing it with the perception of the interviewee's world of SCD patients, I will provide information on how the perspectives, behaviour and overall depiction of the world is comprised from their knowledge. Besides interviewing these individuals, equipping lived experiences is difficult to retrieve through academic papers, in which I trust their knowledge to create a social reality of people living with SCD.

4.1.3 Philosophical orientation

According to Byrne (1998), the ideal interpreter can discover exactly what they believe and desire. The logic of understanding and reflecting people involved in this thesis to be humans against the natural order, having the social science being different from the natural science (Bryman 2016). Analysing human behaviour and changes of these relying on qualitative measurements, herein their feelings towards a subject will be the purpose of this research. Working as an interpretivist I will interpret and understand the different social, behavioural, and cultural variables of the data collected as to understand the reasons of their rationale to the performed behaviour. By interpreting reality as culturally derived and historically situated (Moon and Blackman 2014), utilizing hidden meaning and understanding of language in texts, practices, situations etc., together with understanding the human behaviour, a hermeneutic approach will be taken during this thesis paper. With the involved people having a social understanding, I will interpretate their social scientific frame and interpretation of the subject of SCD awareness (Bryman 2016). I understand that how I interpret the interviews and the

informal campaign can differ to others interpretation of the conducted interviews and informal campaign.

4.1.4 Qualitative researcher

By being a qualitative researcher, I will conduct investigations that use qualitative methods to collect, analyse and interpret data. I will gather rich, detailed information which can provide insight into the meanings, concepts, definitions, characteristics, and description of things in regard to SCD. I will gain a deeper understanding of human behaviour, lived experiences, and social contexts of my data. Showing empathy with interviewees talking about sensitive topics through their lived experiences was respected, and interviewees were recommended to sign a release form in advance of the interview as an ethical consideration of using their shared experiences and information. Emotions and motives from participants are important and highly valued assets for the data collection and should be mentioned when present. Having different personalities and perceptions of SCD, I wanted to have different and alternative interpretations and explanations in order to avoid the research to become rigid in its framework.

When inquiring people for this research, the goal is to use their experiences as a resource, as my knowledge and understanding is limited in lived experiences of people affected by SCD. The key is to tell a good story, focusing on meaning over measurement, making us human and makes us gain knowledge and experience from lived experiences.

The purpose of being a qualitative researcher for this research is to collect a different and newer perspective on the awareness opportunities and lived experiences with SCD, whether for patients, healthcare professionals, or students, educating to become doctors, scientists etc. in the US, British Isles, and SSA, as I want to discover their potential cultural perspectives and differences, herein understanding their lived experiences, how they interpret the future opportunities for people affected by SCD, together with shared behaviour towards living situations aligned with SCD.

4.1.5 Ethical issues and considerations

As I will be using a mixed method qualitative approach by analysing and comparing the ‘Believe It’ campaign and five interviews, asking questions on a sensitive topic and personal experiences, ethical concerns should be remembered. Having SCD affected talking about their experiences and crises are a soft topic to dive into, and in order to recruit candidates for interviews, a release form (Appendix 1) stating the ethical considerations was presented and submitted with and by interviewees, herein

using pseudonym and deleting evidence of transcription and/or video recording immediately after 26 June 2024. Additional use of their experiences has been individually agreed whether possible to use in further research.

4.2 Data collection methods

In this research, interviews of five people and an information/awareness campaign was collected. These will supplement each other, diving further in to understanding the relation between the lived experiences of interviewees and the lived experience shared in the information/awareness campaign.

4.2.1 Interviews

I performed five interviews with participants from British Isles, SSA and US. These were performed during April 2024 and was made through online platforms of Zoom, Microsoft Teams and WhatsApp calls. The interviewees were chosen through specific criteria. Firstly, and most importantly, they had to be a person living with or highly affected by SCD, as for them to give their own lived experiences of being affected by SCD. Secondly, being a patient advocate, having a voice to share during press conferences or workshops for other SCD patients was important, as they then had experience sharing experiences. Thirdly, finding people with expertise in different countries and how it works in their surroundings and culture was necessary to choose them as relevant.

The recruitment process consisted of two processes. The first process consisted of myself reaching out to SCD advocates, finding these through Internet research and past workshops I had participated in regarding SCD, in which I then reached out to multiple on LinkedIn, having few of the people reaching out to confirm their interest in my research. The second process consisted of me reaching out to Sickle Cell Society UK to receive candidates for British Isles and SSA. This included email conversations with a representative, who then requested a one-pager (Appendix 15) to be made and sent to their WhatsApp group for them to learn about earlier mentioned purpose of this research. From there, they received contact information and reached out to me, sending them the same mail to these as did with those accepting through LinkedIn.

Following guidance of Brinkmann and Kvale (2018) on how to conduct a semi-structured interview, when reaching out to interviewees, the formulation of my research question and my theoretical clarification of the theme investigated was provided, herein the purpose of my study, my obtained knowledge about the subject matter in advance, together with sharing the relevant topics of the interviews to contain. Knowing that the interviewees were talking about sensitive topics, I made sure

that they could use as much time as needed, together with having a calm and open beginning and ending of the interviews, making sure that they were agreeing with using the interview was fine by them.

Questions such as “Have you experienced any type of stigma regarding SCD, and if so, how did you manage to make people understand and eliminate these stigmas” and “Can you share a good and bad experience of HCP-Patient experience?” were asked in order to answer my research question and help broaden the later interpretation of the ‘Believe It’ campaign. While the interviews were conducted, a confirmed recording and transcription section was done during the interview. When recording the interviews, live transcription by Zoom, Microsoft Teams and WhatsApp programmes was made and used. With these transcription tools being flawed to a degree, I myself afterwards watched the recordings again, familiarizing and coding the interviews, correcting words and specific types of speech, indicating parts which were unrecognizable (herein quotations).

This data consisted of five interviews with interviewees being between age 21-70s. The interviews lasted a total of 6 hours.

In regard to the recruiting process, payments for using their time to talk about SCD and lived experiences were made. There was no funding for this work, personal funding throughout this work process was made.

4.2.2 Believe It Campaign

A campaign can vary and consists of different purposes and can be political, advertising, project, informal to mention a few. Political campaigns contain the ideas in which the candidate wants to share with voters, whereas advertising is a series of advertisement messages that share a single idea and theme. Public awareness campaigns can be associated with advertisement campaigns, but instead of product, it is instead targeting a purpose and information rather than selling a product. These campaigns tend to require significant funding and might end up not being effective (Seymour 2018). The campaign “Believe It!” by Sickle Cell Disease Association of America Inc. (SCDAA) and Forma Therapeutics were used for the purpose of answering the research problem and comparing with lived experiences shared by interviewees mentioned above. The campaign consists of two types of public service announcements (PSA) with the goal being to drive awareness and create meaningful understanding of SCD to people in the US (Business Wire 2022). This campaign was chosen as it targets both experiences in daily life at school but also the experience and awareness at hospitals towards SCD affected.

This campaign was made for the US public and were not representative worldwide. The two PSAs were available for the public on different platforms. The first PSA is a radio PSA (Appendix 3), which was shared at radio stations across US for a limited time. The second PSA was a digital, broadcasted commercial of respectively 30 seconds and 100 seconds (Appendix 2). The 30 seconds PSA has been excluded, as it is collection of clips from the 100 seconds PSA. The PSAs were collected and analysed to compare the similarities or differences that is described in the PSAs, additionally analysing the campaign, and judging whether the content of the campaign was correctly made for its audience in making people aware of SCD together with taking action. Visual and textual analysis was made with the use of a reflexive thematic analysis, which I will describe below.

The radio PSA was shared on Soundcloud (Soundcloud 2022) on 26 September 2022 and radio stations across the US for a limited time. It consists of an audio message with four questions and four statements, ending with a reference to learn more on scdbelieveit.org. The 100-second video PSA was shared on 21 November 2022 on YouTube (YouTube 2022) and depicts John, a student living with SCD, showing how a normal day could look like for a person living with SCD. We follow the development of having a pain crisis, from its beginning to being treated at a hospital bed. Both of these were accessed during March 2024, and was free of charge.

This campaign was chosen because it depicts a lived experience of a SCD patient, together with informing viewers and listeners, both affected and unaffected, how living with SCD is, trying to eliminate the cause of stigmas, discrimination and more associated with affected and unaffected people.

4.3 Data analysis

Reflexive thematic analysis was used to access the two campaign texts and the five interview transcripts. By having two thematic analyses of two different data, a qualitative comparative analysis was also used to find similarities in lived experience and shown/presented lived experience.

4.3.1 Reflexive thematic analysis

The reflexive thematic analysis by Braun and Clarke (2006) will be used for this research. Reflexive thematic analysis is a theoretically flexible interpretative approach to qualitative data analysis, identifying the patterns and/or themes in one's data set (Byrne 2022; Braun and Clarke 2012).

A thematic analysis can be semantic or latent, and inductive or deductive (Caulfield 2019; Braun and Clarke 2006; Braun and Clarke 2019). I will in this research make use of an inductive approach,

together with a latent coding. Through an inductive approach, my focus is to adopt a data-driven approach to produce codes which solely reflects the content of the data I collected, without any pre-conceived theory or conceptual framework taken into consideration. Braun and Clarke (2013) suggest the coding of the data to be open-coded, in order to represent the meaning of communication from participants to its best, in which I followed throughout the analysis of the data. The themes I identified are strongly linked to the data with data collected specifically for this research. Coding the data without fitting it into pre-existing coding frame or my preconceptions were performed in order to follow an inductive approach. By using an interpretive approach to the analysis of the data collected, reflection on the dataset, the theoretical assumptions of the analysis, together with the analytical skills and resources of the researcher is considered (Braun and Clarke 2019; Byrne 2022).

I will follow Braun and Clarke's (2006; 2019) processes in a reflexive thematic analysis, which consists of six steps: (1) familiarisation with the data, (2) generating initial codes, (3) generating themes, (4) reviewing potential themes, (5) defining and naming theme, and (6) producing the report.

The familiarisation phase derives on collection of the data, whether collected interactive or by oneself. Re-reading the data is time-consuming but should be prioritised in order to fully understand what has been said, the connections between different data etc. Creating an early overview and marking potential themes is essential, as the coding will develop and defined throughout, and not in the moment. Transcription of interviews are mentioned as the key phase of data analysis within interpretative qualitative methodology (Braun and Clarke 2006; Bird 2005), in order to get a firm understanding of the data, herein the interview(s).

My familiarization process is divided in two parts, a generated transcription section of interviews, whereafter correcting spelling issues and familiarization with the text was done, still managing to familiarize myself with the interviews. During the corrections of the interviews, potential themes were being marked during this part.

Phase two is generating initial codes. Here, after the familiarisation, an initial list of ideas on what to use in the data and the interest about them will happen. This process of coding is part of the analysis, where organisation of the data into groups will occur. Themes will then be developed in the next phase when initially coded.

With different perspectives, locations, and cultural heritages being familiarized, finding similarities of themes with a latent approach was performed, trying to identify and examine

underlying ideas, assumptions, and conceptualization. It became interesting to see the difference in experiences comparing the difference in genotypes, with all interviewees still being relevant, but with different associations to the awareness, experiences and stories of SCD.

Third phase is searching for themes. Here, the themes found in the coding will be analysed and considered whether relevant or not, which sub-themes there might be within the larger themes, which themes connect with each other etc. Relationships between codes can be essential to figure which are more relevant than others for this paper, the relationships between codes, themes, and different levels of themes.

With the interviews being conducted and transcribed during, in order to keeping the interview fresh, searching for themes was done separately in every interview after conducting each single one, mapping the themes and relevancy, in order to prepare for phase four.

Phase four is the reviewing of themes. Here, themes have been selected and placed as candidates and non-candidates for the research problem. Themes that were thought of being themes can be reviewed as non-relevant during this phase and consists of two levels of reviewing and refining themes. It involves reviewing the coded data extracts for each theme and the entire data set. The essence of this phase and re-reading the entire data is to figure out whether the themes work in relation to the data set and whether potential themes have been missed or misinterpreted.

Mentioned in phase three, different thematic maps were created after each individual interview and when last interview was conducted, finding the overall themes were performed with some being more relevant than others, and some being problematic for the research. These were then removed and not considered to dive into any further.

Phase five is defining and naming themes. This means identifying the essence if each theme, present and identify the interest of the themes and why. A detailed analysis should be written to each theme, creating a *story*. The individual themes create a small story within the overall story, which is telling in the data and its relation to the research question/problem, having not too much overlap between themes. At the end of this phase, the themes should be clearly defined.

Multiple themes were found, and sub-themes to these were also created, with some themes being defined as bigger and broader than others, but necessary to depict the overall story to be told.

Phase six is the production of the report and starts when all of the aforementioned phases have been completed and ready to be written in the analysis section. The analysis should provide: "*a concise, coherent, logical, non-repetitive and interesting account of the story the data tell / within and across themes.*" (Braun and Clarke 2006, 93). The data extracts need to be embedded within an analytic narrative which illustrates the general story, in which should be told about the data, going beyond description of the data.

For the campaign 'Believe It' by SCDAA and Forma Therapeutics, a reflexive thematic analysis with focusing on text, visuals and sounds was used. Focus on familiarizing the use of sound and visual effects, combined with textual framework, searching, reviewing and defining themes was done separately between the video and radio PSA. The outcome of finding themes by using visual effects and language will be supporting the themes of aforementioned themes that will be found in the interviews conducted.

4.3.2 Qualitative comparative analysis

A qualitative comparative analysis will be made by comparing the themes found in the interviews and campaign, giving insights and similarities between and amongst interviewees and the campaign. This is important to understand in order to analyse what would be the way to frame awareness in the future, and whether the lived experiences were depicted in the campaign.

Charles C. Ragin (1987) proposed the use of theories to interpret and analyse qualitative data of causal relationships. Qualitative Comparative Analysis (QCA) is used to define commonalities in an observed data set, investigating different combinations of causally related condition variables, which are essentially related to a certain outcome (Ragin 2008). QCA assists in finding patterns and sense-making of the variables in the collected data (Selcuk and Cizel 2023), that was mentioned earlier in the method section. This helps in going deeper into understanding the change of planned behaviour that might be happening for people living with SCD.

From the findings made in the previous mentioned thematic analysis of the interviews with SCD patients, parents, and HCP professional, together with an enhanced thematic analysis analysing textual and visuals elements in the SCDAA and Forma Therapeutics video and radio PSA, will these two cases be compared to find causal inferences, finding relations between the variables on these collected data and findings.

With the two conditions of being able to make a comparative analysis (two or more cases, and an attempt to explain rather than only describing) (Pickvance 2001) being fulfilled, multiple objects of comparative analysis can be done. Pickvance mentions two strategies: 'Variable-oriented strategy' and 'case-oriented strategy'. I will follow the case-oriented strategy, appreciating complexity, unravelling historical conditions which produces different historical outcomes, seeking invariant statements relevant to more narrowly defined categories of phenomena, and assumes presence/absence causation (Ragin 1987; Pickvance 2001). By comparing the PSA's and the interviews, I should remind the readers of this thesis paper that the context and location of the PSA's is US, and with multiple different interviews across the world conducted, comparing the similarities and differences should be taking into consideration when referring to a specific interview, which environment is normal for that person, as it might be completely different from the PSA's, creating more independent and uncontrolled variables (Pickvance 2001). The comparative analysis will examine the relationship of both cases, whether they correlate with each other, explaining similarities between the two findings.

By using this methodological framework, I will present the findings from this reflexive thematic analysis to answer this paper's research question through my use of philosophy of science towards this research. By interpreting the hidden meanings and underlying ideas, assumptions, and conceptualizations of the interviews, the aim of this research is to share a more personal perspective on how SCD awareness is, and how different themes within SCD is relevant to depict and touch on in this and future research to gain understanding and knowledge about the research problem.

5. Analysis

The following analysis will make use of the aforementioned data collected in the methods section. With few interviews conducted from specific places around the world, a critical reminder to state is that as it is only few experiences, stories and awareness ideas shared, these should be and will be analysed with an interpretive approach, together with using Braun and Clarke's (2006) reflexive thematic analysis. A visual and textual thematic analysis approach for the 'Believe It' campaign will also be done. Finally, a comparative analysis of these analyses will round up this analysis section, before answering the research problem stated in 1.1. This analysis/findings section will finish by answering the research question looking at the interpreted awareness on sickle cell disease (SCD) from lived experiences.

5.1 Interviews

By making use of a reflexive thematic analysis method, six main themes were analysed by coding through all five interviews. The themes which were found and will be analysed are (1) Overall SCD Awareness (2) Paediatric v adult care (3) Attitudes and Beliefs, (4) Systemic Racism (5) Experienced Stigmatization, and (6) Experts through experience, being US and UK specific.

5.1.1 Overall SCD Awareness

Having three different locations across the world to analyse in this research, diving into the awareness in specific areas should be utilized too. Therefore, in the beginning of this section, I will analyse specific awareness experiences in British Isles, the US and Sub-Saharan Africa (SSA) shared by interviewees. After I have dived into these, I will analyse the overall Patient/HCP/Employer relationship and education, how this will play a role in the future of change of behaviour.

5.1.1.1 British Isles Awareness

The awareness on SCD is experienced as being lower and more centralized to certain places and areas across the British Isles, specifically larger cities (Appendix 11 16:52-18:00). The interviews of two SCD affected, each with different experiences and age difference (parent v young adult), the contrast of previous experiences and the potential change in lack of focus on awareness is interesting. The interest from HCPs is presented as being limited mainly due to limited time in the emergency room (ER) (Appendix 11 21:38-22:00), having to be on the move all the time being a hindrance for the educational understanding and awareness around SCD whenever they enter the ER. Additionally, there is limited interest in learning about SCD.

Both have experienced students and/or younger personnel being interested in learning about SCD, but as Nicole and Julia mentions: “A student who came and just have a chat with me and it’s difficult because it is a case of … if you do meet someone, at what point do you have a conversation about what are you doing, do you suffer” (Appendix 11 57:30-58:01) and “I did have [experience] was a bit more pleasant. The doctor actually asked me …, “I don't know much about sickle cell” … Can you please explain a bit more about it to me” (Appendix 10 25:38-25:57). This indicate that there are few healthcare professionals (HCPs) interested in SCD together with lack of awareness and understanding of SCD, together with few encounters for the HCPs with SCD patients (Appendix 11 48:45), making it difficult to learn through patients.

Lack of funding makes it difficult to have larger focus on SCD on the British Isles. British Isles does have Sickle Cell Units in the bigger cities (Appendix 11 45:28-45:39), but are due to fundings, herein which medical areas should be prioritised, SCD areas are not staffed 24hr (Appendix 11 27:18-28:00). Nicole shares the experience of having to go to the ER on a weekend as: “I was left for … 36 hours in A&E, and I worsened in A&E when I went in. I mean it was my fault as well because it is known that more people die at the weekend … if you go into hospital in the weekend … you don’t the same attention treatment” (Appendix 11 18:01-18:50).

A case of experiencing death while in the emergency department was also shared, depicting the lack of awareness on the seriousness of SCD affected experiencing a crisis. Julia tells: “the worst experience that I had … associated with my sister … she was in the emergency department for hours … had terrible symptoms, legs felt really numb and heavy vomiting, diarrhoea, high fever … I don’t think it was tended to as urgently as it should have been … less than 24 hours she’d passed away” (Appendix 10 1:06:58-1:07:27). Because of experienced bad incidents in sickle cell lives, together with stories being shared, containing consequences on the lack of awareness, has raised the awareness in Ireland (Appendix 10 11:08-12:05), indicating that multiple bad situations happening to people affected by a specific disease creates attention towards it. Indicating that sharing cases of previous bad experiences to the public can make awareness and attention towards SCD better by sharing the issues and experiences.

5.1.1.2 US specific Awareness

SCD is affecting 100,000+ Americans and is also one of the most common RD in the country (1 in 3,300 American) with 1 in 365 African American living with SCD and 1 of 13 having a sickle cell trait (SCT). With campaigns being made specifically highlighting the SCD, together with having a

higher history of interest in screening newborns (Benson and Therrell 2010), the attention and awareness of SCD is still low in the US. Justin argues that: “awareness has gotten better, but it’s not nearly what it needs to be … how patient … people in the community understand what the risk factors are … I have teenagers coming up [at events] that don’t understand what [SCT] status means” (Appendix 12 51:21).

NCAA has implemented mandatory SCT testing for Division III and above players, making sure of making athletes, coaches and organizations aware in regard to medical conditions associated with SCT (Appendix 12 55:30-55:58). The possibilities of making people more aware of SCD and SCT is more plausible with multiple options in terms of direct-to-consumer advertisements promoting, information campaigns, with using products to treat SCD and the opportunities there are to handle the daily life as a SCD patient better.

The medical system is non-beneficial in the US for SCD patients having a crisis, as hospitals are working to get patients out as quickly as possible (Appendix 12 17:32), for them then to be re-admitted when getting the symptoms again a few days later. Furthermore, Justin mentions that: “We don’t in the US have a lot of adult centres for sickle cell patients specifically … the situation is still being viewed as drug seeking or having delays in our treatment … having a lot of unnecessary lab tests … even with the increased awareness” (Appendix 12 12:33-13:31). They are connected to the overall stereotypical assumptions due to lack of knowledge and awareness. Lack of awareness can be indicational on the lack of education about SCD with medical students given less than a day of training (Appendix 12 13:20-13:31). Justin calls for the attention towards better awareness for the public to understand living with SCD and SCT, as he experiences many not knowing what to be aware of when being a carrier of SCT. Justin explains the concerns of awareness by mentioning that: “even if both parents have the trait and they have one or two children already that don’t have the disease they think I can’t have a child with the disease … they don’t realise that chance of having a SCD baby” (Appendix 12 58:28-58:58). The lack of attention towards people with other native language besides English should also be considered to need more attention, with campaigns and talks about SCD being on a different language than their native language, an issue that is problematic for both patients and HCPs in providing the needed information and treatment (Appendix 12 1:02:46).

5.1.1.3 SSA specific Awareness

Awareness in SSA should, compared to awareness in British Isles and US, be completely different on who should be targeted and how they should create the best awareness in this place in the world. This

correlates together with two different aspects: (1) The number of people affected by SCD, and (2) culture/religion. Mentioned earlier in the introduction, most people living with SCD are born and raised in SSA, and with a more religious perspectives to daily situations, believing that God has a reason to why Africans are victims of SCD is one viewpoint (Appendix 14 22:41 and Appendix 13 4:10).

Differentiating from awareness in British Isles and the US, SSA countries should target awareness campaign towards the entire population and those affected by SCD. With religious conspiracies involving SCD, making people and communities understand the situation of SCD will help them limiting the accusations towards those living with SCD, comparing those with a SCD child as witches. Face-to-face campaigns, targeting the religious population at churches, travelling to districts and provinces outside of greater societies is the best way to target the population sharing knowledge, challenges, and understandings of SCD and being a carrier of SCT (Appendix 14 50:42). The face-to-face campaigns should be done with international collaboration between developed countries or organizations and countries in Africa, as the costs and funding for travel expenses and treatments is one of the biggest problems in Africa. SSA is a lucrative market for World Health Organization (WHO) to place a greater focus in order to lower the prevalence of under-5 mortality rate, educating families on how to treat children with SCD, and preventing those with a sickled gene to spread the gene malfunction to their future children (Appendix 14 08:46-11:42).

SSA are lacking personnel compared to the number of affected. A better awareness system should be implemented towards making people understand how to handle living with SCD and SCT, eliminating any religious stigmas towards people living with SCT. The overall interest in SSA as a market in general is portraited as problematic with lack of organizations interested in helping being evident.

5.1.1.4 Patient/HCP/Employer relationship

A common problem for those living with SCD is the distance and location from the SCD patient to the place where treatment is provided which can cause patient/HCP relationship issues. For SCD patients they will have to find a paediatrician, haematologist or general practitioner (GP) that they trust and are able to contact if they are going to the ER (Appendix 10 24:03) or have other complications, in order for them to gain a trust to the medical system (Appendix 14 22:41).

It is shared that HCPs find SCD patients to be difficult to deal with. With SCD patients experiencing different reactions to pain crises and similar, knowing when they are ready to be sent home, wanting to have them in the hospital only when in pain, having it recurring afterwards can create tension

between the patient and the HCP. With limited attention and understanding of the disease from many HCPs, SCD patients rely on meeting someone in the ER, who understands the seriousness of their condition, in order for them to get faster treatment. There is no specific fast track for SCD patients as there is for instance with cancer (Appendix 11 25:21), and if the HCP does not understand the potential serious issues that might be present for the SCD patient, long waiting time with no/limited treatment while waiting to be called up does occur then (Appendix 12 10:15-10:51; Appendix 11 16:52-18:00).

With SCD patients being expert in what type of medication they need, they can be interpreted and labelled as manipulating the system into trying to receive specific drugs, with HCPs questioning their trust in the patient (Appendix 10 19:19). Coming up to the HCP or paediatrician and ask for a specific drug and type of over-the-counter medication can be difficult for them sometimes without having the HCP showing mistrust in the SCD patient having them being *drug seeking* people (Appendix 12 29:22, Appendix 13 16:15) or not believing that they have sickle cell (Appendix 12 9:11-9:35).

The relationship between the SCD patient and place of employment/school is also crucial for the SCD patient. Those living with SS genotype will find it more difficult in their youth due to their more frequent and longer time with recovering from a sickle crisis, having more and longer hospital visits, and if not talked about with school principal, teachers etc. falling behind in the system will occur. With SCD problems occurring mainly as young adults, attention towards extra educational awareness around exams should be presented. The situation is the same for adults who are employed. Those who are paid hourly is more affected by hospitalization and pain crises as they are not paid if ending up in a situation where they cannot work (Appendix 12 37:01 – 37:40), and those who have longer recovery time can end up quitting or losing their job due to lack of presence and work (Appendix 13 22:54), which is more common for these compared to those having shorter time of sickle sickness. Having the individual sharing that they have SCD can help them in having coworkers, principals and teachers understand the issues that might occur to the individual.

5.1.1.4.1 Mental and psychological challenges

Mental and psychological challenges are heavily present for those living with SCD, whether themselves or as a parent. The unconsciousness of not knowing that it's not normal to be hospitalized occasionally (Appendix 10 5:55), or the overall experience of missing out on events, friend groups or similar in their youth can scarce backlash into how they become who they are. SCD patients lack the opportunity of independency, not being able to travel around the world without having in mind of the

risk of having to visit a completely new hospital/ER section, which have limited to no knowledge about SCD (Appendix 10 24:03, Appendix 11 51:39, Appendix 14 22:41). They will have to consider every action they take, and life development in what they have to go through at school, work and similar situations can become worse for these. They will have to balance their stress levels in order not to get pain crises (Appendix 13 4:10), and with education and doing tests and exams, their SCD might be more affected in their youth. Having these seeming controllable Christie and Melissa shares how stress in school is problematic.

"*And in the university, you know, even some get to the university finish exams final year and they die … when he was in final year when he was taking the exams. You know, when they have overworked themselves. Too much stress exam*" (Appendix 14 46:10) and "*I remember back in university; I missed so many exams because as a result of stress … I was so aware of my condition, so I was barely existing in probably was and most of the time was during exams. So it was exam stress that triggered me a lot*" (Appendix 13 4:10).

Experiencing death in family due to SCD, herein bad pain management, does also create trauma and anxiety into going into emergency if same symptoms happen to the individual (Appendix 10 1:07:06-1:07:44). Having the psychological thought that the disease might not be treated as life threatening is a lived experience SCD affected have to live with. Christie provides an example of a lived experience from a parent with a SCD patient, stating the mental difficulties: "when he came back the son died … the man couldn't live. He couldn't come out of his room … man went into crisis … son was blaming him … you people knew that you were both carriers, you brought me to the world to suffer … Parents can't stand the pain which their children are going through, adding that "he wanted to commit suicide because looking at his child going through this kind of [problem]" (Appendix 14 47:35 and 48:47).

With all of the interviewees living with SCD being Sickle Cell Advocacies, they also explore a lot of mentally and psychological draining challenges by helping families and sharing understandings and medical information about SCD. Justin (26:59-27:30) refers to the loss of other advocates as a mental struggle as they look up to those who are older than them to know the future opportunities. They have to help those who are unconscious of what to do with SCD, how they can live with it and what the future looks for them, things that should be explained and taught at an earlier stage at paediatric care when they got to understand that they will be affected by SCD in the future. The shift from paediatric to adult care also affects the advocates in taking care and potentially helping those people making this

change financially getting through their situation, which is physically and mentally draining them (Appendix 12 49:02, Appendix 13 1:01:19, Appendix 14 08:46), showcasing the need of medical experts in these circumstances.

Mental health is not being paid enough attention to and should in the future be considered to have a higher focus. Justin mentioned: “I don’t think mental health is talked enough about with sickle cell … mental health is definitely needed for sickle cell patients” (Appendix 12 25:07 and 29:07), indicating the lack of understanding the mental struggles, in that “African Americans have really embraced mental health” (Appendix 12 29:54), which can correlate to religious reasoning. There is a larger attempt to try and understand the physical effects and share how the behaviour and actions to help them, creating awareness that whenever a person mentions or is looked up in the system to have SCD, they should be taken better care of, and receive proper attention, similar to other critical diseases. Although people affected directly or indirectly by SCD might have extended experience living with SCD, the mental state is an unconscious state, which should be investigated and helped with in the future, as the mental health gets drained by lived experiences, whether as a SCD parent or patient.

Furthermore, the mentioned focus on SCD awareness in SSA might also be challenged for women living with a malfunctioned genotype, herein not being haemoglobin AA. If the man finds out that sickled genes are the reason to the birth of a SCD child, they separate from their women, leaving them with the burden of the child, causing mental and psychological challenges towards that person (Appendix 14 1:04:48). Calling themselves warriors instead of patients (Appendix 13 0:30), they want to fight this disease themselves with as little help as possible from the hospital, as they then feel that they are losing to the disease. “I’ve always felt that when I have to call the ambulance or when I have to go into hospital, I feel like its defeating me” (Appendix 11 15:47) + “I’ve been indoors for about almost two months, so it’ll take me a while to recover … I’m just going hospital-home-hospital-home … it affects me mentally and psychologically … I get to overthink, I get to cry, say why me?” (Appendix 13 1:57). Combining the amount of effort, they put in self-care and self-efficacy, for them then going to the hospital needing help is challenging, as they feel like they lost by losing control on handling the disease. Trying to establish a strong psychological mindset, SCD patients tend to wait until necessary to receive the medical help needed. They want to have a reasoned action to go to the hospital, and their attitude, together with their perceived norm and perceived behavioural control connects with their experiences of the healthcare system, HCPs, when to go to the clinic etc. The degree of mistrust in the system is imminent and combinations between good and bad stories about

other experiences with HCPs, ER etc. creates a difficult and challenging understanding of reason to mistrust and can come down to many aspects such as stigmatization, systemic racism, lack of education and adult care.

5.1.1.5 Education – Change of Behaviour

The education level of SCD compared to other diseases lack in the medical curriculum (Appendix 10 18:14-18:45). Being a medical student, Julia argues that the reason is due to the type of people it affects, only affecting marginalised groups of African descents, Indians, Caribbeans etc. A call to action in the curriculum is mentioned to be needed as the world is constantly evolving (Appendix 10 20:51-21:02). Usual standard training in US concerns less than a day of training on how to take care of SCD patients (Appendix 12 13:20), with overall limited training in RDs, unless pursuing to become a specialist. Low amount of education regarding SCD also appears in British Isles and SSA, with SSA having low numbers of medical doctors who are eligible to treat for SCD.

The way of educating and making people and HCPs aware of SCD are different. It should be stated that in terms of creating awareness and educating people should continue being different in the future of different societies, continued focus on what works best in the different cultures should be applied. There is still a lack of overall awareness and education regarding SCD in the US, but it is becoming better with larger focus on what SCD is through advertisements, but also with more local PAGs being created, making a larger society and hereby a better understanding of SCD, how to live with it and more.

There should be a larger focus on tests and learning about living with SCT or SCD from an early age, when they already know from the newborn screening whether they have a SCT, SCD or nothing. This can become a major factor in preventing the sharing of SCT as it is a genetic disorder which can be shared to children if at least one of the parents have a sickle cell (herein AS, SA, SC, SS and rarer haemoglobin genetic issues). The attention towards those living with SCD and not at those who have SCT, and their future repercussions is a futuristic issue that should be paid more attention and awareness towards. Multiple interviewees (Appendix 11 58:18, Appendix 13 9:53-11:22, Appendix 14 17:00, Appendix 10 40:09) states that people need to know one's genotype. Knowing your genotype before getting children can prevent the trait and disease from spreading, making personal education important to lower the prevalence.

Creating awareness on SCD to the general public is challenging. Nicole mentions about public awareness: "on the other side [of overall awareness] you don't want people to be stigmatised … you

can't [do only one side] … it would be more the young people that would suffer" (Appendix 11 1:15:54-1:16:30).

Africans believe there is a reason why their nutritious possibilities combined with the prevention of crises and living a "better" life with SCD, that God did this for a reason. Whereas US and British Isles does seem to take much care on children, SSA depends on commute opportunities for them to being treated (Appendix 13 19:55, Appendix 14 50:42). Having people sharing with their communities about the disease will minimize the stigmatizations around the parents and families connected to the SCD child, of the family being witches etc. (Appendix 13 8:33, Appendix 14 22:41).

Interviewees explain the need of more education towards treatment and risk factors, in order to prevent the cycle of newborns with SCD (Appendix 12 59:17-59:33). Furthermore; "I don't think it's gonna take like a whole week … it probably could take a whole week to talk about sickle cell if specialising … basic level of sickle cell, what it is, consequences, how to deal with it … done in an hour" (Appendix 10 36:11-36:42).

5.1.2 Paediatric v Adult care

There is more focus on paediatric care than adult care when concerning SCD. The attention and care for SCD patients is problematic, especially when becoming an adult (either 16 or 18 years old depending on hospital system). Julia describes about investment: "emergency care specialists and general practitioners, at least I think those are institutions worth investing in … a [children hospital in Ireland] have a rule that sickle cell patients should be triaged within 10 minutes … but then you go to the adult section and nothing like that is there" (Appendix 10 52:33-53:25).

Once they are an adult in the system the burden of getting certain tests, reaching out to specialists, psychiatrists etc. has to be conducted and paid by the SCD patient themselves. Because of this, many SCD patients re-consider whether some things that could help them in taking better care of themselves is necessary, as these usually costs money, and not everyone can afford this (Appendix 12 49:02-49:56). Even though the awareness of SCD is not as hoped, the care for children is still experienced as better in the US, British Isles and SSA. Utilizing known literature about when crisis usually occur in SCD affected, these were found present in Melissa and Julia, both having crisis around the time of interviewing them, being between 18-30 years of age (Brousseau et al. 2010).

The switch from paediatric to adult care was also presented as lacking in all interviewees. The roles and responsibilities, new insurances and other things that were taken care of in the past is now presented to be their own issue, as focus is mainly on helping the children with SCD.

The lack of parental awareness towards children is a concern to lowering the prevalence of under-5 mortality rate. As Christie mentioned: “They don’t see it as a disease, they attributed it to so many things … they felt it was the work of the evil spirit which tormented the young one … because before age five they die if you don’t take care of them well” (Appendix 14 4:13). If these children do not receive the attention immediately and does have a long trip to receive the necessary help, together with longer waiting periods, the risks of dying increases. Speaking about the condition to those relevant and necessary for the SCD child to have a better childhood creates a better futuristic picture for that child, eliminating stigmas in the relevant areas.

By getting used to daily routines and not having any larger indications of serious issues or crises, the SCD patients with few SCD related issues in their adult life can miss the understanding of when they have a serious issue before actually going to the hospital. Supporting the work on previous experiences having a role, self-care and self-efficacy towards the disease is being proven by following routines to avoid going to the hospital. The behaviour being difficult for HCPs to understand due to not being able to see a crisis, can result in the negative affect on health outcomes such as pain management experience as Jenerette et al. (2014) proclaimed. A SCD affected not remembering how to manage a crisis can also become an issue and could be understanded to why SCD is difficult for HCPs to understand, as people differentiate in behaviour, attitude and similar beliefs when visiting the ER or hospital.

Three interviewees (Appendix 11 18:01, Appendix 10 1:06:58, Appendix 12 46:28) shared situations of serious cases not being dealt the attention and focus it needed, having to wait longer period of times before receiving medical help. Timely intervention, herein the access to quality care is experiences to lack in SSA, having to commute longer distances to receive needed treatment, in which they might not know is needed. Melissa points that “timely intervention, this is something the community people might say “let’s just give them hot water … hot soup [to soothe the pain]. They don’t know they should rush to the hospital … they don’t have finance … it’s the lack of awareness and finance” (Appendix 13 41:52-43:36).

The medical care is shared to be great without complications in paediatric care, but when becoming an adult, the care and help is limited, and own actions and medical, mental and psychological focus

is in its entirety on the SCD affected when becoming an adult. The lack of learnings about the transition seems to be present, with people not knowing what to do, placing them in a difficult situation on taking care of themselves.

5.1.3 Attitudes and beliefs

There are different attitudes and behavioural beliefs when analysing the different lived experiences of interviewees. These were mainly associated with religious interventions and burdens of being affected by SCD. Awareness towards the disease was mentioned as creating potential change of behaviour and attitude from both HCPs and SCD affected.

5.1.3.1 Religions interventions

The care and view on SCD depend on the awareness, learnings and understanding of this. In SSA, if districts and communities does not know about SCD, if one person has a baby who will experience pain crises, problems and potentially die, accusations and assimilations of witchcraft will be assumed to be placed on the entire family if one in that family has a SCD child (Appendix 14 22:41 and Appendix 13 4:10 and 8:33). SSA still lack experts within SCD, and with it being an issue to travel to the bigger cities to learn about SCD, what treatments is possible and medication there is needed, there are different, and quite effective resources to be used. One resource that would be more suitable for SSA is campaign for awareness and other use of resources besides medication in districts, herein nutrition, how to cope with life, but also educating them in what SCD and SCT is.

With limited people in such organizations travelling around the countries in SSA, educating people about SCD in churches with awareness campaigns, informing societies and districts about SCD and SCT is preferred in SSA and in other more religious areas. Preventing SCT to get out of control, raising awareness in regions with higher prevalence is needed, targeting to eliminate religious stances towards SCD.

SSA and families with African descents have African knowledge about how to treat the disease. They stick to what they know can prevent and/or ease pain crises that occurs to them, whether they know or do not know that they have SCD (Appendix 11 5:05, Appendix 13 41:52). Adapting these learnings and knowledge to patients and families who cannot afford medication which treats the issues in the same way should be taught at an early stage so that SCD patients are not positioned to over-drug themselves with too much medication that might have been easy to prevent. Although being taught at

an early age, living standards and what to consume during the day should be paid higher attention to in order to prevent and limit the amount of pain crises.

Regarding losing, the African community is also depicted as a fundamentally difficult due to how they handle the news of having a sickled child. Men in SSA usually find a new woman who is AA, if getting a child with SCD. Christie shares an experience: “his first wife was an AS, and he was AS, their two children died so he had to leave the woman … he married his recent [looking for] an AA … My husband also left me and went and married an AA so we suffer much from it” (Appendix 14 1:04:48). They do not want to take part of the responsibilities of having to deal with a “demonic” child, putting in the amount of work and time that is needed.

5.1.3.2 Burdens

Burdens are respected differently according to cultures, and the financial, societal, and cultural situation has everything to say with this. Parents taking care of their child places a lot of burden and responsibility on the parents into raising their child as best as possible (Appendix 12 6:50-7:54, Appendix 10 6:56, Appendix 11 33:57, Appendix 14 38:45 + 46:10). This will in regular cases mean they will have to either have a flexible job where they can leave if a crisis happens, and their child gets hospitalized. This is not the case for everyone, and some parents might even need to quit their job, being a full-time carer of their child(ren) (Appendix 12 5:34). Physical and mental concerns due to the burdens are unavoidable and their careers, societies etc. are possible to change due to these burdens of taking care of their child(ren) living with SCD. Financial expenses on medication and potential treatment are also a larger burden placed on SCD patients (Appendix 13 17:54). Burdens on the SCD patient themselves includes having a more troubled childhood, being excluded from societies and later, when coming into dating, does showcase these burdens they have by living with SCD. SCD patients when growing older wants to be independent and might not want to place too much burden on their parents. Julia mentions that when going to the hospital they feel guilt of bringing their parents into this situation, causing them to stress and worry about what is about to unravel, and being away from parents, having smaller pain is talked to be burdens on themselves, as knowing others might be stressed and worried about the individual might be problematic for the individual experiencing the issue (Appendix 10 46:22 – 47:42).

5.1.4 Systemic Racism

With SCD being a “non-Caucasian” disease, indirect evidence on lack of interest in SCD regarding awareness and attention to care from this society that is not affected. Other diseases such as cystic

fibrosis (Appendix 10 09:40), cancer and Alzheimer's (Appendix 11 1:10:50) are being paid more attention towards, with these more commonly affecting Caucasian societies. When those affected by SCD does not follow usual treatment plans, going to physicians and doctor's appointment on a regular basis, the SCD community helps creating a stronger and for them worsened systemic racism, as doctors will find them difficult to work with (Appendix 12 17:32), creating worsened health outcomes due to beliefs and other factors. The indirect behaviour and attitude from HCPs who sees these patients as regularly people of African descents can strengthen the systemic racism behaviour if regular problems and difficulties with SCD patients become a regular thing without knowing the effects of SCD. The systemic racism is not only presented in an institutional structure with having accusations of drug abusers going to hospital, but them also taking drugs when having a crisis (Appendix 12 46:28, Appendix 14 22:41) or being problematic and difficult to handle, societal structures are interpreted to be present for SCD patients. Not knowing the issues associated with SCD creates both an intentional and unintentional racialisation on people, although they might or might not be aware of their actions. There is a sociopolitical systematic issue of having SCD patients being labelled as problematic, together with lowered hierarchical presence compared to other diseases and issues.

The level of racism can depend on many things being present in the subconsciousness of a person, whether normal or working as an HCP. It regards the repertoire the African community has received for the individual approached. The level of systemic racism depends on how each individual looks at the communities, whether collectively or individually created. Those who have a strong and negative collectively created opinion on those of African descent will predominantly showcase a high level of systemic racism towards anyone, both unintentional and intentional, but mainly intentional. By highlighting cases of long waiting hours, questioning SCD patients as drug abusers by knowing specific medication etc., these can be represented by systemic racism. It cannot although not be finally concluded that told experiences of longer waiting times through a pain crisis, low attention towards them, together with lack of knowledge, understanding and interest in the disease is due to systemic racism, but the assumptions of drug abusers, vampire etc. can have a social and institutional systemic racism connected, and most likely is on a personal level, rather than a collective level.

5.1.5 Experienced stigmatization

When questioning the interviewees, everyone confirmed that there are many stigmatizations connected to SCD, and that it can be associated with the lack of awareness on the disease. Being

stigmatized as a general disease associated with African descended families, it is from all perspectives that these stigmatization can be analysed. HCPs have overall stigma on SCD patients as being troublesome and non-collaborative (or at least less collaborative), individuals stigmatizing the disease as an early death sentence (Appendix 10 09:20). The stigmatization is present in the childhood and adult life. Justin mentions that family events could be difficult for him: "they would have events that we wouldn't even be invited … didn't know if one of us would be sick or not" (Appendix 12 21:06), but also in school: "we just had a hard time making friends and keeping friends because we're viewed as being different, being sick" (Appendix 12 21:38) and "I thought that it was normal that children have to go to the hospital every few months … I thought all kids had to take medication" (Appendix 10 5:55). Having symptoms (Appendix 12 21:53) and misperceptions of how SCD spreads (Appendix 10 11:05) are other experienced stigmatization the interviewees have lived with. Having physical symptoms of jaundice was also presented as an issue to stigmatization.

In SSA where many are affected by SCD, Melissa and Christie mention that it should become the norm of understanding what SCD is, and whether or not they are carrier of SCT or not, knowing the increased risk there might be for their future children if both have a SCT (Appendix 14 17:00, Appendix 13 9:53). In those cultures, and places where this is not the case, in order to lower the stigmatization, a targeting awareness campaign should be considered, for them to then spread the awareness to those interested by participating in SCD related events, or by becoming friends with other families or people who shares interest in knowing more. In those countries, the focus on eliminating the stigmatization should be on HCPs and education to become this, as they then can become ambassadors in spreading the awareness regarding SCD.

Early death sentence is still one of the most common stigma regarding SCD patients, and making people aware of people living longer lives, future treatments, medication alternatives etc., both people affected and not affected by SCD can prevent early death sentences to become the norm of SCD. Melissa mentions: *"*A lot of my colleagues would tell me you will soon die, you are on a deathrow" (Appendix 13 4:10) which encapsules the stigmatization experience she has experienced herself, adding that: "… my mom got tired of [stories being told that Melissa lives with SCD]. If you want to die, die, I'm tired just giving up on my own daughter" (Appendix 13 32:59). Christie adds that: "when the children have swollen hands and legs, they think they are not human beings. They throw them into the river" (Appendix 14 59:30), indicating that the under-5 mortality can change if being de-stigmatized through awareness.

Utilizing the longer waiting time to be treated when going to hospitals also indicates the stigmatization of the pain that SCD patients experience is not as bad as they present it.

5.1.6 Experts through experience

Sharing their experiences, patients showcased how their experiences have created them into becoming experts of SCD, being able to tell their HCP or ER person on what exactly is needed for them to be treated, herein which type of medication (Appendix 12, Appendix 13 12:40), need for morphine (Appendix 11 7:30) and how they should treat themselves and others living with SCD. The attitude and belief in these patients are the crucial part for the restoration of problematic stigmatization that is associated with SCD affected people from HCPs. HCPs are not the one affected by the disease, and because the body changes (Appendix 11 10:08), the medication that might be presented for the practitioner wanting to prescribe medication to a SCD patient should be aware and conscious of the reactions from patients and learn from their experiences, meaning if one treatment does not work for them, they should provide them new additional medication (Appendix 11 07:30 and 10:08). The lack of complacency regarding the disease and symptoms are present from both patients and HCPs, and although it is difficult for the patients to know exactly when the issue becomes dangerous, HCPs should not question whenever SCD patients enter the hospital, as they then will have a greater idea that they are in a difficult situation as should be treated quickly.

The self-efficacy of SCD patients is shared in the lack of trust to the people in the system (Appendix 11, Appendix 12, Appendix 13, Appendix 14), and that they in many situations prefer themselves taking care of the situation, rather than having others take reasoned actions for them. Mentioned earlier in terms of indicating a problematic behaviour towards HCPs, helping each other as much as possible should be the aim. If the behaviour of the patient, presenting themselves as an expert and expecting to be the one telling what to do, HCPs might find it troublesome and annoying, losing their control belief in the act of their job.

Summing up the experience as expertise, talking about the mentality of HCPs or specialists: “You can come across a patient that has a unique experience and I don’t think your years in treating that condition should override your willingness to learn more about that condition … specialists need to remember that patients are also experts in their conditions because you can’t think that one-size-fits-all everyone’s condition effects them differently … you can’t treat everyone the same based on your experience” (Appendix 10 1:10:51-1:11:58).

5.2 Believe It Awareness Campaign

In late September 2022 (September being Sickle Cell Awareness Month), Forma Therapeutics in a collaboration with Sickle Cell Disease Association of America, Inc. (SCDAA) launched “Believe It”, a campaign focusing on showcasing a living experience of a SCD patient, creating awareness and understanding on SCD. The campaign consisted of two public service advertisements (PSA): A video PSA (both as a 30 second and 100 second video), and a radio PSA, consisting of 30 seconds. The awareness campaign was launched in United States televisions and radios, with the radio PSA sending people listening to visit the website scdbelieveit.org at the end of the PSA.

5.2.1 Video PSA

The 100 seconds video PSA (YouTube 2022) was watched, and the content of speech was textualized to find themes relevant to dive into (Appendix 2) The video PSA contains different themes worth diving into in order to understand the content and purpose of the video PSA about SCD. The following themes will be dived into: (1) Patient-HCP/normal person experience, (2) Word associations and seriousness of SCD, and (3) Sounds and Visuals, herein use of sounds, music and colours throughout the video to change perspective and focus.

5.2.1.1 Patient-HCP/normal person relationship

We see two POV’s in the video PSA: (1) the conscious world perspective (what everyone sees), and (2) the unconscious perspective of John (what only he experiences), reflecting the patient/HCP experience of a person with SCD. It becomes imminent that Forma Therapeutics and SCDAA wants to share conscious and unconscious of perceptual life with normal colouring when presenting the pain and situation from an outside perspective. John’s personal experiences throughout time in class are presented as more sombre, as depicted through faded use of lighting, together with showing and having sounds with static noise and reactions in his arms and hands during short scenes (Appendix 4-6), with walking towards the front of the class feeling like walking on burning fire stones (Appendix 7 + 8).

The advertisement is split into two acts. The first act happens in a classroom, the second at a hospital. In the first act, the relationship is between SCD patient John and the teacher. The relationship between them can be interpreted in many ways. In the very first clip, the teacher can be interpreted to be annoyed and/or surprised that John is attending class, which can correlate to limited understanding of SCD and/or indirect racism towards John. By targeting John, both the care of him together with a potential stigmatization of him being lazy can be pointed to be the case. The teacher might not have

a larger social understanding of John's life living with SCD, as she is aggressive, and when she questions whether he is fine, the fact that John does not express his current issues indicates the lack of knowledge of him having SCD, but sees him as a regular student, creating the showed behaviour and attitude as rude towards John. The lack in interest and knowledge is represented and understood as to provoke a real discussion into showcasing situations in the real lifeworld of a SCD patient going to school but having no interpersonal relationship with the teacher.

The facial expressions by the teacher can indicate stigma towards John as an African American attending classes, maybe with the purpose to exaggerate the fact that he might not be in classes often, targeting back to the hospitalizations that SCD patients experience. With a lack of awareness and understanding of his condition, creating potential mental and psychological stress towards John can end up worsening his situation, which can be depicted with him having to present in front of the class. We see the entire cycle of him not wanting to lose the battle with SCD, and does not want anyone's help, as they might not understand his behaviour and beliefs.

In the second act, we experience John in a hospital bed. The potential SCD affected/HCP relationship is non-existing only having what can be interpreted as John's parents as present. The second act indirectly begin in the first act as John is wearing a hospital gown, indicating walking into the ER aisle (Appendix 7 0:40 – 0:55), herein not receiving the attention needed because the pains are hidden for the outside perspective of seeing a pain crisis on a SCD patient. Whereas the first act focused on the attention and care given in school, the second act focuses on the lack of attention towards SCD patients, having parents being caretakers of their child together with the missing attention and awareness of SCD affected and HCPs, as we do not see any HCPs during the second act, indicating the family dealing with it themselves. The burden of parents having to take care of their child when hospitalized is presented in the video PSA. It can indicate the seriousness of the SCD issue John is experiencing with his father rushing into the room, showing a sort of relief when looking at the mother, whose expression indicates that it seems to be fine, but not as hoped.

5.2.1.2 Word associations and seriousness

Focus on use of text can provide hidden connections associated to the purpose of a campaign. The topic of the class can be interpreted to have such hidden message. With the topic being concerning the 4 P's (Product, Price, Place and Promotion), these can be placed into boxes of SCD. *Product* being the disease itself, being different genotypes of SCD, *price* being symptoms, burdens, stigmas etc. associated with SCD, *place* being centralised around US and the African American community, and

promotion in terms of this advertisement, promoting the awareness and causes there is with having SCD. Forma Therapeutics and SCDAA wants to create a picture of people living with SCD in US, which the topic of the class indicates.

Diving further into the use of words in the video PSA advertisement prioritised focus on specific words was performed through tempo of speech. When John has to leave his seat to present in front of the class, he talks about how it is to live with SCD, creating a storytelling by stating: "Living with sickle cell disease … it's torture … everyday tasks can be filled … with pain … life with sickle cell disease isn't easy" (Appendix 7-8 0:45 – 1:11), creating dramatic pauses, having the audience paying attention to his words. Furthermore, the pauses of storytelling can also indicate current struggles on John, having issues with the ability to speak. Without any personal lived experience with this, small tasks besides talking in front of the class can be connected to the phrase "everyday tasks can be filled … with pain", even the figure of speech and presentation being a challenge for SCD patients.

During the video PSA where the teacher tells John to share with the class, calling his name over and over tells us about the lack of awareness and attention shared with John by the teacher, creating an experience of the pain crisis triumphing any other relations and focus. The different tones during the calls of his name in these scenes also indicates the focus and attention that SCD patients might have during a crisis, blocking everything outside their own world when having a crisis.

Taking understandings from told experiences earlier in the findings, the water bottle present at John's desk is also a sign of awareness of how SCD patients themselves are aware of the consequences of living with SCD. They are aware that they need to consume water, more than anyone else to keep the pain crisis limited.

5.2.1.3 Sounds and visuals

The video PSA makes use of different focus on sounds through static noises and tempo in music. Furthermore, it makes a greater use of specific visuals, depicting two worlds already described earlier in this part of the findings. The effect that these have will be analysed below.

5.2.1.3.1 Music

The background music in the video PSA pictures a story to be told on the situational changes from him having a crisis to him being fine. At (0:14) the crisis begins, with the music indicating that something is about to happen by having higher octaves and creating louder noise. This then is presented with static sounds from (0:25 – 0:40), indicating that even though the teacher is calling, the

main focus is still on John, and that the pain crisis is gaining control distancing John from the real world. The mysterious music, combined with the dimmed and dark lighting creates a sounding disturbance and issue, if which the listener can understand that something is wrong, in this case a pain crisis. In the moments when John speaks about the number of people that are affected by SCD in America, the music becomes more positive and might create a sound of positivity and curiousness making one believe that by doing something, whether it is friends, family or HCPs taking action creating a future where the aim is to lower the prevalence of SCD, which by the music sounds possible.

The music creates the attention on the mistrust that is present between SCD patients and HCPs. The mysterious music in the ending can also be analysed as an opening to possibilities and opportunities regarding SCD awareness and limitation on stigmas. It upbrings the positive possibilities of what can become a better future for SCD patients if the trust, care, and awareness of SCD can be prioritised.

5.2.1.3.2 Colour changes and POVs

The normal perspective is presented as positive with lighter colours, with the SCD patient perspective portraited as dark and gloomy. These we see for a longer period five times (Appendix 4-8), combined with three rapid flicks portraying these darker colours in few of the clips, one before John stepping up towards the class, one before we see him in a hospital bed and last when we switch to a side angle of him and his family at the hospital bed.

There are specific clips in the PSA where SCDAA and Forma Therapeutics want to highlight the life of a SCD patient. The first time we see John indicating pain is at 0:21 when his hands are glowing up, indicating that a pain crisis is about to happen in his arms and hands. The camera comes closer to John, darkening the light and putting him into focus with a close-up angle. After a short switch back to a normal perspective, John looks confused at his arms, before we few seconds later, after he has been asked to present, see John's crisis getting worse, together with him presented wearing a hospital gown. The crisis is getting worse and can be interpreted as him getting nervous or stressed at having to present in front of the class, triggering his body. John is a patient while the darker shots in the PSA are presented, which can already be seen in the first shot in which we have John point-of-view, having a hospital wristband with his name and case number presented. The pains are shared while the visuals are darkened and tampered making us aware of how it is experienced by himself and his body, but switches during his walk to the front of his class from the personal perspective of John showing his

pain crisis as walking on lava stones and the outside perspective of seeing John slowly and collected walk to the front of the class next to the teacher.

The PSA has two important times where it flicks between multiple clips. The first is the switch between John in front of the class, and John laying in a hospital bed, assuming to be together with his mother and father. The visuals in this switch share the process happening for John, having many of his red blood cells sickling, combined with the beeping from an ER machine, indicating that his heartbeat is abnormal. The scenes of John laying in the hospital bed (1:18 – 1:38) shows the ongoing crises with visuals still showing his arms light up, but also the burden that is present from a SCD patient. It also indicates the burden that lays on the parents, with both his mother and father being present. With the dad only entering the room placing a glance at the mother, shows how sudden the reaction has been, with the dad possibly just arriving from work to check whether everything is going "fine".

We get different camera angles during the video PSA, indicating differences in power. We see John from multiple angles, with bird perspective twice looking down at his arms in pain, showing himself as weak and scared. We also see him at the end when laying in the hospital bed, indicating his weakness towards the disease in this case.

The usage of frog perspective indicates the subject as strong, powerful and threatening. These are only shown when John is looking down at his hands in pain, and when presenting him walking towards the lava stones.

Finally, close up looks are focusing on his pain he is/has to going/go through during the video PSA. These indicate and focus on the feelings and experiences that the person might think during the situation, here with John depicting the pain and stress.

5.2.2 Radio PSA

The radio PSA was broadcasted on radios across the US and had a different angle compared to the video PSA. Whereas the televised PSA could focus more specifically on the visuals and sound effects of how living with SCD is like, the radio PSA tries to describe SCD differently. The radio PSA consists of one speaker, asking four indirect questions, before factchecking the numbers of people living with SCD and how people can support their cause of the campaign.

It starts by referencing American football, the national sport of United States, getting the attention from people listening to the radio. The use of the short grunts and background noises used after "Am

I trying to sack the quarterback every snap?" (Appendix 3 0:00-0:02) is a physical expression to indicate the seriousness of the disease on what they are going through. Questioning "Do I have SCD?" (ibid 0:04-0:05) afterwards, together with "Pain crises that causes irreversible organ damage … And when I walk into the ER asking for help for my pain?" (ibid 0:08-0:10) it makes the listener aware of the pain of the described SCD patient, the description and expression of tackling each other to sack the quarterback. It reflects back to the grunts after the first question asked, connecting the sounds and sentences together. Having Americans as the target group, the reference is fitting to the purpose of sharing what might be the closest to how it is to experience a pain crisis through the use of their national sport.

A common use of words is 'Believe it/me', with this being mentioned four times, once after each four questions. Focusing on this indicates the issues that is connected towards SCD stigmatization on people not believing what people with SCD tells individuals. The pain might not be possible to see from the outside, needing HCPs and individuals to behave and trust the person when meeting them and talking about lived experiences. Mistrust and stigmatization of patients by HCPs can be interpreted through this emphasis on mentioning that they should believe the patient, whenever they mention that they have SCD, that they are in pain that can cause irreversible organ damage, indicating seriousness.

The use of "Am I trying to sack the quarterback every snap?" can be understood as the small bumps and challenges that SCD patients experience. Before the game, everything is fine, but when the game starts and you are on the field, each game can be different, and can depict the different lives of SCD patients. If, for instance, trying to sack a quarterback, herein being a defensive player, if your team is good defensively, less snaps and more breaks are happening throughout the game, **their** game. The team offense (which can refer to controlling and keeping up with regulations to not get pain crisis), can be either good or bad. These can to a larger degree describe how living with SCD is. If your crises are worse than usual, your defence is pretty bad, but if your awareness for how to help relieve the problems, whether contacting the people at hospital, knowing that your medication does not work, or keeping a healthy diet, the person tries to prevent it to become as worse as it could be. If the defence is good, but the offense is bad, many smaller pain crises can happen, which the SCD patient might be aware of, but does not consider an issue for the person to live as the person wants to.

5.2.3 Believe It!

The phrase “believe it/me” refers to the patients being experts, knowing what to do, what to believe in etc. When they tell something about their disease, their voice and talk about the disease should be understood and noted, eliminating stigmas and creating knowledge and change of behaviour towards SCD patients.

SCDAA and Forma Therapeutics are targeting throughout their PSAs to express the stigmatization that might be interpreted from those who are not SCD positive. SCD has received limited focus due to the stigmatizations and non-relevancy to the public and those not affected, not understanding the level of pain that are present for those experiencing a pain crisis.

The mistrust between SCD patients and people, families, and (mainly) HCPs is the focus of the campaign. The target groups can be narrowed more specifically to African Americans and HCPs. This is because of the last sentences mentioned in the PSA, focusing on statistics of 1 in 365 African Americans. Although it can easily be argued that the PSA targeted the entire population of US, by explaining how it is to live with SCD, creating attention, awareness, and understanding of how it is to live with SCD, but also by making people aware of the high number of 100,000 affected by it. The misbelief is crucial for SCD societies to eliminate, as those affected by SCD are experts in how to manage the different crises and complications that occurs to them. The radio PSA is compared to the video PSA a statement and call to action towards HCPs and ER staff that should meet these patients in the hospitals to believe them when they tell the personnel what to do.

5.3 Comparative analysis

Lastly, a comparative analysis on all interviews and both PSAs in the 'Believe It' campaign is made. This comparative analysis is to depict whether the campaign was targeted true stories, comparing it to what SCD patients and parents have shared of lived experiences. There will also be a future aspect related to the comparative analysis, analysing through interviews how SCD campaigns should be focused on the future.

5.3.1 Grimaces and fighting

First noticeable topic to compare is the expressions, grimaces and fighting that both John and the interviewees have shared.

The expressions of John are a true depiction of shared lived experience of people living with SCD. Interviewees have indicated how important it is to share the disease with teachers in school, managers at work etc., an experience that was presented to be non-existing in the video PSA.

Grimaces are something that might give people a clue to experiencing someone fighting a pain crisis, but this has been shared as difficult for people to see; "I'm feeling really badly in pain … you probably noticed that grimace here and there" (Appendix 11 39:20). Nicole mentioned to a colleague about a situation, in which that person could not understand and analyse that something wrong was going on, similarly depicted in the video PSA, where John multiple times shares pain with facial expressions and grimaces. Because the general pain is internal and cannot be seen by others, the pain and reactions depicted in the video PSA correlates to told stories from interviewees about swollen hands (Appendix 14 59:30), potential leg ulcer/pain (Appendix 12 4:28) and other pain related issues. The experienced episodes of Melissa regarding the fire stone experience correlates perfectly together with: "I've been feeling some pain for the past couple of weeks and I can barely walk around. I've been indoors for about almost two months … I'm just going hospital-home-hospital-home" (Appendix 13 1:57). Presentation nerves and stress was also depicted by Christie to be true (Appendix 14 46:10).

The connection between video PSA and interviewees lived experience is also the focus of the patient's wanting to fight the disease alone, not wanting to lose to the disease by needing help or more care than they themselves can do.

The usage of football reference might be obscure to use, as SCD patients normally cannot be physically active. Having mentioned this, the challenges that SCD patients experience with issues

towards their body, the reactions of going through a pain crisis feeling like getting pushed by (an expected 250+ lbs) man regularly can be interpreted having pain crisis.

5.3.2 Focus on parents and HCP role

The advertisement depicts a greater perspective to real life problems. With the lack of attention to signs and details in SCD patients through grimaces etc. when fighting a crisis, the presence of how the natural environment is for these people and families. The lack of presence of an HCP person when John is hospitalized resonates with experiences of uncertainty, level of attention and seriousness SCD has for HCPs, not only in the US, but merely across the world. The scene also unravels many sign of comparisons with told stories about parents and their role. By having a child with SCD, the burden on the parents taking care of their child, having to stop what they are doing to be present and take care of their child is something that real life patients can agree on, the burden and effort families have to put on the person with a mere uncontrollable disease. The reactions on the parents as sad can describe them in many ways, from concerned and nervous, to angry and disliking, having less to no apathetic reaction. These scenes describes both the reactions from the interviewees living in US and British Isles, having parents being worried, nervous and concerned, wanting to have some sort of control of how to treat their child for them not to be hospitalized, have them learn their living standards (Appendix 10 46:22, Appendix 11 6:51, Appendix 12 5:34), with those in SSA creating a picture of losing interest in dealing with this anymore, from throwing in the river and let them die to just let them suffer, be distanced from the family.

The worrying part of having larger burden placed on parents until the SCD patient is an independent adult is depicted fairly well in the campaign. Watching John fighting the pain on his own in the beginning, not wanting to cause any major burdens on others and only himself depicts both how little they trust the hospitals and HCPs, but also the focus on not involving others to become part of their world problems. With the lack of understanding on how much attention should be given on SCD patients while in a hospital bed, having their heart rate monitored etc., it becomes difficult to know whether the parents are also concerned of the lack of awareness towards the hospital and HCP(s) dealing with John. With the viewers not knowing the age of John, the amount of care is presented as a young adult, with little to no attention to him besides family, correlating to told experiences of difference between paediatric and adult care.

Bad experiences were shared by interviewees on how they felt mistrusted and mistreated while in the ER, or at least have heard experiences of this, crying and being the expert themselves, telling them

what to do, having a misbelief in the interviewee. This can be the reason onto why John did not want to share his pain, both wanting to fight the crisis himself, together with understanding that the care in the hospital might be worse than if he dealt with it himself.

5.3.3 Portraying a real life-world experience

The campaign portraits the real life-world experience of SCD patients feeling that they are managing the disease themselves in their own society and created world. SCDAA and Forma Therapeutics PSAs undoubtedly creates the nuance of how the experiences are for SCD patients, depicting both the experiences of the lack of consciousness and understanding of the situation in which SCD patients are situated in during a crisis, together with how the lack of reasoned action from both the SCD individual and the people surrounding this person. As depicted in the interviews, having SCD patients sometimes questioning the seriousness of the pain crisis, it cannot be pictured in a better way possible by showing the pains of John in the awareness campaign, together with hearing experiences of similar cases by interviewees. They all have in mind that they should be able to control their attack but ends up hearing about the seriousness of the issue while hospitalized. It depicts one of the crucial dynamics there is present for those living with SCD, that because they have struggled with it their entire life, they might understand themselves as being experts into knowing when the pain is too much or whenever they can handle it themselves.

The campaign wants to attack the stigmatizations that are associated living with SCD. It shows how little attention and awareness SCD is receiving, even with the number of African Americans it affects in the US. The campaign targets to ridicule the absurdity the lack of awareness around SCD is, with 1 out of 365 African Americans living with it. It ridicules in the video PSA by having no one in class paying attention to John, having no interest on who he represents in the video PSA, SCD itself. With none of his classmates feeling interested in SCD, it can be aligned with the told experiences shared by lived experiences of the interviewees. It targets the stigmatization people experience with being ignored and left out by others when learning that they as an individual have SCD, the systemic racism towards lack of institutional interest when hospitalized, the amount of care and burden family has to provide when the SCD patient is hospitalized, with uncertainty of whether surviving or not due to the lack of understanding the severity of the crisis/attack. It centralizes the fact that it is something that the African community has larger awareness towards, as SCD is depicted to be a disease only affecting their communities. The lack in perceived behavioural control, together with the norms and attitudes towards SCD patients, not believing in them and leaving it up to themselves is all perfectly

depicted in the campaign, having the seriousness of the severity lacking in both PSAs and in the interviewees and other SCD patients when conducting people that does not understand SCD and what they experience. They are living in a bubble that is gradually affecting larger areas unless awareness from other descents is happening in the future.

5.4 Sub-conclusion

The research question/problem formulation was as follows:

Looking into awareness of Sickle Cell Disease (SCD), how can SCD awareness be raised in British Isles, US and SSA to lower the prevalence and under-5 mortality of SCD patients, together with educate the public and HCPs in SCD education to lower stigmas and discriminations?

Throughout the process of collecting and analysing the data with the perspective of my aforementioned theoretical and methodological frameworks, multiple interesting findings were discovered.

Firstly, it was found that awareness marketing of SCD differs from who to target, as the same type of people and professions should be targeted awareness in countries in SSA, US and British Isles. These can be connected to the number of SCD prevalence per person. In SSA countries where the percentage rate is highest, awareness around the disease should be targeted every district with focusing more on educating people on SCT as much as overall SCD. US should target African American and migration communities to raise the awareness of SCD. With British Isles having smaller numbers of prevalence, more focus on educating future and current young HCPs to learn about SCD in depth, in order for them to share the knowledge and awareness with patients when meeting them. Educating people into lowering stigmatization, shared opinions of both targeted and overall awareness was mentioned. SSA should target the overall population, In the US and British Isles, with already high attention on racism in the country, people of relevancy should be educated into erasing future stigmatization of SCD.

Secondly, sharing lived experiences of living with SCD should be prioritised for those of influence for the person with SCD, having their established friend groups, institutions etc. understanding the disease for them to have a better childhood and receiving better attention and care when showing different attitude and behaviour. This will create a greater share of awareness towards SCD, and might even help getting rid of stigmas in these societies in which the individual lives in.

Thirdly, the attitude and behaviour towards SCD patients is problematic. With issues of pain etc. for SCD patients, believing them when telling HCPs about their condition, the attitude and behaviour

from HCPs is lacking in trusting the patient. Mistrust to the system in correlation with stigmas, attitude and behaviour, and lack of knowledge are connected to the respect and consideration from SCD patients in asking for help or dealing with the disease themselves. Systemic racism can furthermore be attached to the lack of attention HCPs and the overall hospitals pays on SCD patients. With the lack of an HCP in the video PSA, radio PSA indicating no belief in people living with SCD, completed by interviewees describing experiences of waiting for medical help for too long indicates a systemic racism towards those living with SCD.

Lastly, the lack of awareness towards mental health and living with SCD as an adult has been presented as problematic and something that should receive greater attention when educating people in the medical world and those affected by SCD. Letting these people of on their own can become problematic if no help with the transition is made. Furthermore, more educational focus on SCD should be prioritised in the future, not only having HCPs available for paediatric care, but also for adults when having to visit the hospital because of complications. With paediatric care being fairly great, opposite can be said about adult care, and it is a larger issue as the bigger crises usually happen when they become young adults.

6. Discussion

In the following discussion section, the findings of this research will be discussed, how these findings can contribute to the literature performed within awareness, more specifically on sickle cell disease (SCD). From the findings, future angles and strategies would be suggested in this section, onto how SCD awareness can become greater, how we can lower the prevalence of SCD and sickle cell trait (SCT), and finally lower the amount of stigma associated with SCD.

6.1 What are the future strategies?

SCD is a not only becoming a problematic size in Sub-Saharan Africa (SSA), but with internationalization of societies, the lack of focus on SCD and not being taken fully serious by governments, pharmaceutical companies and people educating to become a healthcare professional (HCP), psychiatrist, general practitioner (GP) and similar. Having SCD being a rare disease (RD) is certain in many countries around the world, but by itself counting approx. 20 million people living across the world with the disease it can be argued that a strategy around SCD, creating awareness, lowering the prevalence, and getting more people interested in SCD is to diverge relevant people from going by the 20 million people affected by SCD, and instead think about the 300 million people worldwide carrying a SCT. Instead of thinking only 1 in 405 (or 0,24 %) lives with SCD, thinking that 1 in 27 are carriers of a single gene which, if both parents have this, gives a 25 % chance of having a child living with SCD. Similarly, another gene disorder in Down syndrome, every occurring 1 of 700 100-1250[1] births occur to have a chance/risk of getting a children with Down syndrome (Department of Health 2024). The problem with using these metrics is that, although both are genetic disorders, one happens as a mere lottery, with the other being passed by generations. Furthermore, 1/3 of all these are living in SSA, making the strategies more relevant for those certain areas, but should also consider developed countries with immigration in future strategies. Although no global metrics on when a disease should be registered as a RD, with 1 in 405 in the entire world living with SCD, its worrying that SCD have not received more attention and awareness that it has. Even though it is not a strong argument, the high and rapid attention Covid-19 received when becoming a global pandemic, efforts towards this degree in the most critical areas, herein SSA would be beneficial. There is still a larger lack of knowledge and awareness towards a disease that are affecting 300,000 babies is worrying. With those being poor being larger contributors to under-5 mortality rate (Houweling et al. 2005; Burke et al. 2016), placing greater focus on SSA in giving access to learnings and

[1] Depending on age. Age 25 is 1 in 1250, age 31 is 1 in 1000, age 35 1 in 400 and about 1 in 100 at age 40

understandings of how to cope with SCD. Although previous research (Burke et al. 2016; Chao et al. 2018) indicates lowered under-5 mortality, it still quantifies that poor families and places in Central and Western Africa are similar to what they were in 1990, having local areas being more affected than national areas. This correlates to the programs and campaigns Melissa and Christie mentioned that should happen in the future to decrease the under-5 mortality more significantly.

6.2 Education and Care centres

Education on SCD and other more common RDs should become more relevant in the overall medical curriculum, both in universities, but also for educated HCPs. With other genetic disorders like haemophilia, cystic fibrosis, Alzheimer's disease receiving more international attention and awareness, although a rough estimate of approx. 300 million worldwide are carriers of SCT (Ashorobi et al. 2024), with 1/3 living in SSA, a greater focus on lowering the prevalence should be to educate people in SSA. Although WHO's response regarding SCD is (1) increasing awareness of the global burden of the disorder, (2) promotion of equitable access to health services, (3) provide technical support to countries for the prevention and management of these disorders, and (4) promote and support research to improve quality of life for those affected, there seems to be a lack of clarification on how these can be done. Shared experiences by multiple researchers of interest in primarily SSA (Esoh et al. 2021; Ally and Balandya 2023; Brousse et al. 2014) indicates that there still is a need for better management of testing and having a better system of SCD centres, together with a greater overview to prevent babies born with SCD, together with therapeutic interventions at an early stage of their childhood. Having these centres already established, focus have only been to have these in larger cities or capitals, creating issues for those living far away, and who economically cannot commute between their district and the SCD centres. Where previous research has focused on SCD research and guidelines for care in SSA (Esoh et al. 2021; Tluway and Makani 2017), the lack of focus on burdens living with SCD is undiscovered to a higher degree due to limited personal research with people affected by SCD. Although having a total of 40 reported SCD treatment centres (Esoh et al. 2021), these covers only 15 countries, and with a rough estimate of over 300,000 babies born with SCD each year with almost 80% of these occurring in Africa, there is a large global burden in SSA, herein the need for more education on SCD to both future and current HCPs, communities and the general public is needed in order to lower the prevalence of SCD births. If more care centres for SCD affected, together with more programs locally, having advocacies and SCD prevention organizations spread across the country in smaller cities closer to districts, the potential overload of SCD babies born and the personal burdens, herein mental, financial and psychological burden can be prevented.

SCD should get as much attention in SSA as malaria and dengue fever in order to lower the prevalence and under-5 mortality rates for the future as most of the people dying before the age of 5 are children living with SCD.

6.3 Preventing stigmatization and discrimination

Diving into racial diverse markets of US and UK, the overall representation of blacks is relevant when SCD patients has to deal with something regarding medication, without seeming suspicious to the stigmatised HCP. Whether visiting emergency rooms in hospitals sharing the pain and/or problems, physical presence and other indications can be difficult to comprehend from the HCP POV. Racism and other negative attitudes and behaviours towards SCD patients with African descents becomes implicit attitude from own preferences but did not form the content of the reason (Levy 2013, 223) which is creating perceptions as to stigmatise these patients. Previous research (Henderson et al. 2014) on anti-stigma efforts amongst HCPs have indicated education, improved knowledge and follow-up assessments as being factors to reduce stigmatization and discrimination. Studies has also had focus on internet-based stigma, and with SCD depending on the different experiences, genotype, and many other reasons, stigmatization of SCD online can be a miscoloured picture of what SCD does to each specific individual affected by SCD.

With African communities being open to learn about SCD (which was learned through SCD interviewees), understand the responsibility and burdens there comes with having a child affected by SCD, the stigmatization and discrimination will fade. This correlates to previous research (Long et al. 2011) indicating that awareness on potential issues for a future child is sought for, but also with concerns for potential risks being raised. Sartorius (2010) argues that interventions to reduce stigma and discrimination will require a long-term, sustained commitment. Education should target youth, and the long-term impact might not be as efficient unless programs to sustain these learnings are kept. Created images of people living with SCD should be changed, and utilizing sickle cell advocacies as voices when talking about SCD can make people create a different picture of those affected by SCD. With challenges between SCD patients and HCPs being depicted in the interviews, cooperation between these should be better in order to create a stigmatized picture of SCD patients being difficult to work with.

6.3.1 Difference of race majority

This thesis paper implies that systemic racism is present for those affected by SCD, and as SCD mainly affects people of minority ethnicity in Western world countries, this supports the overall

racism in healthcare environments. Supporting the inadequate quality of healthcare and viewed as less desirable healthcare users (Morel 2019), and with difference in healthcare outcomes on patients of minorities and whites are widely recognized (MacIntosh et al. 2013). Ben et al. (2017) added that with interpreted experienced racism adding lack of trust to healthcare, this research adds to the systemic racism scope that was focused on with US and British Isles specifically. In their research on racism in healthcare, Hamed et al. (2022) empathized a greater focus on the lack of articles regarding specific healthcare racism. With as much focus I wanted to put on the racist aspect, knowing that finding people living in European countries with SCD would be slim, utilizing the example of Julia, currently living and studying in Poland, was still applicable to use due to background and place to live, giving a different edge to a necessary aspect for the future of SCD communities in a European context, both educational and awareness wise. Greater personal research focus on SCD patients in minorities statuses should be paid greater attention to understand the Western world systemic racism role in healthcare systems and how these can be improved from individual POVs.

6.4 Improved management

A greater focus on creating an improved management of people living with SCD will help in the future to contain and put efforts into these people. Going a step further, with newborn screening tests being accessible at multiple facilities, detecting carriers of SCT and SCD should be prioritised, creating a system where those with SCT also learning about how they should be aware of their condition, what the risks potentially could be in the future etc.

Mentioned in the analysis, newborn screenings and overall tests can be difficult to conduct and manage in regard to people of religious and financial reasons. In US, concerns on newborn screenings due to financial burdens and responsibility of parenting a sick child (Long et al. 2011) makes newborn screening difficult to convince and conduct on future parents, rather not wanting to know their trait status. A newer phenomenon that should be considered in improving the management of SCD and SCT people in the country, people migrating to a country, coming from a country with high risk of carrying SCT should be mandatory tested in order to control the disease awareness. The phenomenon on migration with those trying to avoid the new governmental health systems, can complicate the management for developed countries, until the migrated people register themselves in a hospital with a complication. The system is present with HCP and other medical people to collect data of a person when coming to the hospital etc. but the lack of using this to create a better management understanding for SCD people can be interpreted as lacking in finer margin. Not having greater

control when typing people in the system, understanding the seriousness of a SCD patient coming to the hospital with a crisis, a better system to quicker reaction through better management system should be done in the future. Having an international system, being able to manage every person living with SCD could be an initiative for those travelling or living abroad, in which easy access to their history of SCD experiences can be tracked outside the country if a need to go to the hospital due to a crisis is needed, helping their security and emotions to believe in the system.

6.5 Larger focus on SCT

Going into this research, my own familiarization with SCD at a fine level. I also understood through readings of former research that the majority of SCD patients are people of African, Middle Eastern, Caribbean and Indian heritage being minorities and usually lacks in attention and receiving help regarding medical interventions, as these are non-Western world countries.

Research in the past (Williams 2016) has pointed out the need in specifically SSA to urge towards having expanded and increased focus in African clinicians, researchers and health ministries to focus on getting a better service system and focus on the early detection of SCD, together with a larger roll-out of disease-modifying therapies. Although I will not deny that this is still crucial for prevailing the disease to spread any further than it already does, creating less focus on government actions and more focus on public awareness and learnings, especially in SSA does seem more necessary than receiving medicine to treat the disease. It should not be ignored that penicillin is a great preventer to SCD in lowering and controlling the under-5 mortality, but in regard to other medications and treatments that might be a financial burden to those living in poor societies, focus on preventing it in advance should be the target. This research and the findings also indicate that there is a greater need in general to know and understand the SCT and contain this instead.

With a combination of immigration, together with becoming more familiar of the potential futuristic health issue SCD can become, counting and structuring the management and containment of SCD patients, having them registered in their systems should be targeted in the nearer future. Better attention into knowing the haemoglobin genotype in order to lower the prevalence of SCD should be prioritised in countries with high prevalence per person. National Collegiate Athletic Association (NCAA) are being marketers for sickle cell testing for student-athletes being required in US, with thereto fact sheet for those affected by it, a professional attempt by associations in US are implemented. With SCT still being a risk-factor of sudden death, although the carrier is not necessarily affected by carrying the trait, the genetic and environmental influences should still be

considered (Key and Derebail 2010), hence the need to enhance the attention and awareness towards trait carriers too. The number of people not knowing their trait status is worrying and should be considered for future tests when making either prenatal or newborn screening, making parents also becoming aware of their trait status. The issues that might be avoided or can still happen is the personal aspect of wanting to target AA people more than those who have a sickled gene, which can then lean back towards mental and psychological struggles for those having a sickled gene, being a world-wide concern, and most probably an issue in non-common SCT countries.

A call to action towards funding herein governmental and national health institutional focus on making testing of genotypes more affordable if not necessary for the communities and those not having the test due to newborn screening being mandatory after their date of birth. Although certain tests in a social context have been presented as potentially critical for those being carriers of the trait, providing necessary information of those carrying a trait, herein the percentages of risk of giving a specific gene to one's child should be prioritised in the future in predominant cultures and populations such as Africans, Arabs, Indians and Hispanics.

6.5.1 Too much focus on R&D

The purpose of this research was to dive into the perspective of the SCD patients and advocacies and how they interpret the efforts being done by companies, organizations etc. how they are influential, how they are portraited in their efforts on creating awareness around SCD, and what they should focus on in the future for the SCD prevalence to lower. For now, R&D focusing on expanding and perfecting cures for treating SCD, limiting the cost and training people in this type of cure would be suggested and instead place focus on the general SCD affected people, helping with education and awareness. Especially in poor societies, focus on how to prevent crises etc. themselves would be more beneficial in the future. With current expenses for gene transplants and bone marrow treatment being unfairly high for the standard affected, focusing on R&D will be beneficial in the long run, but for the short run, focusing on the accessible testing of newborns, together with creating centres for management and education being fundamentally the target for now. Genotype tests for those who are still able to have children should be considered to be rolled out too.

6.6 What can this be used for?

This research creates a fine picture on the difference between high density areas of SCD affected, herein SSA, to low-middle density areas such as British Isles and US. My findings and discussion in its mere are fairly small in quantity, but high on quality, giving broader intellect into past, present and

future experiences of SCD patients in these given areas. This is beneficial for the overall understanding on where SCD awareness is and can move towards.

Although being less documented with few interviewees, I have shared light on different intercultural angles that should be considered in future research regarding SCD awareness. The lack of focus on burdens that are part of dealing with SCD, herein financial, mental, psychological, mental health on SCD affected, and time management (commuting) have been presented. Religious aspects and reasoning have had limited focus in previous research on SCD awareness, and having a higher religious focus on how their beliefs and attitudes could change from God having a reason to why people are being born with SCD, the awareness in the future should include focus on God providing them this information to stop the future of the disease to worsen. Furthermore, in regard to burden, this also calls for governmental interactions in better care of the secondary struggles of mental health and personalised insurances for SCD in the future, as living with SCD has higher risks of being forgotten and neglected by the system.

For a better understanding in the future on what specific strategies should be conducted in the future, aiming at countries in SSA where the prevalence is high should be done in order to get a more direct picture of how management and control of prevalence should be handled in regions, looking at each country's infrastructure and health system. I tried to shed light on the future to create more centres in other areas than larger cities, and with previous examples of lowered prevalence in certain areas of Africa, but stagnating prevalence in other areas, mostly in districts where no help close to people are accessible, the cycle of SCD prevalence will continue if only focus is in the populated areas.

Future research would need SCD patients, advocacies and HCPs sharing experiences from specific areas in their respective country have them share their ideas on how to create a better system to lower the prevalence and control SCD in the future. With SDG goals of lowering the prevalence of under-5 mortality rate, more focus and a call to action in educating people in SCD and SCT in SSA and other countries with total affected being denser per capita and lack of accessories and treatments available to help these should be prioritised.

7. Conclusion

This research has focused on answering the problem statement on how to make people more aware of SCD to lower prevalence and under-5 mortality of SCD patients, but also educate the public and HCPs in SCD, trying to eliminate stigmas and discriminations associated with SCD, a lot of opportunities and conclusions can be made.

Firstly, it can be confirmed that there is a larger lack of awareness on SCD in British Isles, US, and SSA. It is obvious that there are differences in how SCD is being managed when looking at the different areas of British Isles, US and SSA, and different themes were more important and relevant to touch on when diving into the interviewees and their experiences of being affected by SCD.

Systemic racism was evident in both US and British Isles, having discrimination towards SCD people of African descent sharing problematic healthcare in previous experiences, both personally and second-hand stories. Having shared an example as an African American being perceived as a drug-user when having a pain crisis showcases the systemic racism present in US. Because of the systemic racism and lack of knowledge when visiting ER and HCPs, people living with SCD are taking more care of themselves, and does not want to be helped by others, an issue that can create problems in pain management for both patient and HCP.

Stigmas was presented to be different from cultural and religious reasons. Stigmatisation towards understanding the labelled as difficult in medical research and hospital visits as adults, together with hidden stigmatization in childhood by not being invited to family gatherings and other children being afraid of the SCD person due to symptoms of jaundice and swollen hands, people in US and British Isles cultures are experiencing these stigmas and discriminations. SSA shares religious stigmas and discrimination towards entire families of SCD patients, with parents being cursed as performing witchcraft, making demon babies come into the world if they have SCD. The stigma of letting your child to die when being born and receiving confirmation of being a SCD patient, provides a great indication onto why under-5 mortality is very high in SSA compared to other places. Early death sentence is still a common issue and focus making people aware of the treatments and examples of people living a long life can help eliminate and get rid of this stigma. Educating people in districts and suburbs in SSA, making them aware on what they as parents or affected by SCD can do, will help create a better future for SCD patients. The process of awareness, herein eliminating SCD stigmas should be considered as a long-term process and should not be expected to become better on a short-

term process. Having advocacies speaking up about this to the public would help these stigmas to lower.

With 20 million living with SCD and approx. 300 million with a SCT, testing of genotype in countries with higher prevalence of SCD patients, herein SSA, should be prioritised in the future in order to lower SCD affected. Furthermore, in order to contain SCD and lower the under-5 mortality rates, poor societies should receive more help in the future. Expanding care centres to also target outside larger cities will also help lowering the under-5 mortality rate on people with SCD. Educating locals in churches to express the religious reasoning of providing knowledge about SCD should be made in SSA in order to manage under-5 mortality better in SSA as districts are struggling more than larger cities.

The target groups of SCD awareness are depicted as different. In cultures and places in the world where there currently is low prevalence of people affected by SCD, awareness towards the public should not be the aim, but instead have more awareness on education and knowledge on designated society groups of African descents being a priority, together with students educating to become HCPs or physicians in the future. In the US, utilizing the options to promote awareness through televised and/or radio advertisement could be a greater awareness project to the overall society, but having targeted awareness likewise as in British Isles on people of African descents. In SSA, awareness campaigns of people reaching out, visiting districts and villages should be prioritised to lower the prevalence by testing people in any relevant age. There is a greater lack of SCD care centres in the markets of the research, and in order to lower the prevalence and number of newborns with SCD, educational focus together with fundings and economical help is necessary.

There are multiple burdens presented as an issue for people affected by SCD. Financial burdens with medication being rather expensive cause them to spend money which they might not have. Social burdens in parents and patients being distanced from people when learning about their disease can affect both their lives. There are still mistrust in the SCD patients/HCPs relationship when going to the hospital or ER, knowing the uncertainty of misunderstandings, stigmatization, systemic racism and lack of knowledge about SCD, making SCD patients believe that they should act as experts while being in an emergency state. Informing colleagues, school principals and other relevant people to know about the condition should be a norm for those living with SCD. This will make those who are relevant in a given situation to provide help and share the knowledge in the future, so that they can help if needed. Selfcare and self-efficacy is something that SCD patients take seriously, and the less

help needed from others is wanted, creating struggles for others to understand and help before it might be too late. They see themselves as warriors, but wanting people to help, going to the ER feels like a loss to SCD, herein having much focus on their selfcare in order to prevent this.

There is a high focus on paediatric care which helps lowering the under-5 mortality, but there is a gap in helping and providing those becoming adults and living with SCD to transition from receiving paediatric care to receiving adult care. There is an overall lack of focus on SCD patients who are young adults, even though most people living with SCD experience issues and pain crisis when they are young adults around 18-30 years old.

The campaign made by SCDAA and Forma Therapeutics are correlating with the depiction of an American child living with SCD and the burdens that comes living with this disease, the burdens of the parents, the child, and how the society cannot depict the pains and sufferings that the people affected by SCD are dealing with. With their two PSAs they created attention and awareness to SCD, both visually and vocally descriptive by sharing and showcasing different challenges that people with SCD goes through. A comparative analysis of both of the PSAs and SCD interviewee participants was made, and although being advertised only in the US, it depicts the overall picture of how SCD children going to school are living, how a sudden crisis can change the entire reality. The experiences SCD patients goes through can be misunderstood and proclaims the attitudes and behaviours towards the awareness and trust there is in SCD patients when speaking up about them living with the disease. The lack of belief in the behaviour and attitude from people living with SCD is presented as lacking, which interviewees could agree to, not depending on country, culture and/or medical systems.

Future focus on having a better system with those living with SCD being treated better with more and quicker care should be done in the near future. Feeling unwanted and taken care of in the adult life is something that should be looked deeper into in the future in order to create a better SCD patient/HCP relationship. If own and second-hand previous experiences of going to the hospital or ER are negative with deaths and long waiting times as the standard norm, a greater mistrust will be created, having behaviours, beliefs, attitudes and actions towards the HCP from the SCD patient and vice versa being problematic.

Finally, this thesis paper should be used as a stepping stone for researchers to dive deeper into understanding the different angles on awareness from HCPs, haematologist with expertise in SCD, parents of an affected SCD child, and the people living with SCD, herein Sickle Cell Advocates, creating the best possible opportunities and strategies in order to make people become more aware of

SCD, and committing into lowering the prevalence of patients born with SCD in the future across the world.

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