Winston Churchill Memorial Trust Fellowship Report

Management of Sickle Cell Disease

By Norris Igbineweka
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Acknowledgement

I would like to thank the Winston Churchill Memorial Trust (WCMT) for helping realise this journey which took me to Nigeria and the USA investigating the management of sickle cell disease (SCD). I would personally like to thank Professor Akinyanju and all the staff at the National Sickle Cell Centre (Lagos, Nigeria), Dr Akinsulie and all staff at Lagos University Teaching Hospital (Lagos, Nigeria), Dr Temi Nwamkpele (Massey Street Children’s Hospital, Lagos, Nigeria) Professor Kola Abimbola (Seattle University, Washington), Dr Temi Abimbola (University of Warwick, UK), Mr Kunle Atinwore and his staff at the University College Hospital (Ibadan, Nigeria), Mr Kehinde Opatola, Miss Adenusi Toyin, Babalawo Adeboye, Chief Ogundiya Iroko Abimbola, Olu Amurie (Benin City, Nigeria), Osaretin (Lagos, Nigeria), Jackson (Lagos, Nigeria), Dr Ify Osunkwo (Children’s Healthcare of Atlanta, Georgia, USA), Dr Yang (Children’s Healthcare of Atlanta, Georgia, USA), Dr Peter Lane (Children’s Healthcare of Atlanta, Georgia, USA) Dr New (Children’s Healthcare of Atlanta, Georgia), Mrs Bose Omorogbie, Mrs Gwendolyn Wiley & Mr Wiley (St Christopher’s Hospital, Philadelphia, Pennsylvania, USA) and Dr Lewis Hsu (St Christopher’s Hospital, Philadelphia, Pennsylvania, USA). I am grateful to these people for helping towards the completion of this Fellowship and making me feel extremely welcome in my respective visits.
Itinerary Outline

Friday 18\textsuperscript{th} July 2008 – Arrival in Lagos, Nigeria
Monday 21\textsuperscript{st} July 2008 – Massey Street Children’s Hospital Lagos, Nigeria
Tuesday 22\textsuperscript{nd} July 2008 (am) – Lagos University Teaching Hospital (LUTH), Lagos, Nigeria
Tuesday 22\textsuperscript{nd} July 2008 (pm) – Sickle Cell Foundation, Lagos, Nigeria
Wednesday 23\textsuperscript{rd} July 2008 – Tuesday 29\textsuperscript{th} July 2008: LUTH & Sickle Cell Foundation
Wednesday 30\textsuperscript{th} July 2008 – University College Hospital Ibadan
- Herbalist 1 – Ilobo, Oshun State, Nigeria
- Herbalist 2 – Oyo State, Nigeria
Tuesday 5\textsuperscript{th} August 2008 – Benin National Museum, Benin City, Nigeria
Thursday 14\textsuperscript{th} August 2008 – Children’s Hospital of Hospital, Atlanta, Georgia, USA
Thursday 15\textsuperscript{th} August 2008 – Friday 22\textsuperscript{nd} August: Children’s Healthcare of Atlanta, Georgia, USA
Monday 25\textsuperscript{th} August 2008 – St Christopher’s Hospital, Philadelphia, Pennsylvania, USA
1. Background to choice of Fellowship Project: Why Sickle Cell Disease?

Sickle Cell Disease (SCD) is the first ‘molecular disease’ identified in humans and represents, for me, a prism of health and disease. As well as the molecular, it is a disease that carries significant psychological, social-cultural and environmental dimensions. Furthermore, advancements made in the understanding of SCD have helped to develop other areas of medicine, for example, in the field of genomics. Consequently, this is a very interesting condition! Despite this, since its discovery in 1910, there is still no universal cure for the disease.

1.1 Introduction

What is SCD?

SCD is a group of inherited conditions that affect the blood and various organs in the body. Central to the disease process, red blood cells, under certain conditions, acquire a sickle-shape (crescent moon shaped) and break down more easily (See Fig. 1). This results in various problems such as episodes of severe pain, infections and organ complications. In between episodes of illness, however, people with SCD can feel well.

In adults, the oxygen carrying component in blood - haemoglobin, consists of building blocks α and β-globin chains. SCD affects the β-globin chains. SCD encompass various subtypes such that sickle cell anaemia (HbSS) (the most common type of SCD) and sickle cell/C (HbSC) both produce structural malformations of the β globin chains. Whereas other subtypes such as sickle cell/beta thalassaemia demonstrate a quantitative deficiency in the amount of β-globin produced. All these different sub-variants show similar clinical symptoms but HbSS is generally considered as worse in terms of severity.

Fig. 1

![Fig. 1](image-url)
SCD is an inherited genetic disorder. If one parent possesses an abnormal gene, they are termed as carriers or having sickle cell trait. This is not the same as having the disease itself and they are no associated symptoms. To have SCD itself, he/she must inherit an abnormal gene from each parent. If both parents have the abnormal gene, there is 1 in 4 chance of them having a child with SCD. If one parent has SCD and the other partner has the trait then the risk of having an affected child is 1 in 2.

The gene mutation in SCD is caused by a substitution between the DNA bases (GTG for GAG) culminating in amino acid glutamic acid (GAG) being substituted for valine (GTG) (Fig. 2). The effect is that the production of sickle haemoglobin has increased tendency of forming polymers (rope-like structures) particularly under conditions that lack oxygen or water. Microscopically, the RBC becomes easily deformed, takes up its characteristic sickle shape and develops an inflexible structure poised with great difficulties passing through capillary blood vessels.

1.2 Genetic Epidemiology

The sickle gene (HbS) possesses an advantage: it protects carriers from succumbing to the severe cerebral malaria infection. The type of malaria infection associated with SCD is *Plasmodium falciparum* which is transferred by the female anopheles mosquito.
As a result, SCD is common in regions endemic with malaria i.e. Africa, Asia, Mediterranean basin and the Middle East (See Fig. 3a & b). Globally, sickle cell disorders and thalassaemias account for the most common and fastest growing genetic diseases. In the UK, approximately 12,500 people have SCD and in many urban areas in England, about 1 in 300 babies are born with the disease. In today’s globalised world, wars, migration and trade continue HbS gene dissemination and making it an increasingly important global health problem.

Fig. 3a & b

Distribution of the sickle cell gene

Distribution of malaria

1.3 Clinical Symptoms

Sickled RBC's are inflexible and adhesive. These two properties result in two major mechanisms which underlie SCD clinical symptomatology: a) vessel blockage (vaso-occlusion) and b) premature breakdown (haemolysis) of RBC's.

Vaso-occlusion causes the painful episodes so familiar to a person with SCD. Sickle RBC's get stuck in small vessels particularly of bones which causes the pain. The pain itself can range from mild to severe and can occur suddenly. However, this vaso-occlusive pain tends to occur intermittently such that episodes of illness are interspersed with times of feeling well.

In babies, symptoms do not begin until aged 3-6 months (before that age, newborns have a different type of haemoglobin called foetal haemoglobin (HbF) which is not affected by the sickle gene). Nevertheless, after this age, a common symptom of babies and young children is pain and swelling in the small bones of fingers and toes – termed ‘dactylitis’. Further symptoms related to the vaso-occlusive process in SCD are a susceptibility to infections like pneumonia due to malfunction of the spleen. In addition, ‘acute chest syndrome’ is a potentially fatal manifestation of SCD where the patient can present with chest pain, fever and shortness of breath due to blocked blood vessels in the lungs.
The other pathway behind symptoms in SCD is the premature breakdown of RBC's. This can lead to ‘anaemia’ where there is a shortage of RBC’s in the body. Symptoms of anaemia include tiredness, shortness of breath, faintness/dizziness and skin pallor particularly around the fingernails, eyelids and tongue.

There are also extra complications related to SCD such growth, development and nutritional problems in children, strokes and spleen problems. SCD can also affect blood vessels in back of the eyes, affect organs such as the lungs, heart and kidneys and may also lead to gallstones, leg ulcers and unwanted painful erections (priapism) in teenage boys and men.

There is a great deal of individual variation in symptoms e.g. quantity and frequency of pain episodes. Some people with SCD have frequent symptoms whilst others have it moderately and finally some in whom their SCD is hardly noticeable.

1.4 How is SCD diagnosed?

The diagnosis can usually be made by a blood test. This blood sample can be analysed with electrophoresis to determine which type of haemoglobin is present in the blood. In countries like the UK and USA, prenatal and antenatal screening programme for SCD are available.

1.5 What is the treatment for SCD?

Bone marrow transplantation can be an option of cure depending on resources, a matched donor and severity of disease. Here, normal blood making cells from a donor is given to a patient with SCD and as a result, the patient can continue to make normal RBC’s with normal haemoglobin for life. However, there are a number of significant limitations to this method of treatment such as the suitability of the donor, immune rejection and severe infection. Consequently, majority of SCD cases cannot be cured. In these circumstances, general treatment goals can be based on staying healthy and avoiding factors that may trigger sickling. These factors include lack of oxygen, dehydration, cold, hard exercise, fever and infection. It is therefore advised that people with SCD drink plenty of fluid, avoid over-exertion, avoid getting cold and prevent infections through antibiotic prophylaxis. When painful episodes are being experienced, treatment is directed with good hydration and a myriad of painkillers ranging from mild painkillers like ibuprofen taken at home to strong opiates like morphine administered in hospital.

In some situations blood transfusions provide a useful alternative particularly in severe anaemia or acute chest syndrome. The transfusion adds normal RBC's to the blood thus correcting the anaemia and preventing sickling. It cannot be used on a routine basis however, as side effects such as iron overload, immune reactions and infections are possibilities.
Another preventative measure in SCD management is the utilisation of hydroxyurea. This drug, when taken regularly, helps some people with SCD reduce their number of pain episodes and risk of getting acute chest syndrome. There may be serious side effects to this drug as it is a form of chemotherapy. Nevertheless, it does provide an interesting option to SCD management. Part of my Fellowship looked at patients with SCD attitudes to this drug and also finding out whether it is beneficial in the respective countries.

1.6 Outlook for SCD

Without intervention, people with SCD can die in childhood from problems like infection. Developing nations like Nigeria are more likely to experience this outcome as issues of malaria and malnourishment are more prevalent. However with management programmes in place such as a newborn screening programme and antibiotic prophylaxis, have significantly increased life expectancy such that average life expectancy in the UK as of 2003 for a person with SCD is 50 years. Indeed, clinicians are beginning to see patients live even much longer than this.

2. Aims

The purpose of my fellowship was to explore sickle cell disease (SCD) and its management in Nigeria and the United States of America.

The following points were specifically addressed:

- How individuals with SCD perceive and cope with their condition in their various communities.
- To gain an understanding of how SCD is perceived by its relevant population.
- Resources or services are offered to people with SCD and their families on a local and national level.
- Various techniques of managing SCD looking at both existing therapies such as hydroxyurea (hydroxycarbamide) and potential therapies.
- Suggested improvements towards the management of SCD from what was learned.
3. Demographics of Countries Visited: USA & Nigeria

Table. 1 Demographics of USA & Nigeria

<table>
<thead>
<tr>
<th></th>
<th>United States of America</th>
<th>Nigeria</th>
</tr>
</thead>
<tbody>
<tr>
<td><strong>COUNTRY</strong></td>
<td></td>
<td></td>
</tr>
<tr>
<td>Capital</td>
<td>Washington DC</td>
<td>Abuja</td>
</tr>
<tr>
<td><strong>Head of State</strong></td>
<td>President Barack H. Obama</td>
<td>President Umaru Yar’adua</td>
</tr>
<tr>
<td><strong>Land</strong></td>
<td></td>
<td></td>
</tr>
<tr>
<td>total: 9,826,675 sq km</td>
<td>total: 923,768 sq km</td>
<td></td>
</tr>
<tr>
<td>land: 9,161,966 sq km</td>
<td>land: 910,768 sq km</td>
<td></td>
</tr>
<tr>
<td>water: 664,709 sq km</td>
<td>water: 13,000 sq km</td>
<td></td>
</tr>
<tr>
<td><strong>PEOPLE</strong></td>
<td></td>
<td></td>
</tr>
<tr>
<td>Population</td>
<td>307,212,123</td>
<td>149,229,090</td>
</tr>
<tr>
<td>Age Structure</td>
<td>0-14 years: 20.2%</td>
<td>0-14 years: 41.5%</td>
</tr>
<tr>
<td></td>
<td>15-64 years: 67%</td>
<td>15-64 years: 55.5%</td>
</tr>
<tr>
<td></td>
<td>≥65 years: 12.8%</td>
<td>≥65 years: 3.1%</td>
</tr>
<tr>
<td>Population Growth Rate</td>
<td>0.977%</td>
<td>1.999%</td>
</tr>
<tr>
<td>Ethnic Groups</td>
<td>white: 79.96%; black: 12.85%</td>
<td>More than 250 ethnic groups. Most populous are: Hausa &amp; Fulani 29%, Yoruba 21%, Igbo 18%, Ijaw 10%, Kanuri 4%, Ibibio 3.5%, Tiv 2.5%</td>
</tr>
<tr>
<td></td>
<td>Asian: 4.43%; Amerindian, Alaskan or Hawaiian native: 1.15%. Hispanics (may be of any race/ethnic group): 15.1%</td>
<td></td>
</tr>
<tr>
<td>Language</td>
<td>English (official). Spanish, Hawaiian &amp; other languages are spoken by minority groups.</td>
<td>English (official), Hausa, Yoruba, Igbo, Fulani</td>
</tr>
<tr>
<td>Religion</td>
<td>Protestant 51.3%</td>
<td>Muslim 50%</td>
</tr>
<tr>
<td></td>
<td>Roman Catholic 23.9%</td>
<td>Christian 40%</td>
</tr>
<tr>
<td></td>
<td>Mormon 1.7%</td>
<td>Indigenous beliefs 10%</td>
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<tr>
<td></td>
<td>Other Christian 1.6%</td>
<td></td>
</tr>
<tr>
<td></td>
<td>Jewish 1.7%</td>
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</tr>
<tr>
<td></td>
<td>Buddhist 0.7%</td>
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</tr>
<tr>
<td></td>
<td>Muslim 0.6%</td>
<td></td>
</tr>
<tr>
<td></td>
<td>Unspecified 2.5%</td>
<td></td>
</tr>
<tr>
<td></td>
<td>Unaffiliated 12.5%</td>
<td></td>
</tr>
<tr>
<td></td>
<td>None 4%</td>
<td></td>
</tr>
<tr>
<td><strong>HEALTH</strong></td>
<td></td>
<td></td>
</tr>
<tr>
<td>Life Expectancy</td>
<td>male: 75.65 years</td>
<td>male: 46.16 years</td>
</tr>
<tr>
<td></td>
<td>female: 80.69 years</td>
<td>female: 47.76 years</td>
</tr>
<tr>
<td>Infant Mortality Rate</td>
<td>male: 6.9 deaths/1,000 live births</td>
<td>male: 100.38 deaths/1,000 live births.</td>
</tr>
<tr>
<td></td>
<td>female: 5.51 deaths/1,000 live births</td>
<td>female: 87.97 deaths/1,000 live births.</td>
</tr>
<tr>
<td>Fertility Rate</td>
<td>2.05 children born/woman</td>
<td>4.91 children born/woman</td>
</tr>
<tr>
<td>HIV/AIDS</td>
<td>Prevalence = 0.6%</td>
<td>Prevalence = 3.1%</td>
</tr>
<tr>
<td>People Living with HIV (2007)</td>
<td>1.2 million people</td>
<td>2.6 million people</td>
</tr>
<tr>
<td>HIV/AIDS Deaths (2007)</td>
<td>22,000 people</td>
<td>170,000 people</td>
</tr>
<tr>
<td>Total Expenditure on Health as % of</td>
<td>15% (World Health Organisation – 2006)</td>
<td>4.1% (World Health Organisation – 2006)</td>
</tr>
</tbody>
</table>
Nigeria

Nigeria is located in West Africa. It shares borders with Benin, Niger, Chad and Cameroon, and has a coastline to its west with the Atlantic Ocean. The Atlantic and Pacific Oceans both surround the USA East and West respectively (See Fig. 4a & b).

Fig. 4a & b

Whilst in Nigeria, I was mainly based in the south-western region in the city of Lagos but I also made visits to Oyo State and Edo State.

3.1 Nigeria & SCD

Lagos University Teaching Hospital (LUTH) is a 40-year old tertiary service teaching hospital and is the largest of its kind in Nigeria. Here, I was kindly welcomed by Dr Akinsulie and his haematology team. I participated in wards rounds seeing patients medically admitted, multi-disciplinary grand round presentations, teaching and outpatient clinics.
I visited outpatient clinics within local communities such as that of Massey Street Children’s Hospital which helped toward managing SCD in the local community. These were just a few of the public services for the management of SCD in Lagos, Nigeria. It was interesting to have the opportunity to visit The National Sickle Cell Centre (NSCC) lead by Professor Akinyanju was also an interesting place to visit. It is a non-governmental, non-profit making organization dedicated to the care and control of SCD in Nigeria. Here, I was given a tour of all the services provided such as its laboratory equipment, genetic diagnostic tools, counselling and voluntary services. Finally, I was able to visit the University of Ibadan in order to arrange a meeting with two herbalists who manage SCD for their local communities.

3.2 Main Findings

My experience of Nigeria was that it is a very lively country. Being a developing country, there were clearly infrastructural related problems like the lack of constant electricity. However, cities like Lagos carried a natural ‘buzz’ in the air. Generally, people did not wait for their government to provide for them but rather took a pro-active approach to develop a means of self-sustenance e.g. starting one’s own business. Consequently, faith also played large part in people’s lives. I generally had a great time in Nigeria taking time to enjoy its history, local food and scenery.

The prevalence of SCD in Nigeria is reflective of its large, dynamic population. One in every four people are healthy carriers of the sickle cell gene (HbAS).
This equates to 40 million Nigerians who are carriers in numerical terms and 150,000 babies are born each year. Institutions like LUTH and the NSCC were attempting to address these needs. LUTH provided services like beds for patients with acute painful crises and outpatient clinics with SCD. Massey Children’s Hospital, which was affiliated to LUTH, provided facilities for the Nigerian government to employ public health initiatives as well as easier access to healthcare for a population who were not situated near LUTH. Another positive strategy was the co-ordination of the private sector i.e. banks and mobile networks in funding medications like folic acid and anti-malarials making them affordable or even free for patients with SCD.

Another impressive aspect was that of the NSCC which responds to the World Health Organization’s (WHO) request for special dedicated centres for the prevention and treatment of SCD in areas where SCD is common. This centre pioneers interventional programs like prenatal and genetic diagnosis of SCD as well as providing collaboration with LUTH in areas such as research, counselling, educational and training courses.

One of the biggest challenges in the Nigerian setting was the issue of infection and malnourishment. Some of the patients which I saw had less than average body weight compared to that of unaffected individuals. Secondly, the impact of various kind of infection such as malaria had a worsening effect on the clinical severity of the patient’s disease. There was also the issue of socio-economic status which had an impact on infection risk and malnutrition. Improvements can be made in hospital to safeguard the patient’s infection risk e.g. providing mosquito nets for hospital windows and enhancing nutritional status. With this recommendation comes the ever present need for more government health funding in Nigeria. A large amount of equipment in LUTH needed replacing as they were old. This was in contrast to NSCC which had state of the art equipment. Questions also remain as to how malaria impacts individuals with SCD and to what extent the sickle cell carrier state protects against malaria?

My visit to the herbalists was very insightful. This was because it provided an alternative viewpoint to the understanding and management of SCD. With the help of a Yoruba-English interpreter, I was able to video-interview two herbalists separately on their role in managing SCD in their communities. The first herbalist lived in a small town named Illobu in Osun state. The herbalist named SCD as “lakuregbée” in which SCD was not seen as a disease of the blood but rather as a rheumatological condition “pain in the bone”. He explained concepts of how SCD is linked to what he called “abiku” in which children who have SCD are attached to malevolent spirits which are born and die soon after (repeater children). On the otherhand, the second herbalist, who lived in Oyo state, believed that SCD is linked to ‘unclean and dry’ blood. Both herbalists treat SCD with a combination of ‘medicines from the bush’ and incantations. It was interesting to see that these alternative views existed alongside medical science. From my meeting with these herbalists, I could see similar parallels with medical science perspective on SCD. However, there
were also stark contrasts. Despite this, both herbalists agreed that there is a role for medical science and herbal medicine to work in partnership. What is more interesting is that both herbalists individualise their treatment for each of their clients as they believe that each person has unique ‘blood’ therefore treatment should be tailored towards this. This sounds similar to the direction of western biomedicine with the progression of gene therapy for diseases like SCD.

### 3.3 USA

Geographically, the USA is 38° North and 77° West of the Equator and Greenwich Meridian respectively. It shares borders to the North with Canada, and to the south with Mexico. The Atlantic and Pacific Oceans both surround the USA East and West respectively (See Fig. 6a & b).

The USA is ranked 13/182 on the United Nations Development Plan Human Development Index. It is one of the most developed countries in the world with a GDP of $46,400 per capita and a life expectancy of 78.17. The health status in the USA compared with other countries is excellent with 15% of its total GDP going towards health. Its overarching goals are to 1) increase the quality and years of healthy life and 2) eliminate health disparities. Despite this, 13.5% of the population remained below the poverty line, 16% of the population had no health coverage in spite of government health insurances like Medicaid, and the life expectancy of people of black ethnicity still averaged 6 years less than those of their non-hispanic white counterparts.

Fig. 6a & b

There is always so much to do in the States. I was taken on sightseeing tours by some wonderful people. I was particularly impressed with US history and
the UK’s involvement as well as scientific discoveries like that of electricity by Benjamin Franklin.

3.4 USA & SCD

My time in the USA was spent in Children’s Healthcare of Atlanta in Georgia and St Christopher’s hospital in Philadelphia. Children’s Healthcare of Atlanta is a consortium of three hospitals in Atlanta with 520 staffed beds which serves more than half a million patients annually. It is also the largest Medicaid provider in the state of Pennsylvania. Specifically, its division for blood disorders serves more than 1500 patients with SCD. Whilst there I participated in patient evaluations and got to speak to individuals with SCD and their parents. I observed patient counselling, sickle cell education and the designated 24 hour acute care for SCD. Children’s Healthcare of Atlanta is also renowned for its cancer centre which has performed more blood and bone marrow transplants (BMTs) for SCD than any other program in the country. To date, 25 children with SCD have been treated through matched-sibling BMTs with a 96% disease free survival rate.

St Christopher’s is a 189 bed paediatric hospital which treats over 70,000 patients annually. I was attached here to a sickle cell team in which I was able to observe the coordination of acute care, routine and follow-up deduction. The family centred psychosocial support available to patients, I found was very positive. This program recognised that SCD extended far beyond biological remits and attempted to address the psychological and social burden SCD can place on individuals and families.

I also observed the utilisation of the drug hydroxyurea within the outpatient setting. Hydroxyurea has been proven to reduce the frequency of hospitalizations, incidence of painful crises, acute chest syndrome and the need for blood transfusions. However, there is existing concern related to possible long term side effects like cancer development. It was interesting to see the negotiation of this risk within the outpatient setting. At the time, St Christopher’s hospital had about 100 out of 600 patients on hydroxyurea of which it was estimated to be effective in about 70-80%. The remaining patients, in which effects were not seen, fell into categories of either being a biological non-responder to the treatment or not concordant with the treatment. Amidst the uncertainties over long term toxicities, it was clear to see how “impressive” the impact hydroxyurea was having on many patients quality of life. Often this was termed in the States as the ‘hydroxyurea smile’. On a psycho-social level, hydroxyurea helps the individual with SCD reduce the amount of interruption to their life due to pain crises, which is significant progress for the individual with SCD. Despite this, questions remain over whether hydroxyurea is being offered and utilised enough in the clinical setting? If not, what exactly are the barriers hindering its utilisation? From my observations, it seems like it is not only patient and family concern over long term toxicity but rather how comfortable the physician feels in prescribing hydroxyurea and their own interpretation of risk.
4. Conclusion

My time in Nigeria and USA was enjoyable and informative. It was interesting to note that individuals with SCD I spoke to in both Nigeria and USA, all felt that they possessed a stigmatizing disease and felt stigmatized by society. In Nigeria, SCD’s relationship with traditional concepts like “abiku” persist amongst certain community members and carry stigmatizing power. SCD’s relation with ethnicity and socio-economic factors in USA also present issues of stigma. I witnessed attempts to raise awareness for the condition in both countries. However, campaign strategies to raise awareness, which might be beneficial in countries similar to the USA, may have to be altered to suit the social context in countries like Nigeria. In Nigeria, simply raising awareness may lead to further stigmatisation and marginalisation of people with SCD. For example, increased awareness can trigger attitudes like “you must avoid marrying a sickler” or exaggerated ideas such as “SCD is a dreadful disorder to be avoided at all costs”. These were some of the concerns voiced by individuals with the condition in Nigeria. Consequently, because of the paradox in raising SCD awareness in different countries, cultural context must be appreciated when promoting public health campaigns.

I asked individuals in SCD both in Nigeria and USA whether they viewed themselves as a ‘sick’ or ‘healthy’ person. From my analysis, most individuals saw themselves as both ‘sick’ and ‘healthy’ persons. However, individuals living in the USA were more likely to view themselves as ‘healthy’ whereas in Nigeria, individuals were more likely to view themselves as ‘sick’. This suggests that clinical severity or perception of clinical severity of SCD may be influenced by both biological and environmental indices.

My time spent with the herbalists emphasised the need for individualised therapy in SCD as each patient has different genetic make-up and environmental factors which influence the clinical course of their disease. With progress in gene therapy, SCD management will possibly be tailored towards the individual. Also listening to herbalist viewpoint on treatment with SCD might shed some light on western biomedical management of SCD.

In both Nigeria and USA, there have been dedicated centres for SCD and this has only been a good thing, for example, in terms of optimising acute care or prenatal genetic diagnosis in SCD. I also saw the importance of multidisciplinary involvement in SCD in both countries and the provision of joint speciality clinics like SCD and orthopaedics, is vital.

As was mentioned earlier, the use of hydroxyurea has had immensely improved the quality of life for patients with SCD in the USA and is currently the only approved pharmacologic therapy for SCD. Questions arise as to what is the applicability of hydroxyurea in the global health setting? With therapies like bone marrow transplantation expensive, can hydroxyurea be effectively utilised in countries like Nigeria where the bulk of the burden of disease is? If not, what are the barriers hindering this and how can we address this?
However, it is clear that more experience is needed with the use of hydroxyurea and it is still unclear to what extent the associated risks with hydroxyurea are. Either way, it may be beneficial to have a regional or possibly worldwide registry monitoring the effects of hydroxyurea, keeping a profile on laboratory markers and side effects.

I would like to thank WCMT and all my hosts for an enlightening experience exploring this very interesting global health disease. It is important that the gap between high and low resourced countries towards the management of SCD is reconciled. This can be achieved through long term collaboration between these regions appreciating cultural nuances, thus impacting research and clinical care.
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