



# A Proposed Model for the Use of Technology in Managing Sickle Cell Disease in Children between the Ages of Five to Fifteen.

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## ARTICLE INFO

### History:

Received: 1<sup>st</sup> May, 2022

Accepted: 5<sup>th</sup> June, 2022

Received in revised form:

10<sup>th</sup> July, 2022

Published: 28<sup>th</sup> August, 2022

## Keywords

Model, Managing SCD, Sickle cell disease, SCD in Children, Technology and SCD

## ABSTRACT

This research work, presents the ideas and a simple proposed model that can be used to develop a system worthy of managing and monitoring Sickle cell disease (SCD) in children between the ages of five to fifteen years. It is an interdisciplinary research involving life sciences and computer science (software engineering), as it is that technology has moved into almost all fields thereby simplifying tasks. Medical practitioners are involved in knowing the basic factors of the disease (symptoms, severity, cure, and other factors related to the disease) alongside, reported literatures found in text, journal magazines and other sources. Data was also collected from few selected clinics aiding more understanding about the disease. In addition, the work gives a comprehensive review on the general knowledge of the disease and promotes awareness by involving technology in the management of the disease. Use of phones and tablets/palmtops is now very common among children of the said range, which can help in operating the application when implemented. Whereas the children may be left bored or involved in playing games that may have negative impacts on their health and education. The disease itself sometimes deals with psychology and mindset of the children, so they need to be strengthened.

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## 1.0 Introduction

Sickle cell Anemia, is partly hereditary disease that can be inherited from parents whom might have the SS/AS blood genotype, where the offspring stands a high chance of becoming a carrier (possessing the genotype SS). It is a serious health disorder, with high mortality rate especially in Africa and in children before their adolescence/adulthood; this can be at the result of poor access to medical care or lack of awareness on the community side. With the advent of technology, internet, mobile applications, TV and Radio stations, are now used for public enlightenment thereby getting up to rural settlers.

Sickle anemia is now seen as a disorder and of generic type which affects the red cells components, having serious morbidity and mortality rate. The sickle hemoglobin is termed as a structural variant normally affecting the adults' hemoglobin according to Williams and Chakravorty (2015). Sickle disorder usually encompasses series of pathological genotypes yielding from the inheritance of HbS. It affects millions of people worldwide, and much number of infants born with the disease in Africa die before the age of five (Aygun and Odame, 2012). It is projected that more than 200,000 children are born with the disease annually in Sub-

Saharan Africa (Makani et al., 2011). The UN General Assembly has now identified the disease as a global public health concern for the high morbidity and mortality caused by the disease as well significant social and economic impact that it results (United Nations General Assembly, 2009).

The disorder was first reported by Herrick (1910) even though reports suggest prior description of the disorder; which is the result of homozygous and compound heterozygote inheritance of a mutation in the beta-globin gene. Single pair point mutation (GAG to GTG) results in the substitution of the amino acid glutamic acid (hydrophilic) to Valine (hydrophobic) in the sixth position of the beta-chain of hemoglobin referred to as hemoglobin S (HbS) Inusa (2019). Phenotypic variation in clinical presentation is a unique feature of the disease despite a well-defined *Mendelian* inheritance, which is the first to be molecularly classified as described by Pauling and Anecdotal (2004) and confirmed to be due to a single amino acid substitution by Ingram, V.M. Anecdotal (2004) almost 60 years ago.

## 2.0 Managing the disease

Sickle cell disease can be managed by several ways including microscopic imaging or biochemical approach. Sickle cell disease is

commonly monitored by taking sample of blood and examining it under the influence of suitable laboratory devices. However, the method has several notable shortcomings including time-consumption and difficulty in capturing important changes that may lead to permanent cell damage, and further, these changes can't be monitored in real time fashion. Most of the clinical centers are aimed at minimizing, avoiding and managing the effects of the disease especially in rural areas and in children at teenage ages. However, they lack effective support from government and individuals in achieving this. Hence, there is need for educating the public and their subjects. Information and Communication Technologies such as TV programs, Websites, Mobile applications, Social media, Telecommunications, etc. are required in management of sickle cell anemia.

The aim is to come up with a simple model for handling Sickle Cell Anemia in children between the ages of 5 to 15, which can later on be used in developing a full-fledged system and may be expanded to capture even adults suffering from same disease. The objective of this study is to investigate causes, severity, symptoms of various cases of the disease, as relevant data is collected from some clinics defined for the purpose of the studies, and to come up with a model that will guide parents in reducing, managing and proper handling of the affected/victims of the Sickle Cell disease.

### 3.0 Related Literatures/studies on the disease

UN general assembly usually acts like an intervention/aiding body for all diseases identified to a global treat. As quoted in their 2009 release: “the disorder is identified to be a global public health concern due to the morbidity and mortality caused by the disease and the significant social and economic impact that it results (United Nations General Assembly, 2009). Normally, the flexible, round red blood cells move easily through blood vessels. In sickle cell anemia, the red blood cells are shaped like sickles or crescent moons. These rigid, sticky cells can get stuck in small blood vessels, which can slow or block blood flow and oxygen to parts of the body (Sickle Cell Anemia - Symptoms and Causes, 2021). The disease is also caused by a mutation in the gene that tells the body to make the iron-rich compound that makes blood red and enables red blood cells to carry oxygen from the lungs throughout the body (hemoglobin). In sickle cell anemia, the abnormal hemoglobin causes red blood cells to become rigid, sticky and misshapen. Both mother and father must pass the defective form of the gene for a child to be affected. If only one parent passes the sickle cell gene to the child, that child will have the sickle cell trait. With one normal hemoglobin gene and one defective form of the gene, people with the sickle cell trait make both normal hemoglobin and sickle cell hemoglobin. Their blood

might contain some sickle cells, but they generally don't have symptoms. They're carriers of the disease; however, it means that they can pass the gene to their children” UN general assembly, 2009 issue).

### 4.0 Sickle Cell versus Children

It is observed that there is rapid increase of nocturnal enuresis in children with sickle cell, especially male with HbSS; and the reason is not yet known. Children with the disease, frequently passes large quantities of dilute urine and possess *nocturia* (Hann et al., 2012). According to Robin and Miller (2018), children with SCD may experience all or some of the following complications:

1. Poor count of red blood components, or anaemia. With symptoms including: paleness, often seen in the skin/body surface, lips, or nail beds, weakness, dizziness, short of breath, feeling lightheaded, so irritable, difficulty in paying attention, and rapid heartbeat
2. Possessing jaundice (skin and whites of the eyes looking yellowish). Usually happens because the sickle-shaped red blood cells break down faster than other cells.
3. High risk for problems like leg pains, bone or joint damage, gallstones, kidney damage, eye damage, and retardation in growth.
4. Children with sickle cell disease can have an issue that requires immediate care by a physician, such as: Acute chest syndrome, aplastic crisis, Hand-foot syndrome, Infection, Priapism, Splenic sequestration crises, Stroke.

### 5.0 Methodology, Data Collection and sample analysis

In this research work, interviews, and questionnaires, were adopted in obtaining data and relevant information from patients and medical practitioners in addition to available secondary data on relevant websites, journal articles, textbooks and other sources. Some sample questions that were included in the semi-structured questionnaire were the following: How many registered cases of sickle cell for a defined period, number of males and female victims, level of education for parents and the children, awareness level of the disease before marriage, treatments offered, successful cases, methods applied, suggestions/counseling's offered, etc.

#### 5.1 Sample Selection

Sample of primary and secondary data were collected from the few selected clinics (names withheld for confidentiality purposes). Purposive sampling was used to develop the sample of the research under discussion; it is quoted from Freedman saying: “the method, which belongs to the category of non-probability sampling techniques, sample members are selected on the basis of their knowledge, relationships and expertise regarding a research subject

(Freedman et al., 2007)”. In the current study, the sample members selected have relationship with the phenomenon under investigation, sufficient and relevant work experience in the field of sickle cell management, active involvement in several sickle cell management initiatives and partnerships, as well as proven research background and understanding of raw data concerning the disease.

### 5.2 Data Analysis

Here the concept of “content analysis” was chosen to analyze the data which was gathered from personal interviews. According to Moore & McCabe (2005), “this is the type of research whereby data gathered is categorized in themes and sub-themes, so as to be able to be comparable, however, human error is highly involved in content analysis, since there is the risk for researchers to misinterpret the data gathered, thereby generating false and unreliable conclusions Krippendorff and Bock (2008)”

### 5.3 Interpretations on the data

#### 5.3.1 Number of Registered Cases of Sickle Cell considered from a particular clinic, Between the Ages of 5 to 15 years.

About 200 cases of sickle cell were considered from the clinic in the span of 10 years. The following are the details. The table shows that high number goes to male gender as compared to female gender, and that female gender responds to treatments faster than the male gender. (See the figures and the percentages in the table above).

Table 1: Cases by gender and response to treatments

Gender	Number of cases	Percentage (%)	Successful treatment	Unsuccessful treatment
Male	110	55%	67	43
Female	90	45%	75	15

Table 2 shows the collected data on the age range, registered cases percentage and status (weak/strong) with respect to the patients (Children). Table 3 shows the SCD conditions versus number of cases and respective percentages, where by Sequestration crises is realized having high number of registered case and Deaths with least number. The total number of death cases was found to be 34, having mortality rate of 17%. Children with Painful (**Vaso-Occlusive**) crisis were realized to have 56 cases at 27.2% of the overall number of cases. The number of cases with Sequestration crisis found to be 70 at 35%, lastly those with Aplastic cases were 40 at 20% of the total number.

Table 2: Presentation by Age Group of the Registered Cases from 5 to 15 years

S/N	Age (5-15)	Registered cases	Percentage	Status
1.	5	27	13.5%	Strong
2.	6	23	11.5%	Weak
3.	7	18	9.0%	Strong
4.	8	21	10.5%	Weak
5.	9	19	9.5%	Weak
6.	10	19	9.5%	Weak
7.	11	17	8.5%	Strong
8.	12	18	9.0%	Weak
9.	13	9	4.5%	Weak
10.	14	18	9.0%	Weak
11.	15	11	5.5%	Weak
<b>ToT</b>		<b>200</b>	<b>100%</b>	

Table 3: SCD Condition, number of cases and percentage

Condition	No. of cases	Percentage
<b>Deaths</b>	34	17
<b>Painful (Vaso-Occlusive) Crisis</b>	56	28
<b>Sequestration Crisis</b>	70	35
<b>Aplastic Crisis</b>	40	20
<b>Total</b>	<b>200</b>	<b>100</b>

### 5.4 Commentary from Experts

According to experts, “there is a recently developed micro technology, offering simple, rapid, and affordable screening of SCD and have the potential to facilitate universal screening in resource-limited settings and developing countries. On the other hand, monitoring of SCD is more complicated compared to diagnosis and requires comprehensive validation of efficacy. Early use of novel micro devices for patient monitoring might come in especially handy in new clinical trial designs of emerging therapies”(expert bulletins on SCD 2020). Table 4 shows some recently developed models for SCD screening/testing. Figure 1 shows the operation of the emerging technologies for Sickle cell diagnosis.

In the year 2012: “Tsai developed a microfluidic platform with an inner surface coated by a confluent monolayer of endothelial cells for modeling vascular occlusion and thrombosis in hematologic diseases, such as SCD, and studied the flow behavior of whole blood samples from two patient populations, receiving (HU+) and not-receiving (HU-) hydroxyl-urea treatment, revealed that HU+ blood achieved higher average velocities within the channels.

Table 4: Recently developed models for SCD screening/Testing

Technology	Pros	Weakness/cons	Efficiency	Reaction	Accuracy	LOD
<b>Paper-based hemoglobin solubility</b>	“Simple fabrication, ease of use, uses natural color of blood, no batching”.	“Failure to distinguish between HbSC and HbAS. Blood clotting interferes with the test. Interpretation of results is susceptible to human error”.	19.5 mins	93.5%a	96.7%a	NA
<b>Lateral flow immunoassay (HemoTypeSC)</b>	“Ease of use. Absence of auxiliary equipment”.	“Complex fabrication. Interpretation of results is susceptible to human error. Hb quantification is not supported”.	1.9 mins	98.9%c	98.8%c	HbA: 41%; HbS: 1%; HbC: 2%
<b>Lateral flow immunoassay (Sickle SCAN)</b>	“Ease of use. Rapid results. Absence of auxiliary equipment”.	“Complex fabrication. Interpretation of results is susceptible to human error. Hb quantification is not supported”.	2.1 mins	98.9%c	98.9%c	HbA: 41%; HbS: 1%; HbC: 2%
<b>Lateral flow immunoassay (HemoTypeSC)</b>	Ease of use. “Absence of auxiliary equipment”.	“Complex fabrication. Interpretation of results is susceptible to human error. Hb quantification is not supported”.	21Mins	99.9%f	99.9%f	HbA: 2.7%; HbS: 3.3%; HbC: 1.3%
<b>Microengineered Electrophoresis (HemeChip)</b>	“Low cost. Ease of use. Robustness. Rapid testing. Results comparable to standard electrophoresis tests. Possible integration with mobile devices. Works on the principles of clinical standard electrophoresis test”.	“High HbF concentration present in newborns less than 4 weeks of age may affect test results”	<9 mins	88–99%i	83–90%i	HbS: 10%; HbF: 10%; HbA:10%; HbC/A2: 3%

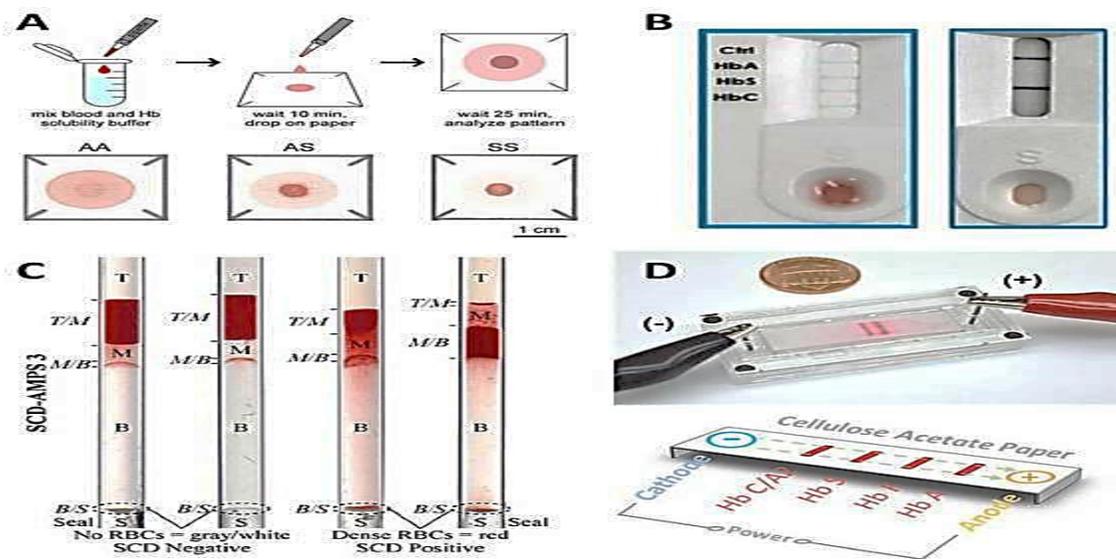


Figure 1. The operation of the emerging technologies for sickle cell diagnosis (Tsai, 2012)

Moreover, HU– samples caused greater levels of obstruction indicating the efficacy of the drug in SCD. Furthermore, interaction of TNF- $\alpha$  activated Human Lung Micro-vascular Endothelial Cells (HLMVEC) with TNF- $\alpha$  activated leukocyte resulted in a significant increase in the obstruction of flow as well as a decrease in flow rate. Despite their advantages in high fidelity mimicking of microvasculature, endothelium-based models are not yet widely applicable to large numbers of patients. This is because of the requirement for labor-intensive and technically challenging tools and a need for a constant supply of cultured endothelial cells” Tsai (2012). Figure 2 shows Emerging POC technologies for SCD monitoring.

### 5.5 Interventions for Sickle Cell Disease

According to Lobitz (2016), “Sickle cell disease is a very complex disorder, alluding to virtually all the areas of medical sciences. Nevertheless, basic preventive and therapeutic interventions in patients suffering from SCD are extremely simple. Minor behavioral changes may reduce the incidence of several complications. Wearing warm clothes prevents pain crises. Vaccinations and penicillin prophylaxis virtually eliminate life threatening bacterial infections. Patients with febrile illnesses require urgent medical care. Parents who are able to palpate spleen size can diagnose splenic sequestration at home at a very early stage and seek medical attention immediately. Most patients who have

internalized this simple code of conduct show a great improvement in their condition” Lobitz, (2016). He further added that, “The groundwork is laid in childhood. It is up to the pediatricians to communicate this information and knowledge during childhood and adolescence. Experience has taught us that the transition to adult care is often inadequate and that those patients who get lost at this critical stage of care have not understood the gravity of their individual situation resulting in serious consequences for their health. However, in everyday life it is sometimes virtually impossible to support children and adolescents to self-manage their condition. For the first time ever, DHIs provides new opportunities to support self-management behaviors Lobitz, (2016)”. On another narration “DHIs may facilitate daily and recurrent routines such as drug intake or appointments through simple reminders. However, at an advanced level, they may also help the patient to cope better with their disease. This may be realized through a number of modes of delivery such as mobile training programs, disease specific social networks using secure communication channels, diaries and blogs Gray et al. (2014)”. Figure 3 shows identification of digital health interventions in management of SCD.

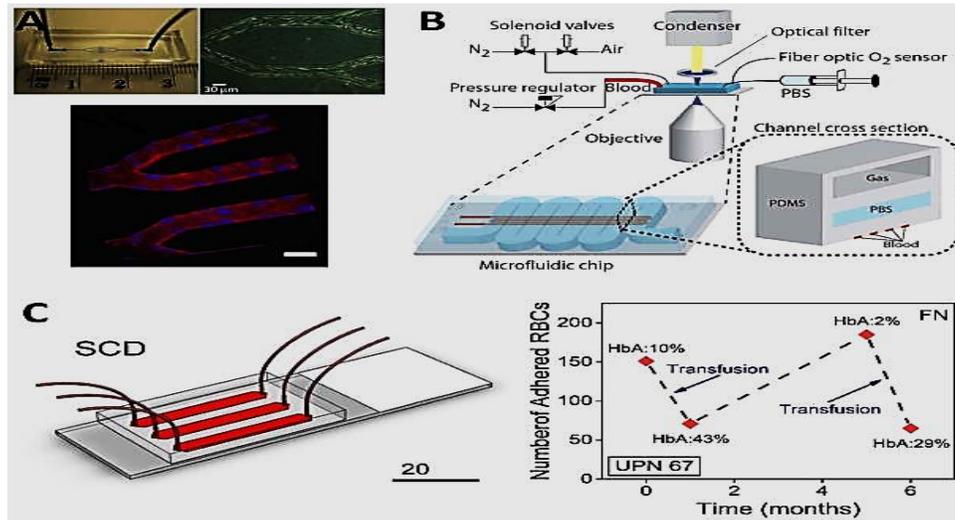


Figure 2. Emerging POC technologies for SCD monitoring (Tsai, 2012)

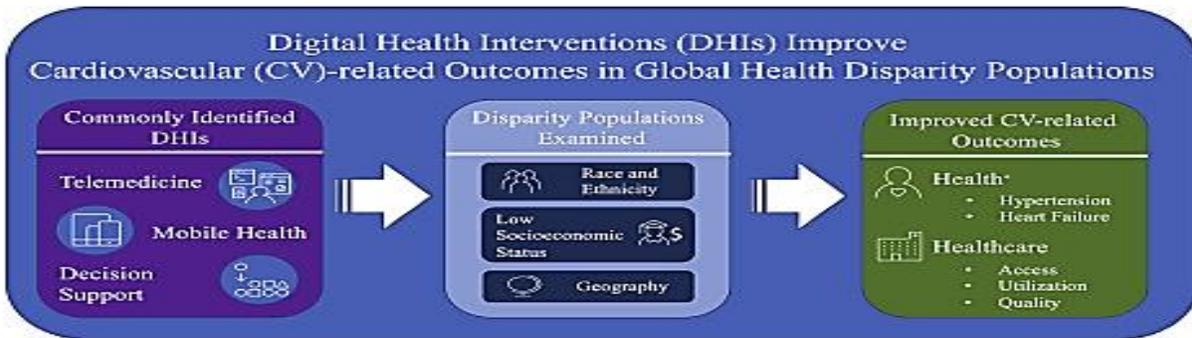


Figure 3. Identification of digital health interventions in management of SCD (Steele Gray, 2014).

Steele Gray (2014) observed that: “It is an absolute prerequisite to awaken a patient’s interest and motivation in their own disease to establish understanding and create awareness for disease specific needs. DHIs, in particular apps, for children and adolescent with SCD aim to create an improved sense of self and disease in the very first instance”. While according to Creary (2014): “Despite the enormous prevalence of SCD, there are still a limited number of SCD apps available to patients suffering from SCD, their families, peers and caregivers as well as a paucity of publications on SCD apps. Nevertheless, so far research has shown promise for the acceptability and usability of SCD apps aimed at tracking multiple symptoms such as pain and tiredness Steele Gray et al. (2014) facilitating reminders for medication Creary et al. (2014)”, enhancing communication with healthcare providers and general

health management (Steele Gray et al., 2014) and delivering therapeutic interventions such as cognitive behavioral therapy (Schatz et al., 2015). “Indeed, research and industry led apps have chosen diverse approaches to address SCD. Consequently, the diverging SCD related apps on the market pursue a variety of objectives. Most apps have several functions and behavioral targets, but they can be classified on the basis of their primary objective (Schatz et al., 2015)”.

### 6.0 Proposed model for the use of technology in managing SCD in children of 5 to 15 years.

Figure 4 shows the proposed model for the use of technology in managing SCD in children of 5 to 15 years.

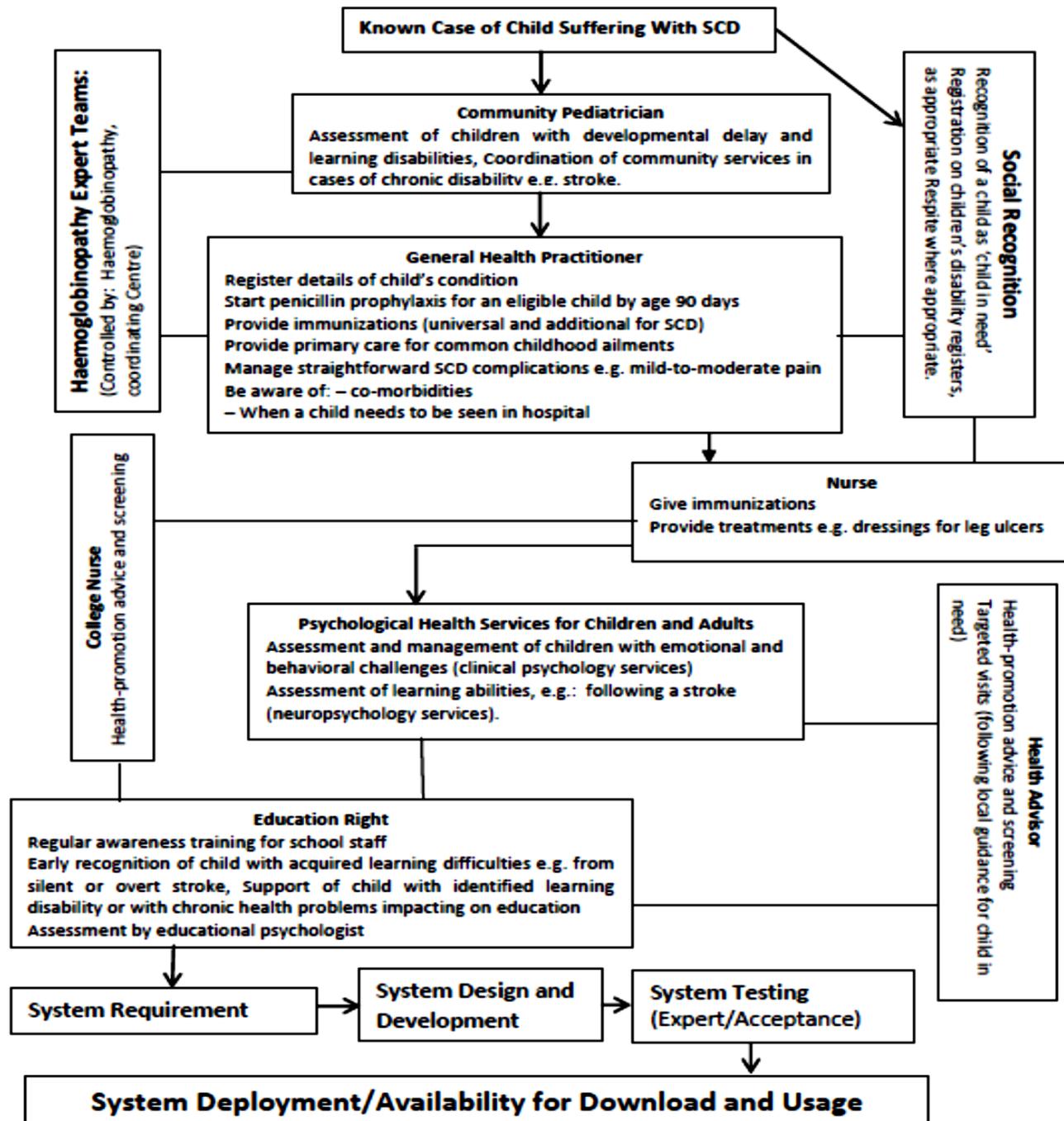


Figure 4: Proposed model for the use of technology in managing SCD in children of 5 to 15 years.

The model above presents different components including: **Haemoglobinopathy expert teams:** for blood examinations, **Community pediatrician:** responsible for assessment of children with developmental delay and learning disabilities, coordination of community services in cases of chronic disability e.g. stroke, **Social Recognition:** responsible for handling child needs and registration, **General health practitioner:** tackling registration details of child, issuance of medications, primary/first aid care, and managing complications of the disease, **Nurses:** tasked for giving immunizations and enforcing treatments, **Psychological health services for children and adult:** Assessment and management of children with emotional and behavioral challenges (clinical psychology services) assessment of learning abilities, e.g.: following a stroke (neuropsychology services). Others includes: Health advisor, Education right and last portion dealing with components of system development stages. The model can be suitable for the cases considered during the process of this work from some selected clinics in Jalingo, Taraba State, Nigeria (To be verified by stake holders during development, testing and deployment).

ICT activities for management of patients with sickle-cell anemia should be based at the primary health-care level, with emphasis on programs that use simple, affordable technology in reaching a large proportion of the community. Examples include: public education, detection of genetic risks in the community by recording family history and on every contact of the patient with the health system paying due attention to that history, health and genetic counseling, and immunization against infections. The personnel involved in medical care will most likely be primary health-care practitioners with task-oriented basic training in sickle-cell anemia. In order to accomplish these activities, primary health-care providers should have adequate links with secondary and tertiary levels of care for consultation and adapting ICT tools such as website, mobile application, SMSs, emailing etc. It is a known fact now, that many people (a good number of the population, young and old) can operate mobile phones and by extension android phones capable of hosting simple and complex applications, implying that implementing the above model into a full-fledged system may not have issue with acceptance and operability to the target populace.

In the next paper/report, other components of the model including: Class diagrams, activity diagrams, sequence diagram, and all functionalities of the system will be provided. Other requirements like: Database, host environment, programming language, and required platforms will also be presented. Usability test, expert/acceptance reports, screens of the working system will also be captured.

## Conclusion

It is obvious that technology is in every aspect or profession with the soul aim of simplifying task from analog world to digital world or simply manual to automated form, with the advantage of being fast, affordable/cost effective (results/solution), accuracy among other unlisted advantages. Converse effect of the disease, due to ignorance and poor medical accessibility, leads to its mortality and morbidity rate increasing very high especially in rural areas. Looking up to the proposed model, stake holders must come together in managing the disease especially: Educationist, general health practitioners, medical psychologist and health advisors, in addition to medical and hemoglobin expert teams. On their side (System analyst and Developers), should consider coming up friendly and useful system that can capture children of the said range, having suitable functions (medical and social) that can optimally help in managing the disease in consideration.

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