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Sickle Cell Disease (SCD) and Stem Cell Therapy (SCT): Implications for Psychotherapy and Genetic Counselling in Africa

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Additional information is available at the end of the chapter

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1. Introduction

Sickle Cell Disorder (SCD) is an inherited disease of red blood cells which has no widely available cure (Bernaudin, Socie, Kuentz, et al., 2007). While current medical therapies can make a significant difference in short-term effects (i.e. to relieve pain symptoms, prevent infections and manage complications such as eye damage, and strokes; and control complications), the progressive deterioration in organ function results in increased mortality and decreased quality of life among affect persons in Nigeria. Presently, blood and bone marrow stem cell transplant appear to be the only viable option for its eliminating. This option is hugely expensive and unaffordable for the vast majority of the affected Nigerian families since most of them could barely provide for the general routine medication therapies of the patient. Little attention is being given to the management of this disorder in Nigeria as compared to diseases such as malaria and polio myelitis. Institutional research attention and international funding support towards the search for ways to predict the severity of and for curative therapies of this disorder are also limited in Africa.

Globally, sickle cell disorders (SCD) affect millions of people of all races throughout the world. About 80% of affected children are born in developing countries and about 50 – 80% of children with SCD die each year in low – middle income countries. Nonetheless, its magnitude in Nigeria and Africa on the whole is alarming. Nigeria has the largest burden of SCD in Africa (*see table 1 for a presentation of the progress report*). At least 40 million Nigerians are carriers (AS) versus 2 million Americans. Over 150,000 Nigerians are born each year with sickle cell anaemia (SS) versus 2,000 in America (Akinyanju, 2009). Numerous families

in Nigeria have lost loved ones to this red blood cell disorder. About 80,000 people are living with SCD in USA versus estimated $\pm 1,000,000$ in Nigeria (Akinyanju, 2009).

1916 - 1945	Virtually nil > 4y
1946 – 1965	Virtually no adults. Known as paediatric disease
1966 – 1985	Many adolescents and young adults
1986 – 2010	Many adults as parents and in workforce

Table 1. Progress of Nigerians with SS

Molineaux et al (1979) noted that there is no other known inherited disorder present at such high frequency in a large population and of comparable severity as sickle cell anaemia in Africa. With rising standards of living and control of malaria, sickle cell anaemia may become an immense medical, social and economic problem all over Africa (see table 2 below).

Indigenous	Imported
<ul style="list-style-type: none"> • Sub Saharan countries north of Zambesi River • Eastern Saudi Arabia • Some States in India 	<ul style="list-style-type: none"> • Mediterranean Basin e.g. Greece, Sardinia • USA & Canada • Brazil, Belize Columbia etc • Cuba, Jamaica, Haiti, Barbados, Trinidad etc • UK, France, Holland,

Distribution and Names of Indigenous Sickle Gene

Haplotypes

Region	Haplotype
<ul style="list-style-type: none"> • Western West Africa • *Cameroon (Ekona) • Central West Africa • Central & East Africa • East Saudi Arabia & India 	<ul style="list-style-type: none"> • Senegal – 3 • Cameroon – 17 • Benin – 19 • Bantu – 20 • Arab/India – 31

Note: * Most Cameroonians have the Benin haplotype. The Ekona haplotype is a recent discovery among the small population of the Ekona ethnic group

Table 2. Where SCD is Found

The symptoms of SCD are seen predominantly in one-third of all aboriginal inhabitants (or their descendants) of parts of tropical and sub-tropical regions where malaria is or was common and in people from parts of the Middle East, Central India, Spanish-speaking regions (South America, Cuba, Central America); Saudi Arabia; and countries bordering the Mediterranean Sea, especially Turkey, Greece, and Italy (Akinyanju, 2009). This is because in

areas where malaria is common, there is a survival value in carrying only a single sickle-cell gene (sickle cell trait) (Akinyanju, 2009).

In the US, SCD affects around 72,000 people, most of whose ancestors come from Africa (Benjamin & Payne, 2007). It occurs in about 1:500 African-American births and 1:1000-1400 Hispanic-American births. About 2 million Americans, or 1:12 African Americans carry the sickle cell trait. Its occurrence among the Hispanic-Americans is about 5%, their median survival based on 1991 national data was 42 years for males, 48 years for females (California Institute For Regenerative Medicine, 2009). By twenty years of age, about 15% of children with SCD suffer major strokes and by 40 years of age, almost half of the patients have had central nervous system damage leading to significant cognitive dysfunction. These patients suffer significant damage to lungs and kidneys as well as severe chronic pain that impacts on quality of life. In Brazil, SCD is considered the most common monogenic disease seen predominantly in the black population as well as among individuals from parts of the Middle East, Central India and countries bordering the Mediterranean Sea, especially Italy and Greece.

2. What is Sickle Cell Disease (SCD)?

Sickle cell disorders (SCD) is a group of inherited autosomal recessive disorder characterized by production of abnormal of haemoglobin (Hb), resulting in anemia, susceptibility to pneumococcal and other infections, pain, stroke, and multiple organ dysfunctions. Normal red blood cells are soft, smooth, round and flexible and last about 120 days. It flows easily through blood vessels, but the abnormal hemoglobin which causes the red blood cells to be hard and sticky looks like a C-shaped farm tool called a sickle under the microscope (see figure 1).

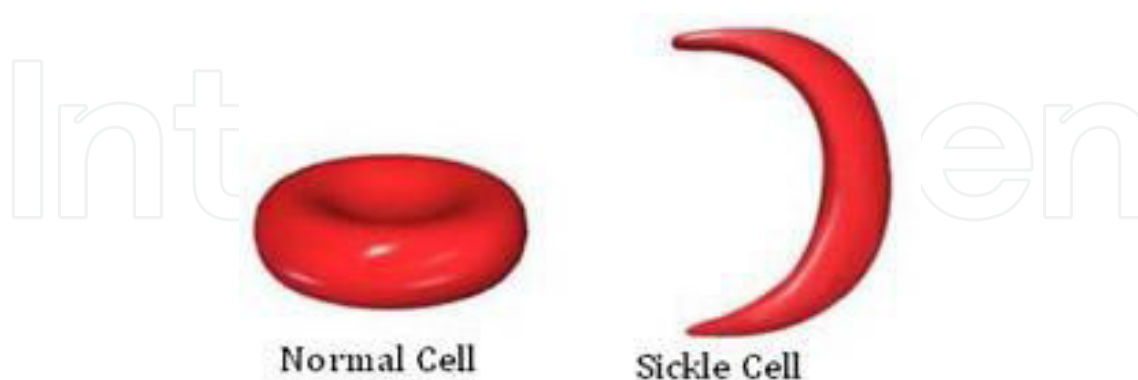


Figure 1. Shapes of Red Blood Cells

Sickle cell disorders, there are two main types of haemoglobin S- fetal Hb (F) in the unborn child (fetus) and adult Hb (A) after birth. It encompasses > 960 variants of sickling syndromes caused by abnormal sickle hemoglobin. Some are harmless; few are incompatible

with life and some like Hb S can make life more challenging. The most common and most severe variant of SCD is hemoglobin SS (homozygous) disease. Other forms include Sickle cell/C disorder (Hb SC), Sickle cell/ β thalassaemia (Hb S β^+ thal or Hb S β^0 thal), SD- Punjab, SO Arab, S Lepore and SE disease (NIH, 2010a, b).

Sickle cell trait (also known as being a carrier) occurs when a person has one gene for sickle hemoglobin and one gene for normal hemoglobin. Approximately one in ten African-Americans carries sickle cell trait. People who are carriers generally do not have any medical problems and lead normal lives. If you are a carrier you cannot develop sickle cell disease.

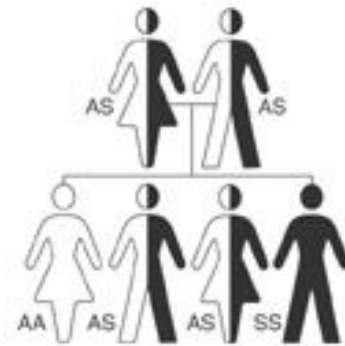


Figure 2. Types of Genotype Status

3. Symptom of Sickle Cell Disease: Related to painful and painless complication

Symptoms of sickle cell disease vary, ranging from mild to severe or life-threatening crises. It may occur without warning, and may go away and then come back many times. The signs and symptoms are linked to anaemia, pain and disease's complications. SCD anaemia related symptoms (lack of RBC) range from mild to very severe symptoms. The symptoms of anemia are fatigue (feeling tired or weak), shortness of breath, dizziness, headaches, coldness in the hands and feet, and paler than normal skin or mucous membranes (the tissue that lines your nose, mouth, and other organs and body cavities).

The pain related symptoms are debilitating pain episode or crisis which can affect bones, lungs, abdomen, and joints; as well as damage organs and increase the risk of stroke. The pain can be acute or chronic, but acute pain is more common. Acute pain is sudden and can range from mild to very severe. The pain usually lasts from hours to as long as a week or more. Chronic pain often lasts for weeks or months. Such pains can be limiting, unbearable and mentally draining. The painful crises are the leading cause of emergency room visits and hospital stays for people who have sickle cell anaemia.

Disease complication related of SCD crises are painful episodes (crises), acute chest syndrome, anemia (low hemoglobin), organ damage due to iron overload, infections, lung

problems, leg ulcers, bone damage, strokes, and premature death. It can cause hand-foot syndrome which is a blockage of the small blood vessels in the hands and feet in children (usually those younger than 4 years of age) leading to pain, swelling, and fever. SCD can also initiate splenic crisis in the abdomen, and pulmonary hypertension.

4. Etiology of SCD crises

The exact cause of episodic painful crisis is unknown. However, more than one factor is involved. First, the crises can occur whenever sickled red blood cells form clumps or abnormal curved shapes called sickles in the bloodstream. These clumps of cells stick to small blood vessels and block blood flow and oxygen to the limbs and organs. This can result in pain and damage to body organs, such as kidneys. It can trigger a stroke and other medical problems. For instance, the Hand-Foot Syndrome usually occurs whenever sickled RBCs block the small blood vessels in the hands and feet in children (usually those younger than 4 years of age). It can lead to pain, swelling, and fever. Swelling often occurs on the back of the hands and feet and moves into the fingers and toes. One or both hands and/or feet may be affected at the same time.

Gallstones usually develop in the gallbladder whenever there is too much bilirubin in the body. Gallstones may cause steady pain that lasts for 30 minutes or more in the upper right side of the belly, under the right shoulder, or between the shoulder blades. The pain may happen after eating fatty meals. People who have gallstones may have nausea (feeling sick to the stomach), vomiting, fever, sweating, chills, clay-coloured stools, or jaundice (a yellowish colour of the skin or whiteness of the eyes).

Ulcers on the Legs (sores) usually begin as small, raised, crusted sores on the lower third of the leg. Leg sores may occur more often in males than in females. These sores usually develop in people who are aged 10 years or older. The cause of sickle cell ulcers isn't clear. The number of ulcers can vary from one to many. Some heal quickly, but others persist for years or come back after healing.

Splenic Crisis can also occur whenever the spleen traps red blood cells that should be in the bloodstream. This causes the spleen to grow large and leads to anaemia. Acute Chest Syndrome may be caused by infection or sickle cells trapped in the lungs. People who have this condition often have chest pain, shortness of breath, and fever. They also often have low oxygen levels and abnormal chest x-ray results. Pulmonary Hypertension occurs as a result of damage to the small blood vessels in the lungs which make it hard for the heart to pump blood through the lungs. This causes blood pressure in the lungs to rise. Increased blood pressure in the lungs is called pulmonary hypertension (PH). Shortness of breath and fatigue are the main symptoms of PH. Priapism which is painful, unwanted erections may occur whenever sickle cells block blood flow out of an erect penis. Over time, priapism can damage the penis and lead to impotence.

SCD crises may also be caused by factors such as dehydration, infections, hypoxia, cold temperature, surgery and emotional stress. Dehydration often increases the risk of a sickle cell

crisis. Drinking plenty of fluids can lower the risk of a painful crisis. Other factors include bacterial infections. Infants and young children with sickle cell disease are especially vulnerable to serious infections, such as those that cause meningitis (infection of the lining of the brain) and blood infection.

5. Treatment goals

The goals of treatment options in SCD are symptom control; prevention of infections and stroke; detection and management of disease complications such as vaso-occlusive crisis, chronic pain syndromes, chronic hemolytic anemia, pulmonary hypertension, and the various organ damage syndromes. Accepted treatment options include narcotic pain killers, drugs, chronic blood transfusions, hydroxyurea, and stem cell transplantation (SCT) for selected children and young adults. Narcotic pain killers are used to treat the severe pain. Drugs are used to stimulate production of additional blood cells. Transfusions are used to treat the anemia and to dilute the sickle cells with normal red blood cells.

6. Stem cell transplantation treatment for SCD

Stem cells are parent cells found in all tissues and organs of the body, such as the bone marrow, skin, muscles, brain, peripheral blood, umbilical cord blood, and, rarely, fetal liver. The early cells nurtured in the bone marrow or less frequently from umbilical cord blood that mature into red and white blood cells and platelets are called multi-potent stem cells or immature cells. Stem cells produce erythroid cells, granulocytes, lymphoid cells, megacaryocytes and monocytes by a number of differentiation steps. Stem cells maintain normal cell populations in a healthy bone marrow controlled by haemopoietic growth factors, and stem cells have the capacity for self-renewal. Haemopoietic growth factors include erythropoietin, interleukins, glucocorticoids, sex hormones and thyroid hormones.



Figure 3. Stem Cell

Stem cell transplantation (SCT) refers to transplantation of the hematopoietic stem cells (HSCs) from a donor into an individual. Transplanted human bone marrow or stem cells are

dynamic biological entities that interact intimately with—and are influenced by—the physiology of the recipient. It is a very risky procedure. Before they are transplanted, cultured human stem cells are maintained under conditions that promote either the self-renewing expansion of undifferentiated progenitors or the acquisition of differentiated properties indicative of the phenotype the cells will assume (The National Institutes of Health resource for stem cell research, 2010). After incompletely differentiated human stem cells are transplanted, additional fine-tuning occurs as a consequence of instructions received from the cells' physiologic microenvironments within the recipient.

The goal of SCT is elimination of the sickle erythrocyte and its cellular progenitors and replacement with donor hematopoietic pluripotent stem cells which give rise to erythrocytes that express no sickle hemoglobin (HbS). This will eventually reduce Hb S levels to those associated with the trait condition. It has the possibility of preventing serious complications from SCD which can cause extensive morbidity and early death.

The donor sources of hematopoietic stem cells transplantation (HSCT) include cells obtained from another person (sibling or unrelated donor), termed allogeneic transplant; an identical twin, termed syngeneic transplant; or the patient, termed autologous transplant (Samavedi, 2011). The autologous HSCT (using the individual's own stem cells) involves using peripheral blood stem cell transplantation (PBSCT) to treat disorders such as multiple myeloma, non-hodgkin lymphoma, hodgkin disease, acute myeloid leukemia, neuroblastoma, germ cell tumors, autoimmune disorders (systemic lupus erythematosus [SLE], systemic sclerosis) and amyloidosis (Samavedi, 2011). It has not been used in the treatment of SCD.

Myeloablative allogeneic hematopoietic stem-cell transplantation (HSCT) is the only potentially curative treatment option for selected individuals with sickle cell anemia or thalassemia major (Samavedi, 2011; Krishnamurti, 2008; Walters, 2004). According to Samavedi (2011) and Doubek, Folber, Koristek, et al. (2009), it can also be used in the treatment of conditions such as, leukemia, myeloproliferative disorders and myelodysplastic syndromes. Successful allogeneic SCT not only eliminates the sickle-cell-induced vaso-occlusive symptomatology, but also leads to reversal of some of the end organ damage that occurred prior to the procedure.

In allogeneic HSCT, it is preferable for donors to have a human leukocyte antigen (HLA) type that is identical to the recipient. Matching is performed on the basis of variability at three of more loci of the HLA gene (e.g., HLA-A, HLA-B, HLA-DRB1). Usage of a non-myeloablative conditioning regimen prior to allogeneic SCT for transplantation of pediatric patients with SCD have been largely unsuccessful due to high rates of graft rejection (Bernaudin, Vannier, et al., 1997). Thus, current opinion is that children with high-risk SCD and a suitably matched donor should be offered allogeneic SCT using a conventional myeloablative conditioning regimen. To date, nearly all transplants have utilized HLA-identical sibling donors, which have limited the number of eligible sickle cell patients.

With HLA variability and lack of appropriate donors, there are increases in transplant-related morbidity and mortality, including graft rejection and graft-versus-host disease (GVHD). All donor-to-patient stem cell transplants use material which contains donor T-cells. These

donor T-cells react to the patient's body as foreign and causes GVHD which is a significant cause of illness and even death in stem cell transplants. It is generally avoided by using a donor as closely matched to the patient as possible. Usage of closely matched donors reduce will the risk of GVHD. In the normal population, a patient has about a 30% chance of having a matched sibling donor. However, SCD is a genetic disease, passed on from parents to children. A brother or sister who is a close match to the patient is very likely to also have SCD, making them inappropriate as a donor. The chance that an SCD patient has a matched sibling donor is less than 15%.

The optimal timing for marrow transplantation in the course of SCD remains uncertain because of the unpredictable nature and clinical heterogeneity of the disease (Walters, 2005). Selection criteria for optimal candidates continue to evolve; however, children and young adults, generally before the age of 21 years are considered the most appropriate candidates. Indications for HSCT have been empirically determined from prognostic factors derived from studies of the natural history of SCD. The most common indications for which patients with SCD have undergone HSCT are a history of stroke, recurrent acute chest syndrome, or frequent vaso-occlusive episodes (Novelli, Kato, Ragni, Zhang, Hildesheim, Nouraie, Barge, Meyer, Hassett, Gordeuk, Gladwin & Isenberg, 2012). Children and young adults who have severe complications (e.g. stroke, recurrent acute coronary syndrome [ACS], refractory pain) and have a human-leukocyte antigen (HLA)-matched donor are the best candidates for transplantation (Panepinto, Walters, Carreras, Marsh, Bredeson, Gayle, et al 2007). Very few adults are considered for transplantation due to existing comorbidities and toxicity of treatment (Walters, 2005).

7. Indications for stem cell transplantation

Sickle cell disorder (SCD) has no widely available cure. Its current medical therapies have only been relieving pain symptoms, preventing infections and managing complications such as eye damage, and strokes; and control complications. The progressive deterioration in organ function has been resulting in increased mortality and decreased quality of life. Some severe cases are resistant to existent therapies and can cut life even shorter.

Presently, Blood and Marrow Stem Cell Transplant appear to be the only viable option for eliminating SCD, especially in high risk patients. Patients with SCD are characterized as high-risk if they have central nervous system pathology (clinical or subclinical stroke, seizures), recurrent severe acute chest syndrome, chronic unremitting pain, or early evidence of end organ damage such as pulmonary hypertension. The appropriateness of SCT can be more firmly established in the presence of these high-risk features.

The bone marrow nurtures stem cells, which are early cells that mature into red and white blood cells and platelets. By destroying the sickle cell patient's diseased bone marrow and stem cells and transplanting healthy bone marrow from a genetically-matched donor, normal hemoglobin may be produced. Clinical studies using a few carefully selected patients

have reported very successful results (Harvey Simon, 2009). Unfortunately, only about 7% of patients with sickle cell meet the criteria for transplantation, including those who:

- Are age 16 or younger (generally considered the better candidates, but patients in their 20s have had successful transplants)
- Have severe symptoms but no long-term organ or neurologic damage
- Have a genetically matched brother or sister who will donate their marrow

The clinical indicator for stem cell application for SCD is based on stem cells' biological properties of self-renewal and their capability to give rise to differentiated cell progenies that maintain tissue homeostasis in physiological and pathological conditions (Lindvall and Kokaia, 2010; Orlicchio et al., 2010; Sendtner, 2009; Yu and Silva, 2008). Thus, neural stem cells in the subventricular zone of the lateral ventricles and the subgranular zone of the dentate gyrus in the hippocampus of the mammalian brain maintain the capability to generate new neural cells throughout the lifetime (Conti and Cattaneo, 2010; Ma et al., 2009; Galli et al., 2008).

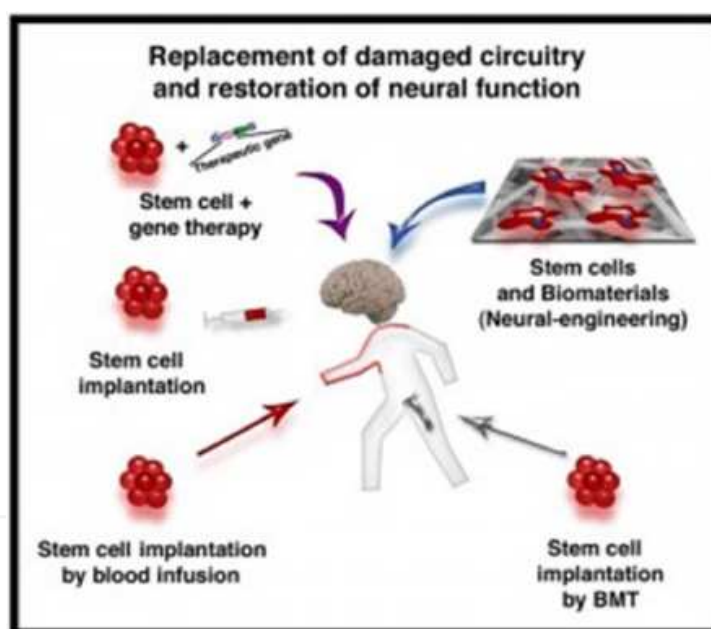


Figure 4. Stem cells replacement therapy for neurological diseases. Cartoon schematizes the different strategies for stem cell delivery in order to repair the degenerated tissue.

8. Risks and benefits of stem cell transplantation for sickle cell disease management in Nigeria

There are two major barriers to stem cell transplants to treat SCD. First is the risk of serious illness associated with donor-to-patient stem cell transplant; and 2) the lack of appropriate

donors. For these reasons, only about 300 of these transplants have been performed to date. All donor-to-patient stem cell transplants use material which contains donor T-cells. These donor T-cells react to the patient's body as foreign, causing graft-versus-host disease (GVHD). GVHD is a significant cause of illness and even death in stem cell transplants, and is generally avoided by using a donor as closely matched to the patient as possible, and by appropriate care after the transplant to quickly address symptoms of GVHD when they arise.

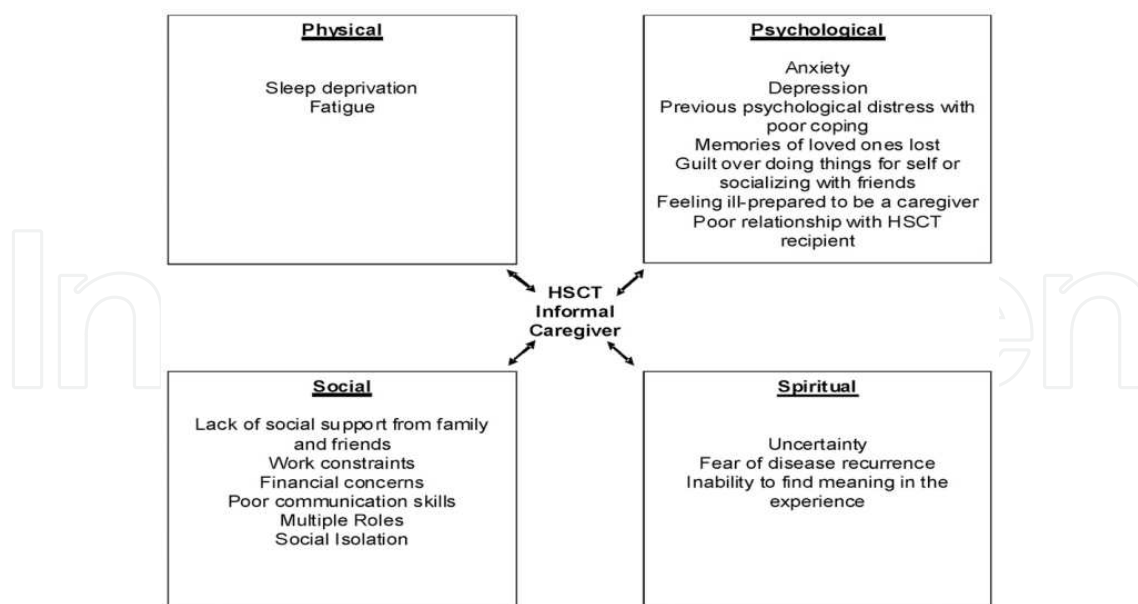
Another problem with stem cell transplant for SCD is a lack of donors. Usually, a patient's brother or sister who is a close genetic match to the patient is the preferred donor. Using closely matched donors reduces the risk of GVHD: the closer the donor cells are to the patient's cells, the less severe the immune reaction is likely to be.

9. Emerging therapies and promising research in Nigeria

In Sub-Saharan Africa, human stem cells offer new opportunities and promise of a remarkable array of novel therapeutics for the management of Sickle Cell Disorders (SCD) (The National Institutes of Health, 2009). In Nigeria, the conventional pharmacotherapeutic treatment options for SCD have been extremely limited in part to the management of its painless and painful crises. That is the current medical therapies for SCD only have significant short-term effects on affected persons in Nigeria. Nonetheless, recently a couple of medical Scientists at the University of Benin Teaching Hospital (UBTH), Benin-City, led by Nosakhare Bazuaye recorded a major scientific breakthrough in having a successful stem cell transplant in October 2011 on a 7 year old patient with sickle cell anemia (who had suffered stroke) after an appropriate allogeneic 14-year sibling donor was identified. This feat was the first of its kind in Nigeria and third in Africa. It came on the heels of earlier ones carried out in Egypt and South Africa (Sun Editorial, 2011).

10. The silence of literature on stem cell therapy for SCD in Africa

Numerous reasons could be advance for the silence of literature in Africa on stem cell transplantation. Firstly, Africa is a continent consisting of many developing nations. In most of these nations, particularly the tropic region, falciparum mosquito is highly endemic. The focus of such nations like Nigeria and Kenya in research and governmental policies has been on malaria. Although SCDs are highly prevalent in these nations, the attention of the government and policy makers have not been fully gained for the management of SCDs. Even among the medical practitioners, sheer magnitude of SCDs induces apathy and or feeling of helplessness. The availability of limited resources for the management of sickle cell crises and complications in these nations could also be attributable to the silence of literature in Africa on stem cell therapy for the management of SCDs. Currently, in Nigeria, attention is being drawn towards genetic counselling for affected individuals and their relations as well as to those intending to marry each other.



Scheme 1.

The political instability in most African nations has made it difficult for policy makers to present bills on the effective management of sickle cell disorders in Africa. The economic and emotional burden of this disorder is huge on the affected individuals and families in Africa. Many of the affected families are poor. Poverty is another reason for the relative silence of stem cell transplantation in Africa. This is coupled with the inequitable distribution of resources such as money, education, information and health care services in African nations. There is also the issue of lack of Respect and Support for research - molecular, clinical and operational- in Africa. The governments and corporate bodies in Africa do not fund research. There is usually low political will (conflict too) and no funding of research related to SCD management in Africa.

11. Implications for psychotherapy and genetic counselling

Although stem cell transplant has curative potentials for sickle cell anaemia or thalassemia, the physical side effects and psychological distress related to this treatment could be severe and even life threatening for the patients, the donor, and family members. First, for the patient, the transplant procedure is very risky and may be psychologically devastating and traumatizing. It can lead to serious physical and psychological side effects or even death. Approximately 5 percent of patients do not survive and it is used only in very severely affected children and young adults for whom there is a donor who is an appropriate genetic match. For instance, Greenfield (2007) in his eloquent essay and personal reflection as both a psychologist and transplant patient described the reality that the “powerful experience” of transplant caused him to re-experience psychological vulnerabilities despite years of psychoanalysis and therapy to address his past issues.

Secondly, bone marrow transplant carries its own dangers and limitations, especially for patients who do not receive a bone marrow transplant from a well-matched brother or sister donor. About 10% of those who have bone marrow transplants die from the treatment. In patients who do not receive a bone marrow donation from a matched sibling, the transplanted cells from a donor (called allogeneic grafts) may attack the patient's own tissues, a potentially fatal condition called graft-versus-host disease (GVHD). Drugs that destroy bone marrow and suppress immunity must be administered before the procedure so that the body's immune system does not attack the transplanted tissue. Still, this does not always prevent the problem.

Other very serious complications include bleeding, pneumonia, and severe infection. Those who live but are not cured face long-term problems caused by the drugs used in transplantation and by the disease itself. Even in those who are cured, long-term consequences may include a higher risk for cancer and infertility.

Psychologically, all the physical complications that patients face after transplantation may have significant impact on their daily and cognitive functioning. The patients may experience significant global psychological distress encompassing areas of existential concerns, obsessive-compulsiveness, loneliness, and ongoing health concerns such as memory loss (Rusiewicz, et al, 2008). They may also experience post-traumatic depression as a result of chronic graft versus host disease (cGVHD), long-term issues of ongoing medical appointments, and side-effects of medications (Sherman, Cooke & Grant, 2005; Syrjala, Langer, Abrams, et al., 2004). They may also experience challenging cognitive changes and post-transplant sexual difficulties such as vaginal dryness for women and erectile dysfunction in men (Sherman, Cooke & Grant, 2005). The patient may be unprepared for post-transplant life. In line with the time trajectory of HSCT as indicated in the table below, the physical and distressing psychological effects of stem cell transplantations have serious implications for genetic counseling and psychotherapy.

Diagnosis	HSCT	Short-term follow-up	Long-term follow-up
Decision to transplant	In-hospital treatment	Frequent controls	Quality of life
Donor search	Side effects, Toxicity Engraftment	Tx-related mortality GvH disease	Return to "normal life" Relapse

Table 3. Time Trajectory of HSCT

For the psychotherapists, genetic counsellors, psychologist, (particularly clinical psychologists) and informal help-givers including the spiritual help-givers., the pathological basis of SCD and stem cell transplantation have generated such questions as: Are the psychotherapists and genetic counselors in Nigeria sufficiently equipped to adequately meet the psychological needs of the individuals living with SCD in Nigeria after stem cell transplantation? Are the professionals (such as psychologists and counsellors) seeing the need to explore the possibility of blending pharmacotherapy with culturally accepted psychotherapeutic interventions for pain coping and increase of steady state among individuals living with SCD in

this part of the world? As a result, professionals should focus on the development of cross-culturally relevant psychotherapeutic measures that will address specific needs of patients, donors and their families prior to and after stem cell transplantation. Such cross-culturally sensitive psychotherapeutic programmes will take into consideration the psychological, cultural and spiritual aspects of individuals living with SCD in order to provide them with holistic care. In other words an eclectic but harmonious combination of behavioural techniques (therapeutic interventions) and cross-cultural therapeutic techniques could be more potent in achieving desirable therapy- outcomes.

There is also the urgent need for research that would assess the level of genetic counselling and psychotherapy being offered individuals living with SCD during their crisis state and steady state whether they are appropriate and whether they are being implemented properly. Appropriate psychological interventions can profoundly alter sets of beliefs, ways of thinking, affective states and patterns behaviour.

Added to this is the urgent need to train enough genetic counselors and psychotherapists with special focus on SCD and stem cell transplantation across sub-Saharan Africa where the disorder is prevalent, and most especially Nigeria which has the largest burden of SCD globally.

The hall mark of psychotherapy and genetic counselling in Nigeria for SCD and SCT shall be meeting the psychosocial health needs of the patients, the donor and family members. Because of the unique nature of the transplant experience, psychosocial assessment and interventions should be a high priority. The transplant procedure itself is complex and although the mortality has improved over the years since transplants began in the 1970's it continues to be a significant stressor. The recovery after transplant can come with prolonged physical and psychological set-backs, and extreme social strain on the patient's caregiver, friends and family members. In addition, the transplant experience can include multiple hospital readmissions for acute complications, slow recovery and long-term issues (Eldredge, Nail, Maziarz, Hansen, Ewing & Archbold, 2006).

12. Conclusion

Sickle Cell Disorder (SCD) is an inherited disease of red blood cells characterized by pain episodes, anemia (shortage of red blood cells), serious infections and damage to vital organs which vary greatly from one person to the next. This disorder has no widely available cure in Nigeria. Allogenic stem cell transplantation is the only treatment option with curative potentials, but is not readily affordable to most SCD affected families in sub-Saharan Africa.

Generally, hematopoietic cell transplantation (HCT) for sickle cell disease (SCD) has a strong track record of efficacy and there is growing appreciation that its benefits exceed its risks in selected individuals. The results of transplantation are best when performed in children with a sibling donor who is HLA-identical. Globally, Nigeria has the largest burden of this disorder. Government's commitment and strong political will are needed to support

and fund all activities and research geared towards effective management of SCDs in Nigeria so that stem cell transplantation can become a clinical curative and affordable reality for patients with sickle cell disorders and their families.

While there appears to be a considerable benefit to those who survive with stable engraftment of donor cells, there are also significant health risks to those who undergo this treatment. Therefore, engagement of trained psychotherapists and genetic counselors with focus on SCD and HCT with the patients, donors and their families should be conducted to ensure informed consent for this procedure. Presently, HCT is reserved for patients who have experienced significant complications of sickle cell disease, such as stroke, recurrent episodes of acute chest syndrome or intractable vaso-occlusive pain. Consequently, Nigeria is in dire need of strong institutional support for and training of psychotherapists and genetic counselors for the psychosocial management of patients with SCD, allogenic stem cell donors and their families.

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