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Review Article

## SICKLE CELL DISEASE AFRICAN KILLER: BIOLOGISTS ALTERNATIVE

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This seminar paper is done towards the elimination of sickle cell disease in our environment. Sickle cell anemia is an inherited disease of the red blood cells in which the red blood cell become sickle-shaped (crescent shaped) and has difficulty passing through small blood vessels. This process produces periodic episodes of pain and ultimately can damage tissues and vital organs and lead to other serious medical problems. Symptoms include fatigue, joint and abdominal pain, irritability, yellow discoloration of the skin and eyes, leg sores, gum disease, frequent respiratory infections, blindness later in life, and periods of prolonged, sometimes painful erections in males. People with sickle cell anemia can have episodes of severe pain in the arms, legs, chest, and abdomen that may be accompanied by fever, nausea, and difficulty breathing. These symptoms occur only in people who inherit copies of the sickle cell gene from both parents. Regular health maintenance is critical for people with sickle cell anemia. Today, proper nutrition, good hygiene, bed rest, protection against infection, regular visits to physician, many people with sickle cell anemia are in reasonably good health much of the time and living productive lives. Before starting a family, a simple blood test can reveal if one or both parents is a carrier. In conclusion, before starting a family, a simple blood test should be done because a child that receives the defective gene from both parents develops the disease. Therefore, marriage between carriers should be discouraged because sickle cell is better not produced than experienced.

**Keywords:** Sickle cell anemia, African killer, Red blood cells

### INTRODUCTION

Sickle cell disease is an inheritable blood disorder that affects red blood cells. People with sickle cell disease have red blood cells that contain mostly hemoglobin S (S for sickle), an abnormal type of hemoglobin. Sometimes these red blood

cells become sickle-shaped (crescent shaped) and have difficulty passing through small blood vessels (Gladwin *et al.*, 2008). When sickle – shaped cells block small vessels, less blood can reach that part of the body. Tissue that does not receive a normal blood flow eventually becomes

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damaged. This is what causes the complications of sickle cell disease. There is currently no universal cure for sickle cell disease except prevention which is better not produced than experiencing it (Strouse, 2008). Because it will affect every aspect of the child's life.

**Heamoglobin:** Is the main substance of the red blood cells. It helps red blood cells carry oxygen from the air in our lungs to all parts of the body (Brawley, 2008). Normal red blood cells contain heamoglobinA (HbA). HeamoglobinS (HbS) and heamoglobinC (HbC) are abnormal types of hemoglobin. Normal red blood cells are soft and round and can squeeze through tiny blood tubes. Normally, red blood cells live for about 120days before new ones replace them. (Geller, 2008). People with sickle cell conditions make use a different form of heamoglobin called heamoglobinS. Red blood cells containing mostly heamoglobinS do not live as long as normal red blood cells. (Normally about 16 days). (Geller, 2008). They also become stiff, distorted in shape and have difficulty passing through the body's small blood vessels.

## SICKLE CELL HISTORY

Although the HbS gene is most common in Africa, sickle cell disease went unreported in African medical literature until the 1870s. This may be because the symptoms were similar to those of other tropical diseases in Africa and because blood was not usually examined. In addition, children born with sickle cell disease usually died in infancy and were tropically not seen by physicians. Most of the earliest published reports of the disease involved black patients living in the US (Hanh *et al.*, 1927). African tribal populations were all too familiar with the disease and created their own names for it. It is interesting to note that

the tribal names all carrying repeating syllables, possibly to symbolize the repeating painful episodes such names include ahututuo (from the Twi tribe ); chwecheechwe (from the Ga); nuidudui (from the Ewe tribe);and nwiwii(from the fante tribe). Many tribal names were also imitations of the cries and moans of the sufferers or formed such phrases as "body chewing" or "body biting" which describe their terrible torment. In one West African tribe, (the Yorubas) children who died soon after birth were called "Abiku" meaning children who come and go. The tribal people believed that an evil spirit was trying to be born into a family with abiku children, but the babies bravely die to save the rest of the family from the demon. Some tribes had as many as 40% of the people carry the sickle cell gene (Allinson, 1954).

In the USA in 1846, a paper entitled "case of absence of the spleen" (from the southern journal of medical pharmacology), (Herrick, 1910), was probably the first to describe sickle cell disease. It discussed the case of a run away slave who had been executed. His body was autopsied and found to have the strange phenomenon of a man having lived without a spleen. Although the slave's condition was typical, the doctor had no way of knowing this as the disease had not yet been "discovered". The first formal report of sickle cell disease came out of Chicago about 50 years later, in 1910. In 1922, after three more cases were reported, the disease was named "sickle cell anemia" (Herrick, 1904). Herrick (1904), reported "peculiar elongated and sickle shaped" red blood cell in "an intelligent Negro of 20". These sickle cells were discovered by a hospital intern, Dr. Ernest Irons, who examined the patient's blood and sketched the strange cells. The patients had come to Dr. Herrick with abdominal pain, and

aches and pains in his muscles. He also feels tired all the time, had headaches, experienced attacks of dizziness, and had ulcers on his legs. After noting these symptoms, the doctor took samples of his blood. This first sickle cell patient had come to Chicago in 1904 to study dentistry in one of the best school of the country and was likely the only black student there. He was a wealthy man from West Indies; and, despite repeated hospitalizations for his illness, Walter Clement Noel completed his training, along with his classmates, three years later. He returned to Canada and practiced dentistry until he died of pneumonia at the age of 32. Although the disease does not distinguish between the rich the poor, it does single out those from tropical and subtropical climates of the old world (Herrick, 1910).

One long-held theory as to why it was so common in the Tropics was its association with malaria. In the 1940s, E.A Beet, a British medical officer stationed in Northern Rhodesia (now Zimbabwe), observed that blood from malaria patient who had sickle cell trait had fewer malaria parasites than blood from patients without the trait. Following this observation, reported by a physician in Zaire Demmis (1950) that there were fewer cases of severe malaria among people with sickle cell trait than among those without it. Allison (1954) continued to build on this observation and hypothesized that sickle cell trait offered protection against malaria. He suggested that those with the trait did not succumb to malaria as often as those without it; but, when they did, their disease was less severe. It is now known that, when invaded by the malarial parasite normally stable red cells of someone with the sickle cell trait can sickle in a low oxygen environment (like the vein). The sickling process

destroys the invading organism and prevents it from spreading through the body. This apparent ability of a genetic condition to protect carrier is particularly important in infants. Thus, in regions repeatedly devastated by malaria, people who carry the sickle cell trait will have a greater chance for survival than other individuals.

In the following years, evidence began to collect in support of this theory as well as some against it. When studies were restricted to young people, the hypothesis held; the sickle cell trait did offer protection to children but not to adults since they were unable to develop antibodies to the malarial parasite. However, even though their immunity was partial, it did help them to survive but offer little additional advantage. Since the youngsters were not able to produce antibodies to the malarial parasite until their immune systems matured, it was the pre-immune malarial patients whose survival was protected by sickle cell trait. For them as well, although protection was only partial, they did survive longer. Since then, several studies of malarial epidemics have revealed a higher survival rate for sickle cell trait individuals than for those who lack the gene HbS. These study areas included geographical distribution, gene frequency, and transgenic mice (the transportation of genes from one species into another). As reported by Anthony (1954). An English neurologist, Lord Brain, once suggested that a double dose of the sickle cell gene could be fatal; a single gene might increase a person's resistance to a disease. As more research was done, it was discovered that he was right, especially when it came to malaria. However, only those with sickle cell trait, not the disease, are protected against malaria. Those with sickle cell disease would die from blood disorder, or die after coming in contact with malaria because of their

weakened immune systems. (Pauling *et al.*, 1984). But if someone with sickle cell trait contracts malaria, the person's body is somehow shielded from this potential fatal disease. Scientists have found that red blood cell of people with sickle cell trait breaks down quickly when the malaria parasite attacks them. Since the parasite must grow inside red blood cells, the disease does not have a chance to become firmly established. However, not every one sickle cell trait can be protected either. Apparent resistance to disease occurs only in children between the ages of two and four (Pauling *et al.*, 1984). Studies have shown that Africans Americans who have lived in malaria-free areas for as long as ten generations, have lower sickle cell gene than Africans, and the frequencies have dropped more than those of others, less harmful African genes. Similarly, sickle cell gene is less common among blacks in Curacao, malaria-free island in the Caribbean, than in Surinam, a neighboring country, where malaria is rampant, even though the ancestors of both populations came from the same region of Africa. As reported as by (Caboot *et al.*, 2008)

## ORIGIN OF SICKLE CELL

Initially the single mutation theory was postulated in which it was conveyed that a single mutation occurred in Neolithic times in the then fertile Arabian peninsula (Pagnier, *et. al.* 1984). Then, the changing climatic conditions and conversation of this area to a desert caused the migration of people that could have carried the gene to India, Eastern Saudi Arabia and down to Equatorial Africa. This hypothesis was supported by citing the distribution of certain agricultural practices and anthropological evidences. But it is now quite clear that the sickle cell mutation has occurred as

several independent events. By using a series of different restriction endonucleases, different chromosomes structures (haplotypes) are identified and HbS gene has been found to be linked to certain commonly occurring haplotypes that are generally different from those bearing the HbA gene. In Africa the HbS gene is associated with at least three occasions in the African continent and at least once in either the Arabian Peninsula or the central India and from the primary sites the migration to the other region has occurred (Pagnier *et al.*, 1984). This can explain the observation made by many investigators that there is wide spread chromosomal heterogeneity of B gene cluster haplotypes in United State as compared to the homozygous condition in Africa, Arab or Asia. The slaves with sickle cell trait who were exported from various parts of Africa to United States had the specific B gene haplotype found in their region but after arrival in US, Jamaica and Brazil, over the years there has been considerable admixture of African ethnic groups.

Available calculations suggest that this gene has developed between 3000 and 6000 generations, approximately 70000-15000 years ago. The Senegal hyplotype is represented most prominently in Senegal and in the most Western regions of Africa above Niger River. The Benin hyplotype designates those found in Nigeria, Benin and countries in the right of Benin. The Bantu or CAR hyplotype encompasses those hyplotypes discovered in the Central African Republic and countries in South Central Africa. Interestingly, there are small pockets of sickle genes of the African haplotype in the region along India's Western Coast. Sickle cell disease have exist in the descendant of African people who came to India during the mogul period, often as "praetorian guards" for the Indian princes.

## SICKLE CELL DISEASE AND MALARIA

There are several theories as to why people with sickle cell trait have milder cases of malaria. This has to do with their being a host to weaker and fewer parasites.

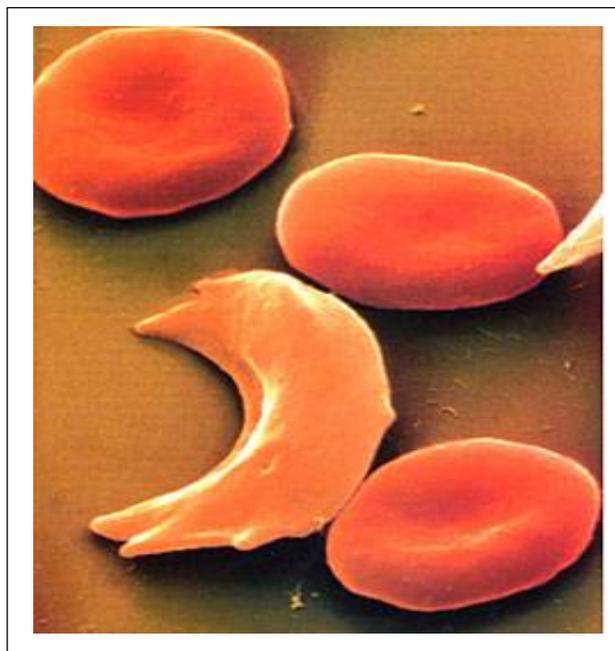
- The parasite inside the red cell produces acid. In the presence of acid, HbS has a tendency to polymerize which causes the cell to sickle. Since sickle cells are destroyed as the blood circulates through the spleen, the parasites are destroyed as well.
- Malarial parasites do not live longer under low oxygen conditions. Since the oxygen is low in the spleen, and since infected red cells tend to get trapped in the spleen, they may be killed there.
- Another thing that happens under low condition is that potassium leaks out of HbS containing cells. The parasites need high potassium level to develop. This may be the reason the parasite fails to thrive in red blood containing HbS. As reported by Carlson *et al.* (1994).

### WHAT IS SICKLE CELL TRAIT?

Sickle cell trait (AS) is an inherited condition in which both hemoglobin A and S are produced in the red blood cells, always more A than S. Sickle cell trait is not a type of sickle cell disease (Ohnishi *et al.*, 2000). People with sickle cell trait are generally healthy.

### GENETICS OF SICKLE CELL

The sickle cell disorder is known to occur when a person inherits two sickle cell genes (one from each parent) or a combination of one sickle cell gene from one parent and any one of several others abnormal hemoglobin gene from the



others (Philips *et al.*, 1992). The sickle cell disorder may be the world's most commonly inherited disorder and is clearly the most common genetic affliction seen in African infants. 1% to 2% of babies born on the African continent are afflicted with sickle cell disorder or one of its variants. The presence of two defective genes (SS) is needed for sickle cell anemia. If each parent carries one sickle hemoglobin gene (S) and one normal gene (A), each child has a 25% chance of inheriting two normal genes and not having the disease; and a 50% chance of being an unaffected carrier like the parents (Philips *et al.*, 1995). The inheritance of a sickle cell related disorder occurs at the moment of conception. As a human egg carrying a sickle trait is initially fertilized by a sperm carrying a similar disorder, the resulting new embryo is irreversibly imprinted with the genetic blueprint that will ultimately and inevitably appear as a sickle all related disorder in the newborn. They are inherited from parents in much the same way as blood type, hair color and texture, eye color and other physical traits.

The type of hemoglobin a person makes in the red blood cells depend upon what hemoglobin genes the person inherits from his or her parents. If one parent has sickle cell anemia and the other is normal, all the children will have sickle cell trait.

## WHY IS SICKLE CELL A DISEASE

The term disease is applied because the inherited abnormally causes a pathological condition that can lead to death and severe complication. Not all inherited variants of hemoglobin are detrimental. A concept known as "genetic polymorphism" (Mehta *et al.*, 2006).

## SICKLE CELL DISEASE PROCESS

- 1) When the sickle hemoglobin loses its oxygen, it forms rigid rods called polymers that change the red blood cells into a sickle or crescent shape (Mehta *et al.*, 2006).
- 2) The sickle –shaped cell stick to the walls and cannot squeeze through the capillaries. Blood flow through tiny blood vessels becomes slowed or stopped in many parts of the body. This deprives tissues and organs of oxygen.
- 3) When this blood flows or stops suddenly in a certain part of the body, the decrease in oxygen (hypoxia) can cause severe pain (the sickle cell crises). Over time, it leads to gradual destruction of organs and tissues throughout the body.
- 4) The higher the concentration of sickle hemoglobin and the more acidic the environment, the faster the sickle cell process.
- 5) When blood cells dry out (hydrate), the density of hemoglobin within the cell increases, thereby spreading the sickling process (Singh *et al.*, 2007).
- 6) Sickle cell has a shorter life span (10-20days) than that of normal red blood cells (90-120days). Everyday, the body produces new red blood cells to replace old ones, but sickle cells become destroyed so fast that the body cannot keep up. The red blood cell count drops which results in anemia. This gives sickle cell disease its more common name, sickle cell anemia (Halasa *et al.*, 2007).

## SYMPTOMS OF SICKLE CELL DISEASE

**General Symptoms in Infants:** In infants, symptoms do not usually appear until late in the baby's first year. Most commonly, they include; as reported by (Chiu *et al.*, 1990):

- 1) Fever.
- 2) Swelling of the hands and feet.
- 3) Pain in the chest, abdomen, limbs and joints
- 4) Nosebleeds and frequent upper respiratory infections.

**General Symptom in Children:** - Pain is the most common complaint. It can be acute and severe or chronic, usually from orthopedic problems in the leg and low back. Other symptoms as observed by (Dunlop *et al.*, 2006) include:

- 1) Fatigue and shortness of breath (signs of anemia).
- 2) Irritability.

- 3) Jaundice (yellowish discoloration of skin and eyes).

**Symptom in Adolescence and Adult:**  
Symptoms of childhood continue in adolescence. In addition, patients may experience:

- 1) Delayed puberty (in young teenagers).
- 2) Severe joint pain.
- 3) Progressive anemia.
- 4) Leg sores.
- 5) Gum disease.
- 6) Vision problems.

## **PAIN AND ACUTE SICKLE CELL CRISES**

The hallmark of sickle cell disease is the sickle cell crises (also sometimes known as vaso-occlusive crises) which is an episode of pain. It is the most common reason for hospitalization in sickle cell disease. The pattern may occur as follows:

- 1) In general, the risk of sickle cell crises is increased by any activity that boosts the body's requirement for oxygen, such as illness, physical stress, or being at high altitude. (Dunlop *et al.*, 2006).
- 2) Episodes typically begin at night and last 3-4 days, accelerating to a peak over several days and then declining.
- 3) The pain is typically described as sharp, intense and throbbing. Severe sickle cell pain has been described as being equivalent to cancer pain. Shortness of breath is common. (Geller *et al.*, 2008).
- 4) Pain most commonly occur lower back, leg, hip, abdomen or chest, usually in two or more

location. Episodes usually recur in the same areas.

- 5) The liver or spleen may also cause nausea, low-grade fever, and increasing jaundice.
- 6) Males of any age may experience prolonged, often painful erections, a condition called priapism.

## **COMPLICATIONS OF SICKLE CELL ANEMIA**

There is still no cure for sickle cell disease other than experimental transplantation procedures, but treatment for complications of sickle cell have prolonged the lives of many patients who are now living into adulthood.

### **ACUTE CHEST SYNDROME (ACS)**

Acute chest syndrome are occur when the lung tissue are deprived of oxygen during a crises. It can be very painful, dangerous and even life threatening. It is a leading cause of illness among sickle cell patients and is the most common condition at the time of death. At least, one whole segment of a lung is involved, and the following symptoms may be present;

- Fever of 101.3f degrees (38.5°C) or above.
- Rapid or labored breathing.
- Wheezing or cough.
- Acute chest pain.

### **CAUSES OF ACUTE CHEST PAIN**

- a) **Infection:** Infection from viruses or small typical organisms (Chlamydia and mycoplasma) is the most common cause of oxygen deprivation that leads to acute chest syndrome.

- b) Blockage Of Blood Vessels: Blockage of blood vessels (called infarction) that cuts off oxygen in the lung is another important cause of acute chest syndrome. Blockage may be produced by blood clots or fat embolisms. (Fat embolisms are particles formed from fatty tissue in the bone marrow that enter and travel through the blood vessels).
- c) Asthma: Asthma can increase the frequency and pain of acute chest syndrome episodes in children.

## INFECTIONS

In general, both children and adult with sickle cell anemia are more vulnerable to infections and have a harder time fighting them off. This is the result of spleen damage or sickle red cells, thus preventing the spleen from destroying bacteria in the blood. Infants and young children especially are susceptible to bacterial infections that can kill them in as little as 9 hours from onset of fever. Fortunately, with screening tests for sickle cell, now required for newborns, and with the use of preventive antibiotics and immunizations in babies who are born with the disease sickle cell infection might be reduced.

**Infections in infants and toddlers with sickle cell disease:** The most common organism causing infection of children include:

*Streptococcus pneumonia* and *Hemophilis influenza* (causing pneumonia, blood infection, or meningitis).

**Infection in children and adults:** Infection is also common among children and adults with sickle cell disease, particularly respiratory infections such as:

- a) Pneumonia

- b) Kidney infections
- c) Osteomyelitis, a serious infection in the bone (the organism causing them; however tend to differ from those in young children. (strouse, et. al 2007)

**a) Infection causing organism in children and adults:** *Clymadia* and *Myloplasma pneumonia*; These are the important infections in acute chest syndrome

**b) Gram-negative bacteria:** These group of bacteria mostly infects hospitalized patients and can cause serious pneumonias and other infection.

## STROKE

After acute chest syndrome, stroke is the most common killer of patients with sickle cell disease, who are older than 3 years old. Between 8-10% of patients suffer stroke typically at about age seven. Patients may also suffer small stroke that may not be immediately noticeable. Strokes are usually caused by blockage of blood vessels carrying oxygen to the brain. Patients with sickle cell disease are also at high risk for strokes caused by aneurysm a weakened blood vessel wall that can rupture and hemorrhage. Multiple aneurysms are common in sickle cell patients, but they are often located where they cannot be treated surgically. (Adams *et al.*, 2005).

## ANEMIA

Anemia is a significant characteristic in sickle cell disease, which is why the disease is commonly referred to as sickle cell anemia. As reported by (Platt *et al.*, 2008.)

**Severe worsening of anemia:** Children, adolescents, and possibly young adults may experience what is called Splenic Sequestration.

This happens when a large amount of the sickle red blood cells collect in the patient's spleen. Symptoms may include pain in the right abdomen below the ribs and a large mass (the swollen spleen) may be felt.

**Chronic anemia:** Because of the short life span of the sickle red blood cells, the body is often unable to replace red blood cells as quickly as they are destroyed. This causes a particular form of anemia called hemolytic anemia. Most patients with sickle cell disease have hemoglobin level of 8g/dl much lower than people without sickle cell anemia. Chronic anemia reduces oxygen and increases the demand on the heart to pump more oxygen bearing blood through the body.

## PREGNANCY AND SICKLE CELL DISEASE

Women with sickle cell disease who become pregnant have higher risk for complications such as miscarriages, premature birth and low birth weight. Sickle cell disease symptom often worsens during pregnancy and pain crises become more frequent. However, with careful prenatal care and monitoring, serious problems can be avoided. Maternal mortality rates have dropped significantly over the past decades. Most women with sickle cell disease can now anticipate favorable pregnancy (Adams *et al.*, 2005).

## DIAGNOSIS

Early diagnosis of sickle cell anemia is critical, so children who have the disease can receive proper treatment (Vishinski, 1981).

**Blood test:** More than 40 states now perform a simple inexpensive blood test for sickle cell disease on new born infants. The test is performed at the same time and from the same

blood samples as other routine newborn screening tests. Hemoglobin electrophoresis is the most widely used diagnostic test. If the test shows the presence of sickle hemoglobin, a second blood test is performed to confirm the diagnosis. These tests also tell whether or not the child carries the sickle cell trait.

## TREATMENT OF SICKLE CELL DISEASE

Treatment goals for sickle cell disease aim to relieve pain, prevent infections, and manage complications. Patients should seek help from doctors who specialize in blood disorder (hematologist), or a clinic that is experienced in treating sickle cell disease.

**1) Bone marrow transplantation:** This is the only potential cure, but it is used in only a small number of cases as few patients are able to find donors who are suitable genetic matches. Blood transfusion is given to prevent worsening anemia and prevent stroke (Adams *et al.*, 2005)

### 2) Drug treatments:

- 1) Antibiotics, usually penicillin, are commonly given to infants and young children, as well as adults to prevent infections.
- 2) Pain relief medications ranging from non-prescription nonsteroidal anti-inflammatory drugs (NSAIDs) are given to control pain.
- 3) Hydroxyurea is prescribed for patients with moderate to-severe disease to help reduce the frequency of pain episodes and acute chest syndrome.

## HYDROXYUREA

Hydroxyurea (Droxia) is a drug that reduces the severity of sickle cell disease by stimulating production of HbF. It is currently the only drug in

general used to prevent acute sickle cell crises (Brawley *et al.*, 2008). HbF, also called fetal hemoglobin is the form of hemoglobin present in the fetus and small infants. Most HbF disappear early in childhood, although some HbF may persist. Fetal hemoglobin is able to block the sickling action of red blood cells. Because of this, infants with sickle cell disease do not develop the symptom of the illness until HbF level has dropped. Adults who have sickle cell disease but still retain high level of hemoglobin F generally have mild disease. Hydroxyurea is not a cure-all. Not all patients respond to hydroxyurea. Many people who can benefit from it are not receiving it (Brawley *et al.*, 2008).

## SIDE EFFECTS

- Constipation
- Drowsiness
- Hair loss
- Inflammation of the mouth

It should not be taken by pregnant women because it can cause birth defects. The long term use of hydroxyurea may increase the risk of developing leukemia. (Brawley, *et al.* 2000).

**3) Transfusion:** Blood transfusions are often critical for treating sickle cell disease. Transfusions may be used either as treatment for specific episodes or as chronic transfusion therapy to prevent life threatening complications ongoing transfusions can also help improve height and weight in children with sickle cell disease (De-franceschi *et al.*, 2008)

**Episodic Transfusions:** They are needed in the following situations:

- To manage sudden severe events, including acute chest syndrome, stroke, widespread

infection (septicemia) and multi organ failure.

- To manage severe anemia, usually caused by splenic sequestration (dangerously enlarged spleen) or aplasia (halting of red blood cell production, most often caused by parvovirus). Transfusions are generally not required for mild or moderate anemia.
- Before major surgeries, transfusions are generally not required for minor surgeries.

**Chronic Transfusion:** (on-going) transfusions are used for

- Stroke prevention for first or recurrent strokes.
- Pulmonary hypertension and chronic lung disease.
- Heart failure.
- Chronic kidney failure and severe anemia.
- To reduce episodes of pain and acute chest syndrome.

## KIND OF TRANSFUSION

- 1) **Simple Transfusion:** Involve the infusion of one or two units of donor blood to restore blood volume levels and oxygen flow. It is used for moderately severe anemia, severe fatigue and also used for acute chest syndrome.
- 2) **Exchange Transfusion:** Involves drawing out the patient's blood while exchanging it for donor red blood cells. It can be done as manual procedure or as automatic one called
- 3) **Erythrocytapheresis:** It may be used when there is any evidence that the patient's condition is deteriorating.

## PRECAUTIONS

- 1) Have regular physical examinations every 3-6 months.

- 2) Have periodic and careful eye examinations.
- 3) Have sufficient rest, warmth, and increased fluid intake (these are critical precautions for reducing oxygen loss and the risk for dehydration).
- 4) Avoid conditions such as crowds that increase risk for infections.
- 5) Avoid excessive demands on the body that would increase oxygen needs (physical over exertion, stress). Low impact exercise (leg lifts, light weights) may be useful and safe for maintaining strength particularly in the legs and hips, but patients should consult physician. As reported by de-Montalembert (2008).

## BIOLOGISTS ALTERNATIVE

**1) Vaccination:** Everyone with sickle cell disease should have complete regular immunization against all common infection. Children should have all routine childhood vaccinations. The following are important vaccinations for everyone with sickle cell disease:

- Pneumococcal vaccines: All sickle cell patients should be vaccinated with pneumococcal vaccines. There are two types of pneumococcal vaccines; the choice between them depends on the age of the patients.
- Vaccination against *Haemophilus influenzae*: The major cause of childhood meningitis, starting at age 2 months.
- Influenza vaccines ("flu shots"): It should be given every winter starting at age 6 months.
- Meningococcal vaccination: for patients age 5 and older.

- Hepatitis B vaccines: All children should receive this vaccine.

**2) Antibiotics:** In addition to regular immunization, preventive antibiotics are the best approach for protection against pneumonia and other serious infections among children with sickle cell disease. Babies diagnosed with sickle cell are given daily antibiotics, starting at 2 months of age and continuing through 5 years of age. Penicillin is usually the antibiotic given, unless a child is allergic to it.

## NUTRITION AND DIETARY SUPPLEMENTS

**i) Food:** Good nutrition, while essential for everyone, is critical for patients with sickle cell disease. Some dietary recommendation include:

- Fluids: Fluids are number one in importance. The patient should drink as much water as possible each day to prevent dehydration. (Singh *et al.*, 2007).
- Diet: Diet should provide adequate calories, protein, fats, vitamin and minerals. Patients and families should discuss vitamin and mineral supplements with their doctors (Al-Momen, 1995).
- Omega-three Fatty Acid: Some studies claim that omega- three fatty acids, found in fish and soybean oil as well as dietary supplements, might make red blood membranes less fragile and possibly less likely to sickle, although no studies have proven this definitively (Mehta *et al.*, 2006).

**ii) Vitamins:** Patients should take daily folic acid and vitamin B12 and B6 supplements. Vitamin B6 may have specific anti-sickling properties. Foods containing one or all these vitamins include

meats, oily fish, poultry, whole grains, dried fortified cereals, soybeans, baked potatoes with skins, water melons, plantains, bananas, peanuts, and brewer's yeast (Muskiel *et al.*, 1991).

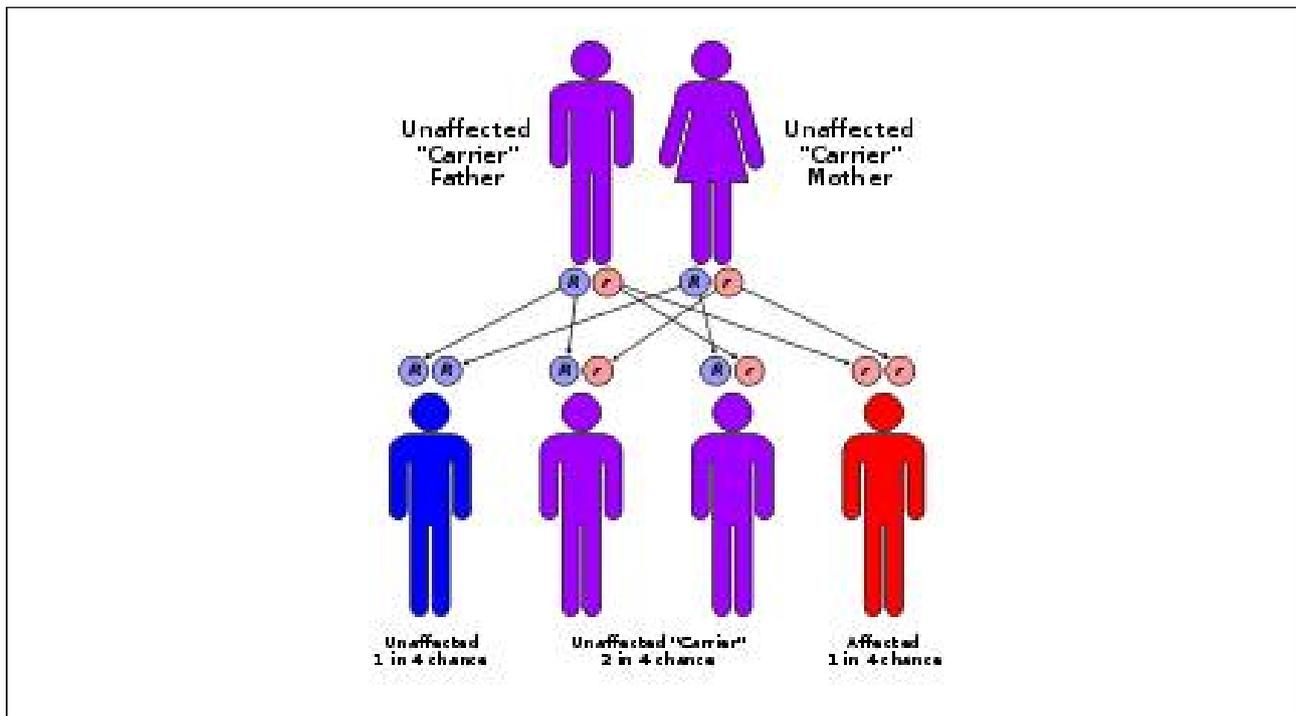
**4) Managing the Emotional and Social Impact:**

In assessing seriousness of the disease, no one should underestimate its emotional and social impact. For the family, nothing is more heartbreaking than watching their child endure extreme pain and life threatening medical conditions. The patient endure not only the pain itself but also emotional strain from unpredictable bouts of pain, fear of death, and lost time and social isolation at school and work. Academic grades among patients average less than C, even in children with a low frequency of hospitalization (averaging 17 days a year).

These problems continue over the years, and both children and adults with sickle cell disease often suffer from depression. The financial cost

of medical treatments combine with lost work can be very burdensome. Any chronic illness places stress on the patient and family, but sickle cell patients and caregiver often great obstacles in finding psychological support for the disease. It is very important for patients and their caregivers to find emotional and psychological support. No one should or can endure this life-long disease alone. Unfortunately, studies indicate that most patients do not receive even basic supportive care that could help reduce the anxiety and intensity of pain that occurs when sickle cell crises erupt.

**5) Marriage:** The child of two carrier parents may inherit a full blown sickle cell disorder. These disorders have no cure and will affect every aspect of the child's life. Therefore, using the Mendel's theory, marriage between two carriers should be avoided because there is 50% probability that every child produce might be a carrier or a patient. So, before starting a family, a simple blood test can reveal if one or both parent is a carrier or a sickle cell disorder patient.



## CONCLUSION

Regular health maintenance is critical for people with sickle cell anemia. Proper nutrition, good hygiene, bed rest, protection against infections, and avoidance of other stresses all are important in maintaining good health and preventing complications. Regular visits to physician or clinic that provides comprehensive care are necessary to identify early changes in the patient's health and ensure immediate treatment. Today with good health care, many people with sickle cell anemia are in reasonably good health much of the time and living productive lives. Infact, in the past 30 years, the life expectancy of people with sickle cell anemia has increased. But not withstanding, marriage between carrier and sickle cell patient should be discouraged because producing a child with this disorder will affect every aspect of the child's life, since sickle cell disease has no cure.

## RECOMMENDATION

Sickle cell disease is better not produced than experiencing it.

## REFERENCES

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