The World Sickle Cell Day

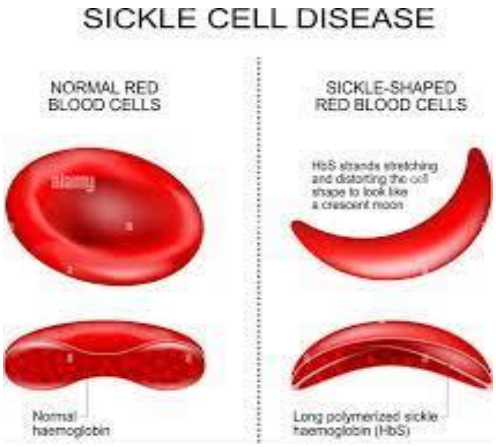
19th June 2023

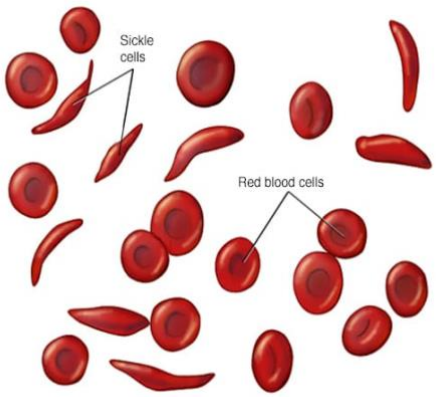
World Sickle Cell Day is observed globally on June 19th each year. It provides an opportunity to shine light on the impact of sickle cell disease (SCD) and to advocate for improved care and support for those affected.

The theme for the year 2023 is "**Building and strengthening Global Sickle Cell Communities, Formalizing New-born Screening and Knowing your Sickle Cell Disease Status**". This is a call to recognise the first step (understanding the genotype in infants and adults) in fighting sickle cell disease, thus breaking its cycle, as it is an inherited disorder being passed down through generations.

Knowing one's status helps to prevent passing down of this disorder to children by getting tested early, especially new-borns, to start early treatment of those having the genotype and fostering communities to providing a safe and affordable space for individuals living with Sickle cell disease (SCD) and their families.

SCD is a group of inherited red blood cell (RBC) disorders. Healthy RBCs are round and move through small blood vessels to carry oxygen throughout the body. But in a person with sickle cell anaemia, RBCs become hard and sticky, looking like a letter "C" or sickle in shape, blocking blood flow and oxygen from reaching all parts of the body. This leads to pain and other serious problems such as: infections, acute chest syndrome and stroke.





Normal red blood cells and sickle-shaped red blood cells

(Mayo Foundation for medical education and research)

SCD is a genetic condition that is present at birth when a child receives two abnormal genes - one from each parent. There are several types of SCD, depending on which gene is passed down from the parents. The specific type of SCD a person has depends on the genes (containing instructions, or code, for abnormal haemoglobin) they inherited from their parents. The following are the most common.

HbSS: People who have this form of SCD inherit two genes, coded for haemoglobin "S", one from each parent. Haemoglobin S causes the RBCs to become rigid and sickle-shaped. This is commonly called sickle cell anaemia and is usually the most severe form of the disease.

HbSC: People who have this form of SCD inherit a haemoglobin "S" gene from one parent and haemoglobin called "C" (a different type of abnormal RBC) from the other parent. This is usually a milder form of SCD.

HbS beta thalassemia: People who have this form of SCD inherit a haemoglobin "S" gene from one parent and a gene for beta thalassemia (another type of haemoglobin abnormality) from the other parent. There are two types of beta thalassemia: **"zero"** designated **HbS beta^{0}** which is severe form of SCD and **"plus"** designated **HbS beta^{+}** which is milder.

There are some other rare types of SCD, such as: **HbSD, HbSE,** and **HbSO** Characterised by inheriting one haemoglobin "S" gene and one gene that codes for another abnormal type of haemoglobin ("D", "E", or "O"), of varying severities.

HbAS: People who have sickle cell trait (**SCT**) inherit a haemoglobin "S" gene from one parent and a normal gene (one that codes for haemoglobin "A") from the other parent. It is rare for a person with SCT to develop health problems. This usually occurs secondary to other stresses on the body, like dehydration or strenuous exercises. SCT patients can pass the abnormal haemoglobin "S" gene on to their children. This is why they are widely known as **SCD carriers**.

Generally, SCD causes severe pain (also known as pain crises), anaemia, infection and other serious health problems that may require treatment by a healthcare provider.

Diagnosis

SCD is diagnosed with a simple blood test, most often done at birth during routine new-born screening tests at the hospital, though it can also be diagnosed while the baby is in the womb, with procedures like chorionic villus sampling and amniocentesis, which can check for chromosomal or genetic abnormalities in the babies. Chorionic villus sampling tests a tiny piece of the placenta, called chorionic villus, while amniocentesis tests a small sample of amniotic fluid surrounding the baby.

Getting Tested

Eradication of SCD by selective mating is very logical and simple. Getting tested prevents the birth of children with SCDs. If two partners are carriers of any of the sickle cell genes, the possibility of their children having SCD makes them incompatible if they intend to have children. Religious bodies and charitable organisations have in the past actively campaigned to implement this by screening young people and instructing those shown to be carriers to avoid choosing spouses who also have any sickle cell gene. Some churches even refused to marry affected partners. The reality, however, is that enforced selective mating of couples has never been shown anywhere in the world to have reduced the incidence of any inherited disorder. Attempts to introduce it by the Church in Cyprus, for the control of thalassemia there, led to increased anxiety and stigmatisation of affected persons and of healthy carriers of the gene (Angastiniotis MA and Hadjiminas MG, 1981; Angastinoiostis, *et al*, 1986). This, in turn, led to widespread denial and falsification of haemoglobin genotype results among carriers who wanted to marry each other. What the Church in Cyprus now does is to ensure that all couples have been counselled on thalassaemia before marriage.

Sickle Cell Disease in Nigeria

After some research in northern Nigeria, Molineaux and co-researchers correctly summarised the SCD situation when they wrote in 1979: **"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 will become**

an immense medical, social and economic problem throughout the continent." (Molineaux *et al,* 1979).

Out of about 50 million people living with SCD globally, Nigeria has about 4-6 million people living with it; Nigeria has the highest number of new-borns with the disease in the world. It is estimated that over 150,000 babies are born with SCD in Nigeria annually which is about 33% of the global burden of SCD, this results in high infant mortality.

Raising Awareness and Dispelling Myths

Through various awareness campaigns, community outreach programmes and media engagement, efforts are made to promote accurate information, debunk myths and address stigmas associated with SCD. By spreading awareness, the aim is to foster a more compassionate and supportive society for individuals living with SCD and continue to discourage carriers having children that would end up with the disease.

Improving Access to Care and Treatment

World Sickle Cell Day provides an opportunity to advocate for improved access to comprehensive care and treatment for individuals living with SCD. This includes affordable healthcare services, access to essential medications, specialised clinics, and multidisciplinary care teams. Additionally, it is necessary to prioritize research and innovation to develop new treatment options and enhance the quality of life for individuals with SCD.

World Sickle Cell Day highlights the importance of providing individuals and their families with psychosocial support, counselling and educational resources. Support groups and community organisations play a vital role in providing a sense of belonging, sharing experiences and empowering individuals to navigate the challenges associated with SCD.

The World Sickle Cell Day awareness campaign stands as a rallying point for Nigeria to join the global effort in raising awareness, supporting individuals and advocating for improved care for those affected by SCD. The joint efforts create a society that embraces inclusivity, provides early diagnosis, ensures access to quality healthcare and offers support to individuals and families living with SCD. We need to seize this opportunity to promote change, foster compassion and empower individuals with sickle cell disease to live fulfilling lives. On this World Sickle Cell Day, we must speak with one voice and united in actions to make a positive impact, improve the lives of those affected by SCD and strive for a future where no one suffers from this debilitating disorder.

REFERENCE

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