Sickle Cell Bulletin

A Publication of the Sickle Cell Foundation Nigeria. Vol. 10 No.2 2018

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Sickle Cell Priapism Control

Timi

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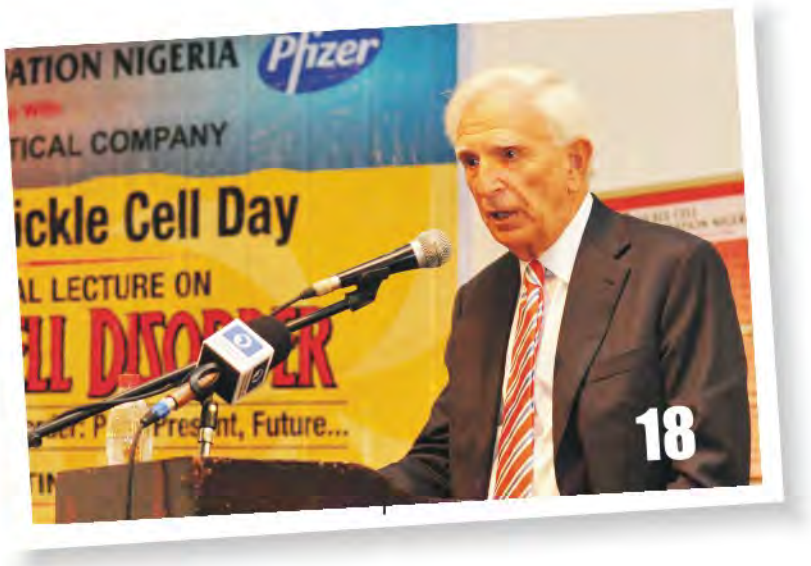
inthisissue

Editorial	4
About Sickle Cell Foundation Nigeria	5
Newsreel	6
Hello! Timi, Nice to Meet You!!	12
Sickle Cell Priapism Control	17
Photoreel	18
Acute Sequestration Crises in Sickle Cell Disorder	23
Prenatal Diagnosis and Preimplantation Genetic Diagnosis of Sickle Cell Disorder	24
Prevention of Infection in Children with Sickle Cell Disorder aged under 10 Years	27
Be Careful; Your Cough can be Worse than you Think	31
Sickle Cell Clubs and Affiliated Organisations in Nigeria	33



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Editorial

I am delighted to be a living witness to the increasing self-confidence of a substantial number of our men and women who are living with sickle cell anaemia. A key pointer to this situation is the fact that they have accepted portrayal on the front cover of several recent Sickle Cell Bulletins, with their opinions published therein.

> **Lately, some successful middle to older aged persons living with SCD are in the process of forming an association that will work towards improving related information...**

This contradicts the uninformed, but popular belief that people with sickle cell disorder are mostly unworthy employees, etc. On the contrary, apart from many who successfully run their own businesses, some have held or still hold excellent positions in the medical, legal, architectural and teaching professions. I also know of one, who is a High Court Judge and another, who was a State Government Commissioner. A colleague of mine also knows a past State Governor who has SCD.

Lately, some successful middle to older aged persons living with SCD are in the process of forming an association that will work towards improving related information, perceptions and research that will improve the quality and duration of their lives.

To this end, the decision of the Editorial Committee to greatly widen the availability and readership of the Sickle Cell Bulletin to interested persons in all states of Nigeria is most welcome and should hopefully provide helpful life information. Happy Reading!

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About Sickle Cell Foundation Nigeria

The Sickle Cell Foundation Nigeria (Limited by Guarantee RC258517) is a non-governmental and non-profit making organisation dedicated to the proper care and control of sickle cell disorder in Nigeria.

The Sickle Cell Foundation Nigeria (SCFN) was registered in November 1994 to address important issues such as capacity building, research, policy development, implementation, monitoring and evaluation, necessary for the sustained management and control of sickle cell disorder in Nigeria. The SCFN operates as a Private Public Partnership (PPP) with the Nigerian government through its affiliations with the Lagos University Teaching Hospital/College of Medicine and the Federal Ministry of Health. In addition the SCFN is associated with the Department of Public Information (DPI) of the United Nations.

Vision: To alleviate the burden of sickle cell disorder on the country and to ensure that all affected persons can live normal pain-free lives.

Mission: To develop and sustain a world class National Sickle Cell Centre and drive the search for effective solutions to the problems associated with sickle cell and related disorders in Nigeria and beyond.

Programmes and Services

Genetic Counselling: Expert genetic counselling is available every weekday for people living with SCD, their families and healthy carriers of the sickle cell or Hb C gene (Hb AS or Hb AC). To book an appointment for counselling, please call 0810 000 2001.

Sickle Cell Clinics: With donor support we supervise and aid the running of 5 dedicated Sickle Cell Clinics across the country in Lagos, Benin, Asaba and Kano. These clinics have recorded improvement in the quality of lives and reduction of death rates in their patients.

Prenatal Diagnosis of SCD: The diagnosis of Hb genotypes in unborn babies is carried out regularly, from about 11 weeks of pregnancy, on pre-booked pregnant women who are at-risk of bearing children with sickle cell anaemia (Hb SS). Training of obstetricians on the safe performance of the procedure (chorionic villus biopsy) is also offered. The fetal Hb genotype diagnostic process involves two or more molecular biological methods in our MTN DNA Laboratory. For enquiries, please call 0803 584 6666.

Reference Haematology Laboratory (RHL): On Mondays to Fridays, our well-equipped RHL provides accurate haematological, Hb genotype and Hb fractions including Hb A_1C (for diabetes control). Update training of laboratory personnel is also offered.

Exchange Blood Transfusion: An automated red cell exchange transfusion service is now available.

Library and Information Services: The Stanbic IBTC Library provides access to current print and digital publications on SCD. The service, which is highly interactive, is open weekdays to healthcare professionals, as well as the general public, who desire reliable and accurate information on SCD. For membership details, call 0803 584 6666.

Stroke Prevention Service: Stroke occurs in about 9% of young children with Hb SS. The detection of children at risk of developing stroke is achieved by Transcranial Doppler (TCD) ultrasound screening. TCD of children aged 2 to 16 years is advocated and done on weekdays. Children identified as being at high risk of stroke are then provided with parental counselling and known stroke prevention measures. For the TCD equipment, we are grateful to the Heineken Africa Foundation and to the AMNI International Petroleum Development Company.

Leg Ulceration: Leg ulceration is a worrisome problem in a minority (6-7%) of our patients with SCD. These ulcers are notoriously slow to heal and most of them would recur spontaneously after healing. Our Leg Ulcer Treatment team works hard to find ways of inducing rapid healing of the ulcers and of preventing their recurrence.

Newsreel

THE NIGERIAN NCD ALLIANCE

As a key member of the NCD Alliance Nigeria, SCFN participated in a workshop to develop the "Civil Society Strategic Plan of Action for Non-Communicable Diseases in Nigeria (2017 – 2021)". NCD Alliance is a partner to governments and UN agencies and an advocate for people at risk of NCDs or living with NCDs.

SCFN presented its Strategic Framework for Sickle Cell Disorder in Nigeria and the report has been adopted and will be part of the broader strategic plan of action for NCDs in Nigeria.

The workshop, which was held at Adna Hotel Ltd., GRA, Ikeja, Lagos from the 27 – 28, April 2018, was well attended and the discussions and presentations were robust, incisive and inclusive of different NCDs in Nigeria.

In addition, SCFN was part of two high profile meetings of the NCD Alliance in Abuja on 17th May, 2018. The meetings were (1) with the Senior Special Assistant (SSA) to the President on Sustainable Development Goals (SDGs) and (2) with the Honourable Minister of Health, Professor Isaac Adewole - at the Federal Ministry of Health. The objective of both meetings was to formally introduce the NCD Alliance to these important government agencies and apprise them of the work of the Alliance as well as proposals for collaboration going forward. The NCD Alliance team was led by its President, Olor'ogun (Dr.) Sunny Kuku, and Executive Secretary Prof. Sanni Malami.

2018 WORLD SICKLE CELL DAY (WSCD) COMMEMORATION ACTIVITIES

SCFN commemorated the 2018 World Sickle Cell Day in June with the following programmes and activities:

(i) **Press Briefing:** The press briefing was organized in collaboration with the Coalition of Sickle Cell NGOs in Nigeria to inform the general public about the Red Umbrella Charity Walk for Sickle Cell – an event planned to bring together all sickle cell NGOs and groups in Lagos State to raise awareness about SCD and advocate for more government and private sector involvement in sickle cell programming. The briefing was held at the Seminar Room of the National Sickle Cell Centre on June 7, 2018. Representatives of Fidelity Bank (Sponsors of the event), member NGOs and journalists from the print and electronic media were all present at the briefing."

The CSR Manager of Fidelity Bank, Mr. Chris Nnakwe, in his remarks commended the Coalition for the initiative which, he emphasized, would go a long way to create the needed awareness at the community level. He said the Bank was happy to partner with the Coalition and looked forward to making the Walk an annual event.

(ii) **Schools Quiz:** The Schools Quiz competition, which is now an annual WSCD event, was held on Thursday, 14th June, 2018. The quiz brought together Senior Secondary School Students from 21 schools in 4 of the 6 Education Districts in Lagos State. The aim

was to raise awareness about SCD among young people in school – a critical demography in the control of SCD.

This year's competition was very keenly contested and tie-breakers were required to determine the winners. At the end of the contest, the first position went to Aladura Comprehensive High School the second position to Top Laurel Grammar School, while Lagos Progressive Senior Secondary School came third.

The event was sponsored by Food Concepts Plc - who provided food and drinks for all the participants and Laroache Leadership Foundation, who provided the gifts and trophies for the top 3 schools.

Present at the event were representatives of the sponsors, Mrs. Otaigbe, Mrs. Adesemoye and Dr. Muideen Bakare. Dr. Bakare, a Consultant Psychiatrist at the Federal Medical Centre, Enugu lives with SCD and has written a book on his experience growing up with the condition to motivate affected persons and encourage them to fulfill their potentials. His book, ***"Aro'mo Le'egun: A Story of Sickle Cell Disease, Stigma, Poverty, Hope and Resilience in Sub Saharan Africa",*** is compelling, educative and motivational. The author interacted with the students during the question and answers session and then donated 200 copies of the book to SCFN for dissemination.

(iii) **The Red Umbrella Charity Walk:** SCFN, once again, joined other Sickle Cell NGOs for a walk to raise awareness about SCD on Saturday, 16th June, 2018."

The walk, which started at Teslim Balogun Stadium, Surulere, traversed Ojuelegba, Itire Road, Ogunlana Drive, Alhaji Masha Street and ended at the point of departure.

The Walk was flagged off by Hon. Desmond Elliot – representing Surulere Constituency in the Lagos State House of Assembly. It brought together people from all walks of life and the turnout was massive and included a delegation from the US Consulate in Lagos. The Lagos State Police Command supported the walk by providing officers for protection while the Lagos State Transport Management Agency (LASTMA) assisted with traffic control.

(iv) **The Annual World Sickle Cell Day Lecture:** This was our Flagship event for the commemoration of World Sickle Cell Day. The Annual Lecture was held on Wednesday 20th June, 2018 at the Agip Recital Hall of the Muson Centre, Lagos. It was delivered by the

distinguished, world-renowned Professor of Medicine, Paediatrics, Pathology & Laboratory Medicine, Dr. Martin Steinberg of Boston University, Massachusetts, USA. The main sponsor for the lecture was Pfizer Pharmaceuticals. The topic of the lecture was **"Sickle Cell Disease: Past, Present, Future"** and it was well attended by the academia, medical practitioners and the general public. Indeed the hall was filled to capacity. Among the distinguished guests were the First Lady of Abia State, H.E, Nkechi Ikpeazu – Founder of Vicar Hope Foundation, Representative of the

Hon. Commissioner for Health, Lagos State – Dr. Babafemi Thomas, Representative of Pfizer – Dr. Margaret Odunvbun.

In addition, there were goodwill messages from H.E. Ikpeazu, and Dr. Ijeoma Akinwumi representing the Nigeria Sickle Cell Support Group. A plaque to commemorate the lecture was presented to Dr. Steinberg by H.E. Ikpeazu alongside our Chairman, Professor Olu Akinyanju.

The event also featured an exhibition by some companies including Pharmaceutical Companies and Medical Laboratories namely, Clina Lancet Laboratories, Bond Chemicals, Pathcare, Medical Art Centre, the Bridge Clinic, etc.; the exhibitors had the opportunity to engage with the lecture participants and showcase their products and services.

Furthermore, some of the art works by our children with sickle cell disorder (from our Art in Medicine Programme) were also on display at the lecture venue and five were purchased at the event.

Tour for Martin Steinberg, MD: Ahead of the WSCD lecture, we were able to take Dr. Steinberg on a grand tour of the National Sickle Centre. He got first-hand experience and appreciation of our facilities and services. He interacted with staff and patients, made useful suggestions on how services could be improved and indicated interest in collaborating with his centre in Boston going forward.

We also organized a tour to Nike Art Gallery in Lekki and the day was rounded up with lunch at Chinaville Chinese Restaurant, Victoria Island with some members of the Board, friends of the Foundation and Pfizer staff in attendance.

Therapy with L-Glutamine reduces pain in patients with sickle cell disease

https://www.ucsf.edu/news/2018/07/411221/new-study-shows-l-glutamine-decreases-sickle-cell-pain-crises-hospitalizations

CSF Benioff Children's Hospital Oakland clinical researchers, in conjunction with other sickle cell centers and scientists at Emmaus Life Sciences, Inc., have demonstrated that therapy with L-Glutamine reduced the frequency of pain episodes in both paediatric and adult patients with sickle cell disease (SCD). The results of the 48-week phase 3 clinical trial were published in the July 19, 2018, issue of *New England Journal of Medicine.*

The paper, **"A Phase 3 Trial of L-Glutamine in Sickle Cell Disease,"** documented the effects of taking Endari™, a prescription-grade, pharmaceutical form of L-glutamine, as compared to placebo, for 230 patients aged 5 to 58 years of age with sickle cell disease. Endari™ was approved in July 2017 by the U.S. Food and Drug Administration based on the safety and efficacy data from this study. Glutamine is an amino acid that is involved in several biochemical reactions.

The study showed that whether administered alone or with hydroxyurea, L-glutamine reduced the frequency of sickle cell pain crises by 25 percent (a median of three events per patient in the L-glutamine group and four in the placebo group) and hospitalizations by 33 percent (a median of two hospitalizations in the L-glutamine group and three in the placebo group). Additional findings showed lower cumulative hospital days of 41 percent and a lower incidence of dangerous acute chest syndrome (ACS) by more than 60 percent.

"This study validated research on the safety of pharmaceutical grade L-glutamine which has antioxidant properties that improves the NAD redox potential in sickle cell patients. Safe neutraceuticals are of major importance to the sickle cell

community," said Elliott Vichinsky, MD, Director of Haematology/Oncology at the Northern California Sickle Center at UCSF Benioff Children's Hospital Oakland. "Our clinical trial found that L-glutamine, which does not require any routine laboratory monitoring, decreases pain events in patients by itself or in combination with hydroxyurea. It is a major advance in therapy for sickle cell disease and offers families safe, new therapeutic options."

Sickle cell disease is a genetic blood disorder that causes a distortion in the shape of red blood cells. This leads to the many symptoms and medical problems affecting children and adults with sickle cell disease, including pain, anaemia, and bone, kidney, lung and neurologic problems.

"Endari, is the first approved treatment for sickle cell disease in paediatric patients 5 years of age and older and the first in nearly 20 years for adults," said study co-author Yutaka Niihara, MD, CEO and Founder of Emmaus, which produces the product. "Our hope in sharing the results of this data from the *New England Journal of Medicine* is to increase awareness of sickle cell disease, a lifelong hereditary blood disorder which commonly affects those of African descent, as well those from Central and South America and people of Middle Eastern, Asian, Indian and Mediterranean descent. It is important for patients to know that they have a treatment option for this debilitating disease."

The double-blind trial evaluated the efficacy and safety of pharmaceutical-grade L-glutamine administered twice daily by mouth, as compared with placebo, in reducing the frequency of pain crises among patients with sickle cell anaemia or sickle β^0-thalassemia and a history of two or more pain crises during the previous year. Patients who were receiving hydroxyurea at a dose that had been stable for at least 3 months before screening continued that therapy through the 48-week treatment period.

Patients were randomly assigned, in a 2:1 ratio, to receive L-glutamine (152 patients) or placebo (78 patients). Those in the L-glutamine group had significantly fewer pain crises than those in the placebo group ($P = 0.005$) and fewer hospitalizations ($P = 0.005$). Approximately 54 percent of the patients were female and 66 percent of the patients in both trial groups were already receiving hydroxyurea. The study found that low-grade nausea, non-cardiac chest pain, fatigue and musculoskeletal pain occurred more frequently in the L-glutamine group than in the placebo group.

Funding for this trial was provided by Emmaus Life Sciences, Inc.

Study authors for the Phase 3 Trial of l-Glutamine in Sickle Cell Disease include Yutaka Niihara, M.D., M.P.H., Emmaus Medical, Torrance and University of California at Los Angeles; Scott T. Miller, M.D., State University of New York-Downstate Medical Center; Julie Kanter, M.D., Medical University of South Carolina, Charleston; Sophie Lanzkron, M.D., M.H.S., Johns Hopkins Hospital, Baltimore; Wally R. Smith, M.D., Virginia Commonwealth University Healthcare Systems, Richmond; Lewis L. Hsu, M.D., Ph.D., Victor R. Gordeuk, M.D., University of Illinois at Chicago, Chicago; Kusum Viswanathan, M.D., Brookdale University Hospital and Medical Center; Sharada Sarnaik, M.D., Children's Hospital of Michigan, Detroit; Ifeyinwa Osunkwo, M.D., Carolinas HealthCare System, Charlotte, NC; Edouard Guillaume, M.D., Interfaith Medical Center; Swayam Sadanandan, M.D., Brooklyn Hospital Center; Lance Sieger, M.D., Osbourne A. Blake, M.D., Los Angeles, Kaiser Permanente Medical Center, Inglewood; Joseph L. Lasky, M.D., Eduard H. Panosyan, M.D., Harbor–UCLA and Los Angeles BioMedical Research Institute; Tamara N. New, M.D., Children's Healthcare of Atlanta, Emory University, Atlanta; Rita Bellevue, M.D., New York Presbyterian Brooklyn Methodist Hospital; Lan T. Tran, M.P.H., Rafael L. Razon, M.D., Charles W. Stark, Pharm.D., Emmaus Medical, Torrance; Lynne D. Neumayr, M.D., and Elliott P. Vichinsky, M.D., UCSF Benioff Children's Hospital Oakland Research Center, Oakland.

46th Annual National Convention Theme Announced

This year's theme for the conference is ***"Celebrating the Diversity Within the Sickle Cell Community: Commitment, Innovation and Practice."***

With over 600 researchers, physicians, nurses, social workers and individuals living with SCD & SCT in attendance last year's convention was a major success and our largest to date. We are excited to unite again with you at the 46th Annual National Convention in Baltimore, MD on October 10-13, 2018!

https://www.sicklecelldisease.org/2017/03/07/46th-annual-national-convention/

The 4th Annual Sickle Cell Disease Symposium for Carolinas Healthcare System - Atrium Health will be held on **Saturday, October 27, 2018** at the **Speedway Club** in Charlotte, NC. The theme this year is: ***"4th Annual SCD Symposium: Racing to Improve the Access to Care & Outcomes for SCD".***

Edmonton Clinic Health Academy, University of Alberta Edmonton, Alberta, Canada

This event will join our community of physicians, surgeons, researchers, medical practitioners and patients with a deep understanding and commitment to finding the cure for Sickle Cell Anemia.

We aim to bring together multiple voices to support and build inclusive and equitable teaching and learning environments, while increasing knowledge of the continued effects of the Sickle Cell Anemia trait. The Sickle Cell Foundation of Alberta is proud to announce the following expert speakers:

Dr. Lakiea Bailey, of Georgia, USA

Dr. Carl James, of Toronto, ON

Dr. Emily Meier of Indiana, USA

Dr. Greg Guilcher of Calgary, AB

Dr. Santosh Saraf of Illinois, USA

Dr. Courtney Fitzhugh of Maryland, USA

REGISTRATION - Conference registration is now open. A variety of registration categories are available for the conference. Please see the fee details below or register online https://ourscfa.org/conference/

More information can be found on the conference website www.ourscfa.org/conference. Contact us at SCFA@buksa.com

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Edwin
Don't get stuck in the thinking
that you are insignificant and unfortunate...

HELLO! TIMI, NICE TO MEET YOU!!

A Mid-morning Chat with the Editor

Timi is a nice, beautiful, petit lady. I met her in one of her offices on Tuesday, 10th April, 2018. She had very little on by way of make-up, dressed simply, corporate but dignified. She had a warm smile of welcome on her face as I walked in after the formal security checks at the entrance. This was our first meeting.

The office was furnished simply but functionally – spacious room with her work desk, adorned with a desk-top, positioned by the window through which she could see all those coming in even before they reached her door. The natural light doused the presence of artificial lighting. A visitor's chair was placed opposite her; part of the rest of the room space was taken up by the usual file shelves but with plenty of space to move around. Walking in, I could feel a sense of determination and purposefulness - a reflection of her kind of person.

SCB Editor: Good morning! You must be Timi?!

Timi: Yes, please come in and have a seat.

I sat down on the offered chair, introduced myself formally and explained the purpose of my visit. She was comfortable and eager. So we got down to it. Here are excerpts for your reading pleasure.

SCB Editor: Please introduce yourself to our readers and tell us something about yourself.

Timi: My name is Oluwatimileyin, born to Pastor Femi and Pastor (Mrs.) Abimbola Edwin. I was born in Ibadan on the 14th of July, 1987. Most people call me Timi. I am a B.Sc. and MBA holder. I have 10 years working experience and run a Sickle Cell NGO and other business ventures.

SCB Editor: Tell us a bit about your family, how many are you?

Timi: My parents are Christians and Pastors. I am the eldest child of three children and the only female. My family is a mid-income family. I come from a family of love where my parents deprived themselves of a lot of things in order to put us in the best schools. The usual parties that many Nigerian families indulge in were certainly not on the cards. My mum lives with the condition and I can say that having a sickle cell parent helped me a lot, especially with living with the condition myself. My parents (especially my mum) raised me to not see my condition as a disability. My mum is the hardest working person I know so I really have no excuse but to be hard working myself.

SCB Editor: They did well because Education is the best gift any parent can give to his children. So what kind of education did you receive?

Timi: My primary education started in Ibadan, at a school called Life Forte Nursery and Primary School, before we moved to Lagos when I was 7 years old. Then I was enrolled in Dansol High School, Agidingbi and thereafter entered Covenant University when I was 16 years old. My MBA is from Business School, Netherlands.

SCB Editor: What did you study at Covenant and why?

Timi: Well, you know the problems associated with entering Universities in Nigeria. I was also frightened about going to public tertiary institutions because I believed that I would experience bullying like I did in Secondary School. So I made up my mind that I was going to go abroad to study and I wanted to study Law. My parents did not have the funds at the time so, as a stop gap, I sat for and passed the entrance examinations to Covenant University and other private universities then. I went for Mass Communication because they did not have a Law Faculty then. I did not mind because I saw Covenant University as a stop gap. I believed that by the second year my parents would somehow find the funds for me to go abroad.

SCB Editor: Did it happen?

Timi: No. At the end of the first year, my father told me I had better stick with it as it was a good school.

SCB Editor: So you got stuck with Covenant University and Mass Communication.

Timi: Yes, but I have no regrets.

SCB Editor: Good. When did you get to know your genotype?

Timi: I will have to go back to the circumstances of my birth. My mother is SS while my father is AS. My mother knew her status and it was while dating that she got my father to get to know his status. She became pregnant with me before the topic of genotype or marriage was discussed. Even though she was seriously advised to abort me and not have any children, she was determined to keep me as she wanted a shot at motherhood, which she was seriously discouraged against as she herself had serious health complications then. I am happy she made that choice. She told me that it was when I was 9 months that she found out about my genotype. This made her extremely hurt, she said that she went to God in prayers to keep me and that if she ever had any more children, they should be AS.

SCB Editor: What about your brothers?

Timi: Both of them are AS.

SCB Editor: Hm, while you took after your mother, your brothers took after your father.

Timi: Yes.

SCB Editor: What was your experience going through school?

Timi: It was tough, really tough. My mates picked on me regularly and I returned from school every day crying for the first three years of my secondary education. It all started when I failed to do my assignment and the six of us who failed to do the assignment were to be flogged – 6 strokes of the cane each. I was the last and I had vowed within myself not to cry or show any emotions to avoid the jeers and taunts of the other pupils. Even though I was able to mentally block out the pain and was hailed as some sort of an Amazon queen by my classmates then, I fainted at the fifth stroke. I was rushed to the clinic and there it became clear to everyone that I was SS and should never be flogged or punished again. From that point on the Principal took special interest in me. Of course the other students did not know why.

Every time students were to be punished for whatever offence, I would be exempted! This made me receive a lot of backlash from my classmates! It didn't help that I was small and absent from class a lot due to the crises I suffered.

SCB Editor: I can imagine!

Timi: Each time I came home crying, my mother would accompany me to school the next day to try and sort things out. Then one day she told me, "Timi, you have to learn to stand up for yourself. I won't always be there!"

SCB Editor: Good advice! Did you take and act on the advice?

Timi: The advice hit me deeply. I didn't act on it till much later in life. It was not easy and by the time I entered Covenant University I had a very low self-esteem. But, thank God, Bishop Oyedepo helped greatly. In those early days of Covenant as a University, the Bishop held services twice a week and his talks then helped me to build up my self-esteem. I started understanding that I could get more out of life. And I determined I would get more out of life, Sickle Cell or not. In fact it was while in Covenant that I began to see my condition as God-ordained for a reason. My life had new meaning from my point of discovery and I thank God for that.

SCB Editor: What happened after University? Did you apply for a job?

Timi: While in University I prayed that I would not have to wait to get a job even though getting jobs even then was not easy. Fortunately, after University, one of my uncles helped me to get posted to MTN for my Youth Service. The only problem then was that MTN had a policy of not retaining Corpers after the service year. At MTN I was posted to the HR Department. I used to work very hard and sowed into my colleagues.

SCB Editor: What do you mean?

Timi: I used to go to my colleagues and ask them to give me work to do. Anytime they told me I did not know their own jobs, I would tell them to teach me and I am a fast learner. So I did my own jobs and assisted others to do theirs. The result was that I became well known as a hard and an enthusiastic worker. Upon completing the service year the issue of my being retained was discussed. The Company Policy was changed and I was retained by MTN after the service year.

SCB Editor: Fantastic! How long did you stay with MTN?

Timi: Seven years and I worked in different Departments.

SCB Editor: Why did you leave?

Timi: I got bored. And I had become a target of a head hunt. So when an offer came from another company offering to pay more than twice my salary at MTN, I jumped at it.

SCB Editor: Who wouldn't? How long did you stay at the new Company?

Timi: Four months.

SCB Editor: Four months? So short! Why?

Timi: The work conditions were not very good and did not suit my health conditions. Fortunately, while still at MTN, I had started my MBA course, so I used the time after leaving the new job to finish my MBA. After which I got another job. As luck would have it, my boss informed me of an opening at a Consulate and helped me secure that job too.

SCB Editor: You mean you are holding two jobs at the same time and your bosses have no objection?

Timi: Yes. I work here till 2 pm and go over to the other place to take care of my assignments there. Then after 5 pm, I take care of matters of my NGO.

SCB Editor: Waoh! What if something comes up at your other job or the NGO needing your attention while you are still here?

Timi: Of course they can contact me either by email or phone and I then take care of matters. There is no problem.

SCB Editor: Wonderful! Can we talk about your love life?

Timi: I have dated several men, dating has never been a problem. One relationship that stood out was with someone I met in 2012. One of the first things I did, when we started dating, was to make sure we knew his genotype, which turned out to be AA.

SCB Editor: Good news. That meant none of your children would be SS. Did you get married?

Timi: No, he ditched me after two years.

SCB Editor: Really? Why? Did it have anything to do with your being SS?

Timi: I suppose so. I know his mother was rather cold to me at the beginning and later I sensed a feeling of indifference. Although he was initially very supportive he, at a stage, told me to sort out my health problems. Later he made some unpleasant remarks on several occasions and eventually called off the whole thing.

SCB Editor: That is sad.

Timi: I cried a lot after that and for a long time. I was really into him and, for the first time, I felt like I had met someone I could spend the rest of my life with. The rejection hurt so deeply. I was at rock bottom. I have come to understand that sometimes you need to be at rock bottom. Rock bottom isn't such a bad place to be at; it sometimes helps you reroute your life. That is when God comes in and pulls you out while redirecting you, if you let Him. I also was in between jobs then so a friend advised me to look for something to do to occupy my time; anything that would take my mind off the situation. Can I mention I was also suicidal then?

SCB Editor: Certainly. So what did you do?

Timi: That point in my life birthed a sickle cell blog - CrimsonBow(www.crimsonbowng.wordpress.com). The blog idea was suggested by another friend – Greg. My story was the beginning of the blog. Later we put in several things I thought would be of interest to people like me. As time went on, the blog received many visitors and became popular. We were receiving emails from people asking for help – how to cope, what to do, etc.

SCB Editor: Is that what transformed into your NGO? What is it called?

Timi: Yes. It is called **CRIMSONBOW SICKLE CELL INITIATIVE.**

SCB Editor: What does it do?

Timi: We reach out to people whose lives have

been affected by Sickle Cell Disorder as well as people living with the disorder such as parents, relations and caregivers. We offer psychological, medical and material help. We empower warrior youths. We also have members who are passionate about the cause and who do all they can to lend support to warriors.

SCB Editor: How do you empower youths? Do you give them money?

Timi: No, we do not give them money. We organize vocational trainings and also organize events that inspire them.

SCB Editor: That is great! Earlier you told me that your boyfriend told you to take care of your health. Did you? And is that how you came in contact with Sickle Cell Foundation, Nigeria?

Timi: Yes, I did. It wasn't exactly when I came in contact with Sickle Cell Foundation. What I did at the time was to do a lot of research on the sickle cell condition. This research led me to knowing and registering with Asaju Medical Centre, which is owned by Professor Akinyanju. It was in that period that I heard about the Sickle Cell Foundation, Nigeria as I had to conduct some tests there.

SCB Editor: You are obviously a very active person – two jobs and an NGO to run. Tell us what drugs are you placed on?

Timi: Hydroxyurea, Folic acid and Paludrine

SCB Editor: Do you belong to any of the Lagos Area Sickle Cell Clubs?

Timi: No.

SCB Editor: Why not?

Timi: Because my NGO has a support group that I belong to. We offer the same type of support these clubs offer.

If you are in a low, pain-filled or unhappy season, this will pass. Life does get better for those that believe.

SCB Editor: Now considering how busy you must be, tell me: do you have any free time and what do you do in your free time? Do you have any hobbies?

Timi: I do a lot of things really. I watch movies; I hang out with friends and also love to try new food when I am less busy. My hobbies are travelling and participating in Adventure.

SCB Editor: Finally, what word of advice do you have for people out there who are in a similar situation as you?

Timi: This, too, shall pass. You have to believe that life is in phases and seasons. If you are in a low, pain-filled or unhappy season, this will pass. Life does get better for those that believe. Don't get stuck in the thinking that you are insignificant and unfortunate. Change your thinking and also do as much as you can to mix with people, good and successful people. This helps you see life differently and helps you learn how to live beyond your condition. Offer a helping hand to others as well; what goes around comes around.

SCB Editor: Great words, and thank you so much, Timi, for sparing so much of your time. I have enjoyed chatting with you.

Timi: Thank you for your time. The pleasure is mine. I feel honored to have been identified by the Sickle Cell Foundation, Nigeria for this.

Sickle Cell Priapism Control

by **Professor Olu Akinyanju** and **Michael Ottun,** Sickle Cell Foundation Nigeria

Priapism (i.e. unwanted erection of the penis) is most common in young men (35%) with sickle cell anaemia (Hb SS). **Minor priapism**, the commoner of the two varieties, occurs predominantly in the early hours of the morning, during sleep, with an average duration of about two hours. However, it may recur on several nights during the week (hence referred to as stuttering priapism), disturbing sleep and causing considerable pain and discomfort.

By definition, priapism persisting **over 4 hours** is termed **major priapism**. Major priapism may persist for several days and is often preceded by a long history of minor priapism. Unlike minor priapism, major priapism often requires surgical intervention for resolution, with a risk of subsequent erectal failure or impotence. For these reasons, attempts to control or prevent minor priapism are very important.

Daily ingestion of *etilefrine* had been reported to be effective, but the discontinuation of its manufacture led to the trial of *ephedrine* tablets for the same purpose. However, contrary to an earlier report, sole or uncombined *ephedrine* tablets are unavailable in most Nigerian pharmacies. For this reason, we were eager to find other more accessible adrenergic agonist drugs for the control of priapism. In this regard, the drug we opted for was *salbutamol* (a β_2-adrenergic receptor agonist) an anti-asthmatic which, given the high prevalence of asthma in Nigeria, is widely available.

We prescribed and anecdotally found *salbutamol* tablets effective in reducing the frequency of minor priapism in some patients when taken at bed time and, sometimes, if also repeated at the onset of priapism. The effective dose was variable but its short duration of action led to our trial, in a few patients, of a longer-acting sustained release salbutamol capsule, marketed as *Ventmax SR* (by Chiesi Ltd.) but unavailable locally. Sadly, not only did we have to order the capsules from London, but its production was discontinued early in 2016.

Given the large population of patients with sickle cell disorder (SCD) in Nigeria, we requested for its local production and/or availability. Happily, a local pharmaceutical company recently provided us with 4mg and 8mg tablets of sustained release *salbutamol*, the impact of which is herewith reported.

SCD patients with priapism were invited to the National Sickle Cell Centre, Lagos, for a trial of the impact of the sustained release *salbutamol* tablets. A total of 13 patients, all with sickle cell anaemia and aged 18 to 28 years reported with stuttering priapism. They were counselled and asked to take the prescribed drug nightly before retiring to bed. The arbitrary doses were 12mg nightly for the 11 patients aged 20 or more years and 8mg for the 2 patients aged 18 years.

As shown in Table 1, seven patients responded promptly with much reduced or absent priapism in the subsequent weeks. Three patients responded slowly over the following 4 to 5 weeks, while 2 patients were lost to follow up. There were no reports of unpleasant side effects.

The last patient, 21-year old E.A., had stuttering priapism on the 5 nights following the medication and **major priapism** for 5 hours, from 1am to 6am when his mother called us. We prescribed another 12mg of the drug which he took at 9am and asked him to meet one of us at the Emergency Department of the Lagos University Teaching Hospital. Happily, by the time we met him and his mother at LUTH at about 12 noon, his priapism had resolved, most likely due to the morning dose of the drug.

The results are encouraging and hence we look forward to more trials across Nigeria with hopeful succour to the affected patients.■

Table 1: Weekly Frequency of Priapism Pre and Post Treatment

S/N	Age y	Pre Wk	Post Wk1	Wk 2	Wk3	Wk4	Wk5
1. G.O	21	6	5	3	2	0	
2. B.A	18	5	0	0	0		
3. R.A	18	1	0	0	0		
4. T.B	26	3	1	0	0		
5. A.R	20	6	1				
6. M.A.	28	3	0	0	0		
7. C.M.	22	1	0	0	0		
8. Y.O.	21	1	0	0	0		
9. J.A	21	7					
10. O.O.	26	7	7	0			
11. O.B.	21	5					
12. A.A.	21	4	5	4	4	2	0
13. E.A.	21	6	6 *				

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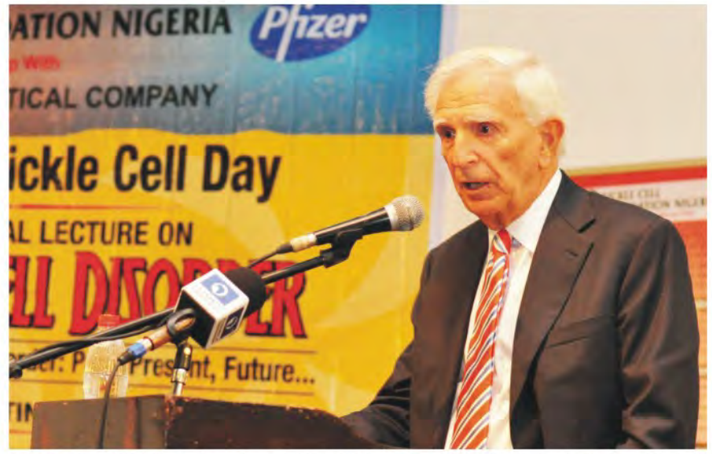


Martin Steinberg, MD delivering his lecture on "Sickle Cell Disease: Past, Present and Future" at the Annual Lecture.

Bridget Augustus, an artiste living with SCD, performing at the Annual Lecture.

The Annual World Sickle Cell Day Lecture

Presentation of Commemorative Plaque to the Distinguished Guest Lecturer by H.E. Ipeazu.

Special Guest of Honour H.E. Nkechi Ikpeazu delivering her remarks at the Annual Lecture to commemorate world sickle cell day 2018.


H.E. Nkechi Ikpeazu, Prof Olu Akinyanju, Martin Steinberg, MD & Dr. Annette Akinsete at the Annual Lecture organized to commemorate World Sickle Cell Day, 2018.

Dr. Akinsete reading the citation of the distinguished Guest Lecturer.

Dr. Margaret Odunvbun, representative of Pfizer, making a presentation at the Annual Lecture.


Participants at the exhibition stands after the Lecture.

Special Guest of Honour, Hon. Desmond Elliot of Lagos State House of Assembly, flagging off the Red Umbrella Walk

Hon. Desmond Elliot at the Red Umbrella walk.

Cross section of participants at the Red Umbrella Walk.

Dr. Muideen Bakare sharing his personal experience with SCD at the Schools Quiz organised to commemorate World Sickle Cell Day, 2018.

Dr. Bakare & Mrs. Osoba (LaRoache Leadership Foundation) presenting a copy of his book to one of the students at the Quiz competition.

Cross Section of participating Schools at the Quiz organized to commemorate World Sickle Cell Day, 2018.

Presentation of copies of "Aro'mo Leegun: A story of Sickle Cell Disease..." authored by Dr. Bakare to Sickle Cell Foundation Nigeria.

Mrs. Otaigbe and Mrs. Adesemoye presenting gifts and certificates of participation to one of the schools at the Quiz Competition.

Dr. Akinsete presenting the Trophy to Aladura Comprehensive Secondary School, Winners of the 2018 Schools Quiz

Representatives of Aladura Comprehensive Secondary School with the Trophy and Gold medals.

Group photograph with Top Laurel Grammar School who came second at the Schools Quiz Competition.

Acute Sequestration Crises in Sickle Cell Disorder

by **Professor Olu Akinyanju, OON,** Chairman Sickle Cell Foundation Nigeria
and
Dr. Chris Otigbuo, Consultant Physician/Haematologist, St. Claire Specialist Clinic

To most people, children and adults with sickle cell disorder (SCD) require the most urgent attention when they have severe bone pain crises. However, although bone pain crisis can be very alarming, other much less painful complications of SCD can rapidly become fatal, if not promptly recognised and treated. The latter include the sequestration crises.

Sequestration means seclusion and is used when blood cells are secluded in an organ in the body, such as the spleen or the liver.

Past publication has confirmed that survival of children with ASS, was much better when their parents had been previously taught to recognise the symptoms...

Acute Splenic Sequestration (ASS)

This is most common in children with sickle cell anaemia (HbSS) and occurs when large numbers of blood cells, especially the red blood cells, are trapped within the spleen. The spleen rapidly enlarges in size and the patient shows signs and symptoms of severe anaemia, such as lethargy, breathlessness on mild exertion, pallor of the palms and soles of hands and feet and of the lower eyelids. ASS is most common in children, especially those aged between 4 months and 5 years, but may also affect adults. Its cause is not known but infection should be sought for and treated. Affected children should be rushed to hospital for treatment (blood transfusion etc.).

After two episodes of ASS, the spleen should be surgically removed (splenectomy) to prevent further episodes. After splenectomy, life-long oral penicillin twice daily should be prescribed for the patient.

A 2-year old girl with sickle cell anaemia once had severe ASS so frequently that her mother had to give up her job to remain at home with her in the capital city of a West African country close to Nigeria. Her grandparents in Lagos informed me, but as her doctor there was adverse to my recommendation of splenectomy, they flew her and her mother to Lagos where we arranged for her splenectomy.

A few years ago at the 90th birthday of her grandfather in Lagos, a young energetic lady approached me and introduced herself as the young child who was flown to Lagos for splenectomy over 2 decades ago. She is presently married, has a child and is a university undergraduate in the United Kingdom. *"Ces't la vie"*, as the French would say!

Past publication has confirmed that survival of children with ASS, was much better when their parents had been previously taught to recognise the symptoms and the enlargement of the spleen. Parents or guardians of young children should ask their genetic counsellors and/or paediatricians to show them how to recognise the symptoms and signs of this condition, including an enlarged spleen.

Acute Liver (Hepatic) Sequestration

Acute hepatic sequestration is commoner in adults with Sickle Cell Anaemia (Hb SS) and, fortunately, it does not frequently recur. It also causes a severe anaemia with pallor, lethargy and breathlessness on mild exertion. The liver is enlarged and tender to touch. It is self-limiting and infection should be excluded as a possible cause. Treatment is as for splenic sequestration except that removal of the liver is not an option. Life without a liver is inconceivable.

Prenatal Diagnosis and Preimplantation Genetic Diagnosis of Sickle Cell Disorder

By

Professor Olu Akinyanju, OON , Chairman, Sickle Cell Foundation Nigeria &
Professor Oladapo Ashiru, OFR Chief Medical Director, Medical Art Centre, Ikeja, Lagos

Prenatal Diagnosis

Prenatal diagnosis (PND) literally means a diagnosis made in a fetus (an unborn child). PND of hereditary disorders was introduced over three decades ago. In most cases, it is used for the diagnosis of Down Syndrome in the fetus of pregnant women who are older than 35 years or of serious hereditary haemoglobin disorders (such as thalassaemia major or sickle cell anaemia) in the fetus of a pregnant woman with a known risk of bearing a child with such a disorder.

Many couples demand PND so they can terminate pregnancies bearing affected children. Others demand it in order to allay the anxiety associated with the uncertainty of whether the unborn child is affected and to prepare their minds for the birth of an affected child.

Publications have shown that **less than 50%** of the women who live in developed countries, with very good health care services, would terminate a pregnancy shown to bear a fetus with sickle cell anaemia. On the other hand, **over 90%** of women living in countries with poor health care and welfare services opt for termination of affected fetuses. In the latter countries, many women who have had unpleasant experiences with previous children with sickle cell anaemia would even terminate any subsequent pregnancy, for fear that it could be bearing another affected child, whereas, if both parents are healthy carriers (with Hb AS) about 75% of these indiscriminate terminations would be of healthy unaffected children (i.e. with Hb AS or AA).

These women would obviously find the option of PND very useful as part of an informed family planning exercise that would prevent the termination of the birth of healthy unaffected children. In summary, parents who tend to terminate affected pregnancies do so because they fear that as a result of poor health care services, they would be unable to provide the care needed to enable their children to survive and live tolerably good quality lives.

PND is available in the developed countries where it can often be obtained free of charge within an equitably funded health service. In the developing countries, where the prevalence of SCD is much higher and therefore the demand would be greater, PND is either not available or is limited to a relatively small number of couples who can afford to pay for the service.

Three methods can be used, namely, **fetal blood sampling (FBS), amniotic fluid sampling (amniocentesis) and chorionic villus sampling (CVS). Fetal blood sampling** is not possible until late in pregnancy when blood can be found in the fetus. For this reason, it is hardly used for PND of sickle cell anaemia.

Amniocentesis is feasible from about 18 weeks of pregnancy, but the amniotic fluid has to be cultured in a suitable sterile medium for a couple of weeks to allow for the growth and multiplication of amniotic cells. These cells are then harvested and their DNA extracted for analysis. PND by amniocentesis is thus a relatively late and slow process during the course of which the sample could become infected and thus, unsuitable.

An **advantage of CVS** is that it can be done early in pregnancy - from about 10 weeks - before the prying and questioning eyes of neighbours can detect the pregnancy. DNA is extracted from the CVS and analysed after amplification by polymerase chain reaction (PCR) to yield a result within a few days. CVS is done under ultrasound guidance and requires skilled and experienced operators. The main disadvantage is the risk of a miscarriage, which is about 1-2%. The chorionic villi (destined to become the placenta) are approached through the cervix of the uterus or through the abdominal wall. General anaesthesia is not required.

Many experienced operators, including ours at the National Sickle Cell Centre in Lagos, now favour the trans-abdominal route because it is associated with a much lower risk of miscarriage of the pregnancy.

Figure 1 illustrates the trans-cervical and trans-abdominal routes.

Availability of PND in Nigeria

PND has long been available at the National Sickle Cell Centre in Lagos. The stalwart who has ensured this achievement has been Dr. Joseph Akinde, a committed consultant obstetrician, who is ably supported by a team including **Mrs. Esther Adesemoye +234 803 319 5177 and Mr. Sola Ojewunmi +234 806 833 2513** to whom service enquiries should be made. Training in PND by CVS is conducted for interested young obstetricians with ultrasound competence. Applications for training, with curriculum vitae, should be forwarded to info@sicklecellfoundation.com. We hope that the service will become available in various regions of the country and would be subsidised to improve access to it and for research that could make less invasive PND feasible.

Preimplantation Genetic Diagnosis (PIGD)

Prenatal diagnosis (PND) can tell you the Hb genotype of an unborn child. On the other hand, preimplantation genetic diagnosis (PIGD) is a process for choosing your child's Hb genotype before pregnancy. A process known as in-vitro fertilization (IVF) is employed. The father's sperm is collected and injected into the mother's previously retrieved eggs, within a sterile container.

The Hb genotypes of the resultant embryos are analysed by DNA after amplification by polymerase chain reaction (PCR). The couple can then have unaffected embryos (i.e. with Hb AA, AS or AC) implanted into the mother's womb. PIGD is particularly desired by couples (e.g. HbAS married to HbSS) who have a 50% chance of having an Hb SS affected child with each pregnancy.

We employ the technique of trophectoderm biopsy, which is an improvement over our earlier publication that biopsied embryos on day 3 with one single blastomere taken out of the eight cells blastomere for analysis. In comparison, trophectoderm biopsy is performed on a day 5 embryo called a "blastocyst". We remove about 4 to 5 cells from the external layer of the embryo and the extracted cells are analyzed for abnormalities. This new process has a higher degree of specificity with a significant percentage increase in the pregnancy success, making it more accessible and acceptable.

The technique of PIGD is applied in conjunction with Assisted Reproductive Technology (ART). ART refers to conception requiring the complex handling or manipulation of both the male and female gametes in-vitro to facilitate pregnancy.

What is IVF?

IVF means fertilization achieved outside of the body. It involves ovulation induction, oocyte retrieval, sperm preparation, oocyte stripping, insemination and fertilization in a culture dish and finally embryo transfer. Oocyte retrieval and embryo transfer processes are done under ultrasound guidance. It is also important to know that gamete handling is done under strict temperature control. Without the development of IVF, PIGD would not be possible.

Currently, at the Medical Art Center in Lagos, we offer PIGD for genetic conditions including sickle cell anaemia. We work in collaboration with Genesis Genetics, a world-renowned genetics institute, and pioneers of PIGD for inherited genetic abnormalities.

Steps Involved for Sickle Cell Disorder

Preimplantation genetic testing can distinguish between genetically normal or affected embryos. Currently, this is the only way to determine the genetic condition of an embryo before pregnancy.

Preliminary work before PIGD: Probe preparation

To go through PIGD for sickle cell anaemia, DNA samples from family members must be obtained to build a probe. This probe which contains specific genetic information received from the collected DNA will be used to test the cells biopsied from the embryos. DNA samples from only direct family members (siblings or the parents of the two partners) are required so there is no need to involve extended relatives. PIGD probe preparation takes 8 to 10 weeks.

Oocyte (Female Eggs) Retrieval

The female partner is given medications to create multiple follicular development and ovulation. Once these follicles are well developed, they are retrieved from the patient under conscious sedation using a unique ultrasound-guided technique. The oocytes are then isolated from the follicles and allowed to rest in the incubator for about 3-4 hours before insemination. This time is used to prepare the sperm for IVF.

Under a very specialized microscope, one sperm cell is aspirated from the very few ones and is injected directly into the egg cytoplasm. This ensures fertilization in significantly high numbers. The inseminated eggs are kept in the incubator in the IVF laboratory for about 5 to 6 days. The fertilized embryo will rapidly divide into the 2-cells, 4-cells, 8-cells and blastocyst stages at which point embryo biopsy can take place.

Yoyo *Figure 1: Trans-cervical and Trans-abdominal Routes*

Embryo Cryopreservation (Freezing)

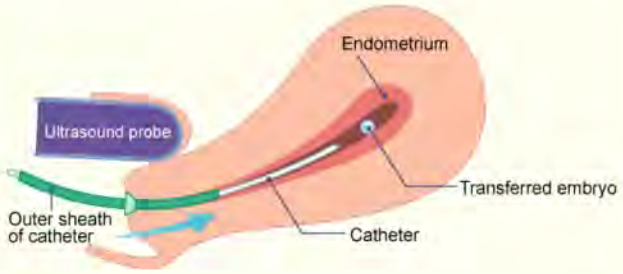
Embryos that have been biopsied are preserved in cryo devices. They are frozen by a technique called vitrification, which is described as very rapid cooling at very low temperatures. They are appropriately labeled and stored in liquid nitrogen and can be thawed when needed. The embryos are safe in the nitrogen tanks for years. Our first sickle cell free delivery was, in fact, a boy conceived from a frozen embryo transferred to his mother.

Once the DNA in trophectoderm cells has been amplified by PCR, they can now be analyzed by a technique known as comparative genomic hybridization.

CGH looks at the entire set of chromosomes to ensure that the correct number of chromosomes is present.

Also, CGH ensures there is a proper balance of chromosome sets providing the genetic integrity of embryos from patients.

With our collaborating center, we usually get the results of the PIGD test within ten days. The results indicate which embryos are Hb AA, Hb AS or Hb SS or otherwise. We consult the patient to discuss the outcome and to plan and prepare for embryo transfer.



Embryo Transfer

Today in Nigeria, sickle cell carrier couples (Hb AS & Hb AS) can now screen their embryos before conception to avoid having a sickle cell (Hb SS) baby. Although this process is quite costly, it is hoped that various groups, including the government, can come to the financial aid of couples in need of PIGD.

More research is required to improve the speed of diagnosis and make the process less expensive. Another major area that deserves investigation and, if possible, a Nobel prize nomination is the study of the understanding of the window of implantation. The reason is that this is the only significant gap in knowledge for us to achieve close to 95% success in IVF. Many scientists have predicted that by 2025 the success rate of IVF should approach 90% if not 95% with more research conducted in this field. ■

For more information visit
www.medicalartcenter.com,
or call +234 1342 9031 or send email to
info@medicalartcenter.com

Prevention of Infection in Children with Sickle Cell Disorder aged under 10 years

by Professor Adebola O. Akinsulie, Consultant in Paediatric Haematology,
Department of Paediatrics, Lagos University Teaching Hospital

Sickle cell disorder (SCD) is a common genetic problem with enormous challenges. It is a major cause of illness and death in millions of children of Mediterranean, Middle Eastern, Indian and particularly of African descent. It poses a great challenge to the affected children especially in Nigeria with the largest concentration of SCD in the world. It is estimated that over 100,000 new births with the disorder occur annually in the country.

The term SCD includes all the conditions in which the clinical manifestations are attributable to the presence of sickle cell anaemia, (Hb SS) or haemoglobin S in association with another haemoglobin variant (e.g. Hb SC). Children with sickle cell anaemia present with the most severe clinical manifestations followed by the heterozygous HbS-$ß^0$ thalassaemia, HbS-$ß^+$ thalassaemia and Hb SC in order of decreasing severity. Children with the sickle cell trait (Hb AS) are healthy.

Affected children are susceptible to frequent infections including malaria, pneumonia, septicaemia and meningitis, particularly caused by encapsulated bacteria such as the pneumococcal bacteria. They are also susceptible to multiple-sited osteomyelitis (bacterial bone infection), particularly those caused by Salmonella bacteria.

Prevention of Infections in SCD

SCD confers an increased susceptibility to infection, especially to certain bacteria, and at the same time, infection provokes a cascade of SCD-specific illnesses. Historically, infection was a major cause of death in SCD, particularly in children, and it was implicated in 20-50% of deaths in prospective cohort studies over the last 20 years. Worldwide, it remains the leading cause of death, particularly in less developed nations. In developed countries, measures to prevent and effectively treat infection have made a substantial contribution to improvements in survival and quality of life, and are continually being developed and extended. However, progress continues to lag in less developed countries where the patterns of illness and death are less well defined and implementation of preventive care is poor.

1. Prevention of overwhelming pneumococcal sepsis.

Children with sickle cell disorder are more susceptible to pneumococcal bacterial infection than other children are, and this has also been

confirmed in sub-Saharan Africa. Pneumococcal disease burden in children includes otitis media (ear infection), pneumonia (chest/lung infection), bacteraemia (blood stream infection) and meningitis (outer brain infection). Fortunately, daily oral penicillin has been shown to be preventive. Furthermore, provision of prenatal diagnosis, new-born screening, antibiotics and immunisation are believed to reduce mortality (death rates) in low to middle income countries from 90% to 50% and from 10% to 5% in high income countries. Evidently, the disorder is most fatal in the poorer parts of the world, which co-incidentally have the highest prevalence. Hence, all children with SCD should receive prophylactic (preventive) oral penicillin twice daily or erythromycin if they are allergic to penicillin. However a study showed no benefit in older children but it is advisable to continue till adolescence as cases of older children and adolescents succumbing to pneumococcal sepsis soon after coming off prophylactic penicillin have been reported. Children should also be vaccinated with a polyvalent conjugate vaccine (PCV) against Pneumococcal bacteria. It is heart-warming that this vaccine has recently been included in the National Program on Immunizaion in Nigeria. Active preventive immunization of infants and children is from 6 weeks until 5 years of age. One such schedule is 2 primary doses at 6 weeks and 10 weeks of age and a booster dose at age of 9 weeks.

2. Prevention of Malaria

In malaria endemic areas of sub-Saharan Africa, prophylactic anti-malaria medication, especially daily Proguanil, are given to children with SCD as they are susceptible to malaria and its severe complications.

3. Maintenance of Wellness in SCD

It is important to maintain wellness and prevent infection in SCD because infection may lead to both bone pain and/or anaemia crises in SCD. This is achieved by offering holistic care management which includes comprehensive genetic counseling, good hydration, balanced nutrition and daily folic acid. It also includes oral hydroxyurea when necessary and prompt management of ailments.

Watermelon is a delicious and refreshing fruit that's also good for you. It contains only 46 calories per cup but is high in vitamin C, vitamin A and many healthy plant compounds.

Helps You Hydrate

Drinking water is an important way to keep your body hydrated. However, eating foods that have a high water content can also help. Interestingly, watermelon is 92% water (1).

What's more, a high water content is one of the reasons why fruits and vegetables help you feel full.

The combination of water and fiber means you're eating a good volume of food without a lot of calories.

SUMMARY

Watermelon has a high water content. This makes it hydrating and helps you feel full.

Contains Nutrients and Beneficial Plant Compounds

As far as fruits go, watermelon is one of the lowest in calories — only 46 calories per cup (154 grams). That's lower than even low-sugar fruits such as berries (2).

One cup (154 grams) of watermelon has many other nutrients as well, including these vitamins and minerals:

Vitamin C: 21% of the Reference Daily Intake (RDI)
Vitamin A: 18% of the RDI
Potassium: 5% of the RDI
Magnesium: 4% of the RDI
Vitamins B1, B5 and B6: 3% of the RDI
Watermelon is also high in carotenoids, including beta-carotene and lycopene. Plus, it has citrulline, an important amino acid.

www.healthline.com/nutrition/watermelon-health-benefits#section1

PUZZLE

O	S	P	L	O	M	A	T	E	C	A	R	A	P
N	U	B	O	R	I	O	N	I	C	H	T	R	R
O	L	C	H	A	S	T	L	I	N	A	U	G	O
E	L	E	C	T	R	O	G	D	A	S	T	C	P
T	I	T	P	H	R	P	O	S	T	E	R	H	E
L	V	A	I	E	R	H	N	S	O	R	E	L	B
E	T	K	B	A	F	O	L	I	C	I	N	P	A
U	C	O	C	T	E	R	T	S	A	O	E	R	N
A	N	B	Y	E	G	E	S	T	C	R	E	F	D
L	I	D	T	R	N	N	Y	M	I	N	D	D	A
O	L	S	E	L	I	S	H	G	D	G	L	O	V
O	P	Z	S	V	R	I	T	A	N	V	E	F	E
W	M	S	I	S	Y	S	S	E	A	L	S	I	N
N	O	T	T	O	C	T	A	R	H	L	S	F	H
S	H	Y	D	R	O	X	Y	U	H	K	O	P	J

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THE GIVE ME FIVE CAMPAIGN

The "Give Me Five" campaign is aimed at achieving adequate and sustainable funding by appealing to thousands of Nigerians to donate monthly, an affordable amount, N5,000 (five thousand naira) only, to the Sickle Cell Foundation Nigeria (SCFN) to fund programmes including countrywide sickle cell research, capacity building of health care personnel and prevention and treatment of the complications of sickle cell disorder (SCD).

We graciously invite you, your friends and colleagues to support the work of the SCFN through our "Give Me Five" campaign. Your support to this laudable project will make a big difference and, given the pervasiveness of SCD in Nigeria, the reflected benefits will be incalculable.

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Be Careful; Your Cough can be Worse than You Think

by **Professor Muuta Ibrahim,** Consultant Paediatrician, Aminu Kanu Teaching Hospital, Professor of Paediatrics, Bayero Universiity, Kano

Sickle cell disorder exposes sufferers to a myriad of conditions that could pose serious threat to well-being and even to life, if not recognized and treated promptly. One of these common conditions is cough. Ordinarily, most people have an attack of upper respiratory infection and attendant cough more than a few times in the year. Children, in particular, get lots of cough and colds throughout the year. The average child may have up to six such attacks annually. The child should recover within a week without further complications. However, what may pass as an ordinary respiratory infection or chesty cough could be a more serious condition in sickle cell disorder sufferers known as "Acute Chest Syndrome". What then is this condition and how can it be recognized? How should a suspected case of Acute Chest Syndrome be handled at home and in a health institution?

Acute chest syndrome (ACS) is an acute lung or chest illness in a child or adult with sickle cell disorder recognized by acute signs and symptoms such as fever, cough, breathlessness or labored breathing, wheeze, chest pain or pain on taking deep breath. It is confirmed by chest x-ray findings of new opacity or shadows in the lung x-ray. Pain on deep breath may be the main finding making the child take shallow breath but breathlessness is also seen. A combination of pain and breathlessness may also be encountered. The cough is often dry with no mucus but could also be productive of sputum.

Acute chest syndrome is a very serious condition and a major cause of death in children and adults with sickle cell disorder. Once it is suspected, the child must be taken to a hospital or good clinic with doctors for prompt diagnosis and treatment.

The causes of acute chest syndrome include:

1. Chest infection
2. Embolism of bone marrow fat to the lungs (the embolus obstructs blood vessels in the lungs causing infarction of lung tissue)
3. Aggregation of sickled red blood cells in pulmonary or lung vessels causing vaso-occlusion and lung infarction.

Infection is a more common cause of acute chest syndrome in children while fat embolism and vasoocclusion are more common in adolescents and adult sickle cell sufferers. What are the recognizable features of acute chest syndrome?

In a child or adult with symptoms of cough or colds, the following features should make one suspect ACS.

- Fever
- Breathlessness
- Cough
- Wheezing or noisy breathing
- Chest pain on deep breathing

- Flaring of the nostrils
- Blood in sputum
- Rapid heart rate as felt on the pulse
- Rapid breathing (more than 40 cycles per minute in a child or 30 cycles in an adult)
- Low oxygen tension seen as blue lips or tongue
- Low SpO_2 <95%
- Progressive restlessness as a result of low oxygen
- Inability to eat

What should be done once ACS is suspected?

The child or adult must be taken to a good hospital immediately. There is no time to waste trying to give home treatment or consulting a 'nearby medicine store or chemist'. This is a medical emergency and must be handled as such in all ramifications. The patient will usually be admitted and placed on oxygen therapy while investigations to confirm the diagnosis are made. The initial treatment may consist of broad spectrum antibiotics and fluid replacement as the patient may have suffered dehydration from breathing with an open mouth and reduced intake of water. However, the maintenance fluid after replacement is kept at about 75% of requirement.

Initial investigations shall include a chest x-ray, which may show opacities or shadows in the lung fields. Other investigations may include a blood culture and a sputum smear, Gram stain, microscopy and culture where the cough is productive. This may show the invading organism.

Treatment may also include an exchange blood transfusion to reduce the quantum of sickled red cells in circulation. Bronchodilators such as nebulized salbutamol may be given to further open up the blocked airways. Analgesics to reduce pain and temperature may be administered. In situations where the doctor suspects fat embolism, anticoagulant therapy using low molecular weight heparin may be administered subcutaneously.

Prevention

It is essential that parents of children and adolescents with SCD should ensure that they receive a full complement of routine immunizations. Furthermore, additional protective immunization against Haemophilus influenza type B and Pneumococcus (Pneumovax) must be given to all SCD patients irrespective of age. This will ensure protection against the most common serotypes of these bacteria causing lung infection and invasive disease. Pneumovax is given in the second year of life and then every 5 years thereafter.

It is essential that parents of children and adolescents with SCD should ensure that they receive a full complement of routine immunizations.

All SCD sufferers should routinely take oral penicillin twice daily. This has been shown to be protective against common bacteria causing illness in them. Making sure that a child with SCD takes oral penicillin twice a day is one of the most important things a parent can do for their child. Children with SCD are 600 times more likely to get pneumococcal infection than other children. This is because of the weakened function of their spleen, an organ that helps the body fight and kill invading bacteria. The dose depends on the age of the patient:

- 62.5 mg twice a day until 1 year of age
- 125 mg twice a day from 1 year until 3 years of age
- 250 mg twice a day from 3 years onwards

Prevention of recurrence

Once the patient is discharged, hydroxyurea therapy is commenced in order to prevent recurrence of ACS. Other measures would be paying attention to use of routine drugs and counselling to avoid extremes of temperature.

Sickle Cell Clubs and Affiliated Organisations in Nigeria

ABUJA

Sickle Cell Aid Foundation
12, Nairobi Street, Off Parakou Street,
Wuse 2, Abuja.
c/o Bukola Bolarinwa
0905 779 6016

ANAMBRA STATE

Orient Sickle Cell Foundation
329, Zik's Avenue, Awka.
c/o Mr. Chidi T. Mokebe
0803 899 5415
tholcylab@yahoo.com

BAYELSA STATE

Yenagoa Sickle Cell Health Watch
Bayelsa State Health Management Board,
Yenegoa.
c/o Mrs. Philomena Aladei
0803 897 7327

CROSS RIVERS

Sickle Cell Club
Medical Centre,
University of Calabar, Calabar.
c/o Prof. Patrick Owai
0806 366 7305

DELTA STATE

Sickle Cell Club Asaba
General Hospital, Okwe, Asaba
c/o Matron Betty Ulueme
0803 724 0686

Sickle Cell Club Sapele
Central Hospital, Sapele
c/o Ms. Ngozi Mabiaku
0803 810 4596

Asaba Sickle Cell Club
Ministry of Health, Asaba
c/o Mrs. Emina O.
0703 875 0015

EDO STATE

Sickle Cell Club Benin City
Sickle Cell Anaemia Centre,
Along Hotel Plaza Road,
Opposite Golf Course, GRA, Benin City.
c/o Prof. (Mrs.) Caroline Omoti
ediomoti@yahoo.com

ENUGU STATE

Cannon Sickle Cell Foundation
118, Agbani Road, Enugu
c/o Charity Aneke
0803 784 4949

KADUNA STATE

Sickle Cell Patient Health Promotion Centre
No. 1A, Badarawa New Extension,
By Wurno Road, Kaduna.
c/o Hajiya Badiya Magaji Inuwa
0803 787 9799, 0808 473 5313
sicklecellpatient@gmail.com
www.sicklecellpatient.org

Sickle Cell Club Kafanchan
Patrick Ibrahim General Hospital,
Kafanchan, Kaduna State.
c/o Mrs. Linda Yari 0806 010 3563
Mrs. Florence Olukokun 0703 808 8908

KATSINA STATE

Malaria and Sickle Cell Control Dept.
Ministry of Health,
Katsina, Katsina State.
c/o Mrs. Halima Idowu – 08039703222
halimamusa@yahoo.com

KANO STATE

Sickle Cell Club
Aminu Kano Teaching Hospital,Kano.
c/o Dr. Dalhatu Gwarzo
0803 624 1535

Sickle Cell Club
Murtala Mohammed Specialist Hospital,
Kano.
c/o Hadiza Dan Isa
0803 349 4894

LAGOS STATE

Sickle Cell Area Club of Apapa
Nigerian Ports Authority Sports Club,
Surulere, Lagos.
c/o Mrs. May Ozor
0803 718 8442

Eko Island Sickle Cell Area Club
Ante-Natal Clinic Hall,
Onikan Health Centre,
Lagos Island.
c/o Mrs. Bisi Olaiya
0805 547 6270

Sickle Cell Area Club of Surulere
Medical Outpatient Department,
1st Floor, LUTH, Idi-Araba,
c/o Prof. M.O. Kehinde
0802 327 1415

Sickle Cell Area Club of Ojo
Nigerian Navy Hospital,
Navy Town, Ojo.
c/o Mr. Obe
0813 477 0264

Sickle Cell Area Club of Isolo
General Hospital, Isolo.
c/o Mrs. Aderonke Adebayo
0701 665 7713
ronkebabe2015@gmail.com

Sickle Cell Area Club of Epe
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0805 350 7254

Sickle Cell Area Club of Ikorodu
General Hospital, Beach Road,
Ikorodu.
c/o Mrs. Damilola Aseyege
0803 731 3865

Sickle Cell Clubs and Affiliated Organisations in Nigeria

LAGOS STATE

Temitayo Awosika Help Foundation (TAHF)
2nd Floor, 3 Olayinka Bamgbose Street,
Off Toyin Street, Ikeja, Lagos.
c/o Alhaja Tawakaltu Giwa
0807 760 0917

Tolu Akinduro Foundation
c/o Mrs. Bambo Akinduro
0803 304 2375
bambo@gibbot.net

Unilag Sickle Cell Club
University of Lagos,
Unilag Health Centre.
c/o Dr. David Ekwoaba
0803 078 0988
ekwoaba2002!@yahoo.com

Sickle Cell Area Club of Badagary,
Government College, Agbara, Badagary.
c/o Mrs. A.S. Abegunde
0803 582 1373

Sickle Cell Area Club of Yaba
c/o Mr. Shogbamu Wasiu
0909 916 2262, 0805 604 5561

Sickle Cell Area Club of Ifako-Ijaye
c/o Mrs. Modupe Popoola
0802 223 6484

Nike Opalemo Sickle Cell Foundation
3rd Floor, Eleganza Plaza,
Billings Way, Oregun, Lagos.
c/o Mrs. Nike Opalemo

Crimson Bow Sickle Cell Initiative
42, Oduduwa Cresent, GRA, Ikeja.
c/o Timi Edwin - 0816 182 6816

Noah's Ark Foundation For Sickle Cell
19, Abdulraman Useni Road,
Off Tobiloba Lawal Road,
Ologolo, Lekki-Epe Expressway, Lagos.
c/o Osasele 0809 317 8452
Ego 0812 117 8749

Soulage Foundation For Sickle Cell
13A, Fagba Crescent, Agidingbi,
Ikeja, Lagos.
c/o Kofo 0814 743 7098

Tonymay Foundation Sickle Cell Aid
6, Esubiyi Street, Mende,
Maryland, Lagos.
c/o Anthony Anerua
0814 825 4985, 0806 341 2027

Sickle Cell Advocacy and Management Initiative (SAMI)
15, Modupe Johnson Crescent,
Off Adeniran Ogunsanya Street, Surulere.
c/o Maureen Nwachi 0807 782 8255

OGUN STATE

Marvel Sickle Cell Foundation
8, Leye Akiboye Street, Mowe.
c/o Stephanie Agbaje
0803 325 1685, 0802 760 6027

Dabma Sickle Cell Foundation
48, Diya Street, Gbagada, Lagos State.
8, Favour Street, Magboro/Makogi,
Ogun State.
c/o Pastor Emmanuel Ibekwe
0803 349 8555

Aglow Sickle Cell Club
State Hospital, Sokenu, Abeokuta.
c/o Mrs. Olufunke Oyeneye
0803 573 1840

Sickle Cell Club of Ijebu Land
Iye Subomi Outpatient Complex,
General Hospital, Ijebu Ode.
c/o Mrs. Taiwo Adebanjo
0805 512 5468, 0803 650 1115
taadebanjo@yahoo.com

Ijebu Ode Sickle Cell Club
Iyesubomi Child Care Centre,
State Hospital, Ijebu Ode.
c/o Dr. N.O. Abimbola

OSUN STATE

LAUTECH Sickle Cell Club Osogbo
c/o Dr. V.O. Mabayoje
LAUTECH Teaching Hospital, Oshogbo.
0803 329 4874

Ile – Ife Sickle Cell Club
OAUTH, Ile – Ife.
c/o Prof. A.O. Akinola
0803 344 7557

OYO STATE

Sickle Cell Club Ogbomosho
Medical Social Services Department
Baptist Medical Centre,
Ogbomosho.
c/o Mr. Amos O. Adeniji
0802 738 3985

PLATEAU STATE

Omega-Cares Foundation
9, Utan Lane, Rock Haven, Jos.
c/o Dr. Kenneth Enwerem
0806 816 0904
kennethenwerem@gmail.com

Kathy Life Builder Foundation
16, Issac Nze Close,
Behind Fototek, Dogon Karfe,
Jos, Plateau State.
c/o Mrs. Nancy Ocholi 0816 498 3875
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