

Management protocols



ARISE

African Research And Innovative
Initiative For Sickle Cell Education

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Juliana Olufunke LAWSON MBBCh, FWACPaed, MSc(Public Health)

Paediatrician

Zankli Medical Centre, Abuja

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Management protocols

Health Promotion and Preventive Care

- Organizing Clinical Care
- Prevention of infection
- Genetic and reproductive counselling
- Nutrition, Growth and Development monitoring in children
- Transition to adult care

Management of Acute Complications of SCD: Anaemia, Pain Crisis, ACS, Acute Splenic Sequestration, Stroke, Priapism, Infections

Screening for Specific Complications of Sickle Cell Disease and Management of Complications of SCD

Stem Cell transplant



Clinical Care

- Network of care : community, primary, secondary and tertiary care. Communication between the different levels of care and Referral system
- Multidisciplinary team
- Frequency of visits
- Education of parents and patients about the disease during each outpatient visit and symptoms that require urgent medical assessment. Assess adherence to medications: folic acid, penicillin prophylaxis, hydroxyurea, up-to-date immunisation and monitoring of Hb level.
- Investigations: FBC, reticulocyte count, and antibody screening and identification during the first visit. Other investigations determined by clinical presentation.
- Adequate records of admissions, number of pain episodes and other complications



Prevention of Infection

- General, Invasive Pneumococcal disease, Malaria, Immunization, Prevention of enteric gram-negative organisms (salmonella, E. coli, klebsiella, etc.)
- Basic hygiene and preventive measures, Insecticide treated nets and indoor residual spraying
- Medical attention for fever ($>38.3^{\circ}\text{C}$ and other signs of infection)
- All routine immunizations
- Immunization of special Interest to SCD: pneumococcus, Hib, Hepatitis B and meningococcus, Salmonella typhi immunization for those at risk of infection
- Oral penicillin prophylaxis
- Folic acid
- Proguanil



Genetic and reproductive health counselling

- Counselling by trained personnel
- Educate all children from school going age with their parents/caregivers and adults routinely about SCD at each visit
- Prompt patients and parents to talk about stigma and their experience with how their communities and others relate to them, dispel myths
- Encourage patients with SCD to have a plan for reproduction including genotype status of potential partner



- **Nutrition and hydration**

- Adequate fluid intake.
- Nutrition advice and growth monitoring
- Early referral to Dietitian

- **Growth and Development monitoring**

- Serial growth measurements should be performed to capture both acute changes and long-term growth velocity using age-appropriate growth charts



Assessment and Management Protocols for SCD

- Vaso-occlusive pain episode (pain crisis) and chronic pain
- Anemia
- Prevention and treatment of infections and malaria
- Acute Chest Syndrome
- Acute Splenic Sequestration
- Prevention and management of stroke
- Priapism
- Complications and the various organ damage syndromes associated with the disease
- Pulmonary hypertension
- Pharmacotherapy



Screening for Specific Complications of Sickle Cell Disease and Management of Complications of SCD

- Stroke Risk
- Renal Disease
- Cardiovascular Disease and Hypertension
- Pulmonary Disease
- Retinopathy
- Avascular necrosis



Other Considerations

- Psychological complications
- Analgesics and addiction
- Transfusion Therapy in Sickle Cell Disease/Iron chelation
- Hydroxyurea Therapy in Sickle Cell Disease
- Transition to adult care
- Hematopoietic Stem Cell transplant : Limited availability of transplant experts and centers, Requirement of a matched donor, Cost and Outcome



CONCLUSION

- There are presently variable guidelines for management of sickle cell disease in Nigeria
- There is need for universally acceptable protocols and guidelines for conditions associated with sickle cell disease to be developed and distributed
- Protocols for screening for various conditions should also be made available
- Institutions from the lowest to the highest cadre should be encouraged to follow these guidelines/guidelines





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