iAHO
Integrated African Health Observatory

World Health Organization
African Region

Analytical Fact Sheet
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NCD Team

Sickle cell disease: the silent killer in Africa

Rationale

Sickle cell disease (SCD), an inherited blood disorder, alters the haemoglobin in the red blood cells, which carries oxygen through our bodies. The red blood cells become rigid and sticky and look like a crescent or 'sickle' shape, struggling to flow smoothly in the bloodstream. Blockages can happen anywhere in the body, bringing various health issues that differ from person to person. These include anaemia, tiredness, swelling in the hands and feet, a high risk of stroke, frequent painful episodes known as pain crises, infections, stunted growth, and vision problems. It also leads to complications such as chronic pain and organ damage. Currently, bone marrow transplantation is the only curative treatment. Treatments are also available to manage the symptoms effectively..

Key messages

- 66% of the 120 million people living with SCD worldwide live in Africa.
- Around 1,000 new babies are born every day with a high risk of SCD, making it the most widespread genetic disorder in Africa region.
- SCD is most prevalent in Africa, affecting about 800 out of every 100,000 people.
- 6.4% of under-five mortality in Africa is attributed to SCD.
- 50-80% of infants in Africa born with SCD die before the age of 5 years.
- 38,403 deaths from sickle cell disease in 2019, a 26% increase since 2000.

SCD overview

Sickle cell disease (SCD) needs urgent attention. In many countries, there are no or limited newborn screening programs. African countries can improve SCD management and control through a comprehensive SCD management approach focusing on prevention, screening, and management strategies. Implementing a national newborn screening program with comprehensive SCD management will significantly reduce the economic and social burden associated with SCD while improving health outcomes and quality of life.

Symptoms and complications

Sickle cell disease symptoms starts early in childhood vary widely from mild to severe, including:

- **Pain crises**: Periodic episodes of extreme pain caused by blockages in small blood vessels. The pain can last for few hours to a few days.
- **Anaemia**: premature death of red blood cells leads to fewer healthy cells, resulting in limited oxygen transport throughout the body. This causes fatigue.
- **Stroke**: results from reduced or blocked blood flow to the brain. The brain function is impaired leading seizures, trouble walking, headache, confusion and loss of consciousness. Ischaemic strokes are common in children, and haemorrhagic strokes are more common in adults.
- **Organ damage**: restricted blood flow, potentially causing various complications such as acute chest syndrome and tissue death (avascular necrosis) in bones and vital organs such as the kidney and liver.
- **Other complications** include blood clots, fever, leg ulcers, vision loss, splenic sequestration, deep vein thrombosis, etc., all related to the disease's effect on blood flow and organs.

Diagnosis and screening

Simple blood tests can diagnose SCD:

- **Blood tests** can identify the specific haemoglobin of SCD in venous blood in both adults and young children.
- **Test usually performed are:**
  1. **Newborn screening**, blood is taken from the baby's heel with a special type of paper and test it in a laboratory;
  2. **Prenatal screening**, before the baby is born; amniotic fluid or placental tissue are used for diagnosis.
- **Genetic test** can determine whether an individual is carrying the SCD gene or if the genetic trait indicates the potential to pass it on to offspring.

Treatment and management

Effective management of sickle cell disease includes :

- **Pain relief** - Pain is common in people with SCD; they can use over the counter or prescribed medicines, heat therapy and relaxation techniques.
- **Hydroxyurea therapy** increases foetal haemoglobin levels, reducing the occurrence of pain crises and complications.
- **Antibiotics** can be used if there is evidence of infection.
- **Blood transfusions** - refill the normal red blood cell count to improve tissue oxygenation.
- **Bone marrow/stem cell transplants** - this procedure can sometimes be used as a potential cure.
- **Additional treatment and care** include treatments associated with specific conditions such as pulmonary hypertension, stroke, and organ damage.

Addressing SCD in Africa

The importance of early detection and management

Early diagnosis in newborns through point-of-care screening allows for early treatments that can save lives, alleviate symptoms, and reduce suffering from pains.

Regular health checks are crucial for adults with SCD to monitor for signs of organ damage. Early detection and effective management are essential improving the quality of life.

- SCD treatment centre provides comprehensive care to manage symptoms, prevent complications, and improve the quality of life for SCD patients.
- Haematopoietic stem - cell transplantation (HSCT) facility offers potential cure for sever cases, by replacing the patient's defected hematopoietic stem cells with healthy ones from matched donors.
- **The number of SCD treatment centres and HSCT is limited compared to the affected population (see figure 1).**
- **Lifesaving early treatment is directly proportionate to the coverage of treatment centres and access to primary care.**
- **Standardized new-born screening and early intervention for children was initiated in twelve Sub-Saharan countries:** Benin, Burkina Faso, Cameroon, Democratic Republic of the Congo, Ghana, Kenya, Liberia, Mali, Nigeria, Senegal, United Republic of Tanzania, and Uganda.

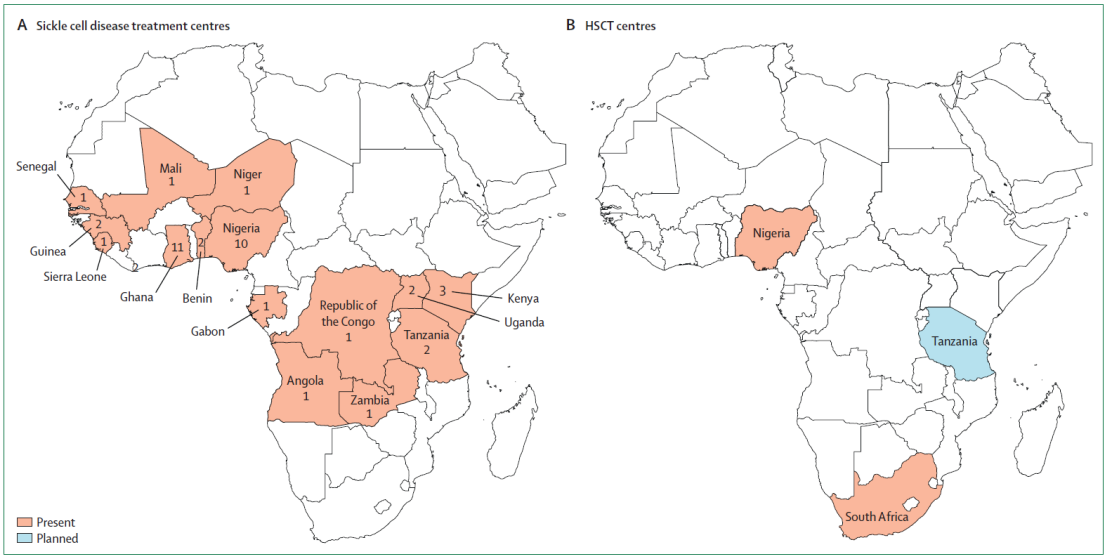

Figure 1: Number of reported sickle cell disease treatment centres and Haematopoietic stem cell transplantation (HSCT) by countries in Africa. Source: Esoh, K., Wonkam-Tingang, E., & Wonkam, A. (2021). Sickle cell disease in sub-Saharan Africa: transferable strategies for prevention and care. The Lancet Haematology, 8(10), e744-e755.

- Recent efforts to initiate pilot projects in sub-Saharan Africa, including new-born screening and comprehensive care, are commendable.
- The challenge is intensified by the absence of new-born screening and ongoing surveillance efforts, leading to unreliable and incomplete data on the SCD prevalence and impact.
- Most national surveys do not include SCD data collection, which hinder the prioritization and resource allocation for the disease. Public health policy support help to reduce the burden of SCD.

Access to and affordability of treatment in Africa

SCD significantly contributed to early death and disease, exacerbated by inadequate investment and efforts to tackle the condition. Despite the urgent need, efforts to manage SCD have been largely neglected in Africa.

Many public health facilities lack basic SCD-friendly services for early diagnosis, disease management and treatment. Improving access to comprehensive health services is essential to reducing the impact of the disease.

- Despite the high prevalence in the region, the lack of skilled staff and resources at primary health care centres limits the effective response to SCD.
- The public health response to SCD in Africa is significantly hindered by the lack of widespread newborn or early screening, leading to many undiagnosed cases and deaths. Patients have limited access to treatment and cannot afford appropriate treatment and care.
- The cost of medication, regular testing and specialized care is particularly unaffordable for those with low income, preventing many from receiving proper management and exacerbating complications.
- Efforts to address these challenges must focus on improving healthcare infrastructure, reducing treatment costs and supporting affected individuals.
- Introducing low-cost SCD diagnostic kits has recently shown promise in field settings across Africa, improving early detection. Meanwhile, countries are moving towards integrating comprehensive care, including potentially curative options such as like Haematopoietic Stem Cell Transplantation (HSCT), into their healthcare systems, aiming for a more holistic approach to managing the disease.

WHO response

- The World Health Organization (WHO) has actively addressed haemoglobin disorders through the adoption of resolutions for SCD and thalassaemia. In May 2006, the WHO focused on sickle cell disease at the 59th World Health Assembly, followed by a resolution on thalassaemia at the 118th WHO Executive Board meeting.
- In addition, in May 2010, the 63rd World Health Assembly adopted a resolution on preventing and managing congenital disabilities, including SCD.
- The PEN Plus regional strategy addresses severe noncommunicable diseases at the first level referral health facilities including SCD as an entry point to PEN Plus. It builds on the World Health Organization's Package of Essential NCD Interventions (WHO PEN) to address the gap for access to care for severe NCDs
- WHO has developed guidance documents for SCD that brings together all relevant materials for the diagnosis and management of sickle cell. The documents are:
    - Guidance Framework for Integrated Sickle Cell Disease Treatment in Sub-Saharan Africa
    - A harmonized guide for the management of the SCD in Africa
- WHO's commitments include raising global awareness, promoting equitable access to health services, supporting countries in disease management and prevention, and supporting research to improve the quality of life for those affected.
- A key initiative launched at the seventy-second session of the WHO Regional Committee for Africa aims to strengthen political, financial, and public support for preventing and controlling SCD throughout the Region. It includes raising disease awareness in schools, communities, and health institutions, advocating for robust health systems that ensure continuous, quality services and equitable access to treatments and innovative tools.

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Sources

The Integrated African Health Observatory supported the production of the factsheets. The graphics are used from the published reports from the references listed above.

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