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SICKLE-CELL DISEASE: A STRATEGY FOR THE WHO AFRICAN REGION

Report of the Regional Director

Executive summary

1. Sickle-cell disease (SCD) is an inherited disorder of haemoglobin. It is the most prevalent genetic disease in the WHO African Region. In many countries, 10%–40% of the population carries the sickle-cell gene resulting in estimated SCD prevalence of at least 2%.

2. The situation in the Region indicates that current national policies and plans are inadequate; appropriate facilities and trained personnel are scarce; and adequate diagnostic tools and treatment are insufficient.

3. Deaths from SCD complications occur mostly in children under five years, adolescents and pregnant women. Strategies and interventions to reduce SCD-related morbidity and mortality should focus on adequate management of these vulnerable groups.

4. This strategy provides a set of public health interventions to reduce the burden of SCD in the African Region through improved awareness, disease prevention and early detection. The interventions include improvements in health-care provision; effective clinical, laboratory, diagnostic and imaging facilities adapted to different levels of the health system; screening of newborns; training of health workers and development of protocols; genetic counselling and testing; accessibility to health care; establishment of patient support groups; advocacy; and research.

5. Success in implementing identified interventions will depend on the commitment of Member States to integrate SCD prevention and control in national health plans, and provide an environment conducive for various stakeholders to contribute to the reduction of SCD prevalence, morbidity and mortality.

6. The Regional Committee is invited to examine and adopt this proposed strategy.

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INTRODUCTION

1. Sickle-cell disease (SCD) is a genetic condition in which the red blood cells contain haemoglobin S (HbS), an abnormal form of the oxygen-carrying protein. Individuals who inherit sickle-cell genes from both parents are homozygotes and develop SCD, while those who inherit the gene from only one parent have the sickle-cell trait (SCT). Those with the trait are carriers, have no symptoms, but can pass the gene on to their offspring.

2. SCD is the most prevalent genetic disease in the African Region.[1] There are different subtypes of SCD in which the abnormal S gene (β^S) coexists with other abnormal haemoglobin genes. Structural studies of the β^S gene suggest that the sickle-cell mutation arose in at least four different places in Africa and a fifth mutation occurred in the Arabian peninsula.[2]

3. The SCT is widespread in the WHO African Region;[3] the β^S gene prevalence in at least 40 countries varies between 2% and 30%, resulting in high SCD-related morbidity and mortality. Deaths from SCD complications occur mostly in children under five years, adolescents and pregnant women.[4]

4. Because there is little genetic counselling available for prospective parents, unions between SCT carriers result in the birth of SCD children. Most countries have inadequate national health policies and plans, and scarce facilities, diagnostic tools, treatment services and trained personnel. There is therefore a need for urgent interventions to address this public health problem.

5. This document provides an overview of SCD in the Region and proposes a strategy for action by Member States and partners. It outlines a set of public health interventions to reduce the burden of the disease through national development or strengthening of policy; early identification; management; and community awareness.

SITUATION ANALYSIS AND JUSTIFICATION

Situation analysis

6. Sickle-cell disease prevalence depends on sickle-cell trait. Where the prevalence of SCT exceeds 20%, SCD is estimated to be at least 2%.[5] The β^S gene concerns the population of at least 40 countries in the Region, and in about 23 countries of west and central Africa the prevalence of SCT varies between 20% and 30%; it is as high as 45% in some secluded areas in western Uganda.[6]

7. Although more than 40 countries are affected, much of the data are still hospital-based and not population-based. Most SCD manifestations are readily amenable to treatment using available interventions; however, the interventions are not accessed by the majority of patients, specifically

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[1] Cook GC, Zumla AI (eds). Manson's tropical diseases, 21st edition. London, WL Saunders, 2003.

[2] Lapouméroulie C et al. A novel sickle gene of yet another origin in Africa: the Cameroon type. American Journal of Human Genetics, 1992, 89:333–37; Cook GC, Zumla AI (eds). Manson's tropical diseases, 21st edition. London, WL Saunders, 2003.

[3] Weatherall DJ et al. Inherited disorders of hemoglobin. In: Disease Control Priorities in Developing Countries. Jamison D et al. New York, Oxford University Press and the World Bank, 2006, pages 663–680.

[4] Dennis-Antwi JA et al. Healthcare provision for sickle cell disease: challenges for the African context. Diversity in Health and Social Care 2008, 5:241–54.

[5] Cook GC, Zumla AI (eds). Manson's tropical diseases, 21st edition. London, WL Saunders, 2003.

[6] Weatherall DJ et al. Inherited disorders of hemoglobin. In: Disease Control Priorities in Developing Countries. Jamison D et al. New York, Oxford University Press and the World Bank, 2006, 663-80; Dennis-Antwi JA et al. Healthcare provision for sickle cell disease: challenges for the African context. Diversity in Health and Social Care 2008, 5:241–54.

the vulnerable groups: children under five years, adolescents and pregnant women. In addition, laboratory facilities for accurate diagnosis are limited.

8. Adequately trained health professionals are few, specialized health care facilities are insufficient and effective medicines, vaccines and safe blood transfusion are very limited. Presently, even in developed countries where stem cell transplantation can be contemplated, there is no widely acceptable public health intervention for the clinical cure of SCD.[7] Consequently, the median survival of SCD patients in Africa is less than five years; about 50%–80% of the estimated 400 000 infants born yearly with SCD in Africa die before the age of five years.[8] The survivors suffer end-organ damage which shortens their lifespan. Thus, to improve management of SCD there is a crucial need for early case identification and implementation of comprehensive health care management (CHCM).

9. Persons with SCD are often stigmatized, and SCD has major socioeconomic implications for affected persons, families, communities and the nation. Recurrent sickle-cell crises interfere with the patient's life, especially regarding education, work and psychosocial development. In the Democratic Republic of Congo, 12% of children hospitalized in paediatric wards have SCD; the estimated annual cost for care is more than US$ 1000 per patient.[9]

10. Despite logistic and economic constraints, neonatal SCD screening along with CHCM have been successfully practised in some parts of Africa. For example, in Benin where neonatal screening and CHCM were practised, the under-five mortality rate of SCD was 15.5 per 10 000, which is ten times lower than the overall under-five mortality rate.[10] These findings are consistent with those from developed countries, demonstrating the benefit of newborn screening and close follow-up of children using CHCM.[11]

11. Research has been done in several countries in the Region to achieve better understanding of SCD,[12] but more remains to be done. The research includes issues related to efficacy of conventional and traditional medicines. The safety, efficacy and quality of some traditional medicines have been evaluated and appeared to be safe and effective in reducing crises associated with severe pain.[13] However there is no substantive documented evidence to support the efficacy of traditional practice or herbal medications in curing SCD.

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7 Rahimy MC et al. Effect of a comprehensive clinical care program on disease course in severely ill children with sickle cell anemia in a sub-Saharan Africa setting. Blood, 2003, 102(3):834–38.

8 Weatherall DJ et al. Inherited disorders of hemoglobin. In: Disease Control Priorities in Developing Countries. Jamison D et al. New York, Oxford University Press and the World Bank, 2006, 663-80.

9 Tshilolo L et al. Neonatal screening for sickle cell anaemia in the Democratic Republic of Congo: experience from a pioneer project on 31204 newborns. Journal of Clinical Pathology, 2009, 62:35–38.

10 Rahimy MC et al. Newborn screening for sickle cell disease in the Republic of Benin. Journal of Clinical Pathology, 2009, 62(1):46–8.

11 Rahimy MC et al. Effect of active prenatal management on pregnancy outcome in sickle cell disease in an African setting. Blood, 2000, 96:1685–89.

12 WHO, Management of Haemoglobin Disorders: report of joint WHO-TIF meeting, Nicosia, Cyprus, 2007; Akinyanju OO. A profile of sickle cell disease in Nigeria. Annals of the New York Academy of Sciences, 1989, 565:126–36; Diagne N et al. Prise en charge de la drépanocytose chez l'enfant en Afrique: expérience de la cohorte de l'Hôpital d'Enfants Albert Royer de Dakar. Med Trop, 2003, 63:513–20; Ogunfowora OB et al. A comparative study of academic achievements of children with sickle cell anemia and their healthy siblings. Journal of the National Medical Association, 2005, 97:405–8; Oshikoya KA et al. Family self medication for children in an urban area of Nigeria. Archives of Disease in Childhood: Paediatric and Perinatal Drug Therapy, 2007, 8:124–30; Tchilolo L. *La drépanocytose en République Démocratique du Congo: apercu sur la situation actuelle et perspectives d'avenir. Congo Médical,* 2003, 3(12):1044–52.

13 Wambebe C et al. Double-blind, placebo-controlled, randomized cross over clinical trial of NIPRISAN in patients with sickle-cell disease. Phytomedicine, 2001, 8(4): 252-61; Sibinga EMS et al. Paediatric patients with sickle cell disease: use of complementary and alternative therapies. Journal of Alternative and Complementary Medicine, 2006, 12:291–98; Oshikoya KA et al. The use of prescribed and non-prescribed drugs in infants in Lagos, Nigeria. Journal of Medical Science, 2008, 8:111–17.

Justification

12. The burden of sickle-cell disease in the African Region is increasing with the increase in population. This has major public health and socioeconomic implications. Despite recent high-level interest in SCD, including commitment from some African First Ladies and the adoption of a UN resolution recognizing SCD as a public health problem,[14] investment in SCD prevention and management using effective primary prevention measures and CHCM remains inadequate.

13. This strategy evolves around existing documents[15] and past achievements in noncommunicable diseases control. The WHO resolution WHA59.20 emphasized the urgency for Member States to design, implement and reinforce in a systematic, equitable and effective manner, comprehensive national, integrated programme for the prevention and management of SCD. The African Union Assembly resolution 1(V) and the United Nations General Assembly resolution 63/237 both recognized SCD as public health problem and urged Member States to raise awareness of SCD. The United Nations Assembly also suggested making June 19 of each year the SCD Day.

14. The SCD strategy for the WHO African Region seeks to increase individual and community awareness about SCD and strengthen primary prevention, reduce disease incidence, morbidity and mortality, and improve quality of life. The Strategy also contributes toward the achievement of the Millennium Development Goals 4 and 5.

THE REGIONAL STRATEGY

Aim, objectives and targets

15. The aim of the strategy is to contribute towards the reduction in incidence, morbidity and mortality due to sickle-cell disease in the African Region.

16. The specific objectives are:

(a) to identify priority interventions for Member States to develop and implement programmes and policies for SCD prevention and control at all levels;
(b) to provide a platform for advocacy to increase resource allocation for prevention and control of SCD through multisectoral collaboration and action;
(c) to establish mechanisms for monitoring, evaluation and research on SCD and apply the findings in policies and programmes.

17. Targets for 2020 are that:

(a) 50% of the 23 Member States with high SCD prevalence should have developed and be implementing a clearly designed national sickle-cell disease control programme within the context of a national health strategic plan;
(b) 25% of the countries in the African Region should have adopted the concept of comprehensive health care management of SCD;
(c) at least 50% of all sickle-cell trait countries will have established a SCD surveillance system with adequately trained staff.

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[14] UN General Assembly. Recognition of sickle-cell anaemia as a public health problem, 2008 (A/RES/63/237).

[15] African Union. Documents Assembly/AU/Dec. 73–90 (V), Assembly/AU/Decl. 1–3 (V) and Assembly/AU/Resolution 1 (V), 2005; WHO. Sickle-cell anaemia, Geneva, World Health Organization, 2006 (Resolution EB117.R3/2006); WHO, Sickle-cell anaemia. Geneva, World Health Organization, 2006 (Resolution WHA/59R20.9).

Guiding principles

18. The guiding principles of this strategy are:

(a) **country ownership, leadership, fairness and community participation** in the implementation of this regional strategy;

(b) **effectiveness, cost-effectiveness and accessibility** of proven interventions and services, especially for the poor and for rural dwellers;

(c) **integrated and evidence-based interventions and prevention-focused population-based approach** for step-by-step implementation of priority interventions as part of the national health plan;

(d) **partnership, team building and coordination** involving all players at various levels; coordination should foster clear definition and understanding of roles, responsibilities and mandates;

(e) **cultural sensitivity, creativity and accountability** involving individuals, patients, civil society and communities in decision-making, planning, implementation and evaluation.

Priority interventions

19. SCD control interventions for Member States of the African Region evolve around Primary Health Care and health promotion approaches to ensure policy development and implementation, legislation and regulation, expansion of primary and secondary prevention. These interventions include:

(a) improvements in health care provision: clinical and laboratory management at all levels of the health system, screening of newborns, training of health professionals, and development of protocols;

(b) genetic counselling and testing;

(c) geographical and financial accessibility to health-care services;

(d) public awareness in schools, communities, health institution, media and associations;

(e) establishment of patient support groups; advocacy; and policies on employment for SCD patients.

The following interventions should be adapted to local settings.

20. **Advocacy for resource mobilization and increased awareness.** Member States should develop and implement effective advocacy interventions to create awareness of SCD and enhance efforts for local and international resource mobilization in order to ensure availability of appropriate infrastructure, equipment, supplies and medicines. WHO and countries should collaborate in developing regional networks and global alliances to help reduce the burden of SCD. High-level advocacy should be explored and encouraged.

21. **Partnerships and social impact.** Partnerships should be fostered between health professionals, parents, patients, relevant community interest groups, nongovernmental organizations (NGOs) and the media. Partnerships will facilitate public education which will increase awareness and encourage screening among members of the community. Partners should support the prioritization of SCD interventions such as widespread provision of screening, laboratory equipment, and specific vaccines that are not part of routine national immunization

programmes; the development of appropriate interventions to strengthen existing health delivery systems; and a multi-disease approach.

22. **Creation or strengthening of national SCD programmes within the framework of noncommunicable disease prevention and control and in harmony with national maternal and child health programmes.** The development of these interventions is the basis for improving the health care of those affected by SCD. Essential areas of work include advocacy; prevention and counselling; early identification and management; data collection, surveillance and research; community education; and partnership. An integrated multisectoral and multidisciplinary team involving health and social workers, teachers, parents and concerned NGOs could be established to work on the practical aspects of care delivery as well as programme implementation and monitoring.

23. **Capacity-building.** Health professionals should be given pre-service and in-service training in SCD control including prevention, diagnosis and management of cases, and complications. The basic requirements to meet these service needs at various levels of the health system should be provided. All members of the health-care team are important for successful programme establishment and implementation.

24. **Supportive activities for special groups–children under five years, adolescents and pregnant women.** Member States should reinforce national SCD supportive activities for vulnerable groups such as children under five years, adolescents and pregnant women who should benefit from financial packages for case management. Other supportive measures include early diagnosis and treatment of complications; special transfusion regimens; surgery; immunization; prophylactic antibiotics, folic acid and antimalarials; and special programmes for prenatal care, psychosocial and professional support to patients, and adaptive educational interventions.

25. **Primary prevention including genetic counselling and testing.** Prevention entails setting up genetic counselling and testing interventions in high prevalence countries to reduce partnering between carriers. Genetic counselling and health promotion activities can lead to substantial reduction in the number of children born with the disease. Widespread community involvement and support are essential.

26. **Early identification and screening.** Ideally, the disease should be identified at birth as part of routine newborn screening or at any subsequent contact the child has with a health facility. Depending on national policy, early identification can be done by universal screening of all newborns, targeted screening of babies born to carrier mothers, and screening of pregnant women. Screening of babies should be done by collecting blood from a heel prick; testing can be done using iso-electric-focusing or high-performance liquid chromatography. Such services should be available alongside counselling and health education since diagnosis raises serious medical, ethical and cultural issues which may differ from one country to another.

27. **Comprehensive health care management for SCD patients of all ages.** CHCM consists of the following: parent and patient education; adequate nutrition; adequate hydration; use of prophylactic antibiotics and antimalarials; folic acid supplementation; use of specific vaccines; continuous medical follow-up; and early detection and management of complications. These measures will reduce morbidity, prevent complications and improve quality of life. In line with the Ouagadougou Declaration, CHCM should also be integrated into health systems using the PHC approach to meet the needs of both rural and urban dwellers, including prevention of complications and patient referral to higher care centres when necessary. Family and community-based care should be integrated into the national programme. Implementation of CHCM requires trained personnel, adequate facilities and interventions adaptable to local needs of communities.

28. **Provision of affordable medicines for SCD management and pain relief.** The use of quality generic medicines as part of the national essential medicines list should be promoted. Subregional economic entities can help in the manufacture and purchase of these medications. Since many SCD patients tend to revert to traditional medicine practices, traditional pharmacopoeias should be fostered after proper testing, validation and standardization. Traditional health practitioners should be involved in SCD management and referral whenever possible.

29. **Strengthening laboratory and diagnostic capacity and supplies with nationwide coverage.** Tools for the diagnosis of SCD should be made available according to their complexity at all levels of the health system starting at primary care level. A system for maintenance and uninterrupted provision of supplies should be developed. Diagnostic and imaging facilities should be made available for early detection of complications.

30. **Initiate and enhance sickle-cell disease surveillance.** Community-based activities including surveillance and supervision, monitoring at all levels of operation, and periodic evaluation at national level should be undertaken to reduce the burden of SCD. The information generated should be disseminated and used as evidence in policy-making as well as in day-to-day decision-making in the management of the programme.

31. **Research promotion.** It is important to describe the history of SCD, its clinical evolution and association with malaria and other diseases. In line with the Algiers Declaration, there is a need to promote innovative research in SCD directed towards basic knowledge and its transformation into new tools such as medicines, vaccines and diagnostic tools; it is also important to identify knowledge gaps and evaluate strategies. It is necessary to promote research in both conventional and traditional medicine to produce evidence of safety, efficacy and quality.

Roles and responsibilities

32. Countries should:

(a) develop, implement and reinforce comprehensive national integrated SCD programmes oriented towards the socioeconomic environment within which the health system operates;
(b) mobilize and allocate resources for SCD programmes;
(c) promote community awareness and involvement in SCD prevention, patient care and support;
(d) integrate SCD surveillance within the national health information system;
(e) improve the knowledge and skills of health and non-health care providers in SCD control;
(f) collaborate with partners to undertake basic and applied research;
(g) support and coordinate national associations working in SCD prevention and control.

33. WHO and partners should support countries by:

(a) mobilizing the international community for prevention and effective management of SCD; and facilitating effective linkage, collaboration and coordination among partners and stakeholders;
(b) advocating for increased resource allocation especially for prevention; provision of adequate infrastructure, equipment and medicines; and research;

(c) providing technical and material support for establishing or strengthening national SCD policies and programmes including monitoring and evaluation;

(d) promoting and supporting partnerships to improve training and expertise of health personnel and to undertake research.

Resource implications

34. The existing level of funding allocated to SCD prevention and control is generally insufficient. Additional internal and external resources will be required to support implementation of this strategy. Specifically there is need to ensure the availability of trained human resources at different levels of the health system along with the provision of medicines and equipment.

Monitoring and evaluation

35. Continuous monitoring and evaluation are crucial to the success of SCD control programmes and should be based on process, outcome and impact measures. These indicators should meet the requirements of national health management systems and be reportable to relevant international forums over the next 5–10 years.

36. Indicators for monitoring progress will include availability and enforcement of SCD control policies, legislation, regulations, programmes and guidelines. Outcome and impact indicators will include reduction of SCD incidence, mortality, morbidity and risk factors; educational achievements; and job security of SCD patients.

CONCLUSION

37. Despite being the most prevalent genetic disease in Africa, sickle-cell disease, along with its serious health and socioeconomic impacts, is largely neglected. SCD ultimately results in multiple organ failure and premature death, occurring mostly in children under five years, adolescents and pregnant women.

38. A comprehensive health care management agenda needs national promoters, committed leadership and effective action at all levels. Strong partnerships must be forged between Member States, WHO and other development partners, communities and individuals.

39. Implementation of the interventions suggested in this strategy would ensure prevention, care and support at all levels and result in improved quality of life and life expectancy of affected individuals. This will contribute towards the achievement of MDGs 4 and 5 and enable affected people to live more productive lives.

40. The Regional Committee is invited to examine and adopt the proposed strategy.