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**SICKLE-CELL DISEASE IN THE AFRICAN REGION:
CURRENT SITUATION AND THE WAY FORWARD**

Report of the Regional Director

EXECUTIVE SUMMARY

1. Sickle-cell disease is a genetic blood disorder that affects the haemoglobin within the red blood cells. The recurrent pain and complications caused by the disease can interfere with many aspects of the patient's life, including education, employment and psychosocial development. Neonatal screening for the sickle-cell trait, when linked to timely diagnostic testing, parental education and comprehensive care, can markedly reduce morbidity and mortality from the disease in infancy and early childhood.
2. In most of the countries where sickle-cell disease is a major public health concern, national programmes for its control do not exist. Basic facilities to manage patients are usually absent, systematic screening for sickle-cell disease is not common practice and the diagnosis of the disease is usually made when a severe complication occurs. As a result, more than 50% of the children with the most severe form of the disease die before the age of five, usually from an infection or severe anaemia.
3. Countries are encouraged to strengthen or set up national programmes which focus on advocacy; prevention and counselling; early detection and treatment; data collection, surveillance and research; and community education and partnership.
4. The Regional Committee is requested to review and adopt the orientations provided in this document.

CONTENTS

	Paragraphs
INTRODUCTION	1–5
SITUATION ANALYSIS	6–12
THE WAY FORWARD.....	13–17
ROLES AND RESPONSIBILITIES	18–19
CONCLUSION.....	20–22

INTRODUCTION

1. Sickle-cell disease is a genetic condition in which the red blood cells contain an abnormal form of the oxygen-carrying protein haemoglobin S. Children who inherit sickle-cell genes from both parents will develop sickle-cell disease, while those who inherit the gene from only one parent will have the sickle-cell trait. Those with the trait have no symptoms but can pass the gene on to their offspring. There are different subtypes of haemoglobin S, and other types of abnormal haemoglobin such as thalassaemia, haemoglobin C and haemoglobin D may coexist with haemoglobin S; hence, clinical presentation depends on the exact combinations inherited.¹

2. Haemoglobin S causes the red cells to become hard, sticky and sickle-shaped, making them fragile and easily destroyed. Unlike normal red cells, which are usually smooth and elastic, sickled cells cannot go through small vessels, thus causing blockage and depriving body organs of blood and oxygen. This results in a chronic slow deterioration of multiple organ systems culminating in recurrent episodes of severe pain, anaemia, serious infections and damage to vital organs. Further complications include stroke, kidney damage and respiratory problems. The term *sickle-cell disease* is preferred because it is more comprehensive than *sickle-cell anaemia*.

3. At its fifty-fifth session in September 2005, the WHO Regional Committee for Africa reviewed the report, *Les premier etats generaux de la drepanocytose*, from a meeting in Brazzaville, Congo in June 2005 attended by experts and first ladies from five African countries. The Regional Committee took cognizance of the meeting's subject and declaration and reaffirmed that sickle-cell disease is an important public health problem which should be discussed again in the near future.

4. The Executive Board, at its one-hundred-and-seventeenth session in January 2006, concerned about the impact of genetic diseases and sickle-cell disease in particular, adopted Resolution EB117.R3. The resolution urges Member States to develop, implement and reinforce comprehensive national integrated programmes for the prevention and management of sickle-cell disease.²

5. This document presents a brief situation analysis, discusses the way forward and suggests what countries and their partners can do to relieve the plight of individuals and communities affected by sickle-cell disease, which remains a grossly neglected health problem.

SITUATION ANALYSIS

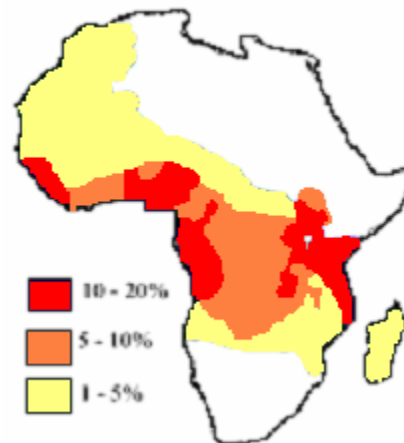
6. The sickle-cell trait is now known to be widespread, reaching its highest prevalence in parts of Africa as well as among people with origins in equatorial Africa, the Mediterranean basin and Saudi Arabia. In Africa, the highest prevalence of sickle-cell trait occurs between latitudes 15° North and 20° South, ranging between 10% and 40% of the population in some areas (*Figure 1*). Prevalence levels decrease to between 1% and 2% in north Africa and to less than 1% in southern Africa. In countries such as Cameroon, Republic of Congo, Gabon, Ghana and Nigeria, the prevalence is

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

² EB 117/34. Sickle-cell anaemia.

between 20% and 30% while in some parts of Uganda it is as high as 45%. In countries where the trait prevalence is above 20% the disease affects about 2% of the population.³

Figure 1: Geographic distribution of sickle-cell trait in Africa



Adapted from: Biogeography and ecology of sickle-cell anemia,
<http://www.ns/c.wustl.edu/sicklecell/prt3/biogeography.html> (last accessed 03-04-2006)

7. The geographic distribution of the sickle-cell trait is very similar to that of malaria. The sickle-cell trait has a partial protective effect against malaria, and this may explain why it has been maintained at such high prevalence levels in tropical Africa.⁴ Those who inherit the gene from both parents do not have this protection. In addition, they suffer from severe effects of sickle-cell disease and many die before they reach reproductive age.

8. In some countries where sickle-cell disease is a major public health concern, control programmes do exist; however, these have neither the national coverage nor basic facilities to manage patients. Systematic screening for sickle-cell disease using a simple blood test is not a common practice, and diagnosis is usually made when a severe complication occurs.

9. Counselling and prevention of causes and infections are simple measures not readily accessible to most patients. As a result, the majority of children with the most severe form of the disease die before the age of five, usually from an infection or severe anaemia. The survivors remain vulnerable to exacerbations of the disease and the complications mentioned above.

10. Sickle-cell disease has major social and economic implications for the affected child as well as the family. Recurrent sickle-cell crises interfere with the patient's life, especially with regard to education, work and psychosocial development.

11. Presently, there is no cure for sickle-cell disease. However, cost-effective treatment exists for the pain and other aspects of the disease. The most important components of this treatment are early intervention with analgesics, antibiotics, rest, good nutrition, folic acid supplementation and high

³ Cook GC, Zumla AI (eds), *Manson's tropical diseases*, 21st edition, London, WL Saunders, 2003.

⁴ Aidoo M et al, Protective effects of the sickle-cell gene against malaria morbidity and mortality, *Lancet* 359: 1311–1312, 2002.

fluid intake. At times, invasive procedures such as blood transfusions and surgery may be needed. Research in some countries in the Region (Benin, Burkina Faso, Nigeria, Togo) has yielded therapeutic agents effective in preventing or reducing the frequency and severity of crises.⁵

12. There is sufficient evidence that neonatal screening for sickle-cell disease, when linked to timely diagnostic testing, parental education and comprehensive care, markedly reduces morbidity and mortality in infancy and early childhood. Nevertheless, simple, inexpensive and cost-effective procedures such as the use of penicillin to prevent infections are not available to most patients.

THE WAY FORWARD

13. **Creation or strengthening of national sickle-cell disease control programmes within the framework of national programmes for prevention and control of noncommunicable diseases** is necessary in affected countries. Essential areas of work should cover advocacy; prevention and counselling; early detection and treatment; data collection, surveillance and research; and community education and partnerships. A multidisciplinary team involving health and social workers, teachers, parents and concerned nongovernmental organizations could be established to work on the practical aspects of implementation and monitoring of the programme.

14. **Prevention** entails setting up sickle-cell screening and genetic counselling programmes in high prevalence countries. Ideally, the disease should be identified during the prenatal period or at birth as part of a routine screening programme. Such services should be available alongside counselling and health education services since diagnosis raises serious ethical and cultural issues which differ from one country to another. Genetic counselling and screening can lead to substantial reduction in the number of children born with the trait.

15. **Management** of sickle-cell disease at different levels of the health-care system should emphasize programmes that use simple, affordable technology and are accessible to a large proportion of the community; such programmes are preferred instead of a parallel system which may be too expensive and unsustainable. The programme should be developed at the primary care level with appropriate technical and patient referral support from higher levels of care. Training of health personnel in prevention, diagnosis and case management should ensure that the health-care system is able to provide the basic requirements of these services. Family and community-based care should be an integral part of the national programme.

16. **Surveillance and research** are important components of the programme. 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. It is also necessary to study the natural history of the disease and its effects on clinical manifestations and transmission of malaria.

17. **Partnerships** should be fostered between health professionals, parents, patients, relevant community interest groups and the media, where appropriate. Partnerships will facilitate public education, identification of genetic risks in the community by recording family disease histories, genetic counselling, awareness and active participation in prevention and care programmes.

⁵ Ekebe GI, Shode FO, Phenylalanine is the predominant antisickling agent in *Cajanus cajan* seed extract, *Planta medica* 56: 41–43, 1990.

ROLES AND RESPONSIBILITIES

Roles of countries

18. Countries should:
- (a) develop, implement and reinforce comprehensive national integrated programmes for the prevention and management of sickle-cell disease;
 - (b) develop capacity to evaluate the situation regarding sickle-cell disease and the impact of national programmes;
 - (c) promote community awareness and involvement in the care and support of persons with sickle-cell disease;
 - (d) collaborate with partners to support basic and applied research on sickle-cell disease;
 - (e) mobilize and allocate appropriate resources for sickle-cell disease prevention and control.

Roles of WHO and partners

19. WHO and partners should:
- (a) increase awareness in the international community of the global burden of sickle-cell disease;
 - (b) provide technical and financial support to national programmes in framing policies and strategies for prevention and management of sickle-cell disease;
 - (c) promote and support partnerships in order to expand the training and expertise of health personnel;
 - (d) support research on sickle-cell disorders in order to increase the life expectancy and quality of life of those affected;
 - (e) create an expert group to advise on various issues concerning sickle-cell disease.

CONCLUSION

20. Sickle-cell disease is the most prevalent genetic disease in the African Region. In spite of the serious impact it has on children, it is still a neglected disease.

21. Although no cure exists for sickle-cell disease, comprehensive programmes could ensure prevention, care and support at all levels and result in both improved quality of life and life expectancy for the patients.

22. The Regional Committee is requested to review and adopt the orientations provided in this document.