



WHO HEREDITARY DISEASES PROGRAMME

PROPOSAL FOR A FEASIBILITY STUDY ON THE CONTROL OF SICKLE CELL DISEASE IN AFRICA

Report of a WHO Informal Consultation

Geneva, 6-7 February 1987



Contents

| | <u>Page</u> |
|---|-------------|
| 1. THE NEED FOR INITIATING THE CONTROL OF SICKLE CELL DISEASE IN AFRICA | 2 |
| 2. RECOMMENDED STRATEGY | 3 |
| 3. SERVICES TO BE PROVIDED BY EACH PROJECT | 3 |
| 4. EDUCATIONAL STRATEGY | 4 |
| 5. ORGANIZATION OF THE PROJECT CENTRES | 5 |
| 6. PRIORITIES | 5 |
| 7. REFERENCES | 5 |
| 8. PARTICIPANTS | 5 |
| ANNEX 1 Clinical manifestations of sickle cell disease | 6 |
| ANNEX 2 Proposed budget for one project spanning 5 years | 7 |
| ANNEX 3 Prospects for the prevention of sickle cell anaemia in Africa | 9 |

This document is not a formal publication of the World Health Organization (WHO), and all rights are reserved by the Organization. The document may, however, be freely reviewed, abstracted, reproduced or translated, in part or in whole, but not for sale or use in conjunction with commercial purposes.

The views expressed in documents by named authors are solely the responsibility of those authors.

Ce document n'est pas une publication officielle de l'Organisation mondiale de la Santé (OMS) et tous les droits y afférents sont réservés par l'Organisation. S'il peut être commenté, résumé ou cité sans aucune restriction, il ne saurait cependant être reproduit ni traduit, partiellement ou en totalité, pour la vente ou à des fins commerciales.

Les opinions exprimées dans les documents par des auteurs cités nommément n'engagent que lesdits auteurs.

1. The need for initiating the control of sickle cell disease in Africa

About 14% of the population of Africa has an abnormal haemoglobin, representing about 65 million people with HbS or HbC.

Each year in Africa about 170,000 children are born with a severe form of sickle cell disease, representing nearly 2% of all births.

Nearly 90% of this problem occurs in the tropical regions of Africa, where up to 30% of the population carry an abnormal haemoglobin.

There is only fragmentary information on the survival of children with haemoglobinopathies in Africa. Certainly a large proportion die in early childhood from infections, including malaria, osteomyelitis and pneumococcal sepsis, and from anaemia. Their infant and childhood mortality is at least 2-3 times that of other children, and sickle cell disease contributes at least 10% of the present early childhood mortality in the whole population. Most of the survivors have life-long clinical manifestations including chronic anaemia, recurrent episodes of pain and other complications, often requiring hospital admission.

There is no known cure for sickle cell disease (except bone-marrow transplantation, which carries an unacceptably high mortality risk). However, regular surveillance and simple protective measures such as regular anti-malarial prophylaxis and folic acid, can greatly improve survival and the quality of life. In addition most complications are susceptible to appropriate medical management, but this often requires specialist services and expertise and prolonged hospitalization. Annex 1 summarizes the commoner complications of sickle cell disease.

Prevention of sickle cell disease is possible. Couples at risk of having affected children can be identified by inexpensive and reliable blood tests and prenatal diagnosis can be reliably performed by chorionic villus sampling (CVS) with DNA analysis, from 9 weeks of pregnancy. Adoption of such measures goes hand in hand with pertinent health education. Experience has clearly shown that discouraging at-risk marriages is ineffective and can be harmful, but that genetic counselling coupled with the offer of prenatal diagnosis can lead to a large-scale reduction in births of affected children. This approach must of course be practised in conformity with the three core principles of medical genetics, i.e., the autonomy of the individual or the couple, their right to adequate and complete information, and the highest standards of confidentiality.

In view of the scale of the public health problem presented by the haemoglobinopathies, a comprehensive approach to control is urgently needed. At present, a large proportion of the African population receives no attention or care for this problem at all. Improved primary health care is improving the survival of affected individuals, and the consequent increased public awareness of the problem, without appropriate education, is leading to stigmatization: so in the absence of any strategy the situation is actively deteriorating. The few centres where facilities for diagnosis and management have existed for a number of years are overburdened with the number of patients requiring regular supervision and periodic treatment. As with all chronic disorders, improved management creates a cumulative demand for more services. It is imperative on the one hand that diagnosis and surveillance be delivered at the community level through the primary health care system; and on the other, that prevention be made available as soon as possible.

While there are many public health needs in Africa, in the case of sickle cell disease the disproportion between the scale of the problem and the small amount that is done about it is extreme.

2. Recommended strategy

To start to at least partially correct the disproportion outlined above, it is necessary to set up a few community-based projects for the control of sickle-cell disease. These should provide education, diagnosis, counselling, surveillance, management and prenatal diagnosis. Such projects will also generate the necessary information on the natural history of the disease in Africa, and the impact of these services upon it.

It is not proposed to build up enough projects to cover each country, but only to introduce them into about four places in different parts of Africa, selected for their diversity. The centres should demonstrate the feasibility of control, develop appropriate technical approaches for Africa, and identify the most efficient ways of introducing these services into the Primary Health Care system. They should therefore provide a model and a guide for the introduction of similar approaches on a wider scale.

The problems associated with sickle cell disease are similar throughout tropical Africa; however, the size and density of the problem in Nigeria together with the existing level of experience indicates that at least one such project should be located there.

A Regional WHO Working Group on sickle cell disease, meeting annually, is an appropriate forum for further developing appropriate approaches, and disseminating them throughout the Region.

In order to sensitize regional policy-makers, a WHO training course on sickle cell disease should be held as soon as possible. The objective would be to demonstrate to senior doctors the social and laboratory requirements for starting the control of sickle-cell disease.

3. Services to be provided by each project

A primary health care unit of about 50,000 people would constitute a suitable study population. Diagnosis of sickle cell haemoglobinopathies in primary health care in the study unit should be seen as part of a wider approach to all forms of anaemia. Facilities should be available on site for accurate determination of:

- Hb concentration
- Packed red cell volume
- Free erythrocyte protoporphyrin assay (for accurate and cheap diagnosis of iron-deficiency)
- Red cell indices, ie, MCV and MCH (desirable but not essential)
- Sickling or solubility screening test
- Haemoglobin electrophoresis

Chorionic villus sampling (CVS) under ultrasound guidance, should be available at an obstetric unit within a reasonable geographical distance.

One DNA diagnostic laboratory could service several projects, and also serve self-referred couples at risk.

The diagnostic strategy will be directed towards the following target groups in the study population:

- (a) Individuals with symptoms suggestive of sickle cell disease (see Annex 1).
- (b) Children less than four years old. This group should eventually involve most infants at or soon after birth.

- (c) Pregnant women as early as possible in pregnancy. There are two objectives for this target group. (i) to identify those with sickle cell disease, who will require special management; (ii) to identify prospectively those at risk for bearing a child with sickle cell disease. Women who are found to have HbAS by electrophoresis will be asked to bring their husband for testing. Cultural and other factors may influence the willingness of husbands to be tested, so the acceptability of testing is itself a worth-while subject of study.

At-risk couples identified in the above manner, together with those who have previously had children with sickle cell disease, will be fully informed and offered prenatal diagnosis. The use they make of it will be monitored, to serve as a guide for future policy.

Screening of pregnant women is recommended as the initial approach for genetic counselling in the study population, since it may be expected to provide the maximum benefit with the minimum risk of stigmatisation or other undesirable consequences. As the knowledge base of the community expands, carrier detection and genetic counselling will be increasingly requested in other situations, and these services should be readily available.

4. Educational Strategy

Success in management and prevention of the haemoglobinopathies depends equally on educational and technical strategies. Educational programmes need to be directed to the following target groups:

- (a) 'Decision-makers' responsible for health and public policy.
- (b) Health professionals: doctors and nurses should have teaching on the inheritance and clinical management of, and counselling for sickle cell disease included in their curriculum. Technicians need training in performing the relevant laboratory tests accurately, and interpreting the results. Counsellors at all levels of the health care system will require training to convey genetic information correctly and sensitively, and in the clinical manifestations and management of sickle cell disease.
- (c) The general public: the ultimate success of the project will hinge on the general level of knowledge about sickle cell disease, and this can only be built up slowly. Therefore, educational resources should first be concentrated on the section of the community where they will have most impact, ie, on those of, or approaching, reproductive age. A particularly appropriate and effective approach may be to incorporate teaching about sickle cell disease in the school curriculum at the secondary school level.

Initially, such educational enterprises should be limited to the project area, as it is a mistake for public education to outstrip the availability of services, since this creates demand that cannot be met.

The organization of educational programmes and materials will be an important function of the recommended Regional WHO Working Group.

Community involvement is a powerful educational force through the formation of "Sickle Cell Associations" of parents and patients to support affected families and provide information to the community. The emergence of such support organizations will be encouraged by each project.

5. Organization of the Project Centres

The proposed study-population of approximately 50,000, would be expected to produce approximately 2,500 births a year: approximately 200 pregnancies a year would be at risk of sickle cell disease, resulting in the birth of approximately 50 affected infants per year. Approximately 10,000 diagnostic blood tests will be required per year.

To deliver diagnosis and counselling to this population, a project centre will be needed with the following: 2 laboratory staff, 2 nurse-counsellors, 2 drivers plus 2 vans, and one medically-qualified project director.

The project director will co-ordinate the work of the staff, maintain productivity and standards, train counsellors, improve health education, and deal with the clinical and diagnostic problems that arise. He/she will also be responsible for analyzing and presenting research data. The nurses/counsellors will be responsible for surveillance of patients and counselling for their families and for couples at risk, and will provide psychological support. The proposed budget which is required for the implementation of the said projects is presented in Annex 2.

6. Priorities

- (a) The initiation of a newborn screening programme with follow-up on patients' progress, the establishment of one of the four projects, and of one of the two DNA diagnosis laboratories.
- (b) The formation of a working group on control of sickle cell disease in Africa.
- (c) The organization of a training course on prevention and management of sickle cell disease in Africa.

7. References

The figures quoted in the report are based on:

- (a) Update of the Progress of Haemoglobinopathies Control: Report of a WHO Working Group 1984 and 1985. Unpublished WHO Document (HMG/WG/85.8).
- (b) Community Control of Hereditary Anaemias: Memorandum from a WHO meeting: Bulletin of the WHO 1983 61: 63-80.
- (c) Sickle Cell Disease: Graham R. Serjeant, Oxford Medical Publications, Oxford University Press: 1985.

8. Participants

Dr O. Akinyanju, Department of Medicine, University of Lagos, Nigeria: Chairman

Dr G. Honig, Sickle Cell Centre, University of Illinois, Chicago, USA

Dr L. Luzzatto, Royal Postgraduate Medical School, London, UK

Dr B. Modell, Department of Obstetrics and Gynaecology, University College London, UK: Rapporteur

Dr M. Rabb, Sickle Cell Centre, University of Illinois, Chicago, USA

WHO Secretariat

Dr V. Bulyzhenkov, Responsible Officer, Hereditary Diseases Programme, Division of Noncommunicable Diseases, WHO HQ, Geneva: Secretary

ANNEX 1

Clinical Manifestations of Sickle Cell disease

The following problems should alert primary health care workers to the possibility of sickle cell disease.

1. Anaemia with or without splenomegaly: in almost all patients with SS disease and many with SC disease.
2. Hand-foot syndrome (dactylitis): occurs from 6 months to 4 years of age: it consists of pain and swelling in various areas of the extremities.
3. Painful crisis: episodes of severe debilitating pain that lasts for a few days on average.
4. Priapism: frequent in pre-adolescent and mature adults due to chronic vaso-occlusion.
5. Leg ulcers: frequent in young adults.
6. Delayed growth and development: particularly common around the age of puberty, usually compensated later.
7. Bone and joint pains ("rheumatism"): especially if recurrent.
8. Unexplained deterioration of vision: may be due to proliferative sickle cell retinopathy.
9. Neurological abnormalities (stroke): particularly in children.
10. Jaundice

ANNEX 2

PROPOSED BUDGET FOR ONE PROJECT SPANNING 5 YEARS (US DOLLARS)

| | <u>Per Year</u> | <u>Per 5 Years</u> |
|--|-----------------|--------------------|
| <u>Staff</u> | | |
| 1 physician (project director) | 20,000 | 100,000 |
| 2 nurse-counsellors | 10,000 | 50,000 |
| 2 laboratory technicians | 10,000 | 50,000 |
| 1 half-time obstetrician | 10,000 | 50,000 |
| <u>Equipment</u> | | |
| 1 microscope | 500 | 20,000 |
| 4 electrophoresis tanks | 4,000 | |
| 2 power packs | | |
| 1 automated blood counter | 15,000 | |
| 1 microhaematocrit-centrifuge | 500 | |
| consumables | 5,000 | 25,000 |
| educational materials | | 15,000 |
| 2 motor vehicles (15,000 each) | | 30,000 |
| 2 drivers (2,500 each per year) | 5,000 | 25,000 |
| maintenance costs of vehicles (3,000 each) | 6,000 | 30,000 |
| ultrasound equipment + personal computer | | <u>200,000</u> |
| TOTAL | | 595,000 |

PROPOSED BUDGET FOR DNA LABORATORY SPANNING 5 YEARS (US DOLLARS)

| | <u>Per Year</u> | <u>Per 5 Years</u> |
|-------------------|-----------------|--------------------|
| <u>Staff</u> | | |
| 1 scientist | 20,000 | 100,000 |
| 1 technician | 10,000 | 50,000 |
| <u>Equipment</u> | | |
| 1 ultracentrifuge | | 50,000 |
| others | | 10,000 |
| consumables | 10,000 | 50,000 |
| maintenance | | <u>10,000</u> |
| TOTAL | | 270,000 |

TOTAL ESTIMATED COSTS SPANNING 5 YEARS (US DOLLARS)

| | |
|--|-----------|
| Four sickle cell control study projects | 2,380,000 |
| Two DNA laboratories | 540,000 |
| Working Group (30,000 x 5 : 1 per year) | 150,000 |
| Training Course (25,000 x 2 : 2 per 5 years) | 50,000 |

ANNEX 3

PROSPECTS FOR THE PREVENTION OF SICKLE CELL ANAEMIA IN AFRICA

BY

OLU AKINYANJU, MD, FRCP
Professor of Medicine
College of Medicine
University of Lagos
P.M.B. 12003
Lagos
Nigeria

ABSTRACTS

The main affected areas of Africa comprise about 34 sub-Sahara countries with a present total population of 370 million expected to rise to 590 million by 2,000 A.D. Heterozygote rates vary widely but may average about 20%. The high birth rate, high child mortality, high unemployment rate and poor economies deny most of the population acceptable standards of living and most children with sickle cell anaemia (SCA) a fair chance of survival. This makes survival rather than SCA prevention the major preoccupation. Improvement in general education, nutrition, population and disease control, would appear to be the logical first essential strategy for these populations. Among the large and growing minority with better standards of living and SCA survival in these countries, prevention of SCA is demanded and is desirable in order to limit the burden of SCA on affected families and prevent the undesirable stigmatization of the heterozygous state which would be an inevitable consequence of not establishing credible education, counselling therapeutic and prevention programmes. There is a need for the development of appropriate control measures, as the WHO strategy for the prevention of haemoglobinopathies, which has been immensely successfully applied to the prevention of thalassaemia in Mediterranean countries, is unlikely to have nearly the same impact on sickle cell anaemia in Africa. Given the present state of knowledge, the prospects of reducing the prevalence of children born with SCA but not of preventing all such births, are quite good on a long term basis. Therefore, the major goal would seem to be to improve the management of children with SCA so much so that parents can perceive their upbringing as a worthwhile alternative to abortion.

INTRODUCTION

It is heartening to learn that the strategy evolved by the WHO for the control of haemoglobinopathies has shown commendable success in the reduction of births of children with beta thalassaemia major in the United Kingdom and in some Mediterranean countries such as Greece, Italy (Ferrara district), Sardinia and Cyprus¹. The difficulties that remain appear to relate mainly to those of logistics in applying the techniques to the larger communities of the United Kingdom, Greece and Sardinia. The strategy used included community education, heterozygote screening, genetic counselling and fetal diagnosis with optimal termination of affected pregnancies. It is the frequent exercise of this option by couples-at-risk which has led to the desired result. An essential component of the strategy is the technical feasibility and accuracy of the method of fetal diagnosis, which is in fact simpler and more reliable for sickle cell than for thalassaemia. Having successfully tested a viable strategy for the control of thalassaemia, it is only right that the WHO should now turn its attention to the problem of sickle cell anaemia (SCA) which is most severe and prevalent on the continent of Africa, especially in those areas lying between the two large deserts - the Sahara to the North and the Kalahari to the South. This paper will therefore examine the prospects for successful control of sickle cell anaemia in these areas of Africa based largely on the author's more intimate knowledge of Nigeria and on the strategy already evolved for the WHO control programme for haemoglobinopathies.

THE SIZE OF THE PROBLEM

The area of Africa under consideration comprises about 34 countries listed in Table I and covering an area of about 19 million square kilometres². Economically, most of the countries are rated as low-income and the rest as low middle-income and which are placed below upper middle-income countries such as Brazil, Greece and Portugal². The total population of these countries in mid 1985 was estimated as 342 million, widely dispersed in remote villages but with increasing concentration in urban centres. The population has a high growth rate of over 3% per annum and is expected to exceed 550 million by the year 2,000 A.D. The population profile shows a pyramidal age structure with many young dependants per working adult².

Polygamous marriages, which in previous ages served to provide a respectable estate for the excess of women in the population and extra hands on the farms, are still widely practised in many areas. Infant and child death rates are high and access to modern health services are generally poor, with low doctor or nurse to population ratios. Recent shortages in drugs and other supplies due to economic difficulties have further limited the benefit to

be derived from medical consultations when these occur. Survival is the major pre-occupation of people in many areas and there is a high rate of illiteracy with limited access to education for all. Communication is generally difficult and superstitious beliefs and remedies for many diseases are widely prevalent. While it is still not easy to obtain accurate basic information, we can surmise that the situation is better than it was 20 years ago as reflected by the significant reduction in infant and child mortality rates since 1965². However, there is also a growing middle class of people in each country at a rate which varies widely from country to country.

The prevalence of the sickle cell gene over the entire area is not accurately known, but an average of 20% is probably conservative. In Nigeria about 25% of the population are heterozygotes^{3,4} and in parts of East Africa heterozygote rates as high as 45% have been reported⁵. These rates vary even within a country because of the heterogeneous derivation of its people. It is important to realize that unchecked falciparum malaria infection, which originally gave a survival advantage to persons with sickle cell trait (Hb AS), is still prevalent in many rural areas of Africa, where the proportion of heterozygotes could still be rising. We are therefore about to consider the control of sickle cell anaemia on the continent in which there are, at present, at least 68 million heterozygotes, which figure will probably rise to over 100 million by the year 2,000 A.D.

AWARENESS AND UNDERSTANDING OF SICKLE CELL ANAEMIA IN AFRICA

Despite the high prevalence of the Hb S gene, the level of awareness of SCA is very low. Actual figures are hard to come by but the clear impression of workers in Uganda⁶ as in Nigeria is that awareness is indeed low. The probable reasons for this are not hard to deduce.

Sicklers (sickle cell anaemia patients) were virtually unknown until recently. They simply did not survive to be recognized, until the fairly recent improvement in infant and childhood mortality rates. This improvement is of course most marked in families of the educated or affluent elite who are mostly urbanized. The picture is different with poor rural dwellers. As recently as 1979, it was found that although 2% of all newborn in a fairly remote Nigerian village had sickle cell anaemia, no survivor could be found among the nine year old children in that village⁷. The poor survival of sicklers was buried within the generally poor childhood survival which can be attributed to infections. It is only where the general childhood mortality rates have fallen drastically that the sickler would stand out as being excessively disadvantaged. The level of understanding of sickle cell disorders, even among those aware, is also poor. That the disease is hereditary, is not at all widely known even by many sicklers. The perception of control of sickle cell anaemia even amongst many highly placed medical doctors is an off-the-cuff one of 'why don't we enforce, or at least encourage the non-coupling of heterozygote partners'. That the implementation of such a policy would lead to severe stigmatization of carriers, especially the females, is not so readily appreciated.

COMMUNITY EDUCATION AND GENETIC COUNSELLING

There are no established effective programmes for education of the public about sickle cell disorders in Nigeria and I suspect in most other African countries. Health educational efforts have rightly concentrated on better sanitation, better nutrition and infection control. The effect of these efforts has of course rebounded beneficially on the survival and well-being of children with SCA. The lack of broad based genetic services or more specialized sickle cell services is now being acutely felt in the urban areas where the greatest survival of children with SCA is occurring. The absence of these services has meant that the needs of the populace for appropriate information is largely unmet. Misinformation and myths therefore abound which further confuse many patients and parents. In order to remedy this situation a non-governmental parent/patient group known as The Sickle Cell Club of Nigeria was formed in 1974 with branches in many towns. Since 1984, the Sickle Cell Club in Lagos has become reasonably active and vocal in enhancing awareness and understanding, and organized the first ever training course for 36 counsellors in August 1986. This as far as can be ascertained was the first ever training course for counsellors in Africa. Trainees included doctors, nurses, medical social workers and teachers, who, by the end of the course, recognized the need for a similar course to be incorporated into the curricula of their own training institutions.

DIAGNOSTIC SERVICES

Diagnostic services are few. They can be obtained from the teaching hospitals and a few of the general hospitals, and some private laboratories. The larger institutions are increasingly unable to perform creditably or at all because of various financial, technical and managerial constraints. The performance of private laboratories is not subjected to any scrutiny or quality control and they are, in any case, too few and too expensive to become effective tools for most diagnosis or population screening. The need for ready access to accurate diagnosis became only too obvious at the end of the recent counsellors' training course in Lagos when the trainees realized its importance as a prerequisite for any counselling session.

PARENT/PATIENT SUPPORT GROUPS

There are very few of these groups and their dormancy rate is very high. The indication, based on our experience with an active group in Lagos, is that their continued existence and value can best be sustained by the parallel establishment of better diagnostic, educational, therapeutic and control services. Otherwise affected families soon lose interest in their activities.

There is nevertheless a rising need for these groups in areas where better survival is increasing the numbers of sicklers. They can serve to enhance awareness and understanding, mobilize public support and action, especially in many African countries where most families can boast of a sickler in the nucleus or extended family.

THE DEMAND FOR PREVENTION

The demand for prevention definitely exists and is strong among the minority who are enlightened about or aware of the disease and have affected children or relatives. This minority is, however, constantly growing but its size is not known. Their demand is largely unmet which is why affluent members of this minority fly off to Europe or America for treatment, counselling and antenatal diagnosis. Most of the demand is coming from couples who discover their risk because of bearing an affected child. Most couples-at-risk have probably not borne affected children and are ignorant of their risk and have therefore not demanded any services. Our recent experience with enhancing awareness is that it can, by creating demands for services that cannot be met, lead to great dissatisfaction and frustration. Having created awareness we have no demonstrable reply to newspaper editorials and to doctors who, unwisely, in our view, advocate the non-coupling of partners with the trait as a means of preventing the disease and, according to them, saves the country money that would otherwise be spent in providing services to sicklers. The need for better services for SCA is therefore most urgent in Africa.

A REALISTIC APPROACH TO THE CONTROL OF SCA IN AFRICA

It should now be obvious that numerous factors peculiar to the African continent, and to individual countries within it, would dictate that appropriate control programmes and technology are evolved to meet the various needs. An obvious need is for better information about prevalence, awareness, understanding, attitudes and health care facilities in the various countries. The size of the present and potential demand for prevention services should also be determined.

Key collaborators with demonstrable interest and energy should be identified in each country, to form the nucleus of an African Sickle Cell Control Group.

Educational seminars should be held for top health policy officials in each country in order to sensitize them to the prospects and implications of control of SCA and to the need for the establishment of sickle cell centres for research and education and for better professional and public education on genetic disorders in general.

Realistic programmes should be prepared based on the prevailing circumstances, and health professions in each country should be fully informed and educated about them. Short training courses in diagnosis and counselling techniques should be taught and introduced into the appropriate levels of the host country's health care system.

Mass education of the public should not precede the establishment of facilities for coping with the consequent demands for services. On the other hand, facilities for rendering a credible service to those who already demand it should be created so as to build up public confidence. To this end fetal diagnosis should be introduced in some centres for the benefit of those who require it. This is the only way to prevent a growing outcry for enforcing or discouraging non-coupling of heterozygotes and consequent stigmatization of the disease. The control of population growth and of communicable diseases should be actively supported and promoted as desirable strategies towards the provision of adequate services for the care and control of SCA. This is in fact a priority for the segment of the population who are poor, unaware of SCA and in any case preoccupied with survival.

CONCLUSIONS

The reduction of sickle cell anaemia births in Africa is desirable as a means of reducing its burden on affected families and meeting the demand by enlightened families who may already have at least one sickler in the family. It will also aid the fight against widespread and catastrophic stigmatization of the disorder. However, numerous problems associated with the developing countries make it necessary to carefully work out appropriate approaches to the problem which should include improvement of the general health and education of the people. While the application of present day technology can assist greatly in reducing the prevalence of SCA, a complete prevention of new births in Africa is hardly attainable in view of the large numbers involved, the poverty, and the likelihood that a significant proportion of parents even if informed might elect to proceed with affected pregnancies.

The financial, technological and human resources required, suggest that the strategy evolved by the WHO for the control of haemoglobinopathies would have less impact on prevention in Africa, than in Europe where the larger communities already show a much slower progress towards a zero affected birth rate. Perhaps it should be remembered that thalassaemia major is far more severe than sickle cell anaemia and some effort should be directed towards educating people about the fact that many sicklers lead meaningful and productive lives and that better control of crises and infection is a viable alternative to outright prevention. Needless to say, some of the attitudes about enforced prevention (not held by sicklers themselves) have been encouraged by the hopelessness created by the lack of appropriate health care facilities in most of Africa. Rectification of this state of affairs is the most urgent requirement.

REFERENCES

1. WHO 1984. Progress in Community Control of Thalassaemia. Report of the Second Annual Meeting of the WHO Working Group on the Community Control of Hereditary Anaemias. Unpublished WHO Document HMG/WG/83.9.
2. Population Growth and Policies in Sub-Saharan Africa. A World Bank Policy Study. Washington D.C., USA, August 1986.
3. Edington, G.M. (1959). Annual Report of the Department of Pathology. University College, Ibadan, Nigeria.
4. Gilles, H.M. (1964). Akufo: An Environmental Study of a Nigerian village. University of Ibadan Press, Ibadan, Nigeria.
5. Ssebabi, E.C.T., and Buluwa, F.M. (1977). Sickle Cell Trait Anaemia in Pregnancy. East African Medical Journal, vol. 4, 258-265.
6. Ndugwa, C.M. (1982). Sickle Cell Anaemia in Pregnancy - Uganda (1971-1980). East African Medical Journal, vol. 59, 320-326.
7. Fleming, A.F., Storey, J., Molineaux, L., Iroko, E.A., and Attai, E.D.E. (1979). Abnormal Haemoglobins in the Sudan Savanna of Nigeria, I. Prevalence of haemoglobins and relationships between sickle cell trait, malaria and survival. Ann. Trop. Med. Parasitol. vol. 73, 161-172.

TABLE I

HEALTH RELATED DATA IN 34 SUB-SAHARAN AFRICAN COUNTRIES

| COUNTRY | POPULATION (MILLIONS) | | GNP (DOLLARS) PER CAPITA | INFANT MORTALITY RATE < 1 YR | CHILD DEATHS 1 - 4 YRS |
|----------------------|--------------------------|-------|-----------------------------|------------------------------------|------------------------------|
| | 1985 | 2000 | 1983 | 1983 | 1983 |
| ANGOLA | 8.6 | 13.2 | NA | 148 | 31 |
| BENIN | 4.0 | 6.5 | 290 | 148 | 31 |
| BURKINA FASO | 6.7 | 9.2 | 180 | 148 | 31 |
| BURUNDI | 4.7 | 7.4 | 240 | 123 | 25 |
| CAMEROON | 10.2 | 16.7 | 820 | 116 | 19 |
| CENTRAL AFRICAN REP. | 2.6 | 4.0 | 280 | 142 | 29 |
| CHAD | 5.0 | 7.3 | NA | 142 | 29 |
| CONGO | 1.9 | 3.4 | 1230 | 82 | 8 |
| COTE D'IVOIRE | 10.3 | 17.3 | 710 | 121 | 20 |
| EQUATORIAL GUINEA | 0.4 | 0.5 | NA | 136 | 29 |
| GABON | 0.8 | 1.2 | 3950 | 111 | 22 |
| GAMBIA | 0.7 | 1.1 | 290 | 200 | 46 |
| GHANA | 14.0 | 23.1 | 310 | 97 | 12 |
| GUINEA | 6.0 | 8.3 | 300 | 158 | 36 |
| GUINEA-BISSAU | 0.9 | 1.2 | 180 | 158 | 36 |
| KENYA | 20.6 | 36.5 | 340 | 81 | 14 |
| LIBERIA | 2.2 | 3.5 | 480 | 111 | 17 |
| MADAGASCAR | 10.1 | 16.9 | 310 | 66 | 10 |
| MALAWI | 7.0 | 11.4 | 210 | 164 | 38 |
| MALI | 7.5 | 11.2 | 160 | 148 | 31 |
| MOZAMBIQUE | 13.9 | 21.7 | NA | 109 | 16 |
| NIGER | 6.5 | 10.5 | 240 | 139 | 28 |
| NIGERIA | 99.7 | 16.3 | 770 | 113 | 17 |
| RWANDA | 6.0 | 10.2 | 270 | 125 | 26 |
| SAO TOME & PRINCIPE | 0.11 | 0.16 | 310 | 62 | NA |
| SENEGAL | 6.6 | 10.1 | 440 | 140 | 28 |
| SIERRA LEONE | 3.8 | 5.4 | 330 | 198 | 54 |
| SOMALIA | 5.4 | 8.5 | 250 | 142 | 30 |
| SUDAN | 21.7 | 33.4 | 400 | 117 | 19 |
| TANZANIA | 22.2 | 36.9 | 240 | 97 | 18 |
| TOGO | 3.0 | 4.9 | 280 | 112 | 17 |
| UGANDA | 14.9 | 24.6 | 220 | 108 | 21 |
| ZAIRE | 31.6 | 49.9 | 170 | 106 | 20 |
| ZAMBIA | 6.7 | 11.0 | 580 | 100 | 19 |
| TOTAL OR AVERAGE | 366.3 | 590.1 | 310 | 125 | 25 |