The Foundation for Research and Education in Sickle Cell Disease

A Prospectus

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I. WHAT IS THE PROBLEM OF SICKLE CELL DISEASE?

SICKLE cell disease is an non-contagious inherited disorder which occurs in the severe form in one out of every 400 black Americans, and in the mild form, or trait, in one out of every 10 black Americans. It is a disease of the blood in which there is a particular kind of hemoglobin in the red cells which causes them in certain specific circumstances to assume a peculiar crescent or sickled shape.

The disease originated in Africa, where its prevalence is even higher in many tribes, particularly in those areas where malaria is endemic. The malaria parasite shuns blood which sickles, and it is this property, the greater ability of individuals with sickle cell disease to survive in these regions of endemic malaria, which probably accounts for its high incidence. It has been disseminated, however, over all those parts of the world where people of African origin have migrated and intermarried. Sickle cell disease is now found in certain parts of Africa, in the Mediterranean countries, India and Pakistan, in the Carribbean and in all parts of South, Central and North America.

The nature of the disease, with its recurrent, painful crises and numerous complications, beginning early in childhood and continuing through adulthood causes profound effects on the individual, on the family and even on society as a whole.

If two people who have the sickle cell trait happen to mate, the chances are that one out of four of their children will have the severe form of sickle cell anemia, two out of four will have the trait and one out of four will have no form of the disease. If one parent has sickle cell anemia, and the other parent the trait, half the offspring will have the trait, the other half the severe form. There are other combinations which give rise to the intermediate forms of the illness.

One chronically ill child in a family, regardless of its economic circumstances, can be devastating. The existence of more than one, or even the threat of its recurrence can be overwhelming. From earliest infancy there is the constant medical problem with suffering and recurrent expense. In the case of school age children constant absences from school, which this illness entails, in addition to the frequent bouts of pain and chronic fatigue, are a severe handicap to school achievement. Ordinary physical activity is limited, competitive sports are not desireable, and the entire educational program selection must be geared to the individual's physical capabilities.

The choice of a vocation for the individual with sickle cell disease should be made with care; certain activities are inadvisable. Individuals with the severe form of sickle cell anemia are limited to sedentary occupations. Employers of patients with sickle cell disease must have an understanding of the illness, and must be willing to accept an employee who may have to lose time from work.

A man with sickle cell anemia may find it difficult to hold a job and to support his family. A woman with sickle cell anemia superimposes a chronic illness on the problem of homemaking. She may have severe difficulty with all phases of her pregnancy and delivery. The life span of individuals with sickle cell anemia is limited—usually they do not survive beyond their forties.

Many medical practitioners are unaware of the disease. Even when it is known, it may not be recognized. Many patients with sickle cell disease are treated for diseases which have similar symp-

toms. Even when detected, some physicians only prescribe to relieve the symptoms of this blood disorder, as one might recommend aspirin for the relief of a headache or rest for a cold. There is no known medical cure for sickle cell anemia.

II. WHAT IS THE FOUNDATION FOR RESEARCH AND EDUCATION IN SICKLE CELL DISEASE?

Some time ago a group of individuals in New York City working with both the research and clinical aspects of this illness met to discuss better methods of combating this disease. This group has now been chartered as the Foundation for Research and Education in Sickle Cell Disease. It is composed of individuals whose professional activities in medicine, education, law and religion are directly concerned with this severe problem of the black community. The Foundation is a charitable, non-profit, tax-exempt organization. The public at large, is unaware of the magnitude of sickle cell disease. The Foundation was formed to overcome this serious gap.[1]

Before a cure is found, certain measures must be taken, i.e. research must be encouraged and supported, the procedures of diagnosis and treatment must be improved, the public must be educated, the medical personnel who handle the problem must be educated and families with this problem must be assisted. Certain specific things need to be done. Members of the medical and allied professions need to include diagnostic tests for sickle cell disease as a regular part of diagnostic procedures. Premarital genetic counseling is greatly needed.

The ultimate goals of the Foundation for Research and Education in Sickle Cell Disease are to coordinate local activities with a national program and to provide for research on all levels.

III. WHAT NEEDS TO BE DONE

As previously stated, the life expectancy of a patient with the severe form of sickle cell disease is less than 40 years. These years are characterized by frequent hospitalizations, restricted activities, loss of school and work time and constant pain and fatigue. If clinical care can be impoved, the frequency and severity of complications can be reduced. Improvement in medical procedures will enable the patients to live a fuller and probably longer life. It is to this end that the Foundation wishes to make known the following basic needs:

1. Research Programs

a) Universal Testing[2]—There is a great need for universal testing of all black individuals, as well as all individuals from other areas of high incidence. The Foundation believes that any black person should know whether they carry the sickling condition or not, and if so, in what form, and all health agencies should know the incidence and forms of this disease in the population they serve.

b) Basic Research—The phenomenon of sickling is only partially understood and the dynamics of the anemia are only partially understood. Much basic hematological research is required here. The genetic factors involved and the means required to influence the pathways of gene combinations or to suppress or enhance gene activity are very fruitful areas to be explored. Support of this type of research with specific reference to sickle cell disease would be of great significance in many other inherited disorders.

c) Clinical Research—Many clinical factors are not known about the disease at present, such as what is the influence of geography and what precipitates crises. The question of therapeutics needs further exploration. Better methods of handling complications is not yet well worked out.

A list of the effects of specific drugs on the illness needs to be compiled. The proper methods of handling transfusions in sickle cell patients is yet to be fully determined. All these techniques, once established, can have broader applications to other diseases.

2. Education

Education of many different groups and individuals is a very definite and important need in the fight against sickle cell anemia.

a) Medical Personnel—Not all physicians are familiar with sickle cell disease, either because they were educated prior to the time when the disease was more widely known in medical circles, or because they see relatively few black patients in their practices. Needed is more information on sickle cell anemia at physicians' conventions, dissemination of literature, composition and distribution of a practical office sickle cell testing kit. Nurses, laboratory technicians, physiotherapists and all paramedical personnel need training and information.

b) Other specific occupational groups which an educational program should reach are:

Teachers who should know what to expect if there is a child with sickle cell anemia in the class.

Guidance and Vocational Counsellors who need to know how to direct sicklers into suitable environments.

Psychologists who may have to handle the many mental hygiene problems of sicklers.

c) Much educational information has to be directed to affected families.

d) The population at large, as an end in itself, requires information about this disease to aid in case finding and acceptance and understanding of the affected person and family.

3. Direct Services to Patients

The care of patients with sickle cell disease is scattered, haphazard and geared to emergency care only, i.e., it ignores comprehensive. continual, preventative care, the type of care that 1) could keep patients with sickle cell disease at their optimum mental and physical condition, 2) reduce to a minimum the crises and complications, and 3) enable them to pursue their education. earn a living, and rear their families.

There needs to be a major improvement in the nature of the medical services provided to people with sickle cell disease. Although individuals suffering from the illness can be, and are, treated in hematology clinics in many hospitals and in the offices of individual physicians, the availability to the sickle sufferer of special centers devoted to sickle cell disease has definite advantages to the individual patient in time of concentration of effect, and availability of the latest knowledge, as well as an overall advantage to all sickle sufferers. It is from centers such as this that major breakthroughs are most likely to come.

Identification of the extent of the disease, in any given population, and identification of the people involved, so that they may receive care, is an important direct service.

The Foundation has already begun free sickle cell testing from its offices. It envisages large programs in appropriate school districts, for the identification of children involved, so that their health and educational problems may be recognized and treated properly.

The Board of Education District Four, New York City, in which the Foundation has its offices, has provided for just such a program involving 20,000 children. The Foundation is assisting in this program. It has requests for funding and assistance from several other districts in the metropolitan area, East Harlem, Red Hook and Southeast Queens. Several groups in Nassau and Suffolk counties have also shown interest in this program.

Patients should have ready access to the latest in medical equipment. Sickle cell disease is one of the most protean diseases and may require much specialized equipment and the services of many individual specialists. There are also needs beyond the physical existence of the clinics, and the physicians who staff them. With today's acute shortage of medical personnel, training must be devised and administered to paramedical personnel for all phases of any medical program; from case finding, which includes outreach into the community (best done by indigenous personnel) through education, appointment making, coordination and follow-up.

Coordination and development of auxiliary services must be undertaken. A well organized blood program is a major immediate requirement. Such a program can also serve to move the black community into the mainstream of already organized efforts to provide blood service for diversified needs.

IV. WHAT THE FOUNDATION HAS DONE SO FAR

1. Since its inception, the Foundation has maintained a Speaker's Bureau, whereby members of the Foundation, upon request, have spoken to civic, community, social and other organizations in the greater New York area, i.e., such as Rotaries, student groups, youth organizations, PTA's and anti-poverty groups. The speakers, which are usually one of the Founders, explain the cause, symptoms and consequences of the disease, and the role of the Foundation in controlling the disease.

2. The Foundation maintains files on: research being done to combat the disease; individuals and organizations who are directly working to overcome the disease; volunteers who have expressed interest in the Foundation; and services available to families and to patients with the disease, such as schools, clinics, etc.

3. The Foundation has distributed over 30,000 brochures on sickle cell disease and the work of the Foundation to physicians, public health centers, schools, social groups, parents' groups and the general public.

4. The Foundation has sponsored educational social functions to introduce the work of the Foundation to the general public and also to persons in the Harlem community. The Foundation has initiated a program of annual awards to community leaders for their efforts in their fight against the disease.

5. The Foundation financed a pilot project for all students at Public School 175 in Harlem (a 95 per cent black student body) where sickle cell testing was included in a complete assessment of individual health status.

6. Through the efforts of the Foundation, "Like It Is," a WABC-TV television program, which describes life in the black community, broadcast a program on the disease and the work of the Foundation. This program, which so far has been broadcast nationwide twice, has stimulated considerable interest in the disease. The Foundation makes this film available to interested groups upon request.

7. The Foundation was instrumental in establishing as a pilot project, the first clinic specifically for sickle cell disease in the metropolitan area at Jamaica Hospital in Queens, a community located in the southeastern area of Queens, where there is the largest concentration of black people in the borough. The Foundation was able to obtain a research grant for the Jamaica Hospital Sickle Cell Clinic to investigate the renal aspects[3] of sickle cell disease. With these funds the Clinic has bought an osmometer and other special equipment and experiments are being conducted, data is being collected and it is expected that results of these experiments will be published in the near future.

8. The Foundation is now involved in the organization of sickle cell clinics at Sydenham and Knickerbocker Hospitals, in the heart of the Harlem community. These clinics are to be centers of research, where the best type of medical care for sickle cell disease is made available.

9. The Foundation has participated in the Harlem Health Day Conference where it set up exhibits, distributed literature and gave 75 sickle prep and sickle dex tests. This was the beginning of the Foundation's program of free testing service in the Harlem community. The Foundation is in the process of evaluating the reliability of the sickle dex,[4] which is a newer method of testing, distributed by Ortho Products. So far, the Foun-

dation has conducted tests in its offices of over two hundred persons.

10. The Foundation has a special testing program for black athletes referred by the Sports Foundation, a Harlem based group interested in promotion of sports and athletics among the youth of Harlem. As a consequence of the Olympic Games held in Mexico in 1968, it came to the attention of this group that athletes with the sickle trait were having difficulties because of the lower oxygen tension of high altitudes. The Sports Foundation consequently wishes evaluation on sickle cell status of its athletes.

11. The Foundation has influenced the New York City Board of Education to undertake a special project in District 4, Manhattan. This project has as its objectives: to test every school age youngster in the District in search for incidence of the sickle cell trait and other physical defects; to provide lecture sessions for patients and community groups about sickle cell disease and the importance of early detection and control; to provide in-service training for teachers in the area that they may become aware of the symptoms of sickle cell disease within the classroom and have knowledge of referral routine; and to provide supportive health services to the youngsters in the area in order that their school attendance, school performance and learning ability will greatly increase.

V. RESOURCES TO MEET THE GOALS

The Foundation has had to rely chiefly on small contributions from individuals, local community groups and charitable donations of social organizations of the black community. Somewhat more substantial amounts have come from civic minded organizations and donations made "in memoriam" of specific individuals. Substantial funding for long range goals continues to be a serious problem. The interest of the government must be aroused and its responsibilities must be pointed out.

We must state again, that "the ultimate goals of the Foundation for Research and Education in Sickle Cell Disease are to coordinate local activities with a national program and to provide for research on all levels." If these goals are to be realized, it requires the knowledge and support not only of the black community, but indeed, all members of the entire American community.

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INCREASE IN MEDICARE INSURANCE PREMIUM

The Department of Health, Education, and Welfare has announced that the voluntary medical insurance premium older people pay for Medicare will be $5.30 a month for the 12-month period that begins next July 1. The present $4 premium rate, set in December 1968, is too low to cover costs during the current premium period and the special Medical Insurance Trust Fund is now drawing on its reserves. Failure to increase the premium rate last December, in accordance with advice from Social Security Administration actuaries, has made it necessary now, in effect, to promulgate two increases at once. Moreover, the depletion of the trust fund that has occurred because of the inadequate rate has made it necessary to provide for a somewhat higher margin of contingency than would otherwise be necessary. About half the increase, 64 cents, is needed just to finance the program at the level of current operations. The other 66 cents of the $1.30 increase in the monthly premium rate will be needed for the following purposes:

- 26 cents to cover an estimated increase of about 6 percent in the level of physicians' fees;
- about 12 cents to cover an estimated increase of 2 percent in the utilization of services under the program;
- about 6 cents because of the $50 deductible which a patient pays will be a smaller proportion of the total covered charges;
- the remaining 22 cents to provide a 4 percent margin for contingencies. This margin is needed because the estimates are based upon minimum reasonable assumptions and because the trust fund out of which this program is financed will be at a low level at the beginning of the premium period on July 1, 1970.

The medical insurance program supplements the basic hospital insurance part of Medicare by helping to pay doctor bills and a wide variety of other medical expenses in and out of the hospital. The premiums paid by people 65 and older who are enrolled in the medical insurance part of Medicare, cover half the cost of their protection. The other half comes out of general Federal revenues. The Medicare law provides for annual review of the costs of the medical insurance program and for any necessary adjustments in the premium rate by January 1. The law requires that the premium rate be sufficient to cover all expenses incurred during each premium period.