The original documents are located in Box 20, folder "Sickle Cell Anemia, 1972-1974" of the Stanley Scott Papers at the Gerald R. Ford Presidential Library.

(Lezar)RP September 30, 1972

SICKLE CELL ARTICLE -- MRS. JULIE NIXON EISENHOWER

There is a story that when the Emancipation Proclamation was taken to President Lincoln for his signature, he picked up his pen, dipped it in ink, moved his hand to the place for his signature -- and then removed his hand and dropped the pen. After a moment's hesitation, Lincoln once again took up the pen -- only to hesitate in signing once again. Immediately, Lincoln turned to Secretary Seward and said:

> "I never, in my life, felt more certain that I was doing right, than I do in signing this paper. But I have been receiving calls and shaking hands since nine o'clock this morning, till my arm is stiff and numb. Now this signature is one that will be closely examined, and if they find my hand trembled they will say, 'he had some compunctions.' But anyway, it is going to be done."

Indeed, Abraham Lincoln's name has gone down in history for that signature. And since that time many other men and women of good will have worked to ensure black Americans the equality of opportunity which should be enjoyed by every American.

It was over 100 years ago that Abraham Lincoln took the first step -- and it has been a hundred years marked by too many promises to black people and too little performance for black people. While there has been much progress there has been too much hesitation as well. And that hesitation can be seen nowhere more clearly than in the continued suffering as a result of the disease called sickle cell anemia.

Although known for centuries in Africa and other parts of the world, sickle cell anemia remained undescribed by doctors in this country until 1910. Even more unfortunate is the fact that only in the last couple of years has the public at large heard of sickle cell anemia. Like so many other Americans, my first awareness of the disease came only as a result of my father's National Health Strategy Message to the Congress of February, 1971, which targeted sickle cell anemia as a critical health problem which deserves special attention on a national scale. He stated that this long neglected disease should receive more attention and proposed a total of $6 million for research, development and treatment of sickle cell anemia. This money represented the first separate Federal effort to fight this disease and meant a five-fold increase over funding

previously available. And later in 1971 this amount was increased, making a total of $10 million for research and community service programs to combat sickle cell disease.

Although national awareness of sickle cell disease has increased, most people know very little about the disease. In recent months I have been fortunate enough to find out more about it and I would like to share some of what I have discovered.

What is sickle cell disease? It is the one most common hereditary blood disease in this country and is found chiefly, but not exclusively, among blacks. It is characterized by the presence of red blood cells which under certain conditions assume a sickle shape. Sickle cell most commonly occurs in one of two forms: either sickle cell trait, a benign form which usually causes no problems, or sickle cell anemia, which is the more severe form of the disease.

Because of the change in shape of the red blood cells caused by sickle cell anemia, those cells have difficulty getting through the smaller blood vessels. They pile up like coat hangers and cause a "log jam" in the blood vessels, thus reducing the flow of blood to the tissues and ultimately resulting in the pain or discomfort of a "crisis."

Patients with sickle cell anemia frequently show the first signs of the disease as early as six to twelve months of age. Those symptoms may include anemia, recurring infections, swelling of the hands and feet, or enlargement of the liver and spleen. Later in childhood, recurrent bouts of pain occur in almost any area of the body, particularly the back, chest, extremities, and abdomen. Leg ulcers frequently develop during the teenage years and the complications of anemia may eventually lead to heart enlargement and heart failure. Victims of sickle cell anemia also have a greater tendency to develop infection, their growth and development may be retarded, and some die at a young age.

What causes sickle cell disease? Although it is a hereditary disease, evidence suggests that it originally developed many centuries ago on the continents of Africa and Southern Asia as an attempt by the body to protect itself against malaria. It is believed that the change in shape of the red blood cells, becoming "sickle" shaped, first occurred as a protective mechanism to engulf and kill malarial organisms which had entered the blood system. What began as a protection mechanism against malaria has today become sickle cell disease. And since malaria is no longer the problem it was years ago, sickle cell disease, first brought to the United States with

black slaves in 1619, has become a major health problem of its own.

Who suffers from sickle cell disease? About 10 percent of the black population of the United States, or 2 and a quarter million persons, have sickle cell trait. About one in 500 black children born in this country suffers from the severe form known as sickle cell anemia. Since sickle cell disease is an inherited blood disease, when two parents with sickle cell trait have children, their offspring may have a 25 percent, or 1 in 4, chance of suffering from sickle cell anemia.

How do you know if you have sickle cell disease? The disease can be diagnosed by a simple blood test which may indicate the presence of the abnormal sickle shaped cells. Through further studies, called electrophoresis, it is possible to discover if a person has sickle cell trait or sickle cell anemia.

How is sickle cell anemia treated? Presently there is no known cure, but physicians can attempt to treat the symptoms which occur. Analgesics are used for pain, antibiotics are used for infection, and patients can be given fluids, oxygen, and blood transfusions, along with other special treatments when necessary.

As I mentioned earlier, I have recently had an opportunity to learn more about sickle cell disease -- particularly by visiting with

the many fine people working to find a cure, and to locate and to help those who suffer from it. The Federal funding which is now being made available is a great help, but in the final analysis it is the efforts of so many fine individuals which will carry the day.

Research activities which have begun include seeking better means to recognize the underlying abnormalities in sickle cell anemia, investigation into the many complications of the problem, and the attempt to find drugs that are effective in the treatment or prevention of the sickle cell "crisis". Recently, several compounds have been evaluated which appear to be useful in treatment and prevention of the painful complications of sickle cell anemia. Hopefully, further research will bring about additional information which will lead to a cure for the disease.

In the community service area, public awareness programs, educational programs, and detection, counseling and referral activities have been begun in an effort to learn what programs are best suited to the varying circumstances found in different localities throughout the country.

On June 29th, I was able to visit one of the first and most successful sickle cell centers in the country, the Deaconess Hospital Sickle Cell Center in Milwaukee, Wisconsin. It is a model medical

facility, but even more importantly, its staff and supporters exhibit a spirit which has brought the Milwaukee community together to fight this disease. The Deaconess Sickle Cell Center represents a coalition of community leadership, professional organizations, agencies and institutions concerned with the health problems of the black community. Extensive sickle cell screening and community education programs have been conducted over the last year and one-half with the cooperation of local news media, the Milwaukee Health Department, the Milwaukee Public Schools, the Medical College of Wisconsin, and a number of community hospitals and social service agencies, as well as the local chapter of the National Foundation for the March of Dimes. In addition, the Milwaukee Advertising Club has selected this as a voluntary program and has begun a wide-ranging media campaign using billboards, newspapers, radio and TV to alert the community to sickle cell disease.

The program was started in 1969 -- well before sickle cell disease began to receive major national attention. I was pleased to go there as a personal representative of the President to recognize their pioneering efforts in treating the disease and bringing it to the attention of the Milwaukee community -- and even more importantly to find out what other communities can do to set up as successful a

program now that the necessary financial resources to begin to fight this disease are becoming available.

The President and the Congress have made it clear that money to launch an all out assault on sickle cell anemia will be made available. In May of this year, the Congress passed -- with bipartisan support -- and the President signed the National Sickle Cell Anemia Control Act, which authorizes the spending of up to $115 million over the next three years for research and community services aimed at combating this disease. The research which this money will make possible can throw additional light on the problem of sickle cell anemia -- and point the way toward better medical treatment. In addition, grants are being made available to help fund 10 comprehensive sickle cell treatment centers and 19 screening and education clinics throughout the United States -- one of which will be the Milwaukee Deaconess Sickle Cell Center.

All this new money will help. But as important as money is, beating sickle cell disease will require much more. It will require community-wide efforts with blacks and whites working together through local clubs and organizations to provide information for patients and the public, to alert health officials and legislators to the needs of the community, to provide training and jobs for sickle

cell patients, to help develop better medical facilities, and to aid in raising the additional money that will be needed if a cure is to be found. At last the possibility of success exists, but people will have to work together to seize the possibility. There has been enough hesitation in the last hundred years. Now is the time for effective community action.

# # #

THE WHITE HOUSE

WASHINGTON

October 4, 1974

MEMORANDUM FOR: STANLEY SCOTT

FROM: JOHN CALHOUN

SUBJECT: Sickle Cell Program

The National Sickle Cell Anemia Control Act (Public Law 92-294) expires in Fiscal Year 1975.

This provides an excellent opportunity to call on the Black Congressional Caucus to help in doing something for blacks through the Legislative Branch. Would advise your approach to them for a liaison meeting and give them an agenda of items in which legislative assistance is needed:

1. Larger budget for OMBE
2. Black colleges
3. Job training programs
4. Etc.

Maybe you could get Blair House for the meeting.

Objective: The Executive and Legislative Branches working together on common needs of blacks.

Educate blacks that Congress also shares part of the responsibility with the President to provide a better life for blacks.

Attachment

THE NATIONAL SICKLE CELL DISEASE PROGRAM

I. BACKGROUND AND HISTORY

Sickle cell anemia, first described in this country in 1910 by Dr. James B. Herrick, is a hereditary blood disorder characterized by the presence of a chronic anemia, jaundice, recurrent bouts of pain called "crises", frequent growth retardation, an increased susceptibility to certain infections, a decreased physical capability, and occasionally a shortened life span. This blood disorder occurs as a result of the presence of genes for sickle cell hemoglobin inherited from both parents. The prevalence of sickle cell anemia in Blacks in the United States is approximately one in six hundred. Some 50,000 or more Blacks are thus affected by sickle cell anemia in the United States.

Sickle cell trait is a healthy state wherein one carries the gene for both sickle hemoglobin and normal hemoglobin. Rarely are there problems associated with sickle cell trait and for the most part individuals with sickle cell trait seldom, if ever, know they carry the sickle gene until tested. The prevalence of sickle cell trait is approximately one in every ten Blacks in this country and numbers approximately two million individuals.

Despite the fact the disorder was first described in 1910, many individuals felt that research and service activities in this problem did not receive adequate attention over the years. In February, 1971, in his Health Message, the President indicated that sickle cell anemia is a national health problem and as such included in his budget an additional five million dollars for research service in this disorder. This amount was later increased to an overall total of ten million dollars in FY 72, fifteen million dollars in FY 73, and sixteen million dollars in FY 74.

The National Sickle Cell Disease Advisory Committee was named by the Secretary, Department of Health, Education and Welfare, and subsequently held its first meeting in August 1971. Through the recommendations of this Committee the National Program was established.

II. OBJECTIVES OF THE PROGRAM

The overall objective of the Federal Sickle Cell Disease Program is to decrease the frequency, morbidity and mortality of sickle cell disease through a program of research and development and demonstration service activities in education, detection, counseling and rehabilitation.

III. NATIONAL SICKLE CELL ANEMIA CONTROL ACT (P.L. 92-294)

On May 16, 1972 the President signed P.L. 92-294, the National Sickle Cell Anemia Control Act. The purpose of this Act is to establish

a national program for the diagnosis, control, treatment of, and research in, sickle cell anemia. The Act provides that these objectives be carried out through a program which includes screening, counseling, information and education, research and development, and training in sickle cell anemia. Congress authorized up to $115 million over a three year period to carry out the provisions of the Act.

IV. COMPONENTS OF THE NATIONAL SICKLE CELL DISEASE PROGRAM, HIGHLIGHTS AND ACCOMPLISHMENTS

Comprehensive Sickle Cell Centers, administered by the Sickle Cell Disease Branch (SCDB), National Heart and Lung Institute, NIH, combine research (basic and clinical), clinical trials, clinical application, training, and community service projects (education, screening and counseling). These Centers focus resources, facilities, and manpower in a coordinated approach to sickle cell disease, bridge the gap between research and service, and bring findings in one area into practical use in the other.

In the first year of operation of the program, ten (10) centers were established utilizing the grant mechanism of support. During the past year an additional five (5) centers were funded bringing the total number of centers to fifteen. It is expected that there will be no further increase over this number in the future.

Specific research projects in progress in the centers include investigation of molecular, cellular, tissue and organ studies in sickle cell disease. Additionally, clinical trials of therapy and continuing education programs for health providers, health educators and other paraprofessionals are in progress. Community service activities include the development of methodology approaches and demonstration projects in community education, screening, counseling and rehabilitation.

Notable accomplishments over the past year include the development of means to better identify abnormal hemoglobins in the presence of large amounts of fetal hemoglobin in cord blood which allows for accurate diagnosis of sickle cell states at birth; the development of a fetoscope which may eventually lead to accurate diagnosis of the presence of abnormal hemoglobins early in pregnancy; the utilization of nuclear magnetic resonance and spin-labelling procedures which will more readily identify the site of structural changes in abnormal hemoglobins and will provide information as to the exact location and mechanism of action of agents which modify hemoglobin structure; the early identification and isolation of retinal changes which herald later serious eye complications and which allow for treatment of lesions before such complications occur; and the trial of several agents which show promise as specific therapeutic approaches for patients with sickle cell anemia.

Continuing education programs have been held by ten (10) centers during the past year and have provided up-to-date information on all aspects of sickle

cell to practicing physicians, investigators and paraprofessionals with activities related to the disorder. More realistic and effective methods of reaching, educating, testing and counseling the general and target populations about sickle cell are in progress. During the past year, technical assistance has been made available to those centers desiring the same to improve their effort in these areas.

The budget for the Center's program during FY 73 was $6.9 million. Of this amount, approximately 40 percent was applied to research activities and 60 percent to demonstration service activities.

Screening and Education Clinics

Screening and Education Clinics are administered by the Sickle Cell Disease Office, Bureau of Community Health Services, HSA. This component of the program serves as a model to determine methods of carrying out service activities such as public awareness, education, screening, counseling and patient referral under various circumstances and in a variety of environments (urban, suburban, rural, etc.).

In the first year of activity, nineteen (19) clinics were funded. This number was increased in FY 73 to an overall total of 26 clinics. The HSA assumed direction of this aspect of the program during the past year and has set up technical assistance and evaluation programs to aid the clinics and facilitate the collection and evaluation of data from these projects.

Preliminary evaluation of clinic operations during this second year of activity indicates increased effectiveness with definitive direction developed for obtaining the desired information of effective means of carrying out service activities in sickle cell in various environments. It is anticipated that in the process of developing and acquiring data, approximately four hundred twenty thousand (420,000) individuals will be screened and forty thousand (40,000) counseled during this fiscal year.

Continuing education programs have also been carried out by several of the screening and education clinics and a closer liaison has been developed between the clinic and center operations. The total budget for clinic operations which included technical assistance and evaluation projects in FY 73 was $3.1 million.

Mission Oriented Research and Development Program

The Sickle Cell Disease Branch, National Heart and Lung Institute, funds and administers the contract-supported, mission-oriented research and development program which has as one of its objectives the study of mechanisms for the prevention and treatment of sickle cell disease. Twenty-seven contracts were awarded in FY 73 totaling $2.1 million. All but one of these awards represents renewal of the previous year's contracts. The one exception is an award made for the formation of a Hemolytic Disorder Study Group for the

study of sickle cell disease and related hemolytic disorders and the preparation of a protocol to test the efficacy of sodium cyanate as a therapeutic approach to decrease the frequency of crises in patients with sickle cell anemia.

Notable accomplishments under this component of the program during the past year include the findings that zinc chloride, alkylating agents and certain amino acids, in addition to the previously described sodium cyanate, will alter the sickling phenomenon. Additionally, the cell membrane appears to play an active role in the sickling process and may be predicated upon changes in the calcium and adenosine triphosphate content of the membrane. Uncontrolled studies utilizing oral and extracorporeal sodium cyanate in sickle cell anemia patients continue to look promising, however, neurological side-effects noted on utilizing the oral route will require additional pharmacological and toxicological studies. Methodologies to identify hemoglobin types in sickle cells obtained from fetal circulation or amniotic fluid are being developed. Success in this area would allow for positive identification of several abnormal hemoglobins in the first trimester of pregnancy. A simple and rapid method of determining hemoglobin-A level in blood samples was developed. This will aid substantially in the identification of Cooley's Anemia states with and without associated sickle hemoglobin. A specific procedure was developed and standardized to measure one of the active defense mechanisms for infections in patients with sickle cell anemia. This may aid in identifying those sickle cell patients with an increased susceptibility to specific infections.

The Hemolytic Disorder Study Group, which consists of a large number of hematologists and other sickle cell investigators across the country, prepared a protocol to test the efficacy of sodium cyanate in patients with sickle cell anemia. This protocol now awaits additional pharmacology and toxicity studies before large clinical trials are initiated.

A National Symposium on Sickle Cell Disease was held late in FY 74 to permit investigators to present papers describing research in sickle cell over the past two years. It is expected that evaluation and analysis of the papers presented will aid in defining promising avenues of sickle cell research for consideration of support in the future.

Biomedical Research Program

The Sickle Cell Disease Branch, NHLI, funds a program of investigator-initiated research under the grant support mechanism. Five (5) grants are presently being supported directly by Sickle Cell Disease Program earmarked funds at a cost of $122,000 with progress of studies reportedly periodically.

Additional research in sickle cell disease is being supported by funds made available by the National Heart and Lung Institute, National Institute of Arthritis, Metabolism and Digestive Diseases, and National Institute of General Medical Science independent of Sickle Cell Disease Program earmarked funds. Grant supported investigation includes studies of the basic structure,

the synthesis of hemoglobin structure, improved detection and identification of abnormal hemoglobins and flow patterns exhibited by cells containing sickle hemoglobin.

Education Program

Education of the general public including health providers, health educators, and the target population about sickle cell has received top priority during the past year. In addition to continuing education programs provided by centers and clinics, the education component has stepped up its efforts to provide factual information to the public through a variety of tools including films, lectures, workshops, seminars, tutorials, pamphlets, and brochures. A communication plan was developed with direction and guidance provided by input from concerns expressed by community organizations and agencies. Emphasis on the comprehensive approach was increased through input by psychologists and social workers. State and laboratory personnel have also had an opportunity to increase their knowledge of sickle cell through the hemoglobinopathy workshops conducted by the Center for Disease Control.

To broaden the education approach further, the Program issued a request for proposals during the year to utilize nongovernmental organizations, agencies, or individuals to carry out programs in public awareness, to aid in the dissemination of factual information, and to conduct educational programs in sickle cell throughout the country. Award of this contract is expected to be made early in FY 75.

The increased effort in education during this year is expected to result in a marked increase in public awareness and a better educated community regarding sickle cell states.

Hemoglobinopathy Detection Training Program

Through an interagency agreement the Hematology Division of the Center for Disease Control (CDC) conducts a training program in the detection of abnormal hemoglobins for personnel of projects funded under the Federal Sickle Cell Disease Program and other state and private laboratory personnel. During the past year, four (4) basic courses and one (1) advance course were held to train personnel in methods in hemoglobinopathy detection. A proficiency testing program was also developed to insure quality control of laboratory techniques in the federally funded screening projects.

The Center for Disease Control also provided reference evaluation service and advice for the identification of abnormal hemoglobins submitted from numerous laboratories across the country.

V. LEGISLATIVE RECOMMENDATIONS

Recommendations relative to the renewal of the National Sickle Cell Anemia Control Act which expires in FY 75 are presently being developed by the Department for subsequent submission to the Congress.