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AN ANALYSIS OF THE FEDERAL SICKLE CELL DISEASE PROGRAM, FY 1971 - FY 1976

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I. INTRODUCTION

Congressional interest in sickle cell anemia peaked during the 92nd Congress. In February 1971, President Nixon emphasized the importance of concentrating Federal resources and public attention on a campaign to conquer sickle cell anemia. On February 6, 1975, President Ford sent a message to the Congress transmitting the Second Annual Report on the Administration of the National Sickle Cell Anemia Control Act. This message commended the Health Services Administration and the National Institutes of Health for their combined effort to bring demonstration service activities to the American Public. The message also stated: "The activity toward Sickle Cell Anemia continues to be of high priority for our Government. . ." 1/ On June 30, 1975, the legislation authorizing the Sickle Cell Disease Program will expire.

This report reviews the development of the Federal Sickle Cell Disease Program. The study begins with a brief overview of the sickle cell disease, and continues with a history of Federal funding for sickle cell anemia projects. This history includes: (1) an analysis of the authorizing legislation, (2) an explanation of the Federal support within the National Institutes of Health and the National Heart and Lung Institute from FY 1970 through FY 1976, and (3) a review of the Congressional interest and appropriations for the Sickle Cell Disease Program from FY 1971 through FY 1976. The report proceeds with an analysis of the current literature to determine the social impact of the Sickle Cell Disease Program, including public reaction to the research, screening, counseling, education, and treatment components of the Program. The conclusion of the report presents suggestions which have been made for improving the Sickle Cell Disease Program. These suggestions are voiced in the literature, and concentrate on the role of the Federal government in funding and reorganizing certain portions of the Program.

1/ Second Annual Report on the Administration of the National Sickle Cell Anemia Control Act - Message from the President of the United States. Congressional Record (daily ed.) v. 121, Feb. 6, 1975: H673.

II. THE SICKLE CELL DISEASE

"Sickle cell disease" is a comprehensive term which includes sickle cell anemia, sickle cell trait, and sickle cell variants. It is a "molecular" disease involving abnormal hemoglobin in red blood cells. Hemoglobin gives normal blood cells their red color, and is an iron-containing protein that transports oxygen from the lungs to the various tissues and organs of the body. The abnormal hemoglobin found in persons with sickle cell disease is designated as "hemoglobin S"; this can be compared to normal adult hemoglobin, designated "hemoglobin A". Hemoglobin S, when deprived of oxygen (and depending on several conditions), causes the cell to "sickle", that is, the cell becomes elongated with pointed ends.

Sickle cell disease is also a genetic disease. According to the laws of heredity, each parent contributes to a child only one gene from each pair of genes. If one parent has a matched set of genes, each member of that set being a mutant gene for hemoglobin S, then that person is said to be HOMOZYGOUS for sickle cell disease and would be subject to the sickle cell anemia. However, if only one gene in the set is a mutant gene, and the other one is normal, then that person is said to be HETEROZYGOUS for sickle cell disease, and would be considered a sickle cell "trait" carrier and not necessarily subject to sickle cell anemia. (Carriers of the sickle cell trait may have sickled cells in their blood, but generally not to the damaging extent of those persons homozygous for the disease.) Sickle cell disease is not a sex-linked disease.

In order for a child to be a trait carrier (heterozygous), he must have inherited a normal gene from one parent and a mutant gene from the other. In order for a child to be subject to sickle cell anemia (homozygous), he would have had to inherit a sickle cell gene from each parent. Therefore, if both parents carry the trait but not the disease itself, there is a 1-in-4 chance that their child will be afflicted with sickle cell anemia (that is, be homozygous), a 2-in-4 chance that he will carry the sickle cell trait (that is, be heterozygous), and a 1-in-4 chance that he will have normal hemoglobin. If one parent is homozygous for sickle cell anemia and the

other has the trait, then there is a 2-in-4 chance that the child will also have sickle cell anemia, and a 2-in-4 chance that he will be a trait carrier.

Sickle cell disease is found predominantly, but not exclusively, in black populations; people of Mediterranean heritage may also be subject to it. Although statistics vary widely, it has been generally estimated that sickle cell anemia occurs in approximately 1 out of every 500 people of African descent. In the United States, it is estimated that sickle cell anemia afflicts more than 50,000 persons. Of course, the sickle cell trait is far more prevalent, and is estimated to occur in approximately 10% of black children. This means that more than two million Americans are estimated to carry the sickle cell trait.

The damage caused by the sickling of the red blood cells in sickle cell anemia can range from mild symptoms to death. When the cells sickle, the body acts quickly to eliminate them from the blood stream. This results in a severe deficiency of red blood, and anemia ensues. Furthermore, blood containing sickled cells is thicker, heavier, and stickier than normal blood, and it does not flow as easily through the smaller blood vessels. The sickled cells begin to stick together, blocking the small vessels and preventing the normal flow of blood. This impediment of the blood flow may cause severe damage, or death, to body tissues.

The clinical symptoms which result from this sickling and the blocking of blood pathways culminate in sickle cell "crises", a direct consequence of the anemia and the damaged tissues. Usually, only those with the disease itself will exhibit the symptoms, while those with the trait generally experience symptoms only rarely and then only during periods of great stress. Recent evidence has shown that in isolated cases, sickle cell trait carriers may experience fatal crises when subjected to severely stressful conditions. Although this potentiality has not been substantially proved, the possibility cannot be discounted; more study is needed.

Severe pain and possible fever characterize the crises, and the patient may become somewhat jaundiced. There may be sores, particularly in the extremities, which do not heal readily. The body may not be as effective in fighting infection.

Persons with sickle cell anemia are often poorly developed and have a short trunk with long arms and legs. In severe cases, the patient may experience weakness, headache, dizziness, ringing in the ears, and spots before the eyes. Nausea and vomiting may be present during a crisis or during periods of severe stress. But the most outstanding symptom of this disease is the pain. This pain is generally most acute in the bones, large joints, the back, and in the abdomen. Severe abdominal pain accompanied by fever may at first appear to be an attack of appendicitis. Vessel blockage in the chest and in the lungs may result in severe chest pains and difficult breathing which also might be diagnosed as another disorder. Strokes and blindness could result from involvement of the vessels of the brain and the eye. Sickle cell anemia has been described as the "great imitator" because of the many symptoms which are associated with the disease --- symptoms which account for a great deal of misdiagnosis and delayed diagnosis.

Screening for sickle cell disease is not a difficult process and is often advised for anyone who is suspected of being susceptible. Those found to have the anemia are placed under continual medical supervision where their prognosis can be significantly improved. Those found to carry the sickle cell trait might be advised of diet and prevention of infection, and genetic counseling might be suggested in order to inform parents of potential sickle cell anemia in their children.

Presently, there is neither effective treatment nor cure for this disease, nor can it be prenatally diagnosed, but medical treatment can be an aid to people afflicted by sickle cell disease. Treatment for the disease is mainly of a supportive nature, since research has yet to discover an effective means to completely control the symptoms. During crises, drugs are used to ease the pain and fluids are administered to counter dehydration. Oxygen and blood transfusions are occasionally administered, but only if absolutely necessary. Analgesics and sedatives are employed for symptomatic relief, and a variety of medications have been introduced, including: corticotropin, adrenal and gonadal steroids, intravenous sodium bicarbonate, anticoagulants, carbon monoxide inhalation, acetazolamide, infusion of low molecular dextran phenothiazine,

and more recently urea and cyanate. Between crises, certain "maintainance" measures can be taken; good nutrition, periodic physical and hematological check-ups, and good mental hygiene are extremely important to anyone afflicted with sickle cell disease.

In laboratory experiments and clinical trials, urea treatments have been shown to reverse the sickling process by breaking the bonds which cause a cell to hold the sickle shape. Until recently there was great hope that urea would prove to be an effective treatment against the symptoms of sickle cell disease. However, during 1974 the National Heart and Lung Institute (NHLI) of the National Institutes of Health (NIH) conducted many experiments and found that urea offered no special advantages over other measures to correct dehydration or excess blood acidity in the treatment of the sickle cell crisis. Neither did urea prove valuable in controlling the pain which accompanies the crisis. Likewise, NHLI found that sodium bicarbonate and dextrose, two other experimental agents, did not perform satisfactorily in these areas.

Presently, one of the most promising new agents for treating sickle cell anemia is sodium or potassium cyanate. This compound has been shown to inhibit effectively the sickling of red blood cells. Cyanate is now being used on a limited number of patients in studies at Rockefeller University, the University of Miami, and John Hopkins University. Side-effects have appeared and include weight loss, nerve disorders, tingling, burning, and other nerve sensations. Physicians working with the drug believe that these effects are dose-related, and as the dose is reduced the side-effects will diminish. One study has also shown that sodium cyanate is effective in decreasing the frequency of painful crises among sickle cell patients. Other promising but clinically untested agents include dimethyl adipimidate (DMA), zinc, and aspirin.

The ultimate goal in sickle cell therapy is to find an effective treatment that exhibits no adverse side-effects. Beyond that, however, the scientist's basic concern is to find ways to overcome the underlying problem of the disorder -- the sickling of the red blood cells -- rather than restricting treatment to the symptoms of the disease.

III. A HISTORY OF FUNDING FOR SICKLE CELL DISEASE PROJECTS

A. Authorizing Legislation

The administration of the Sickle Cell Disease Program is assigned to the National Institutes of Health of the Department of Health, Education, and Welfare (DHEW). The legislative authority of certain sections of the Public Health Service Act provides for the specific funding of the Sickle Cell Disease Program and for general research, research training and fellowships. The list which follows presents the authorizing legislation for each institute supporting sickle cell disease projects.

National Institutes of Health

National Heart and Lung Institute (NHLI)
- Public Health Service Act
  - Title III, Part A, Section 301, General Research
  - Title IV, Part B, National Heart and Lung Institute
  - Title XI, Genetic Blood Disorders, Part A, Sickle Cell Program

National Institute of Arthritis, Metabolism, and Digestive Diseases (NIAMDD)
- Public Health Service Act
  - Title III, Part A, Section 301 General Research
  - Title IV, Part D, National Institute on Arthritis, Metabolism and Digestive Diseases

National Institute of General Medical Sciences (NIGMS)
- Public Health Service Act
  - Title III, Part A, Section 301, General Research
  - Title IV, Part E, National Institutes of Child Health and Human Development and of General Medical Sciences

National Institute of Child Health and Human Development (NICHD)
- Public Health Service Act
  - Title III, Part A, Section 301, General Research
  - Title IV, Part E, National Institutes of Child Health, and Human Development and of General Medical Sciences

The Secretary of DHEW has delegated legislative authority for the funding of the Sickle Cell Disease Program to the National Institutes of Health. The legislative authority for funding under Title III, Part A, Section 301, (General Research, hereafter referred to as "standard general research appropriations" in this report) and Title IV, Parts B, D, and E (NHLI, NIAMDD, NIGMS, NICHD) is indefinite, that is, of a continuing but non-specific level. Sickle cell disease projects which are funded under these titles

of the Public Health Service Act were initiated and continue at levels of support derived from general research or general operations funds.

Only the legislative authority provided by Title XI, Genetic Blood Disorders, Part A, Sickle Cell Disease Program, of the Public Health Service Act is a specific authorization of funding for the Sickle Cell Disease Program. Title XI, Part A (Public Law 92-294) establishes a national program for the control of sickle cell anemia. This title authorizes grants and contracts (1) to assist in the establishment and operation of voluntary sickle cell anemia screening and counseling programs as part of existing health programs, and (2) to support research in the diagnosis, treatment, and control of sickle cell anemia, as well as the development of education, counseling, and testing programs. Title XI, Part A directs the Secretary DHEW to carry out a program to develop and disseminate information and educational materials for the public and for health professionals, and to establish a program through the Public Health facilities to provide for voluntary sickle cell anemia screening, counseling, and treatment. Appropriations authorized in Title XI, Part A include:

Section 1101 — Grants and Contracts for Sickle Cell Anemia Screening and Counseling Programs and Education Programs

FY 1973	$20,000,000
FY 1974	$30,000,000
FY 1975	$35,000,000

Section 1102 — Project Grants and Contracts for research and research training in the diagnosis, treatment, and control of sickle cell anemia, the development of programs to educate the public regarding the nature of the sickle cell trait and sickle cell anemia counseling and testing programs, and other programs for diagnosis, control, and treatment of sickle cell anemia

FY 1973	$ 5,000,000
FY 1974	$10,000,000
FY 1975	$15,000,000

Title XI, Part A, Total Appropriations Authorizations

FY 1973	$25,000,000
FY 1974	$40,000,000
FY 1975	$50,000,000

B. Federal Support for Sickle Cell Disease Projects Within the National Institutes of Health

Currently, sickle cell disease projects are supported by general research or general operations funds within three of the National Institutes of Health (NIH), the National Institute of Arthritis, Metabolism, and Digestive Diseases (NIAMDD), the National Institute of General Medical Sciences (NIGMS), and the National Institute of Child Health and Human Development (NICHD). The provisions of Title XI, Part A of the Public Health Service Act became law on May 16, 1972 (Public Law 92-294), and became effective in FY 1973. However, until FY 1974, there were no appropriations under Title XI for the Sickle Cell Disease Program. The appropriations bills for FY 1973, H.R. 15417 and H.R. 16654, included appropriations for the Program under Title XI in the budget of the National Heart and Lung Institute (NHLI); these bills were vetoed by the President on August 16, 1972 and November 1, 1972, respectively. Funding for programs in NIAMDD, NICHD, and NIGMS continued at a level relatively similar to the appropriations for FY 1972. During FY 1973, support in the NHLI increased by 50% over FY 1972 levels. 2/

In FY 1973, the provisions of Title XI, Part A were implemented, but the support was derived from standard general research appropriations. Subsequently, the Secretary DHEW designated NIH as the agency to coordinate research, and the NHLI as the lead institute for the total DHEW sickle cell disease effort. Also in FY 1973, the Health Services Administration (HSA, formerly the Health Services and Mental Health Administration, HSMHA) was charged with providing service functions in the Sickle Cell Disease Program. These service functions included the screening, counseling, and education portions of the DHEW effort. NIH's research activities in Sickle Cell Disease are primarily the responsibility of the National Heart and Lung Institute. This institute specializes in research to develop possible preventive measures, and treatment of the sickle cell crisis. NIAMDD, NICHD, and NIGMS also have a research role: these institutes specialize in molecular biology research to determine the genetics of sickle cell anemia and the

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2/ DHEW received appropriations at the FY 1972 level of support in these continuing resolutions: Public Law 92-334 (July 1, 1972), Public Law 92-571 (October 26, 1972), and Public Law 93-52 (July 1, 1973).

nature of the hemoglobin molecule.

In FY 1974 and FY 1975, funds were appropriated for the Sickle Cell Disease Program under Title XI, Part A. However, in both fiscal years, this appropriation was combined with the appropriation for NHLI, that is, one appropriation was cited in the appropriatons bills to include funds authorized for NHLI, and for the Sickle Cell Disease Program. Consequently, the exact sum appropriated by the Congress to fund the Program for FY 1974 and FY 1975 cannot be determined.

All support for the Sickle Cell Disease Program outlined in Title XI, Part A of the Public Health Service Act is to be appropriated and obligated in the budget of the National Heart and Lung Institute. The sickle cell disease research performed by the other institutes is to be funded by the National Institutes of Health budget for general research, i.e., this support is not ear-marked as in Title XI, Part A to fund sickle cell anemia research projects. HSA derives support for screening, counseling, and education programs from a reimbursable interagency agreement with the National Heart and Lung Institute: NHLI is essentially "billed" by the HSA for the services it performs. HSA receives no support for the Sickle Cell Disease Program in its own budget.

Table I below presents a history of the U.S. Budget obligations for sickle cell anemia projects by institute.

C. Federal Support for the Sickle Cell Disease Program Within the National Heart and Lung Institute

The Sickle Cell Disease Branch, NHLI, has cooperated with the Bureau of Community Health Services, HSA, in establishing Sickle Cell Disease Centers, and Screening, Counseling, and Education Clinics, as well as collaborative research and development contracts, and biomedical research grants. Table II below presents a breakdown for the levels of NHLI obligated support from FY 1971 through FY 1976 by component of the Sickle Cell Disease Program.

TABLE I: National Institutes of Health, Budget Obligations for Sickle Cell Disease Projects, FY 1970 - FY 1976 (in millions of dollars) 3/

Institute	FY 1970	FY 1971	FY 1972	FY 1973	FY 1974	FY 1975 est.	FY 1976 est.
National Heart and Lung Institute*	0.455 a/	0.935 a/	10.192 b/	15.320 b/	16.115 b/	16.000 b/	16.000 b/
National Institute of Arthritis, Metabolism, and Digestive Diseases	0.766 a/	0.855	0.964	1.120	1.116	1.154	1.154
National Institute of General Medical Sciences	0.100 a/	0.100	0.100	0.100	0.100	0.100	0.100
National Institute of Child Health and Human Development	0.050 c/	0.162 c/	0.214 c/	0.082 c/	0.094 c/	0.136 c/	0.137 c/
---Health Services Administration*	-----	-----	1.975 d/	3.070 d/	3.033 d/	3.500 d/	3.500 d/
TOTAL NATIONAL INSTITUTES OF HEALTH	1.321	2.002	13.445	19.692	20.500	20.890	20.891

3/ Unless otherwise noted, figures quoted in this table are taken from: U.S. Congress, House. Committee on Appropriations. Departments of Labor and Health, Education, and Welfare Appropriations for 1975. Hearings, 93rd Congress, 2nd session, Part 3. Washington, U.S. Govt. Print. Off., 1974. p. 681.

a/ U.S. Congress. House. Committee on Appropriations. Departments of Labor and Health, Education, and Welfare Appropriations for 1974. Hearings, 93rd Congress, 1st session, Part 4. Washington, U.S. Govt. Print. Off., 1973. p. 307.

b/ Quoted from personal communication with Mr. Art Fried, Budget Analyst, National Heart and Lung Institute, February 24, 1975.

c/ Quoted from personal communication with Mr. Dale Dewald, Budget Analyst, National Institute of Child Health and Human Development, March 17, 1975. Figures represent subsidary research, minor research, data analysis, i.e., no primary research projects for sickle cell disease alone are funded. FY 1975 estimate is at rescission level; without rescission, FY 1975 totals $172,000.

d/ Quoted from personal communication with Ms. Jane Peterson, Budget Analyst, Health Services Administration, March 19, 1975. Figures quoted for FY 1975 and FY 1976 do not coincide with figures for HSA from the NHLI Budget, which place FY 1975 at $3.0 million and FY 1976 at $3.0 million for HSA participation in the Program.

* In this table, HSA funding is shown as a separate entry which has already been included in the NHLI column.

TABLE II: NHLI Obligated Funds for the Sickle Cell Disease Program (in millions of dollars) 4/

Program (Administrative Agency)	FY 1971	FY 1972	FY 1973	FY 1974	FY 1975 est.*	FY 1976 est.
Comprehensive Centers for Research (NHLI) and Service (HSA)	N.A.	4.804	6.577	6.762	6.700	6.700
Screening, Education and Counseling Clinics (HSA)	N.A.	1.975	3.070	3.140	3.400	3.400
Collaborative Research and Development Contracts (NHLI)	1.000	2.326	2.251	2.117	2.100	2.100
Biomedical Research Grants (NHLI)		.758	2.561	2.746	2.450	2.450
Administrative Expenses and Intramural Programs (NHLI)	N.A.	.329	.861	1.350	1.350	1.350
TOTAL SUPPORT (NHLI)	1.000	10.192	15.320	16.115	16.000	16.000

4/ Quoted from personal communication with Mr. Art Fried, Budget Analyst, National Heart and Lung Institute, February 24, 1975.

* Figures quoted to include FY 1975 rescissions. If FY 1975 rescissions are rejected by the Congress, the Sickle Cell Disease Program will receive an additional $400,000, for which further allocations among Program components has not been determined.

D. Congressional Appropriations for the Sickle Cell Disease Program

The Sickle Cell Disease Program does not appear as a distinct line-item in the U.S. Budget or in any Congressional appropriations bills. Consequently, unless the Congress makes specific mention of the funding for the Program in reports to accompany appropriations bills or in appropriations hearings, the specific Congressional intent to fund the Program cannot be identified.

Between FY 1971 and FY 1974, only one report to accompany an appropriations bill highlighted the Congressional interest in specifically funding the Sickle Cell Disease Program. In House Report Number 92-1118, it was stated:

> In retrospect, the Committee is surprised and somewhat chagrined that so little attention has been paid in the past to such a serious disorder as sickle cell anemia. In 1971 NIH spent only about $1 million that could be identified as being research directed at this genetic disease. For the current year (FY 1972) about $10 million is being used and the request for 1973 includes $15 million, of which $14 million is in this Institute's (NHLI) budget. The Committee made no change in this amount after being assured by the Assistant Secretary for Health and Scientific Affairs that, "This will make it possible for us to increase the amount of money that is in research so that every reasonably promising lead can be explored with regard to preventing the disease and treatment". 5/

In FY 1975, the Senate Committee on Appropriations again mentioned specific support for the Sickle Cell Disease Program. In Senate Report Number 93-1146, the Committee wrote:

> Although testimony indicated that clinical trials to evaluate the anti-sickling agent urea have shown that it is not effective in the treatment of the sickle cell crisis - an episode of acute, intense pain often followed by deformity of arms or legs - evaluation of other agents that react with the abnormal hemoglobin continues. The NHLI currently (FY 1974) supports 15 sickle cell disease research centers and 23 screening and education clinics. The Committee recommends that up to $25 million be allotted for an expanded program in this important area. 6/

Appropriations hearings by the Congress do not readily provide a concise and detailed accounting of the Sickle Cell Disease Program budget. Explanations of funding for this effort are complicated even more by the piecemeal accumulation of information scattered throughout these hearings in oral testimony and in prepared justifications. However,

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5/ U.S. Congress. House. Committee on Appropriations. Departments of Labor, and Health, Education, and Welfare, and Related Agencies Appropriations Bill, 1973; Report to accompany H.R. 15417. Washington, U.S. Govt. Print. Offl, 1972. (92nd Congress, 2nd session. House. Report no. 92-1118) p. 27.

6/ U.S. Congress. Senate. Committee on Appropriations. Departments of Labor, and Health, Education, and Welfare, and Related Agencies Appropriations Bill, 1975; Report to accompany H.R. 15580. Washington, U.S. Govt. Print. Off., 1974. (93rd Congress, 2nd session. Senate. Report no. 93-1146) p. 46-47.

the intent of the Congress to discover the levels and sources of support for sickle cell disease projects gradually emerges in a review of the appropriations hearings between FY 1971 and FY 1975 (See Appendix I of this report). Budgetary information provided in oral testimony closely resembles the levels of support cited in Tables I and II above.

In FY 1972, oral testimony before the Senate Committee on Appropriations included this statement: "There are some increases in NHLI budget for sickle cell anemia, some modest increases in the area of arteriosclerosis. Because the total budget remains the same, it means the increases in those areas are having to be squeezed out of some other areas." 7/

During the FY 1973 hearings, oral testimony and prepared justification statements included a detailed accounting of support for the Sickle Cell Disease Program by institute and by program component.

Oral testimony in FY 1974 clarified the relative costs of research and service in the Sickle Cell Disease Program: In the Comprehensive Centers for Research and Service, 40% of the cost is applied to research activities and 60% toward service activities. Research and development contracts support controlled studies of therapy, basic studies of molecular structure and various complications of sickle cell anemia. Biomedical research project grants support meritorious research on sickle cell anemia; this research is recommended by other NIH institutes, but is funded by NHLI. 8/

In FY 1973 and FY 1974, justifications statements prepared by NHLI reported that any increases requested for the Sickle Cell Disease Program were completely included and

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7/ U.S. Congress. Senate. Committee on Appropriations. Departments of Labor, and Health, Education, and Welfare Appropriations. Hearings, 92nd Congress, 1st session, on H.R. 10061. Part 6. Washington, U.S. Govt. Print. Off., 1971. p. 3752.

8/ U.S. Congress. House. Committee on Appropriations. Departments of Labor, and Health, Education, and Welfare Appropriations for 1974. Hearings, 93rd Congress, 1st session. Part 4. Washington, U.S. Govt. Print. Off., 1973. p. 307-311.

ear-marked for the Program in the total increase requested for NHLI. 9/ Although there have been increases in the overall NHLI budget between FY 1972 and FY 1974, it appears that the increases in the Sickle Cell Disease Program during this time were derived only in part from increases in the total NHLI budget.

During FY 1975 appropriations hearings, oral testimony included a discussion of the funding, as well as the goals, of the component activities within the Sickle Cell Disease Program. Members of the Senate Committee inquired whether the budget request for this Program's operation in FY 1975 would include an increase to cover rising costs due to inflation. The Acting Director of NHLI (Dr. Ringler) replied: "that level (FY 1975) is the same (as FY 1974), and to whatever extent inflation is a factor, it will be reduced accordingly." 10/ In its justifications for FY 1975, NHLI outlined the research and control goals of the Program: "The research goals of the sickle cell disease program are to develop improved therapy for sickle cell crises; increase knowledge of the fundamental biology of the disease and its complications; and develop effective and acceptable methods for screening and counseling. . . .The goals of the control program are to educate the public about sickle cell disease and Cooley's anemia and to demonstrate techniques for appropriate screening and counseling." 11/

9/ U.S. Congress. House. Committee on Appropriations. Departments of Labor, and Health, Education, and Welfare Appropriations for 1974. Hearings, 93rd Congress, 1st session. Part 4. Washington, U.S. Govt. Print. Off., 1973. p. 246. See also: U.S. Congress. Senate. Committee on Appropriations. Hearings, 92nd Congress, 2nd session, on H.R. 15417. Part 3. Washington, U.S. Govt. Print. Off., 1972. pp. 2956, 2989, 2995; and U.S. Congress. House. Committee on Appropriations. Departments of Labor, and Health, Education, and Welfare Appropriations for 1974. Hearings, 93rd Congress, 1st session. Part 4. Washington, U.S. Govt. Print. Off., 1973. pp. 346, 367, 379, and 383.

10/ U.S. Congress. Senate. Committee on Appropriations. Departments of Labor, and Health, Education, and Welfare Appropriations. Hearings, 93rd Congress, 2nd session, on H.R. 15580. Part 3. Washington, U.S. Govt. Print. Off., 1974. pp. 975-976.

11/ U.S. Congress. House. Committee on Appropriations. Departments of Labor, and Health, Education, and Welfare Appropriations for 1975. Hearings, 93rd Congress, 2nd session. Part 4. Washington, U.S. Govt. Print. Off., 1974. pp. 320, 322-323.

The date on which particular authorizing legislation is cited in hearings is complicated by the date of enactment of the National Sickle Cell Anemia Control Act itself (Public Law 92-294). This Act became Title XI, Part A of the Public Health Service Act on May 16, 1972. Consequently, Title XI, Part A was not cited as authorizing legislation by NHLI during FY 1973 appropriations hearings. From FY 1971 to FY 1973, sickle cell disease support in all the NIH institutes was authorized in the standard general research appropriation. Beginning in FY 1974, and continuing through FY 1975, NHLI cites Title XI, Part A as the legislation to authorize and establish all aspects of the Sickle Cell Disease Program, while the other institutes continue to cite the standard general research authorization for support of basic sickle cell disease research projects.

E. Summary

The history of funding for sickle cell disease projects includes an analysis of (A) the authorizing legislation, (B) the support within the National Institutes of Health, (C) the administration and support within the National Heart and Lung Institute, and (D) the appropriations and interest of the Congress in the Sickle Cell Disease Program.

The legislative authority of Title III, Section 301, (General Research); Title IV, Parts B (NHLI), D (NIAMDD), and E (NICHD and NIGMS); and Title XI, Part A (Sickle Cell Disease Program) of the Public Health Service Act provides for general research, research training and fellowships, and the specific funding of the Sickle Cell Disease Program. In FY 1973, the provisions of Title XI, Part A of the Public Health Service Act (Sickle Cell Disease Program) were implemented, but the support was derived from appropriations under Title III, Section 301, and Title IV, Parts B, D, and E of the Public Health Service Act.

In the National Institutes of Health, NHLI has reported a steady increase in funding the Sickle Cell Disease Program; the other institutes have maintained a combined sickle cell research budget of approximately $1 million. Although there have

been increases in the overall NHLI budget between FY 1971 and FY 1976, it appears that the increases in the Sickle Cell Disease Program during FY 1973 were derived only in part from increases in the total NHLI budget. For FY 1974, NHLI requested a total budget increase of approximately $18 million, with approximately $1 million ear-marked as an increase for the Sickle Cell Disease Program. For FY 1975 and FY 1976, NHLI has begun to stabilize support for the Program at approximately $16 million.

Congressional interest in sickle cell disease peaked during the 92nd Congress. Beginning in FY 1973, and continuing through FY 1975, the Congress attempted to appropriate funds under Title XI, Part A of the Public Health Service Act for the Sickle Cell Disease Program. However, the Program does not appear as a line-item in the U.S. Budget or in Congressional Appropriations bills: unless the Congress makes specific mention of funding for the Sickle Cell Disease Program in reports to accompany appropriations bills, or in appropriations hearings, Congressional intent to fund the Program can only be estimated.

IV. THE IMPACT OF THE SICKLE CELL DISEASE PROGRAM AS REFLECTED IN CURRENT LITERATURE

A. Background

During the past several years, sickle cell anemia has become a well-known issue surrounded by medical, political, and social controversy. The problems causing these controversies have been publicized in medical journals, newspapers, and other published sources. The following analysis explores public reaction to the National Sickle Cell Disease Program as reflected in current literature.

In February 1971, President Richard Nixon publicly indicated special support for sickle cell programs. Before this presidential emphasis, sickle cell anemia was "non-visible", both politically and socially. The majority of the public did not know that such a disease even existed. Some research had been performed; however, the scientific community appeared to regard the disease as an interesting theoretical challenge, not necessarily worthy of high-level government support. Some black people now refer to this era as a period of "indifference" on the part of a white society.

The President's health message marked the advent of a new effort to research this disease. In his message, President Nixon increased the sickle cell budget to $6 million; prior to 1971, only $1 million supported sickle cell research. Some observers viewed this new emphasis as a political move designed, "to win friends for Nixon, who is disliked by many blacks." 12/ The $6 million was to be divided equally between research and services.

On October 8, 1971, Senator John V. Tunney introduced "The National Sickle Cell Anemia Prevention" bill which authorized Congress to appropriate funds for sickle cell research and services. This bill, which was passed and made part of the Public

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12/ Nixon Administration Opens Politics-Laden Attack on Sickle Cell Anemia. Drug Research Reports, v. 14, Sept. 1, 1971: 4.

Health Service Act in 1972, authorized $115 million for a three year program: $25 million for fiscal year 1973, $40 million for fiscal year 1974, and $50 million for fiscal year 1975. As noted earlier, the program is coordinated through NIH's National Heart and Lung Institute (NHLI), and thus it is mainly through NHLI that funds are available for research, studies, and services.

B. Racial/Social Overtones in Research

The major problems with the current Sickle Cell Disease Program are the implicit and expected racial overtones. For example, one argument states that if sickle cell was a white man's disease, research and treatment would have been established long ago.13/ However, an article in Medical World News stated that sickle cell research was actually underway for twenty-five years prior to 1971; racial indifference was not wholly responsible for the slow progress, but rather, the pathology of the disease was extremely difficult to identify. 14/

Dr. Lemuel Diggs, a sickle cell researcher in Memphis, added yet another aspect to the neglect of the disease. He said:

> "Until recently, Negroes themselves were opposed to sickle cell research because it was considered a stigma. There were even Negro doctors who didn't want to teach it or learn about it, because it was looked on as a sign of inferiority. Most white doctors misdiagnosed it. Negro doctors couldn't afford to care for the patients, because they were on welfare and poor just because of it. When you had sickle cell anemia, you couldn't finish school, you couldn't be employed, you couldn't even afford an aspirin." 15/

There is disagreemsnt even within the black population itself regarding the Sickle Cell Disease Program. Some blacks resent the racial references in literature and law, such as ". . .a disease of blacks," ". . .a disease exclusive to Negroes," and demand that these phrases be removed. 16/ Others, such as Colby King, a black employee of DHEW who helped spearhead the interest in sickle cell, are indignant that such

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13/ Cohn, Victor. Black Health Care Lag Cited. The Washington Post, Nov. 15, 1972: A12.

14/ Sickle Cell Anemia. Medical World News, Dec. 3, 1971: 41.

15/ Cohn, op. cit., p. A12.

16/ Ibid., p. A12.

literature and law "fails to note. . .that the disease is found essentially among blacks! Without even an allusion to the racial factor, the educational value. . .is seriously weakened." 17/ Such controversies as these, however, seem minimal in comparison to the accusations of racism, genocide, and the possible abuse of eugenics. 18/

Progressive annual increases of both Federal and private funds for sickle cell research paralleled an increase in national interest. Some felt, however, that this interest did not represent honest concern. William Montegomery, from the University of Pittsburgh, stated that:

> ". . .feelings are being expressed that there are blacks and whites in the community who are only interested in sickle cell anemia because it is only recently that funds have become available. And as funds have become available, numerous people have begun to try to develop various kinds of programs who never had a previous interest in sickle cell before. . .As a result of that, many of them, we feel, are really in it for their own personal gains rather than in the interest of providing some services to people who have sickle cell." 19/

At the end of 1972, and the beginning of 1973, there appeared to be a shift in black criticism from neglect in research to neglect in finding a cure. There was also an evolution of the blacks' definition of "research." Tabitha Powledge, in her well-known article "The New Ghetto Hustle" stated that, "Although sickle cell anemia is now enshrined in textbooks as a classic example of molecular research and Darwinian fitness, the fact remains that there is no very good treatment for the disease, and that is where the charges of neglect begin." 20/ Actually, these are new charges of neglect when compared with the cries of total neglect in research prior to 1971. Therefore, sickle cell research must be defined in terms of time: sickle cell research

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17/ Ibid., p. A12.

18/ Spivak, Johnathan. The Battle Against Sickle Cell Anemia Progresses. The Wall Street Journal, Jan. 4, 1973: 24. See also Tabitha Powledge's The New Ghetto Hustle. Saturday Review of the Sciences, Jan. 27, 1973: 39.

19/ Fraser, Gerald. Disease Programs Scored by Blacks. The New York Times, Apr. 9, 1972: 29.

20/ Powledge, Tabitha. The New Ghetto Hustle, Saturday Review of the Sciences, Jan. 27, 1973: 40.

prior to 1971 has been distinguished as molecular research, while research after 1971 is seen as molecular research and as therapeutic research.

An entirely new trend in thinking became prominent in 1973. A number of blacks complained that the current effort in all sickle cell research is a "cop-out", saying that sickle cell is far less important than other neglected black health problems such as hypertension, malnutrition, iron deficiency anemia, and above all, total health care. 21/ Colby King is one of the leading forces in this more recent attack on the Sickle Cell Disease Program. It was reported in 1971 that 13,500 blacks died as a result of hypertension, in comparison to 340 blacks who died of sickle cell anemia. Furthermore, iron deficiency anemia was reported as high as 40% in some black areas. 22/ In short, many believe that the entire Sickle Cell Disease Program represents a misplaced emphasis, and money could be better spent on total health care programs.

C. Screening and Counseling

The major sociological problems in the Sickle Cell Disease Program stem from the screening and counseling services. Screening involves examining the patient for the sickle cell trait, as well as detecting the actual disease. Counseling is two-fold: 1) medically advising the patient who is found to carry the trait, or the disease itself, and 2) informing these patients of the genetic implications. In order to be effective, counseling should also include financial and rehabilitative advice if the disease is diagnosed.

Education of the public to the facts of the disease seems to be lacking in both the screening and the counseling services, and the suggestion has been repeatedly made that the top priority in the program should be to better educate the susceptible population. It is extremely important that the distinction be made between the sickle cell trait and the disease itself. A report appearing in Medical World News in 1971 said surveys

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21/ Cohn, op. cit., p. A12.

22/ Ibid., p. A12.

showed that only three out of every ten black Americans had any knowledge at all of sickle cell anemia. 23/ Lack of knowldege of the disease creates problems because many of those found to be carriers during screening do not understand that this not necessarily harmful to their personal health. The resulting unwarranted fears often cause unnecessary psychological upsets. Dr. James E. Bowman, a prominent pathologist at the University of Chicago, said that screening centers may be "misguided, mismanaged, and of little value to the black community." He believes that the total effect of these screening programs has been to "create more problems than are solved including inaccurate, misleading, politically motivated propaganda which has left mothers frantic."24/

A professional problem with the screening centers is the increasing hazard of untrained non-professionals as staff. This type of staffing in screening and counseling centers can lead to a decrease in the quality of medical service to which the public is entitled. Some of these centers offer the screening without the counseling, a practice which understandably results in fear and uncertainty on the part of those patients who are told they carry the trait or have the disease.

The social problems stemming from the screening and counseling are compounded by various State regulations. At least 14 States, including the District of Columbia, have enacted screening legislation, some of which made screening mandatory. Often this legislation was enacted without benefit of public discussion or professional medical advice. Many individuals asked why sickle cell screening should be mandatory, when Tay-Sachs disease (a disease affecting a Jewish population) and Cooley's anemia (a disease affecting those of Mediterranean descent) do not require mandatory screening examinations. 25/ Again, this difference in emphasis produced accusations of racism, since there was no precedent for mandatory screening for any other disease on the basis of ethnic differences. Perhaps the most aggravating aspect of the D.C. law and similar State laws was the labeling of sickle cell anemia as a communicable disease, placing it

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23/ Sickle Cell Anemia (Medical World News), op. cit., p. 37.

24/ Bowman, James. Sickle Cell Screening: Help or Hindrance. University of Chicago Reports, v. 21, Fall 1972: 8.

25/ The Row Over Sickle-Cell. Newsweek, Feb. 12, 1973: 63.

in the same category as venereal disease. In the face of this negative public reaction, several States, including D.C., amended mandatory screening to voluntary screening.

Another major legislative issue which became apparent was that many of the screening laws implied that the sickle cell trait was equivalent to the actual manifestation of the disease. The consequences of screening based on this assumption can be psychologically and socially disastrous to a sickle cell carrier. Possession of the sickle cell trait becomes a part of the carrier's permanent health record (this in itself brings up yet another issue -- that of confidentiality of medical information), and this can prevent him from resuming a normal life. Some of the problems which have already been experienced by carriers include:

1. Possible loss of employment.
2. Prevention from obtaining employment.
3. Psychological disturbances/ stigma.
4. Prevention of participation in school sports (in areas where screening is required for children before entering a public school).
5. Problems in obtaining life or health insurance.

In some areas, it is reported that screening is required before obtaining a marriage license, and in this sense, sickle cell is again equated with venereal disease. Obviously, many blacks are greatly incensed by this type of regulation. Furthermore, sickle cell can be, and unfortunately is, often misdiagnosed. In these case, individuals may be forced to experience the aforementioned problems when they are not actually carriers of the sickle cell disease at all. This is particularly a problem where the staff members of the screening center are not skilled professionals.

Many people involved in the Sickle Cell Disease Program believe that screening should be voluntary because of the racial implications resulting from mandatory screening. On the other hand, other sources emphasize that the majority of the susceptible population is not educated to the facts of the disease, and that this awareness is necessary before they can be expected to voluntarily undergo screening and counseling.

It has been said the D.C's original mandatory screening legislation was based on previous experience; that is, a voluntary screening program for lead paint poisoning was initiated, and the response was very poor -- only 500 volunteers appeared for testing. 26/ The reaction to this line of reasoning as to why voluntary screening did not work was that the lead poisoning program was not logically comparable to a voluntary sickle cell program.

Mandatory screening laws place the susceptible black person in a situation where he might be forced to change his life style as a result of having sickle cell disease noted on his health record. Consequently, many blacks rebel against these regulations. Ironically, however, Tabitha Powledge said that most of the screening laws that blacks are rebelling against were sponsored by black legislators and were prompted by black groups. 27/

More disagreement emerges when considering the age at which a "susceptible" person should be screened for sickle cell anemia. As mentioned before, some regulations require that children about to enter school be screened. The argument against this is: if a child is found to carry the trait, what is be to do with the information other than carry it through his school years as an additional burden? More appropriate ages for screening might be: 1) shortly after birth when early detection would enable effective medical supervision, or 2) at child-bearing age when counseling could be appropriately offered. 28/

As mentioned earlier in this section, an extremely sensitive aspect of sickle cell testing is the counseling (that is, in those centers where counseling services are offered along with the screening programs). In 1971, clinicians were careful to

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26/ Frankel, Mark S. Political Responses to Controversial Issues in the Development of Biomedical Technologies. Prepared for delivery before the 1974 Annual Meeting of the American Political Science Association, Aug. 29 - Sept. 2, 1974: 7.

27/ Powledge, op. cit., p. 39.

28/ The Row Over Sickle-Cell, op. cit., p. 64.

refrain from making "recommendations" to affected persons. Instead they tried to explain to the patient the genetic possibilities in transmitting the disease to his or her children. From that point on, it was the patient's right to decide what course of action to take. It appears recently, however, that more definitive counseling is being advised and many, if not most, blacks are taking offense. 29/ From the black viewpoint, it often seems to be a matter of "genocide" when a member of the white population suggests that blacks refrain from reproducing, if they, as parents, carry the sickle cell disease.

One advantage of the new efforts to provide screening and counseling is the opportunity which the program offers for including many black researchers, physicians, and other professionals in a special area of medicine - one in which they might take a more personal interest. 30/

D. Medical Advancements and Concerns

Many researchers often refer to the "miracles" which have resulted from sickle cell research. Due to increased efforts to treat this disease, the following provide reasonable promise for further advancements:

- More is understood about the genetic and molecular bases of sickle cell anemia than any other genetic disease.
- Scientists are hoping that for the first time a genetic disease can be cured with drugs.
- A reliable test exists to identify the carriers.

Contrary to the opinion of some people that nothing is being done to find a cure for the disease, medical journals have been reporting the progress of possible new treatments, including urea, potassium cyanate, carbamyl phosphate, and other new drugs. Several drugs have been found to inhibit the sickling process, but after thorough investigation it is usually found that: 1) they are not as effective as regular invert sugar, 2) the concentrations necessary to affect the sickling are toxic to the patients, or 3) the side-effects are too damaging to make the agent

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29/ Sickle Cell Anemia (Medical World News), op. cit., p. 48.
30/ Culliton, Barbara J. Sickle Cell Anemia: National Program Raises Problems As Well As Hopes. Science, v. 178, Oct. 20, 1972: 286.

worthwhile. In 1974, urea was discounted as an effective treatment on the basis that it did not relieve the acute pain, and that it did not surpass the beneficial effects of invert sugar alone. 31/

Presently, the most promising agent appears to be sodium or potassium cyanate. Cyanate effectively inhibits the sickling process, but does have some side-effects which researchers hope are merely dose-related. 32/ These side-effects include: weight loss, decreased appetite, sensations in the extremities, and other nerve disorders. Cyanate has been shown to decrease the frequency of painful crises, but is still in the experimental stages, along with other possible new treatments such as zinc, aspirin, and dimethyl adipimidate (DMA).

Another idea for treating sickle cell effectively is to construct a machine, along the lines of the kidney machine, where sickle cell type blood could be treated with potent anti-sickling chemicals outside the patient's body, thereby eliminating the risks of toxicity and side-effects. 33/

In common practice now, however, treatment remains basically the same. This means that the symptoms, and not the disease, are being treated. For pain, the patient is given an analgesic; for dehydration, he is given fluids; for acidic blood, alkaline solutions are administered; and for infection, the patient is given antibiotics.

In addition to the clinical concerns in sickle cell treatment, many professionals continue to see a major problem evolving in the manpower factor of medical services. They view the influx of untrained personnel in the screening and counseling centers as the central defect in the overall program and in the quality of medical services available to the public. 34/

31/ Two Studies Discount Urea as Sickle Cell Treatment. The New York Times, May 28, 1974: 52.

32/ Cyanate Holds Promise in Sickle Cell Anemia. Chemical & Engineering News, July 15, 1974: 18.

33/ Antisickling Treatment Machine Foreseen. Medical Tribune, Sept. 4, 1974: 18.

34/ Bowman, op. cit., p. 8.

E. Summary

In the February 8, 1973 issue of the New England Journal of Medicine, Dr. Charles Whitten summarized eleven basic criticisms of the Sickle Cell Disease Program. In spite of the fact that they are the opinions of one man, they nevertheless focus on the controversies surrounding the issue. These opinions are listed below in abbreviated form:

1- Prevention of sickle cell anemia is not emphasized as the ideal program objective of genetic counseling.

2- Testing laws are mandatory, rather than voluntary.

3- The sickle cell trait has often been incorrectly projected as a personal health hazard.

4- Some testing programs are established without the necessary counseling services.

5- Inappropriate priorities exist in some service programs; for example, screening often focuses on young children, rather than on adults and adolescents of child-bearing age.

6- The lack of accurate information has created false and alarming impressions of the disease.

7- Inaccurate diagnoses are made by some physicians.

8- Most testing programs are not taking into account actual paternity.

9- Some sickle cell testing occurs without consent.

10- Fraudulent fund raising hampers the credibility of legitimate efforts.

11- The black community is not receiving a fair share of the funds and recognition in sickle cell anemia programming. 35/

Many critics have centered their objections to the Sickle Cell Disease Program around the opinion that screening for this disease is a "faddish", ethnic health program. They ask how such a program can be justified when, although detection is easy, there still exists no effective treatment or cure. On the other hand, proponents of the screening program argue that in spite of the fact that many of the drugs prove

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35/ Whitten, Charles F. Sickle-Cell Programming - An Imperiled Promise. The New England Journal of Medicine, v. 288, Feb. 8, 1973: 318-319.

unsatisfactory, the number and severity of a patient's crises appear to be reduced - possibly because of better medical supervision and treatment, and because the patients tend to lead more careful personal lives. 36/

To remedy the problems in the Sickle Cell Disease Program, many individuals begin by suggesting that the educational program be strengthened, and that the mandatory screening regulations be abolished. It is thought that voluntary screening and counseling programs could work, if preceded by an effective educational campaign. And, it is also necessary that the distinction be clearly made that possession of the sickle cell trait is not the same as having the sickle cell anemia.

36/ Cohn, Victor. Hopes Fall for Sickle Cell Drug. The Washington Post, Mar. 3, 1974: A12.

V. CONCLUSIONS

The levels of Federal support for the Sickle Cell Disease Program have increased between FY 1970 and FY 1975. These increases have brought a high level of support for molecular research and for screening and counseling efforts. Federal funding for FY 1976 is to be directed into continuing these efforts, as well as into continuing programs of preventive therapy, treatment, and education. However, the support for molecular research, screening, and counseling is much greater than the resources channeled into therapeutic research and public education. For example, in FY 1975 and again in FY 1976, the obligated funds for the Sickle Cell Disease Program (NHLI funds) are estimated to be $16 million. Of this total, approximately 63.0% will support molecular research, screening, and counseling; 28.5% will fund therapeutic research and research on the complications of sickle cell anemia; and 8.5% will pay administrative costs and support the development of educational materials and the Sickle Cell Disease Information Center. In FY 1973, the obligated funds for the Program were estimated to be $15.3 million; at that time, the percentages of the NHLI budget for the above program components were respectively: 62.2%, 31.7%, and 6.1%.

The public reaction to the Sickle Cell Disease Program seems to exhibit a great deal of unfavorable comment. Certain groups offer different kinds of criticisms, but the opinions seem to suggest that some aspects of the program have been over-developed at the expense of necessary support components, for example:

(1) Although molecular research has determined the genetic causes of sickle cell anemia, therapeutic research has yet to provide an inexpensive and safe treatment for the sickling crisis, or an available modality to prevent the sickling crisis.

(2) Public education has been inadequate in the Sickle Cell Disease Program. The basic facts about sickle cell anemia are not generally known, and insufficient distinction is made between those persons who merely carry the trait and those who have sickle cell anemia.

(3) Because public education has not been emphasized in the Sickle Cell Disease Program, the screening and counseling components can cause severe social problems for the patient. The credibility of advice offered in screening and counseling is weakened even more when these services are performed by untrained professional or non-professional staffs.

Published opinion does not suggest that a simple increase in funding now will necessarily improve the Sickle Cell Disease Program. The literature does suggest that:

A. Funds within the Sickle Cell Disease Program should be redirected to increase support for therapeutic research and to strengthen the educational and informational portions of the Program.

B. More emphasis needs to be placed on the distinction between the trait carrier and the individual afflicted by sickle cell anemia itself when such information in contained in an education or information portion of the Program.

C. Mandatory screening regulations need to be abolished.

D. The number and quality of professionals need to be upgraded in the medical, educational, screening, and counseling portions of the Program.

Finally, the National Heart and Lung Institute has summarized its awareness of the suggestions presented in the literature:

> We need to determine the prevalence and distribution of sickle cell disease and Cooley's anemia in this country, particularly the prevalence of carriers of the trait; provide accurate and up-to-date information on these diseases; initiate and expand appropriate community education, screening, and counseling programs; educate the medical and allied health professions about the problems of these diseases; identify and provide guidance for the management of the resulting psychological, sociological and economic problems; and assist individuals with these disorders to attain their maximum potential in life. 37/

37/ U.S. Congress. House. Committee on Appropriations. Departments of Labor, and Health, Education, and Welfare Appropriations for 1975. Hearings, 93rd Congress, 2nd session. Part 4. Wash., U.S. Govt. Print. Off., 1974. p. 323.

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VII. APPENDIX I: INFORMATION ON SICKLE CELL ANEMIA IN APPROPRIATIONS HEARINGS FY 1971 - FY 1975

Page	Content	Support Mentioned
FY 1971		
U.S. House of Representatives a/		
382	NHLI. Justifications. General statement under "Thrombosis and Hemmorhagic Diseases".	---
464	NHLI. Testimony. General Statement under "Hematology".	---
U.S. Senate b/		
2826	NHLI. Justifications. General statement under "Thrombosis and Hemmorhagic Diseases".	---
FY 1972		
U.S. House of Representatives c/		
366-8	NHLI. Testimony. Leads in treating Sickle Cell Disease, Origin of the Sickle Cell Genetic Characteristics.	---
404-5	NHLI. Testimony. Sickle Cell Anemia. NHLI involvement in research on Sickle Cell Anemia. Quotes funding by NHLI and NIAMD.	$5 million increase for NHLI $.450 million quoted as support by NIAMD in FY 1971
433	NHLI. Justifications. General statement under "Thrombosis and Hemorrhagic Diseases".	---
569-70	NIAMD. Testimony. General statement under "Hematology", and under "Coordination of NIH Sickle Cell Anemia Research".	---
864-71	NIGMS. Testimony. General definition of sickle cell anemia as a genetic disease.	---

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a/ U.S. Congress. House. Committee on Appropriations. Departments of Labor and Health, Education, and Welfare Appropriations for 1971. Hearings, 91st Congress, 2nd session, page 3. Washington, U.S. Govt. Print. Off., 1970.

b/ U.S. Congress. Senate. Committee on Appropriations. Departments of Labor, and Health, Education, and Welfare Appropriations. Hearings, 91st Congress, 2nd session, on H.R. 18515. Part 5. Washington, U.S. Govt. Print. Off., 1970.

c/ U.S. Congress. House. Committee on Appropriations. Departments of Labor and Health, Education, and Welfare Appropriations for 1972. 92nd Congress, 1st session, Part 3. Washington, U.S. Govt. Print. Off., 1971.

Page	Content	Support Mentioned
U.S. Senate d/		
3729-30	NHLI. Justifications. General statement under "Thrombosis and Hemorrhagic Diseases". Notes NHLI funding for sickle cell disease in FY 1972.	$3.5 million earmarked by NHLI for sickle cell research in FY 1972
3752	NHLI. Testimony. Sickle Cell Anemia and the NHLI budget. "There are some increases in [NHLI] budget for sickle cell anemia, some modest increase in the area of arteriosclerosis. Because the total budget remains the same, it means the increases in those areas are having to be squeezed out of some other areas."	---
3808	NIAMD. Justifications. General Statement of a new treatment being explored by NIAMD.	---
FY 1973 U.S. House of Representatives e/		
122-3	NHLI. Testimony. Sickle Cell Program attributed to the presidential initiative which identified sickle cell anemia as a high priority disease. Discussed the funding for sickle cell studies, clinics, and centers.	$9 million in FY 1972 NHLI budget $5 million addition requested for FY 1973 NHLI budget $14 million total NHLI request in FY 1973
145-7	NHLI. Testimony. Explanation of $1 million to be used by other institutes in the Sickle Cell Disease Programs. Discusses sodium cyanate as a new treatment for sickle cell anemia.	$.700 million NIAMD in FY 1973 $.300 million NICHD in FY 1973
168-82	NHLI. Testimony. Sickle Cell Screening. American Oil Co. Advertisement on Sickle Cell Disease. Sickle Cell Advisory Committee and its recommendations. Sickle Cell Clinics. Vaso-occlusive crisis explained and a list of 5 research projects in this area listed.	$2.5 million requested for research and service centers; $1 million requested for screening clinics $1.5 million requested for biomedical research $5 million total additional FY 1973 NHLI request

d/ U.S. Congress. Senate. Committee on Appropriations. Departments of Labor and Health, Education and Welfare Appropriations. Hearings, 92nd Congress, 1st session, on H.R. 10061. Part 6. Washington, U.S. Govt. Print. Off., 1971.

e/ U.S. Congress. House. Committee on Appropriations. Departments of Labor and Health, Education, and Welfare Appropriations for 1973. Hearings, 92nd Congress. 1st session, Part 4. Washington, U.S. Govt. Print. Off., 1972.

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184-5	NHLI. Testimony. Private funding for sickle cell disease noted as "seed money" for young investigators.	$.125 million total National Sickle Cell Foundation funds in FY 1972.
207-8	NHLI. Testimony. Activities related to Sickle Cell Disease, discussion of the tentative requests for FY 1973 and compares these to requests for FY 1972.	$4.5 million, FY 1972 and $4.5 MILLION, FY 1973 for research and service centers $1.8 million in FY 1972, and $6.55 million in FY 1973 for screening and counseling clinics $3.7 million, FY 1972, and $3.7 million, FY 1973 for research (2.7 in NHLI budget 1.0 million in other NIH budgets) $.25 million information center in FY 1973
210	NHLI. Testimony. Identification of sickle cell carriers.	---
227	NHLI. Justifications. "Explanations of Changes".	$.5 million of NHLI increase for research grants in FY 1973 $4.5 million of NHLI increase for clinical studies in FY 1973
229	NHLI. Justifications. "Significant Items in House and Senate Appropriations Committee Reports, 1972 House Report, Action to be Taken".	$14 million NHLI total request for Sickle Cell Disease Programs in FY 1973
235	NHLI. Justifications. General Statement. Mentions sickle cell programs as one of NHLI "extended initiatives" for FY 1973.	---
237	NHLI. Justifications. Research Grants Programs Analysis (Sickle Cell Disease).	$3.5 million 1972 est. $4.0 million 1973 est.
244	NHLI. Justifications. Research and Development Contracts. General statement on sickle cell.	---
246	NHLI. Justifications. Increases.	$4.5 million increase of total NHLI increase earmarked for sickle cell studies and clinics

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249	NHLI. Justifications. Research Management and Program Services. General statement on sickle cell therapy.	---
254	NHLI. Justifications. Program Purposes and Accomplishments. Research and Development Contracts. General statement on sickle cell as a new clinical project.	---
256	NHLI. Justifications. Program Purposes and Accomplishments. Research Management and Program Services. General statement on coordinated effort for therapy in sickle cell as a new program.	---
545	NIGMS. Testimony. Discussion of biomedical research on Sickle Cell Disease by NIGMS.	---
586	NIGMS. Justifications. Research Grants, Genetics. General statement on genetics research for sickle cell in NIGMS.	---
1251	NIAMD. Testimony. Sickle Cell Anemia Basic Research as part of Hematology Research in NIAMD. 28 grants for sickle cell disease research supported in FY 1972. Mentions the work of Dr. Makio Murayama.	$.750 million spent annually on basic sickle cell research at NIAMD
1258	NIAMD. Justifications. Laboratory and Clinical Research, Hematology. General statement on inclusion of Sickle Cell Disease basic research under Hematology research.	---
U.S. Senate f/		
2331	NIH. Testimony. Discussion of Budget Increases requested for FY 1973.	$5 million additional request brings total in NIH for sickle cell to $15 million in FY 1973.
2365	NIH. Testimony. Table: 1973 Program Areas of Special Interest, Sickle Cell Diseases.	$9 million spent in FY 1972 for sickle cell in NIH $14 million FY 1973 est. request for NHLI.
2369	NIH. Testimony. Prepared Description and Highlights of Special Emphasis Programs, NHLI, Sickle Cell Disease.	---

f/ U.S. Congress. Senate. Committee on Appropriations. Departments of Labor, and Health, Education, and Welfare Appropriations. Hearings, 92nd Congress. 2nd Session, on H.R. 15417. Part 3. Washington, U.S. Govt. Print. Off., 1972.

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2559-60	Research Resources and General Research Support Grants. Testimony. The Albany Medical College of Union University. Discussion of Project 5: Sodium Cyanate and Sickle Cell Disease.	---
2757	NIGMS. Justifications. Research Grants. Program Plans for 1972-73, Genetics. General statement on research, including mention of sickle cell anemia.	---
2789	NIGMS. Testimony. Genetics Diseases. General statement on the benefits of genetics research, including sickle cell anemia as part of this statement.	---
2956	NHLI. Justifications. Explanation of Changes.	$4.5 million of increase requested for sickle cell clinical studies $.5 million of increase requested for sickle cell research grants $5 million total increase in NHLI ear-marked for sickle cell
2957	NHLI. Justifications. Significant Items in House and Senate Appropriations Committee Reports. 1972 House Report, Action to be Taken.	$14 million total request of NHLI for sickle cell in FY 1973
2963	NHLI. Justification. General Statement. Mentions the sickle cell disease programs as one of NHLI "extended initiatives" for FY 1973	---
2965	NHLI. Justifications. Research Grants Programs Analysis (Sickle Cell Disease).	$3.5 million 1972 est. $4.0 million 1972 est.
2972	NHLI. Justifications. Research and Development contracts. General statement on Sickle Cell Disease as a new clinical program.	---
2976	NHLI. Justifications. Research Management and Program Services. General statement on coordinated effort for sickle cell therapy as a new program.	---
2979	NHLI. Justifications. Program Purposes and Accomplishments. Research and Development Contracts. General statement on Sickle Cell Disease as a new clinical program.	---
2980-81	NHLI. Justifications. Program Purposes and Accomplishments. Research Management and Program Services. General statement on coordinated effort for therapy in sickle cell as a new program.	---

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2983-4	NHLI. Testimony. Sickle Cell Programs. General, prepared statement.	---
2989	NHLI. Testimony. Distribution of Funds and FY 1973 Plans. Summarizes status of the Sickle Cell Program in FY 1972 and for FY 1973. NHLI designated as lead agency in the program. Notes Sickle Cell Disease funding increases as a part of total increases requested in NHLI budget for FY 1973.	---
2995	NHLI. Testimony. Obligations for Sickle Cell Disease Programs in FY 1972. Notes that FY 1973 budget request for sickle cell is $14 million, an increase of $5 million earmarked for sickle cell in the total NHLI budget.	1972 expenditures for sickle cell: $4.5 million, Centers $1.8 million, Clinics $2.7 million, Research $9.0 million FY 1972 total NHLI budget for sickle cell

FY 1974

U.S. House of Representatives g/

11	NIH. Testimony. Research Progress in FY 1973. General statement confirming that the cause of sickle cell anemia is known, and therefore most promising for counseling and developments for treatments.	---
83	NIH. Testimony. Prepared Statement. Appendix 4. Examples of Program orientation, formulation, and implementation: The Sickle Cell Disease Program.	---
263-4	NHLI. Testimony. General Statement. Notes the importance of children in the sickle cell program; also notes that NHLI requests an increase of $18 million to continue research in 4 areas, including sickle cell research.	---
307-11	NHLI. Testimony. Special Report prepared by request of the Committee: Sickle Cell Disease. Notes obligations estimated for all NIH Institutes in the Sickle Cell Program. Explains in detail the Sickle Cell Program. Notes that in the Comprehensive Centers for Research and Service, 40% of the cost is applied to research activities, 60% toward service activities. The entire cost of Screening and Counseling Clinics is applied to service activities. Mission-oriented Research and Development, a contracts mechanism,	$16 million FY 1974 est. request, NHLI $.859 million FY 1974 est. request, NIAMD $.100 million FY 1974 est. request, NIGMS $16.959 million FY 1974 total request for NIH.

g/ U.S. Congress. House. Committee on Appropriations. Departments of Labor and Health, Education, and Welfare Appropriations for 1974. Hearings, 93rd Congress, 1st session, Part 4. Washington, U.S. Govt. Print. Off., 1973.

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	supports controlled studies of therapy, basic studies of molecular structure and various complications of sickle cell disease. Research projects grants support meritorious research on sickle cell disease recommended by other NIH institutes but funded by NHLI.	
319	NHLI. Testimony. General statement on total establishment of the Sickle Cell Disease Program in FY 1973. Concern expressed over State laws enacted to make testing for the sickle cell trait in all people mandatory.	---
320	NHLI. Testimony. Sickle Cell Budget. Contracts are noted as being awarded for controlled studies of therapy. Grants are noted as being awarded for (1) basic studies of hemoglobin and red blood cell membrane shape and structure and (2) basic studies of chemicals as treatments to prevent the sickling crisis or to treat the sickling crisis itself.	$15 million est. expenditures for Sickle Cell Program in FY 1973 $16 million request for Sickle Cell Program Budget in FY 1974.
334-5	NHLI. Testimony. Social Implications of Sickle Cell Programs.	---
346	NHLI. Justifications. Explanation of Changes. Increases. Research and Development Contracts. NHLI budget requests an increase of approx. $18 million: Sickle Cell Disease Program increases will come from Research and Development Contracts.	---
348	NHLI. Justifications. Significant Items in the House and Senate Appropriations Committees Reports. 1973 House Report. Action to be Taken. Sickle Cell Disease current research programs emphasize preventive therapy and treatment. Current services of the Sickle Cell Program are implemented. Data on research and services now being collected.	--- ---
350	NHLI. Justifications. Authorizing Legislation. Cites Title XI, Genetic Blood Disorders, Part A, Sickle Cell Program, as the authorizing legislation for the NHLI Sickle Cell Program. Lists the appropriations for FY 1974.	$40 million authorized in Title XI, Part A for Sickle Cell Program in FY 1974
356	NHLI. Justifications. General Statement. Division of Extramural Affairs. Summarizes the current status of the Sickle Cell Program.	---

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363	NHLI. Justifications. Research Grants. General Statement. The Sickle Cell Disease Program is highlighted in FY 1973-74. The FY 1974 budget request will maintain non-competing commitments (including the Sickle Cell Centers) at the approved levels. Competing renewals and new projects in the regular research grants program would be maintained at the FY 1973 level.	---
367	NHLI. Justifications. Research and Development Contracts. Summary. $4 million of the total Research and Development Contracts Increase ($10.050 million) will provide for blood diseases and resources, including additional Sickle Cell Programs.	---
379	NHLI. Justifications. Program Purposes and Accomplishments. Research Grants. Cites amount ear-marked from increases to supplement the Sickle Cell Center Program	$.500 million FY 1974 increase for the Sickle Cell Center Program
383	NHLI. Justifications. Program Purposes and Accomplishments. Research and Development Contracts. Notes that the Sickle Cell Program funding was increased $5 million from FY 1972 for FY 1973; "Special appropriations have been made to bring about a coordinated effort within DHEW to effect a multi-categorical attack on this disease from the standpoint of research and service." FY 1974 "special appropriations" for the Sickle Cell Program are not specified, but are included in blood diseases and resources increase of $4 million.	---
475	NIAMD. Testimony. Special Report on Cooley's Anemia. Notes that "HSMHA is developing within available resources a genetic blood disorders program within the requirements of Title XI of the Public Health Service Act. The program includes both Part A, Sickle Cell Anemia, and Part B, Cooley's Anemia."	---
798	NIGMS. Testimony. General statement on the common genetic diseases, including sickle cell anemia.	---
FY 1974 U.S. House of Representatives h/		
804-9	HSMHA. Special Report prepared by DHEW which relates to HSMHA. Sickle Cell Anemia. National Center for Family Planning Services. Cites the funding and organization of the services which HSMHA provides in the Sickle Cell Program. Funding is obtained for HSMHA in an interagency reimbursable agreement with NHLI.	$1.975 million in FY 1972 $8.1 million FY 1973 est. $8.6 million FY 1974 est. For HSMHA in NHLI budget

h/ U.S. Congress. House. Committee on Appropriations. Departments of Labor and Health, Education, and Welfare Appropriations for 1974. Hearings, 93rd Congress, 1st session, Part 3. Washington, U.S. Govt. Print. Off., 1973.

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FY 1974 U.S. Senate i/		
1053	DHEW. Overview. Increases in funds for cardiovascular research support "will permit...the continuation of the attack on sickle cell & other blood diseases and on pulmonary ailments."	—
1275	DHEW. Health Services Planning & Development. Regional Medical Program. Justifications. In 1972 RMP supported 2 projects relating to the availability and accessibility of health services for sickle cell anemia. By FY 1974, all RMP programs were planned to be phased out.	RMP support of 2 projects $115,000 (FY 1972)
1705	NHLI. Prepared Statement (Cooper). Emphasizes the importance of children in the Sickle Cell Disease Program. Cites the establishment in 1973 of 10 community centers for research and demonstration and 19 screening and education clinics.	—
1708	NHLI. Testimony. Discusses value of a metropolitan location for the 10 community research centers and the 19 screening and education clinics.	—
1713	NHLI. Testimony. Discusses the cooperative role of NHLI, NIAMDD, and NIGMS in research on sickle cell anemia and Cooley's anemia.	—
1719-20	NHLI. Justifications. Sickle Cell Anemia research grants included as those in the high priority list for NHLI.	—
1722	NHLI. Justifications. Significant Items in the House and Senate Appropriations Committee Reports. House Committee expressed surprise and chagrin that so little attention had been paid to sickle cell disease. Reply includes information on funding, as well as on increased activity in biochemical research and demonstration programs to increase public awareness.	FY 1973 budget = $15 million FY 1974 request = $16 million
1723-25	NHLI. Justifications. Authorizing Legislation. Cites Title XI, Genetic Blood Disorders as the authorizing legislation for the NHLI Sickle Cell Disease Program (Part A). Lists authorized appropriations.	Grants & Contracts (information, education) FY 72 - $20 million FY 73 - $30 million FY 74 - $35 million Grants & Contracts (research and development) FY 72 - $ 5 million FY 73 - $10 million FY 74 - $15 million

i/ U.S. Congress. Senate. Committee on Appropriations. Departments of Labor and Health, Education, and Welfare Appropriations for 1974. Hearings, 93rd Congress, 1st session, Part 2. Washington, U.S. Govt. Print. Off., 1973.

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1729	NHLI. Justifications. General Statement. Division of Extramural Affairs. Summarizes current status of the Sickle Cell Disease Program.	---
1732	NHLI. Justifications. General Statement. Division of Extramural Affairs. "One of the extended initiatives this year is in sickle cell disease. Further research certainly is needed and is planned, but also planned are additional clinics and comprehensive centers to identify the susceptibles, to evaluate new therapy, and to bring information on prevention to the people at risk since there is no cure once the disease is established."	---
1734	NHLI. Justifications. Research Grants, Research Projects. "A program in Sickle Cell Disease is highlighted in 1972 - 1974. This research effort has been designed to uncover new information which can be used to allievate the suffering of a significant percentage of the Nation's black population. The FY 1974 budget request will maintain non-competing commitments (including sickle cell centers) at the approved levels. Competing renewals and new projects in the regular research grants program would be maintained at the FY 1973 level."	---
1745	NHLI. Justifications. Research and Development Contracts. Summary. $4 million of the total Research & Development Contracts increase ($10.05 million) will be to provide for blood diseases and resources, including additional sickle cell programs.	---
1748	NHLI. Justifications. Program Purposes and Accomplishments. Research Grants. Cites amount earmarked from increases to supplement the sickle cell center program.	$500,000 increase for sickle cell centers
1750	NHLI. Justifications. Program Purposes and Accomplishments. Research & Development Contracts. Notes that the Sickle Cell Disease Program funding was increased $5 million from FY 1972 for FY 1973; "Special appropriations have been made to bring about a coordinated effort within DHEW to effect a multicategorical attack on this disease from the standpoint of research and service." FY 1974 "Special Appropriations" for Sickle Cell Disease Program are not specified, but are included in blood diseases and resources increase of $4 million.	---
1964	Research Resources. Justifications. Research Grants. Blood Studies. Normal and abnormal hemoglobins, such as those in sickle cell anemia, are being studied at Carnegie-Mellon University in Pittsburgh. The technique uses a Nuclear Magnetic Resonance spectrometer to elucidate the mechanism by which human hemoglobin carries oxygen.	---

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FY 1974 U.S. Senate i/		
2242-43	NIAMDD. Special Report, Hematology. Highlights of research on the chemical treatments for sickle cell anemia and on detection of the sickle cell gene in utero.	---
FY 1975 U.S. House of Representatives k/		
681-686	DHEW. Special Report on Sickle Cell Anemia prepared by DHEW for the House Committee on Appropriations. Covers period FY 1971 - FY 1975. Provides detailed descriptions of funding for FY 1971 - FY 1975 for NHLI, NIAMDD, and NIGMS. Provides detailed description of on-going programs and projects, and includes a summary of completed research projects. Includes summary of activities of Office of Child Health for FY 1972 - FY 1974.	NIH funding by Institute. NIH TOTAL FUNDING: FY 71 - $1.89 million FY 72 - $11.256 million FY 73 - $16.540 million FY 74 - $17.216 million FY 75 - $17.254 million est.
FY 1975 U.S. House of Representatives l/		
227	NHLI. Testimony. Comments (Cooper) on urea research for treating sickle cell crises: "urea does not appear to be effective in relieving the crises nor preventing the sequelae." Cooper also comments on the lack of available technique to properly inform the layman and the professional about sickle cell disease and its elements of risk.	---
267	NHLI. Testimony. Current extent of the Sickle Cell Disease Program described as addressing this disorder from two standpoints: 1) research and development, i.e., basic and clinical research, and 2) demonstration activities, i.e., education, testing, counseling, and rehabilitation. Funding discussed for FY 1973 and for FY 1975.	Sickle Cell Disease and Related Diseases Research: FY 73 - $14.5 million FY 75 - $15.5 million
277	NHLI. Justifications. Authorizing Legislation. Cites Title XI, Genetic Blood Diserders, as authorizing legislation for the Sickle Cell Disease Program in NHLI.	FY 1975 authorized appropriation: $53.7 million

i/ U.S. Congress. Senate. Committee on Appropriations. Departments of Labor and Health, Education, and Welfare Appropriations for 1974. Hearings, 93rd Congress, 1st session, Part 3. Washington, U.S. Govt. Print. Off., 1973.

k/ U.S. Congress. House. Committee on Appropriations. Departments of Labor and Health, Education, and Welfare Appropriations for 1975. Hearings, 93rd Congress, 2nd session, Part 3. Washington, U.S. Govt. Print. Off., 1974.

l/ U.S. Congress. House. Committee on Appropriations. Departments of Labor and Health, Education, and Welfare Appropriations for 1975. Hearings, 93rd Congress, 2nd session, Part 4. Washington, U.S. Govt. Print. Off., 1974.

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280	NHLI. Justifications. General Statement. The Problem. "Sickle cell anemia and severe hemophilia afflict 50,000 to 70,000 persons."	---
317	NHLI. Justifications. Blood Diseases and Resources. Table description of funding for Sickle Cell Disease and Related Diseases, FY 1974 - FY 1975. (Excludes FY 1973 appropriation restorations.)	Sickle Cell Disease & Related Diseases: FY 74 - $15.5 million FY 75 - $15.5 million est.
318	NHLI. Justifications. Blood Diseases and Resources. Objectives, 1975. Cites Public Health Service Act, Title XI as the authorizing legislation for the Sickle Cell Disease Program. States that the Division of Blood Diseases and Resources "coordinates Federal Sickle Cell Disease activities and operates a clearing-house for information on sickle cell disease."	---
319	NHLI. Justifications. Blood Diseases and Resources. Accomplishments and Objectives, 1973/1974. States that the Program begun in 1972 for sickle cell disease has been expanded; Comprehensive Sickle Cell Centers now number 15; Screening & Education Clinics now number 26. States that basic research centers on hemoglobin.	---
320	NHLI. Justifications. Blood Diseases and Resources. Bleeding and Clotting Disorders. Sickle Cell Anemia and Sickle Cell Trait: "The research goals of the Sickle Cell Disease Program are to develop improved therapy for sickle cell crises; increase knowledge of the fundamental biology of the disease and its compli-cations; and develop effective and acceptable methods for screening and counseling."	---
322-23	NHLI. Justifications. Blood Diseases and Resources. Prevention, Education, and Control. States that NHLI is developing a control program for sickle cell dis-ease. "The goals of the control program are to educate the public about sickle cell disease and Cooley's anemia and to demonstrate techniques for appropriate screening and counseling". Lists a series of "needs", or areas of the program for which data is lacking, i.e., prevalence and distribution of sickle cell disease and of the trait are not known, etc.	---
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328	NHLI. Justifications. Research Management and Program Services. Accomplishments and Objectives 1973/1974. Reviews the mandate for the National Sickle Cell Disease Program as emphasized in the National Sickle Cell Anemia Control Act of 1972.	---

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331	NHLI. Justifications. Program Purposes and Accomplishments. Blood Diseases and Resources. States that in FY 1974 over 70 contracts were to be awarded with emphasis on: 1) development of a nationwide blood system; 2) continuation of sickle cell screening and education; 3) studies of hemoglobin interaction.	---
332	NHLI. Justifications. Program Purposes and Accomplishments. Intramural Laboratory and Clinical Research. Projects on the mechanism of hemoglobin biosynthesis and hemoglobin genetics - which will lead to a better understanding of the molecular basis of the hereditary anemias - were to continue through FY 1974. "In FY 1975 there will be increased support of research in molecular diseases, pulmonary diseases, experimental therapeutics, molecular hematology, pathology, and veterinary research."	---
423	NIAMDD. Justifications. Special Report on Cooley's Anemia and Other Blood Disorders. "It is estimated that there are more than 1,000,000 persons in the United States who suffer from disabling or killing disorders of the blood, including the anemias and bleeding disorders.	---
426-27	NIAMDD. Justifications. Special Report on Cooley's Anemia and Other Blood Disorders. Presents conclusions of NIAMDD research on sickle cell disease among NFL football players; prevalence of sickle cell trait was 6.7%. Study supports evaluation by National Academy of Sciences study that sickle cell trait does not necessarily limit the everyday activities or functions of the trait carrier. Also reports on new technique for mass-screening for sickle cell disease: cost of technique: $.03 per specimen screened.	---
800	NIGMS. Justifications. Genetics. Current Program. Summarizes the work of one Genetic Center on the control, screening, and treatment of sickle cell disease. Specifically mentioned is research on the extracorporeal use of cyanate to treat sickle cell crises.	---
FY 1975 U.S. Senate m/		
929-30	NIH. Testimony. New Initiatives/High Priority Programs. Table presenting appropriations and obligations for the Sickle Cell Disease Program, FY 1973 - FY 1975.	NHLI: FY 73 - $15.320 million; FY 74 - $16.0 million; est. FY 75 - $16.0 million; NIAMDD: FY 73 - $ .933 million; FY 74 - $1.058 million; est. FY 75 - $ .955 million

m/ U.S. Congress. Senate. Committee on Appropriations. Departments of Labor, and Health, Education, and Welfare, and Related Agencies Appropriations. Hearings, 93rd Congress, 2nd session, Part 3. Washington, U.S. Govt. Print. Off., 1974.

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941	NHLI. Testimony. Prepared Statement (Ringler). Mentions the past research on treatment of the sickle cell crisis with urea which proved to be ineffective.	---
965-66	NHLI. Testimony. Presents a list of 15 Sickle Cell Disease Specialized Centers of Research (SCOR), with amounts awarded or committed for FY 1975.	Support cited for each SCOR.
975-76	NHLI. Testimony. Debate on the funding of the Sickle Cell Disease Program. Conclusion that the spending levels for FY 1975 would equal that for FY 1976 with no increase to cover the increased costs due to inflation: "that level is the same, and to whatever extent inflation is a factor, it will be reduced accordingly."	FY 1974 & FY 1975 spending level approximately $16 million. NHLI Plan calls for $20 million in support of the Sickle Cell Program by 1980.
986-87	NHLI. Testimony. Presents information on sickle cell trait and disease prevalence: approximately 2 million persons are believed to carry the trait; approximately 1 in every 500 black babies are afflicted with sickle cell anemia. Notes that the Sickle Cell Disease Program now includes 15 Comprehensive Sickle Cell Centers and 26 Screening, Education, and Counseling Clinics.	---
1001	NHLI. Justifications. Authorizing Legislation. Cites Title XI, Genetic Blood Disorders and authorizing legislation for the Sickle Cell Disease Program within the NHLI.	Authorized Appropriation FY 1975: $53.7 million
1003	NHLI. Justifications. General Statement. The Problem. "Sickle cell anemia and severe hemophilia afflict 50,000 to 75,000 persons."	---
1035	NHLI. Justifications. Blood Diseases and Resources. NHLI: Table Presents FY 1974 and FY 1975 comparable obligations for the Sickle Cell Disease Program. (Excludes FY 1973 appropriations restorations.)	FY 74 - $15.5 million est. FY 75 - $15.5 million
1036	NHLI. Justifications. Blood Diseases and Resources. Objectives, 1975. Cites Public Health Service Act, Title XI as the authorizing legislation for the Sickle Cell Disease Program. States that the Division of Blood Diseases and Resources "coordinate Federal Sickle Cell Disease activities and operates a clearinghouse for information on sickle cell disease."	---
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1046-47	NHLI. Justifications. Program Purposes and Accomplishments. Blood Diseases and Resources. States that in FY 1974 over 70 contracts were to be awarded with emphasis on: 1) development of a nationwide blood system; 2) continuation of sickle cell screening and education; 3) studies of hemoglobin interaction.	—
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